EDITORIAL BOARD
International Journal of Case Reports and Images (IJCRI)

Dr. Achuta Kumar Guddati USA Dr. Hua Zhong USA
Dr. Aditya Gupta USA Dr. Ho-Sheng Lin USA
Dr. Adriar Arboix Spain Dr. Imtiaz Wani India
Dr. Adriana Handra-Luca France Dr. James Cheng-Yi Lin Taiwan
Dr. Afra Hadjizadeh Iran Dr. Jonathan D. Solomon USA
Dr. Ahmed El Said Lasheen Egypt Dr. Kyuzi Kamo Japan
Dr. Ali Soltani USA Dr. Luca Bertolaccini Italy
Dr. Altacilio Aparecido Nunes Brazil Dr. Makoto Adachi USA
Dr. Amin F Majdalawieh UAE Dr. Mehmet Uludag Turkey
Dr. Athanassios Tsakris Greece Dr. Mehmet Inal Turkey
Dr. Antonio La Cava USA Dr. Mohamed Radhi USA
Dr. Asher Bashiri Israel Dr. Mohannad Al-Qudah Jordan
Dr. Aziz Mustafa Kosovo Dr. Morikuni Tobita USA
Dr. Christopher CK Ho Malasiya Dr. Naila Khalil USA
Dr. Claudio Feliciani Italy Dr. Nataliea Semiletova USA
Dr. Daniela Cabibi Italy Dr. Oner Dikensoy Turkey
Dr. Deepa Rastogi USA Dr. Ozlem Guneysel Turkey
Dr. Deepak Sharma USA Dr. Paolo Cardelli Italy
Dr. Emre Karasahin Turkey Dr. Paul Rea UK
Dr. Federico Bizzarri Italy Dr. Parijat Saurav Joy USA
Dr. Gavin A. Falk USA Dr. Petru Matusz Romania
Dr. Gerardo Gomez-Moreno Spain Dr. Pengcheng Luo China
Dr. Gil Atzmon USA Dr. Piyaray Lal Kariholu India
Dr. Giovanni Leuzzi Italy Dr. Piraye Kervancioglu Turkey
Dr. Giovanni Tuccari Italy Dr. Radhika Muzumdar USA
Dr. Gokulakkrishna Subhas USA Dr. Rajesh Parea USA
Dr. Guo Wei USA Dr. Ranjan Agrawal India
Dr. Hajimi Orita Japan Dr. Ranjan Cui China

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

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

Disclaimer
Neither International Journal of Case Reports and Images (IJCRI) nor its editors, publishers, owners or anyone else involved in creating, producing or delivering International Journal of Case Reports and Images (IJCRI) or the materials contained therein, assumes any liability or responsibility for the accuracy, completeness, or usefulness of any information provided in International Journal of Case Reports and Images (IJCRI), nor shall they be liable for any direct, indirect, incidental, special, consequential or punitive damages arising out of the use of International Journal of Case Reports and Images (IJCRI) or its contents. While the advice and information in this journal are believed to be true and accurate on the date of its publication, neither the editors, publisher, owners nor the authors can accept any legal responsibility for any errors or omissions that may be made or for the results obtained from the use of such material. The editors, publisher or owners, make no warranty, express or implied, with respect to the material contained herein. (http://www.ijcasereportsandimages.com/disclaimer.php)
Contents

Cover Image

Magnetic resonance imaging scan of the brain showing a vermian hemangioblastoma associated with 2 small right cerebellar hemangioblastomas.

Case Series

391 Anomalous origins of a coronary artery from the pulmonary artery: A series of three case reports undetected into adulthood
Katie L. Mastoris, Ataul Qureshi, Navin K. Subrayappa, Matthew W. Martinez, James Wu

Case Report

396 An unusual association of von Hippel–Lindau disease and posterior nutcracker syndrome: Management of neurological and urological aspects in a young male
Nadhir Karmani, Faouzi Mallat, Wissem Hmida, Khaled Ben Ahmed, Oussama Karmani, Sidiya Oueld Chavey, Amel Ben Abdallah, Faouzi Mosbah, Hedi Krifa

403 1,25-Dihydroxyvitamin D-mediated hypercalcemia in a patient with malignancy without lymphoma
Katie Mastoris, Sarah Park, Vasudev Magaji

407 A case report of an uncommon large size of prostatic cyst
Jingjin Yang, Xingkai Liu, Yong Zhang

411 Coincident retroperitoneal and sigmoid colon liposarcoma: A rare occurrence
Ayvaz Ulaş Urgancı, Erkan Oymaci, Enver Vardar, Ebru Aıkınçlar, Ömer Engin

416 Scrub typhus vasculitis causing pan-digital gangrene
Suja Lakshmanan, Krishnamoorthy Seetharaman, Ramakrishnan S.R., Sathiyan Sivanandam, Koushik A.K.

422 Adult nephroblastoma

427 Eruptive Collagenoma in a mongol girl: A rare association
Balwinder Kaur Brar, Mahajan B. B., Nidhi Kamra

431 Single coronary artery arising from the right coronary sinus with mid-left anterior descending artery segment courses through the ventricular myocardium: A rare entity
Andrea Romagnoli, Irene Coco, Dominique De Vivo, Eros Calabria, Giovanni Simonetti

436 Cannabis consumption before surgery may be associated with increased tolerance of anesthetic drugs: A case report
Georg Richtig, Götz Bosse, Friederike Artt, Christian von Heymann

440 Primary hydatid cyst of gallbladder: A case report
Proney Kabiraj, Shib Shankar Kuiri, Utpal De

444 Occult hip fracture diagnosed by MRI scan after inconclusive X-ray and CT scan
Bogdan Deleanu, Radu Prejbeanu, Florin Birsasteanu, Dinu Vermesan, Liviu-Ionut Micle, Eleftherios Tsiridis, Vlad Predescu

448 Intramedullary spinal cord abscess by Nocardia: A case report
Khaleed Alishabani, Joseph Adrian L. Buenosalido, Milagros P. Reyes, Ayman O. Soubani

Clinical Images

454 Perianal aggressive angiomyxoma in a male patient
Zainab Taha ALHumoud, Najla Aldaoud, Hussain Abrar, Amro Salem

457 Bowen’s disease involving the dorsal and volar aspects of left thumb: An unusual site diagnostic entity
Shagufia Rather, Peerzada Sajad, Iffat Hassan

Editorial

460 Management of Inflammatory breast cancer: current concepts
Awad Ali M. Alawad
Anomalous origins of a coronary artery from the pulmonary artery: A series of three case reports undetected into adulthood

Katie L. Mastoris, Ataul Qureshi, Navin K. Subrayappa, Matthew W. Martinez, James Wu

ABSTRACT

Introduction: An anomalous origin of the coronary artery from the pulmonary artery (ACAPA) is a rare congenital anomaly resulting in sudden death in 90% of infants during their first year of life. Diagnosis in living adults is particularly unusual, especially for left coronary artery arising from the pulmonary artery (ALCAPA) given the large perfusion distribution that the left system provides to the myocardium. Surgery to correct and restore a bi-coronary arterial circulatory system has historically been the standard of treatment given the lethality of the diagnosis. However, advances in coronary imaging have led to an increased incidence of diagnosis in adulthood and challenges the true association between sudden death and ACAPA. This makes the decision for surgical correction more complex. Case Series: Herein, we presented three cases of coronary anomalies arising from the pulmonary artery that remained undetected until adulthood, only one of which was managed surgically. Conclusion: Currently, the prevalence of ACAPA in adults is low. Hence, there is no strong evidence based recommendations for management of adult patients. With advances in diagnostic modalities and potentially an increase in reported ACAPA cases, more data will become available to help guide the decision of surgical versus medical management.

Keywords: Anomalies, Cardiac surgery, Chest pain, Coronary vessel, Pulmonary artery

INTRODUCTION

Anomalous origins of the left or right coronary artery from the pulmonary artery (ALCAPA or ARCAPA) are rare congenital anomalies. Sudden death from myocardial infarction and congestive heart failure occurs in 90% of infants with this anomaly within the first year of life [1, 2]. For the few that survive into adulthood by way of collateral systems, ALCAPA/ARCAPA may be an important cause of sudden cardiac death. The pathophysiology between the two diseases is similar [2]. However, ARCAPA in adults has been reported more commonly than ALCAPA [3]. This distinction is likely due to the large perfusion distribution that the left system provides to the myocardium. Regardless, both diseases have remained rare diagnoses in adults [3, 4]. We present three cases of coronary anomalies arising from the pulmonary artery that remained undetected until adulthood, which may alter the current correlation...
between coronary anomalies, sudden cardiac death and the immediate need for surgical correction.

CASE SERIES

**Case 1:** A 48-year-old Caribbean male was admitted to the hospital after experiencing chest pain and progressive shortness of breath over a three-month period. Initial ECG illustrated left ventricular hypertrophy with ST-T wave changes compatible with repolarization abnormality. This patient had an abnormal adenosine myocardial perfusion imaging study demonstrating a moderate perfusion abnormality in the inferobasal wall, which prompted a cardiac catheterization. Catheterization verified a dilated left coronary system in the absence of the right coronary artery ostium. Subsequent imaging with a cardiac MRI confirmed ARCAPA (Figure 1) along with a hyperdynamic left ventricle and severe left ventricular hypertrophy. The right ventricle was within normal limits for contractility and no ischemic scars were noted. Three months later, the patient underwent surgical correction to successfully restore a bi-coronary arterial circulatory system (Figure 2). Intraoperative findings were consistent with ARCAPA however significant right ventricular dysfunction and distension, particularly at the intersection of the pulmonary artery, was now noted. The right coronary artery measured 8 mm in diameter with a great deal of collaterals surrounding the heart on the epicardial surface. The surgery consisted of dissecting the right coronary artery from the pulmonary artery and re-implanting onto the aorta. The patient was discharged home after a normal postoperative course. His anginal chest pain completely resolved however some residual shortness of breath continued for the first few months postoperatively.

**Case 2:** A 49-year-old female, suffering from recurrent bilateral breast cancer and awaiting bilateral mastectomies, experienced right-sided chest pain that radiated to her axilla and right arm. Nuclear stress test demonstrated normal diaphragmatic attenuation artifact with no obvious signs of ischemia. Given the magnitude of the surgery planned, a cardiac catheterization was performed and displayed a very large, right coronary artery and a left main artery, which could not be cannulated. A cardiac CTA identified an ALCAPA and a diffuse ectatic coronary vessel with extensive collateral vessels throughout the myocardium and communicate between the right coronary artery and the left coronary artery (Figure 3). Echocardiogram evaluated normal regional wall motion of both the left and right ventricles with normal systolic function. Given the extensive collateral vasculature it was felt that the patient was not at an increased risk for surgery. She underwent the planned bilateral mastectomies successfully without any cardiac complications. Currently, the patient is being managed by a cardiologist; no surgical correction is planned at this time.

**Case 3:** A 63-year-old female with an extensive cardiac history including childhood myocardial infarctions, congestive heart failure, and a presumed diagnosis of endocardial fibroelastosis presented with left shoulder pain radiating to her upper back. She underwent a nuclear stress test, which was abnormal, and an echocardiogram, which showed decreased left ventricular systolic function with heavy calcification of the endocardium in the apical lateral, anterolateral and inferolateral segments. The patient had subsequent cardiac CTA, which showed the presence of ALCAPA with a scarred anterior and anterolateral wall resulting in hypokinesis (Figure 4). Moderate central mitral regurgitation was also identified. Although surgical intervention was initially recommended, the patient declined at the time of diagnosis. She is currently being managed in collaboration with an adult congenital heart disease specialist who also recommended closure of the anomalous left coronary artery through catheterization to prevent the steal phenomenon. The patient however remains satisfied with her overall level of functioning and symptoms and has declined interventions. She continues to be monitored for cardiac arrhythmias.

**DISCUSSION**

An anomalous origin of the coronary artery from the pulmonary artery (ACAPA) presenting into adulthood remains a scarce diagnosis as over 90% of patients die within their first year of life [1, 2]. During fetal life, the pulmonary arterial pressure equals the systemic pressure allowing for antegrade flow into the anomalous coronary artery and perfusion of the myocardium. After birth,
however, the pressure in the pulmonary artery decreases and retrograde flow into the anomalous coronary artery ensues. This is known as coronary steal phenomenon and results in myocardial ischemia, infarction and sudden cardiac death [2].

The proposed mechanism of both ARCAPA and ALCAPA survival into adulthood involves significant collateral circulation to the myocardium originally supplied by the anomalous vessel. ARCAPA has more commonly been reported in adults and thought to be less fatal in infancy than ALCAPA due to the smaller perfusion distribution supplied by the right coronary artery compared to the left [3, 4]. ARCAPA’s overall incidence is estimated to be 0.002% compared to 0.008% for ALCAPA [3].

With advances in technology, the modality of diagnosis has changed over time from autopsy to surgery to catheterization and now angiography [5, 6]. These modalities have led to an increased number of reported adult cases of ACAPA. Many have postulated that the actual prevalence in adult population may be higher than reported and therefore the true association between sudden death and ALCAPA/ARCAPA is lower than described [2].

Historically, due to its association with myocardial infarct, heart failure, ventricular arrhythmias and sudden cardiac death, the standard treatment has been surgical correction [2, 7]. Limited data is available regarding the most effective surgical strategy, however, establishment of a dual coronary artery system with coronary transfer is preferred even if interposition graft is required [8]. Traditionally, plans for surgical correction before the onset of left ventricular function was favored. Owing to the possible lower association between sudden death and ACAPA in adults, the decision for a challenging surgical intervention becomes more complex. Recent literature has suggested that if mild to moderate chronic ischemia and limited necrosis is present, survival without surgical correction is possible [2]. There is even less evidence based recommendations for the medical management of ALCAPA and ARCAPA. Theoretically, optimizing risk factors for acute coronary syndrome and preserving collateral flow should delay the onset of the coronary steal phenomenon.

Figure 2: Intraoperative visualization of severely dilated coronary artery vessels in ARCAPA patient prior to successful surgical restoration of bi-coronary artery system.

Figure 3: Contrast-enhanced coronary computed tomography volume rendered image of the entire heart demonstrating the severely dilated coronary artery vessels in our patient with ALCAPA.

Figure 4: Cardiac CTA in the axial plain illustrating the enlarged coronary artery arising from the pulmonary artery trunk.
Data remains limited on adult manifestations of ALCAPA and ARCAPA and its true association between sudden cardiac death, progression to severe cardiac pathology, and treatment modalities. Our cases represent three symptomatic adult manifestations of ACAPA; only one of which underwent recommend surgical correction to restore a bi-coronary arterial circulatory system. The latter two preferred monitoring and referral to an adult congenital specialist for further guidance and management.

CONCLUSION

Currently, there are no strong evidence based recommendations for management of adult patient’s with anomalous origin of the coronary artery from the pulmonary artery (ACAPA) and strong collateral circulation due to the low prevalence of the anomaly. Our hope is that with advances in diagnostic modalities and an increase in reported ACAPA cases, more data will become available to support the decision of surgical or medical management.

Acknowledgements

Anil Gupta M. D- the referring provider from Pocono Medical Center, Elizabeth Adams D. O. – ADCH specialist at Hershey

Author Contributions

Katie L. Mastoris – 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
Ataul Qureshi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Navin K. Subrayappa – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Matthew W. Martinez – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
James Wu – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

An unusual association of von Hippel–Lindau disease and posterior nutcracker syndrome: Management of neurological and urological aspects in a young male

Nadhir Karmani, Faouzi Mallat, Wissem Hmida, Khaled Ben Ahmed, Oussama Karmani, Sidiya Oueld Chavey, Amel Ben Abdallah, Faouzi Mosbah, Hedi Krifa

ABSTRACT

Introduction: Von Hippel–Lindau (VHL) disease is a complex and systemic entity that can be discovered by neurological complications. Its association with other serious malformative disease like nutcracker syndrome (NCS) was not described, and its complex presentation must not make some other serious problem easily missed or neglected. Case Report: A 27-year-old male patient with 3 intramedullary hemangioblastoma of six months of insidious evolution complicated by medullary compression. The lesions were completely removed with excellent neurological postoperative outcome. Thinking of VHL disease, our investigations were expanded and revealed multiple vermian and cerebellar hemangioblastomas, renal masses, and multiple pancreatic cysts. In addition, an incidentally found of posterior NCS during the abdominal computed tomography (CT) done for VHL disease investigations. Our attitude to both neurosurgical and urological problems (the serious presentation of VHL disease with multiple locations of hemangioblastoma and right renal carcinomas) and for the missed symptomatic posterior nutcracker syndrome were discussed.

Conclusion: The complex presentation of the VHL disease must not makesome other serious problems easily missed or neglected like renal carcinoma or nutcracker syndrome as seen in our case.

Keywords: Hemangioblastoma, Nutcracker syndrome, Renal carcinomas, Total removal, Von Hippel–Lindau (VHL) disease

How to cite this article

doi:10.5348/ijcri-201565-CR-10526

INTRODUCTION

Hemangioblastoma is a rare tumour of the central nervous system characterised by a high vascularization. It accounts for 1.5–2.5% of spinal cord tumors. In the case of von Hippel–Lindau (VHL) disease, hemangioblastoma occurs in 20–30% of patients and is often multiple [1, 2]. Surgical experience is often limited because of the rarity of this tumor. Microsurgical removal is recommended in symptomatic patients or in cases with tumor growth during follow-up [3, 4].

The association of VHL disease with other serious malformative disease like nutcracker syndrome was not described. Herein, we present the case of young male having complicated presentation VHL disease with an incidentally radiological found of a missed symptomatic posterior nutcracker syndrome.
CASE REPORT

A 27-year-old male was hospitalized at the neurosurgery department because of signs of medullary compression with incomplete tetraplegia especially in the right lower limb, appearing insidiously for six months that did not ameliorate.

