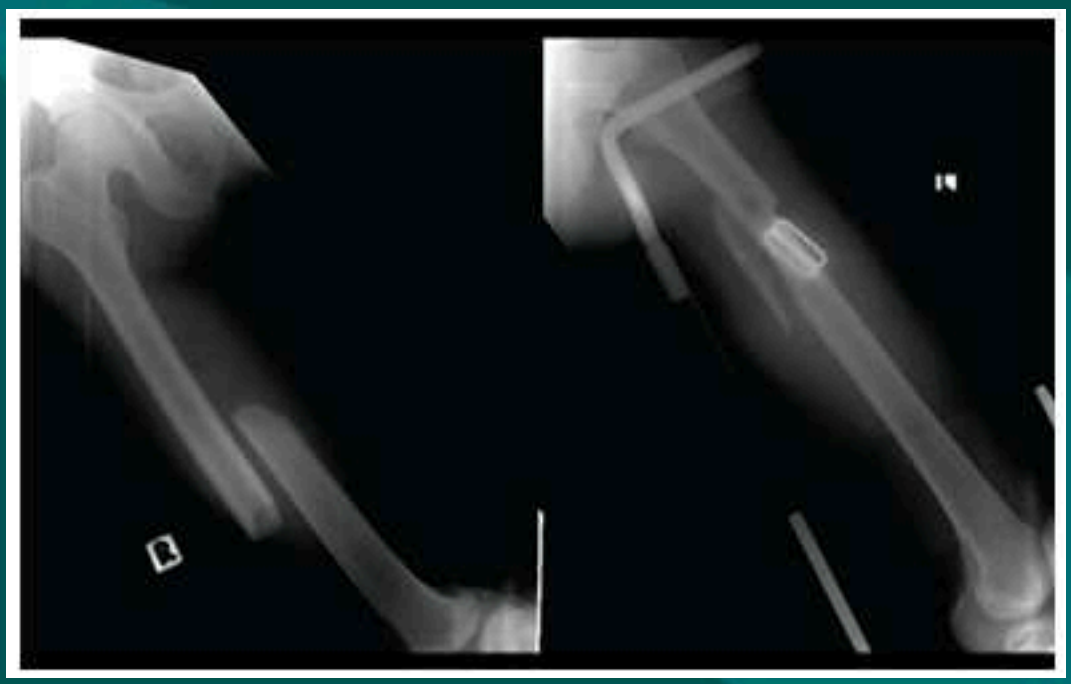
March 2014; Volume 5; Number 3

IJCRI

www.ijcasereportsandimages.com

International Journal of

CASE REPORTS AND IMAGES



Online submission at:
www.ijcasereportandimages.com/submit

ISSN: 0976-3198

International Journal of Case Reports and Images

Contact Details:

Editorial Office

Email: meditor@ijcasereportsandimages.com
Fax: +1-773-409-5040
Website: www.ijcasereportsandimages.com

Guidelines for Authors

Full instructions are available online at:
www.ijcasereportsandimages.com/submit/instructions-for-authors

Manuscript submission:
www.ijcasereportsandimages.com/submit

International Journal of Case Reports and Images

EDITORIAL BOARD

International Journal of Case Reports and Images (IJCRI)

Contact Details:

Editorial Office

Email: meditor@ijcasereportsandimages.com
Fax: +1-773-409-5040
Website: www.ijcasereportsandimages.com

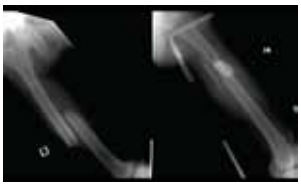
Guidelines for Authors

Full instructions are available online at:
www.ijcasereportsandimages.com/submit/instructions-for-authors

Manuscript submission:
www.ijcasereportsandimages.com/submit

International Journal of
Case Reports and Images

Cover Figure:



All Articles:

Open Access

Contents

Vol. 5, No. 3 (March 2014)

Cover Image

Figure 2: Bilateral midshaft femur fracture.

Case Series

189 Ipsilateral fracture of the supracondylar humerus and forearm in children
Shahid Hussain

195 Acute renal failure in two cases with hypothyroidism related rhabdomyolysis
Davut Akin, Sehmus Ozmen

Case Report

198 Use of diode lasers in treatment of oral submucous fibrosis: A new concept in surgical management
Sanjay Asnani, Uma Mahindra, Rakesh Oswal

201 Medical school finals, nerves and vomiting: Medical student survives Boerhaave's syndrome with recurrence one week after initial presentation
Eshan Oderuth, Kevin Ilo, Munir Somji, Thomas Holme

206 Steatocystoma multiplex of face: A case report
Surej Kumar LK, Nikhil Mathew Kurien, Varun Menon P

210 Damage control orthopedics in a patient with polytrauma complicated with thyroid storm
Rishya Manikam, M Emmed, CS Kumar, Jeffry Amit, AB Sri Latha

214 Pentalogy of Cantrell diagnosed in the first trimester of pregnancy
Yasemin Cekmez, Tülay Tos, Zehra Yilmaz, Nilay Pişkinpaşa, Tuncay Küçüközkan

217 Neurilemmoma of the tongue: A case report
Pallav Kumar Kinra, Jayakumar K, Manoj Joseph Michael

221 Prolonged aphasia and perfusion computed tomography abnormalities in migraine with aura
Anselm Angermaier, Soenke Langner, Michael Kirsch, Alexander V Khaw

225 Stroke in pregnancy: A rare case of protein C and protein S deficiency
Snigdha Kumari, Ashok Kumar Biswas, Sukanta Misra

229 Idiopathic pulmonary hemosiderosis without hemoptysis in an adult: A rare presentation
Jackin Moses R, Nishant Sinha, Madhusmita M, Kisku KH, Manjiri P

234 Benign reactive lesion with atypical mitosis: New example of an old story
Hua Zhong, Marina Chekmareva, Malik Deen, Michael May, Steven Deak, Nicola Barnard

239 An unusual presentation of infected urachal cyst in an adult
Anand Munghate, Ashwani Kumar, Harnam Singh, Mahak Chauhan, Gurpreet Singh, Manish Yadav

Clinical Image

243 Metopic synostosis
Natasha Gupta, Blankstein Josef

246 Anomalous left vertebral artery
Hussein Ahmed Hassan, Caroline Edward Ayad, Tag Eldin Mohamed Ibrahim, Ikhlas Abdelaziz Hassan

SUBMISSION INSTRUCTIONS
All manuscripts, including illustrations, should be submitted at: www.ijcasereportsandimages.com/submit or email to: submit@ijcasereportsandimages.com
Author Guidelines: www.ijcasereportsandimages.com/submit/instructions-for-authors.php
For any questions contact the Editorial Office via e-mail at: info@ijcasereportsandimages.com or Fax: 1-773-409-5040

CASE SERIES OPEN ACCESS

Ipsilateral fracture of the supracondylar humerus and forearm in children

Shahid Hussain

ABSTRACT

Introduction: Simultaneous ipsilateral fracture of the elbow and forearm (floating elbow) is an uncommon injury and treatment recommendations are controversial. The aim of our study was to evaluate the incidence of ipsilateral fractures of the upper limb and to present our experience in dealing with such injuries and to review the literature relating to this topic. The following variables were used: age, gender, side, mechanism of injury, type of fracture, classification, treatment methods, complications and outcome. Case Series: We prospectively followed five children who presented with displaced supracondylar fractures of the humerus associated with a forearm fracture of the same limb. All patients underwent emergency procedures in the form of closed reduction and K-wire fixation/cast fixation. At a minimum follow up of 24 months, all patients were assessed clinically and radiologically and the results evaluated according to a conventional scoring system. Four patients had excellent or good outcomes, and there was one poor result. Conclusion: The floating elbow is an indicator of a high energy injury. The incidence of open fractures, compartment syndrome and nerve injury and the need to perform an open reduction were higher than those recorded for isolated supracondylar or forearm fractures. The existing controversies in the management of such a complex injury and associated problems are outlined. A pertinent literature review is also included.

Keywords: Children, Musculoskeletal injuries, Floating elbow, closed reduction and internal fixation

Shahid Hussain
Affiliations: Registar, Department of Orthopedics, Sheri Kashmir Institue of Medical Sciences and Hospital, Bemina, Srinagar, Jammu and Kashmir, India.
Corresponding Author: Dr. Shahid Hussain, Registar, Department of Orthopedics, Sheri Kashmir Institue of Medical Sciences and Hospital, Bemina, Srinagar, Jammu & Kashmir, India 190006; Ph: 919419003100; Email: drshahid2006@gmail.com

Received: 30 September 2013
Accepted: 24 October 2013
Published: 01 March 2014

How to cite this article

Hussain S. Ipsilateral fracture of the supracondylar humerus and forearm in children. International Journal of Case Reports and Images 2014;5(3):189–194.

doi:10.5348/ijcri-2014-03-470-CS-1

INTRODUCTION

In children, isolated supracondylar fractures constitute 50–70% of elbow injury while the distal forearm bone fractures are equally common injury comprising 75% all forearm fracture [1]. The single bone fractures of radius or ulna or both constitute up to 10% of all pediatric fractures. But ipsilateral supracondylar and forearm bone fracture is an uncommon injury [2, 3]. Some of the textbooks of pediatric orthopedics do not make a mention of floating elbow injury [4]. The incidence of floating elbow injuries is between 3–13% [5]. It is a severe injury and some authors have reported a high incidence of compartment syndrome and in most cases they are associated with high-energy trauma [6–8]. Stanitski and Micheli were the first to use the term *floating elbow* to describe combination of supracondylar and both bone forearm fractures where in the elbow is effectively dissociated from the rest of

the limb [7]. In a study on 3,472 patients, Buckley et al. stated that the second most frequent associations was between radial, ulnar and humeral fractures (nine cases) in concomitant fractures [9]. In his study of 1199 patients with 1722 injuries, Malheiros found combination of fractures of humerus and forearm bones to be most frequent (25 cases) among multiple injured patients [10]. Complications like open fractures, compartment syndrome and neurovascular injuries are more frequent in children with floating elbow injury than in isolated supracondylar fractures [11].

The recommended treatment for this combination of fractures remains controversial. Various combinations of treatment options has been suggested for this injury such as primary closed reduction of both fractures and long arm cast application or olecranon pin traction till swelling subsides and delayed long arm cast application or pinning supracondylar fracture and short arm cast application for forearm bone injury or closed reduction and percutaneous pinning for both injuries [5, 8]. Choosing best management options for floating elbow is challenging. Because of rarity of injury and limited availability of literature.

Although conservative management has been described in literature [12, 13], most authors recommend percutaneous fixation of humeral fractures with pins for better results and to reduce risk of neurovascular complications [7, 14–16]. Templeton and Graham recommended fixation of forearm fractures for neurovascular monitoring and better wound care in open injuries [17]. The aim of the present study was to evaluate the incidence and treatment modalities of ipsilateral supracondylar and forearm fractures in five children who attended our hospital between 2010 and 2012. A detailed review of literature is also included for better understanding of this uncommon injury.

CASE SERIES

Over a two-year period from September 2010 to September 2012 we conducted prospective analysis of 122 pediatric patients at Department of Orthopedics, Sheri Kashmir Institute of Medical Sciences, Srinagar with different musculo-skeletal injuries and came across five (4.09%) children with ipsilateral supracondylar humerus and forearm fractures.

The five children were treated prospectively by closed reduction and percutaneous pinning/external fixation/cast fixation depending upon the fracture anatomy. There were three (60%) boys and two (40%) girls with a mean age of 9.4 years (range 7–13 years). The fractures were right sided in two (40%) and left-sided in three (60%). The cause of injury was a fall from a height in three (60%) cases, road traffic accident in one (20%) and fall from standing height in one (20%). Details of the patients are given in Table 1. There were three Gartland type-III and 2 Gartland type-II supracondylar fractures

Table 1: Details of patients and type of fracture (#-Fracture;SC-Supracondylar; BB FA-both bone forearm; SOR-Shaft of radius; DER-distal end radius; RTA-road traffic accident; FFH-fall from height; FFSH-fall from standing height.

S. No.	Age (Years)	Sex	Side	Mode of trauma	Fractyure pattern	Gartland type	Gustilo Anderson
1	13	M	L	RTA	# SC humerus, # Medial epicondyle, #Olecranon, # capitellum # SOR	3	Type 3A
2	10	F	R (Contralateral DER)	FFH	#SC humerus # bilateral #DER	3	
3	8	M	L	FFH	#SC humerus # DER	2	
4	7	F	R	FFSH	# SC humerus # BB FA	2	Type 1
5	9	M		FFH	#SC humerus # DER	3	Type 1

Table 2: Type of fracture as per Gustilo Anderson and treatment uses.

Type of Fracture		Close Reduction & Pinning	Open Reduction & Pinning	Open Reduction & Pinning + External Fixation	Close Reduction & Plaster	Total
Supracondylar	Closed	3	2		1	5
	Open	2	1	1		
Both bone	Closed					1
	Open	1	1			
Shaft of Radius	Closed	1	1			1
	Open					
Distal End Radius	Closed	4	3			4
	Open					

of the humerus. In the type-III injuries the displacement was posteromedial in four cases. There were four fractures of distal end radius in three patients (with one patient having bilateral injury), both bones of the forearm in one and of the radius alone in one patient. One patient with RTA with compound type 3B elbow injury had, in addition ipsilateral fractures of medial epicondyle, capitellum and olecranon. One patient had ipsilateral fractures of olecranon. Fasciotomy was performed in one patient with ipsilateral both bone forearm fracture who presented with compartment syndrome the type of fracture and treatment used is given in Table 2. One supracondylar fracture was classified as Gustilo Anderson compound type 3B, one supracondylar and one both bone fractures were classified as Gustilo type-1 open fractures [18]. All four distal end radius fractures were Salter Harris type 2 injuries. There were two median nerve injuries. Preoperatively, the radial pulse was absent in two patients. In these patients, the pulses returned after reduction. The forearm both bone (n=1) and proximal radial shaft fractures (n=1) were reduced and stabilized with K-wires. Three distal radial epiphyseal injuries were fixed with K-wires while one fracture was stabilized in plaster cast. The supracondylar fractures were then reduced by

longitudinal traction and manipulation and fixed with two crossed K-wires; one lateral and one medial entry pins (in four (80%) patients). One patient with compound elbow injury needed external fixator in addition to cross K-wire fixation. One patient (20%) was managed with plaster fixation. The lateral pin is inserted first and must traverse the lateral portion of ossified capitellum, cross the physis, proceed up the lateral column and engage the opposite medial cortex. The medial pin is placed through the medial epicondyle and should traverse the medial column and engage the opposite lateral cortex. Care must be taken to protect the ulnar nerve. The elbow is placed short of 90 degree flexion. The nerve may be palpated if possible or a small 1.5 cm incision made over the medial epicondyle. Continuous fluoroscopy was used during reduction of the supracondylar fracture and pinning of both bone fracture. The pins were bent outside the skin to prevent migration and to make later removal easier and an above elbow slab was applied. One patient developed iatrogenic ulnar nerve palsy in immediate post-operative period which eventually recovered at three months follow-up. The two median nerve palsies also resolved with time.

Postoperatively, an above-elbow long arm posterior plaster slab was retained for three weeks. The wires were removed from the elbow at 3–4 weeks and active exercises started. The wires were removed from the forearm six weeks after operation. Patients were recalled for a detailed clinical and radiographic evaluation at between 12 and 24 months from injury and were assessed for pain, stiffness and cosmesis. The range of elbow flexion-extension, forearm rotation, wrist flexion-extension and the carrying angles were measured by goniometers in both the injured and non-injured limbs. Radiographs were obtained of elbows, forearms and wrists. We performed a detailed neurovascular examination and compared the findings with those recorded on admission and after operation. The final outcome was graded according to a combination of Flynn's criteria for isolated SC fractures and a grading of forearm rotation and wrist [16]. The mean follow-up time was 24 months (range 20–27).

We found an incidence of combined fractures of 4.09%, which is similar to the incidence of 3–13% reported in other studies. The left upper extremity was more commonly affected than the right (3:2) and there was higher incidence of open fractures and nerve injuries; which is well documented in literature. Patients who suffered road traffic accidents had multiple fractures, some of which are open injuries. The patients were followed-up at second week, sixth week and third month by the operating surgeon. The final follow-up was at 24th months. Patients were examined for active/passive movements of the elbow and wrist (flexion-extension), measuring loss of motion, and the carrying angle loss (cosmetic factor). Following Flynn criteria (Table 3), there were excellent results in three (60%) cases, good result in one (20%) case and poor results in one (20%) case [16]. One patient with open elbow injury and managed with external fixation developed stiffness of elbow and had less than satisfactory results. There were no cases of failure of fixation and no delayed union or nonunion. We had one case with cubitus varus in a patient managed with closed reduction and plaster. There were two superficial pin-track infections, one in supracondylar and one in forearm

Table 3: Flynn et al. criteria modified by Templeton & Graham. Function is compared with the uninjured limb.

	Loss of elbow flexion/ extension	Loss of forearm supination/ pronation	Loss of wrist flexion/ extension	Change in carrying angle
Excellent	0 to 5	0 to 15	0 to 15	0 to 5
Good	6 to 10	16 to 30	16 to 30	6 to 10
Fair	11 to 15	31 to 45	31 to 45	11 to 15
Poor	>15	>45	>45	>15

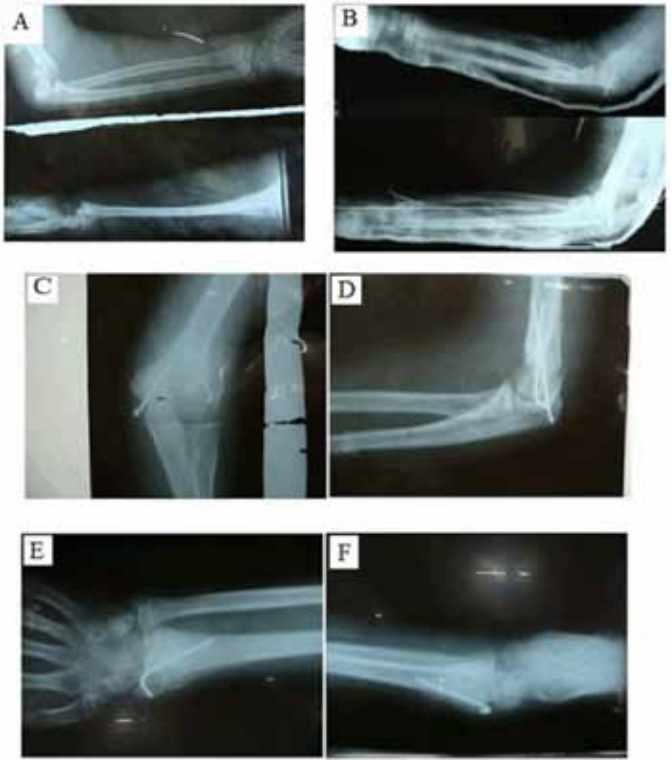


Figure 1: Radiographs showing ipsilateral supracondylar and distal and radius fracture. (A) Preoperative, and (B–F) Postoperative after pinning.

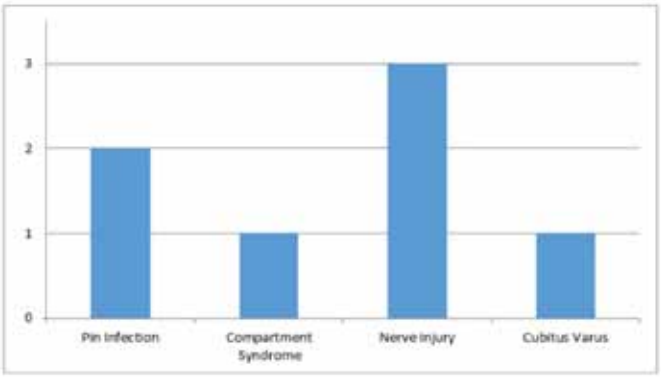


Figure 2: Complications in floating elbow injury.

region which healed after removal of the wires. The nerve injuries were all temporary and recovered by final follow-up. There were one iatrogenic ulnar nerve injuries that resolved with pin removal.

DISCUSSION

The combination of a supracondylar fracture of the humerus and an ipsilateral fracture of the forearm is rare but a severe injury in the growing child. The reported incidence varies between 3–13% [15, 16]. The upper segment injury may include supracondylar, intercondylar, lateral condyle, medial epicondyle fractures. The lower segment fractures may be also at different locations: olecranon, radial neck, Monteggia lesions, and various levels of the bone shaft and distal forearm [19, 20, 11]. Palmer et al. in their analysis of 78 supracondylar fractures found four ipsilateral fractures of the radius and ulna, two ipsilateral fractures of the radius alone and one ipsilateral midshaft ulna fracture [21]. Stanitski coined the term floating elbow for such injury, considered a high energy fracture [7, 20]. The force is so much that single fracture could not dissipate all the energy of trauma [11]. In the true sense floating elbow should include fracture supracondylar humerus with fracture of both bones forearm wherein the elbow is effectively dissociated from rest of the upper limb. However, various reports have included association of single bone fracture also in floating elbow [4, 7, 17]. The incidence of compartment syndrome is higher in these injuries [11, 22] so, the management is sometimes challenging. Blakemore et al. reported an incidence of compartment syndrome of 33% in ipsilateral displaced supracondylar humeral and forearm fractures [6].

In our patients all supracondylar fractures were of the extension type. The mechanism of this injury is usually a fall on the outstretched hand and arm with the wrist dorsiflexed and the elbow extended. The direction of the supracondylar fracture was posteromedial in 80% of our patients which is comparable to an incidence of 75% to 90% in isolated supracondylar fractures [23–24]. In general, the supracondylar component shows complete (Type III) displacement and the forearm fracture is generally seen in the distal one third. This combination of fractures is usually caused by a fall from a height, three (60%) in our series and emphasizes the fact that this association of fractures occurs as a result of high-energy trauma in most cases. Fall from standing height and road traffic accidents accounted for one patient (20%) each. The incidence of associated nerve injuries (40%) and of open fractures (40%) was higher than expected because of the severity of the injury [23, 25]. Tabak AY recommended conservative treatment of the nerve injuries as spontaneous resolution may be expected [26]. While majority of isolated displaced supracondylar fractures of the humerus are managed with closed reduction and percutaneous fixation, the treatment of a supracondylar fracture in the presence of an ipsilateral forearm fracture is still controversial [7, 12, 14, 15, 25]. Though good results have been reported after conservative treatment [15, 16] many authors consider pinning of the supracondylar fracture the best choice [5, 13, 20]. Others recommend percutaneous pinning of both proximal and distal fractures [11]. In 1960s, closed reduction and casting was recommended but an increased frequency of cubitus varus, of up to 25% of cases was observed [5, 14]. Reed in an early series (15 cases) treated all of his patients by conservative methods [5, 19]. Fowles (n=175) reported six cases of this injury, all of them were managed by pinning of the supracondylar fracture and closed reduction and cast immobilization of the forearm fracture [27]. Williamson and Cole managed the supracondylar fracture by traction or manipulative reduction and percutaneous pinning and the forearm fractures were managed by reduction and casting [15]. Reed et al. in a series of 15 patients treated conservatively reported good functional outcome [14]. Stanitski recommends early closed reduction and pinning of superior fracture and closed reduction of inferior fracture and casting [7]. Biyani A et al. reported primarily posterior slab for supracondylar facture and short arm cast for forearm fracture and olecranon pin traction or K-wire fixation for supracondylar fracture only when closed reduction failed [13]. Similarly, Reed et al. also used plaster slab for both injuries after fracture reduction [14]. Both of these studies reported cubitus varus deformity with incidence of 20% but did not find any compartment syndrome. Roposch reported three of his 18 patients with forearm fractures displaced in cast while none of the 29 cases pinned displaced [5]. Harrington et al. stated that they achieved good results from similar treatment for four of their 12 patients with floating elbow [4]. In a series of 21 cases of ipsilateral supracondylar and forearm fractures, Pierce and Hodorski in 1976 concluded that nerve injuries are a predictive factor for poor [28].

We agree with Shaw and Kasser who advocated stabilization of the elbow without exploration unless capillary refill is compromised [29]. Priority of reduction of stabilization of supracondylar fracture or forearm injury first varies among authors though no definitive study has been done in terms of outcome and complications. Templeton and Graham recommended reduction and stabilization of the supracondylar fracture first because they suggested that maintenance of reduction and access to the limb for neurovascular monitoring, dressings and the closure of open fractures may be difficult if the forearm fracture is treated first [17]. The forearm fractures were fixed first in a series by Tabak followed by closed reduction and percutaneous fixation of the supracondylar fracture [26]. This protocol was followed in the Suresh's series. We too reduced and fixed the forearm fractures first because leaving the forearm dangling during reduction of supracondylar fracture can cause more soft tissue injury of forearm and can increase chance of compartment syndrome [30]. In addition, we found subsequent reduction of supracondylar fractures

easier. If the forearm fracture is not reduced and fixed first, it will remain mobile during flexion of the elbow and rotational maneuvers. We managed 1 (20%) supracondylar fracture conservatively who, however, developed cubitus varus. The remaining 4 (80%) were managed with pinning. The distal end radius fractures were managed conservatively with closed reduction and casting (n =1) and percutaneous pinning (n=3). We stabilized the forearm and radial shaft fractures with pinning and did not see any loss of reduction. The rate of remanipulation of the forearm fractures after closed reduction and immobilization in a cast is reported to be between 7% and 15% [31, 32]. Various techniques of K-wire fixation such as crossed K-wires, lateral two K-wires have been described in literature for better biomechanical stability and to decrease chance of potential iatrogenic nerve injury [26]. There was one case of ulnar nerve injury associated with introduction of the medial wire, the nerve function returned after removal of the K-wires. Although fixation with crossed wires is more stable, there is greater risk of injuring the ulnar nerve if the medial wire is passed through the bone blindly [33]. We had no complications such as loss of reduction or neurovascular injury in forearm fractures after K-wire fixation. We therefore recommend stabilization of displaced both bone forearm fractures which are associated with supracondylar fractures. We achieved good results with closed reduction and application of plaster for distal radial epiphyseal injury. The overall results were excellent or good in four (80%) patients but poor in one (20%) patients.

CONCLUSION

Ipsilateral supracondylar and forearm fractures area result of high energy trauma. The injury is uncommon and treatment recommendations are controversial. However, these injuries need prompt management and close observation for early signs and symptoms of development of compartment syndrome. In this series the good results obtained after surgical treatment allow us to state that floating elbow injuries are best managed aggressively with surgical stabilization. However, given the scarce sources in a third world setting as ours, we suggest conservative management in selected cases only.

****

Acknowledgements
I must thank my tender child patients for allowing me to spend time with them to compile this paper. I must also thank Dr Shabir Dhar, consultant orthopedician, who is a teacher and a senior colleague and helped me in compiling this paper in its final stages.

