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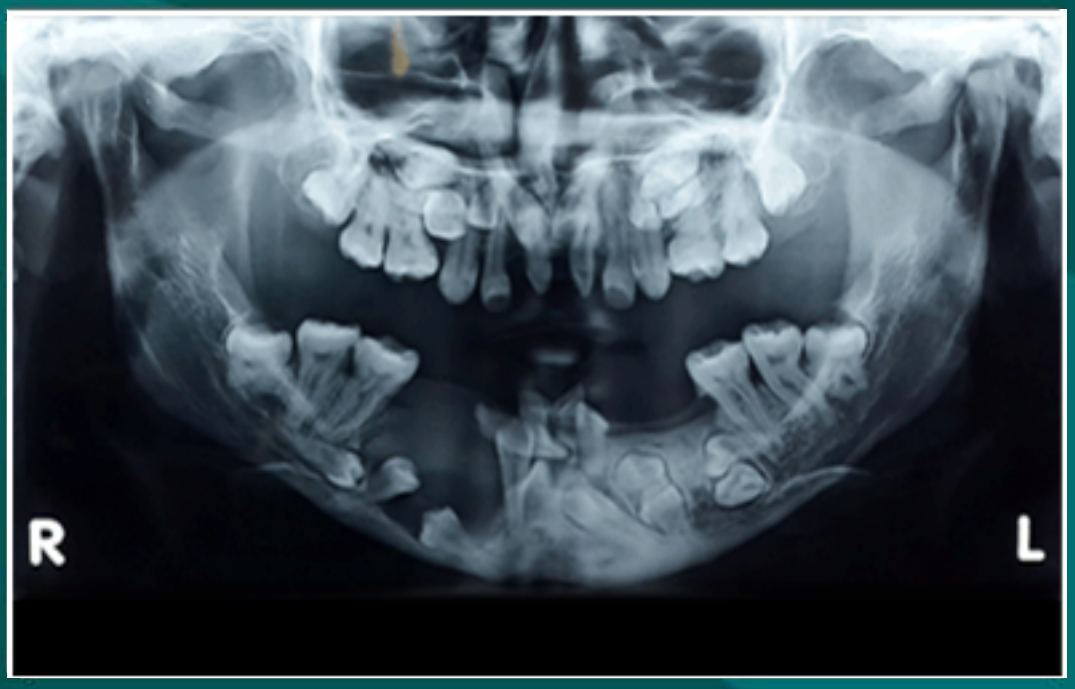

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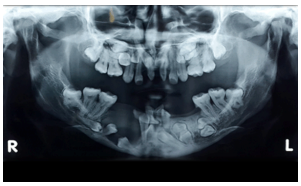

Oral pantomogram showing diffuse radiolucency at the site of interest and multiple impacted teeth.

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CASE SERIES

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Guidewire technique: Removal of a doorknob in tourniquet syndrome

Phee-Kheng Cheah, Xiang-Yun Yang, Azlan Kamalludin, Thian-Lian Soo

ABSTRACT

Introduction: Tourniquet syndrome caused by a constricting band is common, and can be removed either by intact removal or by division. Finding a doorknob as the constrictor is rare and challenging to manage due to its bulky size, larger area of contact with the skin surface, and difficult access to the constricting site. The optimal removal technique has not yet been established. Case Series: This case series explored two different techniques—cutting with a metal saw, and intact removal using guidewire technique which uses the guidewire from a central venous catheter. The first case was an 8-year-old boy who presented with his right index finger stuck to a doorknob, and the team utilized a metal saw to cut open the doorknob in order to release the finger. The second case was a 10-year-old boy also with his right index finger stuck in the doorknob. The team used the guidewire technique and successfully removed the doorknob intact. Conclusion: While both techniques resulted in successful removal with minimal soft tissue injury, the former was more tedious, traumatizing, time-consuming and required technical help that is not immediately available. This can potentially cause delay in removal of the constrictor. We believe that intact removal using our technique is superior to cutting the doorknob because it is simple, cost-effective and ultimately a low risk and time-saving procedure.

Keywords: Constricting band, Doorknob, Guidewire technique, Tourniquet syndrome

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INTRODUCTION

Constricting bands around body appendages is not an uncommon presentation to the emergency department. The constriction can produce a clinical condition known as the tourniquet syndrome, which causes neurovascular compromise to the tissues distally, leading to ischemia and necrosis. This will result in long-term loss of function and sensitivity, or worse, amputation of the affected appendage [1]. Hence, timely removal of the constricting band is of the utmost importance to prevent this complication.

Generally, the methods available can be divided into intact removal, or removal by division [1, 2]. The former can further be classified into five distinct groups: the winding technique, compression technique, the caterpillar technique, twin-thread technique, and glove technique [3–8]. Intact removal techniques work by first reducing the edema distal to the constricting band via different techniques of compression, then followed by distal advancement of the band over the affected area to remove it. On the other hand, removal by division uses cutting devices and will depend on the material and size of the constricting band [2].

Methods to remove a ring from the finger have been extensively described in literature [1–8]. However, no literature is available with regards to methods of removing a doorknob from the finger. Indeed, finding a doorknob as a constricting band around the finger is quite rare. The point of insertion of the finger is into the doorknob hole and through the shank (Figure 1). Metal doorknobs are mostly made out of brass, which is an alloy composed of varying percentages of copper and zinc [9]. Brass is popularly used because of its strength, resistance to rust and formability, hence such durability poses a challenge in its removal.

In this case series, we present two patients who came to emergency department with doorknobs as the constricting band around their fingers.

CASE SERIES

Case 1

An eight-year-old boy presented with his right index finger stuck to a doorknob. The child was playing with an unused doorknob still attached to a door in a house undergoing renovation. Attempts to remove the doorknob by the family members at home had failed. The finger was swollen with the doorknob tightly stuck above the proximal interphalangeal joint. Capillary refill was less than two seconds. There was no cyanosis, pallor or necrotic tissue noted. The child could still move the finger and denied any altered sensation distally.

A home improvement personnel was called in for assistance as we were not familiar with the structure of the doorknob. The patient was administered with a digital block over the right index finger, and removal was attempted using lubricant. It was unsuccessful. We decided to proceed with removal by division as the child was able to cooperate. No sedation was required.

The doorknob structure had to be dismantled into components and had to be sawn into multiple segments, as the hard metal of the doorknob prevented bending of the structure to release the finger. The smooth and rounded surface of the doorknob also proved to be a challenge, particularly to balance the metal saw on it. A few assistants were needed to hold the doorknob steady during the sawing process (Figure 2).

Three cuts were made on the knob, and the segments were bent to overlap each other to enable the release of the finger (Figure 3). The patient tolerated the procedure well and only sustained minimal swelling and a few superficial lacerations around the area due to the prior manipulation attempts to free the finger (Figure 4). There was no reduction in the range of movement. The patient was discharged with normal saline dressing and oral analgesia.

Case 2

A 10-year-old boy was brought into emergency department with his right index finger stuck to a doorknob (Figure 1). The child was playing with a defective door at home. Multiple attempts by the family

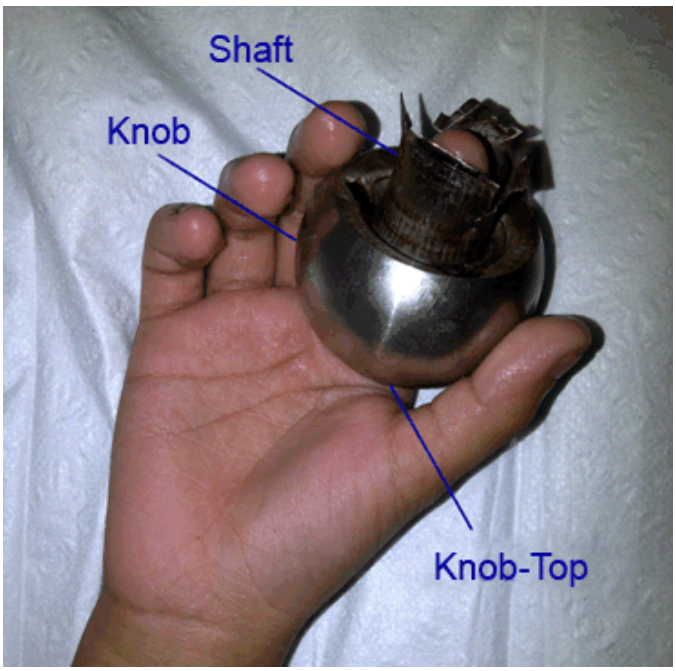

Figure 1: Doorknob as a constricting band around a finger. The finger passes through the doorknob hole through to the shank.

Figure 2: Pieces of the doorknob bent to release the fingers.

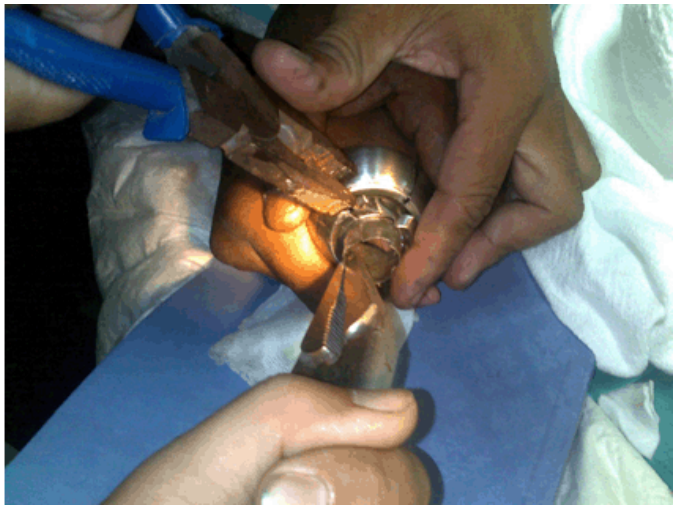

Figure 3: The guidewire was hooked through the doorknob.

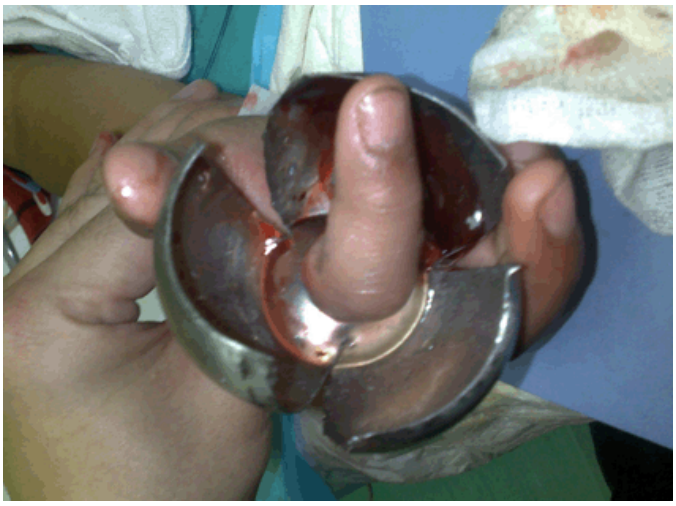

Figure 4: The guidewire was used to maintain a constant traction on the doorknob with one hand stabilizing the doorknob.

members to free the finger from the doorknob were unsuccessful.

In the emergency department, the child complained of severe pain over the affected finger. He was apprehensive of any inspection or manipulation of the finger. The entire length of his finger was covered by the doorknob with minimal swelling noted distally. Capillary refill time was less than two seconds. The division method would be risky in this case as the child was unwilling to cooperate. It was also noted that the doorknob actually has sharp edges near the contact area, which can cause a string, suture or rubber band to snap or tear off during manipulation using the intact removal method. A string with higher tensile strength is more feasible to achieve removal, and therefore, a metal guidewire from a triple lumen central venous catheter was chosen as it is easily available in the emergency department (Figure 5).

Wrist block was administered in this case. The child was also sedated with oral chloral hydrate prior to the procedure.

The guidewire was passed through the constrictor from one end to the other in order to form a "hooking" mechanism through the doorknob (Figure 6). Both ends of the guidewire were held in one hand and constant gentle traction was maintained in order to pull the doorknob away from the affected finger, while the other hand stabilizes the spherical doorknob. The child's hand was kept in position by an assistant (Figure 7). No lubricant was required. The doorknob was then successfully removed within three minutes.

Evaluation post procedure revealed a mildly edematous finger with a circumferential abrasion wound at the site of entry. There was no reduction in finger movement, and no fracture was detected clinically. Simple wound dressing was done and the patient was discharged with oral analgesia. A follow-up via phone call a week later reported full functional recovery of his finger.

Figure 5: Assistance was needed to stabilize the doorknob during the sawing process.

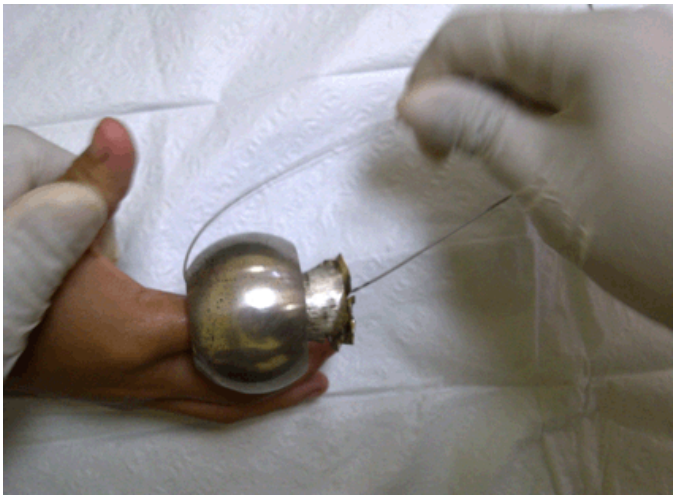

Figure 6: Three cuts were made on the doorknob and the pieces were bent in order to release the finger.

DISCUSSION

In this case series, both intact removal technique and removal by division were used in removing the constricting doorknob. Both techniques resulted in successful removal, with minimal soft tissue injury sustained from the previous removal attempts (Figure 8).

The method employed in Case 1, which was removal by division using a metal saw, has several limitations which has to be addressed. Using a saw would require extra time in the preparation process, such as ensuring the availability of trained personnel in using it, and also the personal protective gears for the staff and the patient. This leads to a potential risk in causing a delay in the removal of the constrictor, which can lead to irreversible complications to the finger.

The doorknob's bulky and rounded physical structure overlies not only the narrowed area of the doorknob hole, but also a large portion of the finger. The wide, smoother and global surface of the doorknob is hard to stabilize especially if the cutting technique is employed. As a result, it is highly likely that the sawing equipment might slip off the cutting surface and cause injury to the patient and also the operator. This poses an extra hazard to the patient and personnel involved.

Significant heat could also be generated while the patient's skin is in contact with the constricting band, causing thermal injuries. Peay et al. found that significant heat was generated by just 5–10 seconds of grinding [10]. In our case, the doorknob is a relatively large sized constricting band as compared to a ring which is the usual mentioned in literature. A longer time would be required to cut the doorknob sufficiently for a finger release. The patient will also have to be cooperative and actively giving feedback regarding the sensation of the affected site, whether it is burning hot or if there is any worsening of the constriction felt. Therefore, this method might not be suitable in patients who are fretful and unable to obey commands, especially children who are already traumatized by the event.

In Case 2, we used a guidewire from a triple lumen central venous catheter to maintain the force of pulling on the doorknob from the finger, which is a modified version of the string technique. Whilst maintaining a constant force of traction, the doorknob itself compresses the immediate point of contact, hence enabling it to progress further distally. In comparison to a string or a rubber band, a guidewire has higher tensile strength and does not snap easily, especially for a large constrictor such as in this case. The guidewire also provides a smooth surface that prevents excessive skin abrasion during manipulation of the constrictor, hence reducing the chances of iatrogenic injury. No lubrication or very minimal lubrication was required in this case, as the constant force maintained over the guidewire enabled the doorknob to surmount the main obstruction, which is the PIP joint.

Such guidewires are also readily available in the emergency department where central vascular catheterization sets are available, as compared to looking for a suitable saw to cut the constricting band open. However, if the finger were to be more edematous, using multiple techniques simultaneously (compression,

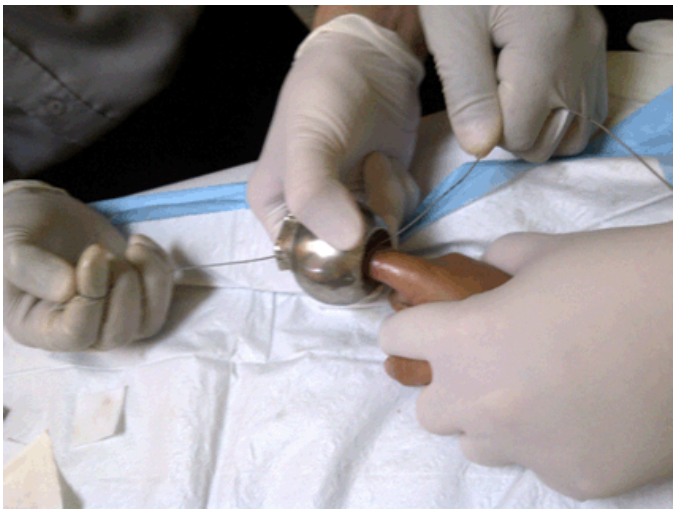

Figure 7: The guidewire from a triple-lumen central venous catheter.

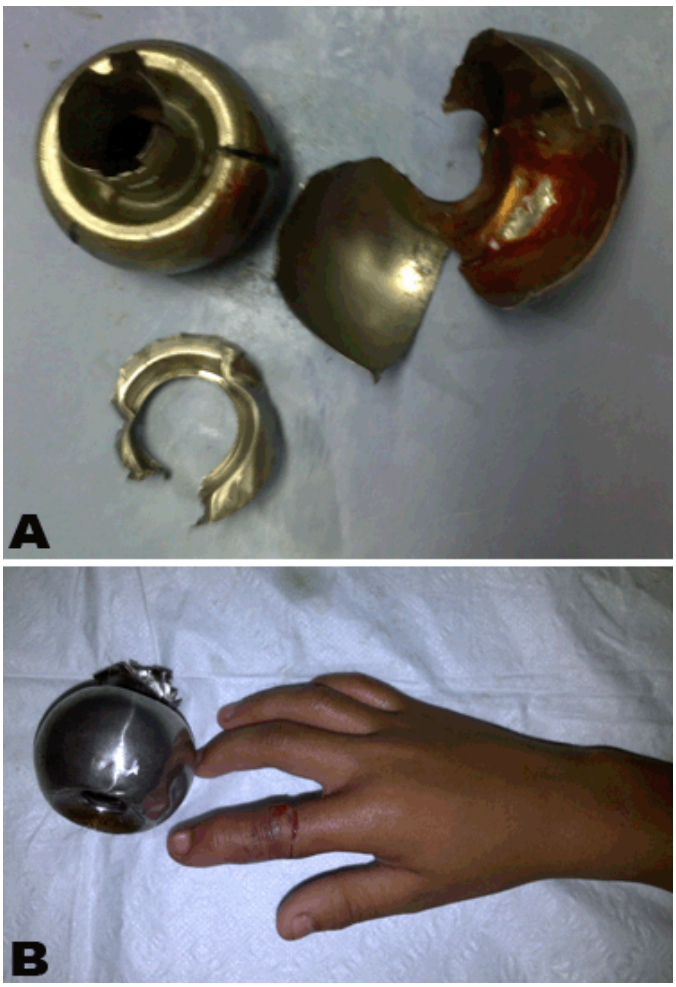

Figure 8: Comparison post removal of the doorknob with both techniques. (A) The dismantled doorknob after cutting (Case 1), and (B) The intact doorknob with the finger post guidewire removal (Case 2).

lubricate and traction) would often increase the chances of success.

Wrist block was administered in this case. The child was apprehensive of any approach by the healthcare staff thus stabilization of the wrist joint would be easier and safer as compared to stabilizing the finger. The bulky size of the doorknob which has covered up most of the finger also leaves minimal surface area for digital block access. Digital block could increase the swelling over the affected site, worsening the tourniquet effect. In this patient, the wrist block is probably a better option of local analgesia as compared to digital block.

Although both techniques successfully removed the doorknob, our guidewire technique using a metal guidewire is simpler and time-saving, with a lower-risk of manipulation-related injury. The guidewire used is a readily available in the most emergency departments, and the technique is easily mastered by novice operators, without the risk of injuring medical personnel themselves. With the absence of loud noises and the threatening appearance of cutting machines, this guidewire technique can be safely applied to most patients, especially the pediatric group. However, in both our cases, the patients only presented with mild edema distal to the constrictor with no open wound or fracture and no signs of ischemia. This technique may only be applicable to such patients.

Kalkan et al. proposed an algorithm in removing a stuck ring in the emergency department [2]. This algorithm can serve as a guide in deciding whether the patient is suitable to undergo cutting or non-cutting technique. However, there is still no review or algorithm proposed for cases of a bulky constrictor, such as a doorknob.

CONCLUSION

It is challenging using either the cutting or non-cutting technique when faced with a doorknob as a constricting band. We believe our guidewire technique will be useful in the removal of large constricting bands, as it is not only readily available in the emergency department, but also poses less hazard to the patient and health staff. This technique can be used as a first attempt before other more complicated methods are undertaken.

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Author Contributions
Phee-Kheng Cheah – 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
Xiang-Yun Yang – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Azlan Kamalludin – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Thian-Lian Soo – 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.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

SGLT2 inhibitor associated diabetic ketoacidosis

Evdoxia Mitsiou, Charalampos Mandros, Kalliopi Kotsa, Frangiskos Koulis, Charalampos Christofidis, Sofia Georgiadi, Theodolinta Testa, Alexandros Anastasiou, Evgenia Efthymiou, Evangelos Potolidis

ABSTRACT

Empagliflozin is a representative of SGLT2 inhibitors, which is used for the treatment of type 2 diabetes mellitus. Common adverse reactions are hypoglycemia and urinary tract infections. We reported a case of 76-year-old female, receiving empagliflozin and being admitted to the hospital because of diabetic ketoacidosis.

Keywords: Diabetes type 2, Empagliflozin, Ketoacidosis

How to cite this article

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INTRODUCTION

SGLT2 inhibitors are sodium-glucose cotransporter 2 inhibitors (SGLT2) in the proximal renal tubules that reduce reabsorption of filtered glucose from the tubular lumen and lower the renal threshold for glucose (RTG). Therefore, SGLT2 is the main site of filtered glucose reabsorption. By inhibiting SGLT2, urine glucose excretion increases and plasma glucose concentration reduces [1]. The SGLT2 inhibitors are generally weak glucose-lowering agents, similar to efficacy to the DPP-4 inhibitors. Empagliflozin is one of the representatives of this category of glucose-lowering agents, which is used for the treatment of type 2 diabetes mellitus, usually in combination with metformin [2] or insulin [3], as an adjunct to exercise and diet to improve glycemic control. Common adverse reactions are hypoglycemia and urinary tract infections [4]. Since approval of the first-in-class drug in 2013, data have emerged suggesting that these drugs may increase the risk of diabetic ketoacidosis [5]. Moreover, in May 2015, the Food and Drug Association issued a warning that SGLT2 inhibitors can increase the incidence of diabetic ketoacidosis [6]. It also identified potential triggering factors such as illness, reduced food and fluid intake, reduced insulin doses, and history of alcohol intake. Our case will be one of the few cases of diabetic ketoacidosis reported in a patient with type 2 diabetes mellitus [7].

CASE REPORT

A 76-year-old female was admitted to our hospital because of increased fatigue and weakness during the last 10 days. At the same time, she started receiving medication for type 2 diabetes mellitus, which concluded metformin 850 (1x1) and empagliflozin 10 (1x1). She did

not receive any other medication. She reported just one fever wave up to 38°C, three days before the admission.