The patient denied any bladder or bowel dysfunction or problems with handwriting, or fine motor skills.

An MRI of the spine showed multiple intramedullary masses. The symptomatic lesions was situated in the terminal cone; the largest mass measured 48×13×10 mm in size. The mass appeared as a cystic lesion with a tissural mural nodule, with hypointense T1 inhomogeneous signal, with increased gadolinium uptake in T1, with perilesional oedema. Just above this mass, there are two other small cystic lesions, up to 1/3 medium of the vertebral body T6; one of the nodules had a subdural extra axial component (Figures 1 and 2).

Based on the characteristics on the MRI, a diagnosis of multiple hemangioblastoma was favored; and thinking of von Hippel–Lindau (VHL) disease, our investigations were expanded:

The patient underwent an MRI of the complete neuro-axis including brain and spinal cord showed a vermician hemangioblastoma associated with 2 small right cerebellar hemangioblastomas (Figure 3). The abdominal CT scan revealed 2 small renal masses, less than 2 cm in size; with multiple pancreatic cysts (Figure 4); without liver lesions, or adrenal masses. An ophthalmologic examination demonstrated none of the stigmatisms associated with VHL. Physical examination did not show any café au lait spots.

At this first step and on neurological level, a diagnosis of medullary compression as a complication of multiple intramedullary hemangioblastoma in a young male with VHL disease was done.

At the second step, and as an incidentally finding, the abdominal computed tomography (CT) scan done for VHL disease investigations, revealed a vascular compression: left renal vein passing posterior to the aorta, entrapped between the aorta and the vertebral column. Left gonadic vein, drained to the second left renal vein, was dilated with multiple pelvic venous collaterals. These aspects were indicative of posterior NCS (Figures 5).

Interviewing the patient after discovering the vascular compression (posterior NCS), he complained of intermittent and various symptoms: urinary (intermittent macroscopic hematuria, intermittent left flank pain and chronic pelvic pain aggravated by physical activity) and systemic signs dominated by chronic fatigue and persistent headache. All these symptoms lasted for his childhood; without definitive diagnosis, despite a long investigational history of several imaging examinations and laboratory tests. In addition, high degree of left varicocele was noted at physical examination; laboratory tests revealed anemia at 10.0 mg/dl, microscopic hematuria, and 24 hour urine collection analysis showed elevated proteinuria.

Based on this clinical presentation and these radiological and biologic finding, a diagnosis of posterior nutcracker syndrome was confirmed.

In summary, we have a young patient with both serious presentation of VHL disease with multiple neurological locations of hemangioblastoma complicated by medullary compression due to spinal hemangioblastoma and right renal carcinomas. Our patient had also missed symptomatic posterior nutcracker syndrome.

Figure 1: Magnetic resonance imaging scan of the spine showing multiple intramedullary masses. The symptomatic lesions was situated in the terminal cone (red arrow); the largest mass measured 48×13×10 mm in size. The mass appeared as a cystic lesion with a tissural mural nodule, with hypointense T1 inhomogeneous signal, with increased gadolinium uptake in T1.

Figure 2: Magnetic resonance imaging scan of the spine (axial) showing the three symptomatic intramedullary masses was situated in the terminal cone.
mobilization and kinetotherapy was done to prevent deep venous thrombosis. Preoperative deficits were gradually ameliorated after the operation.

The MRI scan of the cervical spine performed 8 days after the operation confirmed the total ablation of the 3 tumors and the patient was discharged from the hospital one week later.

With 11 months of follow-up, the patient is doing well, and was neurologically cured. Last MRI of the complete neuro-axis including brain and spinal cord showed same aspects of the vermian and the cerebellar hemangioblastoma, without recurrence or appearance of spinal masses. The patient remained neurologically asymptomatic and had returned to his usual life.

Prior to surgical intervention, the patient was counseled on the risks entailed in the removal of a spinal cord tumor.

After urinary catheter insertion and administration of a steroid-type anti-inflammatory (dexamethasone) and cephalosporin antibiotics, surgical exploration was made through a prone position, and a large median incision.

The dissection in the subdural space and especially the dissection of the tumor were performed using an operating microscope.

The inspection of the medullary surface showed 3 exteriorised masses red, bloody and very well vascularized (Figure 6).

After careful and complete dissection, the tumors were totally ablated (Figure 6), and their vascular pedicle being coagulated and sectioned at the end.

Final histopathological examination confirmed the diagnosis of hemangioblastoma.

Postoperative course was uneventful; There were no postoperative complications; the drainage tube was removed 48 hours after the operation and precocious mobilization and kinetotherapy was done to prevent deep venous thrombosis. Preoperative deficits were gradually ameliorated after the operation.

The MRI scan of the cervical spine performed 8 days after the operation confirmed the total ablation of the 3 tumors and the patient was discharged from the hospital one week later.

With 11 months of follow-up, the patient is doing well, and was neurologically cured. Last MRI of the complete neuro-axis including brain and spinal cord showed same aspects of the vermian and the cerebellar hemangioblastoma, without recurrence or appearance of spinal masses. The patient remained neurologically asymptomatic and had returned to his usual life.

On the urinary and vascular level, our attitudes were:
Concerning the renal masses: tumorectomy was technically impossible; and right nephrectomy was done. Macroscopic examination revealed multiple small tissular masses (more than 3) and histopathological examination confirmed the diagnosis of clear renal cell carcinoma.

Concerning the posterior nutcracker syndrome, our attitude was conservative, only the varicocele was treated and medical treatment of anemia was done. Transposition of the left renal vein was proposed in case of severe complication. At the last follow-up, the varicocele improved significantly, but intermittent hematuria persisted. Laboratory tests revealed hemoglobin at 12.0 mg/dl, with persistent microscopic hematuria and proteinuria. The last abdominal CT-scan showed no recurrence of renal mass in the left kidney.

Prior to surgical intervention, the patient was

After urinary catheter insertion and administration of a steroid-type anti-inflammatory (dexamethasone) and cephalosporin antibiotics, surgical exploration was made through a prone position, and a large median incision.

The dissection in the subdural space and especially the dissection of the tumor were performed using an operating microscope.

The inspection of the medullary surface showed 3 exteriorised masses red, bloody and very well vascularized (Figure 6).

After careful and complete dissection, the tumors were totally ablated (Figure 6), and their vascular pedicle being coagulated and sectioned at the end.

Final histopathological examination confirmed the diagnosis of hemangioblastoma.

Postoperative course was uneventful; There were no postoperative complications; the drainage tube was removed 48 hours after the operation and precocious
DISCUSSION

Hemangioblastoma are rare tumors of the central nervous system characterised by a high vascularization. They account for 1.5–2.5% of spinal cord tumors [1]. They appear spontaneously in most cases; and only 30% of them are associated with von Hippel–Lindau disease and are often multiple [2, 3].

Von Hippel–Lindau disease is a rare autosomal dominant genetic syndrome, characterised by the growth of retinal angiomas, pheochromocytoma, poly cysts, brain and spinal cord hemangioblastoma, renal and pancreatic carcinomas [5]. In our case, hemangioblastoma of the brain and spinal cord, multiple renal carcinomas and pancreatic cysts were found.

More than the half of spinal hemangioblastomas are accompanied by a syrinx [5]. In our case syrinx was not noted.

The initial symptoms of spinal hemangioblastoma are radicular pain, sensory or motor deficit slowly progressing according to the level of medulla involvement as seen in our case that was complicated by medullary compression.

MRI has become the diagnostic tool of choice and is recommended by several authors in the diagnosis of hemangioblastoma; it is also helpful in preoperative planning and the differential diagnosis of spinal cord neoplasms and vascular lesions [6, 7].

On unenhanced T1-weighted images hemangioblastoma appears as an isointense. On T2-weighted images, it appears hyperintense with a homogeneous intense enhancement. Larger hemangioblastoma can appear as a hypointense or mixed hypo-isointense on T1, heterogeneous on T2, and shows an intense heterogeneous enhancement [6]. In addition, cystic components of the hemangioblastoma can be associated with a non-enhancing rim of the cyst.

During surgery and because of the risk of massive bleeding, intramedullary hemangioblastoma surgery pose severe problems [3].

Indications for treatment and timing of surgery of hemangioblastoma are still unclear because of the rarity of these tumors and the relatively limited surgical experience. Most of the recently published series recommend microsurgical removal in symptomatic patients or in cases with tumor growth during follow-up [3, 4].

Using the aforementioned principles of microsurgical removal of spinal hemangioblastoma, microsurgical removal is feasible with a low procedure-related morbidity and can be recommended, especially in VHL patients who were symptomatic or with tumor growth during follow-up.

Occlusion of the feeding arteries, shrinking of the tumor by bipolar coagulation with low power and its en bloc removal are the key points for a safe microsurgical treatment. It is essential that the ablation must be done en bloc.

The most common complications that arise with such lesions are: intramedullary hematoma, epidural hematoma, meningocoele or cerebrospinal fluid fistula and arachnoiditis. Postlaminectomy cervical kyphosis is more common with children and it can be prevented by performing laminoplasty or laminotomy [6]. The postoperative course of our present case was not eventful.

The follow-up was based on both clinical and MRI examinations to detect tumor recurrence every six months after the operation, or in case of multiple localization to evaluate tumor progress. The neurological follow-up was evaluated according to the classification of McCormick [8].

In our case and on the neurosurgical level, we are faced to two serious problems:

The first Problem was the three intramedullary hemangioblastoma complicated by medullary compression: Our attitude was an urgent total ablation. The second problem is non-complicated small vermain and cerebellar hemangioblastoma, that our attitude was conservative, with strict and regular follow-up.

At the last follow-up, the patient remained neurologically stable.

Our patient had in addition to the neurological presentation, urological presentation of the VHL disease as right multiple carcinomas and had also symptomatic nutcracker syndrome.

Posterior nutcracker syndrome results from compression of the left renal vein between the aorta and vertebra, with subsequent development of venous varicosities of the renal pelvis, ureter, and the gonadal vein [9, 10]. The true prevalence of nutcracker syndrome especially the posterior form remains unknown and it might be underdiagnosed and easily delayed [11], as seen in our patient despite several imaging and laboratory investigations.

The computed tomography angiography (CTA) is the procedure of choice of the diagnosis with high sensitivity and specificity [10, 11].

The management of the NCS is controversial and the invasive procedures should be considered only when symptoms are severe or persistent, including severe pain, severe hematuria, renal insufficiency, and failure to respond to conservative treatment [9, 10].

Follow-up, conservative approach and surgical therapy are the different treatment options proposed for this syndrome. The available surgical procedure of the PNCS is the transposition of the LRV. Other options were described including intra-vascular or extravascular stents and renal auto transplantation. The indications for laparoscopic surgery continue to augment for the treatment of the nutcracker syndrome [12, 13].

In this case, we are faced to several urological problems:

The first we have multiple right renal tumors < 2 cm provided to be clear cell carcinomas in the context of a serious and complicated form of the VHL disease; conservative attitude with strict and regular follow-up of right renal parenchyma and the left kidney was our proposed procedure of choice but because the lack of
the ablative techniques in our country (radiofrequency and cryoablation) and the intraoperative difficulties of multiple tumorectomie, a nephrectomy was made.

The second problem was in the left kidney: posterior nutcracker syndrome, our attitude was conservative, only the varicoceles was treated and medical treatment of anemia was done; transposition of the LRV was proposed in case of severe complications.

CONCLUSION

Von Hippel–Lindau (VHL) disease and nutcracker syndrome are rare and severe entities. There association was not described. The complex presentation of the VHL disease must not make some other serious problems easily missed or neglected like renal carcinoma or nutcracker syndrome as seen in our case.

*********

Author Contributions

Nadhir Karmani – 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

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

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

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

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

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

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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES


Copyright © 2015 Nadhir Karmani et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.
 ABOUT THE AUTHORS


 Nadhir Karmani is Resident at Department of Neurosurgery Sahloul Sousse Tunisia, Faculty of Medecine Sousse Tunisia, and Department of Neurosurgery Angers France, Faculty of Medecine Angers France. He earned undergraduate degree neurosurgery doctor from Faculty of Medecine Sousse Tunisia and Postgraduate Degree Form Department of Neurosurgery Angers, Faculty of Medecine Angers France.

 Faouzi Mallat is working in Urology Department, Sahloul Hospital, Sousse, Tunisia

 Wissem Hmida is working in Urology Department, Sahloul Hospital, Sousse, Tunisia

 Khaled Ben Ahmed is working in Urology Department, Sahloul Hospital, Sousse, Tunisia

 Oussama Karmani is working in Radiology Department, Sahloul Hospital, Sousse, Tunisia

 Sidiya Oueld Chavey is working in Radiology Department, Sahloul Hospital, Sousse, Tunisia

 Amel Ben Abdallah is working in Radiology Department, Sahloul Hospital, Sousse, Tunisia
Faouzi Mosbah is working in Urology Department, Sahloul Hospital, Sousse, Tunisia

Hedi Krifa is working in Neurosurgery Department, Sahloul Hospital, Sousse, Tunisia
1,25-Dihydroxyvitamin D-mediated hypercalcemia in a patient with malignancy without lymphoma

Katie Mastoris, Sarah Park, Vasudev Magaji

ABSTRACT

Introduction: Hypercalcemia resulting from calcium homeostasis imbalance is seen in one-third of malignancies. Solid malignancies cause hypercalcemia from high parathyroid hormone-related peptide (PTHrP) and, infrequently, from osteolytic cytokines. Excessive 1, 25-dihydroxyvitamin D, seen mostly in lymphoma, causes hypercalcemia by 1-alpha-hydroxylation of 25-hydroxyvitamin D. This mechanism has also been reported in a few cases of dysgerminoma, myofibroblastic tumors, and gastrointestinal stromal tumors. Case Report: We present an unusual case of 1,25-dihydroxyvitamin D mediated hypercalcemia in a 62-year-old male with colonic adenocarcinoma and mesothelioma. Conclusion: This case emphasizes that elevated 1,25-dihydroxyvitamin D may be a hypercalcemia etiology in non-lymphoma malignancies and should to be considered when workup for elevated PTHrP and osteolytic hypercalcemia is negative.

Keywords: Adenocarcinoma, 1-alpha-hydroxylase, 1,25-dihydroxyvitamin D, Hypercalcemia, Mesothelioma
patient presented to the emergency department with right lower extremity cellulitis leading to hospitalization and septicemia. Due to concern for liver abscess, the liver mass was biopsied. Pathology showed high-grade epithelioid mesothelioma (Figure 2). During the hospitalization, his calcium levels rose (Table 1) and workup for the etiology of hypercalcemia began (Table 2). There was no evidence of osteolytic lesions on imaging, serum and urine electrophoresis were negative, and thyroid-stimulating hormone was within normal limits. There was also no evidence of granulomatous disease or evidence of granulomatous infection on imaging studies. Bisphosphonate administration brought the patient’s calcium level to within normal range (Table 1). His hospital course continued due to septicemia and renal failure requiring dialysis. He was eventually transferred intensive care unit because of hypotension and septic shock from *Escherichia coli* bacteremia secondary to right leg cellulitis. Despite being on 3 vasopressors, antibiotics, and continuous renal replacement therapy, his condition deteriorated and patient died on comfort measures.

**DISCUSSION**

Hypercalcemia resulting from calcium homeostasis imbalance is seen in one-third of malignancies and

<table>
<thead>
<tr>
<th>Normal Range</th>
<th>July 10</th>
<th>Presentation October 22</th>
<th>Hospitalization October 31</th>
<th>Administration of bisphosphonate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum calcium, mg/dL</td>
<td>8.2-10.4</td>
<td>9.8</td>
<td>10.5</td>
<td>11.9</td>
</tr>
<tr>
<td>Corrected calcium, mg/dL</td>
<td>8.2-10.4</td>
<td>-</td>
<td>-</td>
<td>12.4</td>
</tr>
<tr>
<td>Serum albumin, g/dL</td>
<td>3.5-5.0</td>
<td>4.6</td>
<td>4.2</td>
<td>3.4</td>
</tr>
<tr>
<td>Ionized calcium, mg/dL</td>
<td>4.6-5.4</td>
<td>-</td>
<td>-</td>
<td>6.45</td>
</tr>
<tr>
<td>Serum phosphorus, mg/dL</td>
<td>2.5-4.5</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>eGFR, mL/min/1.73m²</td>
<td>&gt;60</td>
<td>96</td>
<td>46</td>
<td>36</td>
</tr>
<tr>
<td>Serum BUN, mg/dL</td>
<td>10-26</td>
<td>19</td>
<td>29</td>
<td>79</td>
</tr>
<tr>
<td>Serum creatinine, mg/dL</td>
<td>0.7-1.5</td>
<td>0.79</td>
<td>1.59</td>
<td>1.95</td>
</tr>
</tbody>
</table>

Abbreviations: eGFR, estimated glomerular filtration rate; BUN, blood urea nitrogen
commonly occurs through increased synthesis of parathyroid hormone-related peptide (PTHrP) or metastatic osteolytic bone lesions. Our patient’s elevated calcium level occurred through excessive 1,25(OH)_2D levels as evident by normal PTHrP, lack of osteolytic lesions, low PTH and elevated 1,25(OH)_2D. Elevated 1,25(OH)_2D results in hypercalcemia through increased calcium absorption in the gastrointestinal tract and by promoting osteoclast differentiation and increased activity.