Author Contributions
Shahid Hussain – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright

REFERENCES

1. Herring JA, editor. Upper extremity injuries. In: Tachdjian's Pediatric Orthopaedics. 4th ed. Vol. 3. Philadelphia: W.B. Saunders 2008.
2. Skaggs DL, Flynn JF. Supracondylar Fracture of the Distal Humerus. In: Beaty JH, Kasser JR, (editors) Rockwood and Wilkins Fractures in Children, 7th Edition Vol. 3. Philadelphia, Lippincott William and Wilkins 2010:487–531.
3. Mann DC, Rajmaira S. Distribution of physeal and non-physeal fractures in 2,650 long-bone fractures in children 0-16 years. J Pediatr Orthop 1990;10(6):713–6.
4. Harrington P, Sharif I, Fogarty EE, Dowling FE, Moore DP. Management of the floating elbow injury in children. Simultaneous ipsilateral fractures of the elbow and forearm. Arch Orthop Trauma Surg 2000;120(3-4):205–8.
5. Roposch A, Reis M, Molina M, et al. Supracondylar fractures of the humerus associated with ipsilateral forearm fractures in children: A report of forty seven cases. J Pediatr Orthop 2001;21(3):307–12.
6. Blakemore LC, Cooperman DR, Thompson GH, Wathey C, Ballock RT. Compartment syndrome in ipsilateral humerus and forearm fractures in children. Clin Orthop Relat Res 2000;(376):32–8.
7. Stanitski CL, Micheli LJ. Simultaneous ipsilateral fractures of the arm and forearm in children. Clin Orthop Relat Res 1980;(153):218–22.
8. Piggot J, Graham HK, McCoy GF. Supracondylar fractures of the humerus in children. Treatment by straight lateral traction. J Bone Joint Surg Br 1986;68(4):577-83.
9. Buckley SL, Gotschall C, Robertson W Jr, et al. The relationships of skeletal injuries with trauma score, injury severity score, length of hospital stay, hospital charges, and mortality in children admitted to a regional pediatric trauma center. J Pediatr Orthop 1994;14(4):449-53.
10. Malheiros DS. Estudo descritivo de lesões ortopédicas em crianças e adolescentes em centro de atendimento

nível II [tese]. Belo Horizonte: Universidade Federal de Minas Gerais. Faculdade de Medicina 2009.
11. Ring D, Waters PM, Hotchkiss RN, Kasser JR. Pediatric floating elbow. J Pediatr Orthop 2001;21(4):456-9.
12. Papavasiliou V, Nenopoulos S. Ipsilateral injuries of the elbow and forearm in children. J Pediatr Orthop 1986;6(1):58-60.
13. Biyani A, Gupta SP, Sharma JC. Ipsilateral supracondylar fracture of humerus and forearm bones in children. Injury 1989;20(4):203-7.
14. Reed FE Jr, Apple DF Jr. Ipsilateral fractures of the elbow and forearm. South Med J 1976;69(2):149-51.
15. Williamson DM, Cole WG. Treatment of ipsilateral supracondylar and forearm fractures in children. Injury 1992;23(3):159-61.
16. Flynn JC, Matthews JG, Benoit RL. Blind pinning of displaced supracondylar fractures of the humerus in children. Sixteen years' experience with long-term follow-up. J Bone Joint Surg Am 1974;56(2):263-72.
17. Templeton PA, Graham HK. The 'floating elbow' in children. Simultaneous supracondylar fractures of the humerus and of the forearm in the same upper limb. J Bone Joint Surg Br 1995;77(5):791-6.
18. Gustilo RB, Anderson JT. Prevention of infection: The treatment of one thousand and twenty-five open fractures of long bones. Retrospective and prospective analyses. J Bone Joint Surg Am 1976;58(4):453-8.
19. John A. Ogden. Skeletal Injury in the Child, 3ed. Springer 2000.
20. Suresh S. Management of "floating elbow" in children. Indian J Orthop 2007;41(4):386-9.
21. Palmer EE, Niemann KM, Vesely D, Armstrong JH. Supracondylar fracture of the humerus in children. J Bone Joint Surg Am 1978;60(5):653-6.
22. Blakemore LC, Cooperman DR. The incidence of compartiment syndrome in ipsilateral fractures of the humerus and forearm in children. In proceedings of the Orthopaedic Trauma Association (OTA) conference (Session VII - Pediatrics): 10 October 1998.
23. Wilkins KE, Beaty JH, Chambers HG, Toniolo RM. Fractures and dislocations of the elbow region. In: Rockwood Jr CA, Wilkins KE, Beaty JH, eds. Fractures in children. Vol 3, Fourth ed. Philadelphia: Lippincott-Raven Publishers 1996:653-904.
24. Kasser JR. Percutaneous pinning of supracondylar fractures of the humerus. Instr Course Lect 1992;41:385-90.
25. Wilkins KE. The operative management of supracondylar fractures. Orthop Clin North Am 1990;21(2):269-89.
26. Tabak AY, Celibi L, Muratli HH, Yagmurlu MF, Aktekin CN, Biçimoglu A. Closed reduction and percutaneous fixation of supracondylar fracture of the humerus and ipsilateral fracture of the forearm in children. J Bone Joint Surg Br 2003;85(8):1169-72.
27. Fowles JV, Kassab MT. Displaced supracondylar fractures of the elbow in children. A reporton the fixation of extension and flexion fractures by two lateral percutaneous pins. J Bone Joint Surg Br 1974;56B(3):490-500.
28. Pierce RO Jr, Hodurski DF. Fractures of the humerus, radius and ulna in the same extremity. J Trauma 1979;19(3):182-5.
29. Shaw BA, Kasser JR, Emans JB, Rand FF. Management of vascular injuries in displaced supracondylar humerus fractures without arteriography. J Orthop Trauma 1990;4(1):25-9.
30. Dhoju D, Shrestha D, Parajuli N, Dhakal G, Shrestha R. Ipsilateral supracondylar fracture and forearm bone injury in children: A retrospective review of thirty one cases. Kathmandu Univ Med J (KUMJ) 2011;9(34):11-6.
31. Voto SJ, Weiner DS, Leighley B. Redisplacement after closed reduction of forearm fractures in children. J Pediatr Orthop 1990;10(1):79-84.
32. Kasser JR. Forearm fractures. Instr Course Lect 1992;41:391-6.
33. Wilkins KE. Fractures and dislocations of the elbow region. In: Rockwood CA, Wilkins KE, King RE, editors. Fractures in children. New York: JB Lippincott 1991. p.509-828.

CASE SERIES OPEN ACCESS

Acute renal failure in two cases with hypothyroidism related rhabdomyolysis

Davut Akin, Sehmus Ozmen

ABSTRACT

Introduction: We describe two cases of acute renal failure due to rhabdomyolysis associated with hypothyroidism. Hypothyroidism usually leads to muscular disorders, otherwise rhabdomyolysis is quite rare. Case Series: This case report describes two cases of acute renal failure due to rhabdomyolysis associated with hypothyroidism in a 72-year-old male and a 36-year-old female in whom muscle enzyme levels were typical of rhabdomyolysis. Other reasons of rhabdomyolysis were excluded. Renal functions were recovered in both cases after treatment. Conclusion: Hypothyroidism must be considered in patients presenting with acute renal failure and elevated muscle enzymes even in absence of additional precipitating factor.

Keywords: Acute Renal Failure (ARF), Hypothyroidism, Rhabdomyolysis

How to cite this article

Akin D, Ozmen S. Acute renal failure in two cases with hypothyroidism related rhabdomyolysis. International Journal of Case Reports and Images 2014;5(3):195–197.

Davut Akin[1], Sehmus Ozmen[2]
Affiliations: [1]MD Assistant Professor, Department of Nephrology, Denizli State Hospital, 32100 Denizli, Turkey; [2]Dicle University School of Medicine and Diyarbakır Memorial Hospital.
Corresponding Author: Dr. Davut Akin, Department of Nephrology, Denizli State Hospital, 32100-Denizli, Turkey; Tel: +90 05055251124; Email: drdavutakin@gmail.com

Received: 28 September 2013
Accepted: 02 November 2013
Published: 01 March 2014

doi:10.5348/ijcri-2014-03-471-CS-2

INTRODUCTION

Rhabdomyolysis is an injury of skeletal muscle resulting in leakage of cell content into bloodstream. Although muscular disorders are usual in hypothyroidism, rhabdomyolysis due to hypothyroidism is very rare and only a few cases have been reported [1–3]. Non-exertional and non-traumatic causes of rhabdomyolysis include drugs, toxins, infections, electrolyte abnormalities, endocrinopathies, inflammatory and myopathies [4]. Most of the cases of rhabdomyolysis due to hypothyroidism and acute renal failure (ARF) had additional precipitating factor [5]. Only a few cases without precipitating factor have been reported [3, 6–9].

We present two cases with ARF due to hypothyroidism associated rhabdomyolysis with no additional precipitating factor.

CASE SERIES

Case 1: A 72-year-old male was admitted to hospital because of confusion, decreased urine output and dark brown urine color for 10 days. A physical examination revealed dry skin and generalized body swelling. His blood pressure was 90/60 mmHg and his pulse rate was 60/min, with an oral temperature 37°C. Abnormal laboratory results were as follows: serum urea 217 mg/dL (10–45 mg/dL), creatinine 11.3 mg/dL (0.6–1.3 mg/dL), K 6.5 mmol/L (3.5–5.1 mmol/L), peak creatine kinase 6024 U/L (29–200 U/L), lactate dehydrogenase (LDH) 449 U/l (125–243 U/l), aspartate aminotransferase (AST) 75 U/L (10–40 U/L), alanine aminotransferase (ALT) 24 U/L (10–35 U/L). Urine analysis revealed dark brown urine with a positive dipstick reaction for blood and granular casts. A history of hashimoto thyroiditis for 40 years was obtained from his medical records. He was not taking thyroxine replacement regularly, but hypothyroidism was detected. Serum free T3, free T4 and

TSH levels were 0.02 ng/dL (1.8-4.6 ng/dL), 0.065 ng/dL (0.8–1.8 ng/dL) and 100 μIU/mL (0.27–4.20 μIU/mL). He has high serum concentrations of antibodies to anti microsomal AB and antithyroglobulin 500 U/mL (0–60 U/mL and 390 U/mL (0–20 U/mL), respectively. Other laboratory tests were normal. Intravenous fluids and L-Thyroxin replacement (100 μg/day) was started. Two sessions of hemodialysis was performed because of uremic acidosis and mental changes. Renal ultrasonography was normal. He experienced a polyuria period after thyroxin replacement. Serum creatine kinase level was normalized in a week of treatment. The final serum creatinine was 1.1 mg/dL.

Case 2: A 36-year-old female admitted with myalgia and decreased daily urine. Her blood pressure 10/70 mmHg and rhythm was normal. Abnormal laboratory results were as follows: serum urea 189 mg/dL, creatinine 10 mg/dL, serum sodium 126 mmol/L, serum potassium 5 mmol/L, AST 337 U/L, ALT 82 U/L, peak CK 4267 U/L, LDH 804 U/L. Urine analysis revealed granular casts no RBC and a positive dipstick reaction for blood. She had a history of subtotal thyroidectomy due to multinodular goiter one year ago. She was not taking thyroxin replacement regularly. Thyroid hormone profile revealed a TSH 100 μIU/mL (0.27–4.20 μIU/mL), FT3 0.1 pmol/l (3.1–7.7 pmol/L), and FT4 2.2 pmol/L (9–18 pmol/L). She had low central venous pressure. Fluids (%0.09 NaCl 120 mL/hr) and bicarbonate infusion (4.2 mmol/ hr) was administered intravenously. No hemodialysis was performed. Thyroxin replacement (100 μg/day) was started. Her final serum creatinine was 1.1 mg/dL, Serum creatine kinase returned normal level.

Both of the patients did not have a past medical history of diabetes, hypertension, or renal disease. Non-exertional and non-traumatic causes of rhabdomyolysis include drugs, toxins, infections, electrolyte abnormalities. Antinuclear antibodies (ANA), anti-Ro, anti-La, anti-Sm, anti-Jo-1 or anti-ribonucleoprotein (RNP) antibodies were negative. Other laboratory tests (such as lipid profile and blood sugar) were normal. Renal ultrasonography revealed normal sized kidneys with normal echogenicity. No additional precipitating factor both of patients could be related with rhabdomyolysis was detected.

DISCUSSION

This case series describes two patients suffering from rhabdomyolysis due to hypothyroidism, with no additional precipitating factor. Rhabdomyolysis was defined as creatine kinase levels above five times the upper limit of normal and renal findings in both cases. Although the main features of rhabdomyolysis are muscular symptoms and increased creatine kinase concentrations, it can become a life-threatening disorder when complicated by ARF. As a cause of rhabdomyolysis, disorders such as collagen disease (e.g., polymyositis), ingestion of massive alcohol, other agents, infection, and trauma were excluded in our cases from medical history.

The exact cause of rhabdomyolysis (abnormal glycogenolysis, mitochondrial oxidative metabolism, and triglyceride turnover, impair muscle function) in hypothyroidism remains unclear [10].

There is a wide variation in the clinical presentation of rhabdomyolysis. The classical triad of symptoms is muscle pain, weakness, and reddish-brown urine. However, these classical features are seen in fewer than 10% of the patients [4]. One of our patients also had muscle pain, weakness, and reddish-brown urine, but the other patient did not report a muscle pain and weakness probably because of confused mental status.

Rhabdomyolysis and ARF due to hypothyroidism is a rare entity. There are a few reported cases in literature [3, 6]. In most of these cases, rhabdomyolysis appears to have been precipitated by additional factors such as vigorous exercise [2, 5], trauma, anti-hyperlipidemic agents, or metabolic disorder. Although hyponatremia is a well-known aetiology that may cause rhabdomyolysis, hyponatremia level in our second case is not as severe as previous papers reporting hyponatremia as cause of rhabdomyolysis [11]. Therefore, hyponatremia may not be the main factor of rhabdomyolysis in our case. To our knowledge only 11 cases with ARF secondary to hypothyroidism-related rhabdomyolysis who had no additional precipitating factor have been reported in literature to date [3, 6–9].

CONCLUSION

In conclusion, hypothyroidism must be considered in patients presenting with acute renal failure and elevated muscle enzymes even in absence of additional precipitating factor.

****

Author Contributions

Davut Akin – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Sehmus Ozmen – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

1. Khaleeli AA, Griffith DG, Edwards RH. The clinical presentation of hypothyroid myopathy and its relationship to abnormalities in structure and function of skeletal muscle. Clin Endocrinol (Oxf) 1983;19(3):365–76.
2. Riggs JE. Acute exertional rhabdomyolysis in hypothyroidism: The result of a reversible defect in glycogenolysis? Mil Med 1990;155(4):171–2.
3. Kisakol G, Tunc R, Kaya A. Rhabdomyolysis in a patient with hypothyroidism. Endocr J 2003;50(2):221–3.
4. Giannoglou GD, Chatzizisis YS, Misirli G. The syndrome of rhabdomyolysis: Pathophysiology and diagnosis. Eur J Intern Med 2007;18(2):90–100.
5. Nelson SR, Phillips AO, Hendry BM. Hypothyroidism and rhabdomyolysis in a marathon runner. Nephrol Dial Transplant 1993;8(4):375–6.
6. Altay M, Duranay M, Ceri M. Rhabdomyolysis due to hypothyroidism. Nephrol Dial Transplant 2005;20(4):847–8.
7. Barahona MJ, Mauri A, Sucunza N, Paredes R, Wägner AM. Hypothyroidism as a cause of rhabdomyolysis. Endocr J 2002;49(6):621–3.
8. Kar PM, Hirani A, Allen MJ. Acute renal failure in a hypothyroid patient with rhabdomyolysis. Clin Nephrol 2003;60(6):428–9.
9. Birewar S, Oppenheimer M, Zawada ET Jr. Hypothyroid acute renal failure. S D J Med 2004;57(3):109–10.
10. Kaminski HJ, Ruff RL. Endocrine myopathies. In: Myology, 2d ed, Engel, AG, Franzini-Armstrong, C (Eds), McGraw-Hill; New York 1994, p,1741.
11. Strachan P, Prisco D, Multz AS. Recurrent rhabdomyolysis associated with polydipsia-induced hyponatremia - A case report and review of the literature. Gen Hosp Psychiatry 2007;29(2):172–4.

CASE REPORT OPEN ACCESS

Use of diode lasers in treatment of oral submucous fibrosis: A new concept in surgical management

Sanjay Asnani, Uma Mahindra, Rakesh Oswal

ABSTRACT

Introduction: Oral submucous fibrosis is chronic, insidious disease affecting the oral cavity and sometimes the pharynx and rarely the tongue. Oral Submucous fibrosis is a well known clinical entity since the time of "Sushruta" as 'Vidari'. Case Report: A 20-year-old patient with oral submucous fibrosis was treated with contact diode laser under local anesthesia and was followed up for six months. Conclusion: Oral submucous fibrosis is an established precancerous condition with increased prevalence in the Indian subcontinent. The treatment of oral submucous fibrosis is a challenging task for a clinician. Different authors have suggested variety of treatment modalities and have claimed success rates; still there is no universally acceptable protocol for the management of oral submucous fibrosis. In this case transection of bands was done by contact diode lasers under local anaesthesia and it offered good results. Diode lasers offered excellent results and had many advantages over conventional surgical treatment.

Keywords: Diode laser, Oral submucous fibrosis, Local anaesthesia

Sanjay Asnani[1], Uma Mahindra[2], Rakesh Oswal[3]
Affiliations: [1]MDS, Department of Oral & Maxillofacial Surgery, YCMM Dental College, Ahmednagar, Maharashtra, India; [2]Professor & Head, Department of Oral & Maxillofacial Surgery, Rural Dental College, Loni, (Bk), Taluka, Rahata, Ahmednagar, Maharashtra, India; [3]Reader, Department of Oral & Maxillofacial Surgery, YCMM Dental College, Ahmednagar, Maharashtra, India.
Corresponding Author: Dr. Sanjay Asnani, Department of Oral & Maxillofacial Surgery, YCMM Dental College, Ahmednagar 414003, Maharashtra, India; Mobile No: 09890050050; E-mail: drsanjayasnani@gmail.com

Received: 22 May 2013
Accepted: 18 July 2012
Published: 01 March 2014

How to cite this article

Asnani S, Mahindra U, Oswal R. Use of diode lasers in treatment of oral submucous fibrosis: A new concept in surgical management. International Journal of Case Reports and Images 2014;5(3):198–201.

doi:10.5348/ijcri-2014-03-472-CR-3

INTRODUCTION

Oral submucous fibrosis is a well known clinical entity since the time of "Sushruta" as 'Vidari' [1]. This condition is predominantly found in the Indian subcontinent. Joshi in 1953 was the first person to describe this entity in India. The highest incidence is found in the state of Kerala with an overall prevalence rate of 2.5% in various states of the country. JJ Pindborg defined it as "an insidious chronic disease affecting any part of the oral cavity and sometimes the pharynx. Although occasionally preceded by and/or associated with vesicle formation, it is always associated with juxta epithelial inflammatory reaction followed by a fibro elastic change of lamina propria, with epithelial atrophy leading to stiffness of oral mucosa and causing trismus and inability to eat" [2].

There is no definite treatment for this condition. The various treatment modalities are mainly medical, surgical, or a combination of both. Conservative/medical modality is the treatment of choice in patients with mild to moderate limitation in mouth opening. The non-surgical line of treatment includes usage of gold [4], iodides, hyluronidase, placental extract, and steroids like hydrocortisone and triamcinolone, vitamins, iron supplements [5]. Whereas surgical treatment is the method of choice in patients with marked limitation of mouth opening or in patients not responding to the conservative management [6]. Many surgical modalities like dermal graft [7], tongue flap [8], nasolabial flap [9] etc. are in vogue to cover the surgical defect created by

transection of fibrous bands in oral mucosa.

Considering the limited success of the various treatment modalities and being aware of the fact that surgical treatment is the only palliative treatment having a fixed, limited target of relieving the trismus to enable the patient to overcome the morbidity, there is perpetual quest for the newer surgical protocols.

We report a case of oral submucous fibrosis treated with contact diode laser under local anaesthesia for transection of fibrous bands.

CASE REPORT

A 20-year-old male patient was reported in the department of oral and maxillofacial surgery with a chief complain of limited mouth opening since last three years and burning sensation on consumption of hot and spicy food. A detailed history was taken regarding tobacco chewing habit and dietary habit. It was revealed that patient consumed tobacco in the form of quid since last five years and also consumed excessive chillies. On examination fibrous bands were palpated on the right and left buccal mucosa extending up to the retromolar region. His preoperative mouth opening was recorded and was 18 mm (Figure 1). Biopsy was taken under local anesthesia to confirm oral submucous fibrosis. After confirmation of biopsy report treatment was explained to the patient and transection of fibrous bands was done bilaterally by contact diode laser under local anesthesia and the raw area was not grafted and was allowed to epithelize on its own (Figure 2). Patient was kept on oral antibiotics and analgesics for 5 days and postoperative physiotherapy exercise was started after 48 hours, atleast for 7–8 times in a day at the interval of one hour. Patient was told to continue the exercise for a period of one year. Mouth opening was recorded at the interval of 1 week, 1 month and 6 months. A significant improvement in mouth opening was recorded at the end of 6 months and it was around 4 cm (Figure 3). Healing of the surgical site was completed in 4 weeks and also there was relief from burning sensation as the patient was kept on oral antioxidants for the same.

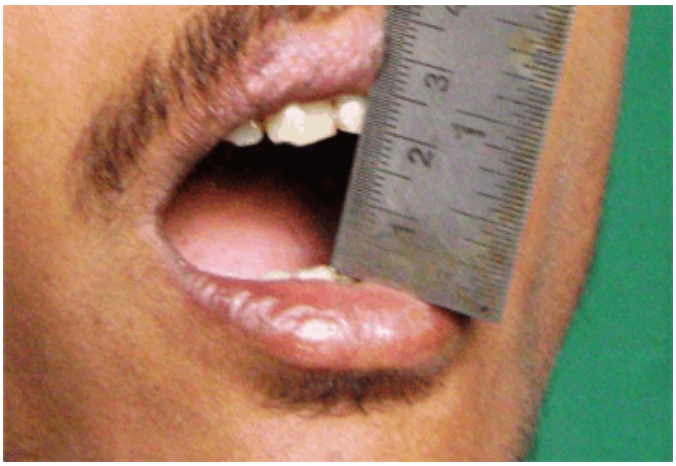


Figure 1: Pre operative mouth opening.

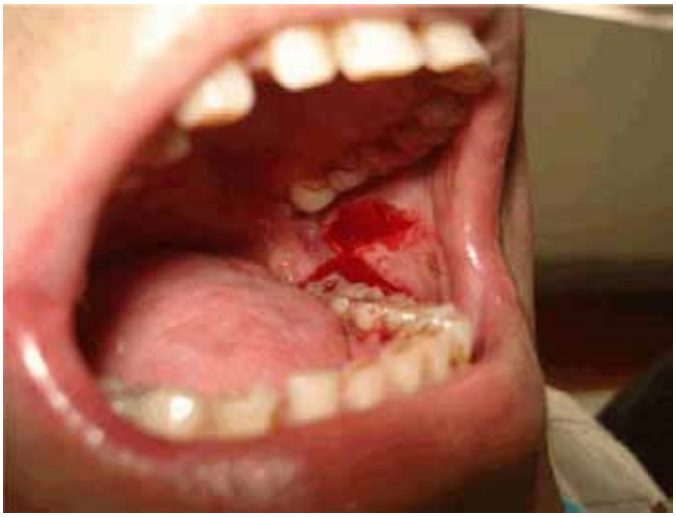


Figure 2: Transection of fibrous bands by diode laser.

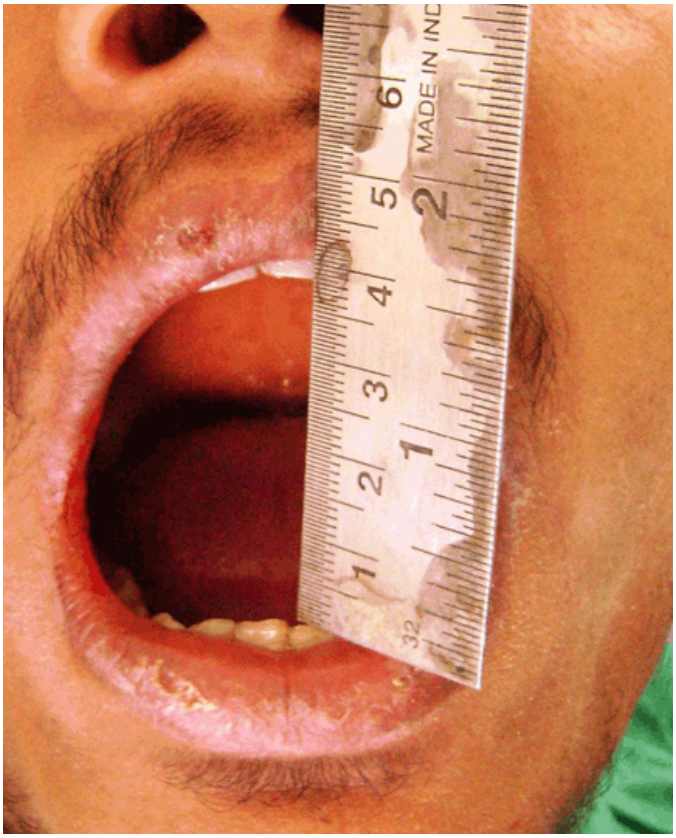


Figure 3: Mouth opening at the end of 6 months.

DISCUSSION

Oral submucous fibrosis is an established precancerous condition with increased prevalence in the

Indian subcontinent. An estimated 2.5 million people suffer from this disease in India [3]. Prevalence rates of oral submucous fibrosis (OSMF) ranging from 0 to 17.6 % have been reported in various population subsets. Betel nut chewing habit is the dominant etiological factor as majority of the patients are habituated to betel nut chewing in one form or the other.

The onset of the disease is insidious and is often of 2 to 5 years duration. The most common initial symptom is burning sensation of the oral mucosa, aggravated by spicy food. Vesiculation, excessive salivation, ulceration, altered pigmentation, recurrent stomatitis, defective gustatory sensation, and dryness of the mouth have also been indicated as early symptoms

The treatment of oral submucous fibrosis is a challenging task for a clinician. All the documented treatment modalities, ranging from medicinal treatment to surgical treatment are purely symptomatic in nature and total cure of the disease is still elusive. Different authors have suggested variety of treatment modalities and have claimed success rates; still there is no universally acceptable protocol for the management of OSMF. This is mainly due to the fact that the etiology of the disease is not fully understood and the disease is progressive in nature.

The basic aim of any type of treatment modality is to relieve the symptoms which include hampered function in the form of trismus, burning sensation in mouth, difficulty in mastication, deglutition and speech. Relieving such type of symptoms makes it a more difficult surgical problem. Surgeon should not only aim to relieve trismus but also should monitor for malignant transformation of this condition.

Recently, diode lasers have been used for excision of soft tissue lesions in oral cavity, also have been used for gingivectomies, curettage of the pockets, debridement of the root canals, for carrying out frenectomies, bleaching, etc. But there are a few literatures available regarding their role in treatment of oral submucous fibrosis. In present case transection of bands was done by contact diode lasers under local anesthesia and it offered good results. There was significant improvement in mouth opening at the end of six months. The diode laser which was used in this case was of 5 watts and was kept on continous mode. Diode laser offered several advantages over conventional surgical procedures like:

(1) The procedure done is a minor outpatient procedure under local anesthesia.
(2) Hemostatic nature of the surgery which allows surgery to be performed more precisely and accurately because of increased visibility and accessibility of the surgical site.
(3) Decreased post-operative pain and swelling.
(4) Causes a reduction in bacterial counts thereby promoting quicker, more predictable healing with minimal post operative infection and inflammation.
(5) Leads to healing with minimal scarring.

The only disadvantage of this technique is availability and its cost effectiveness.

In past KTP 532 lasers have been used for the treatment of oral submucous Fibrosis and have shown good results but in all the studies the procedure was carried out under general anesthesia which required increased hospital stay whereas in our case the procedure was carried out under local anaesthesia which had many advantages as mentioned above.

Diode contact laser system can be used to rehabilitate grade III and early grade IV cases of OSMF. However, further more research is required for this system. A large sample size and long-term follow-up would give better insights for its use.

CONCLUSION

Till today, there is no modality which can be labeled as definitive treatment for oral submucous fibrosis. But this protocol is worth of consideration and may give better results keeping in view the importance of post-operative mouth opening exercises and patient compliance.

****

Author Contributions

Sanjay Asnani – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Uma Mahindra – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Rakesh Oswal – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Mukherjee AL. Oral submucous fibrosis. A search for etiology. Ind J Otolaryngology 1972;24:1:11–5.