On examination she appeared confused and she had tachypnea. She did not show any signs of infection. The temperature was 36.9°C, the blood pressure 90/60 mmHg, the pulse 100 beats per minute, the respiratory rate 30 breaths per minute and the oxygen saturation 96%, while she was breathing ambient air. Blood tests revealed elevated hematocrit (52.8%), white blood cells (15200/ml) and CRP (100 mg/dl), whereas kidney function was normal (creatinine 1.0 mg/dl, urea 50 mg/dl). Serum electrolytes were also normal (serum potassium 4.9 mEq/l, serum sodium 137 mEq/l), serum calcium was 90 mEq/l and serum glucose was 202 mg/dl. Urinalysis revealed 4+ glucose, 1+ albumin, 4+ ketones by dipstick. Blood gas test revealed severe acidosis with pH 7.06, pCO_2 24 mmHg, pO_2 129 mmHg, lac 1.0 mmol/l, HCO_3 9.6 mmol/l, base excess 20.7 mmol/l, SO_2 99%. Urine culture was negative. Anion gap was calculated and appeared to be elevated at 36.4. HbA1c was 10.7%.

The patient was treated as having diabetic ketoacidosis due to type 1 diabetes mellitus. She aggressively received intravenous fluids and insulin and she was covered empirically with a broad spectrum antibiotic because of elevated CRP, even though she did not have any fever during her hospitalization. Urinalysis continued to show elevated glucose levels and ketones, until the day of discharge, on day-8. Blood gas tests stopped to show acidosis on the third day, but the base deficit remained high until the seventh day (HCO_3 16.9 mmol/l, base excess 9.0 mmol/l). The test was normal on the day of discharge.

DISCUSSION

Empagliflozin is an SGLT2 inhibitor usually used in combination with metformin or insulin in order to further reduce serum glucose levels. Lately, apart from the common adverse reactions of hypoglycemia and urinary tract infections, cases of diabetic ketoacidosis come to light. Food and Drug administration issued a special warning about this adverse reaction, which include not only empagliflozin but also all drugs of this category. Based on the pharmacological characteristics of this type of drugs and the physiology of SGLT2, several possible mechanisms could be suggested for the development of diabetic ketoacidosis. Inhibition of SGLT2 causes a rapid increase in urinary volume excretion, which lasts more than 24 hours [8]. Also, the decrease in plasma glucose levels, that is caused by these drugs [9], lead to a decrease in plasma insulin levels and a significant increase in plasma glucagon concentrations, because of a diminished paracrine inhibition by insulin and a decreased SGLT2-mediated glucose transport into α-cells [10]. Therefore, SGLT2 inhibitors seem to be associated with euglycemic diabetic ketoacidosis, perhaps as a consequence of their non-insulin dependent glucose clearance, hyperglucagonemia and volume depletion [8].

CONCLUSION

Patients who are treated with SGLT2 inhibitors should be closely monitored for this adverse reaction and clinicians should also be aware of it. Further research should be done in order to minimize the risk of ketoacidosis due to SGLT2 inhibitors.

****

Author Contributions

Evdoxia Mitsiou – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Charalampos Mandros – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Kalliopi Kotsa – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Charalampos Christofidis – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Evgenia Efthymiou – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Evangelos Potolidis – 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.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Classic case of cleidocranial dysplasia with an infected mandibular cyst

Madhu P. Sivan, Jayakumar K., Jayalakshmi P. S., N. Sruthi

ABSTRACT

Patients with cleidocranial dysplasia commonly present with significant dental problems, such as retention of multiple deciduous teeth, impaction or delayed eruption of teeth and the presence of multiple supernumerary teeth. In fact, the presence of multiple supernumerary teeth is one among the pathognomonic triad for this condition; the other two being partial or complete absence of clavicles, and open sagittal sutures and fontanelles. This case report describes a patient having typical features of cleidocranial dysplasia who had to undergo surgical enucleation and debridement for an infected mandibular cyst. Factors dictating the treatment plan and the current trends in rehabilitation for such a patient have been discussed from a dental surgeon's perspective acknowledging the shift in management paradigm for this condition over the years.

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INTRODUCTION

Cleidocranial dysplasia, is a congenital disorder of bone formation with prominent manifestations being clavicular hypoplasia/agenesis with a narrow thorax, delayed ossification of skull bones, excessively large fontanelles and delayed closing of sutures [1]. One of the earliest description of this condition was given by Martin in 1765 [2]. Marie and Sainton in 1897 used the term cleidocranial dysostosis [3]. It has since been known as cleidocranial dysplasia in recognition of its underlying pathology being more of a generalized skeletal dysplastic condition. One in a million people and both genders are rather equally affected [1].

Genetic mutation as an etiologic factor for this condition was proposed by Kalliala et al. with 20–40% caused by spontaneous gene mutation [4]. Cleidocranial dysplasia follows autosomal dominant pattern of inheritance with high penetrance and variable degree of expression. The locus of the gene is either the long or short arm of chromosome 6p21 [5]. Zheng et al. reported that humans with cleidocranial dysplasia have altered

endochondral ossification due to perturbed RUNX2 regulation of hypertrophic chondrocytes. This gene is essential for osteoblast and dental cell differentiation, and thus for normal bone and tooth formation [6]. Recent studies have indicated that RUNX2 serves as a master gene regulating osteoblast-specific gene expression. The gene is expressed in the cells of osteoblast lineage only, and its expression is regulated by calciotropic agents. In odontogenesis, RUNX2 regulates key epithelial mesenchymal interactions that control the progress of morphogenesis and histodifferentiation of the epithelial enamel organ [7]. Still more recently, CCAAT/enhancer-binding protein beta (Cebpb) which is a key factor of Runx2 expression has been proposed as an additional aetiological factor for cleidocranial dysplasia [8].

Herein, we present a case report of a patient with classical features of cleidocranial dysplasia with symptomatic infected mandibular cyst probably secondary to multiple impacted teeth and its management with surgical enucleation and debridement.

CASE REPORT

A 26-year-old male presented with chief complaint of tender swelling gradually increasing in size in lower right jaw region of 4–6 months duration. Intraoral examination showed a fluctuant swelling approximately 3x2 cm in size in mandibular right canine-premolar region tender to palpation (Figure 1). An active site of pus discharge along the crevices of adjacent molar teeth was seen. Multiple teeth were missing and generalized periodontitis owing to heavy calculus build-up was observed around the few teeth present in the oral cavity. Maxillary incisors were malformed. A total of 16 teeth were present in the oral cavity. A deep and high arched palate was noted.

General physical examination demonstrated a thin build, short stature, slurred speech, narrow thorax and shrugged shoulders which were easily apposable. Prominent forehead with hypertelorism, a depressed nasal bridge, and mid-facial hypoplasia with relative mandibular prognathism were noted. Frontal, parietal and occipital bossing was present, giving the skull a large globular and brachycephalic shape (Figure 2). The panoramic radiograph on examination revealed a diffuse radiolucency in the area of interest suggestive of excessive bone loss due to the cystic lesion along with multiple impacted teeth (around 24 in number) and non-prominent gonial angle bilaterally (Figure 3). Posteroanterior cephalogram suggested non-ossified cranial sutures (Figure 4). Additionally, his elder brother, showed similar external facial and general appearance (Figure 5). Subsequently, a surgical treatment plan involving surgical enucleation and debridement of affected tissue and disimpaction of multiple involved impacted teeth in the area of concern under general anesthesia with supportive systemic antibiotics and analgesics was planned and executed. A difficult airway was anticipated during general anesthesia. Following enucleation, a total of nine teeth in and around the area of interest were disimpacted, tissue debrided and primary closure was achieved. Post-surgical healing was uneventful and satisfactory (Figure 6). Histopathological examination was confirmatory for dentigerous cyst. The patient is under regular follow-up and Prosthodontic rehabilitation is planned in the coming months once the presence of adequate bone support is confirmed clinically and radiographically (Figure 7).

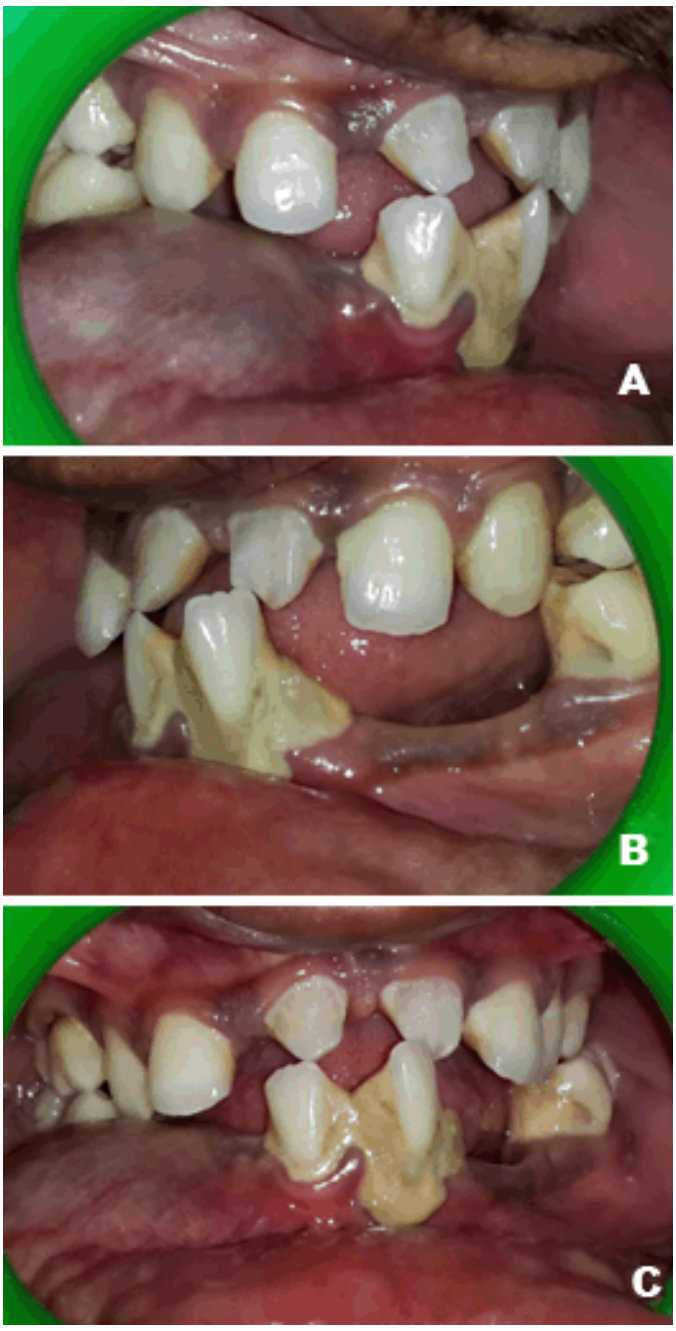

Figure 1: Intraoral views: (A) Right lateral, (B) Left lateral, (C) Frontal.

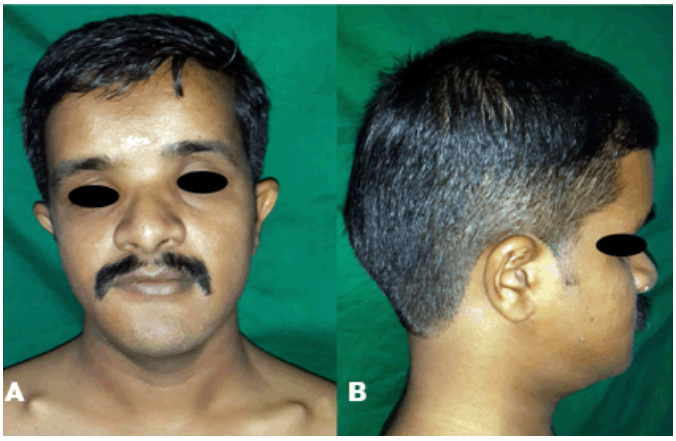

Figure 2: Extraoral views: (A) Frontal, and (B) Parietal.

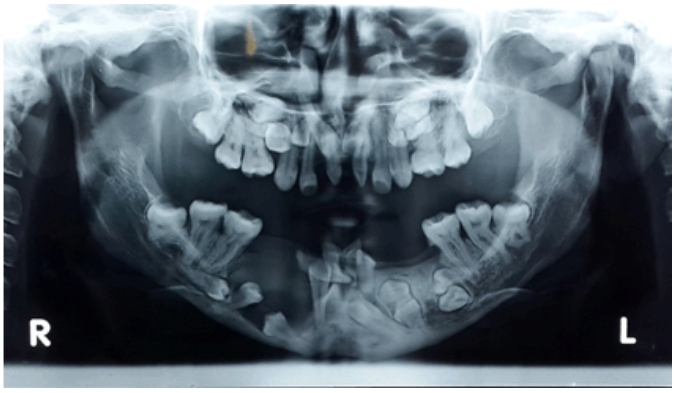

Figure 3: Oral pantomogram showing diffuse radiolucency at the site of interest and multiple impacted teeth.

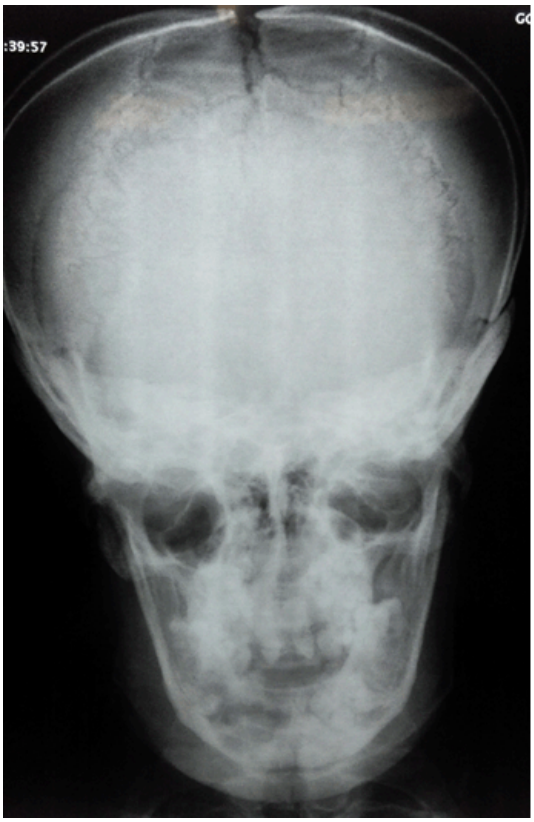

Figure 4: Posteroanterior cephalogram demonstrating non ossified suture lines.

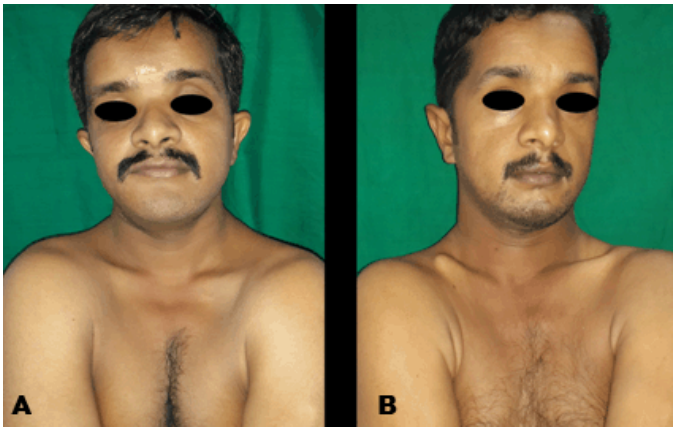

Figure 5: (A) Patient, and (B) Patient's brother, both showing hypermobility of the shoulder.

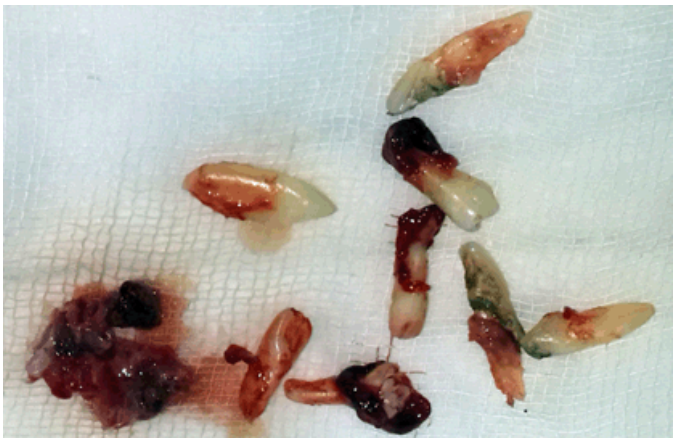

Figure 6: Disimpacted teeth and the enucleated cyst.

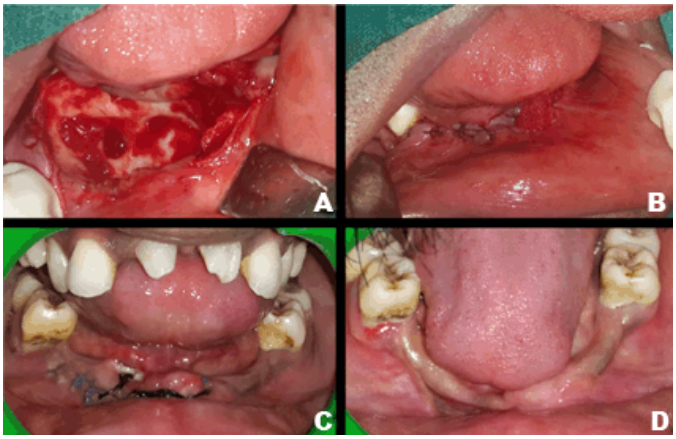

Figure 7: Intraoral views (A) Intraoperative, (B) Immediate postoperative, (C) One week postoperative, and (D) One month postoperative.

DISCUSSION

Cleidocranial dysplasia shows a wide degree of varying manifestations clinically which may or may not affect the quality of life of the patient. They typically have a shorter

stature, brachycephaly with frontal, parietal and occipital bossing, and increased intraorbital distance, wide and flat bridge of the nose, underdeveloped maxilla and relative mandibular prognathism. Hypoplasia of masseter muscle due to discontinuity of zygomatic arches may result in compensatory hyperfunctioning of temporalis muscles. Consequently, anterior border of the mandibular ramus is usually parallel to the posterior border, and the coronoid process is directed upwards and backwards. There is typical delayed closure of the fontanel. Clavicular hypoplasia/agenesis and unusually formed/placed muscles attachments to the clavicles may be seen. Further, unusual positioning of hip joint, abnormalities of the bones of the spine, fingers and hands have been reported. Occasional abnormalities associated with this condition includes scoliosis, extra ribs, small pelvis, cleft palate, recurrent ear infections(otitis media) related to abnormal formation of palate and/or Eustachian tube dysfunction and respiratory problems. These individuals typically have normal ability to learn [9, 10].

The pathognomonic triad for cleidocranial dysplasia were present in this patient. He reported for treatment only when he started experiencing significant discomfort which is often the case elsewhere. It may be assumed that the slow growing cystic lesion associated with the multiple impacted teeth may have been secondarily infected after substantial erosion of the associated bony structure of the mandible.

Since the expression of the disorder shows significant variation and is rarely expressed to a degree causing significant discomfort or disfigurement to the patient, the condition is often diagnosed incidentally or at a much later age when there is an associated secondary pathology. The timing of diagnosis is important in choosing an appropriate treatment plan and in attaining a successful result.

With significant advances in medicine and dentistry clubbed with increase in awareness, accessibility and affordability to rehabilitation procedures for the patient, there has been a significant shift in management protocol for this condition. Many years ago, minimal intervention to address any symptomatic pathology or no intervention was recommended. Over the next few years, rehabilitation procedures to improve the overall quality of life became more prevalent. In early 20th century, orthodontic-surgical intervention for forced eruption, use of partial dentures etc. gained popularity. Still more recently, dental implants and implant supported dentures have been used for oral rehabilitation. Orthognathic surgeries for correction of facial aesthetics have also gained acceptance.

From a dental practitioner's perspective, elimination of clinical pathology followed by achieving a functional dentition and an overall aesthetically satisfying facial appearance should be the ultimate treatment objectives. Psychological support to the patient and parent is often necessary. Multidisciplinary approach is of prime importance. Affected infants should receive their first dental evaluation by one year of age. Extraction of primary teeth does not hasten the eruption of permanent teeth here. Jensen and Kreiborg have suggested that supernumerary teeth form as a result of activation of remnants of dental lamina left unresorbed during odontogenesis. The contributory role of supernumerary teeth to the arrested eruption of permanent teeth is believed to be secondary to that of diminished bone resorption [9, 10].

In late mixed dentition period and permanent dentition period, combination of orthodontic and maxillofacial surgical approaches are recommended. Following the orthodontic phase, skeletal discrepancies (usually a Class III skeletal malocclusion) are corrected with orthognathic surgeries after completion of growth. Prosthodontic rehabilitation with/without the use of dental implants are carried out further to establish a functional dentition and to fulfil aesthetic requirements.

The cystic lesion here was a dentigerous cyst which was enucleated and confirmed histopathologically. Since the bone loss was significant and extended very close to the lower border of the mandible, we handled the soft tissue and hard tissue with care. However, no graft was required. Instructions were given to the patient for adequate care during postoperative period. We have also delayed the prosthodontic rehabilitation until clinical and radiographic evidence for adequate bone presence is established. The first case of cystic involvement in cleidocranial dysplasia was reported by Oatis et al. in 1975 [11]. Since then very few cases such cases have been documented. This is an interesting aspect when we take into perspective the number and frequency of multiple dental impactions seen in cleidocranial dysplasia patients.

CONCLUSION

Planning treatment for a patient with cleidocranial dysplasia is complicated by a host of factors and is often complicated further due to non-coincident dental and chronological age. The patient's perception of the need for treatment may deviate from that of the treating practitioner. It may be difficult to obtain consent to a treatment involving multiple surgical exposures and forced eruption of teeth. The treatment plan thus requires to be tailored to address the individual's chief complaint taking into consideration the extent of motivation of the patient and a host of other clinical factors as discussed above. With proper anticipatory guidance, people with cleidocranial dysplasia usually lead healthy and productive lives.

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Author Contributions

Madhu P. Sivan – 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
Jayakumar K. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Jayalakshmi P. S. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Sruthi N. – Substantial contributions to conception and design, Acquisition of data, 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.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Duplication cyst in the sigmoid colon mimicking a submucosal tumor: A case report

Pyong Wha Choi, Mee Joo

ABSTRACT

Introduction: A duplication cyst in the alimentary tract is a rare congenital anomaly that most often occurs in the small bowel and rarely occurs in the colon. Most cases in the small bowel are diagnosed at childhood due to complications or symptoms caused by the duplication cyst. However, duplication cysts in the colon are often asymptomatic, despite resulting in rare complications, such as obstruction, infection, and inflammation. Thus, duplication cysts in the colon can be detected incidentally through imaging studies or during a colonoscopy, causing them to possibly be misdiagnosed as a submucosal tumor of the colon. Case Report: We present a case of a duplication cyst of the sigmoid colon, which was preoperatively misdiagnosed as a submucosal tumor. A 64-year-old male was referred for treatment of the submucosal mass of the sigmoid colon. A large round submucosal mass was found in the sigmoid colon during colonoscopy, and computed tomography revealed an approximately 4 cm homogeneous enhancing mass of the sigmoid colon without evidence of lymphadenopathy. With a presumed diagnosis of submucosal tumor, laparoscopic anterior resection was performed. However, histologic examination showed that the mass was consistent with a duplication cyst in the sigmoid colon. Conclusion: This case represents an unusual case of a duplication cyst in the sigmoid colon, which could not be preoperatively diagnosed. Although its preoperative diagnosis is difficult given its rarity, a duplication cyst can be considered one of the differential diagnoses of a submucosal colon mass.

Keywords: Colon, Cyst, Duplication

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INTRODUCTION

A duplication cyst of the alimentary tract is a rare congenital anomaly that is defined by its cystic structure that exhibits a common muscular wall and bloody supply with the nearby alimentary tract [1]. While these cysts can develop along the entirety of the gastrointestinal tract, the ileum is the most common site. Meanwhile, the development of a duplication cyst in the colon is rare [2–4]. Duplication cysts can be detected in any age, but pediatric patients comprise most of the cases because of the symptoms caused by the cysts [2–4]. In adults, duplications cysts are often diagnosed by chance during

an imaging study or colonoscopy. Herein, we present a case of a 64-year-old male with a duplication cyst in the sigmoid colon, which was preoperatively diagnosed as a large submucosal tumor.

CASE REPORT

A 64-year-old male was referred to the department of surgery of Inje University, College of Medicine, Ilsan Paik Hospital for the treatment of a submucosal mass in the sigmoid colon. He had undergone a colonoscopy one week ago at a local clinic to screen for colorectal cancer and was referred to the department of gastroenterology for further evaluation and management. He had no past medical history and did not complain of any symptoms related to the mass. Upon admission, his vital signs were stable and his laboratory results were within normal limits, including those of tumor markers. The colonoscopy revealed a large round submucosal mass in the sigmoid colon. The mass was of a hard consistency when pressed with a scope forceps (Figure 1). Abdominopelvic computed tomography scan indicated an approximately 4 cm homogeneous enhancing mass in the sigmoid colon and there was no evidence of lymphadenopathy (Figure 2). With the presumed diagnosis of a submucosal tumor, such as gastrointestinal stromal tumor (GIST), laparoscopic anterior resection was performed. Subsequently, histologic examination was performed, which indicated that the mass was a duplication cyst of the sigmoid colon. The cyst was lined by colonic-type mucosa and had a thin muscular layer partly shared with the adjacent colonic wall (Figure 3). The postoperative course was uneventful, and the patient was discharged on postoperative day-7.

DISCUSSION

Gastrointestinal duplication is a rare congenital anomaly that is mostly diagnosed before the age of two years [3, 4]. Although several theories exist regarding the development of gastrointestinal duplication, the etiology and pathogenesis have not been well established [2]. As the disease name implies, diagnosis is made on the basis of pathologic findings of a tubular or cystic structure that tends to be located on the mesenteric side of the bowel, shares a common muscular wall and blood supply with the bowel wall, and a separate mucosal lining [1]. Communication with the bowel lumen may or may not exist [5]. Although gastrointestinal duplication has been classified according to various pathologic criteria, it is simply divided into tubular or cystic in nature. Most gastrointestinal duplications are cystic structures and the ileum is the most common site [3, 4]. Colonic involvement accounts for less than 20% of cases of gastrointestinal duplication. Of these colonic duplications, 40% are located in the cecum, with the sigmoid being rarely involved [4].

The clinical manifestation of duplication cysts is variable and range from non-specific abdominal pain to peritonitis. The location, size, communication with the bowel lumen, and presence of the heterotopic mucosa, such as ectopic gastric mucosa, are the main determinants

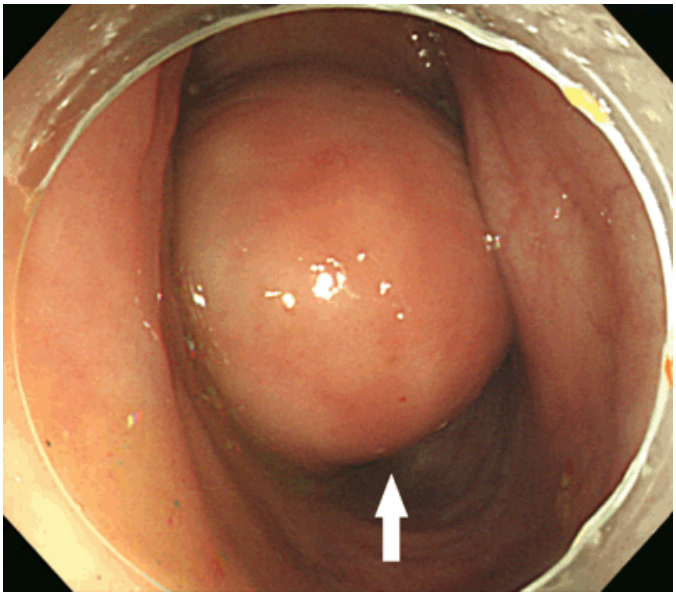

Figure 1: Colonoscopic findings showing an approximately 4 cm, round, submucosal mass (arrow) with a hard consistency is located on the sigmoid colon.

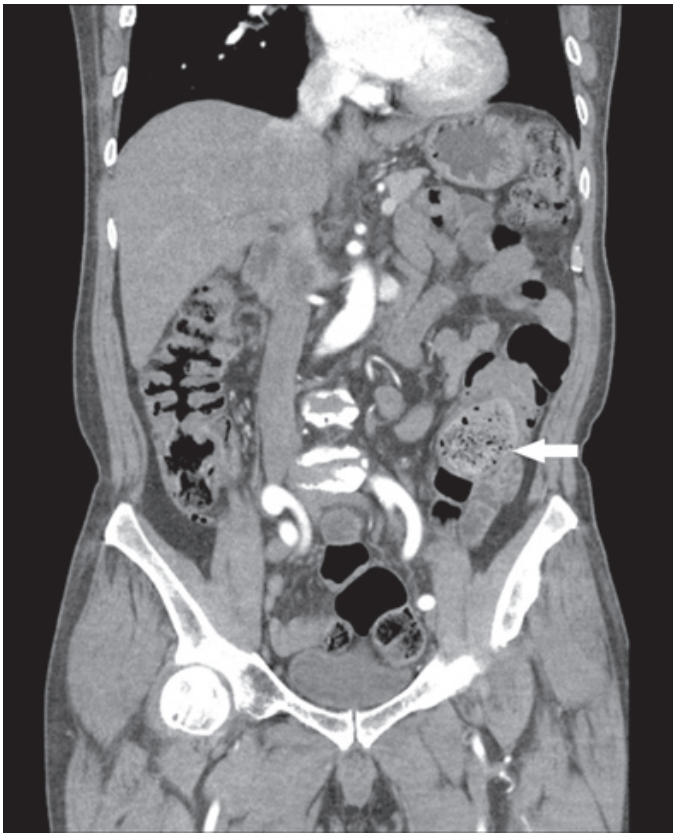

Figure 2: Abdominal computed tomography scan (coronal) indicated a homogeneous enhancing mass in the sigmoid colon without pericolic infiltration (arrow).

for the development of symptoms, including perforation, obstruction, and bleeding [2–6]. Most gastrointestinal duplication cysts are detected during childhood due to complication or related symptoms [2, 3]. Even though colonic duplication cyst may be a leading point resulting in colonic obstruction by intussusception, and infection, inflammation, and ulceration of the cyst may lead to the symptom and sign like diverticulitis and Crohn's disease, colonic duplication cyst tends to remain asymptomatic [3, 6, 7]. Thus, duplication cysts can be incidentally found in adults during colonoscopy or imaging studies for other reasons, such as that which occurred in our case.

The diagnosis of a duplication cyst may be made with imaging studies, such as ultrasonography, computed tomography (CT) scan, magnetic resonance imaging (MRI) scan, and contrast study [2, 3, 8, 9]. Ultrasonography is the imaging modality of choice in childhood patients because it provides highly accurate images and avoids the risk of radiation [3, 9]. However, its accuracy is operator dependent, and it may not be feasible in patients with complication. For cases in which Ultrasonography findings are equivocal or Ultrasonography is not feasible, a CT scan or MRI scan are useful modalities to establish the diagnosis given the nature, location, and extent of the lesion [2,8]. More recently, endoscopic ultrasonography (EUS) and EUS-guided fine needle aspiration have been applied for the diagnosis of colonic submucosal tumor. However, the role of EUS has not been established in the diagnosis of colonic duplication cyst. Contrast studies and colonoscopy are diagnostic if there is sufficient communication to allow contrast or a scope to pass between the true lumen of the colon and the cyst [2]. However, when there is no communication, diagnosis though contrast studies or colonoscopy is limited because the lesion can appear similar to a submucosal tumor. Thus, some authors have suggested that if the diagnosis is unclear, then laparoscopy may be used to confirm the diagnosis [3]. In the present case, although we performed laparoscopic surgery, the final diagnosis was made through pathologic examination because of the rarity of the case. Therefore, duplication of the colon may be considered one of the differential diagnoses in a patient presenting with cystic colonic mass.

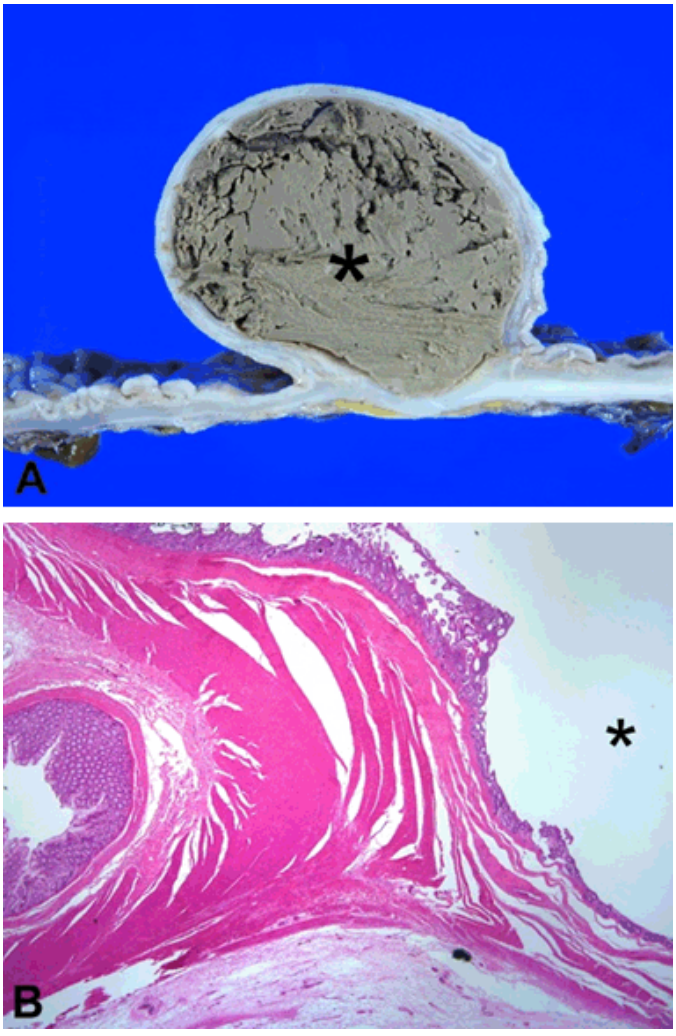

Figure 3: (A) A large, unilocular cyst (asterisk) is protruding into the lumen of the sigmoid colon, which is filled with brownish thick mucoid materials, and (B) Microscopically, the cyst (asterisk) is lined by colonic-type mucosa and has a thin muscular layer partly shared with the adjacent colonic wall (H&E stain, x100).

While the treatment of choice is not well established in asymptomatic patients, the treatment in symptomatic or diagnosed patients is surgical excision [4, 6]. Surgical treatment options range from simple cystectomy to en block resection with the adjacent colon. Since colon cancer may develop in a duplication cyst, and since laparoscopic colon resection is feasible, then en block resection may be a reasonable treatment option in patients with a colonic duplication cyst.

CONCLUSION

Although duplication cysts in the colon are rare, when a degenerative cystic colon mass is detected, a duplication cyst might be considered as one of the differential diagnoses.

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Author Contributions

Pyong Wha Choi – 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

Mee Joo – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, 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.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

A rare presentation of a mesenteric venolymphatic malformation with spontaneous hemorrhage in a newborn infant: A case report

Alaa Mahmoud, Shahla Bari, Dhanashree Rajderkar

ABSTRACT

Introduction: Abdominal venolymphatic malformations are benign rare congenital lymphatic malformations with various presentations. Reported presentations include acute abdomen, intestinal obstruction, torsion, and traumatic hemorrhage. Case Report: We present a case of a 36-week-gestational age female delivered via cesarean section secondary to developing intra-abdominal fluid on antenatal ultrasound. Subsequent imaging, exploratory laparotomy, and pathology were notable for a hemorrhagic left colonic mesenteric venolymphatic malformation with intra-abdominal fresh and coagulated blood. Conclusion: Although rare, previous cases of abdominal venolymphatic malformation have been described. This is the first reported case of spontaneous intrauterine hemorrhage requiring urgent cesarean section.

Keywords: Abdominal, Congenital, Hemorrhage, Prenatal, Venolymphatic malformation

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INTRODUCTION

Venolymphatic malformations are benign rare lymphatic vessel anomalies that occur often in the neck and axilla [1]. Intra-abdominal cystic venolymphatic malformations are exceedingly rare accounting for less than 5% of cases and often arise from the mesentery, retroperitoneum, and visceral organs. Abdominal venolymphatic malformations have various clinical presentations including acute abdomen, intestinal obstruction, torsion, hemorrhage after trauma and asymptomatic presentations [2, 3]. No cases of spontaneous hemorrhage have been reported to our knowledge.

CASE REPORT

A 36-week-gestational age female was delivered urgently via cesarean section due to developing intra-abdominal fluid and bowel distention on ultrasound in utero. The mother was a 27-year-old G2P1001 Caucasian otherwise healthy female. Apgar scores post cesarean section was six and nine at one and five minutes respectively. On delivery, the patient required

transfusion secondary to anemia (20% hematocrit with normal range being 55–68%). Paucity of gas in the left hemiabdomen was noted on plain films. Ultrasound demonstrated a heterogeneous mass in the left abdomen medial to the spleen and anterior to the left kidney measuring approximately 4.8x3.6x4.9 cm (Figures 1–3). This mass lacked flow on Doppler (Figure 4). Ultrasound also was notable for moderate complex ascites with debris consistent with blood products (Figure 2). Computed tomography scan confirmed the presence a large unilocular well circumscribed mass adjacent to the left colon with smooth margins and associated extensive complex ascites (Figures 5 and 6). Our differential diagnosis was a complicated mesenteric cyst versus hemorrhagic ruptured ovarian cyst.

Exploratory laparotomy was performed which revealed a left colonic mesenteric cystic mass with hemorrhage. Intraperitoneal clot and free peritoneal blood was noted. No bowel resection was required. The gross sample consisted of a 2.2x1.9x0.2 cm aggregate of tan-gray membranous friable tissue with clot. Pathology demonstrated benign cystic fibrovascular tissue septa with simple epithelial lining and rare irregular vascular channels filled with lymphocytes consistent with venolymphatic malformation (Figures 7–9). Postsurgically, the patient did well. The patient now is a year old without complications and is as expected developmentally for age.

DISCUSSION

Venolymphatic malformations account for about five percent of all benign tumors in infants and children. Fifty percent of cases involve the head and neck and only 5% are intra-abdominal [1–3]. Other rare anatomic sites have also been reported including the mediastinum, pleura, lungs, pericardium, and bone [4]. Sixty percent of

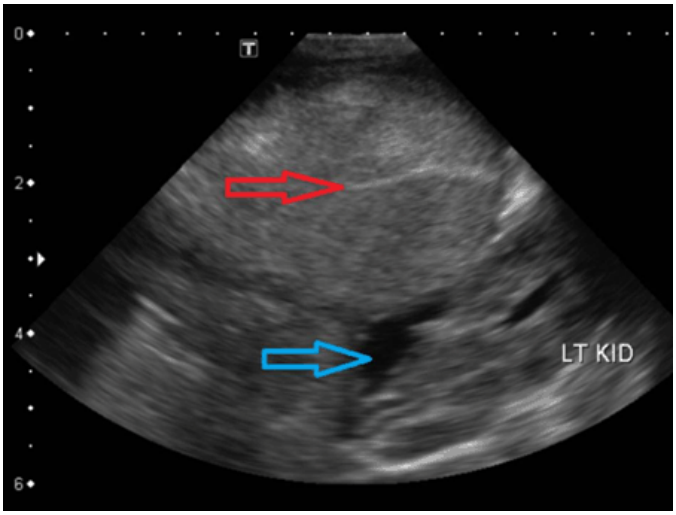

Figure 1: Left lower quadrant mass with septation (red arrow). Moderate volume ascites containing internal low-level echoes (blue arrow).

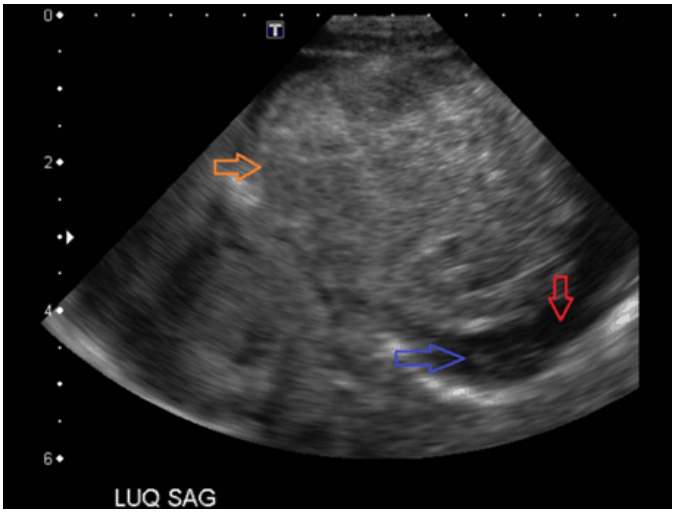

Figure 2: Left lower quadrant heterogeneous mass (orange arrow). Ascites with simple (red arrow) and complex (blue arrow) components.

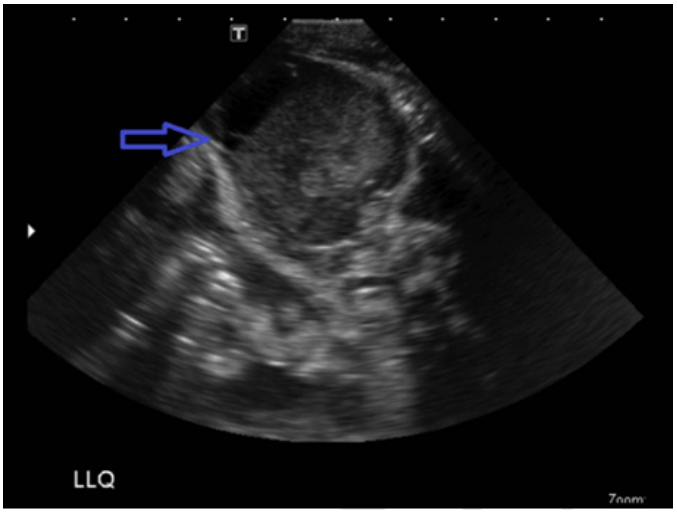

Figure 3: Ultrasound of left lower quadrant showed a heterogeneous mass in the left abdomen medial to the spleen and anterior to the left kidney measuring approximately 4.8x3.6x4.9 cm.

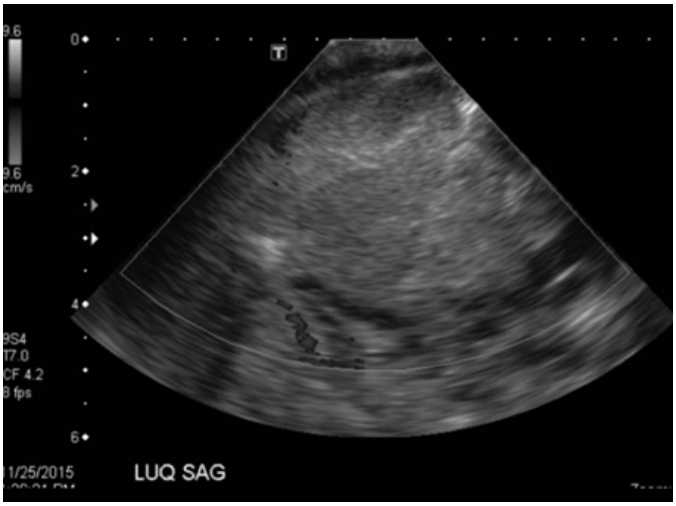

Figure 4: Ultrasound of the left lower quadrant mass with Doppler demonstrates no intralesional flow.

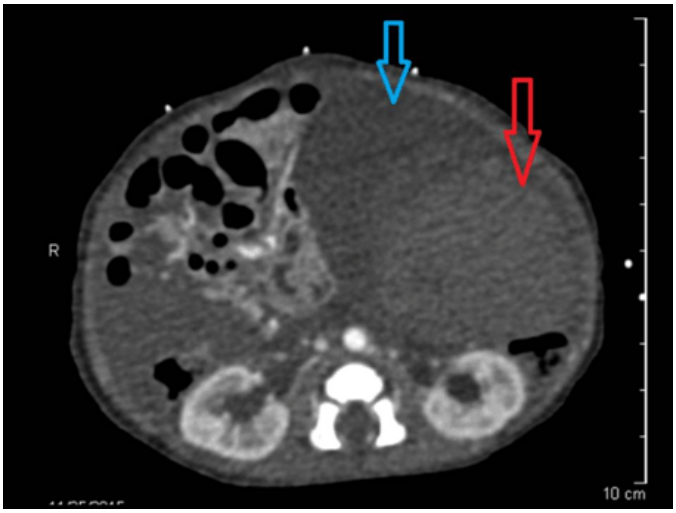

Figure 5: Computed tomography scan of abdomen and pelvis with intravenous contrast showed a large, cystic well circumscribed isodense-slightly hyperdense mass with smooth margins (red arrow) in left lower quadrant with surrounding fluid (blue arrow), displacing the colon. Few very small scattered cysts were noted (Orange arrows).

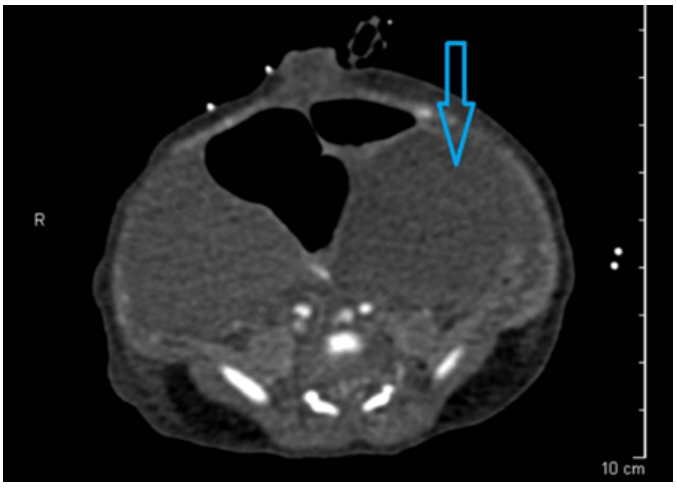

Figure 6: Computed tomography scan of abdomen pelvis with intravenous contrast showing extensive complex ascites (blue arrow).

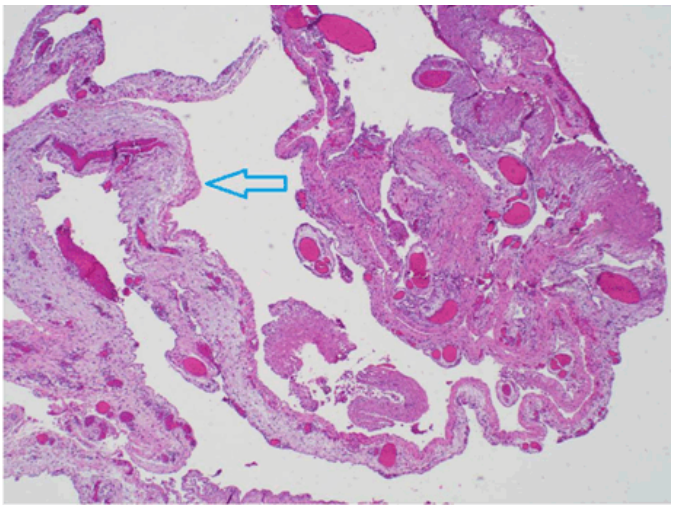

Figure 7: Collapsed cystic lining with fibrovascular septa (blue arrow).

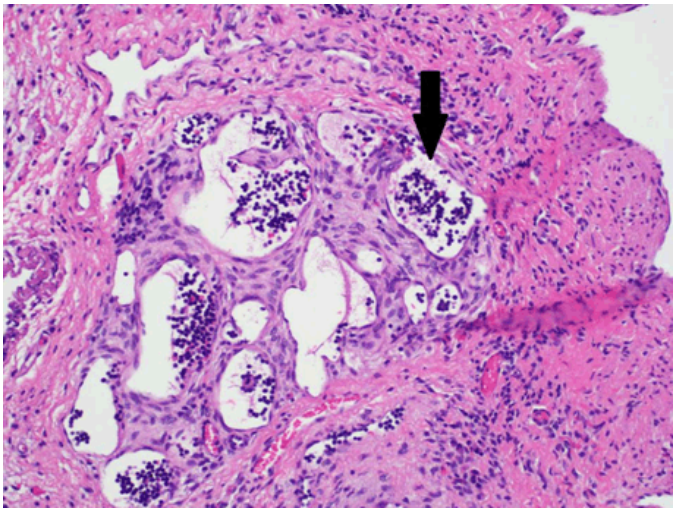

Figure 8: Collapsed cystic lining within the fibrovascular septa, rare lymphocyte filled channels are noted (black arrow) (H&E stain, x200).

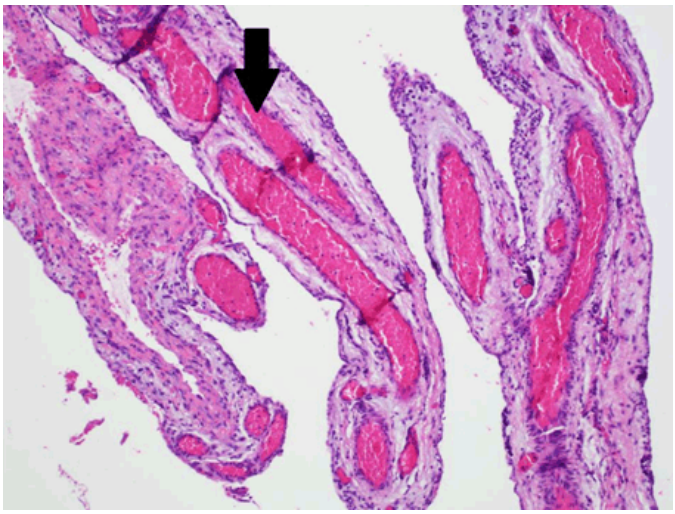

Figure 9: Vascular channels (black arrow) within the septa (H&E stain, x100).

these tumors are noticed at birth. Almost ninety percent are detected by the mean age of two years. Of the intra-abdominal venolymphatic malformations, the majority involve the mesentery of the small bowel. It is believed that these tumors result from an embryological failure of communication between the small bowel lymphatic tissue and the main lymphatic vessels resulting in blind cystic lymphatic spaces lined by endothelial layers.

Clinical presentation is variable and depends on mass size and location. Most abdominal cystic lesions (ACLs) present with a large, slow-growing and mobile mass along with abdominal distention [1]. Abdominal discomfort is common while acute peritoneal symptoms due to rupture, volvulus, hemorrhage or infection occur infrequently.

Ultrasound is the initial, quick and preferred modality to evaluate any suspected abdominal masses in a new

born infant. There are usually multiple thin septations in the multiloculated lesions. If these are complicated by hemorrhage or infection, floating debris, fluid levels and thick septations may be seen. On color Doppler examination, these do not show much internal vascularity.

Computed tomography scan is usually performed prior to surgical planning to know the extent of the lesion and to evaluate for any other organ involvement. On CT, these lesions are well defined, uni/multilocular with internal fluid density [1]. However, there may be areas of hyperdensity related to hemorrhage (as in our patient).

Small lesions with no complications can be observed. They are known to regress on their own. Surgical resection remains the treatment of choice in patients who have large lesions. Bowel resection is sometimes necessary if bowel is involved. Regrowth in the residual lesion is the most common complication but recurrence remains low. The use of sclerosing agents is reserved for immediate decompression in the macrocystic variety [5, 6].

CONCLUSION

Abdominal venolymphatic malformations are known to have various presentations including acute abdomen, intestinal obstruction, torsion, and traumatic hemorrhage. Cases typically present before the age of two. This is the first case to our knowledge of an abdominal mesenteric venolymphatic malformation with spontaneous hemorrhage in the perinatal age.

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Author Contributions
Alaa Mahmoud – 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
Shahla Bari – 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
Rajderkar A. Dhanashree – 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.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Scrotal swelling in a patient with paroxysmal supraventricular tachycardia

Riccardo Bentivegna, Domenico Nobile, Giuseppina Novo, Salvatore Novo

ABSTRACT

We present a case of a patient with paroxysmal supraventricular tachycardia and a history of chronic ischemic disease evolved into dilated cardiomyopathy. During hospitalization, the occurrence of arrhythmic crisis, our patient felt sporadic palpitations associated with dyspnea, distended jugular veins and scrotal swelling, quite unexpected clinical sign that caught our attention. Echocardiography showed dilated left ventricle, severely reduced systolic function, dilated right-sided, dilated inferior vena cava with reduced respiratory excursions. Echography of both testicles showed bilateral expansion of pampiniform plexus with reflux to the functional maneuvers, such as varicocele of grade III–IV. Afterwards coronary angiography, the placement of a metallic stent in left anterior descending artery improved coronary circulation with the net reduction of dyspnea. In later episodes of supraventricular tachycardia the only symptom that the patient felt was mild palpitation associated with scrotal swelling and distended jugular veins. The increased heart rate led to an exacerbation of the underlying pulmonary hypertension resulting in venous stasis upstream the pulmonary capillaries. Restoring sinus rhythm, the heart performs its normal pump function and the pressure within the pulmonary capillaries was decreased, so on physical examination turgor of the jugular veins and scrotal size were strongly reduced.

Keywords: Paroxysmal supraventricular tachycardia, Pulmonary hypertension, Scrotal swelling, Varicocele

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INTRODUCTION

Paroxysmal supraventricular tachycardia (PSVT) is an acronym that includes all of the arrhythmias that originate above the ventricles and that manifest paroxysmally. The causes of these arrhythmias may be different. The most common mechanism is the presence within the heart of a 'short circuit' due to the presence of an anatomical or functional so-called accessory pathway.

The most common forms of PSVT are atrioventricular nodal reentrant tachycardia (AVNRT) and atrioventricular reentry tachycardia (AVRT). The atrioventricular nodal reentrant tachycardia is characterized by the presence of this reentry circuit in the atrioventricular node [1].

The reentry functional circuit is constituted by two parts with different impulse conduction velocity, and tachycardia is generated when the impulse can circulate inside the reentry circuit [2].

The atrioventricular nodal reentrant tachycardia is a benign arrhythmia in most cases, whose main symptoms are palpitations and shortness of breath (although not always present) [3].

CASE REPORT

A 66-year-old Italian male, with a history of high blood pressure and chronic ischemic heart disease (two prior PCI in the circumflex coronary artery) evolved into dilated cardiomyopathy, reaches the emergency room of our hospital because of onset of worsening dyspnea.

On admission, dyspnea was reduced, blood pressure was 130/70 mmHg, arterial oxygen saturation was 99%, blood test demonstrated normal levels of cardiac troponin I (82 ng/ml), elevated creatinine (1.46 mg/dl) with eGFR 51 ml/min/m^2, elevated white blood cell count (12 x10^6/L). Chest X-ray showed no ilo-pleural-parenchymal lesions in active phase.

During the next few hours, the patient becomes progressively dyspneic and asthenic and continuous electrocardiography monitoring showed two episodes of PSVT with heart rate of 160 bpm, both regressed with restoration of sinus rhythm after carotid sinus massage (Figures 1 and 2).

The patient was, therefore, transferred to our cardiology department for the continuation of the diagnostic and therapeutic iter. The patient appeared confused and with loss of memory. Physical examination revealed heart tones in rhythmic succession, systolic murmur 2–3/6L more audible on the mitral outbreak, slightly reduced breath sounds in all lung fields and fine crackles to the basic fields. 12-lead electrocardiography showed: sinus rhythm, previous necrosis in the inferior leads, repolarization abnormalities in the inferior and posterolateral leads.

It was set aspirin therapy, omeprazole, fondaparinux, furosemide, ciprofloxacin, amiodarone, saline 0.9% NaCl infusion at 30 ml/h.

During hospitalization, the occurrence of arrhythmic crisis, our patient felt sporadic palpitations and dyspnea associated with the appearance of distended jugular veins and scrotal swelling (Figure 3), quite unexpected clinical sign that caught our attention, on physical examination.

Considering the clinical-anamnestic context it was performed coronary angiography that showed a critical stenosis of 70% left anterior descending coronary artery (LAD) and good patency of the prior stents in circumflex coronary artery. It was therefore performed a percutaneous transluminal coronary angioplasty (PTCA) with metal stent placement in LAD. Improving cardiac perfusion, ongoing of arrhythmia, our patient does not feel the symptoms described above. However, at the following episodes of PSVT, on physical examination persisted scrotal swelling (already observed before the angioplasty procedure) and the appearance of distended jugular veins.

Echocardiography showed dilated left ventricle (VTD 160 ml - DTD 65 mm) with wall thickness increased; severely reduced systolic function (EF 28%); akinesia of the lower and posterolateral wall, hypokinesis of the remaining segments; severely dilated left atrium (150 ml); regular size of the aortic bulb and ascending aorta ectasia (39 mm); dilated right-sided with reduced TAPSE; dilated inferior vena cava with reduced respiratory excursions; thickened mitral leaflets with symmetrical tethering; sclerosis of the semilunar aortic valves; moderate mitral regurgitation with restrictive pattern, mild aortic regurgitation, moderate tricuspid regurgitation.

Echography of both testicles performed through high-frequency probe (12 MHz) showed normal morpho-volumetric and eco-structural appearance of both didymus and epididymis; regular intraparenchymal vascularization to the color Doppler evaluation; bilateral expansion of pampiniform plexus (maximum right vessel caliber 2.6 mm - maximum left vessel caliber 3.6 mm) with reflux to the functional maneuvers, such as varicocele of grade III-IV. Coexisted diffuse thickening of the subcutaneous soft tissues such as edematous imbibition (Figures 4A–B).

DISCUSSION

The veins that originate from pampiniform plexus,

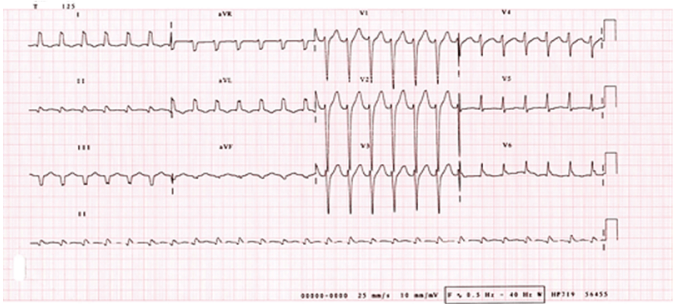

Figure 1: Electrocardiogram registered during paroxysmal supraventricular tachycardia.

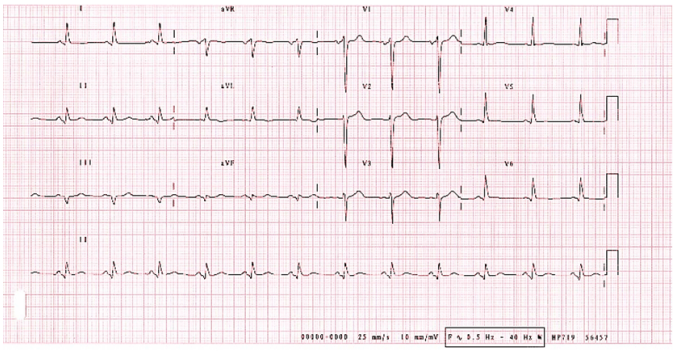

Figure 2: Electrocardiogram showing a restores sinus rhythm .

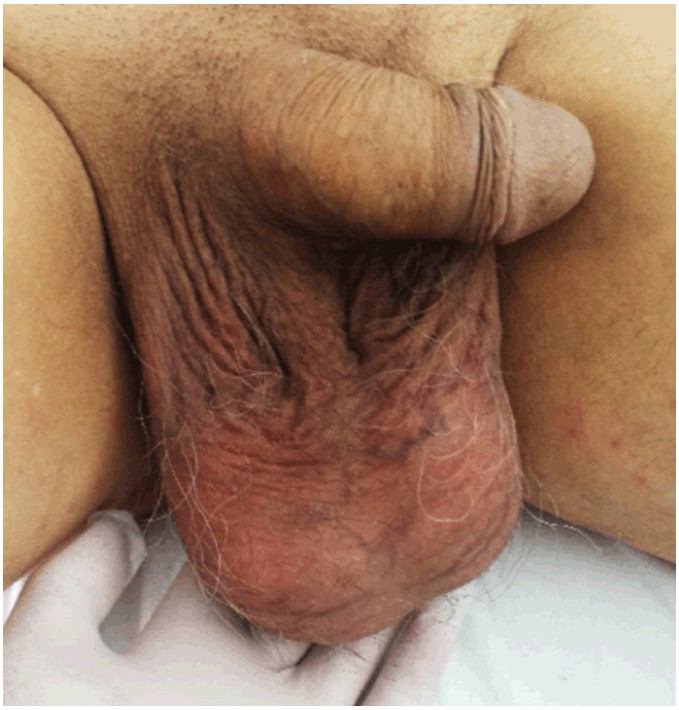

Figure 3: Patient's scrotal swelling.

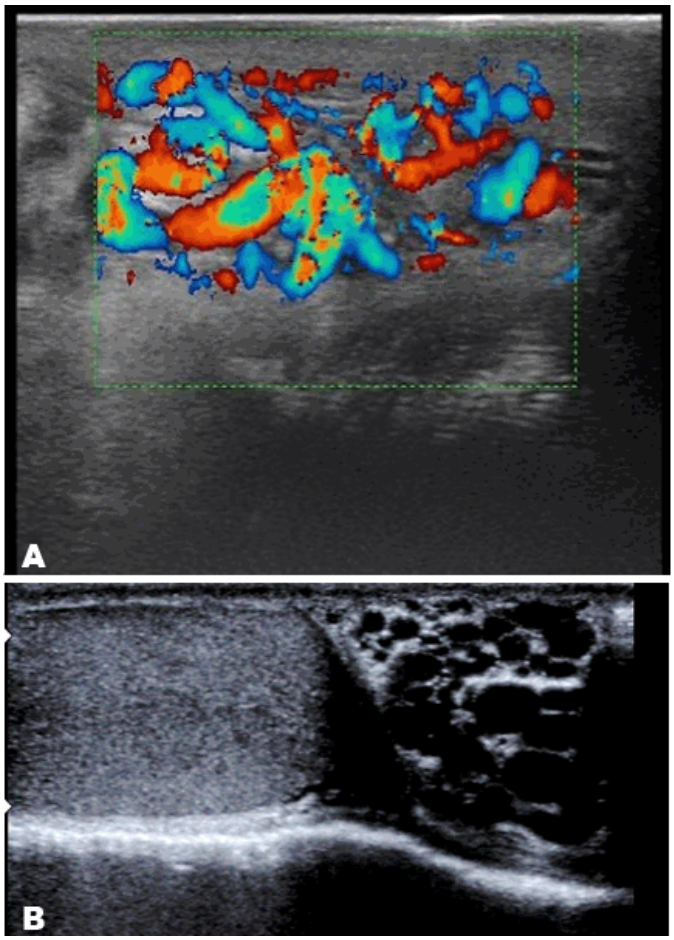

Figure 4 (A, B): Echography images of varicocele.

after the passage along the inguinal canal, join to form a single testicular vein, which opens on the right side into the inferior vena cava (has an high flow) at an acute angle; on the left side into the left renal vein (has low flow) at a right angle [4].

These veins can become incontinent and dilate thereby preventing venous outflow of blood from the testicular veins to the inferior vena cava. This creates a condition of reflux and stasis of blood to the testicle. This is manifested in particular in the left testicle (95%) and rarely in the right testicle (5%) because of the different anatomical features between the two vascular pathways [5].

A complete physical examination in orthostatic position allows highlighting the expansion of veins during the Valsalva maneuver. Instrumental support investigations (ultrasound and testicular color Doppler of the spermatic vessels) allow to determine the extent of reflux and measure the diameters of the testicles [6, 7].

Current guidelines define pulmonary hypertension as a hemodynamic and pathophysiological condition characterized by an increase in mean pulmonary arterial pressure (PAPm) ≥25 mmHg at rest as assessed by right heart catheterization (RHC). Available data have shown that the normal PAPm at rest is 14±3 mmHg [8].

The secondary forms of pulmonary hypertension may result from [9]:

- congenital and acquired heart disease (mitral steno-insufficiency, left ventricular failure)
- vessel disease: pulmonary thromboembolism, vasculitis, etc.
- respiratory diseases characterized by chronic airway obstruction (COPD, emphysema, etc.)

Symptoms of pulmonary hypertension are very unspecific. However, shortness of breath appears to be the most common initial symptom (60% of cases) and is present in 98% of patients at diagnosis. Other disorders, less frequent in the early stages, are often observed at diagnosis, and include fatigue, weakness (73% at diagnosis), chest pain (47%), fainting (41%), syncope (36%), lower limb edema (37%) and palpitations (33%) [10].

CONCLUSION

Scrotal swelling, quite unexpected clinical sign, in our case report was secondary to the patient's pulmonary hypertension. It was present both as a result of cardiac causes (left ventricular failure and moderate mitral regurgitation), both as a result of pulmonary causes (history of COPD). The increased pressure developed in the pulmonary circulation, determines upstream blood stagnation. It was observed instrumentally on echocardiography with a dilated inferior vena cava and poor respiratory excursions, and clinically with distended jugular veins and the sudden and temporary scrotal swelling during the PSVT episode (was confirmed by ultrasound with the detection of varicocele grade IV and the high blood stasis). Restoring sinus rhythm, the pressure within the pulmonary capillaries decreased, so

on physical examination turgor of the jugular veins and scrotal size were strongly reduced.

****

Author Contributions

Riccardo Bentivegna – 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
Domenico Nobile – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Giuseppina Novo – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Salvatore Novo – 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.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Granulocytic sarcoma of epidural mass with acute myeloid leukemia

Garima Singh, Ashutosh Kumar, Mili Jain, Rashmi Kushawa, Arun A. Kumar, Durg Pratap Singh

ABSTRACT

Extramedullary manifestations of acute leukemia include a wide variety of clinically presentation that often pose difficulty in diagnosis and treatment of myeloid sarcoma. We present a case of a five-year-old boy with initial complaints of radicular pain of both lower limbs and urinary retention. On MRI, compression by epidural mass was identified, which was shown to be an extramedullary myeloid sarcoma diagnosed on the basis of Auer rods containing blasts in peripheral blood smear and bone marrow. Diagnosis was confirmed with flow cytometry and induction chemotherapy was started. Initial neurological presentation of paraplegia due to chloroma is extremely rare in myeloid leukemia with very few case reports published earlier.

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INTRODUCTION

Myeloid sarcoma, also termed extramedullary acute myeloid leukemia, extramedullary myeloid tumor, and granulocytic sarcoma have also been referred to as chloroma secondary to their characteristic green color created by the presence of myeloperoxidase [1]. It is a rare manifestation that is characterized by the occurrence of one or more tumor myeloid masses occurring at an extramedullary site with effacing the architecture of tissue. Chloroma are most frequently seen in acute myeloid leukemia (AML) and few cases in myelodysplasia and other myeloproliferative disorders. Myeloid sarcoma present with wide clinical manifestation, granulocytic sarcoma should be considered in the differential diagnosis of an epidural mass along-with other diagnosis. Granulocytic sarcoma virtually can occur in any organ or tissue, most common sites being skin (called leukemia cutis) and bones. Other sites are lymph nodes, mediastinum, epidural sites, small intestine, ovary/testis and brain. Treatment for chloroma consists of systemic chemotherapy for the underlying leukemia, and the lesions frequently respond well. If the lesion is refractory

to systemic chemotherapy, then surgical debridement or radiation therapy may be considered. Here we present a case of five-year-old boy with initial presenting symptom of paraplegia due to granulocytic sarcoma which is extremely rare.

CASE REPORT

A five-year-old boy presented with initial complaints of radicular pain and progressive weakness of both the lower limbs of two months duration followed by difficulty in passing stools and urine for the last one week. There was no history of trauma, significant weight loss, or contact with tuberculosis. On examination, he had pallor; vitals were normal. No lymphadenopathy or hepatosplenomegaly was present. Higher mental function status and cranial nerve examinations were normal. Upper limbs power was normal while in lower limbs, muscle tone was decreased and grade 1/5 power was seen in both the lower limbs. Sensations were intact in both lower limbs. Initial routine blood investigations were done outside and showed hemoglobin 5.7 g/dl, total count 14,800 cells/mm^3, polymorphs 45%, lymphocytes 50%, eosinophil 2% and monocyte 3%. The RBCs mean cell volume (MCV) and mean cell hemoglobin (MCH) and mean corpuscular hemoglobin concentration (MCHC) were reduced as well as platelets count were reduced. Magnetic resonance imaging (MRI) scan showed an ill-defined contrast enhancing lesion in posterior epidural space extending from D2–D10 level, displacing the cord anteriorly with signal intensity alternation and cord compression from D2–D10 level causing compressive neuropathy (Figure 1). Then, peripheral smear was done again in our laboratory which showed (Figure 2) mild reduced of MCV of RBCs. White blood cell count was normal and showed predominantly blast cells with high nuclear cytoplasmic ratio, moderate cytoplasm, hyperchromatic nuclei with multiple nucleoli. Platelet count was decreased. Bone marrow (Figure 3) examination was ordered following the presence of blast cells in peripheral smear and it showed that the marrow elements were completely replaced with blast cells having high nuclear cytoplasmic ratio, moderate amount of eosinophilic cytoplasm, and hyperchromatic nuclei with multiple nucleoli. Focal Auer rods and few maturing cells of the myeloid series were present. The marrow picture was corresponding to acute myeloid leukemia M2. Considering the possibility of granulocytic sarcoma as the cause of paraplegia in acute myeloid leukemia, the patient was started on induction chemotherapy.

DISCUSSION

Myeloid sarcoma (chloroma, granulocytic sarcoma, extramedullary myeloid tumor), may be defined as extramedullary solid tumor mass of immature myeloid cells or monoblast cell which disrupt the architecture of the tissue associate with or without bone marrow involvement [2].

Myeloid sarcoma can be primary, which is very rare and secondary when it is associated with acute myeloid leukemia approximately 3–10%, but rarely it can also associated with myelodysplasia and myeloproliferative disorder.

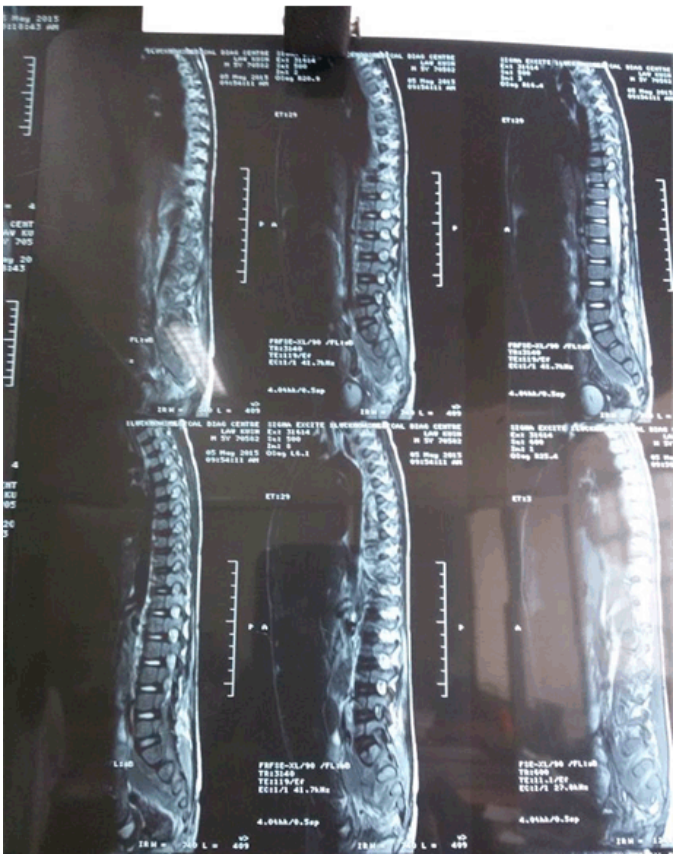

Figure 1: Magnetic resonance imaging scan of the dorsal spine showing an epidural mass lesion D2–D10 level.

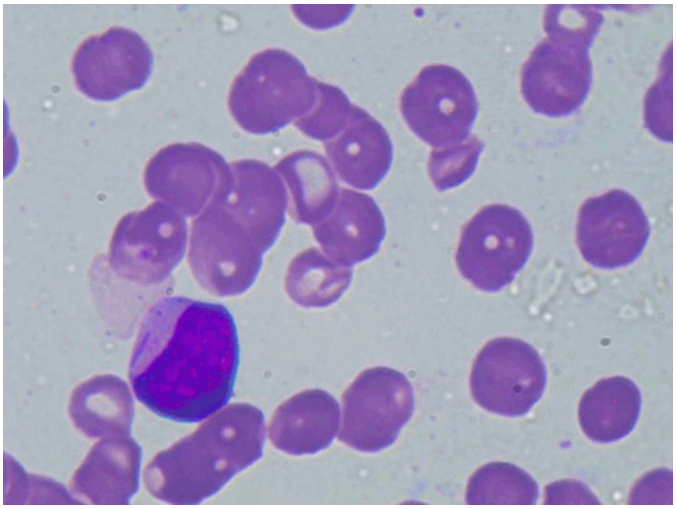

Figure 2: Peripheral blood smear showing blast cells with Auer (Leishman stain, x100).

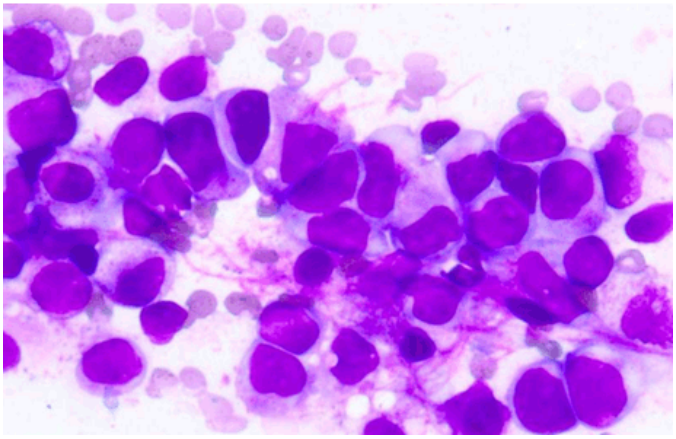

Figure 3: Bone marrow smear showing blast cells suggested of acute myeloid leukemia (Leishman stain, x100).

Myeloid sarcoma can occur in any age group but predominantly 35–50 years of age, as well as it can occur at any site but most common site skin, periosteum, bone, lymph node, soft tissues, beside this numerous other sites has also been reported [3].

Myeloid sarcoma has wide differential diagnosis because of various sites of presentation, along with wide variation of age and symptom presentation. Most common differential diagnosis include round cell tumor such as lymphoblastic lymphoma, medulloblastoma, rhabdomyosarcoma, Ewing/PNET along with other differential such as undifferentiated epithelial cell tumor [4].

Myeloid sarcoma diagnosis requires various diagnostic tools beside morphology in peripheral blood smear, such as special cytochemistry, specific marker for immunophenotyping, and cytogenetic, all play important role in diagnosis of myeloid sarcoma.

On the basis of cytomorphology, myeloid sarcoma classified as blast (which included myeloblasts with little evidence of promyelocyte) immature (shows an intermediate degree of differentiation contains principally myeloblasts and promyelocytes; eosinophil myelocytes are usually present) and mature myeloid cells (primarily consists of promyelocytes and later stages of maturation with abundant eosinophil myelocytes). Blast variant of myeloid sarcoma has differential diagnosis with lymphoblastic lymphoma, carcinoma, melanoma while immature and mature variant of myeloid sarcoma with Hodgkin lymphoma, extramedullary hematopoiesis and infections.

Cytochemistry test include myeloperoxidase, naphthol AS-D chloroacetate esterase and non-specific esterase reaction but their role in diagnosis has been replaced by immunophenotyping methods by flow cytometry. Immunophenotyping panel for diagnosis of myeloid sarcoma includes myeloperoxidase, lysozyme, CD68(KP-1 and PGM-1), CD34 and CD117,CD 43, CD 3, CD 20 are very useful in differentiation for myeloid sarcoma from B cell and T cell lymphoma [5].

Rarely myeloid cell differentiated into erythroid and megakaryocyte linage which can be differentiated by marker for megakaryocyte CD41, CD61 and for erythroid series glycophorin A, hemoglobin A.

Cytogenetic nowadays play an important role in the diagnosis of myeloid sarcoma. However, no specific cytogenetic has been still discovered for myeloid sarcoma. Cytogenetic usually done on blood or bone marrow samples by fluorescence in situ hybridization (FISH). Most commonly cytogenetic abnormality noted in myeloid sarcoma are translocation t(8; 21), inversion of chromosome 16 (Inv 16) along with few rare cytogenetic abnormality are monosomy 7 or 5 and trisomy 8 [6].

Myeloid sarcoma either primary or secondary both possess common treatment regimen conventional AML-type chemotherapeutic protocols. The role of other therapy (radiotherapy, hematopoietic stem transplantation, and targeted therapy) method has not been well established. If local symptoms such as compromise of the spinal cord due to myeloma are present, surgery is considered.

Myeloid sarcoma is rare entity hence it has conflict regarding its prognosis. There has been no variation in prognosis of myeloid sarcoma either as isolated or associated with acute myeloid leukemia reported.

CONCLUSION

This article is yet another classic example showing that myeloid sarcoma can have varied clinical presentations, based primarily on the site of involvement. Early suspicion of the diagnosis, peripheral blood smear, bone marrow and appropriate investigations can aid in early diagnosis of myeloid sarcoma with timely initiation of chemotherapy for the patient.

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Author Contributions

Garima 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

Ashutosh Kumar – 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

Mili Jain – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Rashmi Kushawa – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Arun A. Kumar – Substantial contributions to conception and design, Acquisition of data, Analysis and

interpretation of data, Final approval of the version to be published
Durg Pratap Singh – 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

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Conflict of Interest
Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Coexistence of papillary carcinoma and anaplastic large cell lymphoma in a healthy 26-year-old male: A first case report

Majdi Saada, Mechal Weiler, Yaneev Zohar, Ayelet Raz-Paster

ABSTRACT

Introduction: We present an extremely rare presentation case of simultaneous existence of anaplastic large T cell lymphoma along with papillary thyroid carcinoma in a previously healthy 26-year-old male. Case Report: A previously healthy 26-year-old male presented with a history of fever, headache and subcutaneous nodules. Chemotherapy was initiated after histopathologic examination from one skin nodule revealed findings consistent with anaplastic large T cell lymphoma. A ^{18}F-fluorodeoxyglucose positron emission tomography computed tomography scan (FDG-PET-CT) showed nodal involvement including the nasopharynx and extranodal involvement including disseminated skin involvement and a lung lesion A follow-up FDG-PET-CT which was performed five months after the initiation of treatment, showed resolution of all disease sites but a persistent pathological focal uptake at the right lobe of the thyroid gland. Ultrasound and a following fine needle aspiration revealed findings consistent with papillary carcinoma. The patient underwent successful surgical resection of the thyroid. Conclusion: This rare case illustrates the possibility of existence of concurrent diseases in patients with systemic ALCL.

Keywords: ALCL, PTC, Coexistence

How to cite this article

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INTRODUCTION

Anaplastic large cell lymphoma (ALCL) is a subgroup of the peripheral T cell lymphomas (PTCL) which are a heterogeneous group of generally aggressive neoplasms that constitute less than 15% of all non-Hodgkin lymphomas in adults [1]. The ALCLs are further subdivided into three categories according to clinical criteria and immunohistochemistry: primary systemic anaplastic lymphoma kinase (ALK)(+), primary systemic ALK(–), and primary cutaneous ALCL [2]. Most patients with ALK negative ALCL are adults (age range 40–65 years) with a slight male predominance [3].

Thyroid cancer accounts for only 0.5–1.5% of all malignancies [4]. Papillary thyroid carcinoma (PTC) accounts for over 85% of all malignant thyroid tumors in regions with sufficient iodide intake. Risk factors for the development of thyroid cancer include radiation

exposure, family history of thyroid cancer and hepatitis C related chronic hepatitis [5–7].

Herein, we report a clinical case of a 26-year-old-male with ALK(-) ALCL and papillary carcinoma of the thyroid which appeared after five months following chemotherapy treatment.

CASE REPORT

A previously healthy 26-year-old male presented with a history of fever, headache and subcutaneous nodules on the scalp of three weeks' duration. The initial evaluation including physical examination, blood tests, brain CT and lumbar puncture was non-diagnostic. The patient's skin lesions were suspected to be multiple scalp trichilemmal cysts and the patient was discharged to ambulatory follow-up.

Two months later, he was admitted to the emergency department complaining of a neck pain, chest pain, dyspnea, general weakness and fatigue since two weeks. In addition, he reported a weight loss of six kilograms during a two-month period. On examination, the patient was fully oriented, his body temperature was 38.1°C, the heart rate was 105 beats per minute, the blood pressure was 130/70 mmHg and oxygen saturation was 96% while he was breathing ambient air. Skin examination revealed a firm nodular purple rash on the chest and the abdomen in addition to a soft nodule on the posterior scalp (Figure 1). A lymph node was enlarged on the left inguinal region. Lungs were clear on auscultation, there were no heart murmurs, and the abdomen was soft with no hepatosplenomegaly. Neurological examination revealed a supple neck with no other focal signs, the rest of the physical examination was otherwise normal. A complete blood count showed white blood cells of 10900 with 80% neutrophils, 18% lymphocytes and 0.6% eosinophils. The erythrocyte sedimentation rate was 70 mm/hr, the alkaline phosphatase level was 152 U/L, the lactate dehydrogenase level was 630 U/L, and serological tests for viral hepatitis B and hepatitis C were negative. A lumbar puncture was performed showing no evidence of central nervous system infection.

Computed tomography scan of the chest showed no lymphadenopathy. Computed tomography scan of the abdomen showed enlarged liver and spleen, enlarged left inguinal lymphatic nodes 3–5 cm in diameter. Histopathologic examination from one skin nodule and from the inguinal node revealed infiltration by large T cell lymphocytes which were stained positively for CD3, CD4, CD30 and MUM1 and negative for CD8, TIA1, ALK, CLUSTERIN, CD123, CD56, CD10, CD20 and TDT (Figure 2). A proliferation index of 80% was reported. A bone marrow revealed hypercellular infiltration with T cells consistent with ALK negative ALCL. A ^{18}F-fluorodeoxyglucose positron emission tomography computed tomography scan (FDG-PET-CT) showed nodal involvement including the nasopharynx and extranodal involvement including disseminated skin involvement and a lung lesion (Figure 3). A diagnosis of aggressive ALK negative ALCL stage IVB was made and treatment with CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) and high dose methotrexate was begun.

Following five cycles of the mentioned chemotherapy the patient felt well and there were notable improvement of the skin lesions (Figure 4). The patient underwent autologous hematopoietic stem transplantation with minor complications. A follow-up FDG-PET-CT performed five months after the initiation of treatment, showed resolution of all disease sites and pathological focal uptake at the upper right lobe of the thyroid gland (Figure 3). Ultrasound of the thyroid showed a solid echogenic nodule in the right lobe measuring 5.2x7.7 mm and another small nodule in the lobe of less than 5 mm. A fine needle aspiration showed findings consistent with papillary carcinoma of the thyroid. The patient was referred to a surgeon to undergo a total thyroidectomy. Histopathological examination from the thyroid confirmed the same diagnosis. On subsequent follow-up two months later; the patient reported feeling well with no traces of any of his diseases.

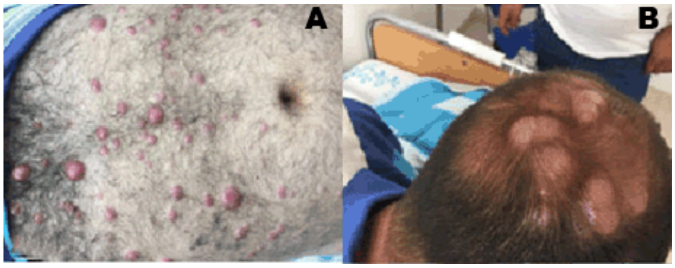

Figure 1 (A, B): The patient had multiple erythematous, tender nodules on the chest and abdomen, as well as several tender nodules on his scalp.

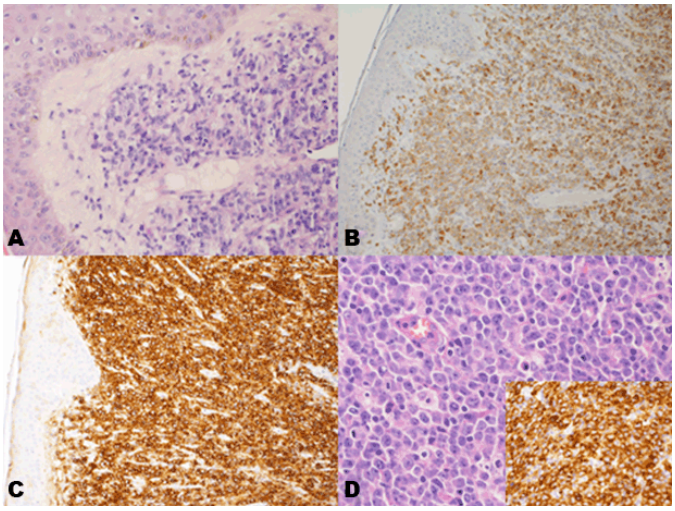

Figure 2: Abdominal skin biopsy showing dermal infiltration by large, anaplastic lymphocytes (A, H&E X40) positive for CD3 (B, X40) and CD30 (C, X40). Excisional lymph node biopsy showing marked infiltration by the same cells (D, H&E X60) which were positive for CD30 (inlet, X60).

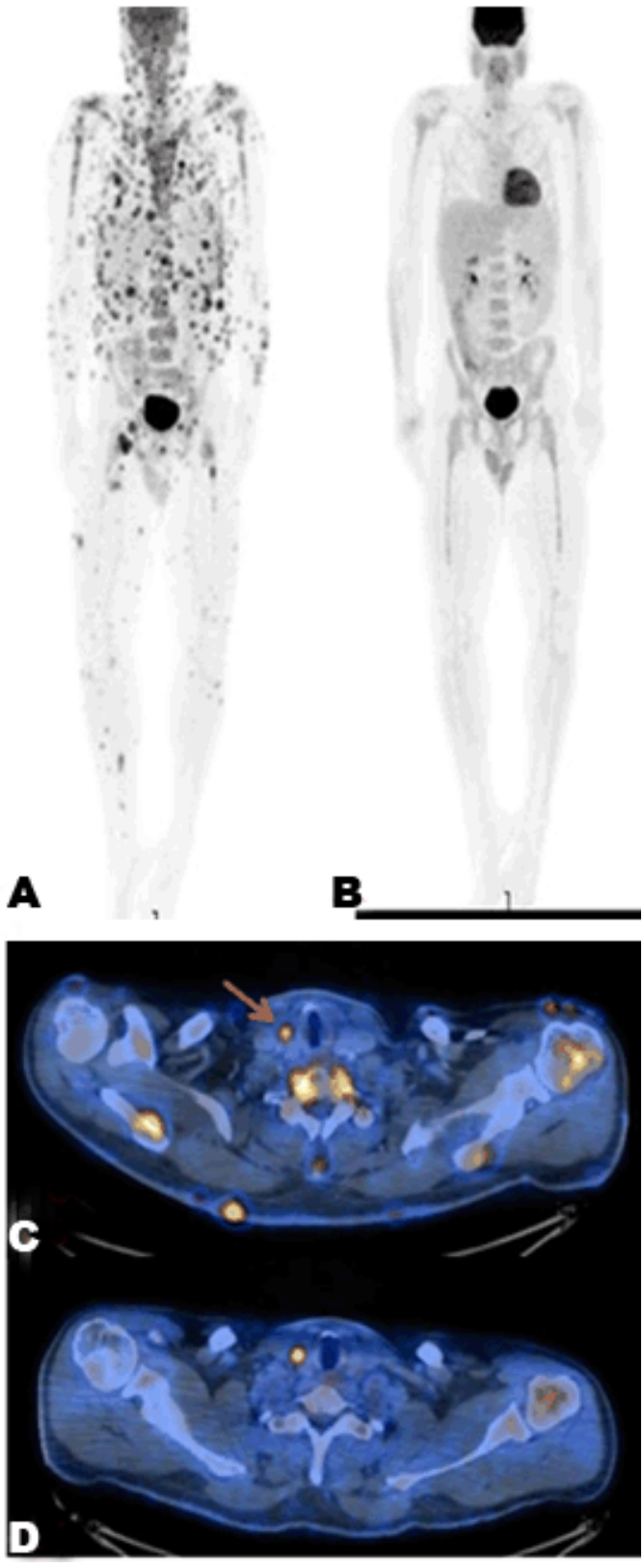

Figure 3: FDG-PET/CT scan at staging and following treatment of ALCL. (A) Maximal intensity projection showing disseminated skin involvement and additional nodal and extranodal sites (B), and resolution of all disease sites following treatment. (C) Axial fused images at the level of the neck show a focus pathological uptake in the staging study (arrow) and (D) following treatment that proved be PTC.

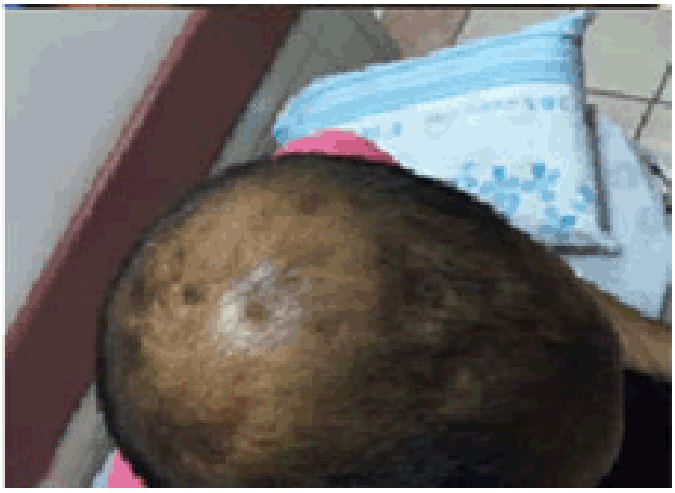

Figure 4: These lesions improved significantly following five cycles of systemic chemotherapy.

DISCUSSION

Papillary carcinomas of the thyroid are the most common malignant growth affecting the thyroid, currently representing 60–65% of malignant thyroid neoplasm. Most patients with papillary thyroid carcinomas present clinically with the discovery of a thyroid nodule during an unrelated physical exam or by the patient or others who notice a lump in the patient's neck [8, 9]. Although the etiology of this neoplasm is unknown, they are thought to be related to neck irradiation, adenoma transformation, Hashimoto thyroiditis, family history of thyroid cancer, occupational and environmental exposures [9], None of which our vignette meets. The simultaneous existence of papillary carcinoma and other tumors had been reported in literature in rare cases, including the coexistence with squamous cell carcinoma in the thyroid and anaplastic carcinoma [10, 11]. The development of papillary carcinoma after chemotherapy is very rare, with one case report describing the development of PTC ten years after treating stage 1A Hodgkin's lymphoma with ABVD (adriamycin, bleomycin, vinblastine, dacarbazine) chemotherapy [12].

Overall, papillary carcinomas represent an indolent group of neoplasm and have an excellent prognosis.

Anaplastic large cell lymphoma is a peripheral T cell derived malignancy, representing around 2–3% of all lymphoid neoplasms. It is an aggressive lymphoma which frequently presents in an advanced clinical stage with systemic symptoms and extranodal involvement [13]. In a case of a systemic ALCL, pathological FDG uptake at the neck region can be seen as a part of the disease itself [14]. The coexistence of PTC with other lymphomas is rare, with 2 cases reporting the concurrent existence of thyroid MALT lymphoma and primary thyroid diffuse large cell lymphoma, respectively [15, 16]. Likewise, CD30+ T cell lymphoma and concurrent solid carcinoma tumor is a rare clinical scenario, with previous cases in literature reporting the co-existence with gastric, esophageal and

renal carcinomas [17–19]. This is the first clinical report of concurrent ALCL with PTC. The pathophysiology of the underlying mechanism is poorly understood, with some theories relating to monotypic expansion of activated T cells that occurs with solid tumors, and it is possible that T cell lymphoma originates from cytotoxic CD30+ T cell expansion [20]. Secondary genetic abnormalities probably lead to the dominance of a T cell clone, leading to irreversible transformation to lymphoma [21]. Whether ALCL predisposes to, or is merely associated with papillary carcinoma, is still a controversial issue which requires a more detailed consideration.

CONCLUSION

Patients with anaplastic large cell lymphoma (ALCL) can frequently present with pathological uptake on PET-CT, thus easily masking an underlying solid tumor. Moreover, this case illustrates the need for further research on the underlying mechanism of the seemingly related tumors.

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Author Contributions
Majdi Saada – 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
Mechal Weiler – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Yaneev Zohar – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ayelet Raz-Paster – 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
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CASE REPORT PEER REVIEWED | OPEN ACCESS

Leiomyoma of hard palate: A rare case report

G. Siva Prasad Reddy, B. Jagannadha Prasad, A. Bhargavi Krishna, P.S.S. Tejaswini, J. Laxmi Sravya, S. Sushma, E. Padmini

ABSTRACT

Introduction: Leiomyomas are benign, soft tissue tumors, arising from the smooth muscle. They usually affect the muscular layer of the gut and body of the uterus. Oral leiomyomas are extremely rare and are thought to arise from the smooth muscle wall of blood vessels. They present as slow growing, well-circumscribed, painless swellings accounting for only 0.42% of soft tissue neoplasms [1] of the oral cavity. Case Report: A 14 years old boy presented to our department with a chief complaint of slow-growing mass since six months duration in the left hard palate. Based on the history, clinical, radiological and histopathological findings the mass was diagnosed as leiomyoma of hard palate. Immunohistochemistry was carried out for a more precise confirmation of the tumor, which showed positivity for SMA and vimentin. The mass was treated by extraction of the involved teeth followed by surgical excision of the tumor. No recurrence was noted during four month follow-up. Conclusion: Oral leiomyomas are very rare, with low recurrence rates. These benign lesions respond well to surgical excision of tumor mass. In this paper, we present a case of leiomyoma of hard palate in a 14 years old boy, along with a brief note on review of literature on leiomyoma.

Keywords: Leiomyoma, Oral leiomyoma, Smooth muscle tumor, Hard palate tumor

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INTRODUCTION

Leiomyomas are benign soft tissue tumors arising from the smooth muscle due to nodular proliferation of muscle cells. Primary leiomyomas of head and neck account for 12% of all leiomyomas [2]. It is more commonly seen in the uterine myometrium (95%), skin

(3%), gastrointestinal tract (1.5%) and less than 1% is seen in the head and neck region [3]. It rarely affects the oral cavity due to absence of smooth muscle except in the wall of blood vessels. Oral leiomyomas are rare (0.06%) and they mostly occur on the lips, tongue, palate and cheek region. Usually, they present as painless, slow growing masses. Occasionally they may cause pain, mobility of adjacent teeth and difficulty in chewing. Due to its unspecific clinical presentation diagnosis is made after histopathological study. Immunohistochemical studies offer a more precise diagnosis in these lesions. Surgical resection of the lesion is the treatment of choice. In this article, we report a case of oral leiomyoma of hard palate in a 14-year-old boy who reported to our department with a chief complaint of painless swelling in the palate.

CASE REPORT

A 14-year-old boy presented to our department with a chief complaint of slow-growing mass since six months in the left hard palate. It was initially small when noticed and steadily progressed to the present size (2x2 cm approx). It was a painless swelling and interfered during chewing food. The patient was otherwise healthy, with no systemic abnormalities and no deleterious or para-functional habits. Clinical examination revealed a sessile, well-circumscribed swelling in the left palate region extending from maxillary first premolar to maxillary first molar (Figure 1). Swelling extended onto the occlusal and buccal surfaces of maxillary second premolar. It was non-tender on palpation and soft in consistency. Overlying mucosa was reddish pink in appearance. There was slight cortical plate expansion in the region of maxillary second premolar. No cortical perforation was noted. Lymph nodes were not palpable.

We have advised fine needle aspiration biopsy and CBCT of upper left maxillary region. Aspiration results were negative, thus ruling out vascular and cystic lesions. CBCT revealed alveolar bone loss in relation to left maxillary second premolar and left maxillary first molar (Figure 2). The case was planned for excisional biopsy under general anesthesia owing to its size and anatomical location. Complete resection of the tumor mass along with the extraction of upper left second premolar and first molar was done under general anesthesia (Figure 3). The resected specimen (Figure 4) was sent for histopathological examination, which suggested the presence of numerous spindle shaped cells (Figure 5) with cigar shaped nucleus and endothelial cells in a sparse stroma without any atypia. To arrive at a more definitive diagnosis, we further subjected the specimen to immunohistochemical studies. The tumor exhibited strong positivity for SMA (Figure 6) and vimentin (Figure 7), thus confirming leiomyoma. No recurrence was noted during the four month follow-up period.

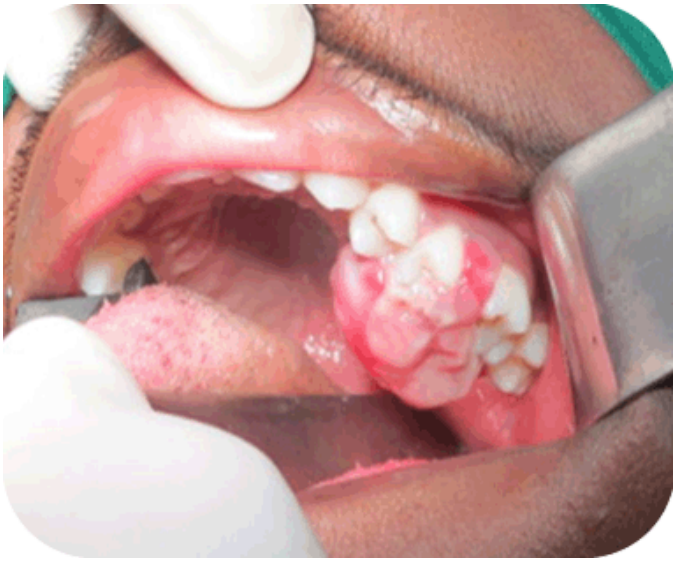

Figure 1: Clinical appearance of the tumor mass.

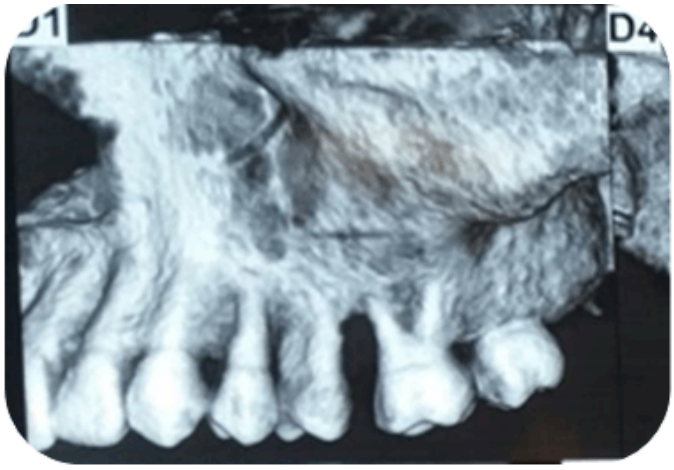

Figure 2: Cone beam computed tomography scan of maxilla showing bone loss around maxillary 2nd premolar and 1st molar.

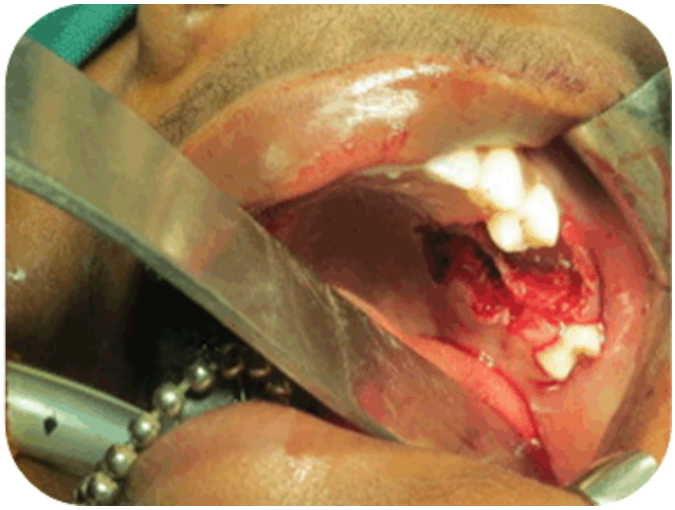

Figure 3: Surgical defect following tumor resection and extraction of involved teeth.

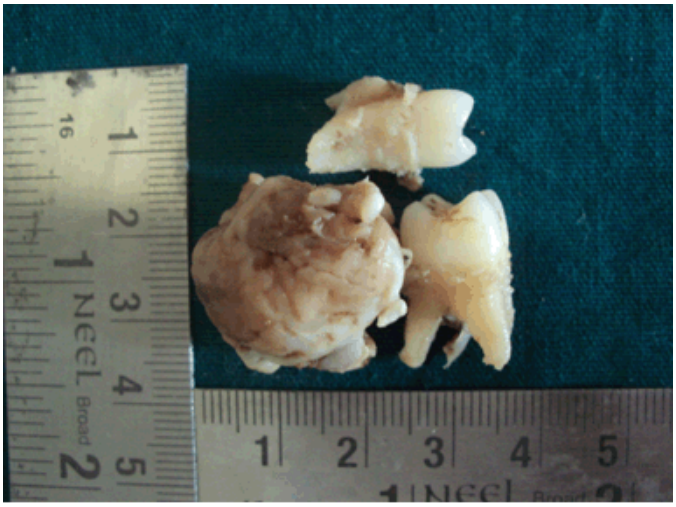

Figure 4: Resected tumor mass and extracted teeth after formalin fixation

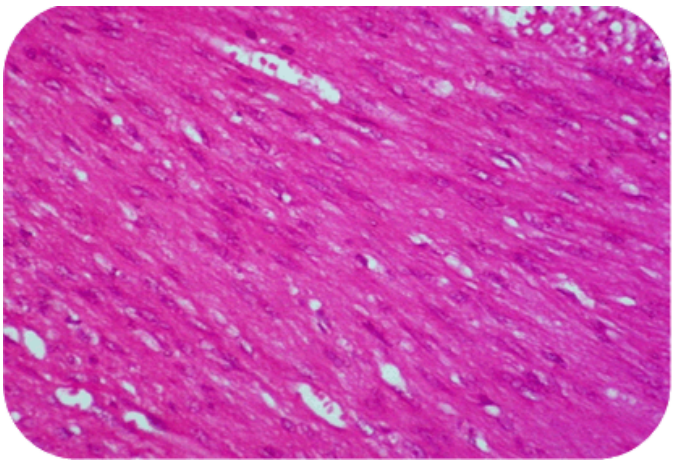

Figure 5: Numerous spindle cells with elongated nuclei (H&E stain, x400).

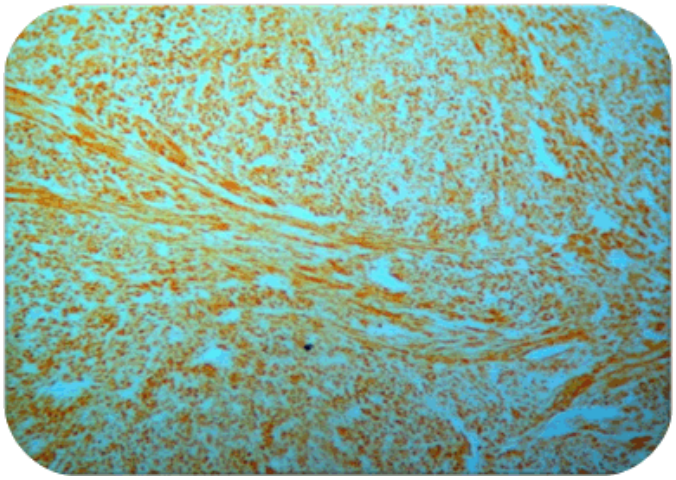

Figure 6: Immunohistochemistry showing positivity for SMA stain (Magnification, x100).

DISCUSSION

Leiomyomas are benign neoplasms that arise from smooth muscle. Virchow [4] in 1854 first described

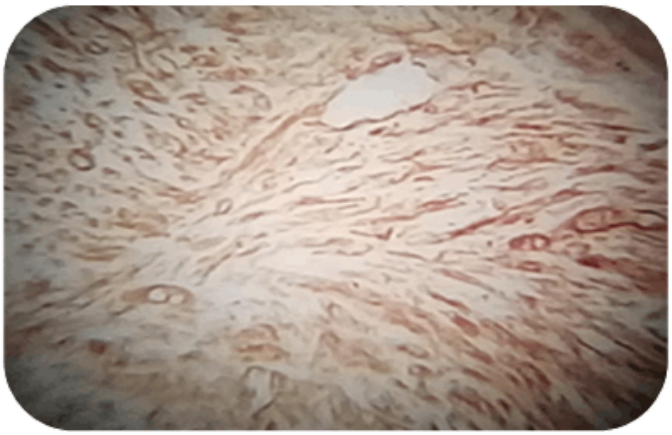

Figure 7: Immunohistochemistry showing positivity for vimentin (Magnification, x100).

this tumor. Although these are rare tumors of the head and neck region, when they occur, they are commonly seen in the oral cavity, nasal cavity and larynx [5]. The hereditary form causing multiple leiomyoomas was noted by Kloepfer et al. [6] in 1958. Smooth muscle tumors are relatively rare in the oral cavity, accounting for only 0.42% of soft tissue tumors because of the paucity of smooth muscle in this region [7]. The only source of smooth muscle in the oral cavity is tunica media as suggested by Scout [8], or the ductus lingualis or circumvallate papillae as suggested by Glass [9]. Brooks et al. reported that, the most frequently affected site was the lip (43.6%) followed by the palate (21.1%), buccal mucosa and tongue (each 9.2%), mandible (8.3%) and buccal and labial sulcus[10]. The WHO classified leiomyomas into three groups angioleiomyoma (74%), solid leiomyoma (25%) and epitheloid leiomyoma (1%). Leiomyoma differs from angioleiomyoma in the degree of angiogenesis [11]. In 1884, the first case of oral leiomyoma was reported by Blanc [12] in a 33-year-old male who presented with a large-tumor at the base of the tongue. Oral leiomyomas are benign neoplasms that present as small, solitary, asymptomatic nodular mass. Highest prevalence is seen in the 4th and 5th decades of life with slight male predilection [13] (1.43:1). However, few reports suggest slight female predominance of leiomyomas in the head and neck region and the authors attribute it to hormonal variation, i.e., progesterone receptor positive and estrogen receptor negative on immunochemical studies [14, 15]. Although most of these lesions are asymptomatic, few authors reported symptomatic lesions that are associated with pain, tooth mobility, difficulty in chewing and swallowing. Pain when occurs can be due to local ischemia causing intra-tumoral vasoconstriction or compression of a somatic nerve by the tumor mass [10]. The average size of these tumors as reported in literature is 1–2 cm with less than 1 year duration. The color of the lesion depends on depth and vascularity. Clinically, it is difficult to differentiate a leiomyoma from other mesenchymal tumors or its malignant counterpart. Hence,

the final diagnosis of leiomyoma is mainly determined by histopathological examination. Leiomyomas are well encapsulated lesions, typically composed of numerous spindle shaped/ fusiform mesenchymal cells arranged in whorls or strands. The nucleus is typically elongated and cigar shaped with eosinophilic cytoplasm on Hematoxylin and Eosin staining. Endothelial cells are seen lining the vascular channels. Unlike other mesenchymal tumors, leiomyomas lack dense fibrous stroma in between the individual mesenchymal cells. As the tumor cells mimic fusiform cells, simple Hematoxylin and Eosin staining cannot differentiate between leiomyomas and other spindle cell tumors. Special stains to identify collagen and muscle cells, such as Von Gieson stain, Masson trichromic acid stain, Mallory's phosphotungstic acid-hematoxylin (PTAH) stain can be used [16]. Von Gieson stain is recommended for muscle. Masson trichromic acid stains the cytoplasmic elements of smooth muscle cells red and collagen and fibroblasts blue or green. Myofibrils are stained by Mallory PTAH stain [17]. Additionally, immunohistochemical studies can be used for a more precise diagnosis. Specific monoclonal antibodies for actin (smooth muscle marker) confirm leiomyoma. Smooth muscle actin that corresponds to the alpha fraction of actin chain is an immuno-marker for smooth muscle but it can have immune reaction to skeletal muscle. S100, vimentin, desmin, antiCD-34 antibody are the other immunomarkers. Vimentin is a structural protein of cytoplasmic elements of mesenchymal cells whereas desmin is a type III intermediate filament near the Z line in sarcomere [18]. CD-34 is a transmembrane protein, expressed by the vascular endothelium and endothelial cells exhibit strong immunoreactivity against antiCD-34 antibody [19]. Leiomyoma should be carefully differentiated from its malignant counterpart i.e., leiomyosarcoma. Clinically, the presence of ulceration may be indicative of malignancy. Cotran [20] et al. described few histological features in uterine tumors, which were suggestive of malignancy like presence of more than 10 mitoses per 40x high magnification field, with or without cellular atypia or 5–10 mitoses per x10 magnification field with atypia . Tumors with 1–4 mitoses per x10 magnification, necrotic areas and atypias are best considered potentially malignant. Presence of fewer than two mitoses in x10 magnification indicates good prognosis. Molecular markers like PCNA, bcl-2, CDK-4, P53, MDM 2 indicate malignancy [21].

In our case, the resected tumor showed numerous spindle cells on Hematoxylin and Eosin staining arranged in parallel bundles with endothelial lining and no atypia. Hence, to differentiate it from other spindle cell tumors, immunohistochemical staining has been done. The tumor cells stained positive for vimentin and smooth muscle actin, thus confirming the diagnosis of leiomyoma. The treatment of choice is local resection with adequate safety margins. Leiomyomas are tumors of vascular origin, but bleeding after resection is not routinely seen. Benign smooth muscle tumors rarely relapse. Brooks et al. reported relapse of two cases , two weeks and nine months postoperatively following resection of hard palate leiomyomas [9].

CONCLUSION

Leiomyomas are benign tumors of smooth muscle origin. Oral leiomyomas are relatively rare. Definitive diagnosis is made after histological and immunohistochemical confirmation. Surgical resection of the tumor with adequate safety margins is the preferred treatment. Recurrence is rarely seen following excision in these tumors.

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Author Contributions

G. Siva Prasad Reddy – 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

B. Jagannadha Prasad – 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

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E. Padmini – 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.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Metastatic transitional cell carcinoma to the inguinal lymph nodes from an unknown primary: A case report

Ghassan Almaimani

ABSTRACT

Introduction: The carcinoma of unknown primary (CUP) is uncommon in the inguinal region, accounting for 1–3% of all CUPs. The diagnostic workup for CUP includes histopathological examination, imaging and, more recently, molecular testing. Nevertheless, the primary site frequently remains unknown. Case Report: Herein, we report a case of 78-year-old male who was referred for evaluation of a painless swelling in the right inguinal region. Histopathological examination of the biopsy and resection specimens revealed transitional cell carcinoma (TCC) but no primary site was identified despite thorough clinical and radiological evaluation. Conclusion: This is the first reported case of pure TCC of the inguinal lymph nodes presenting as a CUP. This may have occurred either from an unidentifiable primary lesion or the primary may have since regressed. Although there are no clear guidelines for the management of such patients, treatment must be multimodal and might include surgical resection and perhaps postoperative adjuvant radiotherapy to achieve optimal outcomes. Investigation with PET would have been useful in this case.

Keywords: Carcinoma of unknown primary, Inguinal lymph node, Transitional cell carcinoma

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INTRODUCTION

The carcinoma of unknown primary (CUP) is an uncommon diagnosis that is offered when diagnostic evaluation fails to identify the primary tumor site. 0.5–4% of all diagnosed cancers are CUPs [1, 2]. The CUPs vary in both presentation and histological type, but the most frequent presentation is as metastatic disease. The CUPs rapidly progress in most cases and often show atypical metastatic patterns for the presumed primary origin. The CUP is more common in the head and neck and axillary regions, with inguinal involvement accounting for only 1–3% of cases [3, 4]. Extensive workup with comprehensive histopathological evaluation (microscopy, immunohistochemistry, electron microscopy, and molecular testing where appropriate) and modern imaging (computed tomography (CT) scan, mammography, positron-emission tomography (PET)) has improved the diagnosis and identification of the primary site [5], which nevertheless remains elusive in most patients even at autopsy. Thus, the diagnosis and treatment of CUPs still challenge the multidisciplinary cancer team (surgeons, pathologists, and oncologists).

CASE REPORT

A 78-year-old male was referred for evaluation of a growing painless swelling in the right inguinal region that he had noticed eight weeks previously (Figure 1). He had a past medical history of left ventricular hypertrophy and hypertension. He reported no weight loss or gastrointestinal symptoms. There was no history of tobacco smoking, chemical exposure, or family history of cancer. Physical examination revealed a 10x10 cm painless mass in the right inguinal region but was otherwise normal.

All routine hematological and biochemical parameters were normal. Serologic tumor markers (CEA, lactate dehydrogenase, alpha-fetoprotein, prostate-specific antigen, human chorionic gonadotropin, CA125, and CA19-9) were within normal limits. An abdominal CT scan with contrast revealed two inhomogeneous masses anterior to the right rectus femoris (proximal 3.4x4.8 and 6.3x10.7; Figure 2) suggestive of enlarged lymph nodes. Chest CT and colonoscopy were normal.

The patient underwent punch biopsy of the inguinal mass, which was suggestive of poorly differentiated transitional cell carcinoma (TCC) (Figure 3A). The decision was made to perform tumor extirpation with right ilioinguinal lymph node dissection. Final histopathological examination confirmed TCC (Figure 3B-C), with immunoreactivity for CK7, CK20, and p16 but not thyroid transcriptase factor-1 (TTF-1), synaptophysin, and CK5/6 (Figure 3D).

A diagnosis of metastatic TCC in the right inguinal lymph node was made on clinical, histological, and immunohistochemical grounds. However, the primary site remained unknown even after a search for a primary tumor using pelvic magnetic resonance imaging (MRI), anorectal examination, and meticulous urological examination including cystoscopy. His inpatient stay was uncomplicated and he was discharged from hospital on the fifth postoperative day. He subsequently underwent adjuvant radiotherapy and received regular follow-up. At the most recent follow-up at 24 months there was no recurrence.

DISCUSSION

The carcinomas of unknown primary are defined as patients who present with histologically confirmed metastatic cancer for which no site of origin can be detected even after comprehensive investigations [1, 2]. Although the exact etiology of CUP is poorly understood, CUP is thought to either represent: (i) a concurrent but undetected primary cancer, or (ii) a spontaneously regressed primary cancer that was either misdiagnosed, ignored, or went unnoticed. The incidence of CUP has declined, mainly due to the evolution of new detection methods and diagnostic modalities to identify the primary site [5, 6].

Histologically, CUPs are categorized into four major subtypes by routine light microscopy:

a) well-moderately differentiated adenocarcinomas (50%)
b) undifferentiated or poorly differentiated adenocarcinomas (30%)
c) squamous cell carcinomas (15%)
d) undifferentiated neoplasms (5%) [1, 2]

Over 50% of CUP patients present with multiple sites of involvement, with a single site affecting the remainder, most commonly the liver, lymph nodes, peritoneum, lung, bone, and brain [1, 2].

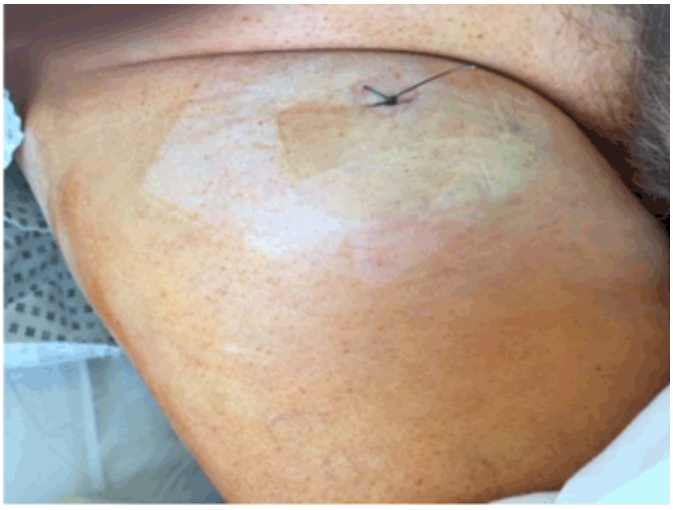

Figure 1: A 78-year-old male with a large right-sided inguinal swelling.

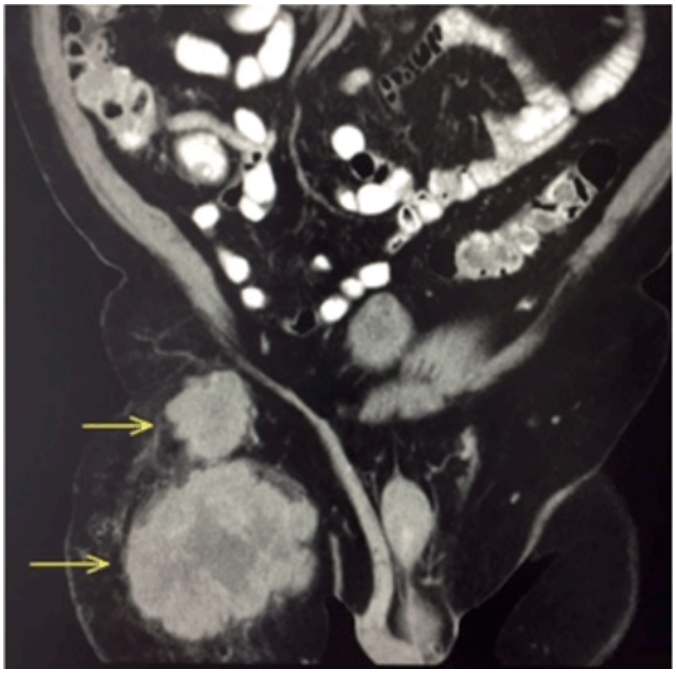

Figure 2: Coronal abdominal computed tomography scan of the abdomen at presentation showing two inhomogeneous masses in the right groin (yellow arrows).

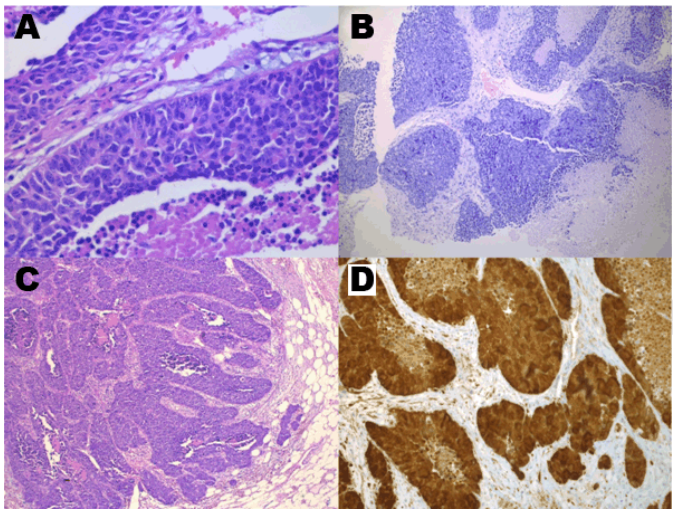

Figure 3: Histopathological examination of the biopsy and resection specimen. (A) Photomicrograph of the inguinal mass biopsy showing poorly differentiated transitional cell carcinoma (TCC) with focal necrosis (H&E stain, x40). (B, C) Photomicrographs of TCC of the inguinal lymph nodes (H&E stain, (B) x20 and (C) x20). (D) Photomicrograph showing tumor immunoreactivity for p16 (magnification: x20).

The inguinal area is a relatively uncommon metastatic site for CUP [7, 8]. The inguinal nodes are affected by a wide variety of metastases, mainly from the pelvis, genitalia, or lower limb but also more distant sites such as the nasopharynx, breast, tracheobronchial tree, salivary glands, and orbit [2, 7, 8]. The diagnostic evaluation of patients with CUP should be directed by the histopathological findings and be multi-modal. Recently, PET scans have proven to be a very useful imaging modality to detect the primary site, but cases in which the origin cannot be found are abundant [5]. In our case, contrast-enhanced CT scan of the abdomen and thorax were performed as PET-CT was not available.

Molecular tumor profiling (MTP) is a powerful diagnostic tool for identifying the primary site. However, validation of the accuracy and clinical value of MTP has been difficult because the anatomic primary site in most patients is never identified [9]. Zaren et al. examined over 2000 patients with metastases in the inguinal lymph nodes and reported that the primary site could not be identified in 22 (1%), whereas Guarischi et al. examined 56 similar patients in whom the primary site could not be identified at all [4, 8]. In our case, even after extensive attempts to find the primary site, the site of the primary tumor could not be determined.

The management of CUP needs to be individualized according to the clinical features. Surgery with adjuvant radiotherapy has been reported as the preferred treatment for inguinal metastasis from a CUP [10]. To our knowledge, this is the first report of pure metastatic TCC in the inguinal lymph nodes from an unknown primary site, although there is one report of mixed transitional and squamous cell carcinoma [7].

CONCLUSION

Carcinoma of unknown primary remains clinically challenging. In spite of new diagnostic tests, the optimal diagnostic algorithm for metastatic tumors with unknown primary has yet to be established. Carcinomas of unknown primary are an important group of tumors to be aware of because their progression differs from other malignant tumors and is difficult to predict. Since the benefits of current therapies are either limited or of uncertain benefit in many patients, the evaluation of novel treatments is essential.

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Author Contributions
Ghassan Almaimani – 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.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

A rare case of large fetal intracranial tumor: Teratoma

Chiranjeev Kumar Gathwal, Devender Kaur, Kulvinder Singh, Jyotsana Sen, Monika B. Gathwal, Parveen Rana Kundu, Gaurav Malik, Kalpana Beniwal

ABSTRACT

Introduction: Fetal intracranial tumors represent only 10% of all antenatal tumors; teratoma being the most common accounting for approximately half of all reported cases followed by astrocytoma, lipoma, choroid plexus papilloma, craniopharyngioma, and primitive neuroectodermal tumor. Usually these tumors clinically manifest in the third trimester of pregnancy, which otherwise nowadays often recognized earlier in pregnancy by ultrasonography and magnetic resonance imaging. Majority of fetal intracranial tumors are supratentorial, however in larger lesions imaging or even autopsy is frequently unable to determine the origin. Case Report: Here we are presenting a rare case of large fetal intracranial teratoma with which the patient presented in the third trimester with fatal outcome. Conclusion: Progress in technology has contributed to early diagnosis of congenital CNS tumors, but the same is not observed with fetal surgery, perhaps because the prognosis of fetal brain tumors remains poor. It is critical to have the most precise information regarding the tumoral nature and the extension of the lesion. Fetal medicine centers should be composed of a multidisciplinary team acting together to provide better assistance for fetuses with congenital CNS tumors and to develop new methods of treatment.

Keywords: Antenatal intracranial tumors, Immature teratoma

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INTRODUCTION

Diagnosis of any fetal abnormality is traumatic event for the family, the presence of a fetal intracranial tumor, along with mental trauma, carries with it additional diagnostic and therapeutic challenges. The prognosis is usually poor with few exceptions. Fetal intracranial

tumors represent only 10% of all antenatal tumors, ranking behind extra cranial teratomas, neuroblastomas, and soft-tissue tumors. Most of the congenital brain tumors arise from the pineal gland, suprasellar area, or cerebral hemispheres. Intracranial teratoma is the most common fetal brain tumor, accounting for approximately half of all reported cases followed in frequency by astrocytomas of varying grades, lipomas, choroid plexus papillomas, craniopharyngiomas, and primitive neuroectodermal tumor [1, 2]. Teratoma is a neoplasm containing tissues foreign to the site of origin and containing more than one embryonic germ cell layer. These tumors most commonly become clinically manifest in the third trimester of pregnancy. Diagnosis earlier in pregnancy, particularly before 30 weeks gestation, portends a particularly deleterious prognosis, with reported mortality rates as high as 96.9% [3].

CASE REPORT

A 22-year-old female G1P0A0L0 with 34 weeks gestational age (by LMP), presented to radiological department for routine antenatal ultrasound and ruling out any gross congenital malformation. As she belonged to rural background, she had not undergone any previous ultrasonography during the course of this pregnancy. On ultrasonography (Figure 1A–B), there was single live intrauterine fetus of 32 weeks gestational age with a large intracranial well defined lobulated heteroechoic lesion measuring 15.6(AP)x12.6(CC)x9.0(T) cm^3 epicentered in midline supratentorial region with right sided extension causing significant dilatation of B/L lateral ventricles (R>L) with surrounding parenchymal thinning. The lesion was large enough for the origin to be exactly defined, however, was limited to supratentorial compartment only with infratentorial compartment and its contents absolutely normal. There were multiple variable sized internal cystic changes with few small calcifications noted within the lesion. No other fetal abnormality was seen. On MRI scan (Figure 2) the lesion was large well defined lobulated of approximately same dimensions as on USG, iso-hypointense on T1/T2 sequences with multiple TI hypo and T2 hyperintense internal variable sized cystic changes with the status of obstructive hydrocephalus same as seen on ultrasonography. Tiny intralesional calcifications as seen on ultrasonography are not well appreciated on MRI scan. On the basis of above mentioned morphology of the lesion, antenatal diagnosis of fetal intracranial tumor possibly teratoma was given. With informed prognosis and proper counseling, termination of pregnancy was decided by parents. Cephalocentesis was performed (as the size of head was very large) followed by vaginal delivery. Parents refused for autopsy of delivered fetus and only the lesion extracted tissue was sent to pathology department for histopathological examination which confirmed the diagnosis of immature teratoma (Figure 3).

DISCUSSION

Fetal intracranial tumors though rare entity, nowadays are often recognized during antenatal period by ultrsound and magnetic resonance imaging, create significant medical and ethical dilemmas. An understanding of the different tumor types and their biologic behavior is necessary for appropriate counseling and care of these patients. Accurate diagnosis has important implications for fetal, maternal, and neonatal care [4]. Majority of fetal intracranial tumor are supratentorial in origin, arising mostly from pineal gland, suprasellar area, or cerebral hemispheres. Fetal intracranial tumors represent 10% of all antenatal tumors, teratoma being the most common accounts for approximately half of all reported cases [1, 2]. In literature, first report of a massive congenital intracranial teratoma was published in 1864 by Breslau and Rindfleisch [5]. Intracranial teratomas are generally large, complex, mixed cystic and solid masses with or without foci of calcification [1]. They typically arise in the midline, predominantly from the pineal gland. They may exhibit very rapid growth and reach massive proportions. Most fetuses die in utero or shortly after birth with only few exceptional long-term survivors [1–3]. They are most often diagnosed in the third trimester by ultrasonography and MRI scan. Often the masses are so large and locally invasive, replacing the intracranial contents, eroding the skull and extending into the mouth, orbit and neck that the precise anatomic point of origin cannot be determined [6].

At prenatal ultrasound, the diagnosis of teratoma should be considered for a complex intracranial mass with calcifications associated with gross distortion or replacement of normal brain tissue by the mass. The differential diagnosis for an ultrasonographically diagnosed large intracranial mass also includes astrocytoma, ependymoma, craniopharyngioma, choroid plexus cyst, and hemorrhage [7]. Intratumoral hemorrhage may also occur and thus in the setting of any fetal intracranial hemorrhage, underlying neoplasm should be considered. On color Doppler, there may be increased vascularity with low-resistance flow; leading to high-output cardiac failure and hydrops fetalis. Vascular flow is also a helpful finding to distinguish from hemorrhage. Computed tomographpy findings of intracranial teratoma include a large heterogeneous mass with coarse calcifications and hydrocephalus. Although the presence of calcifications on CT scan or ultrasound is a helpful diagnostic clue they are not frequently seen on MRI scan [8, 9]. On MRI, findings are highly variable, but often demonstrate a large lobulated multicystic heterogeneous signal mass. Magnetic resonance imaging scan is also helpful to determine the anatomic extent of the tumor, and to differentiate mass from intracranial hemorrhage [10 11]. Several recent case reports have described the use of fetal MRI scan between 25 and 36 weeks gestation in diagnosing intracranial teratoma, and the typical appearance is a large, heterogeneous

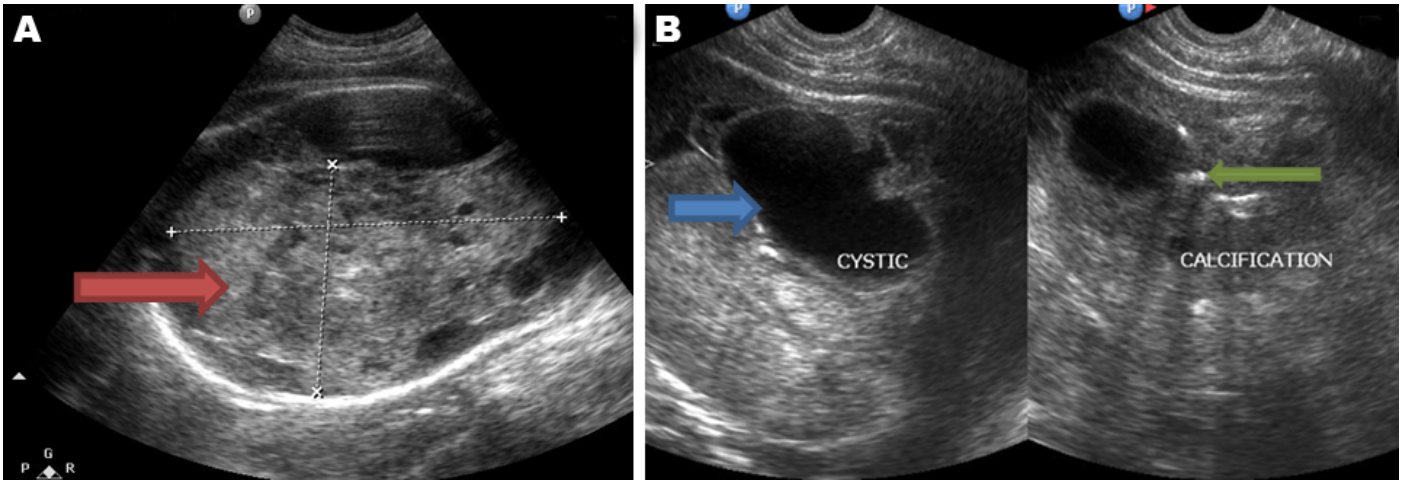

Figure 1: (A) Well defined large lobulated intracranial supratentorial heteroechoic lesion (red arrow) epicentered in midline with right sided extension causing significant dilatation of B/L lateral ventricles, (B) Intra-lesional variable sized internal cystic chnages (blue arrow) and small calcifications (green arrow).

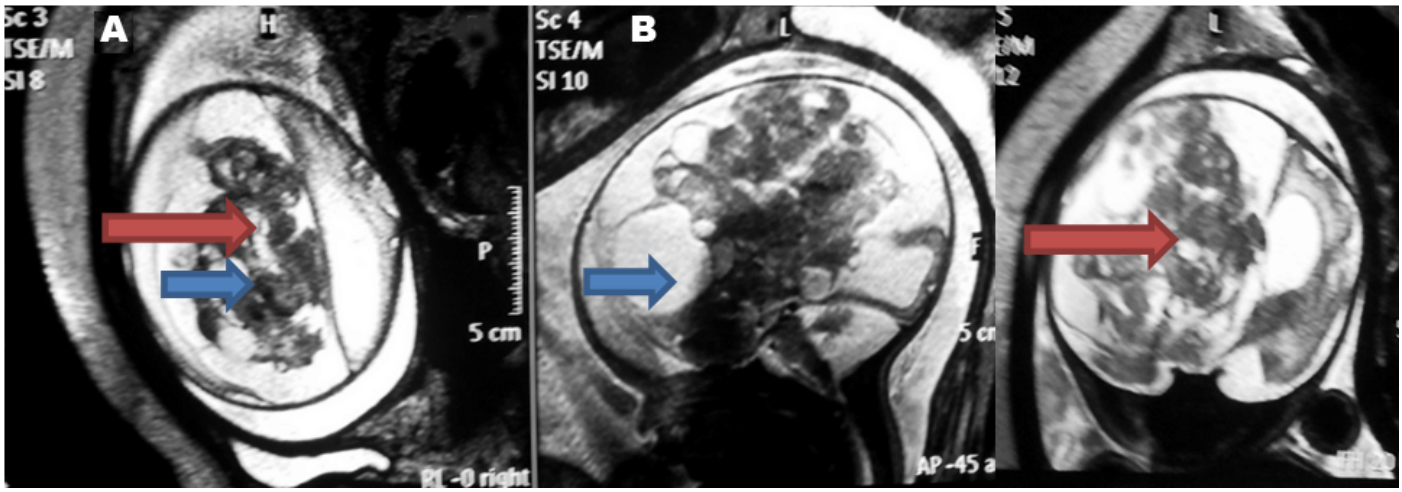

Figure 2: (A) T2 Axial with sagittal, and (B) T2 coronal shows well defined large lobulated intracranial supratentorial iso-hypoechoic lesion (red arrow) epicentered in midline with right sided extension causing significant dilatation of B/L lateral ventricles with intra-lesional variable sized T2 hyperintense internal cystic changes (blue arrow). Tiny calcifications are not well appreciated on magnetic resonance imaging.

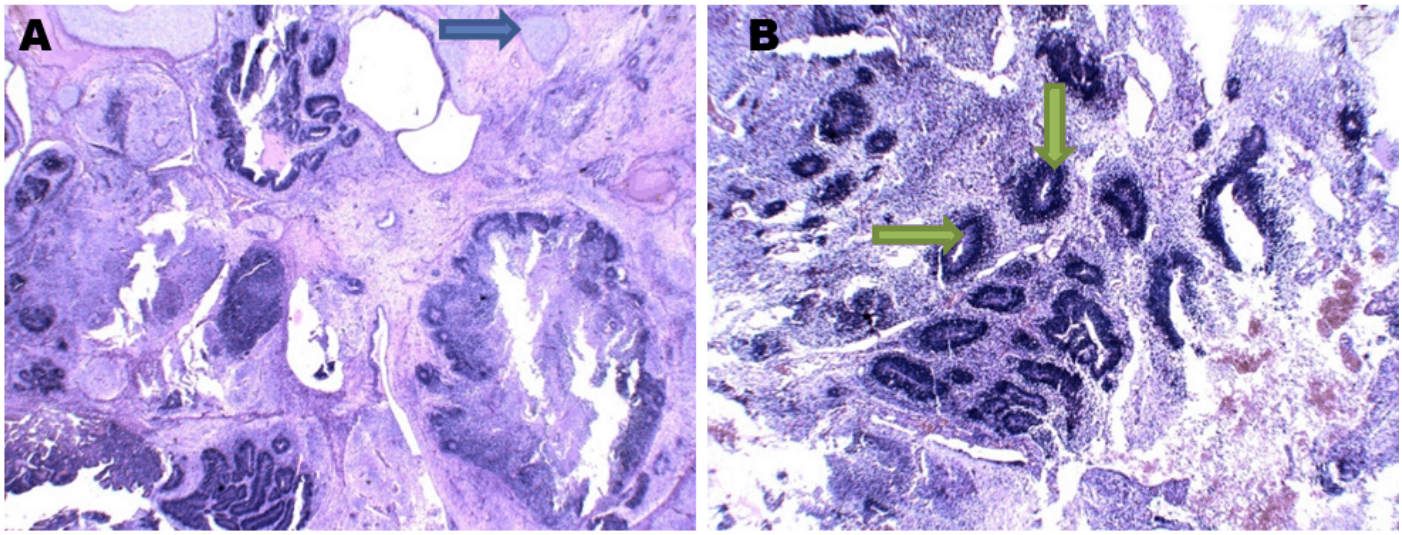

Figure 3: (A) Microphotograph showing immature neural tissue (red arrow), keratin pearl (black arrow) and nodules of cartilage (blue arrow) (H&E stain, x400), and (B) Immature neuroepithelial tubules (green arrow), (H&E stain, x100). Diagnosis: Immature teratoma.

mass with cystic components on MRI scan, with no apparent difference between mature and immature teratomas [10]. Similar features have been described at postnatal MRI scan of mature and immature intracranial teratomas. Fetal MRI scan allows enhanced global imaging of these masses. Anatomical relationships and tissue characteristics are demonstrated by MRI scan with superior detail, except for calcifications (which are better noted with ultrasound and CT scan). As the MRI features of teratomas are relatively nonspecific, the differential diagnosis of congenital supratentorial tumors should also include primitive neuroectodermal tumor, astrocytoma, ependymoma, glioma, craniopharyngioma, and choroid plexus papilloma [12, 13].

The prognosis for congenital intracranial teratoma is extremely poor whether benign or malignant, with an overall mortality rate of 90% rising to 97% if diagnosed prior to 30 weeks gestation [8, 9]. Progress in technology has contributed to early diagnosis of congenital CNS tumors, but the same is not observed with fetal surgery, perhaps because the prognosis of fetal brain tumors remains poor. It is critical to have the most precise information regarding the tumoral nature and the extension of the lesion. In addition, even when the parents do not opt to terminate pregnancy, the precise diagnosis provided by antenatal imaging may help healthcare professionals prepare them for the neonatal outcome. Fetal medicine centers should be composed of a multidisciplinary team acting together to provide better assistance for fetuses with congenital Central nervous system tumors and to develop new methods of treatment. If vaginal delivery is planned, cephalocentesis may be necessary [1]. Caesarean section may be required to prevent dystocia or, in cases of fetal airway obstruction.

CONCLUSION

To conclude, congenital brain tumors represent a diverse group of benign and malignant masses that often have a devastating effect on the fetus and the pregnancy. Each tumor is unique in its natural history, histologic characteristics, anatomic location, and prognosis; and thorough understanding of all these features is necessary to assemble the appropriate multidisciplinary team and to guide patient care. Although rare, there is increasing trend of detecting fetal intracranial tumors prenatally with the ever-increasing use of antenatal ultrasonography in routine obstetric screening. Fetal magnetic resonance imaging is a non-invasive, fast, highly informative examination and problem solving tool and has become a valuable adjunct to prenatal ultrasound in suspected fetal abnormality, especially for those with obesity or oligohydramnios. To our opinion, the use of prenatal magnetic resonance imaging in addition to ultrasound is a valuable tool in utero diagnosis and counseling for a large fetal intracranial mass.

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Author Contributions

Chiranjeev Kumar Gathwal – 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
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Kalpana Beniwal – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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The corresponding author is the guarantor of submission.

Conflict of Interest

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CLINICAL IMAGE PEER REVIEWED | OPEN ACCESS

A case of hepatic portal venous gas: When time is gold

Orlando Artavia, Ahmed Zedan

CASE REPORT

A 67-year-old male with history of type 2 diabetes mellitus, stroke, gastrointestinal bleed secondary to esophageal ulcer and distant cholecystectomy, was transferred to our hospital from outside facility due to one day of severe abdominal pain associated with nausea but no vomit.

Physical examination of the patient showed blood pressure 110/70 mmHg, heart rate 124 beats per minute, respiratory rate 20 breaths per minute, oxygen saturation of 95% on room air. He was awake, alert and oriented in three dimensions. Abdomen massively distended, with diffuse tenderness, rigidity, rebound and no bowel sounds on auscultation. He had a computed tomography scan of abdomen done which showed extensive amount of portal venous gas (Figure 1) associated with extensive dilated fluid-filled small bowel and diffuse pneumatosis of the intestinal wall (Figure 2). His lab work showed lactate 3.9 mmol/L, white blood cell count 20.8x103/µl with 22% of bands, hemoglobin 11.2 g/dl platelets 304x10^3/µl. Sodium 128 mmol/L, potassium 4.3 mmol/L, chloride 101 mmol/L, bicarbonate 15 mEq/L, creatinine 1.4 mg/dl, blood urea nitrogen 15 mg/dl, glucose 432 mg/dl, AST 47 U/L, ALT 28 U/L, total bilirubin 0.4 mg/dl, alkaline phosphatase 95 U/L, total protein 7 g/dl, albumin 3.4 g/dl, calcium 9 mg/dl, prothrombin time 14 sec, INR 1.1. The patient was taken to operating room urgently and exploratory laparotomy showed small bowel ischemia that required removal of 9 feet of medial small bowel, leaving 6 feet of it (3 feet of jejunum and 3 feet of ileum) with primary end to end anastomosis. He was then transferred to intensive care unit, was started on vancomycin and piperacillin/tazobactam, did not require vasopressors.

The patient had no complications during postoperative period, spent uneventful days on intensive care unit and was then transferred to medical floor and eventually discharged.

DISCUSSION

Hepatic portal venous gas (HPVG) is a rare imaging finding first described in 1955 in neonatal necrotizing enterocolitis by Wolf and Evans [1].

As new studies were done on the subject, it was noticed that HPVG was also present in the adult population in cases of bowel necrosis and had a high mortality rate of 90% [2].

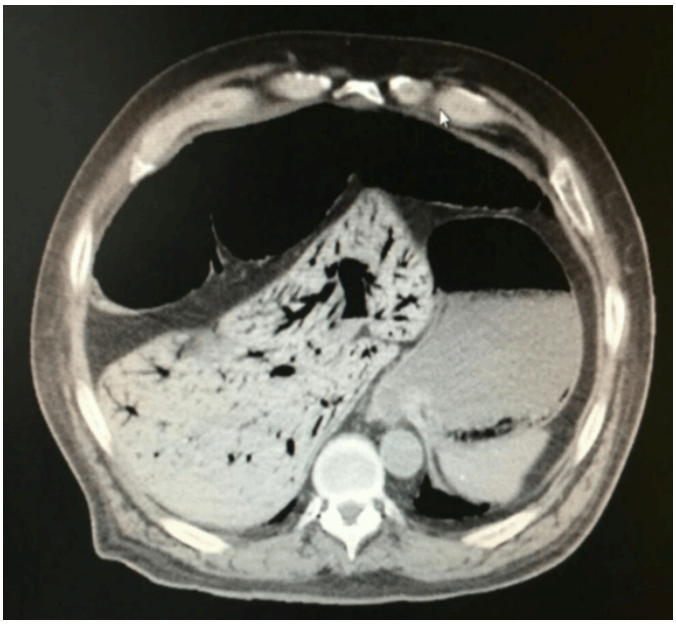

Figure 1: Computed tomography scan of the abdomen showing hepatic portal venous gas with branching pattern and a peripheral distribution.

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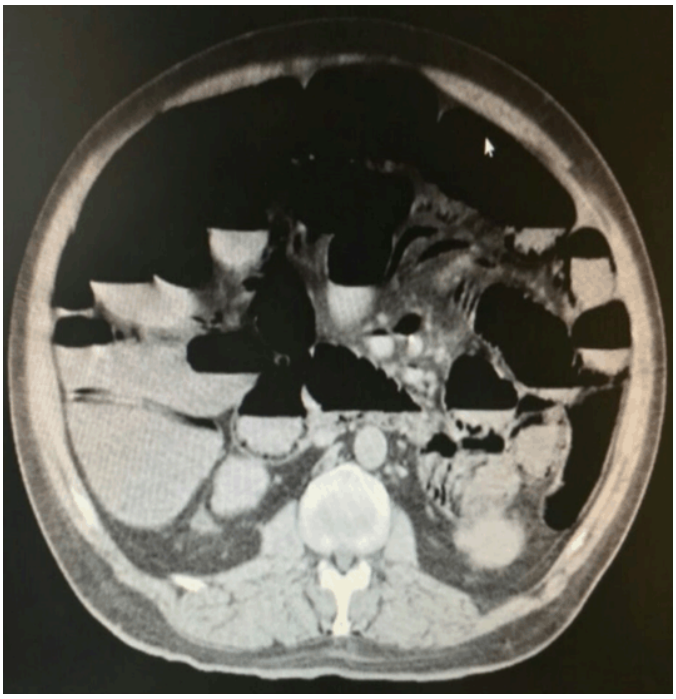

Figure 2: Computed tomography scan of the abdomen showing multiple air fluid levels with evidence of pneumatosis intestinalis at the level of small bowel.

Although HPVG may be diagnosed by conventional radiography, detection is difficult and it is easily overlooked, it fails to detect air in portal system in approximately 80% of cases [3]. The increased use of computed tomography (CT) scan has allowed early and highly sensitive detection of HPVG, and in relation to a wider range of etiologies, such as ulcerative colitis (8%), intra-abdominal abscess (6%), small bowel obstruction (3%) and gastric ulcer (3%), with bowel necrosis still being the most common one (72%) [2–8].

The earlier detection of HPVG and increased relation with other pathologies other than bowel ischemia has led to a decrease in its mortality rate. The earlier detection of it, made possible rapid intervention with laparotomy in cases of bowel ischemia as it happened in the case presented in this paper.

The HPVG is a rare radiological sign that has multiple etiologies. Bowel ischemia is the primary etiology of HPVG (70% of cases) and when associated, they are related with transmural necrosis in 91% of cases and to a high mortality rate (85% of patients) [5]. These facts signify that HPVG is an absolute indication for surgery in the context of mesenteric ischemia.

The pathophysiology of HPVG is not fully understood, the factors that seem to be related with it are intestinal ischemia resulting in damage to the mucosal barrier in association with over-distension of the bowel loops and gas-forming bacterial proliferation, all of these leading to gas moving from the intestinal lumen to the mesenteric veins and flowing through it to the portal system and hepatic parenchyma [2–5].

The composition of portal gas is rich in CO_2, which makes it a highly soluble gas that should rapidly dissolve in the circulation, the reasons by which it would remain in the circulation in cases of HPVG is the constant gas production and also increased intraluminal pressure forcing the intra-luminal gas towards the circulation, as HPVG has been reported as a complication of endoscopic balloon dilation [2, 3, 9].

The presence of HPVG, however, does not provide any information concerning the extent of bowel necrosis. In all cases, CT findings should be correlated with the clinical signs and with laboratory parameters to reach a high sensitivity and specificity level for intestinal necrosis [4], and in the cases where mesenteric ischemia is suspected, such as ours, rapid surgical intervention is required. Conservative management can be applied in settings where bowel ischemia is unlikely [7, 9].

Another important feature of HPVG is that it can be mistaken with gas in the biliary tract (pneumobilia). It is important to know how to differentiate them. The radiologic pattern of HPVG is described as branching radiolucency extending near the liver capsule, within 2 cm of it, due to the centrifugal pattern of blood flow, while gas in the biliary tract tends to remain in central portions of the liver due to the centripetal flow of the bile [3, 9].

CONCLUSION

Hepatic portal venous gas (HPVG) is a rare radiological sign for which computed tomography scan has a high sensitivity. Hepatic portal venous gas can be caused by different conditions, such as ulcerative colitis, intra-abdominal abscess, small bowel obstruction and gastric ulcer. It is, however, more commonly seeing in the setting of bowel ischemia (72%), in which case there is high mortality rate but a positive outcome is possible if good analysis of clinical presentation, laboratory data and imaging its done and prompt surgical intervention is performed.

Keywords: Distant cholecystectomy, Esophageal ulcer, Gastrointestinal bleed, Hepatic portal venous gas (HPVG), Severe abdominal pain, Small bowel ischemia

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Ahmed Zedan – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

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Conflict of Interest

Authors declare no conflict of interest.

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CLINICAL IMAGE PEER REVIEWED | OPEN ACCESS

Urinary bladder cancer showing surface calcification on computed tomography scanning

Yasuyuki Taooka, Yuka Ide, Gen Takezawa

CASE REPORT

An 84-year-old male who was ex-smoker consulted outpatient clinic complained of sudden-on-set of right hemiplegia. Cerebral magnetic resonance imaging scan showed early stage of lacunar infarction of left cerebral basal ganglia, and administration of anti-platelet agent was started. Two days after admission, patient noticed dark color change of urine. There were no other symptoms. Urinalysis revealed moderate occult blood, mild proteinuria, pH 8.5, but urine culture was sterile. Plain abdominal CT scan demonstrated flatter-shaped and round-shaped, tumorous lesion on the right lateral part of urinary bladder lumen (Figure 1 and Figure 2). The surface of tumorous lesion was irregular, and spotted calcification was recognized. Blood laboratory examination showed as following; C-reactive protein 0.4 mg/dL, white blood cell count 5,300/mL, hemoglobin 15.0 g/dL, thrombocyte count 193,000/ mL, CEA 3.0 ng/mL, SCC 1.3 ng/mL, PSA 0.8 ng/mL, albumin 4.2 g/dL, Ca 10.3 mg/dL, P 2.6 mg/dL, intact PTH 48 pg/ml (normal range: 10–65), and PTH related peptide was 1.3 pmol/L (normal range: less than 1.1 pmol/L). Although hypercalcemia was complicated, other symptoms consistent with paraneoplastic syndrome were not recognized. During the treatment of cerebral infarction, urine cytological examination was repeated and showed class V (transitional cell carcinoma mixed with squamous cell carcinoma). There were no metastatic other organs, and endoscopic tumor resection (transurethral resection of bladder) was performed. The size of cancer was 20 mm, and final pathological report was transitional cell

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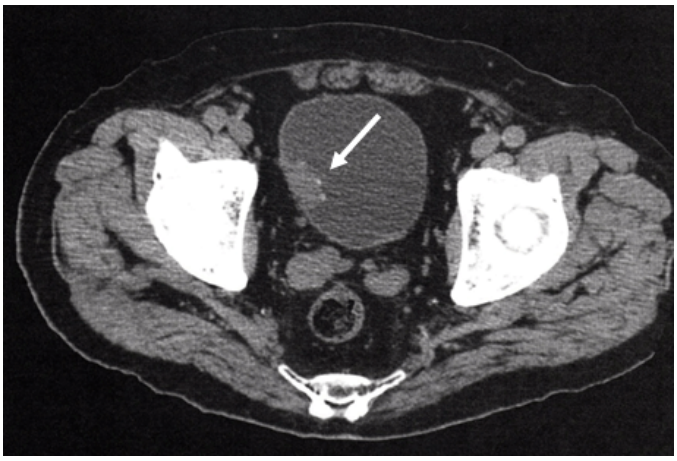

Figure 1: Plain computed tomography scan of pelvic. Arrow head showing surface calcification of urinary bladder cancer.

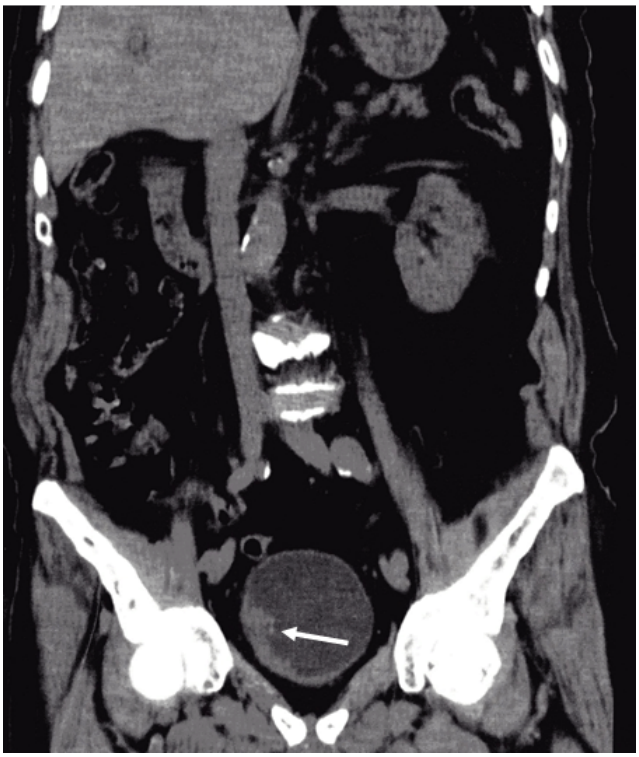

Figure 2: Coronal section of plain abdominal computed tomography scan. Arrow head shows urinary bladder cancer with spotted calcification.

carcinoma mixed with squamous cell carcinoma partially invaded into muscle layer of bladder wall (pT2, pL1, pV0). Grade of severity was G3. Since the patients was rather older and possible risk of cerebral infarction recurrence after surgery was high, surgical removal of whole urinary bladder did not selected as the therapeutic option. Then, additional radiation therapy against urinary bladder was performed. After that, serum calcium concentration recovered into normal ranged. At the last follow-up five years before, no recurrence was confirmed, and the patient was alive.

DISCUSSION

The incidence of urinary bladder cancer is still remained high, and smoking is known as risk factor [1]. But calcification with urinary bladder cancer like this case is rare. According to previous report, incidence of calcification with urinary bladder cancer was less than 1.0% [2, 3]. The characteristic finding of this case was as following, cell type was relative rare, transitional cell carcinoma combined with squamous cell carcinoma, and laboratory data showed hypercalcemia. As the cause of calcification in the present case, the involvement of hypercalcemia was speculated. Hypercalcemia-related paraneoplastic syndrome is known as one of complications of urinary bladder cancer [4]. Fortunately, this case did show complaints related hypercalcemia, and serum calcium concentration recovered into normal ranged after all the treatment of cancer. On the other hand, Moon et al. reported about calcification appearance in blabber cancer and pathological findings [5]. They reported that surface nodular or plaque-like calcification was seen in transitional carcinoma, and multiple fine punctate calcifications were seen in mucinous adenocarcinoma. As other reasons of tumor calcification, repeated urinary-tract infection and parasite infection induced bladder cancer were also reported, but in this case did not consisted with them [2, 3, 5–7].

CONCLUSION

A rare case of urinary bladder cancer was reported. When recognizing tumor with calcification in urinary bladder, possibility of complication with hypercalcemia should be considered.

Keywords: Calcification, Hypercalcemia, Urinary bladder cancer

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LETTERS TO THE EDITOR PEER REVIEWED | OPEN ACCESS

Schwannoma of the median nerve

Ingo Schmidt

To the editors,

A 57-year-old female presented with a two-year history of increasing painful swelling on the flexor surface in the left distal forearm that was associated with progredient paraesthesias in the peripheral area of the median nerve since six months. On physical examination, a non-tender mass was palpable shortly before entry into carpal tunnel, and the Tinel's sign along the distribution of median nerve upon percussion of the tumor was positive. Magnetic resonance imaging (MRI) scan revealed an oval and strictly demarcated tumor within the median nerve with a hyperintense signal relative to skeletal muscle (Figure 1A). The surgical procedure was done through a palmar incision over the carpal tunnel and extending up to the distal forearm with the use of a tourniquet. After that, the carpal tunnel was released. Intraoperatively, a strictly encapsulated tumor within the median nerve was seen (Figure 1B). The tumor was carefully dissected under microscope, followed by an extracapsular excision of the entire tumor with size of 1.5x0.5 cm. After that, excision-related fascicle lesions were not present (Figure 1C). Histological examination revealed a benign Schwannoma. Six months after surgery, the patient remained neurologically intact.

Schwannomas, also known as neurilemmomas, are benign nerve tumors that originate from the cells of the Schwann sheath, and constitute 5% of all benign soft tissue tumors. They are most commonly occur in the head and neck involving the brachial plexus and spinal nerves whereas the upper and lower limbs being affected less often. Schwannomas are the most common benign nerve tumors occurring in the upper extremity, the incidence for appearance in the median nerve is reported to be 14% [1]. They are usually found on the flexor surface of the forearm and hand, and multiple occurrences involving the median and ulnar nerve was observed [2]. Schwannomas of the upper extremity usually occur in patients aged 30–60 years, have no race or sex predilection, usually grow slowly for several years before being diagnosed; and they are sometimes misdiagnosed as lipoma, neurofibroma that grow usually intraneurally with infiltration of the nerve, ganglion or xanthoma [3]. Simple removal of the tumor after careful dissection is recommended if pure sensory deficit is present, because recurrence and/or malignant transformation rate is low [3, 4].

In literature, there is no strict consensus about whether the tumor should become extracapsular

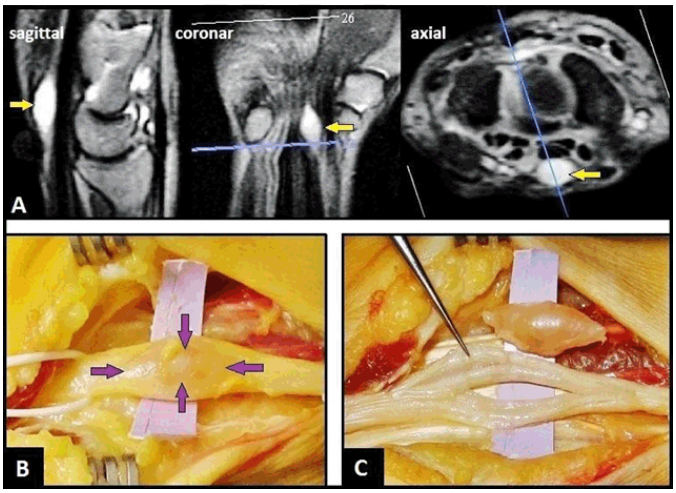

Figure 1: (A) Preoperative magnetic resonance imaging scan demonstrating hyperintense signal of the oval and strictly demarcated tumor within the median nerve, (B) Intraoperative clinical photograph showing the intraneural encapsulated Schwannoma before excision (arrows), (C) Intraoperative clinical photograph showing the complete excised Schwannoma without fascicle lesions after that.

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excised or intracapsular enucleated to avoid fascicle lesions intraoperatively [5, 6]. Even though there are some radiological features that point to the diagnosis of Schwannoma, ultrasonography and MRI scan cannot predict whether the tumor can be completely enucleated when a portion has fascicular involvement leading to residual neurological deficit after excision [4]. If there is a mixed and/or pure motor nerve involvement preoperatively that was found in 62.5% of patients, a sural nerve graft is recommended in younger patients; whereas it is not recommended since the level of neurological deficit is likely insignificant for the decreased level of activities of daily living in the elderly [4]. Recurrence has been seen in different areas of the nerves, but not in operated sites.

Keywords: Median nerve, Neurilemmomas, Schwannoma

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Author Contribution

Ingo Schmidt – 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.

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LETTERS TO THE EDITOR PEER REVIEWED | OPEN ACCESS

A 20-year follow-up after replantation of the right midfoot in an eight-year-old child

Ingo Schmidt

To the editors,

An eight-year-old girl sustained a traumatic amputation of her right foot in the metatarsal level due to a crush injury with an agriculture machine. The midfoot was replanted that was associated with distinctive debridements of soft tissue and metatarsal bones II–IV followed by temporary bony stabilization with the use of Kirschner wires. After that, multiple soft-tissue debridements on the dorsal aspect of midfoot were required (Figure 1), and the soft tissue defect was finally covered with split-thickness skin grafts. Four weeks after replantation, the fourth toe had to be surgically amputated due to a progredient avascular necrosis. Six months after replantation, the medial column of foot was stabilized with reconstruction of the second metatarsal bone loss using a 5-cm long non-vascularized autologous fibular graft that was subperiosteally harvested from her right distal lower leg. The defect of donor site showed completely bony regeneration six months after harvesting, and the fibular graft was completely osseointegrated (Figure 2A). Due to a progredient deviation of the fifth toe in medial direction, a fusion of the fifth metatarsophalangeal joint with use of a 2.0 mm titanium compression screw and a 2-mm titanium plate (Synthes) was required seven years after replantation, and the fusion was completely unioned without any complications (Figure 2A). Twenty years after replantation, a distinctive painful posttraumatic osteoarthritis of the first metatarsophalangeal joint was present, and a total joint replacement using the non-cemented TOEFIT-PLUS™ implant (Smith & Nephew) was performed (Figure 2B). Finally, despite posttraumatic decrease in length of the right foot due to growth disturbance there was a good functional and aesthetic result (Figure 3A), and the 28-year-old female is able to perform high-demand activities such as rock walking and alpine skiing in her leisure (Figure 3B).

Traumatic amputation injuries of the foot represent a challenging problem, and the success of replantation depends on duration of ischemia, microsurgical expertise, and quality in management of soft tissue complications in the presence of severe crushed wounds [1, 2]. In literature, only some case reports could be found in which successful replantations of the forefoot in adults or children have been described [3, 4]. In children, replantation of traumatically amputated forefoot should be always tried because the functional loss is low due to the extrinsically-related retaining of ankle functionality. The main problem after forefoot replantations in children is development of growth disturbance resulting in decrease of foot length that was observed in nearly all reported cases, however, this complication is well tolerated. To our knowledge, this

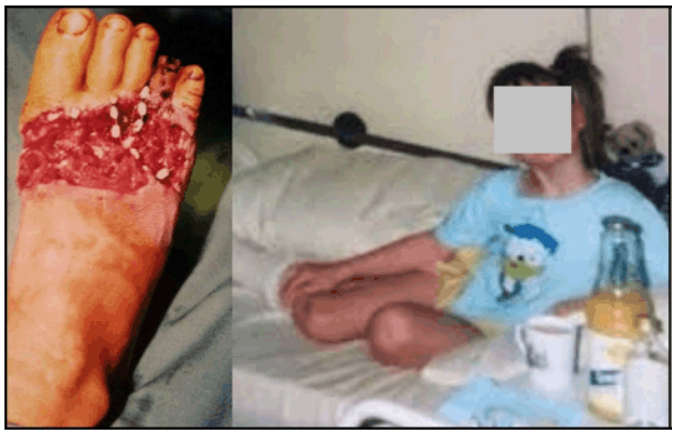

Figure 1: An eight-year-old girl two weeks after replantation of her right midfoot before wound coverage with split-thickness skin grafts and before required surgical amputation of her fourth toe.

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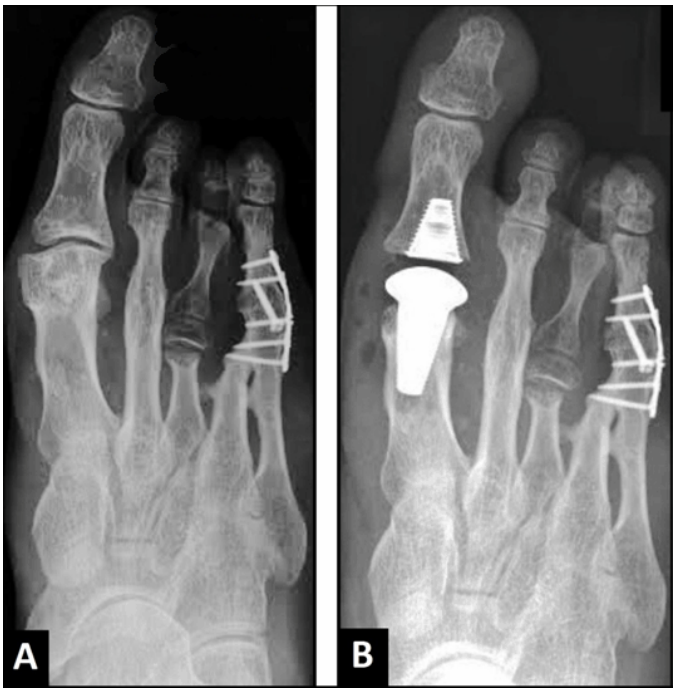

Figure 2: 20-year follow-up (A) Posteroanterior radiograph showing distinctive posttraumatic first metatarsophalangeal joint osteoarthritis, reconstructed second metatarsal with the 5 cm long non-vascularized autologous fibular graft, persistent bone loss of the third and fourth metatarsals distally, fusion of the fifth metatarsophalangeal joint without removal of implants, and posttraumatic synostoses between the third/fourth/fifth metatarsals, and (B) Posteroanterior radiograph showing first metatarsophalangeal joint replacement.

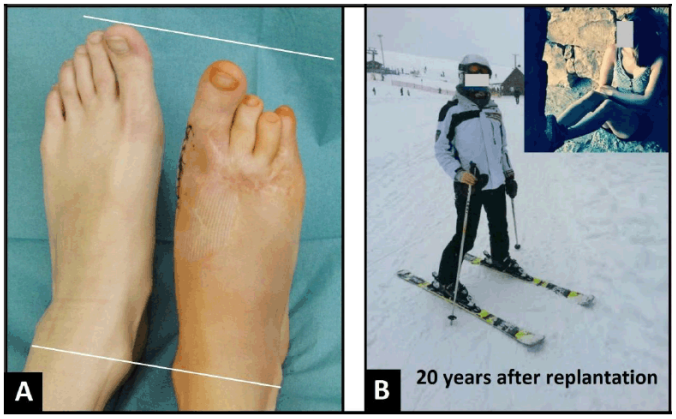

Figure 3: 20-year follow-up (A) Clinical photograph of both foots five days after first metatarsophalangeal joint replacement right demonstrating good aesthetic result despite decrease of foot length (lines), and (B) The 28-year-old female is able to perform high-demand activities in her leisure.

is the first case report that describes a successful midfoot replantation in a child with a 20-year follow-up.

The use of non-vascularized autologous fibular grafts for reconstruction of bony defects was first reported in 1911 [5], and has proven to be a suitable and reliable option. The main advantage is that this procedure does not need microsurgical expertise. It is important that during fibular harvest, the proximal and distal 5 cm are preserved to prevent injury to the common peroneal nerve and to retain the stability of the ankle joint [6, 7], and bony defects with an average length of 7 cm and maximum length up to 15 cm can be reconstructed [8, 9]. When the fibular graft is harvested subperiosteally in children, complete bony regeneration of the donor site is always observed [10].

For treatment of metatarsophalangeal joint osteoarthritis, total joint replacement is the motion-preserving alternative to total joint fusion. The non-cemented TOEFIT-PLUS™ is one implant that is currently in use, but still controversial, and should be indicated restrictively in particular in cases when first ray insufficiency is present [11]. When using this implant, it must be noted that there is high surgical revision rate with 24% at a mean of 33 months postoperatively [12]. For failed metatarsophalangeal joint replacement, metatarsophalangeal joint fusion or an excisional arthroplasty are the salvage options [13].

Keywords: Metatarsophalangeal joint osteoarthritis, Non-vascularized autologous fibular bone graft, Replantation, Total joint replacement, Traumatic midfoot amputation

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Author Contribution

Ingo Schmidt – 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.

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