Elevated 1,25(OH)_2D mediated hypercalcemia has been established in granulomatous diseases and lymphomas. This mechanism occurs through elevated extra renal hydroxylation of 25-hydroxyvitamin D. Normally, regulation of 1,25(OH)_2D levels occurs in the proximal convoluted tubules of the kidney through hydroxylation of 25-hydroxyvitamin D by the 1-alpha-hydroxylase enzyme. Regulation of this enzyme by the parathyroid hormone (PTH) and calcitriol results in normal calcium levels. Extra-renal 1-alpha-hydroxylase, however, is insensitive to normal feedback mechanisms, which leads to uncontrolled hypercalcemia. Extra-renal 1-alpha-hydroxylase has been established in tumor adjacent tissue macrophages in lymphoma patients [1] with very few cases reporting extra-renal 1-alpha hydroxylase or elevation of 1,25(OH)_2D as the cause of hypercalcemia in other malignancies.

Among these neoplasms, dysgerminomas of the ovary leading to hypercalcemia have been reported [2]. Resection of the dysgerminoma resulted in normalization of serum calcium and 1,25(OH)_2D. Similarly, one patient with pleural mesothelioma had increased calcium levels due to elevated 1,25(OH)_2D which again normalized after resection [3]. A case of inflammatory myofibroblastic tumor on surgical pathology associated with hypercalcemia and 1,25(OH)_2D elevation [4] showed abundant messenger RNA (mRNA) encoding for 1-alpha-hydroxylase in the tumoral tissue. While in a case of gastrointestinal stromal tumor with 1,25(OH)_2D-associated hypercalcemia, the patient transiently responded to tyrosine kinase inhibitor and bisphosphonates [5].

Our case demonstrates the rare occurrence of 1,25(OH)_2D-mediated hypercalcemia in a patient with mesothelioma and colon cancer. Moreover, it shows two different malignancies possibly associated with this mechanism. There has only been one prior reported case of pleural mesothelioma associated with hypercalcemia and 1,25(OH)_2D elevation [4]. Interestingly, there is no adenomatous colon cancer case report of 1,25(OH)_2D-mediated hypercalcemia despite ex vivo upregulation of mRNA encoding for 1-alpha-hydroxylase being described in colon cancer [6, 7]. Due to our patient’s underlying septicemia and deterioration, we were unable to explore which malignancy caused the elevated levels of 1,25(OH)_2D.

**CONCLUSION**

We recommend considering further investigation in patients with hypercalcemia when workup for elevated PTHrP and osteolytic hypercalcemia is negative, as elevated 1,25(OH)_2D could be a hypercalcemia etiology in non-lymphoma malignancies.

**Author Contributions**

Katie Mastoris – 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

Sarah Park – Substantial contribution to design, acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Vasudev Magaji – Substantial contribution to design, acquisition of data, Analysis and interpretation of data, Revising the article critically for important intellectual content, Final approval of the version to be published

**Guarantor**

The corresponding author is the guarantor of submission.

**Conflict of Interest**

Authors declare no conflict of interest.

**Copyright**

© 2015 Katie Mastoris et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.
REFERENCES

A case report of an uncommon large size of prostatic cyst

Jingjin Yang, Xingkai Liu, Yong Zhang

ABSTRACT

Introduction: Prostatic cyst is an uncommon disease, especially large one, which need to be differentiated from prostatic neoplastic masses, prostatic non-neoplastic masses and periprostatic neoplastic masses. Case Report: A 63-year-old male who had dysuria for three years and could not urinate for three days. Initially, he was diagnosed as prostatic hyperplasia by color Doppler ultrasound. After further examinations with prostate-specific antigen (PSA), computed tomography (CT) scan, magnetic resonance imaging (MRI) scan, cystoscopy and transrectal ultrasound-guided aspiration, he was diagnosed as prostatic cyst. The size of prostatic cyst was 9.2x4.9 cm. Two weeks after diagnosis patient received an open surgery through perianal incision to remove all content in the cyst and clean this large prostatic cyst. Patient is still seen for follow-up visits every two months to further assess the treatment. Conclusion: Ultrasound, CT scan and MRI scan together help to get final diagnosis of prostatic cyst. The surgery of prostate cyst excision we performed improved outcome although we still need to follow our patient for longer time.

Keywords: Prostatic cyst, Prostatic hyperplasia, Perianal incision

INtroductIon

Prostatic cyst is a rare disease with uncertain origin [1–4], which needs to be differentiated from prostatic neoplastic masses, prostatic non-neoplastic masses, periprostatic neoplastic masses, periprostatic neoplastic masses and cyst of seminal vesicle. Most of prostatic cysts are small and located laterally without spermatozoa [5–8]. To make a correct diagnosis it is needed to perform clinical test, ultrasound, computed tomography (CT), magnetic resonance imaging (MRI) and tissue biopsy. All these procedures can help to understand non-neoplasia or neoplasia, location, size, margins and its contents. Prostatic cysts include utricular and mullerian duct cysts, diverticula of the ampulla, cysts of the ejaculatory duct, retention cyst of the prostate, hemorrhagic prostatic cyst and cyst associated with prostatitis [6, 8, 9].

We noted that only very few cases have been reported worldwide. Herein, we reported an uncommon size of prostatic cyst, which finally diagnosed through all procedures of rectal examination, clinical test, ultrasound, CT, MRI and transrectal ultrasound-guided aspiration of prostatic cyst, and successfully performed an open surgery.
CASE REPORT

A 63-year-old male without urination for three days was admitted to the hospital. The patient had suffered from dysuria, frequent urination, a delay in starting urination, a weak or slow urinary stream, a feeling of incomplete bladder emptying and getting up frequently at night to urinate (4–6 times/night) for three years. He was diagnosed as prostatic hyperplasia by abdominal color Doppler ultrasound without any treatment at his local hospital before coming to our hospital.

All his general physical examinations were normal except rectal examination. His rectal examination showed that his prostate gland was grade II prostate enlargement, the central sulcus swallowed, both sides symmetry increase, firm and smooth without nodules, absence of tenderness, feeling a cystic lesion (~4 cm) above dentate line without clear boundary and pain, and no blood on the glove. Color Doppler ultrasound showed 4.2 x 4.9 x 3.9 cm of enlarged prostate with clear outline, rough internal echo and multiple bright echos.

To further confirm the diagnosis, the patient received PSA test, CT scan, MRI scan and transrectal ultrasound-guided aspiration of prostatic cyst. Patient’s PSA test was negative. CT scan demonstrated that there was a 4.5 x 4.2 cm of even hypodense shadow behind prostate gland resulting in pushing prostate gland to front left. Patient’s prostate gland was enlarged with smooth boundary (Figure 1A–B). MRI scan showed there were irregularly a long T1 and T2 signals (9.2 x 4.9 cm) near the right peripheral leaves and penis cavernous part which extended to perineum and rectum (Figure 1C–D). Figure 1 showing that surrounding tissues were compressed and displaced. We performed transrectal ultrasound-guided aspiration of prostatic cyst and collected 15 ml of viscous milky fluid, which contained many epithelial cells and no sperm and no cancer cells. Bacterial cultures were negative.

Two week after diagnosing, we performed an open surgery through perianal incision to get rid of all content in patient’s prostatic cyst, clean this cyst and try to remove all cyst lining although this surgery was not very easy. Currently, we follow-up this patient after every two months to monitor patient’s condition and further assess our surgery treatment.

DISCUSSION

Patient was diagnosed as prostatic hyperplasia in the local hospital. After admitting to our hospital, we repeated abdominal color Doppler ultrasound plus CT scan. The results indicated that the patient could have the prostatic cyst. The negative PSA indicated that the patient did not have prostate cancer. A previous study suggests that MR imaging is useful in demonstrating the liquid content of prostatic cyst [10]. Further MRI scan indeed showed clear image of prostatic cyst, and was consistent with CT scan. The patient was diagnosed as prostatic cyst, an uncommon large size of the prostatic cyst. The result from transrectal ultrasound-guided aspiration of prostatic cyst further confirmed that the patient was suffered from the prostatic cyst, not prostate cancer and prostatic hyperplasia.

In general, treatments of prostatic cysts include transurethral resection, endoscopic marsupialization, endoscopic urethrotomy and incision, transrectal ultrasound-guided drainage, and open surgery [4, 11]. It suggests that invasive procedures are best avoided [2]. After patient was diagnosed, we had difficult time to decide which treatment would be benefit to the patient because we did not have good experience about the uncommon large size of prostatic cyst. Finally, we performed an open surgery through perianal incision to remove all content in the cyst, clean this large prostatic cyst and try to remove all cyst lining. We hope that the bladder and pelvic-cavity-pressure will push the prostatic cyst and eventually close dead space of the prostatic cyst. After surgery, all infravesical obstructive symptoms were disappeared. Patient is still seen for follow-up visits every two months in order to monitor patient’s condition and further evaluate the effect of our treatment.

CONCLUSION

Ultrasound, computed tomography (CT) scan and magnetic resonance imaging (MRI) scan together help
to get final diagnosis of prostatic cyst. The surgery of prostate cyst excision we performed improves outcome although we still need to follow our patient for longer time.

Acknowledgements
We would like to thank Dr. Changyu Zheng (NIDCR, NIH, USA) for manuscript editing assistance.

Author Contributions
Jingjin Yang – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Xingkai Liu – 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
Yong Zhang – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Jingjin Yang et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES
Coincident retroperitoneal and sigmoid colon liposarcoma: A rare occurrence

Ayvaz Ulaş Urgancı, Erkan Oymacı, Enver Vardar, Ebru Akıncılar, Ömer Engin

ABSTRACT

Introduction: Retroperitoneal liposarcomas are rare malignancies. There are a limited number of liposarcoma cases in gastrointestinal system in literature. There is no known etiological factor in the pathogenesis of liposarcomas yet. The treatment is total resection. Case Report: A 59-year-old male patient detected synchronous liposarcoma both in retroperitoneum and in colon and treated with en block resection. We discussed our case with review of literature. Conclusion: We think that even though it could not be detected in preoperative examinations, this rare togetherness should be considered intraoperatively.

Keywords: Colon liposarcoma, Gastric liposarcoma, Retroperitoneal liposarcoma

CASE REPORT

A 59-year-old male patient admitted after he realized a swelling in his abdomen with asthenia, fatigue and abdominal pain complaints for the last few months. In the physical examination, vital findings were normal. In palpation, a mass was detected filling lower left
quadrant, and extending to the right of midline and umbilicus. In laboratory examinations, white blood cell (WBC):15,400/mm³, Albumin:2.1 mg/dl and HCV RNA (+). Carcinoembryonic antigen (CEA), CA 15-3, CA 19-9, CA 125 were found to be within normal ranges. In magnetic resonance imaging (MRI) scan, a mass lesion with the size of 25x19 cm, filling the lower half of abdomen, having cystic necrotic areas internally. The mass was well circumscribed and pushed the intestinal segments and mesenteric tissues towards periphery (Figure 1). No finding indicating metastatic disease was found.

The patient was taken for operation after preoperative preparation. An approximately 30x30 cm mass originating from left retroperitoneum, pushing sigmoid colon and intestinal segments to the upper right abdominal quadrant was detected in the operation. The mass had no organ invasion but left testicular vessels and left ductus deferens could not be prepared. The mass was excised totally (Figure 2). Afterwards in the exploration, a second lesion, approximately 20x20 cm in mass, that was originating from sigmoid colon wall was detected (Figure 2). It was excised with Wedge resection. Both excised masses were reported to be pleomorphic liposarcoma in the pathological examination of the piece (Figure 3–5). CD-34 was detected to be (+), CD-68 was (-) in pleomorphic cells, S-100(-), actin was (-), desmin was (-) in the mass originating from sigmoid colon. Actin was found to be (-), S-100 was (-), and CD-68 was (-) in pleomorphic cells in retroperitoneal mass. Surgical margins of the resected pieces were reported to be negative. Postoperative period of the patient was problem-free. The patient was mobilized on the 1st postoperative day. Bowel movements were started on the 2nd postoperative day, and oral food was given to the patient on the 3rd postoperative day. Abdominal drains were removed in 6th and 7th postoperative days. The patient was discharged on the 9th postoperative day.

The patient was discussed at tumor committee. Follow-up without any additional treatment was decided. No findings indicating metastasis or recurrence were detected in the three months follow-up of the patient.

**DISCUSSION**

Most frequent intra-abdominal mesenchymal tumors are liposarcomas. They usually manifest with non-specific abdominal pain and abdominal mass [2]. The patients may usually have mild abdominal symptoms, weight loss and usually they are generally diagnosed in late period. In our case, the present symptoms were recognized in later period and the patient was admitted with a big mass. Rarely, neurological findings due to mass pressure, may accompany [6]. Metastasis may be detected in approximately 11% of the patients and metastases are frequently occur in lungs and liver [6]. Despite admitting in later period, no distant metastases were detected in the patient.
Abdominal computed tomography (CT) scan and MRI scan may show the association of the mass with adjacent organs and vascular structures, and may provide information about histological type. There are publications recommending abdominal CT scan or MRI scan as ideal method [6–8]. In our case, the structure of mass and its association with surrounding tissues and adjacent organs were assessed in detail before the operation with MRI.

In our day, the only treatment for retroperitoneal liposarcomas is en bloc Ro resection [2, 5, 6, 9]. There is not enough evidence regarding radiotherapy and chemotherapy modalities [2, 5, 6, 9]. The most important prognostic factors are Ro resection and tumor histology [3, 9]. Multifocality is a rare but important prognostic parameter [6].

Liposarcomas are assessed in five sub-groups such as well differentiated, dedifferentiated, myxoid, round cell and pleomorphic based on their cytogenic and morphologic anomalies [1, 4]. Well differentiated and myxoid liposarcomas are seen much more frequently among primary tumors, and their prognoses are better and metastasis rates are lower than other types. On the other hand, dedifferentiated liposarcomas are seen more frequently in recurrent tumors. The rates of dedifferentiated or pleomorphic subtypes, are higher in recurrent tumors than primary ones [2].

Very frequent local recurrence (66%) and organ invasion were the leading mortality causes of the disease [2, 5, 9]. In wide series, five-year disease-free survival after en-block resection was given as 18% [10]. Recommend treatment is re-operation in recurrent cases. De-bulking and radiotherapy may be recommended in cases for which Ro resection cannot be performed [2, 9].

A second mass was detected in intra-operative observation in our case despite it could not be recognized in preoperative examinations. Liposarcomas are rarely seen in gastrointestinal system. While there are liposarcoma cases detected as multiple foci in retroperitoneum in literature, we did not encounter any case who had synchronous liposarcoma in gastrointestinal system and retroperitoneum. Recurrence is an important criterion for survival in these patients, and the probability of synchronous tumor must also be considered.

**CONCLUSION**

In conclusion, while retroperitoneal liposarcomas are rarely seen as single tumors, synchronous colon liposarcoma occurring together is very rare in the literature. We think that even though it could not be detected in preoperative examinations, this rare togetherness should be considered intraoperatively.

**Author Contributions**

Ayvaz Ulaş Urgancı – 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

Erkan Oymacı – 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

Enver Vardar – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ebru Akıncılar – Acquisition of data, Drafting the article, Final approval of the version to be published

Ömer Engin – Acquisition of data, Drafting the article, Final approval of the version to be published
Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Ayvaz Ulaş Urganci et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES

ABOUT THE AUTHORS

Ayvaz Ulaş Urgancı is working in General surgery Department at Buca Seyfi Demirsoy Goverment Hospital in İzmir-Turkey. He earned undergraduate degree from İzmir Ege University Hospital Turkey and postgraduate degree (general surgery) from İzmir Atatürk Research and Training Hospital-Turkey. He has published 15 research papers in national and international academic journals. His research interests include gastrointestinal surgery, laparoscopic surgery, colorectal surgery.
E-mail: ulasurganci@gmail.com

Erkan Oymacı is working in Gastrointestinal Surgery at Bozyaka Research and Training Hospital in İzmir-Turkey. He earned the undergraduate degree from İzmir Ege University Hospital Turkey and postgraduate degree in Gastrointestinal Surgery from TYIH Research and Trainin Hospital Ankara-Turkey. He has published 40 research papers in national and international academic journals. His research interests include gastrointestinal surgery, laparoscopic surgery, colorectal surgery.
E-mail: erkan.oymaci@hotmail.com

Enver Vardar is working in Patolhogy Derpartment at Bozyaka Research and Training Hospital in İzmir-Turkey. He has published 40 research papers in national and international academic journals. His research interests include gastrointestinal pathology.
**Ebru Akıncılar** is working in General surgery Department at Buca Seyfi Demirsoy Government Hospital in İzmir-Turkey. She earned undergraduate degree from 9 September University Hospital İzmir-Turkey and postgraduate degree form general surgery from İzmir Atatürk Research and Training Hospital-Turkey. She has published 15 research papers in national and international academic journals. Her research interests include Colorectal Surgery, Breast Surgery.

**Ömer Engin** is working in General surgery Department at Buca Seyfi Demirsoy Government Hospital in İzmir-Turkey. He has published 40 research papers in national and international academic journals. His research interests include Gastrointestinal Surgery, Colorectal Surgery.
Abstract

Introduction: Scrub typhus is a mite-borne infectious disease caused by Orientia tsutsugamushi. Farmers account for approximately two-thirds of all reported cases. Incidence rates are highest in people aged 40 to 60 years. The severity of infection can range from mild symptoms and signs to multi-organ failure. The chief target of the organism is the vascular endothelium of various organs. There is inflammation in and around the blood vessels that causes endothelial damage resulting in vascular leakage and organ dysfunction.

Case Report: A 62-year-old female, farmer by occupation who presented to us with features of acute respiratory distress syndrome (ARDS) and meningitis, subsequently developed gangrene of digits of all four limbs. We investigated the patient for infectious and non-infectious causes of vasculitis leading onto gangrene. We attributed the cause for pan-digital gangrene as scrub typhus as evidenced by the presence of eschar and positive serology. Patient recovered with ventilatory support and antibiotics though her digits could not be salvaged which was managed conservatively.