2. Gupta SC. “Mist” an etiological factor in oral submucous fibrosis. Ind J Otolaryngology 1978;30(1):5–6.
3. Shahid R Aziz. Oral submucous fibrosis: case report and review of diagnosis and treatment. J Oral Maxillofac Surg 2008;66:2386–9.
4. Marawetz G, Katsikers N, Weinberg S, Listrom R. Oral submucous fibrosis. J Oral Maxillofacial Surgery 1987;16:609–14.
5. Borle RM, SR Borle. Management of Oral Submucous Fibrosis: A conservative approach. J Oral Maxillofac Surg 1991;49:788–9.
6. Khanna JN, Andrade NN. Oral submucous fibrosis: a new concept in surgical management. Report of 100 cases. Int J Oral Maxillofac Surg 1995;24:433–9.
7. Yen DJ. Surgical treatment of submucous fibrosis. Journal of Oral Surgery 1982 September:269-72.
8. Golhar S, Manohar MN, Narkhede S. Tongue flap in oral submucous fibrosis. Ind J Otolaryngol 1989;41:104–7.
9. Kavarana NM, Bhathena HM. Surgery for severe trismus in submucous fibrosis. British Journal of plastic surgery 1987;40:407–9.

CASE REPORT OPEN ACCESS

Medical school finals, nerves and vomiting: Medical student survives Boerhaave's syndrome with recurrence one week after initial presentation

Eshan Oderuth, Kevin Ilo, Munir Somji, Thomas Holme

ABSTRACT

Introduction: Boerhaave's syndrome is a serious condition that can be fatal due to commonly nonspecific symptoms and delayed treatment. This rare syndrome is a result of a spontaneous esophageal rupture. Case Report: We describe a case of a medical student who developed Boerhaave's syndrome on the day of his finals. Initially, the medical student's symptoms were misdiagnosed when they sought medical attention. Despite the medical student being clinically well, their medical knowledge prompted a surgical opinion and Boerhaave's syndrome was confirmed. Treatment was conservative, however, its recovery was complicated with a recurrence of symptoms shortly after the initial episode. Conclusion: This case highlights and discusses the importance of esophageal rupture to be considered as a cause of chest pain. Prompt diagnosis is of utmost importance. If conservative treatment is commenced, adequate time for healing should be permitted.

Keywords: Boerhaave's Syndrome, Esophageal rupture, Esophageal perforation, Upper third esophageal rupture

Eshan Oderuth[1], Kevin Ilo[2], Munir Somji[3], Thomas Holme[4]
Affiliations: [1]Foundation Year 2, General Surgery, Queen Elizabeth Hospital, London, England; [2]Foundation Year 2, Trauma and Orthopaedics, Queen Elizabeth Hospital, London, England; [3]Foundation Year 2, General Surgery, St Thomas's Hospital, London, England; [4]Foundation Year 2, Accident and Emergency, St Thomas' Hospital, London, England.
Corresponding Author: Eshan Nazir Hassan Oderuth, 37 Harvey Road, Worthing, West Sussex, ENGLAND, BN12 4DS; Ph: +447732376940; Email: eshan786@msn.com

Received: 17 November 2012
Accepted: 28 February 2013
Published: 01 March 2014

How to cite this article

Oderuth E, Ilo K, Somji M, Holme T. Medical school finals, nerves and vomiting: Medical student survives Boerhaave's syndrome with recurrence one week after initial presentation. International Journal of Case Reports and Images 2014;5(3):202–206.

doi:10.5348/ijcri-2014-03-473-CR-4

INTRODUCTION

Physician Herman Boerhaave discovered the first case of Boerhaave's syndrome in 1724 following the autopsy of the General Admiral of the Dutch Fleet [1]. Boerhaave's syndrome is defined as a spontaneous esophageal rupture resulting from high intra-esophageal pressures exceeding that of the intrathoracic cavity [2]. It is a life-threatening condition with high mortality rates (40–60% > 48 hours presentation) often due to delayed presentation, misdiagnosis and severity. Boerhaave's syndrome occurs commonly in the postero-lateral distal third of the esophagus possibly due to the muscle and neurovascular supply to that area [2]. It typically presents with the Meckler Triad of left sided chest pain, vomiting and subcutaneous emphysema. However, atypical presentations are not uncommon [2, 3]. We describe the remarkable case of a medical student being afflicted with Boerhaave's syndrome during his medical school finals complicated with a recurrence of the condition. He was treated conservatively on both admissions.

CASE REPORT

A previously well 24-year-old final year male medical student presented with central chest pain. On morning of his medical finals, he was nervous and vomited once and wretched seven times. He then coughed and experienced a sharp sudden onset retrosternal chest pain which was

constant in nature and rated 6/10 in severity. The patient attended his exam, during which he experienced the uncomfortable feeling of crackling down his throat while ingesting water. He felt unwell after his exam and was unable to fully inspire due to on-going chest pain. The patient palpated his neck which felt like 'bubble wrap'. He then auscultated his chest himself and found his heart sounds resembled crackles (Hamman's crunch). The patient's housemates concurred with the findings, so he attended Accident and Emergency department and was seen by the general physician triage. The patient's past medical history included asthma. He had a history of smoking five cigarettes a day with moderate alcohol intake. Observations were unremarkable. The general physician found no abnormality after auscultating his chest and palpated lymph nodes on neck examination. Throat examination revealed erythema. Despite the patient's and his accompanier's reservations, the general physician diagnosed a viral infection and advised to return in five days if his symptoms would not improve.

The patient returned home. However while taking his dinner, he experienced severe odynophagia describing the sensation like 'swallowing glass'. The following day, his symptoms persisted so he consulted his surgical tutor whose registrar palpated the subcutaneous emphysema in his neck with no lymphadenopathy. An urgent X-ray of the neck revealed air in the subcutaneous tissues. Computed tomography (CT) scan of chest with contrast and Gastrografin swallow revealed a small leak outside the esophagus and pneumomediastinum (Figure 1). Blood tests were unremarkable. The patient was transferred to a tertiary upper gastrointestinal centre with suspected Boerhaave's syndrome. The tear was thought to be in the upper third of the esophagus. He was kept nil by mouth, supplemented with intravenous fluids and given prophylactic daily intravenous tazocin, metronidazole and fluconazole. Gastrografin swallow on the fifth day revealed no obvious leak, his symptoms had resolved. The patient was discharged with advice of a liquid diet for one week building to solids slowly thereafter.

The patient's symptoms returned spontaneously two days post discharge on the eve of his surgical finals. Subcutaneous emphysema was found on examination. Observations, blood tests and arterial blood gas (ABG) showed no abnormality. Computed tomography scan of chest with IV contrast and Gastrografin swallow showed pneumomediastinum but no leak of Gastrografin (Figure 2 and Figure 3). Furthermore, there was the dilemma of whether he would be able to sit his exam which was at the same tertiary centre. He was given a stat bag of intravenous fluids, the aforementioned intravenous antibiotics, kept nil by mouth and allowed home to rest. He was admitted directly after his surgical finals and treated conservatively. Nutritional support was given via a nasogastric tube which was inserted under radiological guidance. The patient's symptoms improved and a Gastrografin swallow study on day-10 was normal. Soft diet was commenced which he tolerated. The patient's second hospital stay was 12 days. He continued with a fork mashable soft diet until his three month follow-up, which was satisfactory, returning to a normal diet thereafter. Thankfully, the patient passed his finals and continues to work as a doctor (Figure 4 and Figure 5).

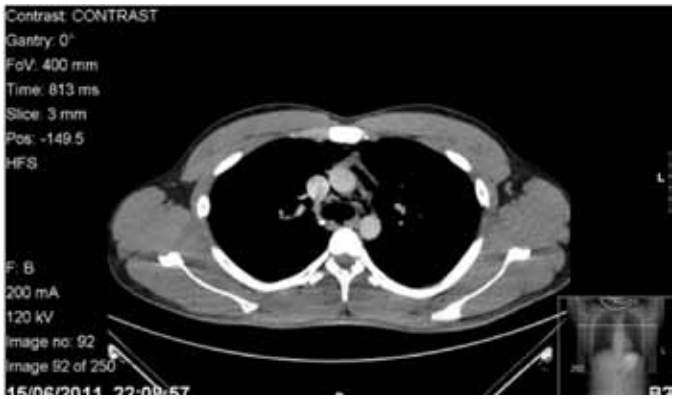


Figure 1: Computed tomography scan of chest with Gastrografin contrast on initial admission illustrating a pneumomediastinum with leak of contrast into mediastinum.

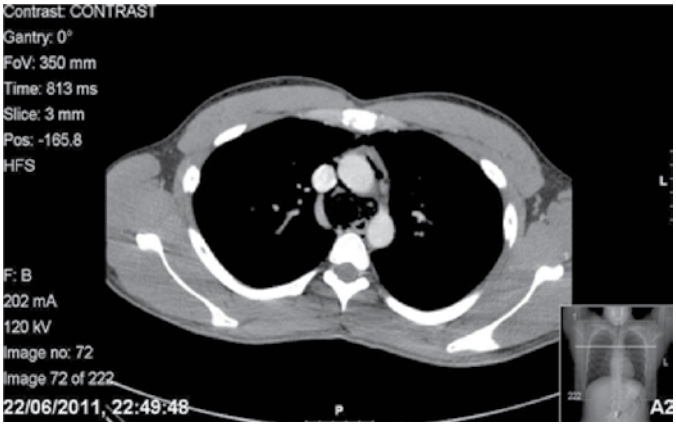


Figure 2: Computed tomography scan of chest with Gastrografin contrast on second admission illustrating a pneumomediastinum with no leak of contrast.

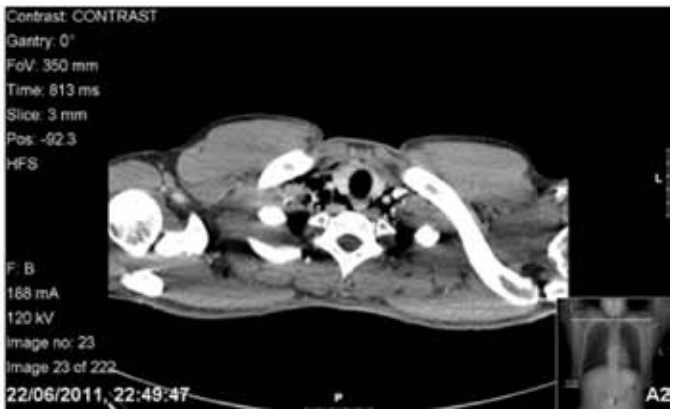


Figure 3: Computed tomography scan of chest with Gastrografin contrast on second admission illustrating surgical emphysema in the neck with no leak of contrast.

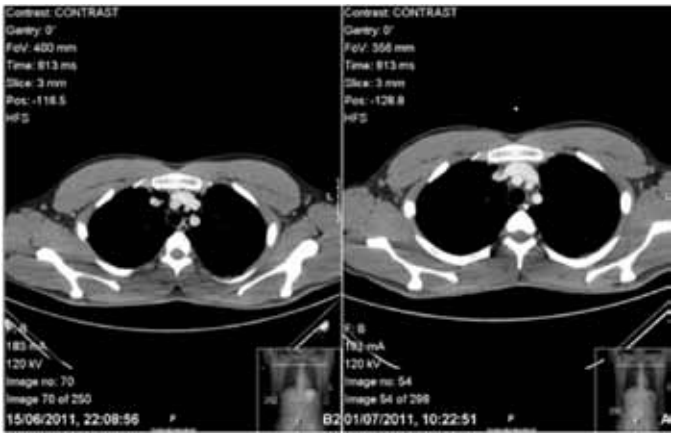


Figure 4: Computed tomography scan of chest with Gastrografin contrast; comparison of image on admission and image after resolution showing no pneumomediastinum.

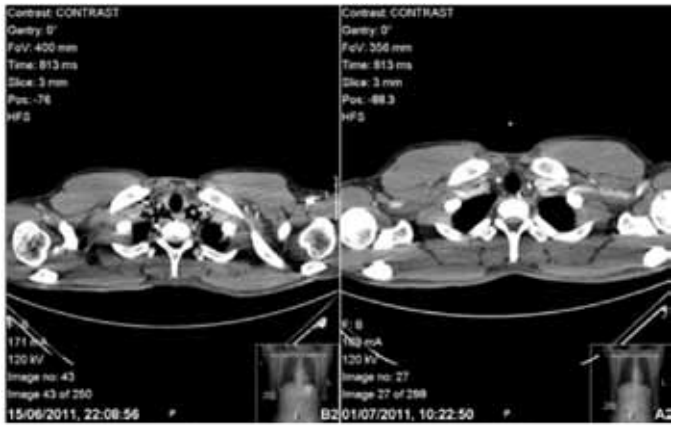


Figure 5: Computed tomography scan of chest with Gastrografin contrast; a comparison of image on admission image after resolution showing no surgical emphysema.

DISCUSSION

This case illustrates how treatment for Boerhaave's syndrome can be delayed and with such high mortality rates this can often be a fatal mistake. The mortality rate is significantly higher for those who are diagnosed after 24 hours of onset of symptoms [5, 7, 8] highlighting the importance of prompt diagnosis and the initiation of treatment of Boerhaave's syndrome. Unusually, this patient had a perforation in the upper third of the esophagus rather than the postero-lateral distal third. This may have contributed to their good outcome despite the diagnostic delay.

The classical presentation of thoracic pain, subcutaneous emphysema and vomiting described by Hermann Boerhaave may not be apparent [8]. Atypical presentations are common often resulting in the sufferer of Boerhaave's syndrome not seeking medical advice with misinterpreted clinical features resulting in misdiagnosis [2]. This case demonstrates the importance of the history and thorough examination in those with chest pain preceded by vomiting. Suspicions should be raised if the past medical history includes alcohol excess and esophageal abnormalities such as varices, esophagitis (reflux, eosinophilic and drug induced), strictures and infection. The differential of Boerhaave's syndrome should be considered no matter how clinically well the patient might be.

A chest X-ray is required when suspecting Boerhaave's syndrome, this may demonstrate a pneumothorax, subcutaneous emphysema or pleural effusion. Most spontaneous ruptures of the esophagus occur in the lower posterior lateral aspect as there is often a contact with the left pleural cavity. Chest drains can be diagnostic due to the presence of food particles, high amylase or gastric juice (pH <6). Contrast radiography can be performed but this has a false negative rate up to 25% [3]. A CT scan is valuable as it can aid in the exclusion of other diagnosis and has been suggested even in the absence of physical findings [5]. Endoscopy can be used to further delineate the extent of the rupture.

The management of Boerhaave's syndrome can be non-operative or operative. Generally, primary repair via a thoracotomy is performed to manage this condition with good results seen within 24 hours after presentation [3]. The best operative results are within 24 hours [7]. Thoracoscopic surgery can also be considered in those with stable vital signs. Whether the rupture is contained or not has been used as criteria for determining surgical and non-surgical management [5]. Sulpice et al. demonstrated equivalent results from conservative surgical management comparing primary suture and T-tube repair in a cohort of 39 patients. Reinforced sutures showed more favorable outcomes reducing postoperative leakage compared to non-reinforced primary sutures [9]. Glue and esophageal stents inserted endoscopically have been used with mixed results, with stent complications including aorto-esophageal fistula and leakage [4]. The optimum management for patients with Boerhaave's syndrome is undecided [7]. Schipper et al. suggested an algorithm following their literature review in which surgical (thoracotomy/laparotomy) and endoscopic intervention should be considered within 48 hours and conservative thereafter with a view to converting to surgery should sepsis develop [4]. Non-operative management can be successful as demonstrated in this case report. One can utilize broad-spectrum antibiotics, percutaneous drainage of pleural fluid/abscesses, enteral feeding and intravenous fluids [5]. There are criteria which have suggested when conservative treatment is appropriate. However, disadvantages of conservative treatment exist such as esophageal necrosis, infected paraesophageal tissues and continuous spoilage leading to deterioration of an initially clinically well patient [6].

There are no published recommendations for how long a patient who is treated non-operatively for Boerhaave's syndrome should remain hospitalized and monitored. Griffin et al. reported no deaths in 17 contained ruptures treated non-surgically, eight of which required drainage of

collections. Their hospital stay ranged between 6–113 days (median 14) [5]. In this case report, the initial treatment period of five days was not sufficient, even after a negative Gastrografin swallow, resulting in re-rupture. However, the second treatment period of 12 days of conservative management was more appropriate. Shaker et al. in 2010 allowed patients to eat and drink, if there was no or minimal radiological evidence of leakage. Their mean times from perforation to eating food were 20 and 48 days in their surgical and non-surgical groups, respectively [7].

Teh et al. found that time to oral intake were medians of 13 days (range 6–28) and 8.5 days (range 3–25), respectively in those treated with primary repair and conservatively [8]. Further published data will aid the decision on an acceptable treatment period for operative and non-operative management before commencement of oral diet. Furthermore, a guideline for diet following Boerhaave's syndrome should be agreed. In this case report, despite radiological evidence of healing, the commencement of even a liquid diet was too much for the minimally healed esophagus therefore leading to re-perforation. One should proceed with caution when considering oral intake even with radiological evidence of healing.

CONCLUSION

Boerhaave's syndrome is a fatal and serious condition which requires the utmost vigilance to diagnose and treat aggressively. An open mind should be kept with patients presenting with the specific and nonspecific symptoms of this condition. Referrals should be prompt and acted on quickly. This case highlights how easy this condition is to miss, the importance of allowing enough time to heal and the fact that nutrition plays an important role during admission and thereafter. Normal imaging may not necessary mean healing is complete. Further case series and randomized controls trials are needed to add to the current literature so that an agreed management and dietary algorithm can be decided upon. However, this will take time as Boerhaave's syndrome represents a moving target with the vast heterogeneity of presentations that have been described and acted on for this condition.

****

Acknowledgements
Many thanks to Sneha Varkey for the images provided in this case report.

Author Contributions
Eshan Oderuth – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Kevin Ilo – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Munir Somji – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Thomas Holme – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

REFERENCES

1. Boerhaave H. Atrocis, nec descripti prius, morbi historia secundum medicae artis leges conscripta, lugduni batavorum, boutesteniana. Medici 1724;60. Translated in: Bull Med Libr Assoc 1955;43:217–40.
2. Szeliga J, Jackowski M. Boerhaave Syndrome. Pol Przegl Chir 2011 Sep;83(9):523–6.
3. Prichard R, Butt J, Al-Sariff N, et al. Management of spontaneous rupture of the oesophagus (Boerhaave's syndrome): Single centre experience of 18 cases. Irish Journal of Medical Science 2006 Oct-Dec;175(4):66–70.
4. de Schipper JP, Pull ter Gunne AF, Oostvogel HJ, van Laarhoven CJ. Spontaneous rupture of the oesophagus: Boerhaave's syndrome in 2008. Literature review and treatment algorithm. Dig Surg 2009;26(1):1–6.
5. Griffin SM, Lamb PJ, Shenfine J, Richardson DL, Karat D, Hayes N. Spontaneous Rupture of the Oesophagus. Br J Surg 2008 Sep;95(9):1115–20.
6. Mahmodlou R, Abdirad I, Ghasemi-Rad M. Aggressive surgical treatment in late-diagnosed esophageal perforation: A report of 11 cases. ISRN Surg 2011;2011:868356.
7. Shaker H, Elsayed H, Whittle I, Hussein S, Shackcloth M. The influence of the 'golden 24-h rule' on the prognosis of oesophageal perforation in the modern era. Eur J Cardiothorac Surg 2010 Aug;38(2):216–2.
8. Teh E, Edwards J, Duffy J, Beggs D. Boerhaave's syndrome: A review of management and outcome. Interact Cardiovasc Thorac Surg 2007 Oct;6(5):640–3.
9. Sulpice L, Dileon S, Rayar M, et al. Conservative surgical management of Boerhaave's syndrome: experience of two tertiary referral centers. Int J Surg 2013;11(1):64–7.

ABOUT THE AUTHORS

Article citation: Oderuth E, Ilo K, Somji M, Holme T. Medical school finals, nerves and vomiting: medical student survives Boerhaave's syndrome with recurrence one week after initial presentation. International Journal of Case Reports and Images 2014;5(3):201–205.

Eshan Oderuth is working a Core Surgical Trainee. He has interests in both general surgery and orthopaedics and hope to choose one as a future career path. He studied at Imperial College London School of Medicine.

Kevin Ilo is looking to proceed with a career in orthopedics in the future. He has studied at Imperial College London School of Medicine.

Thomas Holme hopes to become an orthopedic surgeon once he finishes his surgical training. He has studied at Imperial College London School of Medicine.

Munir Somji is pursuing a career in radiology. He has studied at Kings College School of Medicine.

CASE REPORT OPEN ACCESS

Steatocystoma multiplex of face: A case report

Surej Kumar LK, Nikhil Mathew Kurien, Varun Menon P

ABSTRACT

Introduction: Steatocystoma, an uncommon cutaneous disorder, initially thought to be a sebaceous or retention cyst is recognized as a hamartomatous malformation of the pilosebaceous duct junction characterized by the development of numerous sebum containing dermal cysts with sebaceous glands in the cyst walls. Case Report: A 70-year-old male patient presented with asymptomatic dark patches, bilaterally on malar region. Surgical excision was done under local anesthesia. Histopathology conmfirmed steatocystoma. Conclusion: Steatocystoma is a cosmetic problem and a lifelong condition. Steatocystoma multiplex should be considered as a spectrum with different variations in anatomical distribution which can be treated successfully.

Keywords: Steatocystoma Multiplex, Face, Malar region, Yellow Cutaneous cyst, Hereditary

****

Surej Kumar LK[1], Nikhil Mathew Kurien[2], Varun Menon P[3]
Affiliations: [1]MDS, Professor & HOD, Department of Oral & Maxillofacial Surgery, PMS College of Dental Science & Research, Thiruvananthapuram, Kerala, India; [2]MDS, Reader, Department of Oral & Maxillofacial Surgery, PMS College of Dental Science & Research, Thiruvananthapuram, Kerala, India; [3]Junior Resident, Department of Oral & Maxillofacial Surgery, PMS College of Dental Science & Research, Thiruvananthapuram, Kerala, India.
Corresponding Author: Dr. Varun Menon P, Junior resident Department of Oral & Maxillofacial Surgery, PMS College of Dental Science & Research, Golden Hills, Vattappara, Venkode P O, Thiruvananthapuram-695028, Kerala, India; Ph: +91-8891787360; Fax: +91-472-258 7874; E-mail: varunmp@gmail.com

Received: 11 April 2013
Accepted: 16 May 2013
Published: 01 March 2014

How to cite this article

Kumar SLK, Kurien NM, Menon VP. Steatocystoma multiplex of face: A case report. International Journal of Case Reports and Images 2014;5(3):207–210.

****

doi:10.5348/ijcri-2014-03-474-CR-5

INTRODUCTION

Steatocystoma multiplex was first described by Jamieson in 1873 and the name was coined by Pringle in 1899 [1]. Brownstein described regarding steatocystoma simplex in 1982. Since then a variety of names such as steatocystomatosis, sebocystomatosis and epidermal polycystic disease have been given to steatocystoma multiplex [2].

Steatocystoma, an uncommon cutaneous disorder, initially thought to be a sebaceous or retention cyst is recognized as a hamartomatous malformation of the pilosebaceous duct junction characterized by the development of numerous sebum containing dermal cysts with sebaceous glands in the cyst walls [3]. It, generally, can be classified into steatocystoma simplex when it is a solitary lesion and steatocystoma multiplex when there is multiple, small, soft, movable, yellowish to skin colored dermal cystic papules.

CASE REPORT

A 70-year-old male was presented to our outpatient clinic with asymptomatic dark patches bilaterally on malar region (Figure 1). The lesion had slowly enlarged over two years and physical examination revealed yellow to skin colored, soft movable cystic masses, ranging from 10 mm to 20 mm in size with no punctum. There was no remarkable family history of similar lesions and

the patient had no other cutaneous diseases. The nails, teeth and hair were all normal. Routine laboratory investigations were within normal limits.

Surgical excision was done under local anesthesia (Figure 2). The lesion was sent for histopathological examination (Figure 3A–B). The specimen was stained in Hematoxylin and Eosin and was seen under 100x, the histopathology slide showed mild hyperkeratosis. Multiple cysts were lined by stratified squamous epithelium and granular layers were seen. The lumen of the cysts was filled with laminated keratin and a few of the cysts showed focal ulceration. Adjacent tissue exhibited skin appendages, dense acute and chronic inflammatory cells and numerous foreign body giant cells. A few melanophages were also seen in dermis. There was no evidence of granuloma or malignancy (Figure 4).

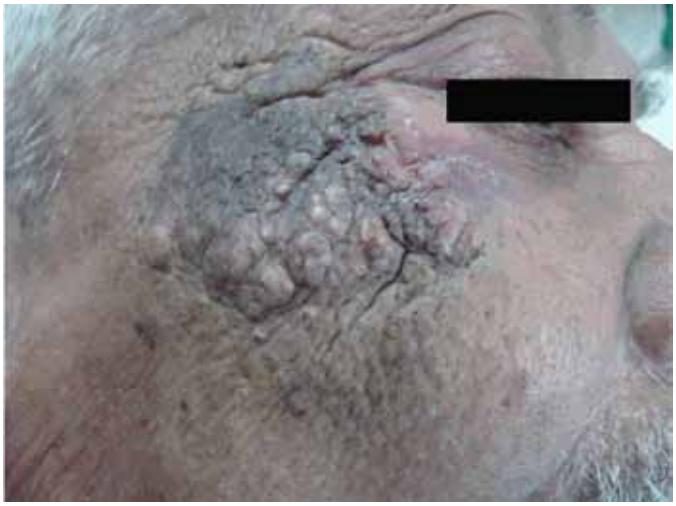


Figure 1: Dark patches seen unilaterally on malar region.

Figure 2: Post surgery of the patient.

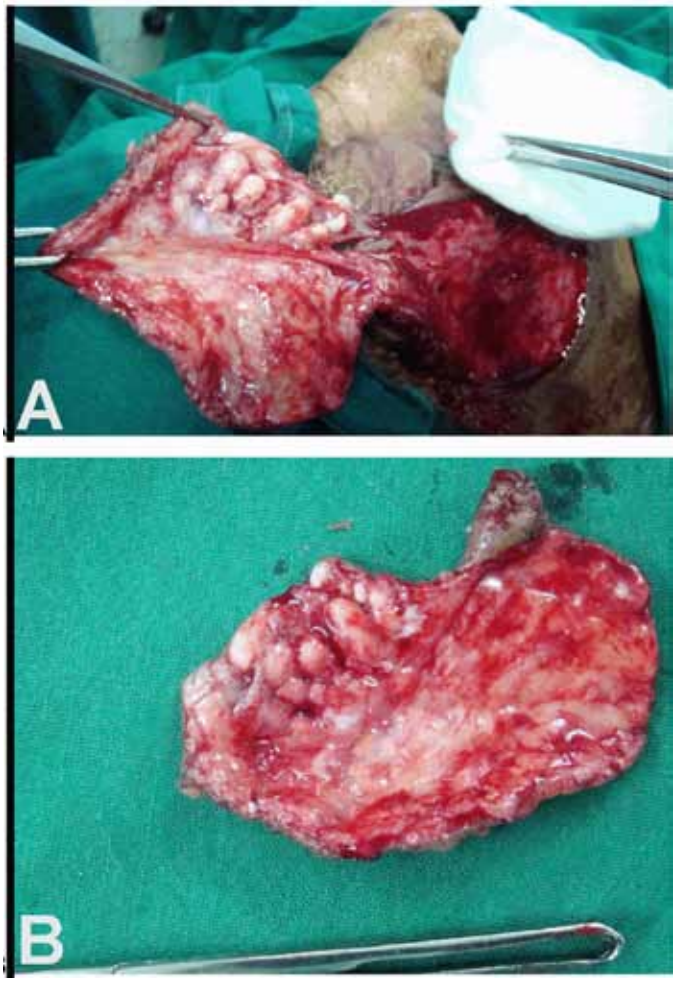


Figure 3: (A) Surgical excision of the lesion done, (B) Excised specimen.