Conclusion: While going through literature we observed that there were no case reports of gangrene caused by scrub typhus. Pan-digital gangrene is an unusual complication of scrub typhus. Hence scrub typhus has to be ruled out in patients presenting with this complication.

Keywords: Pan-digital gangrene, scrub typhus, Vasculitis

How to cite this article


doi:10.5348/ijcri-201569-CR-10530

Introduction

Orientia tsutsugamushi is an obligate intracellular gram-negative coccobacillus which is the etiologic agent for scrub typhus. There are three strains of Orientia tsutsugamushi namely Karp, Gilliam, and Kato strains. Infection with one strain does not confer immunity from infection with another strain. Scrub typhus is endemic in India, China, Korea, Pakistan, Taiwan, Japan, Thailand, Malaysia, and Northern Australia. The vector for scrub typhus are larval trombiculid mites, also called chiggers [1].

After an incubation period of 7–10 days, the disease may start with prodromal symptoms like headache, anorexia, malaise with fever [2]. Rash and characteristic eschar may develop subsequently. The infection can have
varying presentation. Some have mild symptoms, while others can develop multiorgan failure. Approximately, 4% of patients hospitalized have fatal infection [3]. Elderly patients are more likely to have severe illness and complications. Some patients develop a localized necrotic skin lesion (eschar) at the site of their infecting chigger bite.

Generalized lymphadenopathy, acute kidney injury and respiratory complaints are often present. Rarely, acute respiratory distress syndrome may occur. Involvement of blood vessels in the central nervous system may produce meningitis or encephalitis. Herein, we present a patient who developed severe respiratory failure due to acute respiratory distress syndrome requiring ventilatory support and pan-digital gangrene due to scrub typhus.

**CASE REPORT**

A 62-year-old female, farmer by occupation presented to the emergency department with history of high grade fever and chills for three days. She was managed with antipyretics in outside hospital with no benefit. She developed breathlessness at rest for 1 day. Hence she was referred to our institution for further management. On examination, she was febrile (101°F) and tachypneic with a respiratory rate of 40 per minute. Her pulse rate was 106 per minute and her blood pressure was 110/80 mmHg. She had an eschar in her right loin. Her respiratory examination revealed bilateral extensive crepitations. Her jugular venous pressure was normal and she was clinically euvoletic. We proceeded with the following investigations as given in Table 1.

Arterial blood gas revealed type 2 respiratory failure (pH 7.04, pCO2 7.5 mmHg, PO2 57 mmHg). In view of progressive breathlessness despite supportive measures she was intubated and connected to a ventilator. Chest X-ray showed features of consolidation (Figure 1) which was subsequently confirmed by CT scan of thorax (Figure 2).

The patient developed altered sensorium subsequently. CT scan of brain was normal. Cerebrospinal fluid (CSF) analysis showed sugar of 72 mg/dl, protein 67 mg/dl, 140 WBC/hpf, neutrophils 65% and lymphocytes 35%. CSF culture, gram staining, HSV serology done were negative. Etiologic workup for fever, ARDS, and altered sensorium were done (Table 2).

Patient was initially managed with broad spectrum antibiotics – imipenem plus cilastatin and linezolid. After serology report of scrub typhus being positive she was started on doxycycline 100 mg BD and rifampicin 600 mg OD. On day-3 of admission patient developed blackish discoloration of the terminal digits of upper and lower limbs which was progressive in nature and ultimately developed into dry gangrene (Figures 3 and 4) over 4–5 days. However, all her peripheral pulses were felt normally. Vasculitis workup like ANA, dsDNA, antiphospholipid antibody, C-ANCA, P-ANCA, Anti

**Table 1: Laboratory investigations of the patient.**

<table>
<thead>
<tr>
<th>Hemoglobin</th>
<th>9.6 g/dl</th>
</tr>
</thead>
<tbody>
<tr>
<td>MCV</td>
<td>87.9 fl</td>
</tr>
<tr>
<td>MCH</td>
<td>29.7 pg</td>
</tr>
<tr>
<td>Total Count</td>
<td>13900 cells/mm3</td>
</tr>
<tr>
<td>Differential Count</td>
<td>P70 L28 E1 M1</td>
</tr>
<tr>
<td>Peripheral Smear</td>
<td>Tox neutrophil granules with shift to left</td>
</tr>
<tr>
<td>Platelet counts</td>
<td>1.50 lakhs/mm3</td>
</tr>
<tr>
<td>Serum creatinine</td>
<td>1.5 mg/dl</td>
</tr>
<tr>
<td>Serum sodium</td>
<td>141 meq/l</td>
</tr>
<tr>
<td>Total bilirubin</td>
<td>0.98 mg/dl</td>
</tr>
<tr>
<td>ALT</td>
<td>10 U/L</td>
</tr>
<tr>
<td>AST</td>
<td>42 U/L</td>
</tr>
<tr>
<td>Alkaline phosphatase</td>
<td>42 U/L</td>
</tr>
<tr>
<td>Serum albumin</td>
<td>2 g/dl</td>
</tr>
<tr>
<td>Electrocardiogram</td>
<td>Normal</td>
</tr>
</tbody>
</table>

**Table 2: Etiologial workup for fever, ARDS and altered sensorium.**

<table>
<thead>
<tr>
<th>QBC – MP</th>
<th>NEGATIVE</th>
</tr>
</thead>
<tbody>
<tr>
<td>MALARIAL ANTIGEN</td>
<td>Negative</td>
</tr>
<tr>
<td>RT-PCR FOR H1N1: throat swab</td>
<td>Negative</td>
</tr>
<tr>
<td>SEROLOGY FOR SCRUB TYPHUS</td>
<td>Positive</td>
</tr>
<tr>
<td>IGM ANTILEPTOSIRAL ANTIBODY</td>
<td>Negative</td>
</tr>
<tr>
<td>IGM DENGUE ANTIBODY</td>
<td>Negative</td>
</tr>
<tr>
<td>HIV</td>
<td>Negative</td>
</tr>
<tr>
<td>Anti HbsAg</td>
<td>Negative</td>
</tr>
<tr>
<td>Anti HCV</td>
<td>Negative</td>
</tr>
<tr>
<td>Blood culture</td>
<td>No Growth</td>
</tr>
<tr>
<td>Urine culture</td>
<td>No Growth</td>
</tr>
<tr>
<td>CSF culture</td>
<td>No Growth</td>
</tr>
<tr>
<td>NON BAL culture</td>
<td>No Growth</td>
</tr>
<tr>
<td>Echocardiogram</td>
<td>No evidence of IE , EF 64%</td>
</tr>
</tbody>
</table>

**Abbreviations:**

QBC-MP quantitative buffy coat-malaria parasite
RT-PCR real time polymerase chain reaction
HIV Human Immunodeficiency Virus
Anti HBsAg anti hepatitis B surface antigen
ANTI HCV anti hepatitis C virus
Non-BAL non-bronchoalveolar lavage
centromere antibody were negative. Vascular surgery opinion was sought and the patient was started on LMWX and aspirin. Biopsy of the lesion was not attempted as the patient’s relatives did not give consent.

After eight days of doxycycline therapy patient showed gradual improvement in clinical condition and was weaned off from ventilator subsequently. Patient developed severe pain in her upper and lower limb digits around the gangrenous areas. She received several analgesics including tramadol, amitriptyline, paracetamol, NSAIDS with partial relief of pain. She was started on cilostazol, aspirin, pentoxifylline by vascular surgeon after cessation of LMWH. She eventually developed auto amputation of her right lower limb second and third toes followed by second toe of left lower limb and little finger of her right hand over the next 2–3 months.

**DISCUSSION**

Scrub typhus infections may present as mild illness or can lead onto fatal complications like ARDS, meningoencephalitis, acute kidney injury, myocarditis, hepatic dysfunction and multiorgan involvement. Complications usually occur after the first week of illness. Due to non specific clinical features of scrub typhus it is grossly under diagnosed, though it is an growing and
emerging disease. Since scrub typhus can lead to life threatening illness, initiation of treatment should be early and prompt based on clinical suspicion, which later should be confirmed by serology.

*O. tsutsugamushi* proliferates in the endothelium of small blood vessels that results in cytokine release. This cytokine causes endothelial damage leading to leakage of fluid and aggregation of platelets, and proliferation of polymorph and monocytes leading to focal microinfarction. This eventually leads to venous thrombosis and peripheral gangrene. Various organs like skin, kidneys, skeletal muscles, brain lungs, and cardiac muscles are predominantly affected by this process [4, 5].

*O. tsutsugamushi* infection spreads to multiple organs through blood stream and lymphatics. It predominately targets the macrophages present in the spleen and liver [6]. The *Rickettsia* attacks the endothelial cells causing inflammation by a mechanism called oxidative stress leading onto local and systemic vasculitits [7]. Despite the fact that disseminated endothelial infection occurs in scrub typhus, the real incidence of vasculitis leading to vaso-occlusion is quite rare which is the highlight of our case.

Systemic vasculitis causing gangrene can occur due to both infectious and non infectious cause. Among the infectious cause viruses are the leading organisms that lead to systemic vasculitis. Hepatitis B virus is associated with polyarteritis nodosa. Hepatitis C virus related mixed cryoglobulinemia causes vasculitis in only a minority of people who acquired the infection. There are a few other viruses that cause vasculitis like Human immunodeficiency virus (HIV), parvo virus B19, varicella-zoster virus, human T-cell lymphotropic virus (HTLV)-1 and cytomegalovirus.

Bacteria, fungi or parasites also cause vasculitis which could be by direct invasion of blood vessels or by septic embolization. Syphilitic aortitis or cerebrovascular disease and rickettsial diseases are other, more specific, bacteria-induced vasculitides.

The importance of differentiating infectious and non-infectious cause of vasculitis is necessary because the treatment strategies are different. Bacterial, parasitic and fungal infections are treated with appropriate antimicrobial agents. Combination of antiviral drugs and plasma exchanges are effectively tried for HBV infections causing polyarteritis nodosa and HIV related vasculitis. HCV related cryoglobulinemic vasculitis is treated with antiviral drugs combined with low dose steroids [8].

The development of digital gangrene is a sign of systemic diseases. Smoking, hypertension, diabetes, obesity and hyperlipidemia are commonly associated with digital gangrene. Homocysteinemia, vasculitides like primary systemic vasculitis and medium vessel vasculitis such as polyarteritis nodosa commonly presents with digital gangrene. Rheumatoid arthritis and systemic lupus erythematosus cause premature atherosclerosis and gangrene. Thromboangitis obliterans (TAO) or Buerger’s disease can cause claudication and digital gangrene [9].

While looking into the infective causes as in our case, the diagnosis of scrub typhus should be made by excluding other viral infectious diseases like dengue, infectious mononucleosis, HIV and bacterial diseases like typhoid, leptospirosis, meningococcal disease causing ARDS and vasculitis. These infections were excluded with appropriate investigations. As serology for scrub typhus done by ELISA was positive, we thought about the association of scrub typhus as the cause of vasculitis.

The clinical features of this 62-year-old female who presented with fever, breathlessness and altered sensorium who responded to treatment with doxycycline are consistent with scrub typhus. This is confirmed by the presence of eschar and positive scrub typhus serology. We eliminated all other differentials for scrub typhus with multi-organ involvement. The patient subsequently developed pan digital gangrene. We excluded other infective and non infective diseases causing vasculitis. Hence we conclude that the patient developed pan-digital gangrene secondary to scrub typhus vasculitis. To our knowledge this is the first reported case of pan-digital gangrene in scrub typhus infection.

**CONCLUSION**

Although there are so many conditions causing vasculitis, it is essential to differentiate infective from non infective causes as the treatment is entirely different. From the case that we have presented here, we learn that pan digital gangrene may be an unusual complication of scrub typhus and as many cases have not been reported, this has to be ruled out in cases of digital gangrene.

**Author Contributions**

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

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

Ramakrishnan S.R. – Analysis and interpretation of data, Critical revision of the article and final approval of the version to be published

Sathiyan Sivanandam – Conception and design, Analysis and interpretation of data, Drafting the article, and Final approval of the version to be published

**Guarantor**

The corresponding author is the guarantor of submission.
Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Suja Lakshmanan et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES

ABOUT THE AUTHORS


L. Suja is Assistant Professor in Sri Ramachandra Medical College and Research Institute, Chennai, India. She is interested in the field of autoimmune disorders.
E-mail: suja.lakshmanan@gmail.com

Krishnamoorth S Seetharaman is Assistant Professor in Sri Ramachandra Medical College and Research institute, Chennai, India. He earned his undergraduate degree from Calicut Medical College, Calicut University, Calicut Kerala, India and postgraduate degree from Stanley Medical College, Tamil Nadu Dr MGR Medical University, Chennai, Tamil Nadu, India. He has published one research paper in a national journal. His interest include detailed evaluation of rare and interesting cases, comprehensive management of each patient and delivering lectures in various forum.
E-mail: drmoorthykrishnan@yahoo.co.in
Ramakrishnan S.R. is Professor in Sri Ramachandra Medical College and Research institute, Chennai, India. He earned undergraduate and postgraduate degree from Stanley Medical College, Tamil Nadu Dr MGR Medical University, Chennai, Tamil Nadu, India. His research interest is on diabetes mellitus. E-mail: drsrk_71@yahoo.com

Sathiyan Sivanandam is Assistant Professor in Sri Ramachandra Medical College and Research Institute, Chennai, India. He earned undergraduate degree from PSG medical college, Coimbatore, Tamil Nadu, India and post graduate degree from Madurai Medical College, Tamil Nadu Dr MGR Medical University, Madurai, Tamil Nadu, India. He did his super specialization in nephrology at Madras medical college, Tamil Nadu Dr MGR Medical University, Chennai, Tamil Nadu, India. He is interested in renal transplantation and clinical nephrology. E-mail: sathiyan.sivanandam@gmail.com

Koushik A.K. is a postgraduate in Sri Ramachandra Medical College and Research institute, Chennai, India. He has published a couple of case reports in the national journals. E-mail: style.koushik@gmail.com
ABSTRACT

Introduction: Nephroblastoma is rare in adults. It accounts only 0.5% of all renal tumors and it is often diagnosed in advanced stages. Histology, grading and staging are similar to pediatric nephroblastoma (PN) but its prognosis in the same clinical stage is more pejorative. However, some reports use the current pediatric protocols to treat adult nephroblastoma (AN) taking into consideration tumor stage and grade. Others recommend using advanced disease regimens for all stages and grades and no standardized treatment is defined yet. Case Report: A 29-year-old female who was diagnosed with stage IV nephroblastoma. She underwent radical nephrectomy and received multi-agent chemotherapy according to the International Society of Pediatric Oncology (SIOP) protocol No. 9301, a very important reduction of metastasis size occurred after three cycles. Conclusion: Adult nephroblastoma is rare, its prognosis is poorer than that of children when the disease is compared stage for stage, but the outcome for adult patients having this tumor is steadily improving if it is early detected.

Keywords: Adult, Nephroblastoma, Pediatrics, Wilms tumor

INTRODUCTION

Wilms tumor (WT) or nephroblastoma, is one of the most frequent malignant renal tumor in children [1], it is extremely rare in adults, its incidence is 8 per million in children [2], whereas its incidence is lower than 0.2 per million per year in adults, There is no difference between histological features of nephroblastoma in children and in adults, Treatment guidelines for pediatric nephroblastoma are well established but are less clear for adults [1–4], in view of the rare occurrence of this tumor in adults it is extremely difficult to carry out randomized clinical trials and therapeutic guidelines have not been defined yet.

Current therapies are based on trials and studies for children conducted by the Société International of Oncologie Pédiatrique (SIOP) and the Children’s Oncology Group (COG) [3]. The prognosis for adult was reported to be worse than that for children [1, 2] because Adult nephroblastoma has more aggressive behavior and shows worse therapeutic response [2]. In children the overall survival is 90% for stage I and 70% for stage IV, this decrease with 76 % for stage I and with 11% for stage IV in adult [4].

We report a new case of adult nephroblastoma (AN) in which we discuss the diagnosis, treatment and outcome of this rare tumor for which the therapeutic strategies are more developed for pediatric groups and on which there are no large studies.
CASE REPORT

A 29-year-old female was admitted to our department after 5 months history of left loin pain. Physical examination revealed hypertension and a palpable left loin mass. Biologically, the blood analysis were normal: hemoglobin: 12 g/dl, neutrophil: 3.4 g/l, leucocyte: 4.5 g/l, platelets: 340 g/l, creatinine: 58 mmol/l. Computed tomography (CT) scan of the abdomen showed a large left renal tumor with extension into renal vein and para-aortic lymphadenopathy with liver metastases, CT thorax confirmed multiple bilateral pulmonary lesions consistent with metastatic disease. A radical, transabdominal nephrectomy was performed. The operative specimen weighed 1540 g with a size of 13.5x13.5x8 cm surrounded by peritoneal grease with irregular outer surface. The kidney measures 19x17x15 cm and presents a white yellowish upper pole tumor of 13.5x13.5x8 cm encapsulated. It has a soft consistency with necrotic bleeding site estimated at 20%. The next of renal parenchyma measures 5x6.5x3 cm (Figure 1). Histology confirmed a diffuse blastemal and epithelial Wilms tumour, it shows tumor proliferation with dual component: epithelial and mesenchymatous; the epithelial component makes glandular and trabecular structure bored with cylindrical basophile cells with big size, with hyperchromic nucleus and atypical mitosis. The epithelial structure is furrowed with fusiform cells big nucleus size, all are developed in a stroma surrounded by a fibrous tick capsule respecting the adjacent parenchyma. No image of vascular embolism was seen, the renal hilum was unaffected as well as the ureteral cuts and the adrenal (Figure 2). The lymph node dissection was negative 0N/6N. There were no postoperative complications.