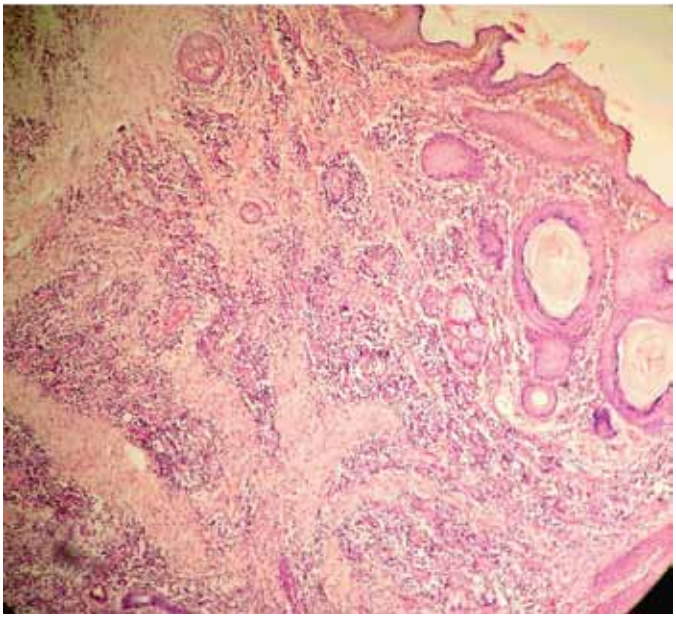


Figure 4: Histopathological examination showing stratified squamous surface epithelium exhibiting melanin pigmentation on the basal layer. Connective tissue shows multiple cysts filled with keratin along with few sebaceous gland and chronic inflammatory cell infiltrate, predominantly lymphocytes and few giant cells (H&E stain, x100).

DISCUSSION

Steatocystoma multiplex is an uncommon, inherited disorder that is characterized by multiple, asymptomatic, variably-sized dermal cysts. It is an uncommon disorder of the pilosebaceous unit characterized by the development of numerous sebum-containing dermal cysts [2] . Occurrence of large sized variant of these lesion clustered in large number in a localized region of body is rare. In our case steatocystoma multiplex was seen bilaterally on malar region, though it is the steatocystoma simplex variant that is more commonly seen on the face.

Steatocystoma simplex is the non-hereditary counterpart to the hereditary steatocystoma multiplex, clinically both are almost identical. Steatocystoma simplex is characterized by solitary, non-heritable growth mostly seen on face while multiplex is commonly seen on the axillae, groin, trunk and proximal extremities. It is rarely seen on face and scalp and is inherited, so is termed steatocystoma multiplex congenita. But some cases of steatocystoma multiplex seen on face and scalp can arise sporadically without any family history, as was the case seen in our patient [4]. It usually begins in late childhood and persists indefinitely, but in some patients the lesions have developed late in life [5]. Steatocystoma multiplex localized in the face and scalp has been divided into the following three types:

(i) Facial papular variant type
(ii) Sebocystomatosis and
(iii) Cysts located exclusively on scalp [5].

The etiological factors of steatocystoma multiplex are still unclear but trauma infection and immunological events have been proposed as some reasons [6]. Steatocystoma multiplex, familial cases have been linked to pachyonychia congenita and ectodermal dysplasia through a mutation in keratin 17.8. Pachyonychia congenita is a rare hereditary disorder characterized mainly by nail hypertrophy and dyskeratosis of skin and mucous membrane PC-2 (Jackson–Lawler form) and is accompanied by steatocystoma multiplex [7]. Steatocystoma is considered as forme fruste of pachyonychia congenita II.

Steatocystoma multiplex has also been associated with ichthyosis, koilonychia, acrokeratosis, Verruciformis of Hopf, hypertrophic lichen planus, rheumatoid arthritis, hypohidrosis, hypothyroidism and hypotrichosis. In our case, there were no such associated findings. Eruptive vellus hair cysts can mimic steatocystoma multiplex clinically and epidermal inclusion cysts have to be excluded. Comparison studies of keratin expression showed that epidermoid cyst expressed K10, EVHC expressed K17, trichilemmal cyst and steatocystoma multiplex expressed both K10 and K17 [8]. Kligman and Kirchbaum postulated that pluripotential ectodermal cells retain the embryonic capacity to form appendages or naevi rather than retention or inclusion cysts [9, 10]. Both steatocystoma multiplex and steatocystoma simplex arise from ducts of sebaceous glands.

Clinically, lesion is characterized by numerous small skin colored or yellowish cutaneous cysts. Colour of the lesion varies from yellowish to skin color depending upon the depth of the lesion, the superficial lesion being yellow and deeper lesions being skin colored [4].

It appears as multiple subcutaneous nodules, occurring anywhere on the body, even intraorally, but trunk and proximal extremities are the more common sites. The size of the nodules range from 0.2–2 cm and has no punctum. It remains asymptomatic but may get inflamed. When an inflamed cyst ruptures it produces steatocystoma multiplex suppurativa. The cyst may contain oily yellow fluid and vellus hairs. Steatoma is due to over production of sebum by one or more sebaceous glands while the outlet is closed. The sebum collected inside may undergo calcareous degeneration after remaining in situ for years destroying the epithelial lining.

The differential diagnosis should include other inherited syndromes of multiple benign adnexal tumors such as cylindromas, trichoepitheliomas, trichilemmomas who also have their counterpart in non-heritable solitary tumors.

The condition is histologically characterized by cystic structure with sebaceous glands within the cyst wall and epithelium that displays an eosinophilic cuticle. Inflammatory cells of macrophage monocyte lineage in connective tissue are also seen [6]. Keratin, oil and hairs in the lumen are also associated findings. Hyperkeratosis, multiple cyst lined by stratified squamous epithelium, lumen filled with lamellated keratin, foreign body giant cells in the lining cells, hair follicle, focal ulceration of cyst and more importantly a hyaline cuticle ,all these features correlated well with our histological study too.

Various treatment modalities have been described each with its own merits and demerits. Oral isotretinoin, a retinoid, tetracycline along with incision and drainage, intralesional steroids , extirpating the contents with aspiration without removing the cyst wall or excochleation of cyst wall with a curette are all methods often tried but with a high rate of recurrence.

Radiofrequency, cryosurgery, carbon dioxide laser, YAG laser are better options but residual scarring, blister formation, pain, postinflammatory hyperpigmentation are problems that have to be dealt with. Wide excision with its removal in toto is still the preferred choice because of the less postoperative complications and its cost effectiveness.

CONCLUSION

Steatocystoma is a cosmetic problem and a lifelong condition. There are no reports of malignant transformation from these benign adnexal tumors and we were able to confirm the unique nature of this lesion and rule out malignancy. Steatocystoma multiplex should be considered as a spectrum with different variations in

anatomical distribution which when properly diagnosed and timely intervened can be treated successfully.

****

Author Contributions

Surej Kumar LK – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Nikhil Mathew Kurien – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Varun Menon P – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Davey MA, Burkhart CN, Morrell DS, et al. Steatocystoma Multiplex. http://emedicine.medscape.com/article/1059725-overview 2012.
2. Belinchón I, Mayol MJ, Onrubia JA. Steatocystoma multiplex confined to the scalp. Int J Dermatol 1995;34(6):429–30.
3. Thomas VD, Swanson NA, Lee KK. Benign epithelial tumors, hamartomas, and hyperplasias. In: Wolff K, Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, editors. Fitzpatrick's dermatology in general medicine. 7th ed. New York: McGraw-Hill 2008. p. 1065–6.
4. Ebling FJG, Cunliffe WJ. Disorders of the sebaceous glands. In: Champion RH, Burton JL, Ebling FJG, editors. Textbook of dermatology. Oxford: Blackwell 1992. p. 9–744.
5. Kumakiri M, Yajima C. Eruptive steatocystoma multiplex on the scalp. J Dermatol 1991;18(9):537–9.
6. Setoyama M, Mizoguchi S, Usuki K, Kanzaki T. Steatocystoma multiplex: A case with unusual clinical and histological manifestation. Am J Dermatopathol 1997;19(1):89–2.
7. Smith FJ, Corden LD, Rugg EL, et al. Missense mutations in keratin 17 cause either pachyonychia congenita type 2 or a phenotype resembling steatocystoma multiplex. J Invest Dermatol 1997;108(2):220–3.
8. Tomková H, Fujimoto W, Arata J. Expression of keratins (K10 and K17) in steatocystoma multiplex, eruptive vellus hair cysts, and epidermoid and trichilemmal cysts. Am J Dermatopathol 1997;19(3):250–3.
9. Kligman AM, Kirschbaum JD. Steatocystoma multiplex: A dermoid tumor. J Invest Dermatol 1964;42:383–7.
10. Hansen KK, Troy JL, Fairley JA. Multiple papules of the scalp and forehead. Steatocystoma multiplex (facial papular variant). Arch Dermatol 1995;131(7):835, 838.

CASE REPORT OPEN ACCESS

Damage control orthopedics in a patient with polytrauma complicated with thyroid storm

Rishya Manikam, M Emmed, CS Kumar, Jeffry Amit, AB Sri Latha

ABSTRACT

Introduction: The concept of damage control orthopaedics (DCO) especially in a polytrauma patient compounded with other medical conditions being vigorously advocated by orthopaedic trauma surgeons. Case Report: Presenting a case of a polytrauma patient with thyroid storm. The decision on DCO in this patient being discussed. Conclusion: The decision about the timing of surgery should be based on the patients' condition as a whole. Careful planning and definition of priorities in each individual patient is essential. This holistic approach will see that patient safety care improves.

Keywords: Polytrauma, Thyroid Storm, Orthopaedics

****

Rishya Manikam[1], M Emmed[2], CS Kumar[3], Jeffry Amit[4], AB Sri Latha[5]

Affiliations: [1]MBBS, Head, Accident and Emergency Department, University Malaya, Kuala Lumpur, Malaysia; [2]Head, Accident and Emergency Department, University Malaya, Kuala Lumpur, Malaysia; [3]MBBS, M.S (Ortho), Senior Lecturer, Orthopedics Department, University Malaya Medical Centre, Kuala Lumpur; [4]MBBS, Senior Lecturer, Orthopedics Department, University Malaya Medical Centre Kuala Lumpur, Malaysia; [5]MBBS, Accident and Emergency Department, University Malaya Medical Centre, Kuala Lumpur, Malaysia.

Corresponding Author: Dr. Rishya Manikam, Head, Emergency and Trauma Academics, University Malaya, Lembah Pantai, 50603, Kuala Lumpur, Malaysia; Tel: +60379494198; Fax: +60379494198; E-mail: rishyaman@yahoo.com

Received: 07 November 2011
Accepted: 26 March 2012
Published: 01 March 2014

How to cite this article

Manikam R, Emmed M, Kumar CS, Amit J, Latha ABS. Damage control orthopedics in a patient with polytrauma complicated with thyroid storm. International Journal of Case Reports and Images 2014;5(3):211–214.

****

doi:10.5348/ijcri-2014-03-475-CR-6

INTRODUCTION

'Operate, operate and operate now' was the mantra of the orthopedic surgeons until the introduction of concepts of damage control orthopedics (DCO) in the early 1990s. Damage control orthopedics is characterized by primary, rapid, temporary fracture stabilization and followed by secondary definitive management, once the acute phase of systemic recovery has passed. In severely multiple injured patients who are in an unstable or in extremis clinical condition, DCO is the current treatment of choice. This is important to avoid 'second hit' or unnecessary additional trauma to the patient which can increase the risk of developing systemic complications and early mortality. With the concurrent development of the emergency medical care, it further enhances this concept where initial stabilization is done promptly. Decision about timing of the fixation requires balancing the risks of operative stress against benefits that can be gained by early fixation. We report a case of bilateral femur fracture associated with occipital skull fracture, subdural hemorrhage over right frontal and left frontotemporal complicated by thyroid storm on admission. Today's concepts and option of treatment of polytrauma patients are discussed in this report.

CASE REPORT

Mr. L (patient) was a 24-year-old Chinese male, who was involved in a motor vehicles accident, in which the motorbike was hit and run by a lorry. He was brought to the Trauma and Emergency department presented with headache, swelling and deformity over both the thighs.

On arrival, he was looking confused and restless with Glasgow coma scale was 14. The blood pressure was stable around 150/90 mmHg but he was tachycardic with pulse rate of 134 beats per minute. He had high body temperature of 39.3°C.

Physical examination revealed tenderness over the back of the skull, swollen and deformity over both the thighs. There was no neurovascular deficit over bilateral lower limbs. He had bibasal crepitation of the lungs. The abdomen and pelvis examinations were normal.

Computed tomography scan brain showed occipital skull fracture and hemorrhage over right frontal and left frontotemporal (Figure 1).

Further history taken from the family members found that patient had history of thyroid problem six months ago, which was defaulted treatment. The emergency physician diagnosed and started treating him as having thyroid storm as his calculated Burch–Wartofsky score was more than 45. The medical team, especially the endocrine unit was consulted for further management. He was successfully resuscitated by the emergency team and transferred to the ward for further management.

On the orthopedics side, he was put on bilateral high tibial pin skeletal traction for fractures shaft femur (Figure 2) while waiting for definitive management later once the general condition becomes more stable. Neurosurgery team was decided to continue conservative management for the head injury.

Definitive management for the bilateral femur racture still cannot be carried out in view of thyroid storm.

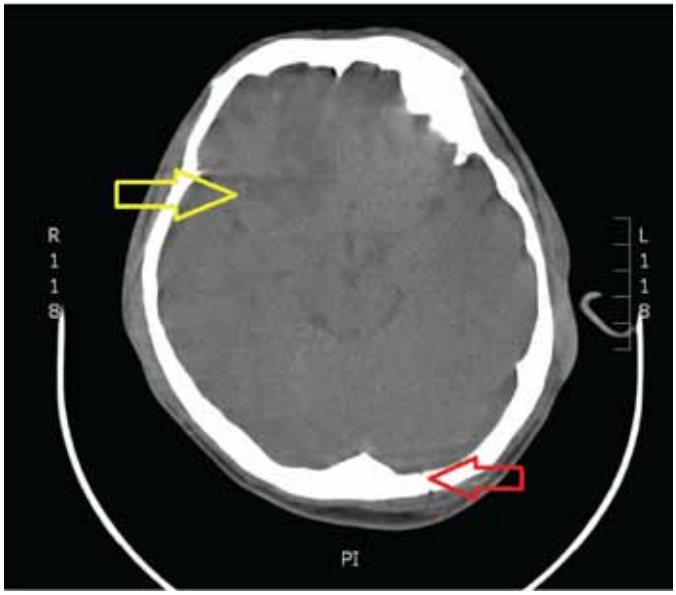


Figure 1: Occipital skull fracture (red arrow) and hemorrhagic contusion both frontal lobe with surrounding oedema (yellow arrow).

Patient was resistant for the thyroid storm treatment, where it took almost one month under medical treatment to recover. During this period, as he was immobile, he developed 1st degree sacral sore which was treated with daily dressing and two hourly turning. Otherwise, there was no other complication occur related to prolong immobilization. Latest radiographs of bilateral femur showed huge callus formation with malunion of left femur (Figure 3). Clinical evaluation showed shortening of the left lower limb around 5 cm. Clinically, fracture already united with minimal tenderness on movement.

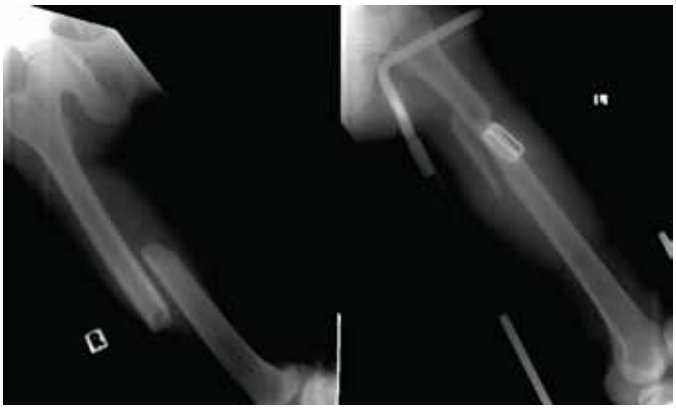


Figure 2: Bilateral midshaft femur fracture.

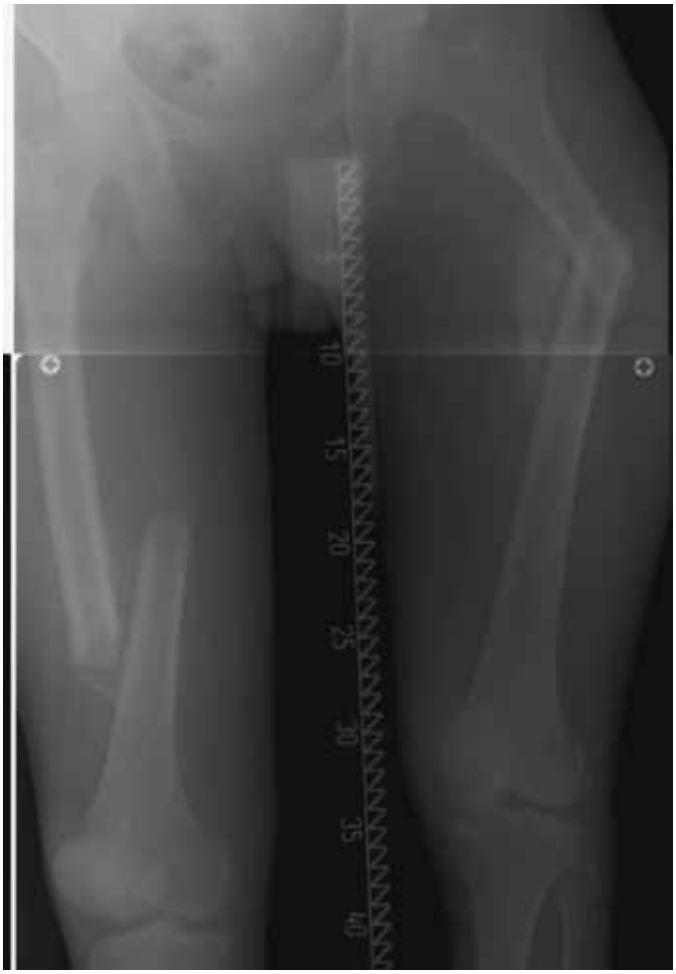


Figure 3: Malunion bilateral femur fracture.

Tibial pin was removed and wheelchair transfer was educated. He was planning for operation to correct the deformity of the left femur once medically fit for surgery.

DISCUSSION

Damage control orthopaedics has evolved as a new approach to minimize the impact of primary surgery. DCO should be regarded as a part of the resuscitation process. By careful choice of surgical technique, blood loss and tissue trauma are minimized and tissue oxygenation maintained. Thereby, the impact of operative fracture stabilization is minimized. In this case, we only choose simple procedure to put bilateral high tibial pin skeletal traction, temporarily to stabilize the long bone fracture.

In polytrauma patients, one of the major aims is rapid stabilization of their extremity injuries. According to Seibel et al, they found an association between respiratory complications, including ARDS and the number of days the patient spent on skeletal traction [1]. Another study done by Bone et al found a high rate of pulmonary complications in patients who had delayed stabilization of fractures [2]. Lozman et al. demonstrated better cardiac function in patients undergoing immediate fracture fixation [3] and Pape et al. found that early intramedullary nail was associated with a significantly lower ventilation time and total intensive care stay, as compared to patients undergoing secondary stabilization [4].

However, recent reports suggest that not all the polytrauma patients will get beneficial from the early total care management. Patients with severe head injury may get poorer outcomes if they went for early surgical intervention [5]. According to Frank et al., primary procedures of greater than 6 hours duration and major surgical procedures at days 2 to 4 should be avoided [6].

Unfortunately, in this case the definitive surgical treatment of the bilateral femur fracture cannot be carried out in view of thyroid storm and was delayed up to 1 month post trauma. As a consequence, fracture femur had united in malunion position. The complication that arises from late definitive surgical treatment however can be corrected later using advance surgical technique once patient fit for surgery, as we know thyroid storm is a life-threatening condition.

CONCLUSION

The decision about the timing of surgery should be based on the patients' condition as a whole. Careful planning and definition of priorities in each individual patient is essential. This holistic approach will see that patient safety care improves.

****

Author Contributions

Rishya Manikam – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

M Emmed – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

CS Kumar – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Jeffry Amit – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

AB Sri Latha – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Seibel R, LaDuca J, Hassett JM, et al. Blunt multiple trauma (ISS 36) femur traction, and the pulmonary failure septic state. Ann Surg 1985;202(3):283–95.
2. Bone LB, Johnson KD, Weigelt J, Scheinberg R. Early versus delayed stabilization of femoral fractures. A prospective randomized study. J Bone Joint Surg Am 1989;71(3):336–40.
3. Lozman J, Deno DC, Feustel PJ, et al. Pulmonary and cardiovascular consequences of immediate fixation or conservative management of long bone fractures. Arch Surg 1986;121(9):992–9.
4. Pape HC, Hildebrand F, Pertschy S, et al. Changes in the management of femoral shaft fractures in polytrauma patients: from early total care to damage control orthopedic surgery. J Trauma 2002;53(3):452–61.
5. Giannoudis PV, Smith RM, Bellamy MC, Morrison JF, Dickson RA, Guillou PJ. Stimulation of the inflammatory system by reamed and undreamed

nailing of femoral fractures – an analysis of the second hit. J Bone Joint Surg Br 1999;81(2):356–1.
6. Hildebrand F, Giannoudis P, Kretteck C, Pape HC. Damage control: extremities. Injury 2004;35(7):678–9.

CASE REPORT OPEN ACCESS

Pentalogy of Cantrell diagnosed in the first trimester of pregnancy

Yasemin Cekmez, Tülay Tos, Zehra Yilmaz, Nilay Pişkinpaşa, Tuncay Küçüközkan

ABSTRACT

Introduction: Pentalogy of Cantrell is a rare syndrome of unknown etiology characterized by multiple structural congenital anomalies. Case Report: We aimed to present an early diagnosed case of pentalogy of Cantrell accompanied by craniorachischisis. Conclusion: Pentalogy of Cantrell can be diagnosed with a careful ultrasonography in the first trimester. Potential side effects of pregnancy termination to the mother can be reduced by the early diagnose of the disease.

Keywords: Pentalogy of Cantrell, Fetus, Ectopia cordis, Craniorachischisis

How to cite this article

Cekmez Y, Tos T, Yilmaz Z, Pişkinpaşa N, Küçüközkan T. Pentalogy of Cantrell diagnosed in the first trimester of pregnancy. International Journal of Case Reports and Images 2014;5(3):215–217.

doi:10.5348/ijcri-2014-03-476-CR-7

Yasemin Cekmez[1], Tülay Tos[1], Zehra Yilmaz[1], Nilay Pişkinpaşa[1], Tuncay Küçüközkan[2]
Affiliations: [1]MD, Department of, Obstrectic and Gynecology, Sami Ulus Medical and Research Hospital, Ankara, Turkey; [2]Professor, Department of, Obstrectic and Gynecology, Sami Ulus Medical and Research Hospital, Ankara,Turkey.
Corresponding Author: Yasemin Cekmez, MD Sami Ulus Medical and Research Hospital 34090 Ankara - Turkey; Tel: 05053339047; Email: yaseminkandicekmez@hotmail.com

Received: 24 June 2013
Accepted: 30 July 2013
Published: 01 March 2014

INTRODUCTION

Pentalogy of Cantrell is a rare syndrome defined by Cantrell et al. in 1958. The syndrome is characterized by the front upper abdominal midline and sternum defects, the lack of diaphragmatic face of the pericardium and the front face of diaphragm, and a variety of cardiac anomalies with unknown etiology [1]. The incidence has been reported to be 1 per 5.5 million live births [2]. Prognosis is associated with the defects defined and severity of cardiac anomalies. We aimed to present a case of pentalogy of Cantrell accompanied by craniorachischisis.

CASE REPORT

A 26-year-old (G4P1) woman was admitted to our hospital at her 12th week of gestation. In the routine ultrasound examination for first-trimester screening, *crown rump length* (CRL) was 40 mm in diameter, compatible with the single live fetus of 10 weeks 6 days. The fetal calvarial bone was deformed (exencephaly) (Figure 1a1). The view of herniated organs out of the abdominal wall defect and the defects in the lower end of the sternum was compatible with omphalocele and ectopia cordis (Figure 1b1). Fetal spine was deformed and short. With these available ultrasonographic findings the diagnosis of Pentalogy of Cantrell was established and the pregnancy was terminated with the consent of mother and father. By the postmortem examination fetal omphalocele (Figure 1b2), ectopia cordis and diaphragmatic hernia was detected. In addition to these findings, we observed an exencephaly continuing with spinal dysraphism as additional anomaly (craniorachischisis) (Figure 1a2). Fetal spine was deformed and short.

DISCUSSION

Pentalogy of Cantrell is a rare syndrome that causes defects involving diaphragm, abdominal wall, pericardium, heart and lower sternum. In a study

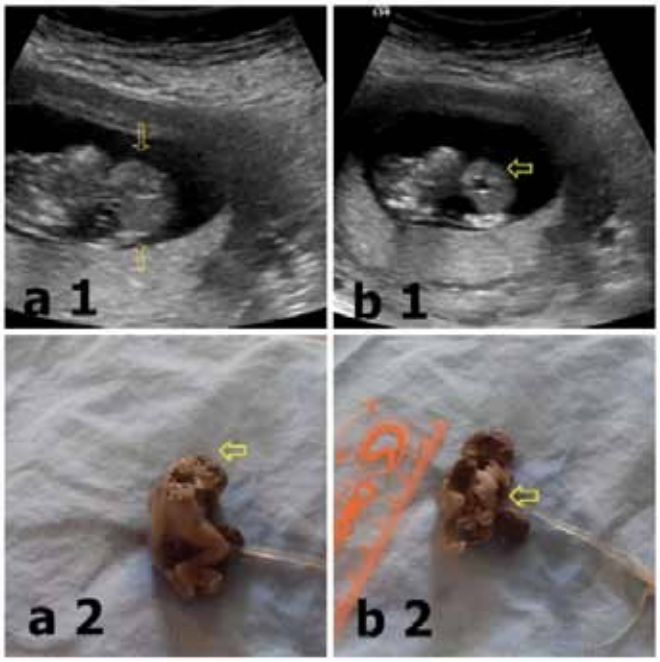


Figure 1: (a1) ultrasonografic view of exencephaly, (a2) exencephaly, (b1) ultrasonografic view of omphalocel and ectopia cordis and (b2) omphalocel.

consisting of 61 patients with Cantrell pentalogy, Toyama has proposed a classification for this syndrome [3]. According to this classification, patients including all five defects (definite diagnosis) were classified as type 1, patients including 4 defects 2 of which were intra-cardiac and ventral wall abnormalities (moderate diagnosis) were classified as type 2, patients including different combinations of incomplete abnormalities (always accompanied with sternal defects) were classified as type 3. Although the early gestational week of our patient prevented the detailed evaluation of intra-cardiac defects, the presence of ectopia cordis and omphalocele provided us to diagnose the Cantrell pentalogy in the first trimester.

The pathogenesis of Cantrell pentalogy is uncertain. The syndrome is thought to arise from an embryological development defect occuring in a segment of lateral mesoderm, 14–18 days after the conception [1]. As the transverse septum of the diaphragm cannot develop, a problem occurs in the ventro-medial migration of upper abdomens mesodermal fold. For this reason, the defects detected in the midline, sternum and diaphragm, consists of the lack of mesoderm migration. Although the pentalogy of Cantrell appears sporadicly, rarely it may be in association with chromosomal abnormalities (trisomy 13, trisomy 18 and triploidy) [4]. In addition, as it was seen in two boys from the same family in literature, a familial component of the syndrome can be considered [5].

In the differential diagnosis of Cantrell pentalogy, thoraco-abdominal ectopia cordis, amniotic band syndrome, limb body wall complex (LBWC), and body stalk anomaly should be considered [6, 7]. Craniofacial and central nervous system defects (cleft lip and palate, encephalocele, hydrocephalus, and craniorachischisis), limb defects (clubfoot, absence of the radius and tibia or hypodactyly), abdominal organ defects (gallbladder agenesis and polysplenia) may accompany to Cantrell pentalogy. Craniorachischisis is a related neural tube defect, in which anencephaly is accompanied by a contiguous open defect of the spine (spina bifida totalis). It is an extreme example of defective closure of the neural tube during early embryogenesis, around 20 to 22 days gestation [8]. In our case, the craniorachischisis was detected as an additional anomaly.

The prognosis of Cantrell pentalogy is related to the severity of defects identified. Ectopia cordis is a standalone component of the penthalogy that has a change of living although the poor prognosis of the disease [9]. As other symptoms that accompany to Cantrell pentalogy is closely associated with the prognosis they should be screened carefully during the ultrasound scan. The disease was detected at 10 weeks 6 days according to crown-rump length in our patient and the pregnancy was terminated.

CONCLUSION

In conclusion detection of this fetal disease is possible with a careful ultrasonographic examination in the first trimester and so the mortality and morbidity to the mother with late termination can be reduced.

****

Author Contributions

Yasemin Cekmez – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final of the version to be published

Tülay Tos – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final of the version to be published.

Zehra Yilmaz – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final of the version to be published

Nilay Pişkinpaşa – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final of the version to be published

Tuncay Küçüközkan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright

REFERENCES

1. Cantrell JR, Haller JA, Ravitch MM. A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. Surg Gynecol Obstet 1958;107(5):602–14.
2. Carmi R, Boughman JA. Pentalogy of Cantrell and associated midline anomalies: a possible ventral midline developmental field. Am J Med Genet 1992;42(1):90–5.
3. Toyama WM. Combined congenital defects of the anterior abdominal wall, sternum, diaphragm, pericardium, and heart: a case report and review of the syndrome. Pediatrics 1972;50(5):778–92.
4. Fox JE, Gloster ES, Mirchandani R. Trisomy 18 with Cantrell pentalogy in a stillborn infant. Am J Med Genet 1988;31(2):391–4.
5. Martin RA, Cunniff C, Erickson L, Jones KL. Pentalogy of Cantrell and ectopia cordis, a familial developmental field complex. Am J Med Genet 1992;42(6):839–41.
6. Romero, R, et al. Body stalk anomaly, In: Prenatal diagnosis of congenital anomalies, Romero, R, et al. (Eds), Appleton & Lange, Norwalk 1998. p. 226.
7. ten Donkelaar HJ, Hamel BC, Hartman E, van Lier JA, Wesseling P. Intestinal mucosa on top of a rudimentary occipital meningocele in amniotic rupture sequence: disorganization-like syndrome, homeotic transformation, abnormal surface encounter or endoectodermal adhesion? Clin Dysmorphol 2002;11(1):9–13.
8. Naveen N, Murlimanju, Vishal K, Maligi A. Craniorachischisis totalis. J Neurosci Rural Pract 2010;1(1):54–5.
9. Yavuz SIMSEK, Oktay KAYMAK, Sinasi YILMAZ. Diagnosis of a Rare Congenital Anomaly, Isolated Ectopia Cordis: A Case Report. Turkiye Klinikleri J Gynecol Obst 2008;18(4):277–9.

CASE REPORT OPEN ACCESS

Neurilemmoma of the tongue: A case report

Pallav Kumar Kinra, Jayakumar K, Manoj Joseph Michael

ABSTRACT

Introduction: Neurilemmomas, also referred to as schwannomas or neurinomas, are benign encapsulated nerve sheath tumors composed of Schwann cells. Neurilemmomas arise when proliferating schwann cells form a tumor mass of unknown etiology encompassing motor and sensory peripheral nerves. We report a rare case of neurilemmoma over the ventral surface of the tongue. Case Report: A 15-year-old child came to our Department of oral and maxillofacial surgery, Govt. Dental College, Kozhikode complains of swelling over ventral surface of tongue since two years. The lesion was freely moveable and asymptomatic. Clinically, the lesion appeared to be a benign soft tissue neoplasm. Excisional biopsy of the lesion was done under local anesthesia and tissue was sent for histopathological examination. Diagnosis is made by histological examination of the lesion. The treatment is surgical excision of the lesion. They do not show recurrence after complete excision. Conclusion: As neurilemmoma is a benign neoplasm, surgical excision is the treatment of choice. The prognosis of neurilemmoma is usually excellent after adequate excision. This case showed no recurrence after the treatment and malignant transformation of a previously benign neurilemmoma is exceedingly rare.

Keywords: Neurilemmoma, Tongue, Oral cavity, Schwannoma

How to cite this article

Kinra PK, Jayakumar K, Michael MJ. Neurilemmoma of the tongue: A case report. International Journal of Case Reports and Images 2014;5(3):218–221.

doi:10.5348/ijcri-2014-03-477-CR-8

Pallav Kumar Kinra[1], Jayakumar K[2], Manoj Joseph Michael[2]
Affiliations: [1]Junior Resident, Department of Oral and Maxillofacial Surgery,Govt. Dental College,Kozhikode, Kerala, India; [2]Associate Professor, Department of Oral and Maxillofacial Surgery, Govt. Dental College, Kozhikode, Kerala, India.
Corresponding Author: Dr. Pallav Kumar Kinra, Junior Resident, Department of Oral and Maxillofacial surgery, Govt. Dental College Kozhikode, Kerala, India- 673008; Ph: 918943560527; Email: pallav_kinra163@yahoo.com

Received: 16 July 2013
Accepted: 19 August 2013
Published: 01 March 2014

INTRODUCTION

Neurilemmomas are benign neoplasms, which are commonly well encapsulated. It usually arises from spinal, cervical, sympathetic, vagus, peroneal or ulnar nerves. Extracranially, 25% of all schwannomas are located in head and neck, but only 1% show an intraoral origin [1]. Intraorally, it shows predilection for tongue followed by palate, buccal mucosa, lip and gingiva [2]. The tip of tongue is the least affected part. They remain asymptomatic unless they attain an appreciable size. Schwannomas may occur at any age but peak incidence is between third and sixth decades. There is no gender predilection. Only 50% of these tumors have direct relation with a nerve [3]. Schwannomas are usually solitary lesions but in unusual instances can occur as multiple, with von Recklinghausen's neurofibromatosis. They show no recurrence if completely excised and almost never undergo malignant transformation. We report a rare case of neurilemmoma on ventral surface of tongue [4].

CASE REPORT

A 15-year-old child came to our Department of oral and maxillofacial surgery, Govt. Dental College, Kozhikode complains of swelling over ventral surface of tongue since two years (Figure 1). Patient had no history of any change in size the of lesion. Patient had no difficulty in chewing, swallowing and phonation and there was no sensory or taste abnormalities by the patient. Color of overlying mucosa was normal. Patient was only concerned about the swelling which was totally asymptomatic. The past medical history of the patient was unremarkable. On clinical examination a 1.5-cm diameter mass was present on ventral surface of tongue just beneath the mucosa. The lesion had rubbery consistency, was freely moveable, non tender and asymptomatic. Rest of the oral cavity was normal. Clinically, the lesion appeared to be a benign soft tissue neoplasm.

Surgical excision of the lesion was done under local anesthesia with adequate surgical margins (Figure 2) after giving bilateral nerve block. During excision lesion was well circumscribed and encapsulated with no infiltration in the surrounding tissues. After excision tissue was sent for histopathological examination. Grossly the mass was well encapsulated, measuring 1.5 cm in diameter and had firm gray white cut surface with small cystic areas. The patient has not shown any recurrence in follow-up period of two years.

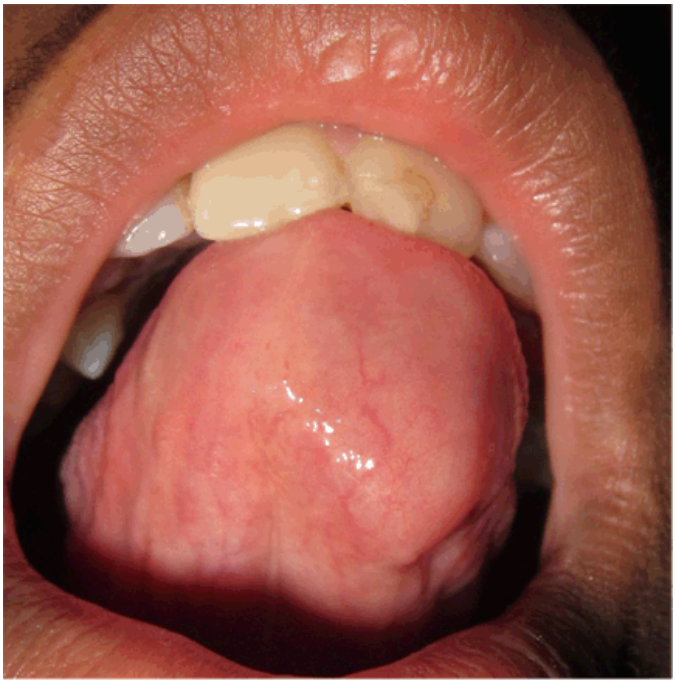


Figure 1: Preoperative view of tongue lesion.

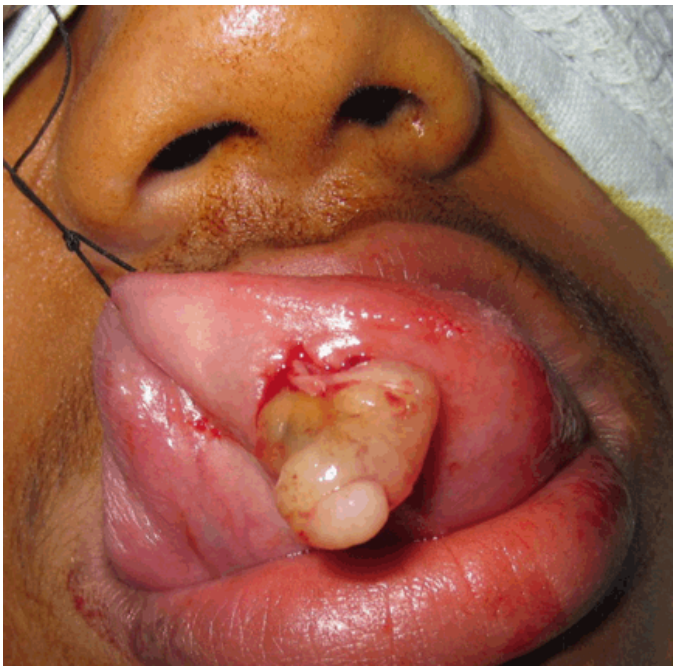


Figure 2: Intraoperative view of the lesion being removed.

DISCUSSION

Schwannomas are benign nerve sheath neoplasms composed of schwann cells. They were first described by Verocay in 1908. Embryologically, schwann cells arise during fourth week of development from a specialized population of ectomesenchymal cells of neural crest [2]. These cells form a thin barrier around each extracranial nerve fibres of motor and sensory nerves with myelin sheath to enhance nerve conductance. Schwannomas being rare in oral cavity, and is not often encountered in clinical practice. Diagnosis is confirmed by microscopic examination (Figure 3A). Two microscopic patterns of schwannomas are known to coexist: Antoni A and Antoni B. In Antoni A, elongated cells with cytoplasmic processes are arranged in fascicles in areas of moderate to high cellularity with little stromal matrix [5]. Spindle shaped nuclei are aligned in parallel rows forming a typical palisaded pattern. Between the rows of cells there are fine cytoplasmic fibrils with acellular, eosinophilic masses called Verocay bodies [3]. In Antoni B, the tumor is less densely cellular with a loose meshwork of cells along with microcysts and myxoid changes similar to neurofibroma [6]. In both patterns, the cytology of the individual cells is similar, with elongated cytoplasm and regular, oval nuclei. S-100 protein was strongly reactive in the spindle cells (Figure 3B). Degenerative changes in schwannomas can occur and include nuclear pleomorphism, xanthomatous change and vascular hyalinization [6]. Robert et al. have documented a schwannoma in a 30-year-old woman arising in the tip of tongue [2].

Lopez and Ballestin in their study of nine intraoral schwannomas found three schwannomas in vestibule, two each in tongue and palate and one each in floor of mouth and lower lip [5]. The main differential diagnosis clinically are other benign lesions which may occur at this site. These include neurofibroma, traumatic neuroma,

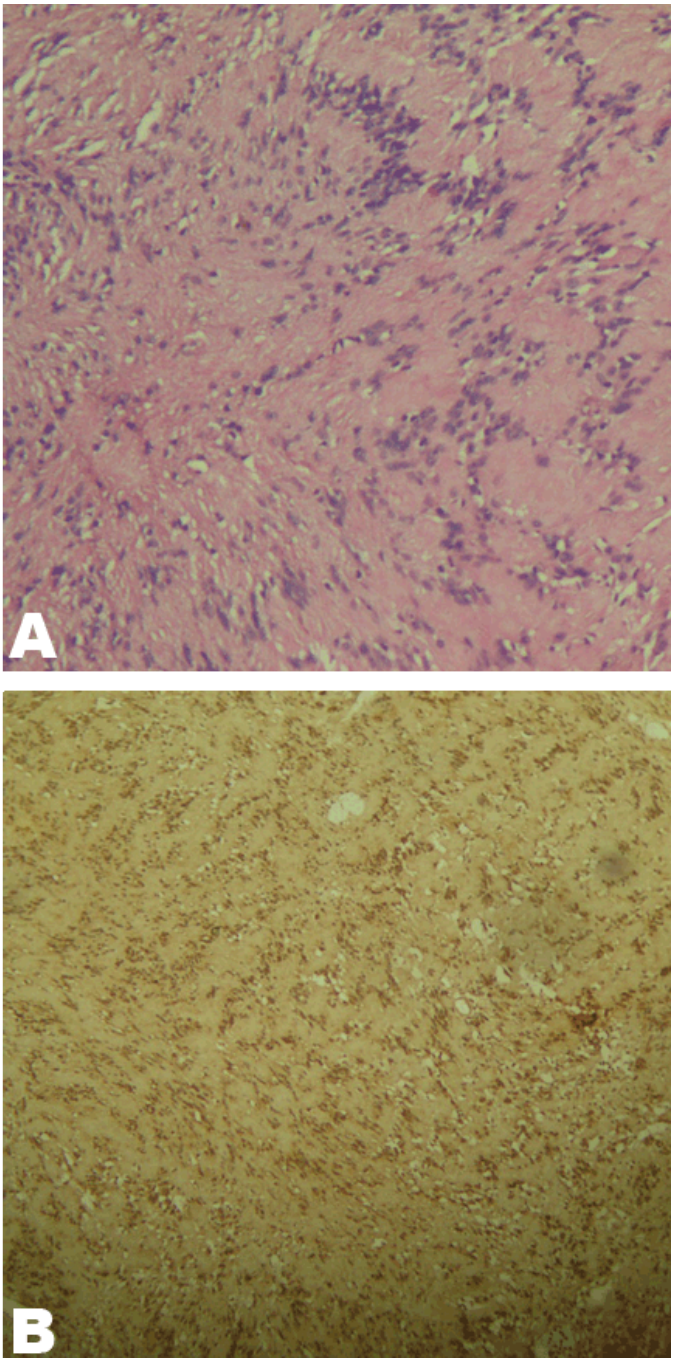


Figure 3: (A) Antoni A pattern with verocay bodies (B): Tumor cells showing S-100 reactivity.

fibroma, lipoma, leiomyoma etc. Diagnosis of the lesion can only be confirmed by histopathological examination and immunohistochemistry. Malignant transformation of schwannoma is exceptionally a rare event. Treatment of the lesion is complete surgical excision.

CONCLUSION

As neurilemmoma is a benign neoplasm, surgical excision is the treatment of choice. The prognosis of neurilemmoma is usually excellent after adequate excision. Diagnosis is only confirmed by histopathological examination. This case showed no recurrence after complete surgical excision and malignant transformation of a previously benign neurilemmoma is exceedingly rare.

****

Author Contributions

Pallav Kumar Kinra – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Jayakumar K – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Manoj Joseph Michael – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

ACKNOWLEDGEMENTS

Dr Soumithran CS, Dr Ayyappan, Dr Akhilesh, Dr Tushar, Dr Ambadas

REFERENCES

1. Gallesio C, Berrone S. Schwannoma located in the tongue. A clinical case report. Minerva Stomatol 1992;41(12):583–90.
2. Pfeifle R, Baur DA, Paulino A, Helman J. Schwannoma of the tongue: Report of 2 cases. J Oral Maxillofac Surg 2001;59(7):802–4.
3. Chiapasco M, Ronchi P, Scola G. Neurilemmoma (schwannoma) of the oral cavity. A report of 2 clinical cases. Minerva Stomatol 1993;42(4):173–8.
4. López JI, Ballestin C. Intraoral schwannoma. A clinicopathologic and immunohistochemical study of nine cases. Arch Anat Cytol Pathol 1993;41(1):18–23.

5. Nakayama H, Gobara R, Shimamoto F, Kajihara H. Ancient schwannoma of the oral floor and ventral portion of the tongue: A case report and review of the literature. Jpn J Clin Oncol 1996;26(3):185–8.
6. Cotran R, Robbins SL. Pathologic Basis of Disease (ed 6). Phila-delphia, PA, Saunders 1999:1352–4.

CASE REPORT OPEN ACCESS

Prolonged aphasia and perfusion computed tomography abnormalities in migraine with aura

Anselm Angermaier, Soenke Langner, Michael Kirsch, Alexander V Khaw

ABSTRACT

Introduction: Migraine with aura is defined as a recurrent disorder manifesting in attacks of reversible focal neurological symptoms that usually develop gradually, last for less than 60 minutes and is followed by characteristic headache and vegetative symptoms. Acute aphasia is a well-known aura symptom. We present a case of an acute focal neurologic deficit in which perfusion imaging proved helpful in rapid decision making for the appropriate treatment by identifying the syndrome as a stroke mimic. Case Report: A 24-year-old male student was admitted with global aphasia and headache precluding any interview for the patient's medical history. Initial perfusion computed tomography scan showed hypoperfusion in the entire left hemisphere, pronounced in the left occipitotemporal lobe and Broca's area. This pattern which was not restricted to a vascular territory and hypoperfusion above critical ischemia guided us in classifying the deficit as a stroke mimic, specifically as migraine aura. Conclusion: In the hyperacute phase of stroke-like symptoms, multimodal computed tomography scan can add valuable information for differentiating ischemic stroke from stroke mimics and support treatment decision-making.

Keywords: Migraine with aura, Aphasia, Perfusion computed tomography (PCT) scan, Cortical spreading depression

How to cite this article

Angermaier A, Langner S, Kirsch M, Khaw AV. Prolonged aphasia and perfusion computed tomography abnormalities in migraine with aura. International Journal of Case Reports and Images 2014;5(3):222–225.

doi:10.5348/ijcri-2014-03-478-CR-9

Anselm Angermaier[1], Soenke Langner[2], Michael Kirsch[3], Alexander V Khaw[4]

Affiliations: [1]MD, Resident, Department of Neurology, University Medicine Greifswald, DE–17475 Greifswald (Germany); [2]MD, PhD, Consultant, Institute for Diagnostic Radiology and Neuroradiology, University Medicine Greifswald, DE–17475 Greifswald (Germany); [3]MD, Consultant, Institute for Diagnostic Radiology and Neuroradiology, University Medicine Greifswald, DE–17475 Greifswald (Germany); [4]MD, Consultant, Department of Neurology, University Medicine Greifswald, DE–17475 Greifswald (Germany)

Corresponding Author: Anselm Angermaier, Department of Neurology, University Medicine Greifswald, Ferdinand-Sauerbruch Str., DE–17475 Greifswald (Germany); Tel: +49 3834 86 6815; Fax +49 3834 86 6875; Email ID: anselm.angermaier@uni-greifswald.de

Received: 07 August 2013
Accepted: 04 September 2013
Published: 01 March 2014

INTRODUCTION

Aphasia is a well-known symptom in migraine with aura, but a major stroke symptom as well [1]. Differentiating between both in the acute setting has tremendous effects on treatment and prognosis. We present a case of a migraine attack with prolonged aphasic aura in which perfusion computed tomography (PCT) scan guided us in classifying the deficit as a stroke mimic.

CASE SERIES

A 24-year-old, right-handed, male student who was living alone, was referred to our emergency ward at 00:30 because of acute speech problems and headache. Exact time of onset could not be determined. The emergency physician reported that the patient called a friend at about midnight indicating speech difficulties. His friend

then called an ambulance. He was able to open the door to EMS and walk. Clinical examination demonstrated severe aphasia. In the hospital the patient was unable to speak, read or write, could follow simple commands, was agitated and indicated by gesture that he was having severe headache, photophobia and phonophobia. Suddenly, he started vomiting. No other focal neurological deficits were found. Differential diagnoses included intracranial hemorrhage or ischemic infarction, cerebral venous thrombosis and migraine with aura. Multimodal computed tomography scan was undertaken according to our hyperacute stroke imaging algorithm. Non-contrast computed tomography showed no early signs of cerebral ischemia and ruled out intracranial hemorrhage. Computed tomography angiography (CTA) demonstrated no arterial occlusion or cerebral sinus thrombosis. However, perfusion-CT showed a modest hypoperfusion of the left hemisphere with an area of more severe hypoperfusion in the left Broca and occipitotemporal region which was not restricted to one vascular territory (increase of time to peak (TTP) up to 21%; decrease of cerebral blood flow (CBF) and cerebral blood volume (CBV) up to 38% and 30%, respectively in comparison to the corresponding contralateral area; Figure 1). Therefore, ischemic infarction was deemed unlikely and consideration of thrombolysis was aborted. At this time the patient's mother contacted the hospital by phone reporting that her son had called her at 21:30 complaining about blurred vision since 19:00 and a bilateral scotoma wandering from the outside to inside of the visual field for five minutes and paresthesia in his right arm and around the right angle of his mouth. Then a severe pulsating bifrontal headache occurred. At 22:00 during a second telephone call his mother noted he developed progressive problems in finding words. Suddenly, he started vomiting. His mother, father and sister were known to have migraine. He had not been diagnosed as having migraine, but during childhood and adolescence he had about six episodes with visual disorders or numbness in his right arm followed by headaches. However, there had never been vomiting, photophobia or phonophobia.

Within 24 hours after analgesics and antiemetic medication aphasia and headache resolved completely. A follow-up magnetic resonance imaging (MRI) scan after four days showed no signs for infarction or perfusion abnormalities.

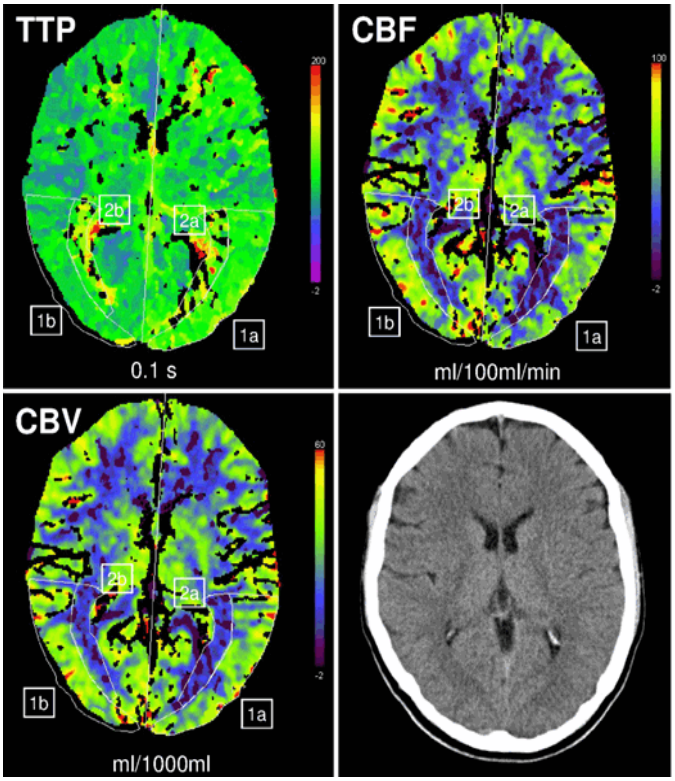


Figure 1: Perfusion computed tomography shows hypoperfusion of the entire left hemisphere, predominantly in the left Broca's and occipitotemporal region (time to peak + cerebral blood flow), while cerebral blood volume is above ischemic thresholds (over 60% of average cerebral blood volume in corresponding contralateral unaffected area). Left regions of interest are used for quantification, mirrored regions of interest on the right side. Abbreviations: TTP: time to peak; CBF: cerebral blood flow; CBV: cerebral blood volume; 1: cortical Region of Interest; 2: subcortical Region of Interest, a: hypoperfused tissue; b: normal tissue. Right bottom: non-contrast computed tomography at the level of perfusion CT

DISCUSSION

Our patient presented with evolving neurologic symptoms of sudden visual and sensory disturbances followed by headache, vomiting and global aphasia which lasted for less than 24 hours. The diagnosis of migraine with prolonged aura was made [2]. A transient ischemic attack was considered unlikely because of the following reasons:

- the evolution of the clinical symptoms
- history of episodes with focal neurological deficits associated with headaches
- positive family history for migraine
- absence of vascular risk factors and
- hypoperfusion in left hemisphere that was not restricted to any one or two vascular territories

However, in the hyperacute phase of admission the patient's severe aphasia precluded communication of the first four of the above-mentioned five criteria.

Perfusion changes during a migraine attack with aura are well known from single-photon emission computed tomography (SPECT) and positron emission tomography (PET) studies [3], but there is also rising knowledge from acute imaging with computed tomography (CT) scan or MRI scan. Nieuwkamp et al. presented a first ever migraine where initial PCT showed modest hypoperfusion of the left hemisphere which the authors attributed to migraine aura [4]. Recently, Floery

et al. showed that patients with migraine aura had hypoperfusion in more than one vascular territory but no diffusion-weighted Imaging (DWI) abnormality [5]. ^{133}Xe-SPECT studies demonstrated 'spreading oligemia' from occipital to frontal which is likely to be a secondary phenomenon to cortical Spreading depression (CSD) [6]. The patient´s aphasic aura is consistent with nonischemic hypoperfusion in the cortex supplied by the middle cerebral artery suggesting a correlation between location of reduced CBF and clinical symptoms. The PCT did not indicate infarction, as CBF and CBV values were above critical ischemic levels [7], and CTA did not show any arterial stenosis as another possible cause of non-critical hypoperfusion. Therefore, we believe that this ictal hypoperfusion occurred secondary to CSD. Another important differential diagnosis for stroke mimics, postictal focal deficit (Todd's paresis), which would have warranted electroencephalography (EEG) after imaging, was abandoned as the decisive clues became available by the mother's information. The role of PCT in epileptogenic conditions is yet unclear, as both hypoperfusion and hyperperfusion have been demonstrated in case reports, possibly depending on whether the underlying pathophysiology was truly postictal or focal status epilepticus, respectively [8, 9].

CONCLUSION

Multimodal computed tomography scan can provide important information for differential diagnosis between migraine with aura and ischemic stroke. This can decisively influence hyperacute treatment, especially if taking medical history is hampered or even impossible.