Chemotherapy according to the SIOP 93-01 protocol was started as soon as possible. After 2 cycles of chemotherapy combining actinomycine D at 15 mg per kg and per injection/3 weeks, adriamycin with the dose of 50 mg/m² each 3 weeks and Vincristine 1.5 mg/m² d1-d8, a very good clinical response was seen and a marked reduction in metastases size occurred (Figures 3 and 4). After six cycles of chemotherapy a complete remission of liver metastases was obtained with a very good reduction of lung metastases, unfortunately two months later the patient died after a sudden crisis headache.

DISCUSSION

Nephroblastoma is rare in adults and could have a more aggressive clinical behaviour than that in children, it is frequently diagnosed at more advanced stages, stage III disease represents 50% of AN.

Clinically, the tumor is often discovered as an incidental abdominal mass either by routine palpation or on a CT-scan or ultrasound done for other reasons. More commonly however, flank pain and/or hematuria are the first indications of the condition. Systemic symptoms such as weight loss, malaise, weakness, and fever are indicative of advanced disease [5, 6]. The most common metastatic sites are the lungs, liver and the bones [3]. The classic histology of nephroblastoma is that of a triphasic embryonal neoplasm containing varying amounts of blastema, stroma and epithelial cells forming abortive tubular or glomerular structures. For diagnosis to be made not all the classic features are necessary, as some Wilms tumors may contain blastemal or epithelial features only [7]. According to all available studies and due to the very small number of reported cases, randomized trials cannot be undertaken and results of pediatric trials should be considered. And Until recently, no standardized treatment for AN was found in literature [3, 8]. There
In 1982, the NWTSG reported 31 patients with AN who were treated between 1968 and 1979 [8, 6]. 51.7% had stage III and stage IV disease. The three-year survival rate was 24%, compared with 74% for patients with pediatric nephroblastoma (PN). These results indicated a need for new therapeutic approaches for patients with AN. In 1990, Arrigo et al. [8, 11] from the NWTSG studied 27 adult patients who were treated between 1979 and 1987. Their results demonstrated a survival rate of 67% at three year. Moreover, the national Wilms Tumor Study (NWTS) recommends a multidisciplinary treatment: for stages III and IV, a large nephrectomy associated with a triple Chemotherapy (actinomycine D, vincristine and doxorubicin) during 15 months then a radiation of tumor bed and a double chemotherapy without radiation for both stage I and II [12].

For our patient we followed the NWTS’s recommendations, after surgery a combination of vincristine, daunomycin and doxorubicin was used, after three cycles a remarkable reduction of about 80% of liver and lung metastases was obtained.

Figure 3: Computed tomography pulmonary scan showing the pulmonary metastases before (A) and after (B) 3 cycles of chemotherapy.

Figure 4: Computed tomography abdominal scan showing the liver metastases before (A) and after (B) 3 cycles of chemotherapy.

In 1982, the NWTSG reported 31 patients with AN who were treated between 1968 and 1979 [8, 6]. 51.7% had stage III and stage IV disease. The three-year survival rate was 24%, compared with 74% for patients with pediatric nephroblastoma (PN). These results indicated a need for new therapeutic approaches for patients with AN. In 1990, Arrigo et al. [8, 11] from the NWTSG studied 27 adult patients who were treated between 1979 and 1987. Their results demonstrated a survival rate of 67% at three year. Moreover, the national Wilms Tumor Study (NWTS) recommends a multidisciplinary treatment: for stages III and IV, a large nephrectomy associated with a triple Chemotherapy (actinomycine D, vincristine and doxorubicin) during 15 months then a radiation of tumor bed and a double chemotherapy without radiation for both stage I and II [12].

National Wilms Tumor Study Group (NWTSG) realises nephrectomy at the time of diagnosis for all resectable primary tumors. Surgery is then followed by chemotherapy and radiation therapy to sites of metastatic or residual disease after surgery [9]. In contrast, The International Society of Pediatric Oncology (SIOP) recommends administering several weeks of chemotherapy before nephrectomy [10].

were only a few reports of small series of patients with AN who were treated with different protocols involving chemotherapy, surgery, and radiotherapy [8]. The results of these early studies are somewhat contradictory. Fortunately, the more current guidelines proposed by the National Wilms Tumor Study Group (NWTS) and The International Society of Pediatric Oncology (SIOP) have helped to direct physicians [8].

National Wilms Tumor Study Group (NWTSG) realises nephrectomy at the time of diagnosis for all resectable primary tumors. Surgery is then followed by chemotherapy and radiation therapy to sites of metastatic or residual disease after surgery [9]. In contrast, The International Society of Pediatric Oncology (SIOP) recommends administering several weeks of chemotherapy before nephrectomy [10].

National Wilms Tumor Study Group (NWTSG) realises nephrectomy at the time of diagnosis for all resectable primary tumors. Surgery is then followed by chemotherapy and radiation therapy to sites of metastatic or residual disease after surgery [9]. In contrast, The International Society of Pediatric Oncology (SIOP) recommends administering several weeks of chemotherapy before nephrectomy [10].
In the study of the International Society of Pediatric Oncology [3], 30 AN were treated: 66% of them were localized whereas 33% were metastatic at the time of diagnosis. Unlike protocols for children, the majority of adult in SIOP study underwent primary surgery; whereas preoperative chemotherapy was administered to only 4 out of 30 patients. The event-free survival was 57% at 4 years and the overall survival was 83% [3].

Terenziani et al. [13] reported their institutional experience regarding AN. Between 1983 and 2001, 17 patients with AN who were older than 16 years were treated according to pediatric nephroblastoma (PN) guidelines. In this series, the overall survival was 62.4% at 5th year.

Due to the frequency of lung metastases in stage III and IV, Hentrich et al. justify a systemic pulmonary irradiation of lung metastases they consider that an irradiation of bed tumor may complete surgery following by chemotherapy (Dactinomycin + Vincristine + Doxorubicin). Lately, in relapsed AN patients, high-dose chemotherapy followed by autologous steam cell rescue has been used as the salvage therapy but only a small number of patients have obtained long complete remission [1, 14]. The use of the new combination chemotherapy regimens based on the SIOP-2001 second-line protocol has not improved the outcome in patients with recurrent AN. The best approach in the treatment of AN still needs to be defined. Abu-Ghosh et al. [15] reported an overall response rate of 82% in 11 patients treated with ifosfamide-carboplatin etoposide chemotherapy for poor-risk relapsed nephroblastoma. Italiano [16] reported one case of a short but striking response to paclitaxel in an adult who had recurrent massive disease. Our patient presented with advanced disease. His clinical follow-up suggests that treating AN like childhood Wilms tumor remains an effective option. New modalities of treatment are being tested. One example is the use of recombinant interferon-a to treat a case of recurrent adult nephroblastoma [17]. Whether these new modalities of treatment will prove to be more effective, they await additional data, hopefully from standardized clinical studies on adults.

CONCLUSION

Wilms tumor or nephroblastoma is an exceptional tumor in adults. This diagnosis, often histological, is generally established at a more advanced clinical stage than in children and the prognosis, for the same clinical stage, is comparatively poorer, it must be kept in mind that only the early detection of the tumor offers patients the best chance for survival.

********

Author Contributions

N. Naqos – 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
Z. Bouchbika – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
A. Taleb – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
N. Benchkroune – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
H. Jouhadi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

© 2015 N. Naqos et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES

Eruptive Collagenoma in a mongol girl: A rare association

Balwinder Kaur Brar, Mahajan B. B., Nidhi Kamra

ABSTRACT

Introduction: Eruptive collagenoma is a rare type of connective tissue naevi with predominant extra-cellular matrix component being collagen. Lesions are characterized by abrupt history of onset. Case Report: An 18-year-old female with a clinical diagnosis of Down syndrome presented with abrupt onset of skin colored nodules on upper back and mons pubis. Lesions were asymptomatic. On examination multiple, firm, discrete, non-tender, flesh colored nodules were present on upper 2/3 of back and mons pubis. Histopathology aided with Masson's trichome stain confirmed the diagnosis of collagenoma. Conclusion: The association of collagenoma has been known with various systemic disorders but its alliance with Down syndrome and localization over mons pubis has still not been reported.

Keywords: Eruptive collagenoma, Down syndrome, Mons pubis, Flesh colored modules

INTRODUCTION

Connective tissue nevi are hamartomas characterized by an excess or deficit in the number of cells and their biosynthetic products including collagen, elastic fibers, and glycosaminoglycans [1]. Connective tissue nevi with predominantly collagen are referred to as Lipschutz type and with elastic component as Lewandowsky type. Multiple collagenomas are invariably present in several distinct syndromes like Buschke-Ollendorf syndrome, eruptive collagenoma, familial cutaneous collagenoma and tuberous sclerosis. Isolated cerebriform collagenoma of the palm and sole is a well-known entity and has been reported in association with Proteus syndrome. Eruptive collagenoma is a rare type of acquired collagenoma that was first reported in 1955 by Colomb [2–5]. Herewith a report of eruptive collagenoma in Down syndrome is reported for its rarity and unusual localization

CASE REPORT

An 18-year-old female with history of delayed milestones, low IQ, and typical Mongolian facies (Figure 1) (typical slant of forehead, and macroglossia) presented to our outpatient department with abrupt onset of asymptomatic skin colored nodules within a period of one month on upper back. The patient did not report any previous history of chicken pox/ trauma at the involved site. Her family history was unremarkable. A review of
the various organ systems (cardiovascular, respiratory, gastrointestinal tract, central nervous system) was within normal limits. Examination revealed multiple, discrete, firm, non-tender, skin colored nodules measuring 0.5 to 2 cm in diameter with no scaling or exudation on the surface. The lesions were present predominantly on upper 2/3 of back and on thorough cutaneous examination similar lesions were present on the mons pubis, which as per patient developed 6 months back (Figures 2 and 3). We kept the differentials of eruptive xanthoma, steatocystoma multiplex and eruptive collagenoma. There was no evidence of hypopigmented macules on wood’s lamp examination or skin lesions suggestive of tuberous sclerosis. There was no significant past medical or surgical history. The hematological and biochemical investigations including complete blood count, renal, liver function tests, urine analysis, lipid profile, electrocardiogram, abdominal ultrasound and chest roentgenogram were within normal limits. Skeletal survey did not demonstrate any evidence of osteopoikilosis. A skin biopsy obtained from the lesion on upper back and mons pubis showed focal acanthotic epidermis and significantly increased density of collagen bundles in the deep reticular dermis (Figure 4). Masson’s trichrome stain confirmed the presence of dense collagen bundles with decreased elastic fibers (Figure 5). Thus a diagnosis of sporadic eruptive collagenoma with Down syndrome was made.

Figure 1: The patient has typical Mongolia facies expression

Figure 2: Multiple, discrete, skin colored nodules on upper back.

Figure 3: Multiple flesh colored nodules on mons pubis.
Collagenomas in FCC are also distributed symmetrically on trunk and proximal extremities, but are more numerous (in hundreds) and are also associated with various cardiac abnormalities like cardiomyopathy and conduction disorders [3, 6]. Shagreen patches (plaques of collagenoma) are present in tuberous sclerosis with other classic cutaneous findings like ash-leaf macules, facial angiofibromas, periungual fibromas (Koenen’s tumor), gingival fibromas and fibrous plaque of forehead. Isolated collagenomas are sporadic and are localized most commonly on palm, sole and labium majus. Cerebriform plantar nevi are considered to be pathognomic of Proteus syndrome (a type of epidermal nevus syndrome) [4, 5, 7]. However, many authors have reported the presence of plantar collagenoma without any co-existent features of Proteus syndrome [8]. Isolated collagenoma on the scalp has been reported and can manifest as cutis verticis gyrata [9]. Collagenomas have also been reported in alliance with pseudohypoparathyroidism and hypogonadism [10, 11]. Though the pathogenesis of collagenomas is unknown, sporadic collagenomas may be related to trauma, since they appear most frequently in areas subject to friction, in Down syndrome too its pathogenesis is unclear. However, elastosis perforans serpiginosa is also well known as a complication of this syndrome, suggesting that Down syndrome may have various accompanying connective tissue disorders [12].

DISCUSSION

Collagenoma (collagen nevi) have been classified into distinct groups on the basis of clinical considerations (Table 1). Depending upon classification of the genetic inheritance pattern, collagenomas are classified as either inherited or sporadic (Table 2) with autosomal dominant inheritance common to all inherited subtypes. Eruptive collagenoma is characterized by abrupt development of multiple asymptomatic skin colored papules, nodules, plaques symmetrically on torso and proximal upper extremities but localization to mons pubis as in our case has not been reported. Collagenoma in eruptive type are smaller than those of familial cutaneous collagenoma.

Table 1: Classification of collagenoma (Clinical variants)

<table>
<thead>
<tr>
<th>Collagenoma Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Familial cutaneous collagenoma (FCC)</td>
</tr>
<tr>
<td>Shagreen patch (Tuberous sclerosis)</td>
</tr>
<tr>
<td>Eruptive collagenoma</td>
</tr>
<tr>
<td>Plantar cerebriform collagenoma</td>
</tr>
<tr>
<td>Linear connective tissue naevus</td>
</tr>
<tr>
<td>Knuckle pads</td>
</tr>
<tr>
<td>Other collagenomas</td>
</tr>
</tbody>
</table>

Table 2: Classification of collagenoma (Genetic inheritance pattern).

<table>
<thead>
<tr>
<th>Genetic Inheritance Pattern</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inherited collagenoma</td>
</tr>
<tr>
<td>1. Familial cutaneous collagenoma</td>
</tr>
<tr>
<td>2. Dermatofibrosis lenticularis disseminate</td>
</tr>
<tr>
<td>3. Shagreen patch (tuberous sclerosis)</td>
</tr>
<tr>
<td>Acquired collagenoma</td>
</tr>
<tr>
<td>1. Eruptive collagenoma</td>
</tr>
<tr>
<td>2. Isolated collagenoma</td>
</tr>
</tbody>
</table>

CONCLUSION

Collagenoma may be a marker of internal disease like tuberous sclerosis, Down syndrome, pseudohypoparathyroidism and it may be present in isolated or eruptive pattern as in our case. To the best of our knowledge, only eight case reports of eruptive collagenoma could be retrieved by searching on PUBMED/MEDLINE, with none being reported in Down syndrome with unique localization to mons pubis.
Acknowledgements
Dr. Asha Kubba, Delhi Dermatology group.

Author Contributions
Balwinder Kaur Brar – 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
Mahajan B. B. – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Nidhi Kamra – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Balwinder Kaur Brar et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES


Single coronary artery arising from the right coronary sinus with mid-left anterior descending artery segment courses through the ventricular myocardium: A rare entity

Andrea Romagnoli, Irene Coco, Dominique De Vivo, Eros Calabria, Giovanni Simonetti

ABSTRACT

Introduction: Congenital anomalies of the coronary artery have an incidence of 1% and isolated single coronary artery without other congenital cardiac anomalies has an approximate incidence of 0.024–0.066% in general population. This rare entity can be diagnosed incidentally during life, however may lead to symptomatic or asymptomatic myocardial infarction and sudden heart attack, even among young athletes. The prognosis varies according to the anatomic distribution, associated atheromatous disease and associated vascular anomalies. Case Report: A 45 years old male with presentation of a single coronary artery from the right coronary sinus with subsequent coursing between the aorta and pulmonary trunk arteries and mid anterior descending artery segment course through the myocardium. Patient presented with non-typical angina symptoms, normal resting electrocardiograph, myocardial perfusion scintigraphy suspect to reduced left ventricular blood flow but negative angiographic evaluation. Conclusion: Thin bridges cannot be demonstrable angiography, so anatomic and panoramic CT-evaluation give more information especially if associated with functional evaluation. Knowledge of physiology, normal and variant anatomy, is most important in managing congenital and acquired disease, and variation in coronary arterial patterning is frequent.

Keywords: Coronary artery anomaly, Cardiac anomalies, Heart attack, Isolated single coronary artery, Myocardial bridge, Ventricular myocardium

How to cite this article
doi:10.5348/ijcri-201572-CR-10533

INTRODUCTION

Congenital anomalies of the coronary artery have an incidence of 1% and isolated single coronary artery without other congenital cardiac anomalies has an approximate incidence of 0.024–0.066% in general population. This rare entity can be diagnosed incidentally during life, however may lead to symptomatic or asymptomatic myocardial infarction and sudden heart attack, even among young athletes [1]. The prognosis varies according to the anatomic distribution, associated atheromatous disease and associated vascular anomalies.
CASE REPORT

A 45-year-old male was admitted in our department with symptoms characterized by intermittent substernal chest discomfort non-provoked by exertion or emotional stress enlarging for over four months. The patient was a smoker of 7–8 packs of cigarettes per week for 10 years. Clinical history was negative for hypertension, diabetes mellitus, hyperlipidemia or family history of premature vascular disease and no risk factors for pulmonary embolism or respiratory distress. Physical examination showed normal blood pressure (BP: 128/88 mmHg), heart rate of 73 beats/min, no signs of vascular disease and normal cardiac examination. Initial laboratory tests showed no particular anomalies with normal blood levels of glucose and cholesterol (LDL: 103 mg/dl; HDL: 35 mg/dl; TG: 198 mg/dl).

Resting electrocardiogram (ECG) showed normal sinus rhythm at 82 beats/min without anomalies in stress examination. Several echocardiographic examinations revealed a moderate enlargement of the left atrium; an apical form of left ventricular hypertrophy; a preserved left ventricular (LV) systolic function (LV ejection fraction, 57%); and moderate mitral valve regurgitation, with no abnormality of the mitral valve apparatus or of LV wall motion.

Three months ago the patient had undergone coronary angiography evaluation which demonstrated an anomalous vascular anatomy with absence of coronary ostium in left cusp of the aortic valve and a single coronary artery (SCA) with origin by the right coronary cusp with one and only common trunk (CT) which gives rise to right coronary artery (RCA), left anterior descending (LAD) and diagonal artery, circumflex branch (CB) and the proximal left marginal artery (LMA). The RCA gives off the posterior descending artery (PDA) and the postero-lateral branch (PL).

Significant flow-limiting stenosis was not detected with regular caliper representation of the three main coronary branches up to their distal segments. Contrast ventriculography confirmed the apical LVH and showed no wall motion abnormality.

In our department, the patient underwent CT scanning, which confirmed the anomalous origin and course of coronary vessels showing also an inter-arterial course of the CT between the pulmonary artery and the aorta (Figures 1–2). A 2-cm segment of the proximal-middle tract of the common trunk was embedded within the inter-ventricular septum, following an intramural course with a relative reduction of tunneled vessel (Figure 3).

No significant atheromatous alterations have been highlighted, if we except for some parietal irregularities at LAD.

Dipyridamole Tl-201 SPECT confirmed a perfusion abnormalities with reversible perfusion defects in the mid anteroseptal wall without an apical abnormality.

![Figures 1: Coronary computed tomography angiography with volume rendering reconstruction (1) and with Curved MPR (2) images demonstrated an anomalous vascular anatomy with absence of coronary ostium in left cusp of the aortic valve and a single coronary artery (SCA) with origin by the right coronary cusp with one and only common trunk (CT) which gives rise to right coronary artery (RCA), left anterior descending (LAD) and diagonal artery, circumflex branch (CB) and the proximal left marginal artery (LMA). The RCA gives off the posterior descending artery (PDA) and the postero-lateral branch (PL).](image1)

![Figures 2: Coronary computed tomography angiography with volume rendering reconstruction (1) and with Curved MPR (2) images demonstrated an anomalous vascular anatomy with absence of coronary ostium in left cusp of the aortic valve and a single coronary artery (SCA) with origin by the right coronary cusp with one and only common trunk (CT) which gives rise to right coronary artery (RCA), left anterior descending (LAD) and diagonal artery, circumflex branch (CB) and the proximal left marginal artery (LMA). The RCA gives off the posterior descending artery (PDA) and the postero-lateral branch (PL).](image2)
The patient was admitted to cardiology center for further evaluation, treatment and periodical follow-up.

**DISCUSSION**

Isolated SCA without other congenital cardiac anomalies is very rare among the different variations of anomalous coronary patterns occurring in approximately 0.024% of the population [1].

Origin of the LCA in the right coronary sinus has been described at a frequency of 0.02% in autopsy series and from 0.05–0.19% in angiographic series [2].

The LCA either has a common ostium with the RCA, or arises independently to the ostium of RCA. These cases are classified according to the course of LCA into four categories: (1) inter-truncal or inter-atrial, between the aorta and pulmonary arteries; (2) anterior, in front of the right ventricular outflow; (3) posterior or retro-aortic, behind the aorta; (4) inter truncal-septal or trans-septal, through the supraventricular crest and inter-ventricular septum. The retro-aortic course of the LCA is an uncommon entity. This anomaly is serious and associated with sudden cardiac death and myocardial infarction, if anomalous LCA passes between the aorta and the pulmonary artery [3, 4].

Although most patients with the anomalous LCA arising from the right sinus of Valsalva are asymptomatic, the therapeutic approach must be individualized in each subject. In asymptomatic subjects, the age of the patient and the type of anomalous artery should be carefully evaluated [6].

Usually, typical angina does not occur with SCA in the absence of coexisting coronary artery disease or aortic stenosis [1].

In our case, the patient’s symptoms could be attributable to the intramyocardial segment of LAD.

The myocardial bridge, occurs when the artery coursing within the myocardium, presents compression to the contraction of the heart muscle to systole, which is clinically silent most of the time [6].

A deepened critical analysis of many autopsy samples was first presented by Geiringer et al. in 1951: clinical interest and systematic research were triggered by the observation of myocardial bridging right along with myocardial ischemia [7].

The rate of angiographic bridging is <5%, linked to thin bridges that provide a light compression. Carrying out provocation tests in subjects presenting normal angiographic coronary arteries may enhance the systolic myocardial compression and could thereby demonstrate myocardial bridges in ≤40% of cases [8].

Myocardial bridges are preferably localized in the middle segment of the LAD [7].

One of the parallel LAD branches frequently keeps an intramural course [7].

Diagonal and marginal branches may be respectively involved in 18% and 40% of cases [6]. By the angiographic side, myocardial bridges are almost exclusively spotted in the LAD. They set at 1 to 10 mm depth showing typical length around ≈10 to 30 mm [7]. Ferreira et al. distinguished two types of bridging: (1) superficial bridges (75% of cases) crossing the artery in perpendicular way or taking an acute angle towards the apex, and (2) muscle bundles arising from the right ventricular apical trabeculae (25% of cases) that transversely, obliquely, or helically cross the LAD before they flow in the interventricular septum. Arterial segments could also be set in a deep interventricular gully. The segment proximal to the bridge frequently, because of hemodynamic forces, shows atherosclerotic plaque formation, although the tunneled segment is typically spared [9]. Neither low value proximal to the bridge stenosis, nor systolic compression of the tunneled segment are allowed by the way to explain a severe ischemia and its related symptoms. When the arterial occlusion was limited to the only systole, phasic coronary blood flow and distal coronary pressure were observed in a considerable delay, contributing to a smaller myocardial oxygen consumption and to the increase of the coronary sinus lactate concentration. Angina, myocardial ischemia, myocardial infarction, left ventricular dysfunction, myocardial stunning, paroxysmal AV blockade, as well as exercise-induced ventricular tachycardia and sudden cardiac death are reported as sequelae of the myocardial bridging. However, following the prevalence of myocardial bridging, these complications are rare. Patients may complain atypical or angina-like chest pain with unsure association between...
the severity of these symptoms and length/depth values of the tunneled segment, or the systolic compression degree [6]. The up-to-date gold standard for diagnosing myocardial bridges remains the coronary angiography presenting the typical “milking effect” and a “step down–step up” phenomenon induced by the systolic compression of the tunneled segment. By the way, these signs give insufficient information about the functional impact on myocardium. Proximal stenosis and myocardial bridging could only be identified by carrying out a percutaneous transluminal coronary angioplasty, because higher intravascular pressure values and reversed hypokinesis can reveal the myocardial bridging [10].

CONCLUSION

Coronary angiography evaluation has demonstrated an anomalous vascular anatomy with absence of coronary ostium in left cusp of the aortic valve. Computed tomography (CT) scanning has confirmed the anomalous origin and course of coronary vessels showing also an inter-arterial course of the CT between the pulmonary artery and the aorta. A 2-cm segment of the proximal-middle tract of the CT was embedded within the interventricular septum, following an intramural course with a relative reduction of tunneled vessel. Thin bridges can be not demonstrable angiography, so anatomic and panoramic CT-evaluation give more information especially if associated with functional evaluation. Knowledge of physiology, normal and variant anatomy, is most important in managing congenital and acquired disease, and variation in coronary arterial patterning is frequent.

Author Contributions

Andrea Romagnoli – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published
Irene Coco – Acquisition of data, Drafting the article, Final approval of the version to be published
Dominique De Vivo – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Eros Calabria – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Giovanni Simonetti – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Andrea Romagnoli et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES

ABOUT THE AUTHORS


**Andrea Romagnoli** is working in Department of Diagnostic Imaging, Molecular Imaging, Interventional Radiology and Radiotherapy, Tor Vergata University Hospital Foundation, University of Rome Tor Vergata, Italy.
E-mail: romagnoli.69@libero.it

**Irene Coco** is working in Department of Diagnostic Imaging, Molecular Imaging, Interventional Radiology and Radiotherapy, Tor Vergata University Hospital Foundation, University of Rome Tor Vergata, Italy.
E-mail: ire.coco@hotmail.it

**Dominique De Vivo** is working in Department of Diagnostic Imaging, Molecular Imaging, Interventional Radiology and Radiotherapy, Tor Vergata University Hospital Foundation, University of Rome Tor Vergata, Italy.
E-mail: dominiquedevivo@libero.it

**Eros Calabria** is working in Department of Diagnostic Imaging, Molecular Imaging, Interventional Radiology and Radiotherapy, Tor Vergata University Hospital Foundation, University of Rome Tor Vergata, Italy.
E-mail: eros.calabria@libero.it

**Giovanni Simonetti** is working in Department of Diagnostic Imaging, Molecular Imaging, Interventional Radiology and Radiotherapy, Tor Vergata University Hospital Foundation, University of Rome Tor Vergata, Italy.
E-mail: giovanni.simonetti@uniroma2.it

Access full text article on other devices

![QR Code](image1)

Access PDF of article on other devices

![QR Code](image2)
Cannabis consumption before surgery may be associated with increased tolerance of anesthetic drugs: A case report

Georg Richtig, Götz Bosse, Friederike Arlt, Christian von Heymann

ABSTRACT

Introduction: Cannabis is the most commonly used illicit drug worldwide in adults. Several studies suggest an interaction of cannabis intake and anesthetic drugs like propofol and thiopental, but the pharmacodynamic interaction between cannabis and commonly used anesthetic drugs is still not well understood. Case Report: A 37-year-old male patient with a history of regular cannabis consumption and cannabis intake the day before surgery was admitted for an arthroscopic shoulder stabilization. Induction of anesthesia required large doses of propofol, thiopental and sevoflurane. Maintenance was achieved with the additional application of nitrous oxide to provide clinical tolerance of the surgical procedure. Return of consciousness and the postoperative course were uneventful. Conclusion: History of cannabis intake should be taken into routine preoperative assessment and patients should be closely monitored by experienced anesthesia staff for adequate anesthetic depth and analgesia.

Keywords: Anesthesia, Cannabis, Interaction, Pharmacodynamics

INTRODUCTION

Cannabis is the most consumed illegal drug worldwide and there is a growing number of ongoing programs to offer a legal source of cannabis to chronically sick patients [1]. Several studies suggest an interaction of cannabis intake and anesthetic drugs like propofol and thiopental [2]. However, this interaction and its implications for anesthetic practice in patients with chronic cannabis intake is not well understood.

Herein, we describe a case of a patient scheduled for a routine surgical procedure with a history of regular cannabis and tobacco consumption, who needed large amounts of different anesthetic agents to provide sufficient condition for intubation.

CASE REPORT

A 37-year-old male patient, 93 kg weight, 206 cm tall, was scheduled for an arthroscopic stabilization of
his right shoulder due to a traumatic glenoid fracture. His medical history revealed an appendectomy at the age of nine with no anesthetic problems. The patient was healthy and in good medical conditions. He reported a daily nicotine usage of 10–15 cigarettes, a cannabis intake of 1 g per week and an occasional use of alcohol. He admitted smoking one cannabis cigarette the day before surgery.

In the operating room non invasive cardiorespiratory monitoring measured a blood pressure of 120/70 mmHg, a heart rate 80 bpm and an oxygen saturation (SpO2) of 98%. Prior to induction the patient was oxygenated with a FiO2 of 1.0 and a flow of 12 L/min for 5 minutes. A regional anesthetic technique like an interscalene block had been declined by the patient.

Over an iv-line on his left backhand (BD Venflon™ Pro Safety 18G, Becton Dickinson, Heidelberg, Germany) 0.2 mg of fentanyl (Fentanyl®-Janssen, Janssen-Cilag, Neuss, Germany) were injected and the patient reported of a dizzy feeling after four minutes. 200 mg of propofol (Braun Propofol®-Lipuro, B. Braun, Melsungen, Germany) were administered without any anesthetic effect as the patient was still speaking with the anesthesia staff. Another two doses of propofol of 100 mg each were injected, but the patient did not show any reduction of his conscious state. The iv cannula was checked for correct position and no signs of extravasation of the anesthetic agents were detected.

The anesthetic regime was changed. We decided to administer 500 mg of thiopental (Thiopental Inresa® 0.5 g, Inresa Arzneimittel, Freiburg, Germany), that finally induced anesthesia. However, the patient was still breathing spontaneously with a tidal volume of 100–150 ml and a respiratory rate of 20/min requiring assisted ventilation using a face mask that was not tolerated by the patient. 4 vol% of isoflurane (Forane®, Abbott, Ludwigshafen, Germany) was added to achieve tolerance of the assisted ventilation.

After three minutes of isoflurane-supplemented assisted ventilation a size 5 laryngeal mask (LMA/Unique™ Single Use, Teleflex Medical, Kernen, Germany) was inserted to further improve ventilation. The tidal volume was increased to 200 ml requiring a peak inspiratory pressure of 30 cm H2O. A further dose of 200 mg of propofol was administered to deepen the anesthesia so that the patient was finally sufficiently ventilated. Due to the need to position the patient on the left side, neuromuscular blockade was achieved with 100 mg succinylcholine and an endotracheal intubation was performed without any complications (Cormack II, tube size 8 mm, Covidien Mallinckrodt™ Hi–Contour, Covidien Germany, Neustadt/Donau, Germany).

After intubation sufficient ventilation was achieved by a tidal volume of 650 ml, a respiratory rate of 16 and an etCO2 of 40 to 45 mmHg. In the operating room anesthesia was maintained with an end-tidal volume concentration (etVol%) of 2.4% sevoflurane (Sevorane®, Abbott, Ludwigshafen, Germany). Nitrous oxide was added with a concentration of 50% and a flow of 1 L/min.

The entire induction period took 15 minutes from the application of fentanyl to the maintenance state with sevoflurane and nitrous oxide at the end. Only after the application of sevoflurane and nitrous oxide the patient had a significant drop in his blood pressure and heart rate. After fractional (4x2 ml) application of akrinor (caderidine hydrochlorid 200 mg and theodrenalinehydrochlorid 10 mg, Akrinor®, CT Arzneimittel, Berlin, Germany) the patient showed a stable blood pressure of around 110/60 mmHg through the whole procedure. For intraoperative analgesia a total amount of 0.5 mg of fentanyl was administrated. 20 minutes before the end of procedure 2 g metamizole (Novaminsulfon-ratiopharm®, Ratiopharm, Ulm, Germany) were applied. Since the patient had a constantly high blood pressure (150/80 mmHg) after extubation and no pain, he also received 60 µg clonidine (Catapresan®, Boehringer Ingelheim Pharma, Ingelheim, Germany). After 30 minutes of observation in the post-anesthesia care unit, the patient was fully awake, showed no signs of persisting anesthetic drug effect and was discharged to the surgical ward. Post-surgical pain was controlled with 500 mg metamizole every eight hours. The patient was discharged two days after surgery.

**DISCUSSION**

Herein, we report a case of a 37-year-old male with a regular cannabis abuse who required excessive doses of different anesthetic drugs for induction of anesthesia. We postulate that this might be related to a pharmacological interaction of cannabis and anesthetic drugs resulting in anesthetic drug dosage.

In literature, the effect of cannabis on anesthetic drugs is discussed controversially.

Flisberg et al. [2] could show, that patients with a history of cannabis consumption needed significant more propofol to successfully achieve the insertion of a laryngeal mask, than the control group. They concluded that regular cannabis users show a more variable response to induction of anesthesia with higher doses of propofol needed [2]. These human data are supported by Brand et al. [3] who reported a dose-dependent antagonistic interaction between tetrahydrocannabinol (THC) and propofol in mice.

In contrast, several experimental studies have also shown an enhancement of the sedative effect of intravenous [4] and inhalational [5] anesthetic drugs by THC, so that an individual dose titration and continuous monitoring of the anesthetic depth is recommended.

In our case, the use of thiopental and propofol did not seem to have the expected effect on the patient’s consciousness. Furthermore, the induction and maintenance of anesthesia with high concentrations of isoflurane and sevoflurane did not achieve adequate anesthetic depth. Only when nitrous oxide had been added to sevoflurane, sufficient tolerance of the surgical anesthesia...
procedure could be provided. In connection with pharmacokinetics, similar findings have been reported by Symons [6] describing a patient with a history of cannabis smoking, who required repetitive doses of propofol (3x50 mg), midazolam (2x1 mg) and ketolorac (10 mg), besides high concentrations of isoflurane and nitrous oxide to maintain anesthesia for a tooth extraction.

A possible pharmacodynamic explanation could be that propofol and thiopental mediate their effects over the gamma-aminobutyric acid type a (GABA (A)-receptor), like the volatile anesthetics, sevoflurane and isoflurane, which may have been influenced by THC and other cannabinoide compounds.

Liu et al. [7] observed that GABA (A)-receptors could form a complex with G-Protein-Coupled-Receptors (Dopamine-5-Receptor) resulting in a reduced efficacy of the GABA transmission.

So a possible explanation for the pharmacodynamic interactions in our case could be the ability of the CB1-Receptor and the GABA (A)-Receptor to form such a receptor complex resulting in a lower GABA neurotransmission. A hint for these theory, could be the fact that in our case no anesthetic - which mediated its effect through the GABA (A)-receptor - could provide optimal conditions for intubation. Only when nitrous oxide, which blocks the NMDA-receptor, was provided to our patient, sufficient anesthesia could be achieved and maintained.