****

Author Contributions

Anselm Angermaier – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
SoenkeLangner – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Michael Kirsch – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Alexander V Khaw – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

1. Vincent MB, Hadjikhani N. Migraine aura and related phenomena: Beyond scotomata and scintillations. Cephalalgia 2007;27(12):1368–77.
2. Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders: 2nd edition. Cephalalgia 2004;24:9–160.
3. Olesen J, Friberg L, Olsen TS, et al. Timing and topography of cerebral blood flow, aura, and headache during migraine attacks. Ann Neurol 1990;28(6):791–8.
4. Nieuwkamp DJ, van der Schaaf IC, Biessels GJ. Migraine aura presenting as dysphasia with global cognitive dysfunction and abnormalities on perfusion CT. Cephalalgia 2010;30(8):1007–9.
5. Floery D, Vosko MR, Fellner FA, et al. Acute-Onset Migrainous Aura Mimicking Acute Stroke: MR Perfusion Imaging Features. AJNR Am J Neuroradiol 2012;33(8):1546–2.
6. Lauritzen M. Pathophysiology of the migraine aura. The spreading depression theory. Brain 1994;117 (Pt 1):199–210.
7. Koenig M, Kraus M, Theek C, Klotz E, Gehlen W, Heuser L. Quantitative assessment of the ischemic brain by means of perfusion-related parameters derived from perfusion CT. Stroke 2001;32(2):431–7.
8. Mathews MS, Smith WS, Wintermark M, Dillon WP, Binder DK. Local cortical hypoperfusion imaged with CT perfusion during postictal Todd's paresis. Neuroradiology 2008;50(5):397–401.
9. Hauf M, Slotboom J, Nirkko A, von Bredow F, Ozdoba C, Wiest R. Cortical regional hyperperfusion in nonconvulsive status epilepticus measured by dynamic brain perfusion CT. AJNR Am J Neuroradiol 2009 Apr;30(4):693–8.

ABOUT THE AUTHORS

Article citation: Angermaier A, Langner S, Kirsch M, Khaw AV. Prolonged aphasia and perfusion computed tomography abnormalities in migraine with aura. International Journal of Case Reports and Images 2014;(3):221–224.

Anselm Angermaier is a Resident in the Department of Neurology, University Hospital Greifswald, Germany. His research interests include treatment of acute stroke and imaging of stroke.

Soenke Langner is chief consultant of the Section of Neuroradiology of the Department of Diagnostic Radiology and Neuroradiology, University Hospital Greifswald. His research interest include interventional therapy of acute stroke and stroke imaging.

CASE REPORT OPEN ACCESS

Stroke in pregnancy: A rare case of protein C and protein S deficiency

Snigdha Kumari, Ashok Kumar Biswas, Sukanta Misra

ABSTRACT

Introduction: Stroke in pregnancy is a rare occurrence, its reported incidence being 11–26 deliveries per 100,000. Ischemic strokes are slightly more common than hemorrhages. Subarachnoid hemorrhage, embolism and cerebral venous thrombosis can also occur, albeit with a lesser incidence. Peak incidence of stroke is in the peripartum and postpartum period. Mortality rate of pregnancy associated stroke is 10.13%. Therefore, rapid recognition and management of these patients are imperative to ensure successful outcome. We report a rare case of stroke during pregnancy which has the protein C and protein S deficiency as its precipitating cause. Case Report: We report a case of a 24-year-old female, with previous cesarean section in her third trimester, suddenly developed weakness of left side of the body and deviation of face to the right. There was no history of head trauma, fever, unconsciousness or seizure. She did not have any skin rash, arthritis or signs of deep vein thrombosis. There was no history of prior fetal loss. Protein C and protein S levels were low. Conclusion: We report a rare case of stroke during pregnancy which has the protein C and protein S deficiency as its precipitating cause. Stroke is a recognized complication of pregnancy, is uncommon but feared. Stroke in young adults aged 15–35 years is more common in women with poorer outcome in terms of disability and dependence. Most pregnancy related stroke occurs in peripartum and puerperium.

Keywords: Stroke, Pregnancy, Protein C deficiency, Protein S deficiency

Snigdha Kumari[1], Ashok Kumar Biswas[2], Sukanta Misra[3]
Affiliations: [1]MBBS, MS. Resident, Department of Obstetrics and Gynecology.Vivekananda Institute of Medical Sciences, Ramakrishna Mission SevaPratishthan.Kolkata, West Bengal, India; [2]MBBS, MD, DNB.Associate Professor, Department of Obstetrics and Gynecology.Vivekananda Institute of Medical Sciences, Ramakrishna Mission SevaPratishthan.Kolkata, West Bengal, India; [3]MBBS, MD, FICMCH.Professor, Department of Obstetrics and Gynecology. Vivekananda Institute of Medical Sciences, Ramakrishna MissionSevaPratishthan. Kolkata, West Bengal, India.
Corresponding Author: Snigdha Kumari, MBBS, Resident, Department of Obstetrics and Gynecology, Main office, Ramakrishna Mission SevaPratishthan, Vivekananda Institute of Medical Sciences, 99 Sarat Bose Road, Kolkata, India 700026; Ph: 919831376252; Email: snigdha.obs@gmail.com

Received: 19 August 2013
Accepted: 03 September 2013
Published: 01 March 2014

How to cite this article

Kumari S, Biswas AK, Misra S. Stroke in pregnancy: A rare case of protein C and protein S deficiency. International Journal of Case Reports and Images 2014;5(3):226–229.

doi:10.5348/ijcri-2014-03-479-CR-10

INTRODUCTION

Stroke in pregnancy is a rare presentation. The highest incidence of stroke is mainly in the peripartum and postpartum period. Ischemic strokes are slightly more common than hemorrhages. Subarachnoid hemorrhage, embolism and cerebral venous thrombosis can also occur, albeit with a lesser incidence. Cerebral venous thrombosis is a rare complication during pregnancy or the puerperium. The cerebrovascular system may be primarily involved in young adults suffering from anticoagulants deficiency. Women with thrombophilia are also more prone to venous thromboembolism in pregnancy and

puerperium. Acquired or hereditary thrombophilia occur in almost two-thirds of women presenting with recurrent miscarriages, preeclampsia, intrauterine growth restriction, abruptio placentae, or stillbirth, which are associated with microvascular thrombosis in placental blood vessels. Women with venous thromboembolism during pregnancy and especially those with thrombophilia need proper management, based on the type of defect, the detailed family history, and the presence of any additional risk factors. These factors are important for antithrombotic drug therapy during pregnancy and the puerperium, and the thrombo prophylactic strategy for future pregnancies. Low molecular weight heparin is effective in improving the outcome of pregnancy in women with previous obstetric complications. Mortality rate of pregnancy associated stroke is 10.13%.Therefore, rapid recognition and management of these patients are imperative to ensure successful outcome.

CASE REPORT

A 24-year-old female, with previous cesarean section in her third trimester, suddenly developed weakness of left side of the body and deviation of face to the right. She was brought to the emergency, examination revealed stable vitals (pulse 90/min, regular, blood pressure 130/84 mmHg, supine), altered level of consciousness (Glasgow coma scale 10), and left sided hemiparesis (power 2/5 in both upper and lower limbs). Deep tendon reflexes were brisk and Babinski sign was present, associated with upper motor neuron palsy of the left facial nerve. Other systemic examination was normal. There was no history of head trauma, fever, unconsciousness or seizure. She did not have any skin rash, arthritis or signs of deep vein thrombosis. There was a history of vertigo, unconsciousness, lower limb weakness, with recovery within an hour, 4 years back for which she received no treatment. There was no history of prior fetal loss.

An urgent non-contrast magnetic resonance imaging scan of brain was done which showed acute infarct in right centrum semiovale and right gangliocapsular region in the right middle cerebral artery territory (Figure 1). Patient's complete hemogram, liver function tests, serum creatinine and electrolytes were normal. Abdominal ultrasound showed a single, live fetus with normal parameters. Echocardiography was normal. She had normal prothrombin and activated partial thromboplastin time. The patient was seronegative for hepatitis B, C and HIV. Antinuclear factor by indirect immunofluorescence method was negative. Anticardiolipin antibodies, antineutrophil cytoplasmic antibodies (pANCA and cANCA) were negative. Blood homocysteine level, Lupus anticoagulant screen time was normal. Mutation of factor V could not be detected by real time polymerase chain reaction. Protein C (0.9 mg/L, reference range 1.67–3.16 mg/L) and protein S (7 mg/L, reference range 55–123 mg/L) levels were low.

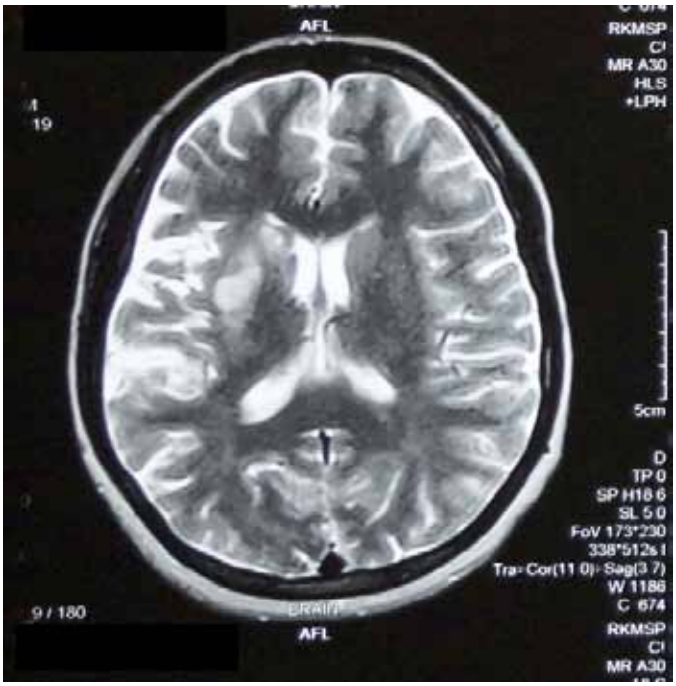


Figure 1: A non-contrast magnetic resonance imaging scan of brain showing an acute infarct in right centrum semiovale and right gangliocapsular region in the right middle cerebral artery territory.

She was diagnosed as a case of protein C and protein S deficiency and was started on low molecular weight heparin (60 mg intravenously twice daily) for 10 days. It was stopped 24 hours prior to elective cesarean section with patient's International normalized ratio 2.0. A three-kilogram healthy male baby was delivered. After 4 hours of cesarean section, low molecular weight heparin (60 mg subcutaneously once daily) was restarted. Subsequently, the patient was maintained with oral anticoagulant (2 mg once daily) with an overlap period of 3 days with a target International normalized ratio of 2.53.

DISCUSSION

Arterial thrombosis is reported in protein C, protein S, antithrombin deficient patients [1]. The incidence of pregnancy related stroke lies between 11–26 deliveries per 100,000 [2]. Prophylactic anticoagulation offers no benefit. Randomized control trials of treatment protocol in women with thrombophilia and pregnancy is needed. Stroke is a recognized complication of pregnancy, has to be taken care of. Stroke in young adults aged 15–35 years is more common in females with poorer outcome in terms of disability and outcome. Most pregnancy related stroke occurs in peripartum and puerperium [3]. Cerebral venous thrombosis is a rare complication during pregnancy or the puerperium [4]. The cerebrovascular system may be primarily involved in young adults suffering from anticoagulants deficiency [5]. Considering

the importance of prothrombotic state, especially caused by deficiency of protein S, any patient presenting with features of cerebrovascular accidents should be thoroughly investigated for any natural anticoagulants deficiency, in whom no other etiologic factors can be determined [6]. Hence, thrombophilia screening might be justified in women with pregnancy loss, and treatment with low molecular weight heparin might be considered for those with pregnancy loss and thrombophilia [7]. Women with thrombophilia are also more prone to venous thrombo embolism in pregnancy and puerperium [8]. Although initial results on use of thrombolysis in pregnancy have been encouraging, further evaluation with regard to maternal and fetal risk is warranted.

CONCLUSION

We report a rare case of stroke during pregnancy which has the protein C and protein S deficiency as its precipitating cause. Stroke in young adults aged 15–35 years is more common in females with poorer outcome in terms of disability and outcome. Hence, women with venous thromboembolism during pregnancy and especially those with thrombophilia need proper management, based on the type of defect, the detailed family history and the presence of any additional risk factors.

****

Author Contributions

Snigdha Kumari – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ashok Kumar Biswas – Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Sukanta Misra – Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

ACKNOWLEDGEMENTS

No grants or funds were needed for this study. We have no conflict of interest.

REFERENCES

1. Soare AM, Popa C. Deficiencies of proteins C, S and antithrombin and factor V Leiden and the risk of ischemic strokes. J Med Life 2010 Jul-Sep;3(3):235–8.
2. Davie CA, O'Brien P. Stroke and pregnancy. J Neurol Neurosurg Psychiatry 2008 Mar;79(3):240–5.
3. Salonen Ros H, Lichtenstein P, Bellocco R, Petersson G, Cnattingius S. Increased risks of circulatory diseases in late pregnancy and puerperium. Epidemiology 2001 Jul;12(4):456–60.
4. Klai S, Fekih-Mrissa N, Mrissa R, Rachdi R, Gritli N. Maternal cerebral venous thrombosis, uncommon but serious disorder, pathologic predictors and contribution of prothrombotic abnormalities. Blood Coagul Fibrinolysis 2013 Apr;24(3):269–72.
5. Martinez HR, Rangel-Guerra RA, Marfil LJ. Ischemic stroke due to deficiency of coagulation inhibitors. Report of 10 young adults. Stroke 1993 Jan;24(1):19–25.
6. Barinagarrementeria F, Cantú-Brito C, De La Peña A, Izaguirre R. Prothrombotic states in young people with idiopathic stroke. A prospective study. Stroke 1994 Feb;25(2):287–90.
7. Pabinger I. Thrombophilia and its impact on pregnancy. Thromb Res 2009;123 Suppl 3:S16–21.
8. Vormittag R, Pabinger I. Thrombophilia and pregnancy complications. Hamostaseologie 2006 Jan;26(1):59–62.

ABOUT THE AUTHORS

Article citation: Kumari S, Biswas AK, Misra S. Stroke in pregnancy: A rare case of protein C and protein S deficiency. International Journal of Case Reports and Images 2014;(3):225–228.

Snigdha Kumari is a Resident Surgeon in the department of obstetrics and gynecology at Vivekananda Institute of Medical Sciences, Kolkata, India, a Postgraduate institute and one of the apex referral centre. She is actively involved in the academic activities of the institute. Her field of interest is induction of labour, management of failed induction of labour, minimal invasive gynecological surgical intervention.

CASE REPORT OPEN ACCESS

Idiopathic pulmonary hemosiderosis without hemoptysis in an adult: A rare presentation

Jackin Moses R, Nishant Sinha, Madhusmita M, Kisku KH, Manjiri P

ABSTRACT

Introduction: Idiopathic pulmonary hemosiderosis (IPH) is defined as a clinical triad of hemoptysis, pulmonary infiltrates, and iron deficiency anemia. It is a diagnosis of exclusion when all other causes of diffuse alveolar hemorrhage have been ruled out. Epidemiologically, IPH is a rare disease, with an incidence of 0.24 per million children per year; overall, 80% of cases occur in children, most being diagnosed in the first decade of life. Case report: We present a 38-year-old female who presented to our casualty with acute respiratory failure which resolved on high dose corticosteroids. Patient on evaluation was diagnosed to have diffuse alveolar hemorrhage with idiopathic pulmonary hemosiderosis. Patient is currently asymptomatic and is on regular follow-up in our hospital.Conclusion: Diffuse alveolar hemorrhage should be a differential diagnosis in a patient presenting with hemoptysis, breathlessness, anemia and radiological evidence of infiltrates. Awaiting detailed work-up, patients should be promptly started on high dose steroids.

Keywords: Idiopathic pulmonary hemosiderosis (IPH), Diffuse alveolar hemorrhage (DAH), Acute interstitial pneumonia

Jackin Moses R, Nishant Sinha, Madhusmita M, Kisku K H, Manjiri P
Affiliations: Pondicherry Institute of Medical Sciences, Kalapet, Pondicherry, 605014.
Corresponding Author: Jackin Moses R, Room No: 535, 4th floor OPD block, PIMS campus, Kalapet, Pondicherry 605014, City: Pondicherry, State: Pondicherry, Country: India; Postal Code: 605014; Ph: +91-7708657414; Email: drjackinmoses@gmail.com

Received: 11 September 2013
Accepted: 18 October 2013
Published: 01 March 2014

How to cite this article

Jackin MR, Singha N, Madhusmita M, Kisku KH, Manjiri P. Idiopathic pulmonary hemosiderosis without hemoptysis in an adult: A rare presentation. International Journal of Case Reports and Images 2014;5(3):230–234.

doi:10.5348/ijcri-2014-03-480-CR-11

INTRODUCTION

Idiopathic pulmonary hemosiderosis (IPH) is defined as clinical triad of hemoptysis, pulmonary infiltrates, and iron deficiency anemia [1, 2]. It is a diagnosis of exclusion when all other causes of diffuse alveolar hemorrhage have been ruled out [3, 4]. Epidemiologically, IPH is a rare disease, with an incidence of 0.24 per million children per year [5]; overall, 80% of cases occur in children, most being diagnosed in the first decade of life [6]. The remaining 20% of cases are adult-onset IPH, most of which are diagnosed before 30 years of age. In adults it is more common in males and in smokers [6]. Our patient had a rare presentation as she is a 38-year-old female, non smoker and had no history of hemoptysis.

CASE REPORT

A 38-year-old female, presented to our emergency room with progressive breathlessness (MMRC grade 2-4) and dry cough for three months. She had history of multiple blood transfusions elsewhere during the prior three months period. She had no history of hemoptysis, hematemesis, menorrhagia, bleeding per rectum or any other obvious focus of bleeding. There was no history of fever. She had no symptoms related to connective tissue disorder. She was not a known smoker and had no history of exposure to toxic gases and fumes. She had no history of exposure to any drugs. Physical examination in the

emergency room revealed the temperature to be 370 °C, pulse rate 130 beats/minute, Blood pressure 130/80 mmHg, Respiratory rate 60/minute and SpO2 was 56% on room air ventilation. Her palpebral conjunctiva was pale. On auscultation fine end inspiratory crackles were heard bilaterally in all lung fields. Physical examinations of other systems were found in normal limits.

Arterial blood gas analysis of the patient showed Ph 7.46, PCo2 30 mmHg, Po2 37 mmHg, SpO2 77 mmHg (P/F ratio of 0.37). Her serum d-dimer done within two hours of admission was low. Electrocardiography and echocardiogram were normal. Her total counts were also normal. She was admitted in intensive care unit (ICU) and was treated with non invasive ventilation and high flow oxygen.

Subsequent investigations revealed corrected reticulocyte count 0.5%, microcytic hypochromic peripheral blood smear, low serum iron (36 µg/dL), low transferrin saturation (28%), normal Ferritin (140.2 ng/mL), normal total iron binding capacity (415 µg/dL) and normal serum vitamin B12 and folic acid levels. Her stool was negative for occult blood or parasites. Her serum creatinine was 0.8 mg/dL, blood urea was 36 mg/dL and urine microscopy was normal (no protein or red blood cells). The liver function tests and coagulation profile were within normal limits. Her retroviral status (HIV I and II by ELISA), hepatitis B surface antigen and hepatitis C antibody were negative. She received one pint blood transfusion in the ICU. Her chest X-ray (Figure 1) which was done in the ICU showed bilateral reticular infiltrates. Computed tomography (CT) scan of thorax was subsequently done which showed bilateral extensive ground glass opacities with interstitial thickening and nodules (Figure 2). A provisional diagnosis of acute interstitial pneumonia/diffuse alveolar hemorrhage was made and she was started on broad spectrum antibiotics with high dose pulse steroids (Inj methylprednisolone 500 mg i.v. daily in divided doses for 3 days). Within 3 days she improved significantly and was weaned off the non invasive ventilator support. Later the steroids were tapered to oral prednisolone at 0.75 mg/kg body weight. Subsequently fibre optic bronchoscopy was done under local anesthesia with conscious sedation. Sequential broncho alveolar lavage (BAL) was done from right middle lobe, it showed increasing hemorrhagic returns of BAL aliquots and BAL counts showed 60% hemosiderophages (Figure 3), confirming the diagnosis of diffuse alveolar hemorrhage. BAL was negative for gram stain and culture showing no evidence bacterial pathogens, acid fast bacilli, fungal organism, Fluorescent antibody stain for *Pneumocystis jeroveci* and cytology for cancer cells. Transbronchial lung biopsy was done from right middle and lower lobes. It showed interstitial fibrosis, hemosiderin laden macrophages in alveolar spaces and interstitium with positive Perl's stain (Figure 4). There was no evidence of vasculitis (capillaritis), granulomas, organising pneumonia and malignancy in the biopsy specimen. Her serum Rheumatoid factor

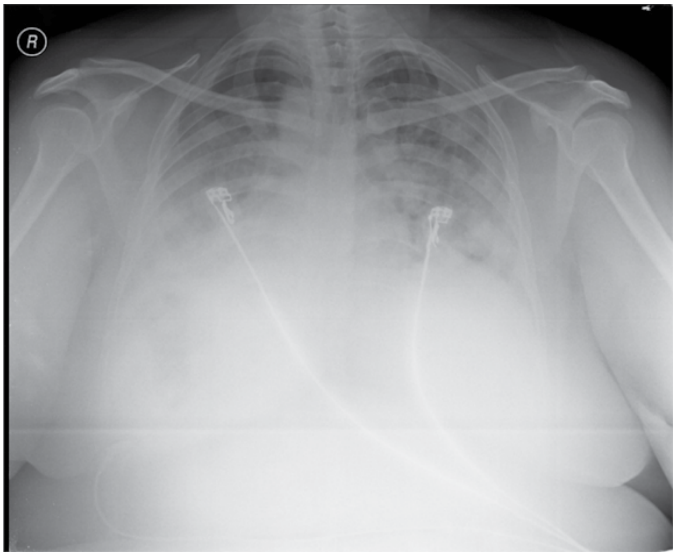


Figure 1: Chest X-ray showing diffuse reticulo nodular shadows involving all segments of both lung fields. X-ray done at the time of admission when the patient was in acute respiratory failure.

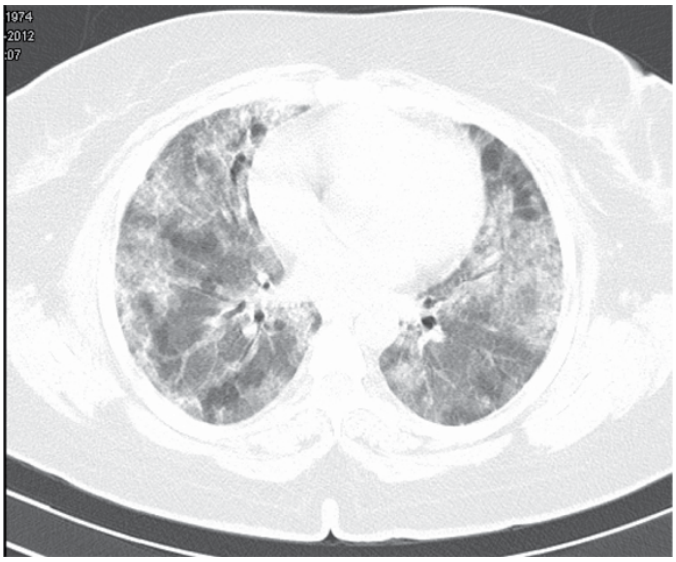


Figure 2: Computed tomography scan of chest revealed ground glass opacities with diffuse interstitial opacities.

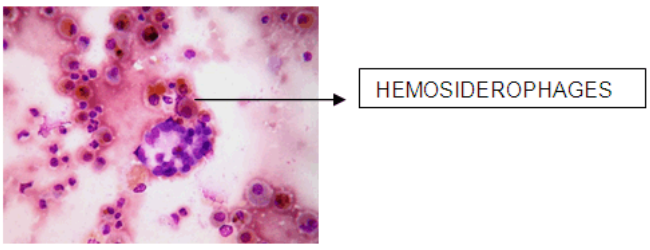


Figure 3: Broncho alveolar lavage showing Hemosiderin laden macrophages.

(RF), antinuclear antibody (ANA), Double stranded Deoxyribonucleic acid (DsDNA), antiphospholipid antibody(APLA), Anti Glomerular Basement Membrane

antibody (anti GBM) and anti U1 RNP were negative. Serum Complements (C3, C4 and Total Complement) and Antineutrophilic Cytoplasmic antibodies(c and p ANCA) were in normal range. After one week of treatment with steroids, she improved and there was no further drop in hemoglobin. At this time spirometry was done and it was normal. Her chest X-ray and CT thorax repeated after one month showed complete clearing of the shadows (Figure 5).

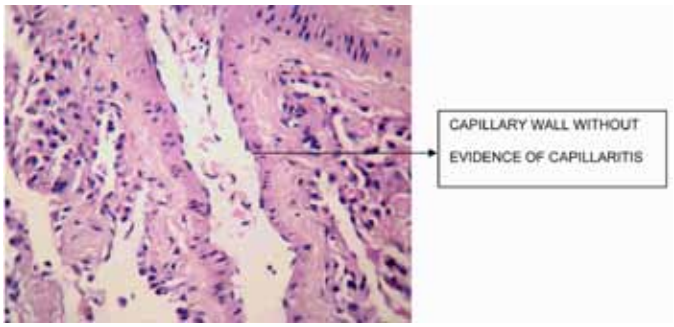


Figure 4: Tans bronchial lung biopsy capillary wall with no evidence of capillaritis.

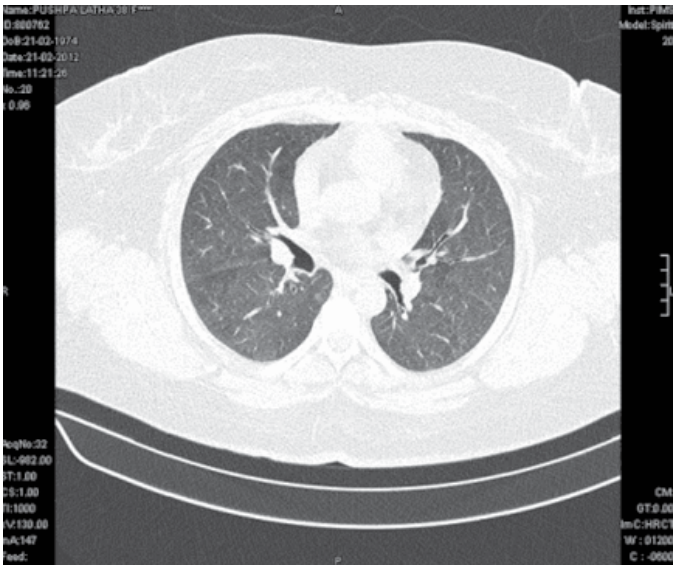


Figure 5: Post treatment computed tomography chest showing significant resolution.

DISCUSSION

The entity of IPH was initially described by Virchow [7] in 1864 as "brown lung induration". Waldenstrom [8] established the first ante mortem diagnosis in 1944. There are a few aetiological hypotheses of IPH including genetic, autoimmune, allergic, environmental and metabolic theory [9]. Environmental exposure to fungi (especially Stachybotrys atra) in water-damaged houses has been mentioned to cause IPH [10]. The clinical course includes two phases. First, an acute phase "IPH exacerbation", corresponds to intra-alveolar bleeding episodes, manifested by cough, dyspnea, hemoptysis and sometimes respiratory failure. Almost 100% of the adults experience hemoptysis during the disease course [9]. Secondly, the chronic phase is characterised by a slow resolution of previous symptoms, with or without treatment. The alternation of the two phases is largely unpredictable. Our patient never had hemoptysis.

The diagnosis of IPH is difficult, it requires the exclusion of other diseases which can be associated, such as cardiac diseases, bleeding disorders, connective-tissue diseases (systemic lupus erythematosus and rheumatoid arthritis), systemic vasculitis (Wegner's granulomatosis or microscopic polyangiitis), or anti-basement-membrane-antibody diseases (Goodpasture's syndrome). Laboratory analysis for ANCA, anti-GBM, antiphospholipid antibody, ANA and RF should so be negative. In our patient, they were all negative.

Association of IPH with celiac disease has been mentioned in literature [11]. However, we have not done serum antigliadin, antireticulin and cow-milk precipitins as she had no symptoms suggestive of celiac disease or milk allergy. Goodpasture's syndrome can present with or without pulmonary capillaritis [3], diffuse alveolar hemorrhage resulting from antibasement membrane antibody disease is usually associated with glomerulonephritis [1]. Immunofluorescence of transbronchial lung biopsy specimen was not done as she had no glomerulonephritis and serum anti GBM antibody was negative. Based on the clinico- radio- pathological interpretation, we feel that the findings are consistent with idiopathic pulmonary hemosiderosis.

A number of therapeutic trials have been tried, including: (1) splenectomy, without significant results (there is no evidence of hypersplenism), and (2) systemic glucocorticoid, commonly started in the acute phase of IPH, with apparent good control and impact on mortality [1]. Among immunosuppressants, azathioprine in combination corticosteroids might be the best therapeutic regimen, especially in preventing IPH exacerbations [12]. Adults with IPH seem to respond more favorably to oral corticosteroid therapy [6] than children, in whom treatment can be problematic. The recommended starting dose is 1 mg/kg/day prednisolone for two months, until the new alveolar infiltrates tend to resolve [13, 14]. In our patient, there was complete radiological clearing at four weeks and repeat CT thorax after four weeks showed no new infiltrates. We treated our patient with oral prednisolone 0.75 mg/kg body weight for 4 weeks, followed by 0.5 mg/kg body weight for another 8 weeks and 0.25 mg/kg body weight for another 12 weeks (Total duration of 6 months). We had planned to add azathioprine if she had recurrence or exacerbation. She is in our regular follow-up and has no recurrence of her symptoms. The long-term prognosis of the disease remains poor, and death may be due to massive diffuse pulmonary hemorrhage or chronic respiratory failure because of pulmonary fibrosis [11].

CONCLUSION

Diffuse alveolar haemorrhage should be a differential diagnosis in a patient presenting with hemoptysis, breathlessness, iron deficiency anemia and radiological evidence of infiltrates. Awaiting detailed work up, patients should be promptly started on high dose steroids. Idiopathic pulmonary hemosiderosis is confirmed when all other secondary causes of alveolar hemorrhage have been ruled out. Adults with IPH respond better to steroid therapy than children.

****

Author Contributions

Jackin Moses R – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Nishant Sinha – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published
Madhusmita M – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published
Kisku KH – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published
Manjiri P – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Soergel K, Sommers SC. Idiopathic pulmonary hemosiderosis and related syndromes. Am J Med 1962;32:499–511.
2. Morgan PG, Turner-Warwick M. Pulmonary haemosiderosis and pulmonary haemorrhage. Br J Dis Chest 1981;75(3):225–42.
3. Amy L Olson, Marvin I Schwarz. Diffuse Alveolar Hemorrhage. Prog Respir Res. Basel, Karger 2007;36:250–63.
4. Lara AR, Schwarz MI. Diffuse alveolar hemorrhage. Chest 2010;137(5):1164-71.
5. Kjellman B, Elinder G, Garwicz S, Svan H. Idiopathic pulmonary haemosiderosis in Swedish children. Acta Paediatr Scand 1984;73(5):584–8.
6. Miwa S, Imokawa S, Kato M, et al. Prognosis in adult patients with idiopathic pulmonary hemosiderosis. Intern Med 2011;50(17):1803–8.
7. Virchow R. Die krankhaften Geshwulste. Berlin, August Hirschwald 1864, p. 26–240.
8. Waldenstrom J. Relapsing, diffuse, pulmonary bleeding or hemosiderosis pulmonum - A new clinical diagnosis. Acta Radiol 1944;25:33–42.
9. Ioachimescu OC, Sieber S, Kotch A. Idiopathic pulmonary haemosiderosis revisited. Eur Respir J 2004;24(1):162-70.
10. Dearborn DG, Smith PG, Dahms BB, et al. Clinical profile of 30 infants with acute pulmonary hemorrhage in Cleveland. Pediatrics 2002;110(3):627–37.
11. Pacheco A, Casanova C, Fogue L, Sueiro A. Long-term clinical follow-up of adult idiopathic pulmonary hemosiderosis and celiac disease. Chest 1991;99(6):1525–6.
12. Airaghi L, Ciceri L, Giannini S, Ferrero S, Meroni PL, Tedeschi A. Idiopathic pulmonary haemosiderosis in an adult. Favourable response to azathioprine. Monaldi Arch Chest Dis 2001;56(3):211–3.
13. Michaeli J, Kornberg A, Menashe M, Lugassy G, Mogle P. Exacerbation of idiopathic pulmonary hemosiderosis in pregnancy. Eur J Obstet Gynecol Reprod Biol 1987;25(2):153–8.
14. Leatherman JW. Immune alveolar hemorrhage. Chest 1987;91(6):891–7.

CASE REPORT OPEN ACCESS

Benign reactive lesion with atypical mitosis: New example of an old story

Hua Zhong, Marina Chekmareva, Malik Deen, Michael May, Steven Deak, Nicola Barnard

ABSTRACT

Introduction: Ischemic fasciitis (atypical decubital fibroplasia) is a benign reactive lesion that may mimic mesenchymal malignancy. Atypical mitosis has never been reported in such a lesion, which adds an additional challenge to differential diagnosis. Case Report: A 94-year-old chronically immobile and debilitated female presented with a slightly painful and rapidly growing mass over her right back. Based on the clinical and histological features of the lesion, the diagnosis of ischemic fasciitis, a benign reactive lesion of the deep dermis, was made. Active mitosis and atypical mitotic figures were demonstrated in the lesion. Conclusion: Ischemic fasciitis may show atypical mitotic figures that can mislead differential diagnosis.

Keywords: Ischemic fasciitis, Atypical decubital fibroplasia, Atypical mitosis, Soft tissue

****

Hua Zhong[1,2], Marina Chekmarva[1,2], Malik Deen[2], Michael May[2], Steven Deak[3], Nicola Barnard[2]
Affiliations: [1]MD, PhD, Rutgers Cancer Institute of New Jersey, New Brunswick, New Jersey, United State; [2]MD, Department of Pathology and Laboratory Medicine, Rutgers Robert Wood Johnson Medical School, New Brunswick, New Jersey, United State; [3]MD, Department of Surgery, Rutgers Robert Wood Johnson Medical School, New Brunswick, New Jersey, United State.
Corresponding Author: Hua Zhong, MD, PhD, 195 Little Albany Street, New Brunswick, NJ08903; Ph: (732)235-8058; Fax: 732-235-6292; Email: zhonghu@rutgers.edu

Received: 12 September 2013
Accepted: 11 October 2013
Published: 01 March 2014

How to cite this article

Zhong H, Chekmareva M, Deen M, May M, Deak S, Barnard N. Benign reactive lesion with atypical mitosis: New example of an old story. International Journal of Case Reports and Images 2014;5(3):235–239.

doi:10.5348/ijcri-2014-03-481-CR-12

INTRODUCTION

In 1892, based on cancer research papers published in the early 20th century, Dr. Stroebe discovered asymmetrical division, or atypical mitosis, in nonmalignant processes with reactive or regenerative changes. Others, however, argued against his discovery and believed that atypical mitosis could only occur in carcinomas, sarcomas or unusual inflammatory reaction in a Cysticercus rat model of which cells with atypical mitosis eventually resulted in malignancy [1]. After over one century, a general consensus has been reached in the surgical pathology society, stating that an atypical mitotic figure may indicate a malignant process while it can also be seen in benign lesions. For example, atypical mitotic figures can be occasionally found in granulation tissue following ionizing-radiation exposur, ischemic colitis, long standing ulcerative colitis, gastric intestinal metaplasia, endometrium associated with chorionic tissue effect, benign cutaneous lesions induced by taxane therapy, and abnormal but non-neoplastic astrocytes [2–8]. Herein, a rare case of ischemic fasciitis, also named as atypical decubital fibroplasia, is reported, which shows typical clinical and histological features, except that atypical mitosis is evident.

CASE REPORT

The patient was a 94-year-old female who was chronically immobile and debilitated with a history of

locally recurrent basal cell carcinoma (left upper lip and face, up to 1.1 cm in diameter) and superficially invasive cutaneous squamous cell carcinoma (right cheek, 0.8 cm in diameter) of the head and neck region. These cutaneous carcinomas were resected with negative margins. The patient presented with a slightly painful and rapidly growing mass over her right back for two months. She denied history of trauma or prior surgical procedure to the region. On physical examination, the skin surface on the right scapular and axillary regions appeared unremarkable. A 3x3.5 cm slightly firm mass was palpated at the lateral edge of the right scapula at right axillary level. The mobility of the mass was limited while the overlying skin was mobile. No other masses or any signs of lymphadenopathy were present. Results of basic laboratory tests were within normal limits. Since clinical and laboratory findings did not suggest infection, a soft tissue sarcoma or a metastatic lesion was suspected. Fine-needle aspiration (FNA) was performed, and was followed by surgical excision of the mass with skin sparing.

The FNA specimen (Figure 1A–B) was basically hypocellular with a background of red blood cells. Smears showed loose aggregates of cells grouped around delicate, branching vascular networks in addition to scant loose cells. Individual cellular constituents included a mixture of spindle shaped fibroblast or myofibroblast-like cells with ample cytoplasm that tended to taper at the ends and with plump ganglion-like cells. Nuclei were large and often dark on smears. In addition to the cellular component, the aspirates demonstrated a background of granular-serous materials and occasional inflammatory cells. The latter included lymphocytes, foamy macrophages and rare neutrophils. Collagen micro-fragments were occasionally seen. Cell block showed fragments of granulation tissue or necrotic tissue with small cysts, which was basically similar histology as demonstrated in the resection specimen (Figure 2A–F). An FNA cytopathology was interpreted as an atypical spindle cell lesion, and deferred to tissue diagnosis.

The surgically excised specimen grossly revealed an irregular, slightly firm, pink-tan and ill-defined mass, 3.5x3.0x2.5 cm in size. Cut surfaces were solid, variegated, red-tan, focally hemorrhagic and myxoid. Histologically, the lesion was situated in the deep subcutis adjacent to superficial skeletal muscle layer. The lesion consisted of a distinct zonal configuration with a central zone of remarkable fibrinoid necrosis forming small cystic and spongy architectures (Figure 2A). The fibrinoid necrosis involved adipose tissue as well as small vessels of which the lumens became obliterated (Figure 2E). At the periphery of the lesion was an irregular and thick layer of granulation tissue with vascular proliferation and prominent proliferating endothelial cells. The area was admixed with plump ganglion-like cells (Figure 2C–D) in a background of collagenous or myxoid stroma. The ganglion-like cells showed enlarged eccentric nuclei with vesicular chromatin, prominent centrally located

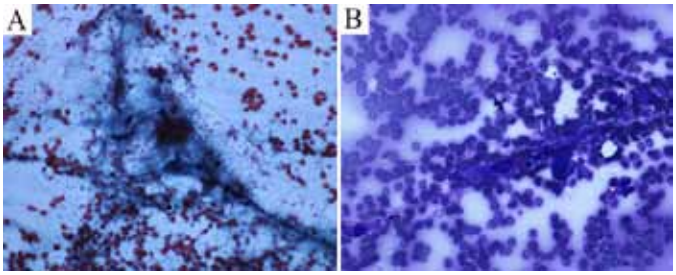


Figure 1: Cytological feature of ischemic fasciitis. Smears of fine needle aspiration were shown in (A) Pap stain, x200, and (B) Diff Quik stain, x400. Loose aggregates of atypical spindle cells were around branching vessels. Nuclei of the spindle cells were enlarged and dark.

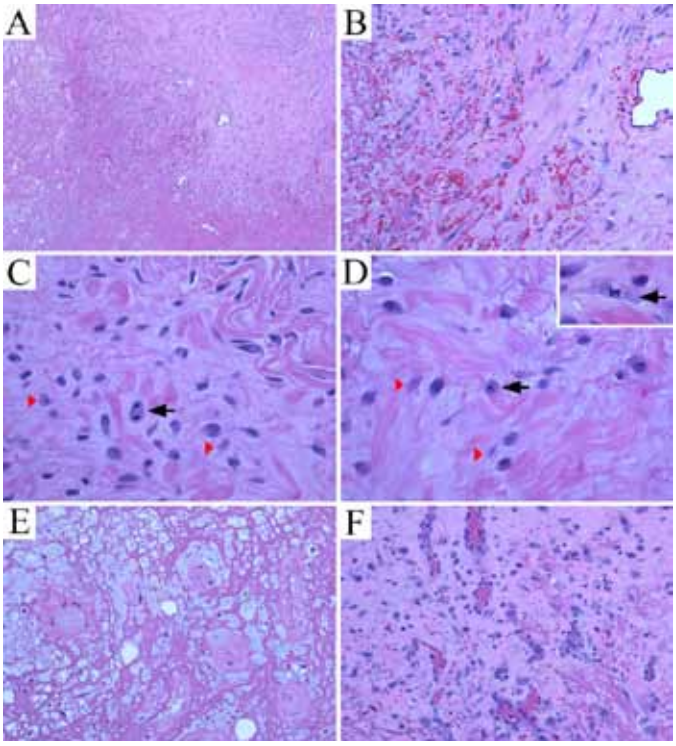


Figure 2: Histomorphological features of ischemic fasciitis. (A) Partly showing zonal configuration with irregular junction between necrotic zone and adjacent granulation tissue (H&E stain, x400), (B) A higher power view of Figure 2A showing extravasated erythrocytes in the junction of necrotic zone and granulation tissue (H&E stain, x200), (C) Showing granulation tissue with plump ganglion-like cells in collagenous stroma (H&E stain, x400). A normal anaphase mitotic figure is shown in the center (black arrow). Cells with lightly smudged nuclei or without appreciable nuclear chromatin materials are shown (red arrow heads), (D) Showing granulation tissue with plump ganglion-like cells in collagenous stroma (H&E stain, x400). Two atypical (abnormal) mitotic figures are shown, a tripolar (black arrow) and a multipolar mitotic figure (insert – from another high power field, x400). Cells with lightly smudged nuclei or without appreciable nuclear chromatin materials are shown (red arrow heads), (E) Showing fibrinoid necrosis involving small vessels, mixed with myxoid stroma and dispersedly extravasated erythrocytes (H&E stain, x200), and (F) Showing granulation tissue with thin wall vessels that are characterized by margination of and extravasation of inflammatory cells (H&E stain, x200).

nucleoli, and abundant eosinophilic cytoplasm. Fairly often, smaller cells with similar cytoplasmic features were shown with lightly smudged nuclei or occasionally with no appreciable chromatin materials (Figure 2C, D). These cells demonstrated strongly diffuse immunoreactivity for vimentin, were weakly for CD68 (cytoplasmic), weakly focal for CD31, but were completely negative for pan-cytokeratin and S100. The mitotic figure count equaled 15 per 50 high power fields (40x objective and 10x ocular). Two atypical mitotic figures were demonstrated (Figure 2D), a tripolar and a multipolar. Scattered mononucleated inflammatory cells and rare polymorphonuclear leukocytes were also seen. Focus of granulation tissue with thin wall vessels characterized by margination and extravasation of inflammatory cells was noted (Figure 2F). Foci of extravasated erythrocytes were frequently evident, mostly located in association with granulation tissue immediately adjacent to the necrotic zone (Figure 2A–B). The lesion was non-encapsulated, generally hypocellular and with an ill-defined margin. Local induration was noted on the surgical site at one month follow-up. The patient was followed-up again at three and six months post-surgery with completely recovery.

Overall findings favored a reactive process— an entity with so-called ischemic fasciitis/atypical decubital fibroplasia that may show active mitosis, whereas atypical mitotic figures have never been reported in such a lesion.

DISCUSSION

Ischemic fasciitis is a rare, but distinctive entity. It is a benign reactive lesion that usually occurs in elderly immobile patients in the bony prominence overlying connective tissue of weight-bearing areas [9]. The pathogenesis of ischemic fasciitis is hypothetically related to intermittent ischemia with resultant fibrinoid necrosis and regenerative changes [9]. The hypothetical pathogenesis does not appear to have yet been reproduced and confirmed in any animal models. The main pitfall in diagnosing the lesion is to clinically and pathologically mistake it for a malignancy of mesenchymal origin [9, 10]. Our case regarding the presentation of atypical mitotic figures in particular raises the consideration of malignancy. The entire histological context, however, does not appear to fit a malignant process. The characteristic clinical and histologic features of ischemic fasciitis have been described in several case reports and case series since 1992 [9]. The histological features of ischemic fasciitis partly overlap with but generally show less cellularity than other fasciitis-type or inflammatory myofibroblastic lesions [9, 10]. Ischemic fasciitis is thought to represent a constellation of fibroblastic or myofibroblastic proliferation, and it is a good histological example in the spectrum of pseudosarcomatous lesions [9]. Thus far, abnormal or atypical mitosis has never been reported in ischemic fasciitis or atypical decubital fibroplasia. Recognizing its typical clinical and histological features is critical in differential diagnosis. As in the current case, although atypical mitotic figures are present, the overall characteristic clinical and histological features favor the diagnosis of ischemic fasciitis.

Atypical or abnormal mitotic figure is one of the morphological changes that imply anaplasia or lack of differentiation [11]. However, demonstrating atypical mitotic figures in a benign reactive lesion is not something new, but is rare finding in both experimental and surgical pathology related to inflammatory or regenerative processes [1–8]. One may argue that some of above mentioned chronic pathological processes, for example, long standing ulcerative colitis [4] and gastric intestinal metaplasia [5], are associated with malignant potential. It is true that the role of ulcerative colitis or intestinal metaplasia as pre-neoplastic lesion is well established, associating with various genetic alterations or chromosomal instability [12, 13]. It is also true that some of but not all these clinical cases will develop malignant tumors. The fact is that there is a lack of or it is difficult to perform clinical studies that support direct malignant association of which a reactive lesion with atypical mitosis eventually develops into a malignant tumor. Nevertheless, ischemic fasciitis is classified as a reactive lesion with unique clinical scenario that is not associated with any malignant behavior. Our current case provides a new example to the old story of which atypical mitosis occurs in a benign reactive lesion.

The mechanisms of atypical mitosis are very complicated, and its biological significance is largely context-dependent. In the early studies reviewed by Dr. Mendelsohn [1], atypical mitosis has been shown in cultured cells subjected to a variety of physical and chemical treatments. Recent studies have demonstrated that abnormal mitosis and DNA damage are reciprocal processes, and that abnormal mitosis can even potentially facilitate the activation of oncogenic stimuli that may promote transformation [14]. The occurrence of atypical mitotic figures, especially multipolar mitosis, is closely associated with centrosome defects and chromosome missegregation [15]. Chromosome missegregation is physically and chemically inducible in primary human fibroblasts [16] and immortalized retinal pigment epithelial cells [17] at a rate lower than neoplastic cells. In addition, induced mitotic defects or chromosome missegregation can be transient and dynamic [17, 18].

The above lines of evidence suggest that chromosomal alterations or atypical mitotic figures can be induced in specific conditions, and that not all atypical mitotic events lead to malignant transformation. In other words, atypical mitosis does not equal to malignancy, although a majority of them do.

CONCLUSION

Ischemic fasciitis/atypical decubital fibroplasia may show atypical mitotic figures that can mislead differential

diagnosis. This report adds one more example to the benign reactive lesions with atypical mitosis.

****

Author Contributions

Hua Zhong – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Marina Chekmareva – Substantial contributions to Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Malik Deen – Substantial contributions to Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Michael May – Substantial contributions to Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Steven Deak – Substantial contributions to Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Nicola Barnard – Substantial contributions to Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Mendelsohn W. The significance of abnormal mitosis in the development of malignancy. Am J Cancer 1935;24:626-636.
2. Weidner N, Askin FB, Berthrong M, Hopkins MB, Kute TE, McGuirt FW. Bizarre (pseudomalignant) granulation-tissue reactions following ionizing-radiation exposure. A microscopic, immunohistochemical, and flow-cytometric study. Cancer 1987;59:1509-1514.
3. Zhang S, Ashraf M, Schinella R. Ischemic colitis with atypical reactive changes that mimic dysplasia (pseudodysplasia). Arch Pathol Lab Med 2001;125:224-227.
4. Rubio CA, Befrits R. Atypical mitoses in colectomy specimens from patients with long standing ulcerative colitis. Anticancer Res 1997;17:2721-2726.
5. Rubio CA, Kato Y, Kitagawa T. Atypical mitosis in gastric intestinal metaplasia in Japanese patients. Jpn J Cancer Res 1993;84:493-494.
6. Arias-Stella J, Jr., Arias-Velasquez A, Arias-Stella J. Normal and abnormal mitoses in the atypical endometrial change associated with chorionic tissue effect. Am J Surg Pathol 1994;18:694-701.
7. Plummer RS, Shea CR. Dermatopathologic effects of taxane therapy. J Am Acad Dermatol 2011;65:592-596.
8. Kuhlmann T, Lassmann H, Bruck W. Diagnosis of inflammatory demyelination in biopsy specimens: a practical approach. Acta Neuropathol 2008;115:275-287.
9. Montgomery EA, Meis JM, Mitchell MS, Enzinger FM. Atypical decubital fibroplasia. A distinctive fibroblastic pseudotumor occurring in debilitated patients. Am J Surg Pathol 1992;16:708-715.
10. Liegl B, Fletcher, C.D. Ischemic fasciitis: analysis of 44 cases indicating an inconsistent association with immobility or debilitation. Am J Surg Pathol 2008;32:1546-1552.
11. Kumar V, Abbas, A, Fausto, N (eds). Robbins and Cotran Pathology Basis of Disease, 7TH ed.: Elsevier Saunders, Philadephia, 2005.
12. Seril DN, Liao J, Yang GY, Yang CS. Oxidative stress and ulcerative colitis-associated carcinogenesis: studies in humans and animal models. Carcinogenesis 2003;24:353-362.
13. Tsukamoto T, Mizoshita T, Tatematsu M. Animal models of stomach carcinogenesis. Toxicol Pathol 2007;35:636-648.
14. Ganem NJ, Pellman D. Linking abnormal mitosis to the acquisition of DNA damage. J Cell Biol 2012;199:871-881.
15. Lingle WL, Salisbury JL. Altered centrosome structure is associated with abnormal mitoses in human breast tumors. Am J Pathol 1999;155:1941-1951.
16. Cimini D, Tanzarella C, Degrassi F. Differences in malsegregation rates obtained by scoring ana-telophases or binucleate cells. Mutagenesis 1999;14:563-568.
17. Thompson SL, Compton DA. Examining the link between chromosomal instability and aneuploidy in human cells. J Cell Biol 2008;180:665-672.
18. Silkworth WT, Cimini D. Transient defects of mitotic spindle geometry and chromosome segregation errors. Cell Div 2012;7:19.

CASE REPORT OPEN ACCESS

An unusual presentation of infected urachal cyst in an adult

Anand Munghate, Ashwani Kumar, Harnam Singh, Mahak Chauhan, Gurpreet Singh, Manish Yadav

ABSTRACT

Introduction: Urachus, median umbilical ligament, is normally obliterated in early infancy. So being remnant, is uncommon in adults. Delay in diagnosis and management can present with complications like drainage from umbilicus, severe abdominal infection, cyst with stone formation, fistula to urinary bladder, peritonitis, lump abdomen or carcinoma. Diagnosis remains challenging due to the rarity of this lesion and the non-specific nature of its symptomatology. Case Report: We report a case of a 35-year-old female presenting with umbilical sepsis with abdominal (suprapubic) pain. Investigations and laparotomy lead us to a confirmative diagnosis of infected urachal cyst. Conclusion: Urachal anomalies are rare in adults. Presentation is atypical therefore a high index of suspicion is required in order to achieve a diagnosis. Complete surgical excision is the treatment of choice due to the risk of malignant transformation.

Anand Munghate[1], Ashwani Kumar[2], Harnam Singh[3], Mahak Chauhan[4], Gurpreet Singh[5], Manish Yadav[5]
Affiliations: [1]MS (General Surgery), Junior Resident, Department of General Surgery, Government Medical College, Patiala, Punjab, India; [2]M.S (General Surgery), Professor, Department of General Surgery, GovernmentMedical College, Patiala, Punjab, India; [3]MS (General Surgery), Assistant Professor, Department of General Surgery,GovernmentMedical College, Patiala, Punjab, India; [4]MBBS, Intern, Department of General Surgery, Government Medical College, Patiala, Punjab, India; 5MS (General Surgery), Junior Resident, Department of General Surgery, GovernmentMedical College, Patiala, Punjab, India.
Corresponding Author: Anand Munghate, Department of General Surgery, Government Medical College and Rajindra Hospital, Patiala, Punjab, India 147001; Ph: +919779396131; +9101752212055; Email: dr.anand24by7@yahoo.com

Received: 08 October 2013
Accepted: 09 November 2013
Published: 01 March 2014

Keywords: Urachus, Urachal cyst, Umbilicus

How to cite this article

Munghate A, Kumar A, Singh H, Chauhan M, Singh G, Yadav M. An unusual presentation of infected urachal cyst in an adult. International Journal of Case Reports and Images 2014;5(3):240–243.

doi:10.5348/ijcri-2014-03-482-CR-13

INTRODUCTION

Urachal is a rare congenital abnormality of abdominal wall defect which results from incomplete regression of the fetal urachus. The urachus is a fibromuscular tubular extension of the allantois that develops with the descent of the bladder to its pelvic position. They are more common in children than in adults, due to urachal obliteration in early infancy [1]. Remnants of the tract may present as a patent urachus, vesicourachal diverticulum, urachal sinus or urachal cyst [2]. The incidence of urachal cyst in adults is rare and it is more common in men than women. In adults, urachal cyst is the commonest variety, with infection being the usual mode of presentation [3]. An infected urachal cyst usually presents with lower abdominal pain, a tender mass, fever, dysuria, voiding difficulty or even with umbilical drainage [4]. Ultrasonography, computed tomography (CT) and magnetic resonance imaging (MRI) scans will all assist in making this unusual diagnosis. Treatment is by complete excision, however, techniques have been debated . We present a unique case of an infected urachal cyst in an adult patient with abdominal pain and umbilical sepsis.

CASE REPORT

A 35-year-old female was presented in outpatient department with history of pain in lower abdomen (suprapubic region) for last 15 days and umbilical sepsis

with periumbilical erythema for eight days. She gave no history of nausea, vomiting or change in bowel or bladder habits. Systemic examination revealed periumbilical erythema and tenderness in suprapubic region with soft abdomen. Erythema subsided with intravenous antibiotic therapy for one week.

Ultrasonography and contrast-enhanced computed tomography (CECT) scan of abdomen and pelvis showed heterogeneous mass of size ~6.4x2.8 cm in midline extending from anterior wall of urinary bladder to umbilicus involving both recti (Figure 1 and Figure 2). The laparotomy was performed, infraumbilical transverse incision was given (Figure 3) which revealed urachal cyst surrounded by an inflammatory mass extending into the dome of the urinary bladder. The infected urachal cyst and urinary bladder dome were excised and sent for histopathological analysis to rule out any evidence of malignancy (Figure 4). Postoperative period was uneventful and patient was discharged in satisfactory condition. Histopathological analysis of the resected specimen showed chronic granulomatous inflammation with no evidence of malignancy. On follow-up, patient is doing well with no episode of pain abdomen or bowel and bladder disturbances.

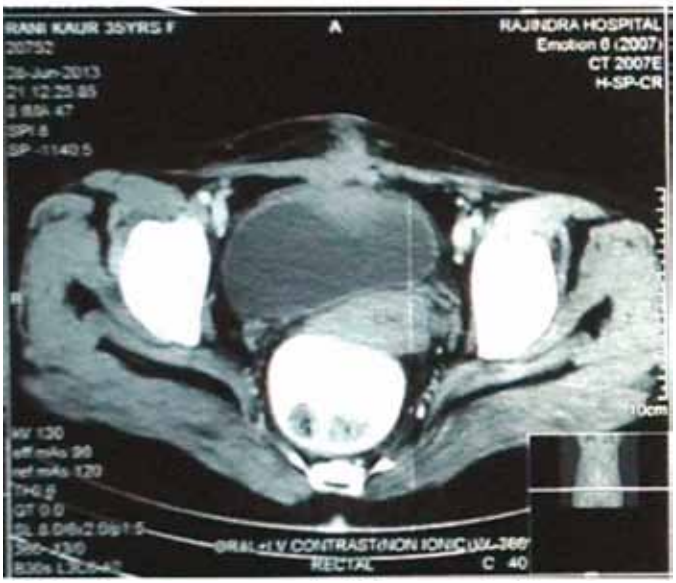


Figure 1: Computed tomography scan of abdomen pelvis showing a large inflammatory mass in midline extending from anterior wall of urinary bladder to umbilicus involving both recti.

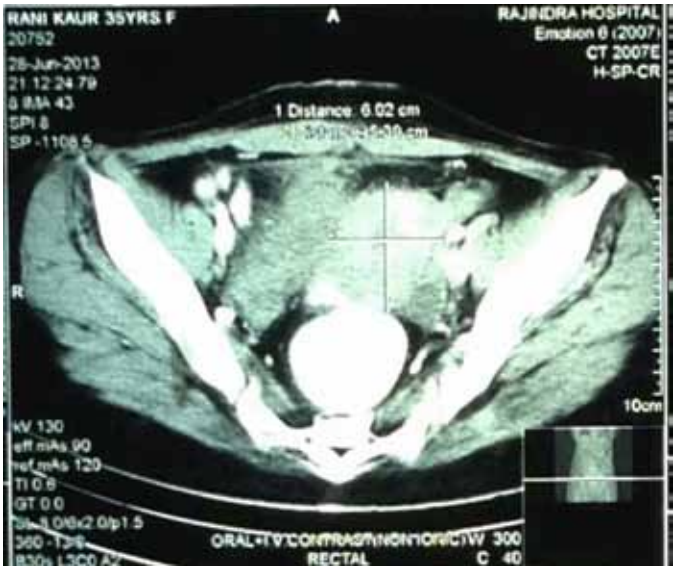


Figure 2: Computed tomography of abdomen pelvis showing a large inflammatory mass in midline at anterosuperior to anterior wall of urinary bladder.

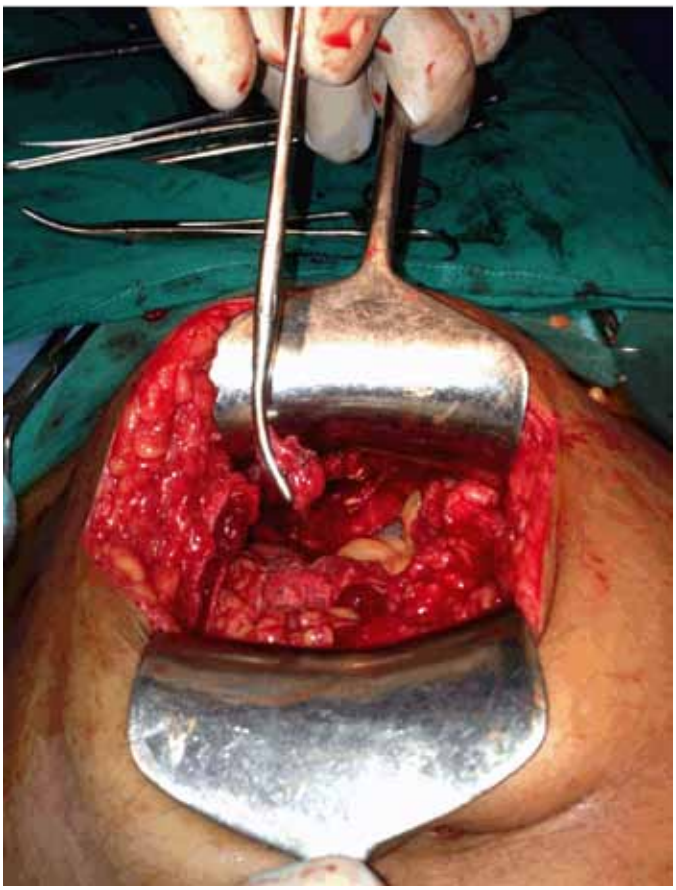


Figure 3: Intra-operative picture showing urachal tract extending up to inflammatory mass at anterior wall of urinary bladder.

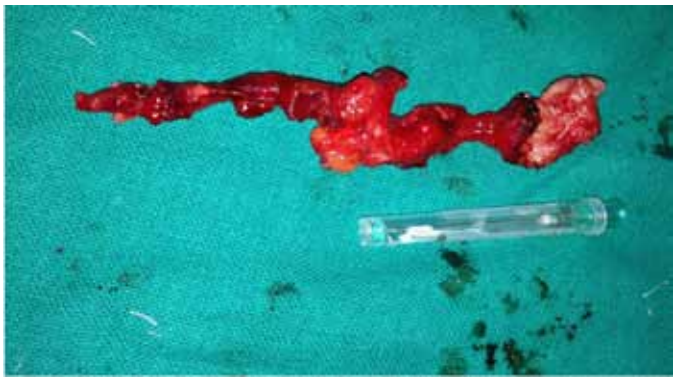


Figure 4: The urachal cyst with tract containing inflammatory mass.

DISCUSSION

Urachus, developmentally is the upper part of the bladder, both of which arise from the ventral part of the cloaca and allantois. Descent of the bladder from the fifth month of development into the fetal pelvis pulls the urachus with it resulting in the formation of the urachal canal. The lumen of this canal progressively obliterates during fetal life, with eventual formation of a fibrous tract in early adult life [5]. Histologically, it is composed of three layers; an innermost layer of modified transitional epithelium similar to the urothelium, the middle layer of fibroconnective tissue and outermost layer of smooth muscle continuous with the detrusor [1, 5]. Incomplete regression of the urachal lumen results in the following abnormalities:

i. patent urachus~ 50%; in which the entire tubular structure fails to close
ii. urachal cyst ~30% ; in which both ends of the canal close leaving an open central portion
iii. urachal sinus ~15%; which drains proximally into the umbilicus
iv. vesicourachal diverticulum ~5%; where the distal communication to the bladder persists [2].

The incidence of urachal cyst in adults is rare and it is more common in men than women [3, 6]. In adults, the most common variety is urachal cyst, with infection being usual mode of presentation, otherwise the condition usually remains asymptomatic [3]. Infected urachal cyst usually presents with lower abdominal pain, a tender mass, fever, dysuria, voiding difficulty or even with umbilical drainage [4]. The route of infection is hematogenous, lymphatic, direct or ascending from the bladder. The commonly cultured microorganisms from the cystic fluid include *Escherichia coli*, *Enterococcus faecium*, *Klebsiella pneumonia*, *Proteus*, *Streptococcus* viridians and *Fusobacterium* [4, 6].

Our case highlights the potential complications related to congenital urachal anamolies in patient presenting with complaints of umbilical sepsis with suprapubic abdominal pain. Diagnosis of an infected urachus cyst was made after radiological investigations.

The risk of urachal malignancy in adults is high and the prognosis is poor. Histologically, the innermost layer of the urachus is mainly transitional cell. Adenocarcinoma is the predominant histological type and most are mucinous. This is probably due to metaplasia arising from chronic inflammation. The prognosis for urachal adenocarcinoma does not differ significantly from non-urachal adenocarcinoma and is relatively poor, with a five year survival of 37% and a 10 year survival of 17% [7]. CT scan or MRI scan is essential for confirming diagnosis because of the nature of the condition radiographic evaluation of urachal cyst by ultrasonography. Ultrasound scan can help to make diagnosis in 77% of patients [4]. As in our case, ultrasound finding raised the differential diagnosis of urachal cyst showing collection/mass anterior to urinary bladder and involving abdominal wall. Diagnosis is often made following exploratory laparotomy for an unexplained acute abdomen. The treatment of choice for urachal cyst is by complete primary excision. However, Yoo et al. [4] in their study suggested a two-stage procedure involving initial incision and drainage followed by later excision of the urachal remnant. Complete excision is important because malignant transformation of the remnant is possible [8]. Traditionally, open excision has been the approach of choice. However, a laparoscopic approach is also an attractive alternative in recent years [9]. The advantage of this approach is good view and the minimal risk of incomplete excision of the urachal remnant [10].

CONCLUSION

Urachal anomalies are rare in adults. Presentation is atypical, therefore, a high index of suspicion is required in order to achieve a diagnosis. A triad of lower midline mass, umbilical discharge and sepsis is suggestive. However, radiological investigations such as ultrasonography, magnetic resonance imaging and computed tomography scans confirm the diagnosis and defines the surrounding anatomical relationship. Complete surgical excision is the treatment of choice due to the risk of malignant transformation.

****

Author Contributions

Anand Munghate – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ashwani Kumar – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Harnam Singh – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Mahak Chauhan – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Gurpreet Singh – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Manish Yadav – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright

REFERENCES

1. Hammond G, Yglesias L, Davis JE. The urachus, its anatomy and associated fascia. Anat Rec 1941;80:271–4.
2. Nimmonrat A, Na-ChiangMai W, Muttarak M. Urachal abnormalities: Clinical and imaging features. Singapore Medical Journal 2008;49(11):930–5.
3. Risher WH, Sardi A, Bolton J. Urachal abnormalities in adults: The Ochsner experience. South Med J 1990;83(9):1036–9.
4. Yoo KH, Lee SJ, Chang SG. Treatment of infected urachal cysts. Yonsei Medical Journal 2006;47(3):423–7.
5. Begg RC. The Urachus. Its anatomy, histology and development. J Anat 1930;64(Pt 2):170–83.
6. Ashley RA, Inman BA, Routh JC, Rohlinger AL, Husmann DA, Kramer SA. Urachal anomalies: A longitudinal study of urachal remnants in children and adults. J Urol 2007;178(4 Pt 2):1615–8.
7. Grignon DJ1, Ro JY, Ayala AG, Johnson DE, Ordóñez NG. Primary adenocarcinoma of the urinary bladder: A clinicopathologic analysis of 72 cases. Cancer 1991;67(8):2165–72.
8. Yohannes P, Bruno T, Pathan M, Baltaro R. Laparoscopic radical excision of urachal sinus. Journal of Endourology 2003;17(7):475–9.
9. Groot-Wassink T, Deo H, Charfare H, Foley R. Laparoscopic excision of the urachus. Surg Endosc 2000;14(7):680–1.
10. Cutting CW, Hindley RG, Poulsen J. Laparoscopic management of complicated urachal remnants. BJU Int 2005;96(9):1417–21.

CLINICAL IMAGES OPEN ACCESS

Metopic synostosis

Natasha Gupta, Blankstein Josef

CASE REPORT

A 35-year-old female delivered a male infant prematurely at 33 weeks. Neonate weighed 1930 grams at birth and was noted to have a keel like protrusion of the forehead, hypertelorism, narrow temples and fused metopic suture. All the other sutures were noted to be open. A detailed karyotype assessment of the neonate was normal, thus no genetic etiology could be identified. This was considered to be a case of isolated metopic synostosis or single suture craniosynostosis, also referred to as nonsyndromic craniosynostosis. We managed this case with surgical repair which is the mainstay of treatment in craniosynostosis. We present here the 3D reconstructed images from computed tomography scan of the neonate's skull (Figures 1 and 2).

DISCUSSION

Metopic synostosis is a type of craniosynostosis where metopic suture of the fetal skull fuses prematurely leading to a skull malformation called trigonocephaly (triangular forehead) and low volume of anterior cranial fossa. This leads to impaired skull and brain growth. It is associated with advanced maternal age, male fetuses, multiple births, preterm gestation and low birth weight infants [1]. Incidence of metopic synostosis has been increasing over last few years, although incidence of other craniosynostosis including sagittal, unicoronal and multisutural has remained unchanged [1]. The increase

Natasha Gupta[1], Blankstein Josef[1]
Affiliations: [1]MD, Department of Obstetrics and Gynecology, Mount Sinai Hospital, 1500 S California Ave, Chicago
Corresponding Author: Natasha Gupta, Department of Obstetrics and Gynecology, Mount Sinai Hospital, 1500 S California Ave, S Washtenaw Ave, Unit 1, Chicago, IL, 60608.; Phone No: 248-464-0451; Fax: 773-257-6359; Email: docnatasha3@gmail.com

Received: 21 December 2012
Accepted: 03 August 2013
Published: 01 March 2014

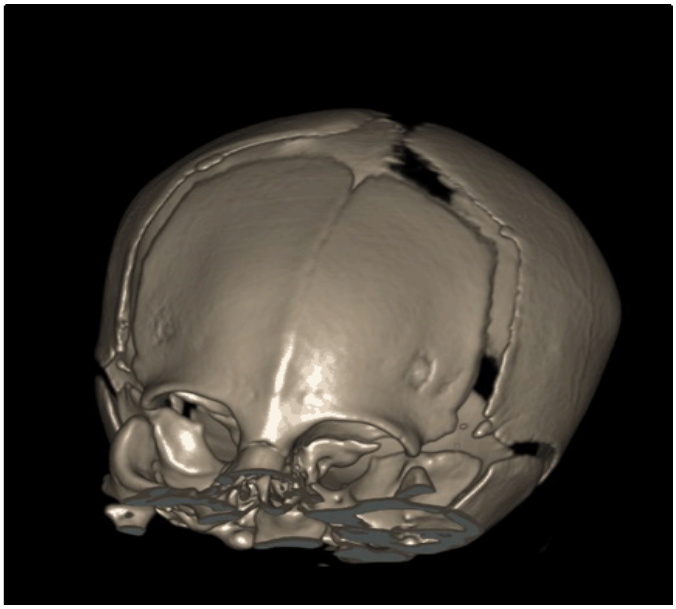


Figure 1: 3D reconstruction of skull from computed tomography scan showing prematurely fused metopic suture.

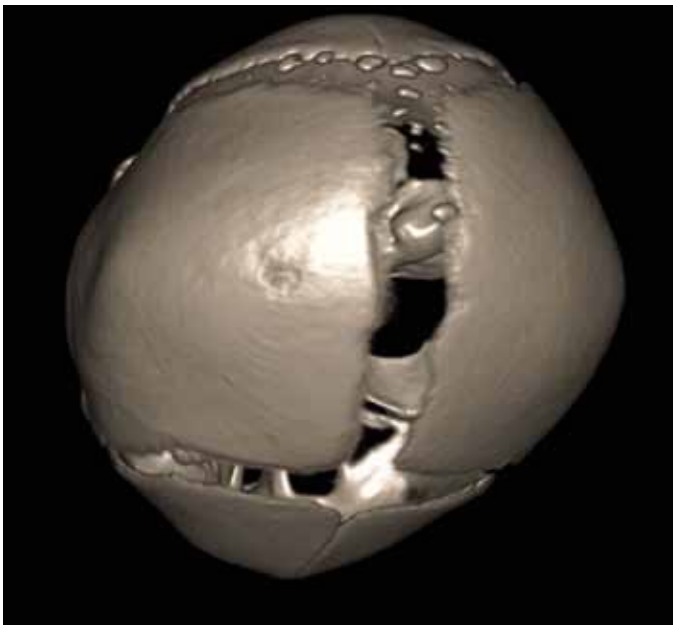


Figure 2: Computed tomography scan showing other sutures including sagittal, coronal and lambdoid open and a closed metopic suture.

in incidence of metopic synostosis may be attributable to increasing incidence of these risk factors. The primary modality of treatment is surgical repair, performed prior to 1 year of age, which traditionally consists of bifrontal craniotomy, skull contouring, bilateral fronto-orbital advancement and placement of a bone graft to increase the width of fronto-orbital bar. This surgical approach is safe with minimal complication rate, short length of hospital stay and great aesthetic outcome [2]. A newer endoscopic approach with all the acknowledged advantages of a minimally invasive procedure over an open surgery is also emerging [3].

CONCLUSION

Uncorrected synostosis may cause increase in intracranial pressure and progression of craniofacial deformity.

How to cite this article

Gupta N, Josef B. Metopic synostosis. International Journal of Case Reports and Images 2014;5(3):244–246.

doi:10.5348/ijcri-2014-03-483-CL-14

****

Author Contributions

Natasha Gupta – Conception and design, Drafting the article, Revision of the article, Final approval of the version to be published
Josef Blankstein – Analysis and interpretation of data, Revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Lee HQ, Hutson JM, Wray AC, et al. Changing epidemiology of nonsyndromic craniosynostosis and revisiting the risk factors. The Journal of craniofacial surgery 2012 Sep;23(5):1245–1.
2. Engel M, Thiele OC, Mühling J, et al. Trigonocephaly: Results after surgical correction of nonsyndromatic isolated metopic suture synostosis in 54 cases. J Craniomaxillofac Surg 2012 Jun;40(4):347–53.
3. Hinojosa J. Endoscopic-assisted treatment of trigonocephaly. Childs Nerv Syst 2012 Sep;28(9):1381–7.

ABOUT THE AUTHORS

Article citation: Gupta N, Josef B. Metopic synostosis. International Journal of Case Reports and Images 2014;5(3):243–245.

Natasha Gupta is Chief Resident in Department of Obstetrics and Gynecology at Mount Sinai Hospital, Chicago. Her research interests include Gynecologic Cancers. She intends to pursue a fellowship in Gynecologic Oncology after completing her residency.

CLINICAL IMAGES OPEN ACCESS

Anomalous left vertebral artery

Hussein Ahmed Hassan, Caroline Edward Ayad, Tag Eldin Mohamed Ibrahim, Ikhlas Abdelaziz Hassan

CASE REPORT

A 40-year-old Sudanese female was referred to the magnetic resonance imaging (MRI) department presented with upper limbs weakness, urine incompetence and general weakness evaluation. Brain MRI axial and sagittal T1, T2, post contrast series, fluid attenuation inversion recovery (FLAIR), magnetic resonance angiography (MRA) were done.

Images show anomalous left vertebral artery impressing into the cord substance at cervico–medullary junction causes cervico-medullary stenosis, no evidence of cerebral tonsil herniation or syrinx formation, in right parietal sub cortical white matter; two small high signal intensity foci were seen on T2 and FLAIR images suggestive of ischemic foci. Brain MRA confirmed malformation in the left vertebral artery, images of MRA showed the left vertebral artery entering foramen magna with medial tortuous kinking to the medulla oblongata.

Normal MRI scan appearance of the basal ganglia, thalami, internal and external capsule on both sides were detected also the ventricular system and extra axial cerebrospinal fluid (CSF) space are within normal limits, no mass effect or midline shift. Normal configuration of optic chiasm, optic nerves, pituitary gland and its stalk, Normal features of the cerebellar hemispheres, vermis, peduncles and brain stem with no abnormal intensities. Normal MRI features of the cerebello pontine angles and internal auditory canal, no evidence of space occupied lesion (SOL), fresh or sub dural collection was detected (Figure 1).

Hussein Ahmed Hassan[1], Caroline Edward Ayad[1], Tag Eldin Mohamed Ibrahim[2], Ikhlas Abdelaziz Hassan[1]
Affiliations: [1]PhD, Diagnostic Radiologic Technology, Sudan University of Science and Technology, Radiology Department, College of Medical Radiological Science, Khartoum, Sudan; [2]MD, Consultant Radiologist, Radiology Department, Modern Medical Center, Khartoum, Sudan.
Corresponding Author: Caroline Edward Ayad, Sudan University of Science and Technology, College of Medical Radiological Science, Khartoum, Sudan, P.O. Box 1908; Ph: +249 0922044764, Email: carolineayad@yahoo.com

Received: 14 June 2013
Accepted: 11 July 2013
Published: 01 March 2014

DISCUSSION

Anatomical variation is defined as the normal flexibility in the topography and morphology of body structures [1]. Anomalous origin of vertebral arteries,

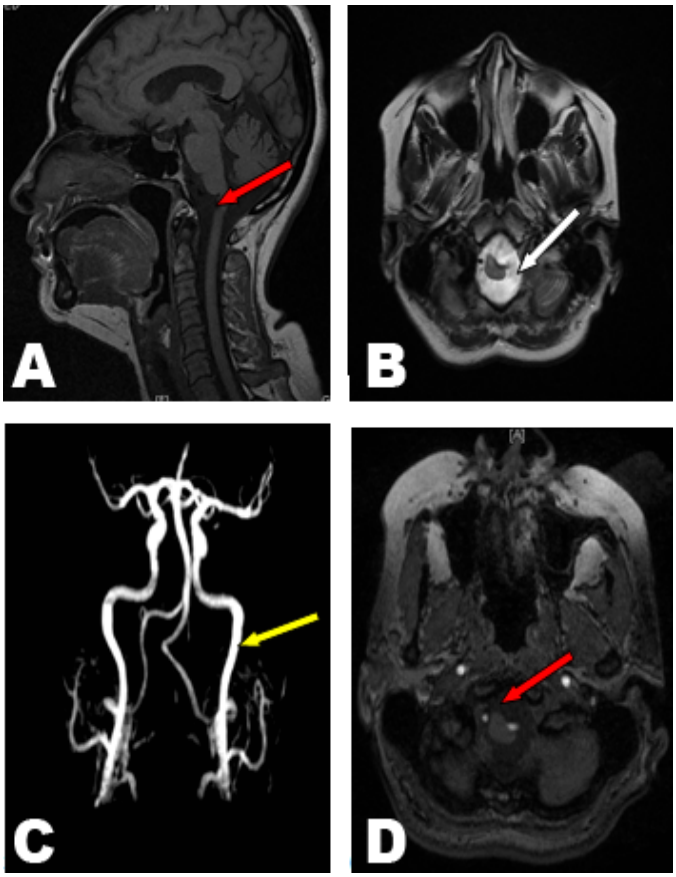


Figure 1: (A) Magnetic resonance imaging sagittal T1 showing spinal cord cervico medullary stenosis (red arrow) due to anomalous vertebral artery, (B) Axial T2 showing cervico medullary sub arachnoid space enlargement, (C) MRA showing kinking left vertebral artery (yellow arrow), (D) Time of flight axial image showing the anomalous vertebral artery (red arrow).

is of relation with the embryologic development of the aortic arch and the brachiocephalic arteries [2].

Anomalous origin of the vertebral artery is rare and is mostly seen in the left vertebral artery which originates directly from the aortic arch between the left common carotid artery and the left subclavian artery [3], with a reported prevalence of 2.4–5.8% in a large autopsy series [4].

Also anomalous origins of both vertebral arteries are uncommon. There are only unreliable cases in the radiology literature which discuss the conventional angiographic findings [5]. The advances in the imaging modalities in recent years have required more accurate knowledge and a greater understanding of the normal anatomy and variation and their clinical importance [6]. Studies were done to ensure the incidence of anomalous origin of the left vertebral artery citied in the radiology literature, and suggest that the current increased frequency of cross-sectional imaging; could elevate the observed incidence of this anomaly in practice [7]. With modern multi-detector computed tomography technology, supra aortic arteries can be well delineated with excellent image quality. Reporting of anomalous origin of the left vertebral artery is important; as the anomaly has significant clinical and surgical implications during endovascular treatment of aortic arch injuries and during angioplasty and stent procedures in conditions [8].

Diagnostic cerebro vascular imaging can be achieved either by catheterization during angiography or by computed tomography angiography (CTA), MRA or Doppler sonography [9]. A thorough identification of anomalous origin of vertebral artery is paramount when performing both diagnostic and interventional angiography. If the vertebral arteries are not identified in their normal position, this can be misinterpreted as the vessel being congenitally absent. This information is important for endovascular or cardiothoracic surgeries in head and neck regions [10].

Anomalous origin of the vertebral artery did not result in clinical symptoms [11, 12], but In some cases, patients may complain of symptoms of dizziness [13]. Anomalous origin effects hemodynamic and may lead to intracerebral malformation [10]. The knowledge of a potential vertebral artery origin variant appears to be obligatory for planning vascular surgery and endovascular intervention [14].

CONCLUSION

The true value of detecting anomalous origins of vertebral artery is the diagnostic gain prior to the surgery of supra aortic arteries. Using magnetic resonance imaging grants an excellent diagnostic value, as it is noninvasive with expansive imaging appearance of both vessels and soft tissues, it has been anticipated as diagnostic imaging modality for detection of anomalous vertebral artery.

How to cite this article

Hassan HA, Ayad CE, Ibrahim TEM, Hassan IA. Anomalous left vertebral artery. International Journal of Case Reports and Images 2014;5(3):247–249.

doi:10.5348/ijcri-2014-03-484-CL-15

Author Contributions

Hussein Ahmed Hassan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Caroline Edward Ayad – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Tag Eldin Mohamed Ibrahim – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ikhlas Abdelaziz Hassan – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Satti SR, Cerniglia CA, Koenigsberg RA. Cervical vertebral artery variations: An anatomic study. AJNR Am J Neuroradiol 2007 May;28(5):976–80.
2. Moore KL. The Developing Human. 3rd ed. Philadelphia: Saunders 1982.
3. Lemke AJ, Benndorf G, Liebig T, Felix R. Anomalous origin of the right vertebral artery: Review of the literature and case report of right vertebral artery origin distal to the left subclavian artery. AJNR Am J Neuroradiol 1999 Aug;20(7):1318–21.

4. Albayram S, Gailloud P, Wasserman BA. Bilateral arch origin of the vertebral arteries. AJNR Am J Neuroradiol 2002 Mar;23(3):455–8.
5. Lemke AJ, Benndorf G, Liebig T, Felix R. Anomalous origin of the right vertebral artery: Review of the literature and case report of right vertebral artery origin distal to the left subclavian artery. AJNR Am J Neuroradiol 1999 Aug;20(7):1318–21.
6. Bergman RA, Afifi AF, Miyauchi R, "Opus II: Cardiovascular system vertebral artery variations," in Illustrated Encyclopedia of Human Anatomic Variation, Bergman RA, Afifi AF, Miyauchi R, Eds.
7. Braham van der Merwe, Christelle Ackermann, Shaun Scheepers, Sulaiman Moosa S Afr Is anomalous origin of the left vertebral artery indeed a rare finding? J Rad 2012;16(4):144–6.
8. Ka-Tak W, Lam WW, Yu SC. MDCT of an aberrant right subclavian artery and of bilateral vertebral arteries with anomalous origins. AJR Am J Roentgenol 2007 Mar;188(3):W274–5.
9. Albayram S, Gailloud P, Wasserman BA. Bilateral arch origin of the vertebral arteries. AJNR Am J Neuroradiol 2002 Mar;23(3):455–8.
10. Sadaf Nasir, Manzar Hussain, Sikandar Ali Khan, Muhammad Ayub Mansoor, Salman Sharif. Anomalous origin of right vertebral artery from right external carotid artery.Journal of the College of Physicians and Surgeons Pakistan 2010 Mar;20(3):208–10.
11. Ka-Tak W, Lam WW, Yu SC. MDCT of an aberrant right subclavian artery and of bilateral vertebral arteries with anomalous origins. AJR Am J Roentgenol 2007 Mar;188(3):W274–5.
12. Lemke AJ, Benndorf G, Liebig T, Felix R. Anomalous origin of the right vertebral artery: Review of the literature and case report of right vertebral artery origin distal to the left subclavian artery. AJNR Am J Neuroradiol 1999 Aug;20(7):1318–21.
13. Nishijima M, Harada J, Nogami K, Endo S, Takaku A. Operative correction of kinking and coiling at the origin of vertebral artery and stellate ganglionectomy in patients with severe vertigo and dizziness. No Shinkei Geka 1989 Mar;17(3):255–61. [Article in Japanese].
14. Albayram S, Gailloud P, Wasserman BA. Bilateral arch origin of the vertebral arteries. AJNR Am J Neuroradiol 2002 Mar;23(3):455–8.