It also needs to be mentioned, that plasma and blood concentrations of THC in frequent cannabis users (four times per week a cannabis cigarette with 54 mg THC content) are detectable up to more than 30 hours after consumption [8]. Moreover it has to be considered, that the amount of THC depends on the type of plant species, cultivation, climate and soil and ranges from 1–20% in herbal cannabis [9, 10]. Therefore, we would recommend, that surgical patients refrain from consuming cannabis as long as possible prior to surgery, since the way of interaction with anesthetic drugs is not safely predictable. The interaction of THC with anesthetic drugs is complex, possibly resulting in increased sensitivity [4] or greater tolerance to anesthetic drugs, the latter with a higher need of anesthetic agents to induce anesthesia. In patients with greater tolerance of anesthetic drugs nitrous oxide, could be one possible solution to avoid possible interactions between GABA (A)-receptor signaling and THC. Since the mode and the effect of interaction is not predictable history of cannabis intake should be taken into routine preoperative assessment.

Limitations of our reported case are, that we did not have any monitoring of depth of anesthesia applied, a relatively low fentanyl induction dose (200 µg, 2 µg/kg), no THC blood level measurements and no knowledge of any genetic mutation of the patients GABAA-receptor.

CONCLUSION

Patients with regular cannabis consumption are recommended to closely monitored for adequate anesthetic depth and analgesia, treated by experienced staff only and regional anesthetic techniques should be favored. Patients are recommended not to consume cannabis as long as possible before the surgical procedure.

**********

Author Contributions

Georg Richtig – 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

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

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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

© 2015 Georg Richtig et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES

Primary hydatid cyst of gallbladder: A case report

Pronoy Kabiraj, Shib Shankar Kuiri, Utpal De

ABSTRACT

Introduction: Hydatid cyst is a common clinical entity in India. Liver and lungs are the common organs involved. No organ is immune to infection. Case Report: We report a case of primary hydatid cyst of gallbladder presenting as a gallbladder lump. Conclusion: In endemic regions hydatid cyst should be considered in patients presenting with gallbladder lump.

Keywords: Echinococcus granulosus, Echinococcus multilocularis, Gallbladder, Hydatid cyst, Liver, Lungs

INTRODUCTION

Hydatid cyst in humans is an accidental dead end infestation caused by Echinococcus granulosus. It is endemic in sheep rearing countries. In India, Jammu and Kashmir is an endemic region though reports of hydatid disease has been reported from almost whole of the Indian subcontinent [1]. Liver (75%) and lungs (15%) are common sites of infection in humans [1, 2]. Primary extrahaepatic intra-abdominal hydatid cyst is a rare clinical entity. This article reports and reviews a case of primary gallbladder hydatid (Table 1).

CASE REPORT

A 16-year-old female patient admitted suffering from intermittent mild pain in the right hypochondrium and epigastrium associated with nausea since six months. There was no history of fever or jaundice. Physical examination revealed mild tenderness in the right upper quadrant of the abdomen. On deep palpation a globular lump (4 cm x 3 cm), moving with respiration was noted in the right hypochondrium. The upper margin of the lump was impalpable while the other margins were well defined. Murphy’s sign was positive.

Base line hematological examination was unremarkable. Plain abdominal X-ray was normal. The ultrasound examination showed that her gallbladder was distended with a localised thickening of its wall. A single cystic lesion was noted inside the gallbladder lumen giving it an impression of double gallbladder. There was no image of gallstone.

An abdominal CT scan was performed showing inflammatory gallbladder wall with a cyst within cyst appearance (Figure 1). Hepatic parenchyma and other abdominal organs proved absolutely normal.

Anti-Echinococcus antibodies were not found in serum. The diagnosis of primary hydatid cyst of gallbladder was made and surgery was decided. The patient underwent right subcostal laparotomy. Intraoperatively, the gallbladder was found to be tense and hugely...
distended (Figure 2). The wall appeared oedematous and inflamed. After proper precaution the gallbladder wall was opened (Figure 3). A single cyst with germinal layer was delivered intact. No other cysts were found in the liver and peritoneal cavity. Cholecystectomy was performed. It permitted a total removal of the cyst without rupture. A peroperative cholangiography searching daughter cysts in the common bile duct proved unremarkable. The patient’s postoperative course was uneventful and she was discharged on tenth postoperative day after stitch removal. The patient was put on albendazole (10 mg/kg) in divided doses. The histopathology confirmed the presence of hydatid cyst (Figure 4). At sixth month follow-up, the patient was well and had no recurrence of hydatid disease.

**DISCUSSION**

Human hydatosis is a zoonosis caused by *Echinococcus granulosus* and *Echinococcus multilocularis* (malignant hydatid). *Echinococcus granulosus* predominates in humans. Cattles (sheep, goat, dogs) are primary (definitive, sexual cycle) host. Human beings are secondary dead end (asexual cycle) host [1–5]. Human infection is caused by fecal-or oral route or ingestion of contaminated vegetables or meat [1–9]. Cystic transformation of *Echinococcus granulosus* in human organs results in varied symptoms. No organ is immune to echinococcal infection [5]. Liver (first filter) and Lungs (second filter) are the commonest organs involved worldwide. In children, due to unknown cause lung infection (41–70%) is more common than liver (42–48%) [5–7]. Other organs are involved secondary to pulmonary or hepatic involvement. Among the unusual primary sites, in order of frequency, central nervous system (4–5%), musculoskeletal system (0.5–4%) and cardiovascular system (0.5-2%) are involved [2, 5, 6, 9].

The incidence of primary extrahepatic intra abdominal hydatid cyst is 6.5% [1–9]. Spleen (2–3%) is the predominant organ involved [1, 3–5]. Gallbladder is primarily involved in 0.6%. GB cysts may be located intraluminally or on the surface [1–9]. As a rule luminal cysts are single and parietal cysts are multiple containing daughter cyst [2, 5]. Luminal cyst may attain enormous size before they are symptomatic due to distensible capacity of the gallbladder. On the contrary parietal cysts produce early symptoms. The pathogenesis of gallbladder infection is not well documented. The hypothetical routes of spread include portal circulation, biliary, lymphatic and peritoneum [1–9].

Univariate presentation of gallbladder hydatid include acute cholecystitis with cholangitis, gallbladder

<table>
<thead>
<tr>
<th>Serial No</th>
<th>Author(s)</th>
<th>Title</th>
<th>Journal</th>
<th>Year</th>
<th>Pubmed PMID</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Pautov FN.</td>
<td>A case of primary alveolar echinococcosis of the gallbladder</td>
<td>Med Parazitol (Mosk).</td>
<td>1964</td>
<td>14250141</td>
</tr>
<tr>
<td>2</td>
<td>Rigas et al.</td>
<td>Primary hydatid cyst of the gallbladder</td>
<td>Br J Surg.</td>
<td>1979</td>
<td>466022</td>
</tr>
<tr>
<td>3</td>
<td>Kapoor et al.</td>
<td>Sonographic diagnosis of a ruptured primary hydatid cyst of the gallbladder</td>
<td>J Clin Ultrasound.</td>
<td>2000</td>
<td>10602107</td>
</tr>
<tr>
<td>5</td>
<td>Pitiakoudis et al.</td>
<td>Primary hydatid disease in a retroplaced gallbladder</td>
<td>J Gastrointestin Liver Dis.</td>
<td>2006</td>
<td>17205152</td>
</tr>
<tr>
<td>6</td>
<td>Krasniqi et al.</td>
<td>Primary hydatid cyst of the gallbladder: a case report</td>
<td>J Med Case Rep.</td>
<td>2010</td>
<td>20205877</td>
</tr>
<tr>
<td>8</td>
<td>Noomene et al.</td>
<td>Primary hydatid cyst of the gallbladder: an unusual localization diagnosed by magnetic resonance imaging (MRI)</td>
<td>Pan Afr Med J.</td>
<td>2013</td>
<td>23504393</td>
</tr>
</tbody>
</table>
lump, obstructive jaundice or peritonitis with shock due to rupture. A combination of the above symptoms is not infrequent. Gallbladder hydatid with daughter cysts in the common duct producing obstructive symptoms may give rise to a false positive Courvoisier’s law [2, 5–9]. So in endemic areas this should be included in the differential diagnosis of patients with obstructive jaundice.

Abdominal sonography is highly sensitive and specific for diagnosing hydatid cyst but accurate extrahepatic organ localization is at times doubtful [5, 8, 9]. Computed tomography, and magnetic resonance imaging, are adjuncts to sonography for definitive diagnosis. Serological (ELISA) and hematological (eosinophilia) tests may clinch the diagnosis along with radiology [4, 6–9].

Cystoprostatectomy (cholecystectomy) is the treatment of choice [2–5, 8, 9]. Apart from the usual peroperative precautions gentle handling of the gallbladder is of utmost importance. An intraluminal rupture may
result in spillage into common bile duct. Manual handling instead of instrumental (application of Moynihan’s clamps) holding of the gallbladder during dissection is warranted [1, 7]. Cystic duct should be identified early and stay suture passed in order to prevent accidental spillage of contents into CBD in the event of rupture of cyst. Careful ligation of cystic duct and artery together with removal of gallbladder from liver bed along with a portion of the liver tissue prevents rupture and spillage. Postoperative treatment with albendazole (10–15 mg/kg) in divided doses should be continued for a period of four months to prevent recurrence [1–9].

**CONCLUSION**

In endemic countries, hydatid cyst should be considered in the differential diagnosis in patients presenting with gallbladder lump.

*********

**Author Contributions**

Pronoy Kabiraj – 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

Shib Shankar Kuiri – Substantial contributions to conception and design, Drafting the article, Critical revision of the article, Final approval of the version to be published

Utpal De – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

**Guarantor**

The corresponding author is the guarantor of submission.

**Conflict of Interest**

Authors declare no conflict of interest.

**Copyright**

© 2015 Pronoy Kabiraj et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

**REFERENCES**


Occult hip fracture diagnosed by MRI scan after inconclusive X-ray and CT scan

Bogdan Deleanu, Radu Prejbeanu, Florin Birsasteanu, Dinu Vermesan, Liviu-Ionut Micle, Eleftherios Tsiridis, Vlad Predescu

ABSTRACT

Introduction: Hip fractures are common, most of them being easily diagnosed with the clinical examination and X-ray. Few cases need a computed tomography (CT) scan and even fewer an magnetic resonance imaging (MRI) scan. Here we report a case that was diagnosed with occult femoral neck fracture only after undergoing a pelvic MRI scan. Case Report: The patient has presented a normal X-ray at the first exam with minimal clinical signs. The emergency CT scan shown no injury and the patient was left home. Three days after, the patient returned to hospital with increased pain and limited mobility to the left hip. Following the emergency MRI scan a femoral neck fracture has been revealed. The patient was soon operated and we used a total hip cemented prosthesis on the affected hip considering the age and the condition of the patient. Conclusion: Occult hip fractures should be suspicioned in all elderly patients who have a history of hip trauma even if X-ray and CT scan do not reveal a fracture. If it’s available, the use of MRI scan is the best option for diagnosis of occult hip fractures.

Keywords: CT-scan, MRI-scan, Occult hip fractures

INTRODUCTION

Hip fractures are one of the most frequent types of fractures seen in orthopedic practice [1, 2]. Considering that life expectancy increased more and more in the past few decades the elderly population increased too [3], so much more attention is needed to properly diagnose hip fractures which occur more often in this segment of population [4]. Standard anterior-posterior X-ray may help us to diagnose displaced fractures but in non-displaced fractures a CT scan or an MRI scan is recommended [5, 6]. Studies show that occult hip fractures represent 2–10% of total hip fractures [5, 7], and the MRI scan has the best accuracy in detecting these fractures [5–9]. It is considered that late diagnose of occult hip fractures occur in 2–9% of the cases [10]. Holder et al. showed that CT scan performed in less than 3 days after trauma has 93% sensitivity and 95% specificity, so there is a significant percent of occult hip fractures missed by CT scan [11, 12].

How to cite this article

Here we report a case of occult femoral fracture that was diagnosed using a pelvic MRI scan 3 days after an X-ray and a CT scan shown no injury. The purpose is to show the importance of MRI scan in diagnosing occult hip fractures.

CASE REPORT

A 72-year-old female had a history of left hip trauma after tripping and falling at home. She was brought immediately to the emergency department and an anterior-posterior hip X-ray was done (Figure 1). As seen in the X-ray there was no sign of fracture but the clinical examination revealed pain at the left hip and limited mobility. Thus, considering the age of the patient, a pelvic CT scan was requested which also revealed no injury to the left hip (Figure 2). The patient was discharged and left home with anti-inflammatory and pain medication, being programmed to return after 7 days for a follow-up.

After three days the patient returned to emergency department with increased pain to the left hip and almost no active mobility possible to the affected hip. We decided to do a pelvic MRI scan which revealed an occult left femoral neck fracture (Figure 3).

The second day the patient was operated. We have chosen a hip direct lateral approach (Hardinge) with the patient in supine position. Considering the age of the patient and her associated conditions we opted for a cemented total hip arthroplasty in order to have the best results (Figure 4). The second day postoperative the patient started rehabilitation with bed side exercises and at third day postoperatively weight bearing walk was possible. Follow-up at 45th day, the patient had an overall good recovery with a hip Harris score of 78.

DISCUSSION

The best imaging option for diagnosing hip fractures is still a subject of discussion. Obviously this discussion applies to the undisplaced and occult fractures.

X-ray exam is a primary investigation used in pre-operative planning but by definition an occult hip fracture is not visible on X-ray exam so using only this exam is not helpful in diagnosing such cases. Due to increased cost and limited availability in some hospitals the MRI-scan is not a primary imaging option, as seen in this case.

Studies show that MRI scan has the best sensitivity and it should be used as a primary choice for diagnosing this type of fractures [6–9, 13]. Both orthopedic surgeons and radiologists agree that MRI scan is the gold standard in diagnosing occult hip fractures [5–9, 13]. It is recommended that MRI scan to be done in the first 24 hours after the hip trauma. If not available, a 72-hour CT scan or MRI scan is recommended.

CT scan is another imaging option which can be helpful but as seen in this case the results may be inconclusive or can give false-negative results. The CT scan has the advantages to be widely available with a lower cost comparative to MRI scan [5, 14], but even so the MRI is preferable. The advantages of MRI scan over CT scan are the better sensitivity (100%) and no exposure to harmful radiation [15]. The literature also mentions the bone scintigraphy using technetium polyphosphate with
available remains the MRI scan and it should be the first to be used in cases with hip trauma and no X-ray signs, especially in the elderly population. Preferably the MRI should be performed in the first 24 hours after trauma to decrease the complications and obtain the best curative results.

**********

Author Contributions
Bogdan Deleanu – 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
Radu Prejbeanu – 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
Florin Birsasteanu – Analysis and interpretation of data, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Dinu Vermean – Analysis and interpretation of data, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Liviu-Ionut Micle – Analysis and interpretation of data, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Eleftherios Tsiridis – Analysis and interpretation of data, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Vlad Predescu – Analysis and interpretation of data, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Bogdan Deleanu et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.
REFERENCES


Intramedullary spinal cord abscess by *Nocardia*: A case report

Khaled Alshabani, Joseph Adrian L. Buensalido, Milagros P. Reyes, Ayman O. Soubani

**ABSTRACT**

Introduction: *Nocardia* is a Gram-positive bacteria that usually cause opportunistic infections but can affect the immunocompetent host. The central nervous system can be the primary site of infection in up to 40% of the cases but the involvement of the spinal cord is extremely rare. Case Report: We present a case of a middle age man with history of alcoholic liver cirrhosis and diabetes mellitus who presented with acute lower back pain that is associated with right leg weakness and numbness. Magnetic resonance imaging (MRI) scan showed a spinal cord intramedullary abscess. The cerebrospinal fluid analysis was consistent with meningitis and the organism was eventually identified with CSF culture as *Nocardia Farcinica*. Patient was treated with prolonged intravenous trimethoprim/sulfamethoxazole and meropenem. No neurosurgical intervention was done and the patient had full neurological recovery in few months. Discussion: *Nocardia* species are able to cause different kinds of disease in man. Spinal cord abscess are extremely rare and high index of clinic suspicion is required for diagnosis. Initial combination intravenous antibiotics treatment is essential and should be continued for at least three to six weeks. Treatment can be switched to oral and continued for a minimum of six months. Surgical intervention might be indicated in some patients. 16s rRNA gene sequencing allows earlier identification and thus adjustment of antibiotics. Conclusion: This report illustrates that a diagnosis can be made by neuroimaging and cerebrospinal fluid (CSF) 16s ribosomal RNA sequencing, allowing early and effective antibiotic therapy and obviating the need for a high-risk neurosurgical intervention.

Keywords: Gram-positive bacteria, Intramedullary abscess, *Nocardia*, Spinal cord

How to cite this article


**INTRODUCTION**

*Nocardia* is known to be ubiquitous in the environment and is able to cause different kinds of diseases in man, including skin and soft tissue infections, pneumonia, central nervous system (CNS) infection and bacteremia [1]. *Nocardia* infections commonly affect immunocompromised patients and those who are on corticosteroid therapy. The lungs are usually the first organs that are infected, but the CNS eventually becomes...
infected in around 40% of cases if the primary infection is left untreated [2]. Brain abscess formation is the most common presentation of Nocardia CNS infection. Spinal cord involvement with intramedullary abscess formation is exceedingly rare. We present a rare case of such neurological involvement as the initial presentation of Nocardia infection. We also review literature (Table 1) for this rare presentation [3–12].

CASE REPORT

A 53-year-old Caucasian male with past medical history of alcoholic liver cirrhosis, diabetes mellitus, and hypertension presented to the emergency department with acute onset lower back pain and inability to walk for two days. He also had numbness and weakness in the right leg, urinary retention and constipation. Physical examination was remarkable for weakness in the lower extremities that was worse on the right side, diminished pinprick and vibration sensation below the mid-thighs bilaterally, and hyporeflexia in both legs. He had mild tenderness over the lower lumbar spine.

Initial blood tests included complete blood count, basic metabolic profile, Erythrocyte sedimentation rate, and C-reactive protein were all within normal limits except for thrombocytopenia.

The patient was admitted to the general medical floor and started on intravenous (IV) dexamethasone. Computed tomography (CT) scan of the spine showed no cord compression. However, given the high clinical suspicion, a magnetic resonance imaging (MRI) showed a signal abnormality and enhancement of the spinal cord, including the conus medullaris, consistent with intramedullary abscess (Figure 1).

A lumbar puncture was performed and cerebrospinal fluid analysis (CSF) showed a red blood cell (RBC) count of 1621 RBCs/µL, nucleated cells of 4822 WBCs/µL with neutrophilic predominance (84% neutrophils), protein of 306 mg/dL, and glucose < 1 mg/dL. The Gram stain was negative. The patient was then started on broad spectrum IV antibiotics.

The initial CSF culture showed Gram-positive bacilli partially acid fast with some branching. Mycobacterium tuberculosis PCR was negative and patient was initiated on intravenous trimethoprim/sulfamethoxazole (TMP/SMX) and meropenem for Nocardia double coverage. Patient was evaluated by neurosurgery and it was decided that it was in his best interest not to intervene on the abscess as the risks outweighed the benefits. On day 10 of admission, and using 16s ribosomal RNA gene sequencing, the organism was identified as N. farcinica.

The CSF culture was finalized on day 20 of his admission and showed Nocardia farcinica, sensitive to TMP/SMX, amikacin, and imipenem, but resistant to ceftriaxone. Patient weakness started to improve slowly towards the end of his admission. He was eventually discharged to a subacute rehabilitation center on IV antibiotics. A six-month follow-up MRI scan showed resolution of the intramedullary abscess.

DISCUSSION

Nocardia is a Gram-positive bacteria of the actinomycetes group. It is responsible for nocardiosis, an uncommon infection. Nocardia species can cause localized or systemic disease usually in immunocompromised patients as an opportunistic infection, but one third of the cases occur in healthy individuals [13]. Nocardia is defined as a complex that includes N. asteroides sensu stricto, N. farcinica, N. nova, and N. transvalensis complex. Nocardia species are not part of the human flora. They are known to be ubiquitous in the environment [1]. Inhalation is the most common mode of entry and that explains the lung involvement in a majority of the infections. It can also result from trauma-related introduction of the organism [12].

Immunosuppression is a major risk factor for attracting the disease. HIV, malignancies, corticosteroid therapy, organ and hematopoietic stem cell transplantation are the most common causes of immunosuppression leading to Nocardia infections. Diabetes mellitus is a prominent risk factor. Alcoholism and COPD are important risk factors for pulmonary nocardiosis [13]. Nocardia species are able to cause different kinds of disease in man, including skin and soft tissue infections, pneumonia, CNS infection and bacteremia. The most common site of primary infection is the lung (up to
70%) [5]. CNS involvement is common in disseminated nocardiosis, but can also be the primary site [9]. CNS involvement can also occur in up to 40% of untreated primary lung lesions [2]. Brain abscess is the most common presentation of CNS nocardiosis [10]. Nocardia epidural abscesses have also been described.

Spinal cord involvement with intramedullary abscess formation is exceedingly rare and only ten cases have been reported in literature [3–12]. Our case is number 11. Yenrudi et al. reported a case of disseminated Nocardia, but we excluded it because the spinal cord involvement was secondary to thromboses of several blood vessels resulting in widespread myelomalacia and not by direct infection by the organism [14]. Of these cases, six patients were males. The mean age was 58.27 years (SD +/- 16.11). Comorbid conditions were reported in seven cases besides our patient, and included hypertension, diabetes mellitus (in four patients), end-stage renal disease, renal transplant, liver cirrhosis, chronic obstructive pulmonary disease, and ulcerative colitis. One patient was mistakenly diagnosed with allergic alveolitis, but her symptoms were secondary to the pulmonary nocardiosis as he improved dramatically with Nocardia treatment. Table 1 summarizes the main features of these cases.

Antimicrobial susceptibility patterns are variable between different studies, countries and species. Therefore, formal antimicrobial susceptibility testing is always necessary to ensure optimal therapy. There are rapid tests that can identify the organism much faster than culture and knowing the organism itself with these tests, can allow us to pattern our two empiric antibiotics by relying on studies that show the susceptibility profiles of specific species [1]. 16s ribosomal RNA gene sequencing is also a quicker method for coming up with the specific species of Nocardia, and thus allowing an earlier and improved empiric therapy while cultures are pending [15], as we did in our case.

Treatments strategies are based on cumulative retrospective experience, as well as in vitro studies, as no prospective randomized trials have been performed to determine the most effective treatment. Given the variable antibiotics resistance of Nocardia, empiric coverage with two or three agents in a patient with severe disease is warranted. TMP/SMX is a first line and standard therapy and should be included in these regimens. Combination

<table>
<thead>
<tr>
<th>Author and year reported</th>
<th>Patient's age and gender</th>
<th>Extension of nocardiosis</th>
<th>Type of Nocardia</th>
<th>Diagnosis</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kampfl [4] 1992</td>
<td>63 y/o male</td>
<td>Primary CNS: multiple cerebral and cervical spinal cord abscesses</td>
<td>N. asteroides</td>
<td>CSF consistent with bacterial meningitis + high titers of serum Nocardia IgG + complete resolution of radiological findings and partial clinical recovery with treatment</td>
<td>TMP/SMX + amikacin</td>
<td>Partial recovery 3 months after discharge</td>
</tr>
<tr>
<td>Hiller [5] 1999</td>
<td>56 y/o female</td>
<td>Disseminated: Lung + brain + single spinal cord lesion</td>
<td>N. asteroides</td>
<td>Spinal cord biopsy (consistent with abscess) + BAL. Culture positive for Nocardia</td>
<td>IV TMP/SMX</td>
<td>Partial recovery in two months</td>
</tr>
</tbody>
</table>
therapy is warranted in severe infections, defined as CNS involvement, disseminated disease, and infections involving more than one site in immunocompromised patients. Agents available for combination therapy include amikacin, imipenem, meropenem, third generation cephalosporins, minocycline, extended spectrum fluoroquinolones (e.g., moxifloxacin), tigecycline, and dapsone [1, 16].

Initial treatment should be administered intravenously for at least three to six weeks and/or until clinical improvement is documented. Treatment can be switched to oral and continued for a minimum of six months and up to 12 months in immunocompromised patients and those with CNS involvement.

Surgical intervention might be indicated in some situations, such as abscesses that do not respond to antibiotic therapy [12]. Prognosis is good in general, even in disseminated cases, if therapy is initiated early and continued for appropriate duration.

High index of suspicion of Nocardia infection is necessary in patients with suggestive neuroimaging. Spinal tap and stains and cultures for Nocardia are important part of the work up, but 16s rRNA allows earlier identification of the specific species, allowing adjustment of antibiotics, given the distinctly variable antibiotic susceptibilities of Nocardia species. A multidisciplinary approach that includes infectious disease, neurosurgical and neurological specialists is essential for early diagnosis and appropriate management of these patients.

CONCLUSION

This report describes a rare case of Nocardia intramedullary spinal abscess. It illustrates that a diagnosis can be made by neuroimaging and CSF 16s ribosomal RNA, with subsequent CSF culture confirmation. Appropriate antibiotic therapy was
effective and resulted in significant recovery and obviated the need for a high-risk neurosurgical intervention.

Author Contributions
Khaled Alshabani – Substantial contributions to conception and design, Acquisition of date, Analysis and interpretation of date, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Joseph Adrian L. Buensalido – Substantial contributions to conception and design, Acquisition of date, Analysis and interpretation of date, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Milagros P. Reyes – Substantial contributions to conception and design, Analysis and interpretation of date, Revising the article critically for important intellectual content, Final approval of the version to be published
Ayman O. Soubani – Substantial contributions to conception and design, Analysis and interpretation of date, Revising the article critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Khaled Alshabani et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES
Perianal aggressive angiomyxoma in a male patient

Zainab Taha ALHumoud, Najla Aldaoud, Hussain Abrar, Amro Salem

CASE REPORT

A 45-year-old male presented with a swelling in the perianal area that had begun to grow four months previously. He complained of pain, and problems defecating. Physical examination located a soft lump in the perianal area, with ill-defined edges near to the posterior midline (Figure 1). Although it is a rare condition, one of our preliminary differential diagnoses was lipomas. Other differential diagnosis was possible Desmoids Type Tumor.

Magnetic resonance imaging (MRI) scan of the pelvis showed a large flask shaped mass sagging from the perineum, with fat lobules apparently originating from perineum, extending into the intergluteal cleft. There was concern about the swirled appearance noted post contrast study, which suggested possible diagnosis of a myxoid type tumor and less likely a lipoma (Figure 2A–B).

Surgical excision of the mass was performed. The gross appearance of the specimen was a disoriented mushroom shaped mass, about 380 grams in weight, with a fatty cut surface (Figure 3). Microscopically, it showed an ill-defined tumor composed of small spindle cells embedded in an abundant myxoid stroma interspersed with a thick wall and hyalinized blood vessels. There was no evidence of mitotic activity or nuclear atypia (Figure 4A–C). Based on these histological features, a diagnosis of aggressive angiomyxoma (AAM) was confirmed.

DISCUSSION

Aggressive angiomyxoma (AAM) is a rare benign soft tissue tumor of mesenchymal origin, characterized by its...
locally infiltrative nature and high recurrence rate. It was
given this name due to the pathological findings of the
stellate and spindled cells, along with variable-sized blood
vessels, which were intricately entwined within a myxoid
matrix [1]. This presentation occurs predominantly
in woman of reproductive age (female to male ratio of
approximately 6:1), and exclusively in the pelvi-perianal
region. To our knowledge, only 43 cases occurring in men
have been reported in literature [1]. In men, AAM usually
involves the scrotum (38%), spermatic cord (33%), and
perineal region (13%) [2].

Although clinical diagnosis of AAM may be difficult, the
typical MRI features of AAM are swirled strands, aligned
with the craniocaudal axis. This particular imaging feature
is caused by a stretching of the fibrovascular stroma [3].
Confirmation of diagnosis is based on a histopathology
study. AAM should be distinguished from other benign,
potential lesions with low local recurrence, and malignant
tumors with widespread metastatic potential.

Surgery is the mainstay of treatment for AAM, although
achieving negative resection margins is complicated,
because of the infiltrative nature of the tumor. Local
recurrence rate falls between 36% and 72% and is usually
seen in the first three years. Recurrences of the disease
are usually controlled with surgery [1]. Several reported
attempts at using chemotherapy and radiotherapy as
part of the treatment protocol for AAM have proved
disappointing; probably due to the low mitotic activity/
growth fraction of cells. Primary treatment with GnRH
agonists has proved successful, as many of angiomyxoma
are positive for estrogen and progesterone receptors [4].
However, the duration of the responses and an optimal
treatment schedule are still unknown [1].

CONCLUSION

In conclusion, whether the treatment of aggressive
angiomyxoma (AAM) is surgery, hormonal therapy or
a combination of the two, it is apparent that adequate
management of AAM necessitates close, and long-
term follow-up at sixth month intervals, to monitor for
recurrence. Multiple relapses can occur but metastases
are unusual.

How to cite this article

ALHumoud ZT, Aldaoud N, Abrar H, Salem A.
Perianal aggressive angiomyxoma in a male patient.
doi:10.5348/ijcri-201522-CL-10077

*******
Acknowledgements
We would like to thank Dr. Thabet Ghazal who was involved in the Management of the case.

Author Contributions
Zainab Taha ALHumoud – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Najla Aldaoud – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published
Hussain Abrar – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published
Amro Salem – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Zainab Taha ALHumoud et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES

SUGGESTED READING
Bowen’s disease involving the dorsal and volar aspects of left thumb: An unusual site diagnostic entity

Shagufta Rather, Peerzada Sajad, Iffat Hassan

CASE REPORT

Herein, we report a case of a 40-year-old normotensive, non-diabetic, euthyroid male patient who presented with a non-healing erythematous slightly scaly plaque involving the volar and dorsal aspects of left thumb from last eight months (Figures 1 and 2). There was history of pruritus and slight burning sensation. There was a history of application of potent topical steroids for few months with no response. A punch biopsy was done which confirmed the diagnosis of eczema psoriasis and Bowen’s disease. On histopathology, the patient showed thickened epidermis with full thickness dysplasia of squamous epithelium and atypical keratinocytes with numerous mitotic figures. The basement membrane was intact with no focus of invasion or solar elastosis (Figures 3 and 4). Thus a diagnosis of Bowen’s disease was made. The patient was put on topical 5-fluorouracil and is doing well.
Bowen's disease (BD) is a form of intra-epidermal or in-situ squamous cell carcinoma with a small potential for invasive malignancy, and commonly involves chronically photo-exposed areas, especially head and neck region. Commonly a persistent, non-elevated erythematous scaly and crusted plaque is seen. Bowen's disease may occur at any age in adults, but is rare before the age of 30 years; most patients are aged over 60. Any site may be affected, although involvement of palms or soles is uncommon. Bowen's disease occurs predominantly in women in whom about 60-85% of patients have lesions on the lower leg, usually in previously or presently sun-exposed areas of skin. Chronic ultraviolet radiation exposure, arsenic exposure, human papillomavirus and immunosuppression are the various aetiological factors [1–3].

Histopathology is characterized by full-thickness dysplasia of the epidermis, with loss of the normal maturation of its components. Keratinocytes are atypical and disorderly, often described as having a windblown appearance. Basement membrane is intact. Topical 5-fluorouracil, imiquimod, photodynamic therapy, cryotherapy and excision are the various treatment modalities [4].

**CONCLUSION**

The head, neck, and extremities are the most commonly affected anatomic locations in men, while the lower limbs and cheeks are most commonly affected in women. Involvement of palms and soles is an unusual site of occurrence, but it should be considered in the differential diagnosis of non-healing erythematous scaly plaques.

**DISCUSSION**

Figure 3: Histopathological picture (x400) of Bowen’s disease.

Figure 4: Histopathological picture (x100) of Bowen’s disease.

How to cite this article

doi:10.5348/ijcri-201523-CL-10078

**********

Author Contributions
Shagufta Rather – 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
Peerzada Sajad – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Iffat Hassan – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2015 Shagufta Rather et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original
author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES


Inflammatory breast cancer (IBC) is considered as the most aggressive type of locally advanced breast cancer that carries an appreciably poor prognosis. Sir Charles Bell described the first case of IBC in literature. It was reported and published in 1814 by Sir Charles Bell [1]. Clinically, IBC is defined by characteristic features, including rapid onset within short time, erythema, edema of the breast, and a “peau d’orange” appearance to most areas of breast skin. Moreover, patients presented with positive metastatic lymph node involvement and up to one fourth of patients have distant metastases at time of diagnosis [2]. Pathologically, the presence of dermal lymphatic tumor emboli is considered as the hallmark of IBC. Previously, single modality treatment to manage IBC had failed. The majority of patients developed recurrence and/or metastases within short time, and 5-year survival rate was less than 5%.

Today, the general consensus is that patients with IBC without proof of metastases at the time of diagnosis should receive neoadjuvant chemotherapy followed by surgery followed by radiotherapy. For patients with human epidermal growth factor receptor (HER2) disease, trastuzumab (an antibody targeting HER2) is recommended. For patients with hormone receptor–positive disease, hormonal therapy is indicated. Regarding chemotherapy, the sequence of taxane-based chemotherapy followed by anthracycline-based chemotherapy is the cornerstone of primary chemotherapy for IBC [3]. Among patients with IBC, response to neoadjuvant chemotherapy plays a comparable prognostic role to that observed among patients with non-IBC.

Mastectomy is generally considered an important part of the multimodality treatment of IBC. The only method of definitive surgery offered to patients with IBC following neoadjuvant chemotherapy is modified radical mastectomy. Skin-sparing mastectomy and breast conserving surgery are contraindicated for patients with IBC [4]. Postmastectomy chest wall radiotherapy (RT) is generally indicated for patients with inflammatory breast cancer who are treated with neoadjuvant chemotherapy.

Understanding the biological characteristics of the disease has allowed for the development of targeted therapies (e.g., trastuzumab and lapatinib) that are improving the outcome of this aggressive disease. Human epidermal growth factor receptor 2 (HER2) positive patients should receive HER2 targeted therapy with neoadjuvant chemotherapy. Trastuzumab should be continued after surgery to complete one year of treatment [5]. Lapatinib is an orally active small molecule that reversibly inhibits the tyrosine kinase component of both HER2 and epidermal growth factor receptor-1 (ErbB-1). A recent research showed that lapatinib in conjunction with chemotherapy reduces the risk of disease progression by 50% in women with inflammatory breast cancer whose disease had progressed on trastuzumab-containing chemotherapy regimens [5]. Other agents that are presently being evaluated for the treatment of IBC include antiangiogenic agents and Ras pathway inhibitors. IBC is known to be highly vascular that express a number of angiogenic factors such as vascular endothelial growth factor (VEGF). This encouraged a number of studies looking at the role of anti-VEGF agents (e.g., bevacizumab) combined with chemotherapy in the treatment of IBC, with hopeful results [5, 6].

Combinations of neoadjuvant chemotherapy, mastectomy, and radiotherapy have led to an improved prognosis. However, the overall five-year survival rate for patients with IBC is still very low, at ∼20% [7]. Proper management of IBC requires close coordination between the surgical, medical, and radiation oncology teams.
among surgical, radiation oncologists, radiologists, and pathologists. It is important for health institutions to pool resources by establishing a tumor registry for collecting data from patients with IBC worldwide to deal with this fatal disease because of the infrequency of IBC.

How to cite this article


doi:10.5348/ijcri-201502-ED-10002

*********

Author Contributions

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

© 2015 Awad Ali M. Alawad. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES