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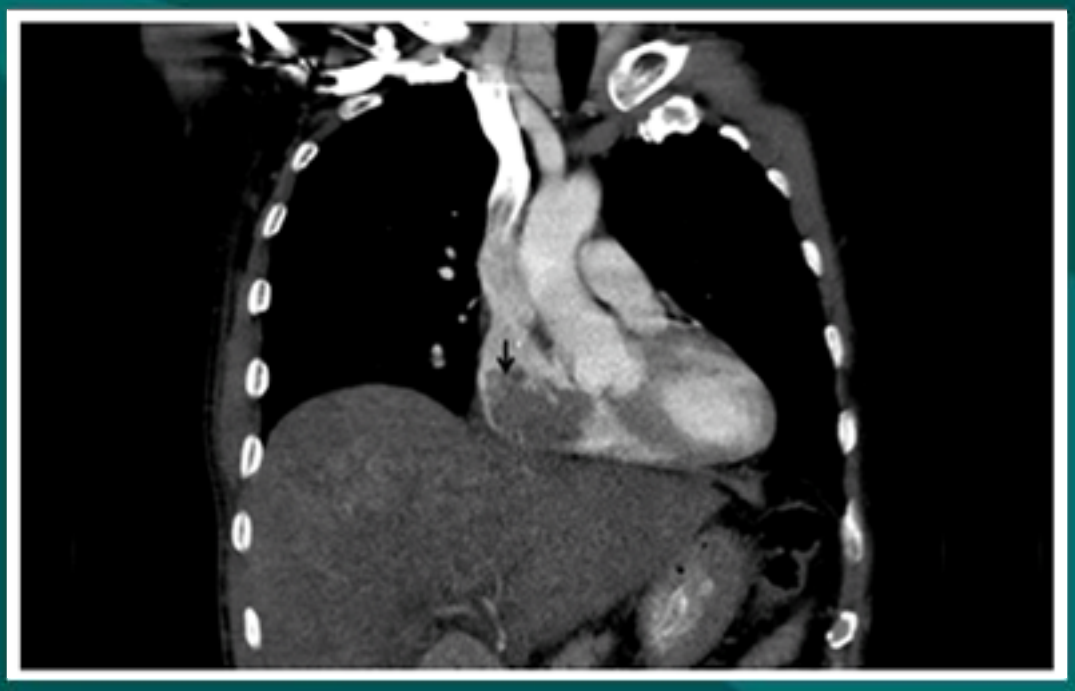

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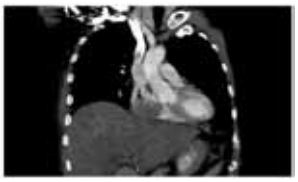

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Contents

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CASE SERIES

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Pilomatricoma—unveiling the ghost story: A case series

Ranjan Agrawal, Parbodh Kumar

ABSTRACT

Introduction: Pilomatrixoma are benign skin neoplasms of hair follicle origin but often are mis-diagnosed. They present as superficial masses of the head and neck especially in children. This present study is aimed at pointing to awareness among the clinicians as well as the pathologists about the varied presentations and morphological features of the entity. Case Series: Five cases were included in the present study, of which four were females and one male. The sites involved were right thigh, right elbow, forehead, right eyebrow and subareolar area in the right breast. Conclusion: Diagnostic pitfalls have been discussed along with the clinical differential diagnosis.

Keywords: Basaloid cells, Calcifying epithelioma of malherbe, Ghost cells, Pilomatricoma

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INTRODUCTION

Pilomatricoma is a relatively uncommon benign skin neoplasm arising from the hair follicles. With more morphologic features and clinical presentations being described there has been a better identification of this neoplasm. Probably due to lack of awareness or otherwise a preoperative diagnosis of pilomatricoma is rarely considered by the clinicians [1].

CASE SERIES

Five cases were included in the present study. Four (80%) of these were females and one (20%) male. The site of involvement in all the patients were different (Table 1). Two cases (40%) occurred in the head and neck region with face as the predominant site involved. The other sites involved were lateral aspect of right thigh, right elbow and beneath the areola of the right breast. All the patients presented with a painless swelling. On examination the swellings were non-tender and freely mobile.

The clinical diagnosis in all the cases was suggestive of epidermoid/dermoid cyst. However, subsequent histopathologic examination confirmed these cases as pilomatricoma. All the lesions were well encapsulated. H&E sections showed dual population of cells comprising the peripheral basaloid cells and the ghost or shadow cells in the centre (Figure 1). A transition of basaloid to ghost cells was noted in many areas. Sections from the right eyebrow swelling posed great difficulty in diagnosis since there were mainly ghost cells present and almost negligible basophilic cells representing stage 4 (Figure 2). The remaining four cases (80%) belonged to the third morphological stage. Two cases showed a rich infiltrate of lymphoplasmacytic cells and numerous foreign body giant cells near the shadow cells (Figure 3). Three cases showed areas of calcification.

Table 1: Showing Age, Sex and Site distribution of Pilomatricoma cases.

Case	Age	Sex	Site involved	Presentation	Duration	Morphological Stage
1	30	F	Lateral aspect of right thigh	Painless swelling	1 year	3
2	11	F	Right elbow	Painless swelling	18 Months	4
3	37	M	Forehead	Painless swelling	9 Months	3
4	33	F	Right eyebrow	Painless swelling	8 Months	3
5	16	F	Subareola right breast	Painless Nodule	13 Months	3

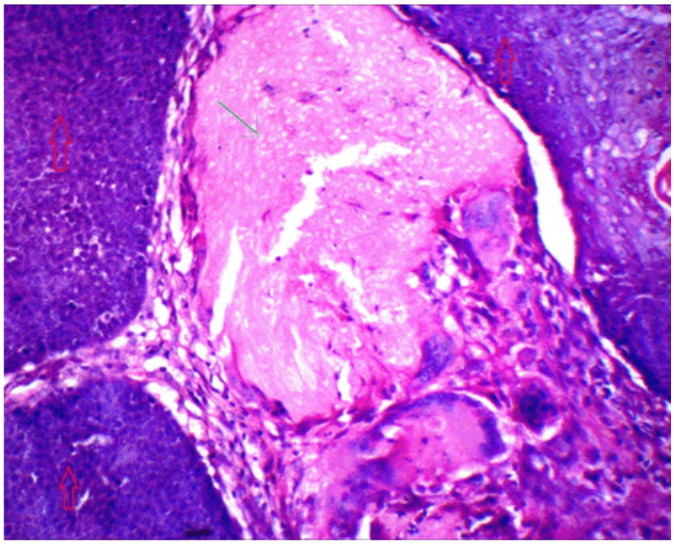

Figure 1: Uniform population of small basophilic cells (⇑) and another area of large shadow/ghost cells filled with keratin and cell debris (↑). (H&E stain, x100).

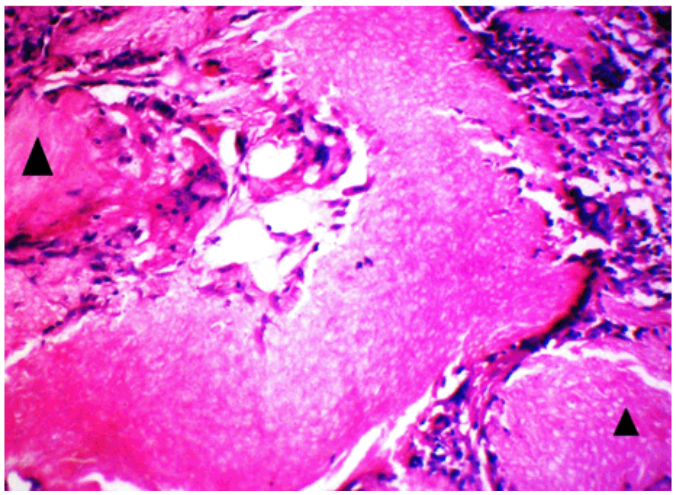

Figure 2: Ghost cells with keratinization (▲). Basophilic cells are almost absent (H&E stain, x400).

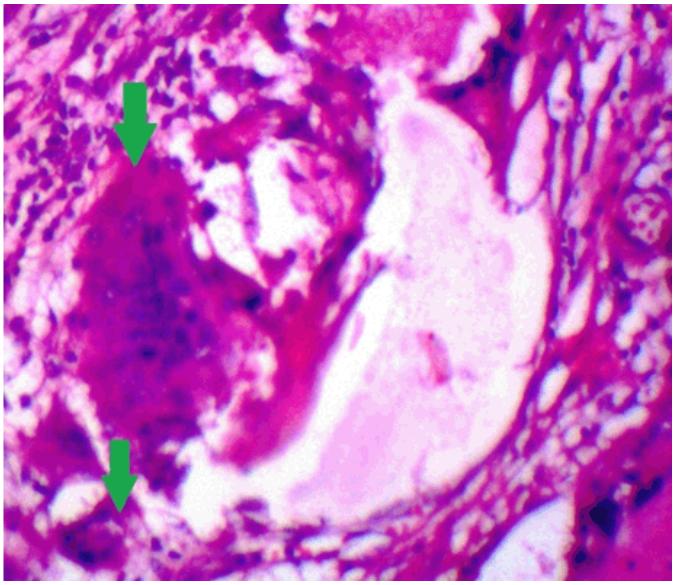

Figure 3: Photomicrograph showing rich lymphoplasmacytic cell infiltration and numerous foreign body giant cells (↑) (H&E stain, x400).

DISCUSSION

Pilomatrixoma, or calcifying epithelioma of Malherbe, was first described in 1880 by Malherbe and Chenantais as a benign subcutaneous tumor arising from the sebaceous glands. In 1922, Dubreuilh and Cazenave described the unique histopathologic characteristics of this neoplasm, including islands of epithelial cells and shadow cells. Turhan and Krainer in 1942 stated the origin of this neoplasm to be from the hair cortex cells. In 1961, Forbis and Helwig proposed the term pilomatrixoma denoting their origin from cells of hair matrix and avoiding overlapping nomenclature with malignancy as suggested by the previously used term “Epithelioma”. In 1977, the name was corrected etymologically to pilomatricoma.

The incidence of pilomatricoma is estimated to be 1 in 2000 surgical specimens [2]. The most frequent anatomical location is the head and neck region, followed by the upper extremities, trunk and the lower extremities in decreasing order of frequency [1]. The sites of occurrence reported in the head and neck region are cheek (36%), neck (20%), periorbital region (14%), scalp (9%) and remaining as multiple lesions [1–8]. Involvement of palms, soles, genital region or lymph nodes has not been reported [1]. Reports of involvement of breasts are also

available [9]. A rare case of pilomatricoma following split thickness skin graft on the lower extremity is available in literature [10]. The female to male ratio reported is 1.5:1 [1, 7]. Female predominance was observed in the present study as well. Pilomatricoma can develop at any age, demonstrating bimodal peak presentation during the first and sixth decades of life. About 40% of cases occur in patients younger than 10 years of age and about 60% of cases within the first two decades of life [1]. In the present study, all five patients were below 40 years of age with 2 below 20 years.

Pilomatricoma mainly presents as a solitary cutaneous nodule with an average size of 1 cm rarely exceeding 2 cm in diameter and is covered by normal or hyperemia skin [11, 12]. All the cases in the present study had similar presentation. Four cases measured less than 1 cm and one was 1.5 cm in diameter. They are usually asymptomatic, deep seated, firm, non-tender and adherent to the skin but not fixed to the underlying tissue. Multiple occurrences and familial cases are also known, in association with disorders like Gardner's syndrome, Steinert disease, Sarcoidosis, Rubinstein-Taybi syndrome, Turner syndrome and Myotonic dystrophy [1, 7, 13]. No such association was observed in any of our cases. Stretching of the skin over the tumor shows the "tent sign" with multiple facets and angles, a pathognomonic sign for pilomatricoma [1, 14]. In addition, pressing on one edge of the lesion causes the opposite edge to protrude from the skin like a "teeter-totter". Another characteristic feature is the blue-red discoloration of the overlying skin differentiating it from epidermal inclusion or dermoid cysts [1].

The accuracy rate of preoperative diagnosis of pilomatricoma reported ranges from 0–30% probably due to the lack of familiarity with this tumor [15]. Radiologic imaging is of little diagnostic value for pilomatricoma. Fine-needle aspiration cytology has also a limited role since the results can be misleading if there are no ghost cells present in the aspirate [1]. The major factors contributing to misdiagnosis include cystic lesions with varying consistency, punctum like appearance (due to skin tethering), atypical location and the absence of clinically recognizable calcification [16]. Another dilemma is the differentiation of this tumor from other benign masses frequently seen in clinical practice such as epidermal inclusion cyst, sebaceous and dermoid cyst, branchial cleft remnants, pre-auricular sinuses, foreign body granulomas, lipoma, calcific lymph nodes, fat necrosis, degenerating fibroxanthoma, osteoma cutis and ossifying hematoma [1, 6–8]. Inclusion cysts have a diffuse yellow color when filled with keratin, are softer, palpable and rare in children. Dermoid cysts are firmly attached to the underlying tissue with covering skin moving freely over the lesion and do not exhibit irregular nodules on the surface [1].

The aetiology of pilomatricoma is unknown. Recently, role of an overactive proto-oncogene called BCL-2 is proposed which suppresses the normal process of cell death and mutations in CTNNB1 suggesting loss of regulation of a protein complex called beta-catenin/ LEF which is an effector in the WNT signaling pathway leading to differentiation and proliferation [6, 8, 17].

Histopathologically, pilomatricoma are well encapsulated and composed of islands of cells separated by fine, fibrovascular connective tissue stroma. The cell islands reveal two distinct cell populations comprising the basaloid cells and the ghost (shadow) cells. Early lesions show a predominance of basophilic cells grouped in islands at the tumor periphery. With tumor maturation, the basophilic cells acquire more cytoplasm and eventually start losing their nuclei to become eosinophilic shadow cells. These latter cells constitute the central portion of the tumor and may calcify gradually imparting the lesion a bony hard consistency. The basaloid cells are darkly stained, round-to-ovoid, have ill-defined cell borders with minimal cytoplasm and vesicular nuclei with most of them exhibiting prominent nucleoli. The ghost cells show well-defined cell borders abundant, pale, eosinophilic cytoplasm with a central clear area. A transition of basaloid cells to ghost cells is noted in many areas. There can be areas of keratinization, calcification, ossification, melanin deposition and focal lymphocytic infiltration along with a foreign body giant cell reaction in close proximity to the ghost cells. Secondary changes such as hemorrhage, ossification, myxoid change, edema and stromal fibrosis may occur [2].

Four distinct morphological stages of pilomatricoma reported include: (a) Early: small and cystic lesions, (b) Fully developed: large and cystic, (c) Early regressive: foci of basaloid cells, shadow cells and lymphocytic infiltrate with multinucleated giant cells, (d) Late regressive: numerous shadow cells with absence of basaloid and inflammatory cells. These stages reflect the evolution of the tumor from a matrix cyst to a calcified and ossified nodule with no visible epithelial component [1, 15]. They do not have treatment or prognostic implication.

Complications of pilomatricoma are rare. However, occasionally they may grow to several centimeters in diameter. Transformation to pilomatrical carcinoma is very rare. Pilomatrix carcinoma needs to be distinguished from proliferating pilomatricoma, which is a benign tumor exhibiting a relative symmetry, sharp circumscription and a distinct lobular proliferation of basaloid cells with small foci of shadow cells along with variable nuclear atypia and mitosis [2, 18].

Since there is no spontaneous regression, malignant transformation is rare and the lesions are well encapsulated, the standard treatment of pilomatricoma is complete surgical excision [1, 6].

CONCLUSION

This study illustrates the diagnostic pitfalls of pilomatricoma with special emphasis on its presentations and differentials. The diagnostic pitfalls have been

highlighted in the present study. We would reiterate the significance of the knowledge of these cases and also to make the clinicians aware of this entity especially as a cause of solitary skin nodule on the head, neck, breast or the upper extremities.

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Author Contributions

Ranjan Agrawal – 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
Parbodh Kumar – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Primary retroperitoneal benign mucinous cystadenoma in a male

Malek Barka, Faouzi Mallat, Mohamed Ben Mabrouk, Chaima Mrad, Khaled Ben Ahmed, Wissem Hmida, Ghassen Tlili, Sidiya Ould Chavey, Sarra Mestiri Sonia Ziadi, Faouzi Mosbah

ABSTRACT

Introduction: Retroperitoneal mucinous cystadenoma is extremely rare, prevailing specifically in female gender, with unclear histogenesis. Only three cases concerning male patients have been reported. Case Report: We report a case of a 75-year-old male presented with right flank pain and a palpable mass. An abdominal computed tomography scan revealed a retroperitoneal tumor resulting after the histological examination to a primary retroperitoneal mucinous cystadenoma. Conclusion: This is the fourth case of primary retroperitoneal mucinous cystadenoma of benign type in a male reported in literature. Several hypotheses may explain the histogenesis of this pathological process. Radical resection is the treatment of choice.

Keywords: Cystadenoma, Male, Mucinous, Retroperitoneal

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INTRODUCTION

Retroperitoneal tumors account for less than 0.2% of all neoplasm [1]. Primary retroperitoneal tumors of mucinous type is an extremely rare entity that affects, almost exclusively women [2] and can be subdivided into benign, borderline or cystadenocarcinoma [3]. Since 1965, only 51 cases have been reported in literature [4]. We report a case of a 78-year-old male presented with primary retroperitoneal mucinous cystadenoma of benign type. In our knowledge, this is the fourth case reported in literature.

CASE REPORT

A 75-year-old male admitted to our hospital complaining of right flank pain and a palpable mass that was constantly growing up during the last three months. There were no associated features, and no aggravating or relieving factors. His past medical history included a lung hydatid cyst operated three years ago. Abdominal examination revealed a firm, sizeable and hard mass at the right abdomen extending from the inferior ribs to the right iliac crest. Biological analyses were normal.

Ultrasonography of the abdomen demonstrated a 20-cm well defined cystic mass. Abdominal computed tomography (CT) scan revealed a large cystic lesion

occupying the right retroperitoneal space and measuring 20×14 cm (Figure 1).

The patient underwent laparotomy and a tumoral exeresis has been performed. The patient made an uncomplicated postoperative recovery. Microscopic examination reported a retroperitoneal mucinous cystadenoma of benign type (Figure 2).

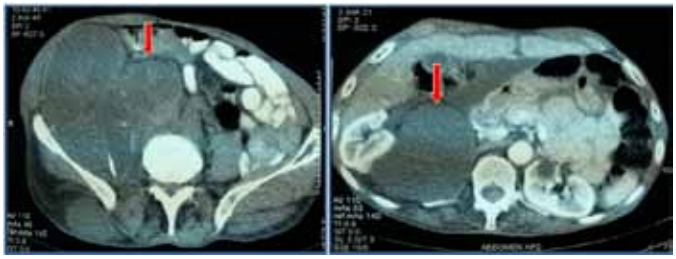

Figure 1: Axial computed tomography images showing the size of the tumor and its mass effect to the right kidney and the small intestine.

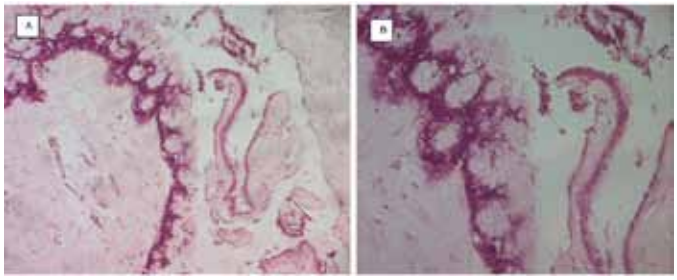

Figure 2: (A, B) Microscopic findings: cystic wall lined by a monostratified mucinous type epithelium without cytologic atypia (H&E stain, x100, x200).

DISCUSSION

This is the 23rd reported case of a benign primary retroperitoneal mucinous cystadenoma in literature (Table 1) and only the fourth primary retroperitoneal mucinous cystadenoma (PRMC) diagnosed in a male [5–26].

This entity was first described by Handfield–Jones in 1924 in his study on retroperitoneal cysts [26]. Only three cases of pure PRMC in a male patient were reported in literature. The retroperitoneal location of these tumors is rare because of the non-existence of epithelial cells in this area so the histogenesis of PRMC remains unclear; one theory involves that that these tumors may arise from a coelomic epithelium which normally during the embryogenesis converts to peritoneal mesothelium and ovarian germinal epithelium but that become trapped in the retroperitoneum [27–28]. Some immunohistochemical and ultrastructural similarity with ovarian mucinous tumors as positive match to cytokeratin 7 and cytokeratin 20 antibodies supports this hypothesis [29]. A second hypothesis involves the seeding of ectopic ovarian tissue in the retroperitoneum [30]. Another histogenesis suggests the proliferation of mucinous epithelium in retroperitoneal teratoma [31]. Clinical signs and symptoms are non-specific including predominantly an abdominal mass, chronic abdominal pain or both. Radiologically, this entity presents a cystic formation repressing the organs around and there are a wide range of differential diagnoses including lymphangioma, cystic teratoma, lymphocele, urinoma and cystic mesothelioma [32]. In our case, because of a history of lung hydatid cyst, a second localization in the retroperitoneum was suspected. Ultrasonography is not specific [17].

Table 1: Cases of primary retroperitoneal mucinous cystadenoma of benign type reported in the literature.

Ref	Case	Age	Sex	Presenting complaint	Investigations
[5]	Navin (2012)	30	F	Pain	US, MRI, Lap
[6]	**Fujita (2012)**	**71**	**M**	**Mass**	**US, CT, MRI, FNA, Lap**
[7]	Tapper (2010)	37	F	Mass	US, MRI, Lap
[8]	Del Gobbo (2010)	52	F	Pain	US, MRI, Lap
[9]	Rifki-Jai (2009)	43	F	Mass	US, CT, MRI, Lap
[10]	Abedalthegafi (2009)	44	F	Mass	US, MRI, Lap
[11]	Yan (2008)	29	F	Mass/Pain	CT, Lap
[12]	**Prabhuaj (2008)-**		**M**	-	-
[13]	Kassab (2007)	80	F	Mass	US, CT, Lap
[14]	Min (2004)	38	F	Pain	CT, Lap
[15]	Isse (2004)	18	F	Flatulence	US, CT, MRI, Lap
		85	F	Pain	-
[16]	Arribas (2004)	39	F	Incidental	-
[17]	**Lai(2004)**	**52**	**M**	**Mass**	**CT, Lap**
[18]	Tamura (2003)	14	F	Incidental	CT, MRI, Lap
[19]	Erdemoglu (2003)	39	F	Pain	US, Lap
[20]	Balat (2001)	44	F	Mass	-

Continued...

Table 1: *(Continued)*

Ref	Case	Age	Sex	Presenting complaint	Investigations
[21]	Subramony (2001)	25	F	Incidental	US, CT, Lap
[22]	Kehegias (1999)	21	F	Mass/Pain	US, CT, MRI, FNA, Lap
[23]	Yunoki (1998)	45	F	Mass	US, CT, Lap
[24]	Park (1991)	40	F	Mass/Pain	US, CT, Lap
[25]	Pennell (1989)	19	F	-	-
	Our case	**75**	**M**	**Mass/Pain**	**US, CT, Lap**

F female, M male, US ultrasonography, CT computed topography, MRI magnetic resonance imaging, Lap laparoscopic investigation, FNA fine-needle aspiration.

The diagnostic value of computed tomography and magnetic resonance imaging scan is similar to differentiate between a cystic teratoma and cystadenoma through detection of calcification within the cyst and the description of the tumor in relation to soft tissue and radiological evidence of its origin [33]. No blood profile abnormalities are specific to PRMC; previous cases have demonstrated an increase in carcinoembryonic antigen (CEA) and CA 19-9 [34–36]. Surgery is the preferred treatment for most cystic retroperitoneal masses [37] by a complete enucleation [38].

CONCLUSION

We present a rare case of primary retroperitoneal mucinous cystadenoma of benign type, the fourth in a male patient. Several hypotheses may explain its histogenesis. Surgical excision is advised for diagnostic assessment and management. Owing to the small number of reported cases and the insufficient surveillance data, prognosis of recurrence is unknown.

****

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Author Contributions
Malek Barka – 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
Faouzi Mallat – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellrctual content, Final approval of the version to be published
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Sonia Ziadi – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Faouzi Mosbah – 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

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CASE REPORT PEER REVIEWED | OPEN ACCESS

First case reporting histoplasmosis presenting as tubal abscess

Parin Pearl Rimtepathip, Alex Erickson, Harold Katner, Younes Bakri

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Anaphylaxis and Kounis syndrome after using amoxicillin and clavulanic acid

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ABSTRACT

Introduction: Kounis syndrome is the fortuitous occurrence of acute coronary syndrome consisting of hypersensitivity reactions related to allergic or hypersensitivity activation of inflammatory cells. Case Report: An 84-year-old male admitted to our emergency department with anaphylaxis symptoms after his intake of a 1000 mg amoxicillin and clavulanic acid (ACA) tablet. Diabetes, coronary artery disease and hypertension had been encountered in the patient's medical history. During the physical examination, uvula edema, wheezing, rales and rhonchi were present. Adrenaline, pheniramine and methylprednisolone were given to the patient with a diagnosis of anaphylaxis. During his observation, ECG changes and troponin elevation were detected and he was diagnosed as type 2 Kounis syndrome. After 24 hours follow-up, he was discharged without any complication. Conclusion: A single dose of ACA can cause anaphylaxis and may result in Kounis syndrome. It should be taken into consideration in all types of allergic reactions which treated by adrenaline.

Keywords: Anaphylaxis, Clavulanic acid, Coronary syndrome, Drug allergy, Kounis syndrome

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INTRODUCTION

Allergic reactions due to use of drugs may be seen as situations ranging from simple urticaria to severe angioedema. The admission rate to the emergency service because of anaphlaxis comprise 0.4–1% in various studies. Medications comprise 7.7–34.6% of anaphylaxis. Amoxicillin has the highest proportion (40%) in those drugs that cause severe anaphylaxis [1, 2].

Kounis syndrome is the coincidental occurrence of acute coronary syndromes with hypersensitivity reactions involving activation of interrelated and interacting inflammatory cells and including allergic or hypersensitivity and anaphylactic oranaphylactoid insults [3]. To date, three types Kounis syndrome have been described. Type 1 includes allergic mediators-induced coronary vasospasm in patients without coronary artery disease oratherosclerotic risk factors. Type 2 includes vasospasm, plaque erosion or rupture inpatients with coronary artery disease. After the investigation of thrombi on drug-elutingstents demonstrate the presence of eosinophils and mast cells, this situation is referred to type 3 [4].

Herein, diagnosis and treatment of a patient related to allergic reaction and type 2 Kounis syndrome after single dose of amoxicillin and clavulanic acid (ACA) are discussed.

CASE REPORT

An 84-year-old male admitted to emergency department after intake of a 1000 mg ACA tablet,

which had been recommended for upper respiratory infection, with shortness of breath and numbness in the throat complaints ongoing for 30 minutes. Past medical history revealed diabetes, coronary artery disease and hypertension. On admission, TA 110/50 mmHg, pulse rate : 105/min, fever 38.9°C and blood glucose 224 mg/dL. On physical examination, uvula edema, wheezing, rales and rhonchi were present and other systematical examinations were normal. The 12-lead electrocardiogram (ECG) revealed sinusal tachycardia and negative T wave on lead aVL (Figure 1).

We administered 0.5 mg adrenaline intramuscularly, 45.5 mg pheniramine, 80 mg methylprednisolone intravenously to the patient, concerning the diagnosis of anaphylaxis. Paracetamol 1 g used for fever management.

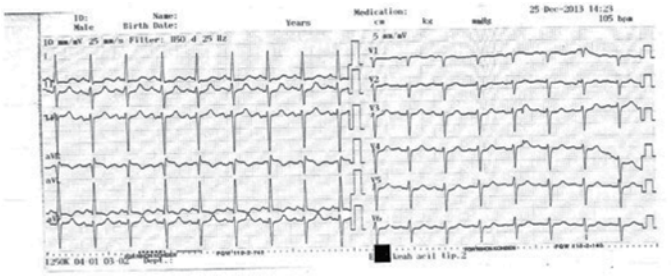

Figure 1: Patients first electrocardiogram. Sinusal tachycardia and negative T wave on lead aVL.

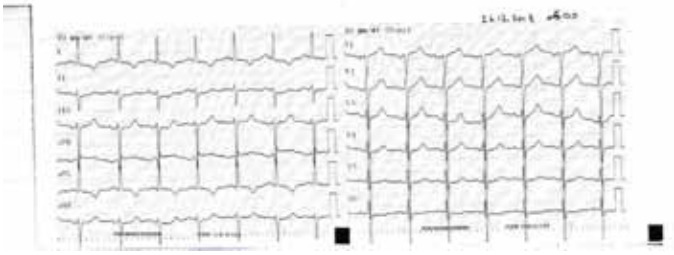

Figure 2: Negative T wave on leads DI, aVL and V6 on 4th hour control electrocardiogram.

For bronchoconstriction, 2.5 mg salbutamol was given to him via nebulizer. Laboratory findings are given in Table 1.

During follow-up, increase in troponin levels and changes in ECG (Figure 2) were observed and the patient was consulted to cardiologist with a pre-diagnosis of Kounis syndrome. Echocardiographic examination revealed normal left ventricular systolic function, diastolic dysfunction grade 1 and EF %60. Troponin levels were lower at follow-up, was not planned coronary angiographic intervention. Uvula edema regressed, no fever or ECG changes detected and Troponin levels decreased and patient was discharged after a 24-hour follow-up without any complication.

DISCUSSION

In developed countries, cardiovascular diseases are the central cause of death. And this stuation is not expected to change significantly by 2020 [5]. Acute coronary syndromes (ACS) are the acute manifestation of cardiovascular diseases that can lead to death. Kounis syndrome is one of the rare causes of ACS. Kounis syndrome is also called and known Allergic angina or Allergic myocardial infarction in different sources. The main pathophysiology of Kounis syndrome is the activation of mast cells by allergic stimulation and release of biological amines, neutral proteases, arachidonic acid derivatives and platelet activating factor. Histamine increase thrombocyte activation, coronary vasoconstriction and tissue factor synthesis. Neutral proteases cause plaque erosion and rupture as a result of matrix metalloproteinase activation. And also they increase vasoconstriction by raising angiotensin-2 levels [3].

Table 1: Laboratory findings

Hours	0.	60.	12.	15.	Normal ranges
CK-MB	4.5 ng/mL			6.2 ng/mL	0.6 - 6.3 ng/mL
Troponin- I	0.43 ng/mL	1.36 ng/mL	1.37 ng/mL	1.24 ng/mL	0 – 0.04 ng/mL
Glucose	235 mg/dl				74 – 106 mg/dl
Ph	7.38				7.35 – 7.45
PCO2	40.3 mmHg				35 – 48 mmHg
PO2	43.8 mmHg				83 – 108 mmHg
HCO3	23.4 mmol/L				22.5 – 26.9 mmol/L
Base Excess	-1.5 mmol/L				-2.7 - +2.5 mmol/L

Abbreviations: CK-MB: KreatinKinaz-MB, PCO_2: Partial pressure of carbon dioxide, PO_2: Partial pressure of oxygen, HCO_3: Bicarbonate

A wide variety of allergens was reported in the course of Kounis syndrome. Drugs that have been reported to induce Kounis syndrome were antibiotics, contrast media, antineoplastics, intravenous anesthetics, NSAIDs, thrombolytics and anticoagulants, skin disinfectants and some other drugs including allopurinol, enalapril, esmolol, insulin. Antibiotics have been reported to date are ampicillin, amoxicillin, amikacin, cefazolin, cefoxitin, cefuroxime, cephradine, cinoxacin, lincomycin, penicillin, sulbactam/cefoperazone, sulperazone, trimetophrim/ sulfamethoxazole and vancomycin. Almost all of the cases related with ACA have been reported as type 1 Kounis syndrome (rarely type 2 or type 3) [3].

The contribution of assays for serum histamine, specific IgE antibodies, tryptase, complement proteins (C4- and C1- esterase inhibitor) and evaluation of eosinophilia to diagnosis is not certain. Negativity does not exclude diagnosis because of the short half-life of these mediators, [6]. There are no certain information and guide regarding the treatment of Kounis syndrome coming from case reports and case series [7]. Kounis [6], Cevik [7], Biteker [4] and Ridella [8] make offers treatment for Kounis syndrome. The review by Ridella et al. demonstrated that steroid, H1 blocker, nitroglycerin, adrenaline and acetyl salicylic acid were administered [8]. We got a very positive response after use of adrenaline, H1 blocker, steroid and hydration. Troponin levels decreased and symptoms of anaphylaxis improved after this treatment. So, this treatment method can be used at similar conditions. Cevik et al. emphasized whether or not all medications may be useful. Nevertheless pharmacological management should be considered individually [6].

CONCLUSION

Even if typical chest pain or coronary artery disease history does not exist, yet anaphylaxis and Kounis syndrome should be taken into account.

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Author Contributions

Gokhan Eyupoglu – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Mehmet Tatli – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Kerem Dost Bilmez – Conception and design, Drafting the article, Critical revision of the article, Final approval of the version to be published
Ozlem Guneysel – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

An unusual presentation of encapsulating sclerosing peritonitis

Emmi Khoo, Dharmenaan Palamuthusingam, Clyson Mutatiri

ABSTRACT

Introduction: Sclerosing encapsulating peritonitis (SEP) is a rare condition. Underlying cause of SEP is believed to be multifactorial. It can occur secondary to several conditions like prior episodes of severe peritonitis, use of beta-blockers, history of peritoneal dialysis, autoimmune disease, intra-abdominal malignancies, exposure to chemicals such as silicosis or asbestosis, endometriotic cyst, uterine leiomyomas, ovarian tumors, sarcoidosis, or tuberculosis. Case Report: We report an unusual case of a 74-year-old male patient of Dutch descent presented with intermittent watery diarrhea, recurrent ascites and bilateral lower limb lymphedema. Conclusion: Symptoms of the patient at initial presentation were non-specific. Bilateral lower limb lymphedema has not been previously identified in literature as a presenting symptom of this disease. Clinicians need to have a higher index of suspicion for the diagnosis of SEP in those that present with bilateral lower limb lymphedema and non-specific abdominal symptoms. The etiology of SEP in this patient remains unclear, which is consistent with most of the cases reported in literature to date.

Keywords: Ascites, Bowel obstruction, Lymphedema, Sclerosing encapsulating peritonitis

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INTRODUCTION

Sclerosing encapsulating peritonitis (SEP) is a rare condition with varying prevalence around the world. An Australian study identified 54 cases in 14 years, indicating prevalence of 0.7% [1]. This compares with a prevalence of 3.3% described in a UK series [2].

Underlying cause of SEP is believed to be multifactorial [3]. It can occur secondary to several conditions like prior episodes of severe peritonitis, use of beta-blockers, history of peritoneal dialysis, autoimmune disease, intra-abdominal malignancies, exposure to chemicals such as silicosis or asbestosis, endometriotic cyst, uterine leiomyomas, ovarian tumors, sarcoidosis, tuberculosis or it can be idiopathic [4]. There is no significant association between SEP, age and gender [5]. However, notably, idiopathic types are found more commonly among young adolescent female in tropical and sub-tropical areas [4].

CASE REPORT

A 74-year-old male lived his life in Australia and very rarely traveled overseas. He has never smoked tobacco or consumed alcohol. He has a known history of transient ischemic attack four years ago, left knee osteoarthritis,

depression, and hypercholesterolemia. His medications include escitalopram 20 mg mane, lorazepam 1 mg nocte and rosuvastatin 5 mg mane. Examination revealed normal cardiovascular and respiratory system and his abdomen was soft, with no masses but was positive for shifting dullness. He also had bilateral pitting edema up to his mid–thighs.

Initial biochemistry, full blood count and liver function tests were unremarkable (Table 1). An echocardiogram showed normal right and left ventricular function and normal valves. An ultrasound (USS) of the abdomen confirmed more than 1 L of ascites but normal liver and spleen dimensions with no ultrasonographic evidence of portal hypertension. The USS of his lower limbs did not demonstrate deep venous thrombosis. Computed tomography (CT) scan of his abdomen showed mild

Table 1: Initial investigations performed

Investigations	Value	Units	Ref. Range
Hemoglobin	131	g/L	120–180
White cell count	4.7	x 10^9/L	3.5–11.0
Platelet	196	x 10^9/L	140–400
Creatinine	90	mmol/L	64–108
GFR (estimated)	72	mL/min/1.73m2	>60
Protein	63	g/L	60–83
Albumin	42	g/L	35–50
Alkaline Phosphatase	217	U/L	56–119
Gamma-GT	29	U/L	<55
Alanine Transaminase	10	U/L	<45
Aspartate Transaminase	14	U/L	<35
Lactate Dehydrogenase	170	U/L	150–280
Calcium (Alb. Corr.)	2.30	mmol/L	2.15–2.60
Phosphate	1.08	mmol/L	0.81–1.45
Magnesium	1.04	mmol/L	0.70–1.10
INR	1.0		0.9–1.2
CRP	2.3	mg/L	<5.0
CA 19.9	<5.0	kU/L	<35
Carcinoembryonic Ag	<1.0	ug/L	<5.0
ePSA	0.14	ug/L	<6.5
Ferritin	172	ug/L	30–300
Transferrin	1.8	g/L	1.6–3.4
Vitamin B12	172	pmol/L	133–680
Serum Folate	29.4	nmol/L	>7.0
25-Hydroxy-Vitamin D	66	nmol/L	50–150
AST (Red cell)	6.2	U/g Hb	3.0–7.0
Ceruloplasmin	1.78	mmol/L	1.33–2.22
Serum Zinc	12	mmol/L	8–18
C3	1.51	g/L	0.90–1.80
C4	0.52	g/L	0.10–0.40
Rheum. Factor (Neph)	<20	IU/ml	<30
Anti-CCP	0	U/ml	<6
Antinuclear Antibody	Negative	Titre	
Extractable Nuclear Ag	Negative		
Anti dsDNA	4	IU/ml	<7
Monoclonal Protein	Not Detected		
IgG	4.8	g/L	7.0–16.0
IgA	0.3	g/L	1.0–4.0
IgM	0.2	g/L	0.4–2.3
Kappa FLC (N Latex)	10	mg/L	7–22
Lambda FLC (N Latex)	16	mg/L	8–27
K/L Ratio (N Latex)	0.62		0.31–1.56
Urine Bence Jones Protein	Not Detected		
Urine Creatinine: Protein	<18	g/mol creat	<15
Urine Cytology	No malignant cells found		
Stool Alpha-1-Antitrypsin	0.5	mg/g dry wt	<1.5
Stool Occult Blood	Negative		

heterogeneity of the greater omentum in the upper abdomen, some circumferential thickening of the sigmoid, and few sclerotic foci in axial skeleton.

A abdominal paracentesis performed was cloudy in appearance but cytology and biochemistry were unremarkable. The surgeons performed a sigmoidoscopy and subsequent biopsies revealed normal large intestinal architecture.

A diagnosis remained elusive and the patient was treated symptomatically with bumetanide 1 mg mane but there was no clinical improvement.

Ultimately, a radionuclide lymphoscintigraphy confirmed bilateral distal 'dermal backflow', consistent with bilateral lymphedema. A provisional diagnosis of lymphangitis was made based on these findings he was commenced on prednisolone 50 mg daily.

After four weeks on steroid treatment, his edema had reduced significantly and his bowel symptoms improved.

Despite improved symptomatology, he was revisited by the surgeon in clinic, who continued to be suspicious for intra-abdominal malignancy and insisted on diagnostic laparoscopy being performed. At the time of the operation, the small bowel looked normal with white plaque-like patches in some areas. The surgeon converted to an exploratory laparotomy via a midline incision and noted the cecum feeling and appearing fibrotic and having some whitish areas resembling scarring in pelvic peritoneum. The sigmoid colon was adherent to pelvic sidewall, and the omentum was shrunken with obliteration of lesser sac. Several peritoneal biopsies were taken.

The histology of peritoneal biopsy revealed tissues containing bland spindle cells separated by collagen, some of which is thick and eosinophilic; with rare mitotic figure noted but no atypical figures were identified. Omental tissue biopsies showed thin submesothelial zone of collagenous connective tissue containing cytologically bland spindle cells, similar to those seen in the peritoneal biopsy. These histological features were consistent with sclerosing peritonitis. However, at this point in time the diagnosis SEP was overlooked for multiple reasons. Firstly, the experienced surgeon did not feel that at the time of the operation, the macroscopic appearance of the abdomen was not consistent with the typically described 'cocoon-like' appearance. Secondly, the patients' presentation was not a characteristic manifestation of SEP that had been previously documented in literature.

Unfortunately, the patient developed acute bowel obstruction and died from sepsis eight months later.

DISCUSSION

Underlying cause of SEP is believed to be multifactorial [3]. It can occur secondary to several conditions like prior episodes of severe peritonitis, use of beta-blockers, history of peritoneal dialysis, autoimmune disease, intra-abdominal malignancies, exposure to chemicals such as silicosis or asbestosis, endometriotic cyst, uterine leiomyomas, ovarian tumors, sarcoidosis, or tuberculosis [4]. Or it can be idiopathic. There is no significant association between SEP, age and gender [5]. However, notably, idiopathic types are found more commonly among young adolescent female in tropical and subtropical areas [4].

Common clinical features at presentation of SEP are abdominal pain, nausea, vomiting, loss of appetite, constipation and weight loss [5]. Other than symptoms of bowel obstruction, SEP may present with features of fluid overload like ascites and edema. In those undergoing PD this is attributable to the progressive loss of ultrafiltration. This is a result of peritoneal scarring and adhesions by fibrocollagenous membrane which leads to the loss of surface area for ultrafiltration [6]. Repeated episodes of peritonitis are believed to accelerate the process [7].

Preoperative diagnosis is non-specific. Imaging techniques are helpful but not diagnostic. Plain abdominal X-ray may show signs of bowel obstruction [8, 9]. Small bowel intestine loops congregated in a single area in the peritoneal cavity, encased by a soft tissue-density mantle may be seen on abdominal CT scans [10]. Other features like signs of obstruction, agglutination and fixation of intestinal loops, mural thickening, peritoneal thickening or calcification and reactive adenopathy may also be present [10]. Ultrasonography may illustrate ineffective peristaltic contractions, dilated fixed intestinal segments matted together and tethered posteriorly, intraperitoneal echogenic strands, trilaminar appearance of the bowel wall, and loculated free fluid [9, 11, 12]. The "cauliflower sign" is the conglomeration of bowel loops that appear to adhere to each other surrounded by a sac-like structure. This along with delayed transit of contrast medium are characteristics of SEP and can be found on contrast study including barium X-ray and contrast CT scab [8].

Ultimately, the diagnosis of abdominal cocoon is based on intraoperative and histopathology findings. During surgery, encapsulation of entire or partial intestine in thick white fibrous "cocoon-like" membrane is seen; and histology of membrane reveals proliferation of fibrocytes and enrichment of collagen fiber, with non-specific inflammatory reaction and vascular proliferation [8].

Surgical adhesiolysis is the gold standard in treatment of patients with recurrent bowel obstruction secondary to SEP [8, 11]. Other alternate treatment options for milder cases include TPN and nasogastric decompression [11].

Many reports in literature describe the beneficial effects of immunosuppressive agents on the progression of SEP [12–14]. Patients with SEP have been successfully treated with corticosteroids alone or in combination with azathioprine [11, 14, 15]. Examples of other immunosuppressants like colchicine, tamoxifen, renin- angiotensin inhibitors, phosphatidylcholine and antifibrotic agents have also been trialed in management of mild disease and outcomes were inconclusive [11, 12]. A study in Japan showed that two-year survival among patients treated with steroids was 73%; while among patients not receiving such treatment, the two-

year survival was only 48% [11]. It was proposed that immunosuppressive therapy could slow the progression of SEP by reducing the production of inflammatory mediators, which promote fibrinogenesis; hence, formation of fibrous capsule [12]. Due to the sporadic nature and low incidence of the disease, there is a lack of literature to conclude on the significance of long-term immunosuppressive therapy and treatment durations have varied between 3–4 months to life-long treatment [11, 14, 15].

This patient's symptoms at initial presentation were non-specific. His symptoms centered mainly on bilateral lower limb non-pitting lymphedema with ascites and some weight loss, with no indication of bowel obstruction. In a case series by Ping et al., all five of their patients experienced symptoms between 2 weeks to 10 years, before a diagnosis was confirmed. Those symptoms included recurrent episodes of intestinal obstruction, colicky abdominal pain, non-bilious vomiting, abdominal distension and constipation [16]. In our case, underlying malignancy had been considered high on the list of differentials. In retrospect, there were some initial features which were suggestive of SEP but this diagnosis had not been entertained preoperatively. His first episode of acute bowel obstruction occurred 10 months after this.

The aetiology of SEP in this gentleman remains unclear, which is consistent with most of the cases reported in literature to date.

CONCLUSION

This case report highlights that clinicians need to have a higher index of suspicion for the diagnosis of sclerosing encapsulating peritonitis (SEP) in those that present with bilateral lower limb lymphedema and non-specific abdominal symptoms.

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Author Contributions

Emmi Khoo – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Dharmenaan Palamuthusingam – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Clyson Mutatiri – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

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Conflict of Interest

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Iatrogenic saline toxicity complicated by malnutrition

Muhammad Uneib, Parin Rimtepathip, Harold P. Katner

ABSTRACT

Introduciton: Hypotension is a common presenting symptom in elderly patients with malnutrition and dehydration, often being treated with a continuous volume of 0.9% normal saline. Since normal saline is slightly hypertonic, prolong continuous treatment with normal saline can result in hyperchloremic non-anion gap metabolic acidosis. Patients with a low albumin state are also at risk for third spacing that can lead to generalized edema. Case Report: We present a 63-year-old Caucasian female with history of schizophrenia, traumatic brain encephalopathy, and chronic kidney disease stage III who presented to the emergency room from a nursing home due to altered mental status and low blood pressure. The patient received continuous infusion of normal saline despite failure of improvement in blood pressures due to the wrong diagnosis of the cause of hypotension. The patient ended up with iatrogenic normal saline toxicity. Conclusion: It is important for healthcare professionals to recognize signs and symptoms of normal saline toxicities especially in elderly patients with many chronic illnesses. Hypotension treated with continuous normal saline will eventually led to hyperchloremic metabolic acidosis with edema from third spacing in patients with low albumin secondary to malnutrition.

Keywords: Hyperchloremic metabolic acidosis, Malnutrition, Normal saline, Toxicity

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INTRODUCTION

Infusing large volumes of 0.9% sodium chloride (saline) to patients can cause toxicities of both hyperchloremic metabolic acidosis and edema of the surrounding tissues [1]. The most commonly used resuscitation fluid in the United States especially for patients presenting with signs and symptoms of dehydration is 0.9% normal saline. Malnutrition and dehydration present very similar among the elderly patients, especially those with altered mental status and underlying causes such as psychiatric disturbances, stroke, or chronically ill. Both the chronic hypoalbuminemia that accompanies critical illness and the acute dilutional hypoalbuminemia that accompanies rapid crystalloid infusion effectively reduce the upper limit of the normal range of the anion gap because approximately 75% of the normal anion gap is composed of the negatively charged albumin molecule [2]. Malnutrition patients who are often misdiagnosed as dehydration often get treated for dehydration with large volumes of 0.9% saline but with lack of clinical improvement. Health care professionals need to properly

screen and assess for malnutrition especially in vulnerable population. Without the right treatment for the cause, high volume of normal saline can be hazardous to those patients because it can cause metabolic derangement. We describe a case of 63 years old Caucasian female with a history of schizophrenia, traumatic brain encephalopathy, and chronic kidney disease stage III, who presented to the emergency department from a nursing home due to altered mental status with hypotension.

CASE REPORT

A 63-year-old Caucasian female with history of schizophrenia, traumatic brain encephalopathy, and chronic kidney disease stage III, presented to the emergency department due to altered mental status and a decline in blood pressures. Upon admission, the blood pressure was initially 107/56 mmHg but dropped to 73/56 mmHg, her heart rate was 75 bpm, respiratory rate 16 bpm, and temperature 36.9 °C. The patient was given norepinephrine 8μg/min IV infusion and IV fluids in the emergency department for stabilization. Her laboratory values were obtained which showed sodium 136, potassium 3.2, chloride 100, and bicarbonate 31. coagulations, liver enzymes, urinalysis, and chest X-ray were within normal limits. On day-2 of admission, the patient continuously received 0.9% saline IV fluids for hypotension with the blood pressure of 84/45, heart rate of 76, respiratory rate of 13, and oxygen saturation of 100 on nasal canula. Laboratory examination on day-2 showed Na^+ 142, K^+ 3.7, Cl^- 112, $HCO3^-$ 26, and albumin 2.6. Blood cultures were positive for *Staphylococcus capitis* in one of the four cultures. The patient was treated with oxacillin. On day-3 of admission, despite the continuous 0.9% saline IV and treatment of bacteremia, patient was still hypotensive with blood pressure of 85/43. Serum chemistries on day-3 showed Na^+ 135, K^+ 3.7, Cl^- 116, $HCO3^-$ 18, and albumin of <1.5. Physical examination showed new findings of pitting edema on her extremities. Computed tomography scan of the abdomen and pelvis was obtained due to possible internal bleed as the cause of hypotension. The patient was scanned through the abdomen pelvis with no intravenous of oral contrast. The study did not reveal any retroperitoneal or intraperitoneal hemorrhage. However, there were small bilateral pleural effusions, small ascites, and small free fluid in the pelvis. There was mild diffuse mesenteric edema, mild edema at the pelvic floor and in the perirectal soft tissues. On day-4, infection disease department was consulted due to failure of improvement of the blood pressure despite antibiotic treatment with additional onset of pitting edema and new findings on the CT scan. Per consultation; *Staphylococcus capitis* bacteremia was not the cause of hypotension due to only one out of the four cultures was positive with normal white blood cell count and lactic acid level. Laboratory values also showed normal anion gap with increased in chloride level (Table 1). *Staphylococcus capitis* was probably positive on blood culture due to contaminant. The consultation described the low albumin level as possible malnutrition due to normal liver enzyme with no protein in the urine as patient was not swallowing her food per discussion with the nurses plus the history of being an elderly with AMS and schizophrenia. The patient was then replaced with 25 g albumin infusion (25% solution) with monitored blood pressure. On day-6 of admission, the hypotension resolved with albumin with blood pressure of 116/75 mmHg. Patient was considered for peg tube placement for malnutrition with stoppage of the 0.9% normal saline due to normal saline toxicity.

Table 1: Laboratory examination results

Lab Values	Na+	K+	Cl-	HCO3-	AGAP	Albumin
Day-1	136	3.2	100	31	8.2	n/a
Day-2	142	3.7	112	26	7.7	2.6
Day-3	143	3.3	116	18	10.3	<1.5
Day-6	139	3.2	105	26	11.2	increase

DISCUSSION

Malnutrition is defined as imbalance of nutrients mainly due to poor oral intake often seen in the elderly patients, especially those with underlying causes such as psychiatric disturbances, stroke, and chronically ill. Physical factors that affect malnutrition include oral health (i.e., decrease oral intake), physical impairment, early satiety, and taste and smell changes [3]. As the research statistics indicate, not only is malnutrition prevalent in the elderly, but also frequently misdiagnosed or unrecognized. Many health care professionals are not properly screening or assessing malnutrition in the elderly [4]. This case described an elderly patient with history of schizophrenia, although initial presentation was consistent with dehydration from abnormal laboratory values, the lack of improvement after normal saline infusion for a prolonged time course, suggested needs for additional diagnosis. After the exclusion of bacteremia as the cause of hypotension, abnormally low albumin level should have been considered as the culprit for hypotension, especially with no proteinuria and normal liver enzymes. Correct treatment is absolutely dependent on differentiation of hyperchloremic (non-anion gap) acidosis from lactic acidosis [5]. Aggressive attempts to improve organ perfusion, based on misdiagnosis of lactic acidosis, could prove harmful just like our patient with normal lactic acidosis and non-anion gap metabolic acidosis. Normal saline is considered as hypertonic solution because it has an osmolality of 308 mOsm/L, with electrolyte composition consisting of 154 mmol/L of

Na^+ and 154 mmol/L of Cl^- [6]. While normal saline is slightly hypertonic and does not create a huge imbalance in body fluids or electrolytes, there are few major precautions. Two major complications of normal saline toxicities are hyperchloremic non-anion gap metabolic acidosis and the edematous state from third spacing especially in patients with low albumin level. The index patient showed signs of normal saline toxicity with both two major complications proven with CT scan of abdomen pelvis and abnormal laboratory values. It is important to point out that aggressive fluid therapy should be carefully monitored as our patients were almost "flood" with excessive fluid. Prolonged course of uncorrected hypotension with normal saline solutions accompanied by laboratory values of hyperchloremia, non-anion gap metabolic acidosis, no proteinuria, normal liver function, and low albumin should prompt a differential diagnosis of malnutrition. Albumin played an important role by maintaining intravascular oncotic pressure and homeostasis. Decrease in albumin level will eventually led to decrease in oncotic pressure with increased in capillary pressure. A prolonged decreased in oncotic pressure and increased in capillary pressure will eventually resulted in dehydration due to the escape of fluid from intravascular to interstitial space. These patients need to be taken off the fluid immediately with prompt administration of albumin as a correct treatment for their dehydration.

CONCLUSION

In conclusion, elderly patients are at the pinnacle upon risk for malnutrition. Malnutrition is often unrecognized and under-treated by healthcare professionals. When these patients presented with clinical symptoms of severe dehydration with no improvement upon normal saline infusion, electrolytes, anion gap, and albumin level should be acquired. These laboratory values especially the anion gap and chloride level should be taken advantaged of due to their availability to assist in diagnosing underlying reasons for hypotension in malnutrition versus bacteremia in elderly patients especially those with history of physiological and mental incapability.

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Author Contributions

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

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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

What lies beneath? A case of a rare complication of orthopedic casting

Christina A. Fleming, Shane C. O'Neill, Prasad Ellanti, Paul Moroney

ABSTRACT

Introduction: Orthopedic casting is a fundamental technique in orthopedic practice. While orthopedic casting is effective it is not without complications. Case Report: We discuss a case of a 65-year-old male who following a scaphoid fracture, developed a severe acute allergic contact dermatitis to fiberglass orthopedic cast. He had no previous history of atopy and had orthopedic casting previously, 20 years ago. He was treated symptomatically initially without cast removal but deteriorated. His condition was successfully managed conservatively with a combination of cast removal and use of a canvas splint, intravenous antibiotics, oral antihistamine and topical steroids. He achieved full fracture healing clinically and radiologically. Conclusion: A high index of suspicion should be kept when patients with an orthopedic cast present with any symptoms to the emergency department. Any symptoms beneath a cast should prompt cast removal and full examination.

Keywords: Cast dermatitis, Contact allergic dermatitis, Orthopedic casting, Scaphoid fracture

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INTRODUCTION

Orthopedic casting is a fundamental technique in orthopedic practice. It is used for the immobilization of fractures both as definitive or postoperative management. While orthopedic casting principles date back several centuries, the use of modern casting techniques was first described in 1851 by Mathijsen culminating in the introduction of synthetic casting tapes (45% polyurethane resin and 55% fiberglass) in the 1970s [1]. While orthopedic casting is effective it is not without complications including stiffness, pressure sores and compartment syndrome [2]. All of these complications may be avoided with correct casting technique and regular cast review. We describe a case of a rare orthopedic cast complication in which a previously well patient with no atopic background developed a severe acute allergic contact dermatitis to fiberglass cast application.

CASE REPORT

A 65-year-old male presented to the emergency department with a painful right wrist, subsequent to a fall ten days earlier on to his outstretched hand. On clinical assessment he had a painful wrist and tenderness over his anatomical snuffbox that was suspicious for a scaphoid fracture. An undisplaced scaphoid waist fracture was confirmed on plain film radiographs (Figure 1). He was placed in a Plaster of Paris (POP) back slab and an orthopedic follow-up was arranged. At the orthopedic

review, it was decided that non-operative treatment in a below elbow "Colles" cast was the treatment of choice. A standard 3M (TM) fiberglass cast was applied with a further clinical and radiographic review scheduled. Over the following two days the patient began to develop pain, pruritus and felt the cast getting tighter. He presented to the emergency department where a further radiograph demonstrated no fracture displacement and it was felt that the symptoms would resolve with elevation as the arm was not too swollen. The following day he noted progressive edema of his hand and fingers and pustular lesions on his right arm, spreading to his left arm (Figure 2). There were similar lesions on his right trunk and abdomen where he had rested the cast in direct contact with skin (Figure 3). The pain and irritation became so unbearable that the patient removed the cast with a saw himself before re-attending the emergency department. On arrival he was apyrexial, normotensive and without any symptoms or signs of systemic infection or overt anaphylaxis. There was no past medical history of atopy or dermatological conditions. His right forearm was edematous with erythema and numerous bullous eruptions. Many lesions were impetiginized and cellulitic. He had a similar rash on his trunk and left forearm. Routine bloods showed normal white cell count of $8x10^9$/L, normal eosinophils $0.15x10^9$/L and a normal C-reactive protein of 4 mg/L.

The patient was admitted to hospital with a working diagnosis of acute allergic contact dermatitis (ACD) secondary to fiberglass cast application. Swabs were taken from pustular lesions and sent for microscopy, culture and sensitivity analysis to out rule infection. He was commenced on empirical flucloxacillin (500 mg six hourly) by mouth and subsequent microbiological results returned negative for the presence of any micro-organisms. He was urgently reviewed by the dermatology team who concurred with a diagnosis of acute ACD and further commenced on oral prednisolone 10 mg once daily by mouth, cetrizinechloram 4 mg three times daily by mouth and topical agents including Fucibet© (betamethasone and fusidic acid) and jelonet dressings. The patient was nursed with his arm elevated in a sling support and showed signs of improvement within twenty four hours. On day-3 he was switched from oral steroids to topical steroid cream (dermovate). The antibiotics and antihistamines were continued for a total of ten days.

He was discharged home on day-5 with the wrist in a canvas splint with thumb extension for management of the scaphoid fracture. By six weeks the skin had greatly improved and the scaphoid fracture had healed by eight weeks (Figure 4). The patients skin had normalized by three months and he has been discharged from follow-up. He remains pain free and without any functional limitation.

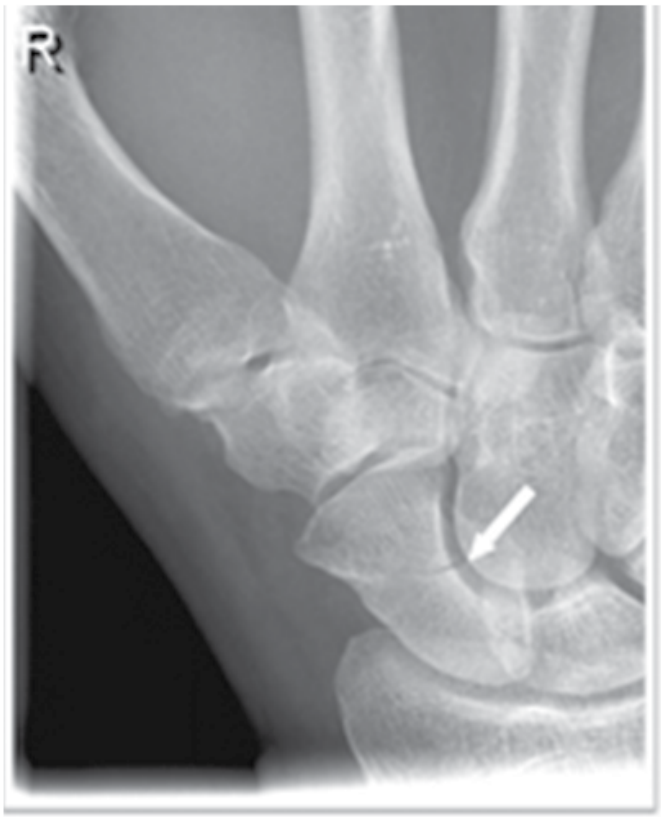

Figure 1: Radiograph demonstrating an undisplaced scaphoid waist fracture (white arrow).

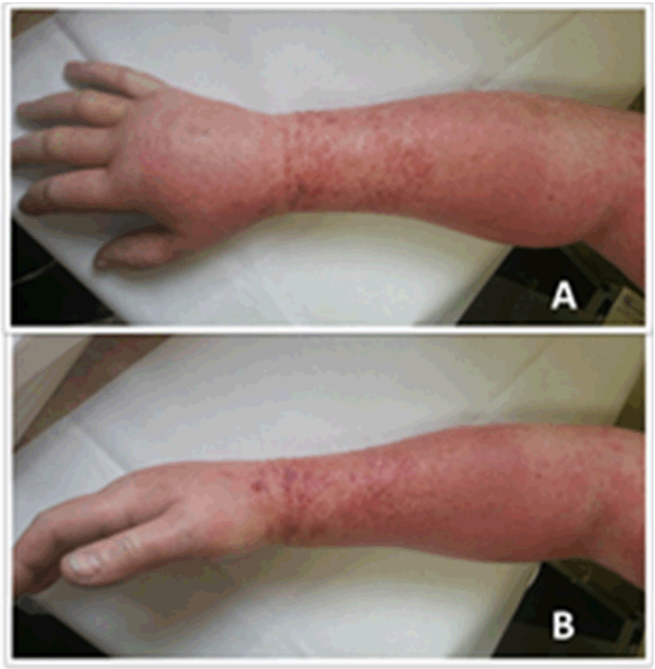

Figure 2: Clinical photographs of the right fore arm demonstrating the extensive skin involvement and pustular lesions.

DISCUSSION

This case report describes acute allergic contact dermatitis (ACD) secondary to fiberglass orthopedic cast

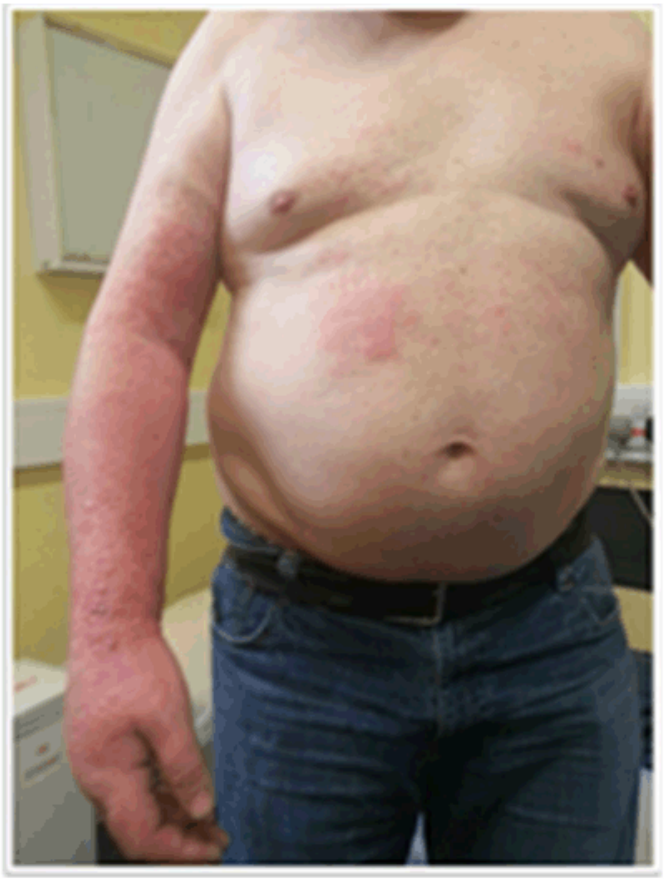

Figure 3: Clinical photograph demonstrating the allergic contact dermatitis involvement of the forearm, arm and abdomen.

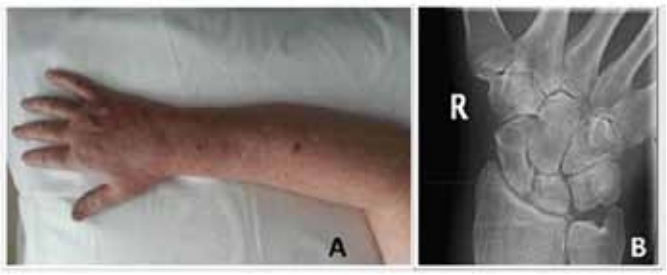

Figure 4: (A) Clinical photograph demonstrating resolution of allergic contact dermatitis, (B) Radiograph demonstration healed scaphoid fracture.

application. While previous case reports have reported allergic skin reactions to POP, the development of ACD to newer fiberglass cast agents have not been reported in the literature to date. Allergic contact dermatitis is the result of a T cell mediated, delayed-type hypersensitivity response to an exogenous agent on direct contact [3]. Hapten-binding is the initial step in the development of ACD and after initial sensitization CD 4+ and CD 8+ T cells as well as natural killer T cells and T regulatory cells are critical participants in the immunogenic response [4]. Clinically, it presents as erythematous, indurated skin often with edema and bullae formation [5]. Secondary skin changes may be seen in the form of excoriation or impetiginization as was seen in this case also [6]. While lesions are typically seen in areas in direct contact with the irritant, diffuse distributions may also occur depending on the nature of the allergen or transfer of the allergen from the primary site of contact to distant skin areas [7]. Steroids and anti-histamine therapies provide the foundation of management along with treatment of superimposed infection.

3M(TM) Scotch cast (TM) tape currently is the most commonly used orthopedic casting agent and was used in this case. It contains a resin embedded in woven fibreglass and can be molded to the required shape for immobilization. Chemical constituents include fibrous glass, 4,4'-diphenylmethane diisocyanate-polypropylene glycol polymers and dimorpholino diethyl ether [8]. Patch testing for these agents may identify atopy and confirm diagnosis but clinical diagnosis is sufficient to recommend future avoidance of this material in patients showing sensitivity to this material. We must make note of the clinical difficulty in this case between fracture management and skin management. The scaphoid, due to its retrograde blood supply is vulnerable for non-union if not appropriately managed. However, due to the extent of the patient distress form the significant cutaneous involvement, skin management was prioritized. Splint immobilization was sufficient to manage the fracture.

CONCLUSION

A high index of suspicion should be kept when patients with an orthopedic cast present to the emergency department with persistent pain or other unexplained symptoms under an orthopedic cast. It is often due to the underlying fracture itself or the subsequent edema of the affected limb but occasionally may be more sinister. While radiographic assessment is important, cast removal must be performed as a matter of urgency in any patient who presents with unexplainable pain, persistent burning or neurovascular symptoms to allow for full examination and ensure there is no evidence of compartment syndrome. An urgent orthopedic opinion must be sought on these patients.

****

Author Contributions

Christina A. Fleming – 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

Shane C. O'Neill – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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

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

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Local invasion of jaw osteosarcoma

Roberto Fiori, Dominique De Vivo, Angela Lia Scarano, Silvia D'Onofrio, Eros Calabria, Simonetti Giovanni

ABSTRACT

Introduction: One of the most common primary malignant bone tumor is osteosarcoma that counts high percentages in the level of incidence the 5% of all tumor in children, and the 9% of all sarcomas. Osteosarcoma are malignant bone tumors developing from mesenchymal tumor cells that growth as disorganized immature bone or osteoid tissue. Patients affected by osteosarcoma show a peak incidence related to puberty (for young female 12±2 years and for young male 16±1.8 years). Regardless of the favorable biological behaviour, the patients of jaw osteosarcoma usually exhibit advanced tumor as it often goes unnoticed by the dental professional thus stressing on the need for early diagnosis of the lesion. Case Report: We report an unusual case history of a 34-year-old male with swelling and blood loss in anterior gingival of maxilla with loss of teeth. Conclusion: There are three main methods to diagnose the tumor based on clinical and radiographic characteristics before confirmation by histology and to bring attention to dental professional to approach cases diagnosing them at an early stage leading to better prognosis.

Keywords: Bone tumor, Jaw, Maxillofacial neoplasms, Mesenchymal neoplasia, Osteosarcoma

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INTRODUCTION

One of the most common primary malignant bone tumor is osteosarcoma that counts high percentages in the level of incidence the 5% of all tumor in children, and the 9% of all sarcomas. Osteosarcoma are malignant bone tumors developing from mesenchymal tumor cells that growth as disorganized immature bone or osteoid tissue [1, 2]. Patients affected by osteosarcoma show a peak incidence related to puberty (for young female 12±2 years and for young male 16±1.8 years) [1].

Osteosarcoma is an osteoid-producing malignant mesenchymal tumor and accounts for 15–35% of all primary bone tumors although only 6–10% of osteosarcomas occur in the craniofacial region [3]. The international ranks consider the mandible as the common site of involvement, immediately followed by maxilla and skull [4, 5].

The gold standard is an early diagnosis in order to prevent its continuous infiltrative growth [6].

Osteosarcoma usually metastasizes through the blood such as mesenchymal malignancy. The aim of this case report is to highlight how malignant neoplasm with continuous autonomous growth, and to make a comparison between osteosarcoma in general and osteosarcoma of the jaw that is known to have a relatively low incidence of metastasis and a better prognosis [7].

CASE REPORT

A 34-year-old male was admitted to our emergency department with the complaint of a painless swelling of the cheek gradually enlarging for over nine months and difficulty in swallowing solids and liquids and progressive loss of teeth in the upper maxilla.

On physical examination, the patient presented mild displacement of the eye, difficulty in opening the mouth, gingival swelling, tears from eyes and loss of teeth on the upper maxilla. He showed pain on palpation, the swelling was firm, hard and immovable and was fixed to the underlying bone.

At intraoral examination, the patient had a great mass extending intraoral into the hard palate as an ulcerated, painful mass invading the nasal cavity.

Blood pressure, pulse and respiratory rate and temperature were normal.

After admitting to emergency department the patient underwent contrast-enhanced computed tomography (CT) scan which revealed tumor with a major component like soft tissue, central calcification/ossification, originating in the left upper quadrant of the maxilla and invading the nasal cavity, the ethmoid cells, the sphenoidal and frontal sinuses, the right upper quadrant of the maxilla, the left alveolar process, the hard palate and the base of the orbit without involvement of the orbital adipose tissue.

Computed tomography scan examination before and after injection of contrast medium also showed the involvement of the tumor mass to the parotid gland, the masseteric fossa and the infra-temporal fossa bilaterally with multiple increased lymph nodes on mandibular angle and in lateral cervical region the invasion (Figures 1 and 2). We elaborated three-dimensional images to better define the extension of the lesion (Figure 3). In the suspicion of distal localization the patient underwent CT whole body which was negative.

A biopsy under local anesthesia proved coexistence of osteoblastic and chondroblastic osteosarcoma (Figure 4). So-called osteoblastic osteosarcoma is dominated by the production of extracellular bone matrix. This picture demonstrates immature bone deposition into which tumor cells are incorporated. So-called chondroblastic osteosarcoma. This picture is dominated by cellular hyaline cartilage with wisps of osteoid formation.

The patient was admitted to oncological center for further evaluation and treatment.

Chemotherapy was performed and nowadays the patient is in a periodical follow-up.

Osteosarcoma is the second primary malignant tumor of the bone after multiple myeloma although its localization in the orofacial region is rare [8]. Among the possible symptoms that patients usually present there are pain, paresthesia and swelling of the area similar to osteomyelitis, ossifying fibroma, periostitis, osteoblastoma, suppurative osteomyelitis, and even fibrous dysplasia [7, 9].

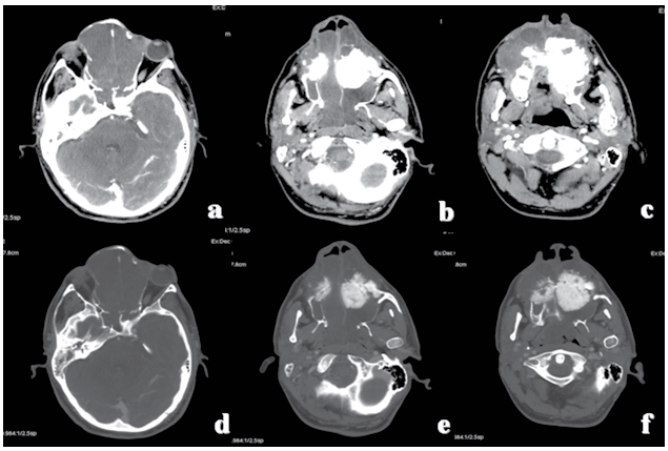

Figure 1: Computed tomography scan after injection of contrast medium. Axial images evaluated (a–c) with soft tissue window and (d–f) with bone tissue window (d, e, f) revealed tumor with a major component like soft tissues, central calcification/ossification, originating in the left upper quadrant of the maxilla and invading nasal cavity, ethmoid cells, sphenoidal and frontal sinuses, the right upper quadrant of the maxilla, left alveolar process, hard palate and the base of the orbit without involvement of the orbital adipose tissue.

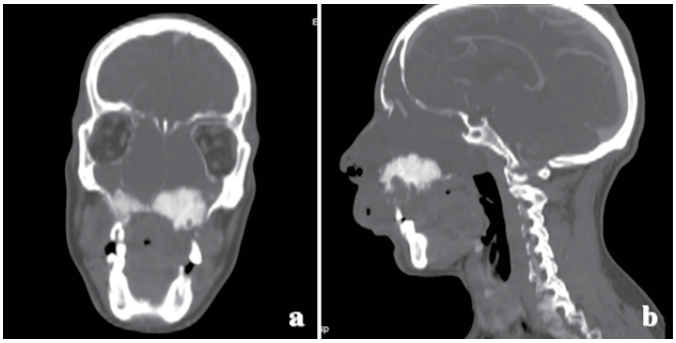

Figure 2: Computed tomography scan after injection of contrast medium. (a) Coronal and (b) Sagittal images demonstrate the lesion originating in the left upper quadrant of the maxilla and invading the nasal cavity and the sphenoidal and frontal sinuses.

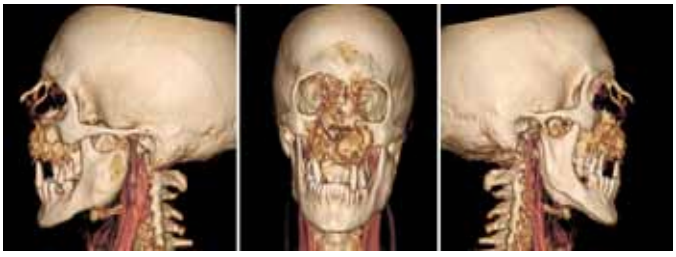

Figure 3: Computed tomography 3D reconstruction showing the mass in the upper maxilla, invading the left alveolar process, the hard palate, the nasal cavity and the base of the orbit.

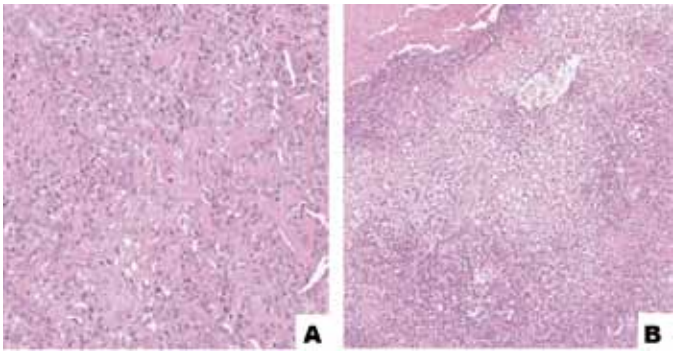

Figure 4: A biopsy proved coexistence of (A) osteoblastic and (b) chondroblastic osteosarcoma. (A) So-called osteoblastic osteosarcoma is dominated by the production of extracellular bone matrix. This picture demonstrates immature bone deposition into which tumor cells are incorporated. (B) So-called chondroblastic osteosarcoma. This picture is dominated by cellular hyaline cartilage with wisps of osteoid formation. (H&E stain, x100).

Table 1: Osteosarcoma Variants (WHO 2002).

Central (Medullary)
1 Conventional osteosarcoma
Chondroblastic
Fibroblastic
Osteoblastic
2 Telengiectatic osteosarcoma
3 Intraosseous well-differentiated osteosarcoma
4 Small cell osteosarcoma
5 Low-grade central osteosarcoma
6 High-grade central osteosarcoma
7 Secondary osteosarcoma
Surface (Peripheral)
1 Parosteal (juxtacortical) well-differentiated osteosarcoma
2 Periosteal osteosarcoma
3 High-grade surface osteosarcoma

DISCUSSION

Osteosarcoma is a frequent primary malignant bone tumor and it rarely appears in the orofacial anatomical region.

The patients usually refer pain, swelling and sometimes bleeding from the interested area. In a relatively short time the affected patients present paresthesias, initial mobility and then loss of the tooth. These clinical non-specific symptoms are similar to some benign tumors, malign tumors and infective disease (osteoblastoma, ossifying fibroma, fibrous dysplasia and infective pathologies like osteomyelitis with proliferative periostitis, suppurative osteomyelitis) [1–7].

For these reasons radiologic and histopathologic characterization in association with clinical examination are necessary in detecting the kind of disease in order to plan a correct management of the case. On a radiographic examination osteosarcoma can present as radiopacity, radiolucency or a mixed aspect [4]. In some cases, there should be widening of the periodontal ligament space also known as *Garrington sign* [10]. A plain radiograph usually demonstrates a destructive mass associated to irregular, spiculated periosteal reaction [11]. An integrative CT scan confirms irregular mass with periosteal and endosteal reaction even if osteosarcoma and fibrous dysplasia have a similar presentation even if as “sunray spiculations” in soft tissue could indicate osteosarcoma.

For these reason the gold standard is a histologic evaluation through a biopsy. The histological appearance of osteosarcoma is highly variable, in the jaw the most frequent variant is the chondroblastic pattern. Deep biopsy should be recommended to avoid the histological characterization of the periosteal reaction (Table 1).

In osteosarcoma, there is a direct formation of osteoid from a sarcomatous tissue variable from a sclerotic osseous tumor to diffuse osteoid aspect. The stromal cells may be osteoblastic, chondroblastic, and/or fibroblastic. The most common form is osteoblastic growth but in the jaws localization the most common aspect is chondroblastic pattern.

In case of osteosarcoma, a not rare event is the recurrence or metastases appearances. The most frequent target organ is the lung, for this reason a chest X-ray or a chest CT scan is mandatory for surgical and or oncological planning. In our case, the patient was in an advanced stage of the disease and there was a high probability of metastatic lesions. The patient underwent cycles of chemotherapy and surgical intervention in another hospital and he is actually undergoing trimestral clinical and radiological follow-up. A late diagnosis do not allow good life expectancy and the prognosis is very poor.

CONCLUSION

In conclusion, osteosarcoma of the jaw is an aggressive bone tumor that should be treated at referral centres, by a multidisciplinary team (radiologist, oncologists, maxillofacial, and head and neck surgeons, radiotherapist, maxillofacial prosthodontist including plastic surgeons) with a experience managing these lesions because an early diagnosis could allow a better prognosis for the patient consisting in a complete tumor resection and planning maxillofacial reconstruction.

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Author Contributions

Roberto Fiori – 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

Dominique De Vivo – Analysis and interpretation of data, Drafting the article, Revising it critically for important

intellectual content, Final approval of the version to be published
Angela Lia Scarano – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Silvia D'Onofrio – Analysis and interpretation of data, Drafting the article, 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, Revising it critically for important intellectual content, Final approval of the version to be published
Simonetti Giovanni – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Late infantile metachromatic leukodystrophy in a two-year-old boy: A case report

Jillalla Narsing Rao, Swathi Chacham, Uppin Narayan Reddy, Janampally Ravikiran, S. Pratap Rao, Battula Sugunakar Reddy

ABSTRACT

Introduction: Metachromatic leukodystrophy (MLD) occurs due to cerebroside sulfatide accumulation in the extraneural and neural tissues. Arylsulfatase A (ARSA) enzyme deficiency leads to progressive focal or generalized demyelination. This rare disorder involves both central and peripheral nervous system. Case Report: We report a two-year-old boy, born of consanguineous marriage presenting with recurrent seizures from 13 months of age, followed by regression of milestones and spasticity from 15 months of age. Neurodegenerative disorder was considered initially and the child underwent neuroimaging followed by enzyme level assay. Magnetic resonance imaging scan (MRI) of brain revealed demyelination involving both cerebral cortex in the periventricular white matter with sparing of subcortical 'u' fibers and tigroid appearances / leopard skin sign, hallmark of metachromatic leukodystrophy. Normal β-galactosidase enzyme activity along with undetectable (ARSA) enzyme levels confirmed the diagnosis of late infantile variant of metachromatic leukodystrophy. Conclusion: A two-year-old boy presented with recurrent, generalized seizures, regression of milestones along with characteristic MRI findings and untraceable ARSA activity suggesting late infantile metachromatic leukodystrophy.

Keywords: Arylsulfatase deficiency, Degenerative disorder, Infantile, Metachromatic leukodystrophy

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INTRODUCTION

Metachromatic leukodystrophy (MLD), a rare neurodegenerative metabolic disorder occurs with an incidence of 1 in 40, 000 to 1,60 000 individuals, worldwide [1]. Arylsulfatase A (ARSA) (a lysosomal

enzyme) deficiency results in sulfatide accumulation within the myelin sheath of the central and peripheral nervous system, leading to progressive focal or generalized white matter degeneration [2, 3].

CASE REPORT

A two-year-old male child born of third-degree consanguinity, presented with the history of generalized, recurrent seizures from 13 months of age. The developmental milestones were normal till one year of life. The developmental age in all quadrants (motor, language and social) was appropriate. The child had attained neck holding at third month and was able to sit with support by sixth month. Sitting without support was attained by eighth month and was walking with support by nineth months. At the age of one year, the infant was able to stand without support. The child was speaking mono-syllables by sixth month and bi-syllables by nine months. He showed stranger anxiety by 10th month. At the age of 13th month, the child developed generalized, tonic-clonic seizures which were associated with fever and minor respiratory illness. Seizures lasted for 20–25 minutes and required administration of two anti-epileptic agents. At the time of admission, there were no meningeal signs and CSF analysis (WBC 3 cells/mm^3, neutrophils nil, glucose 56 mg/dL, protein 21 mg/dL, chloride 121 mmol/l, LDH 158U/l, ADA 4U/L, negative for acid-fast bacilli, culture and sensitivity— no bacterial growth) was not suggestive of meningitis. Hence the child was discharged on two anti-epileptic agents. At the age of 15th months, he developed recurrent seizure episodes, which necessitated administration of 3rd anti-epileptic agent. This episode was succeeded by regression of milestones which was progressive in nature. He developed progressive inability to walk, sit and speak with in a time span of one to two months. The child required frequent hospitalization for recurrent seizures and required multiple anti-epileptic agents. The antenatal period was uneventful and the infant was delivered by spontaneous vaginal delivery with birth weight of 3 kg. No history of perinatal asphyxia, neonatal seizures and neonatal jaundice. No maternal history of fever, rash in the antenatal period, which could be suggestive of toxoplasmosis, rubella, cytomegalovirus, and herpes simplex (TORCH) infection. No family history of similar complaints. On examination, there were no neurocutaneous markers and no facial dysmorphism. Head circumference was 46 cm. Cardiovascular system and respiratory system were unremarkable. However, central nervous system revealed generalized spasticity, exaggerated deep tendon reflexes and extensor plantar response. Retinal examination was unremarkable. Initial differential diagnosis of infantile stroke, cerebral palsy, and neurodegenerative disorder was considered and the child was further investigated. MRI scan of brain was abnormal and the β-galactosidase levels were normal. However, undetectable arylsulfatase A activity in the child confirmed the diagnosis of MLD. MRI brain image showed Tigroid appearances /Leopard Skin sign, suggesting metachromatic leukodystrophy(as shown with arrows in Figure 1). There was demyelination of periventricular white matter of cerebral cortex on both sides along with sparing of subcortical 'u' fibers(as shown in Figure 2). The patient was treated with supportive care together with physiotherapy. However, bone marrow transplantation, a newer modality of treatment was not feasible due to financial constrains.

DISCUSSION

Aryl sulfatase A, a lysosomal enzyme aids in degradation of sulfated glycolipids, especially galactosyl sulfatide. Classical type MLD results from this arylsulfatase A enzyme deficiency, which leads to accumulation of sulfatide in central and peripheral nervous system, progressive demyelination, motor and cognitive dysfunction. MLD has been categorized depending on the age of presentation as late-infantile type (onset before three years of age), juvenile type (onset before 16 years) and adult type [4]. Late infantile and the juvenile variants are characterized by rapid motor decline, while adult form presents with cognitive and behavioral

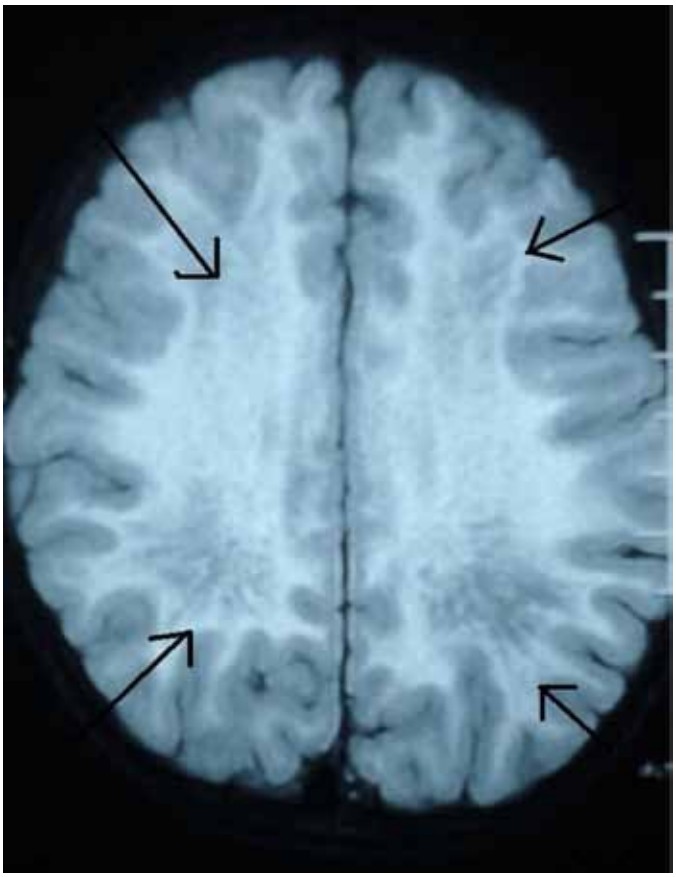

Figure 1: Magnetic resonance imaging of brain revealing tigroid appearances/leopard skin sign suggesting metachromatic leukodystrophy.

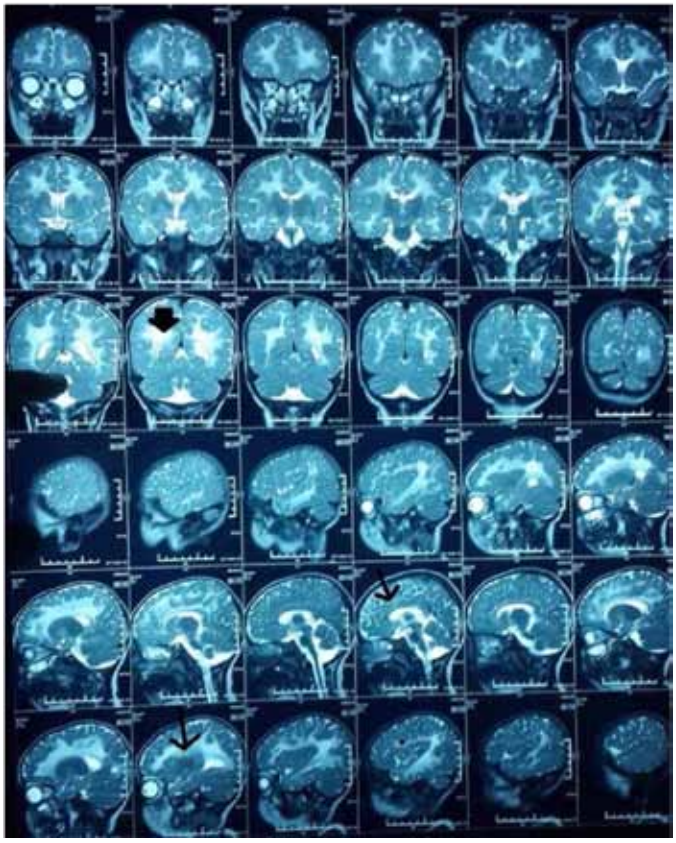

Figure 2: Magnetic resonance imaging of brain showing demyelination of periventricular cerebral cortical white matter on both sides with sparing of subcortical 'u' fibers.

problems. The index case is considered as late infantile variant of MLD, based on the age of onset and clinical presentation. The index child presented with typical regression of milestones following refractory seizures. Another characteristic feature of MLD is recurrent seizures and their incidence increases with duration of illness, as in the index child. Generalized seizures are common in infantile type (as in the present case), while partial seizures are more common in juvenile type. Older children present with gait abnormalities, seizures, behavioral changes and deterioration of scholastic performance. Along with regression of milestones, spasticity (a characteristic manifestation of MLD) was also present in the index child [5, 6]. The important diagnostic modalities used to confirm this degenerative disorder are arylsulfatase A enzyme activity, molecular genetic testing of arylsulfatase A, estimation of urinary sulfatide and detecting metachromatic lipid deposits in the nervous tissue [7]. Gene sequence analysis of arylsulfatase A is an important tool for prenatal diagnosis. As MLD progresses with age and the neurodegeneration worsens with time, there is no definitive treatment till date. Newer treatment modalities include stem cell transplantation, bone marrow transplantation along with genetic engineering and these might halt the progression of neurologic dysfunction [8–10]. Recombinant human ARSA administration, an experimental treatment can be a promising option in future, although it lacks universal recommendation and adaptation.

CONCLUSION

A two-year-old boy born of consanguinity, manifesting with generalized refractory seizures followed by regression of milestones and spasticity. Characteristic MRI findings, tigroid appearance/leopard skin sign coupled with undetectable arylsulfatase A activity suggests late infantile variant of metachromatic leukodystrophy.

ABBREVIATIONS

ARSA (Aryl sulfatase A), MLD (Metachromatic Leukodystrophy), MRI (Magnetic resonance imaging), TORCH (Toxoplasmosis, Rubella, Cytomegalovirus and Herpes simplex)

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Rhabdomyolysis and acute kidney injury in Salmonella gastroenteritis: A case report

Ziqiang Zhu, Sina Aghaie, Andrei Bandarchuk, Anjula Gandhi

ABSTRACT

Introduction: Although infection by *Salmonella* has been commonly associated to diarrhea, complications such as cholecystitis, pancreatitis, acute pyelonephritis, osteomyelitis, myocarditis, encephalopathy, and multi-organ failure have been rarely reported. Case Report: Herein, we report a case of *S. enteritidis* gastroenteritis complicated by rhabdomyolysis and acute kidney injury, which was successfully treated with hydration and antibiotics without dialysis. In this report, we reviewed all the previously published cases of rhabdomyolysis associated with *Salmonella* infection in English literatures and discussed the prevalence, pathogenesis and treatment. Conclusion: Rhabdomyolysis was thought to be a rare complication of *Salmonella* infection. However, due to the variable manifestations that depend upon the extent and severity of muscle damage, rhabdomyolysis is perhaps under diagnosed as an extra-intestinal manifestation of *Salmonella* infection. Therefore, it is critical to recognize the condition promptly, initiate early antibiotics treatment and provide good supportive care. A high index of suspicion may help to reduce significant co-morbidities associated with the development of rhabdomyolysis in *Salmonella* infection.

Keywords: Acute kidney injury, Gastroenteritis, Rhabdomyolysis, *Salmonella* infection

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INTRODUCTION

Salmonella is estimated to cause more than 1.2 million illnesses every year in the United States [1], with 19,000 hospitalizations and 380 deaths every year. Among over 2500 serotypes of *Salmonella*, only a few of them can cause human infection [2], which can be divided into two categories–those cause typhoid fever and those cause gastroenteritis. *Salmonella typhimurium* and—*enteritidis* are the two most common serotypes in the United States. *Salmonella* infection may have different clinical presentations. The elderly, children, and immunocompromised patients are more likely to have severe disease. The most common manifestation of salmonellosis is gastroenteritis, including fever, diarrhea, and abdominal pain 12–72 hours after infection, which accounts for 70% of cases. Extraintestinal manifestations are not uncommon and have been reported both in immunocompromised and immunocompetent patients involving a variety of organ systems, including acute cholecystitis [3], pancreatitis [4], acute pyelonephritis [5], osteomyelitis [6], myocarditis [7], encephalopathy [8], disseminated intravascular coagulation (DIC) and

multi-organ failure [9]. Rhabdomyolysis secondary to *Salmonella* infections was thought to be a rare complication and has only been reported a few times. Here, we present a case of rhabdomyolysis and acute kidney injury with *Salmonella enteritidis* infection in a middle-aged male.

CASE REPORT

A previously healthy 42-year-old African-American male presented to the emergency department with 20–30 watery stools daily for three days, accompanied by nausea and vomiting. Three days prior to admission, the patient ate lunch at a local restaurant and later that night, he developed fever, chills, muscle cramping and watery diarrhea. There was no dysuria, melena, no history of recent illness or antibiotic use or contact with persons with similar illness prior to admission. He denied any recent travel outside the United States. Only past medical history was asthma. The patient was a healthy and active man with no history of alcohol, tobacco or illicit drug use.

On physical examination, he appeared ill-looking and dehydrated. He was afebrile, with blood pressure of 122/89 mmHg. His pulse was weak but regular at 87 beats/min. Bowel sounds were present and active. Generalized abdominal tenderness without guarding or rebound or organomegaly was noted on palpation. No skin rash was appreciated. No muscular weakness was elicited.

Hematological laboratory findings and blood chemistry values on the day of admission and subsequent days are given in Table 1. Pertinent laboratory data on admission included leukocytosis $13.3x10^9$/L, and hemoglobin 16.6 g/dL. The serum creatinine level was elevated to 10.5 mg/dL, with an estimated glomerular filtration rate (GFR) of 5 mL/min per 1.73 m^2, suggesting acute kidney injury. The patient was found to have pure metabolic acidosis (anion gap 35). ALT and AST were mildly elevated. The creatinine kinase (CPK) level significantly elevated at 21 481 U/L. Urinalysis showed a specific gravity of 1.010 and a pH 5.5. A spot urine test showed urinary sodium of 9 mEq/L and creatinine 113 mg/dL, sample was positive for myoglobin. Rapid influenza and HIV tests were both negative. *Salmonella Group D enteritidis* was isolated from stool, while the blood and urine culture remained sterile. An abdominal ultrasound showed normal liver, spleen and kidney. Electrocardiogram showed sinus tachycardia and no ST-T wave changes.

The patient initially was managed by intravenous hydration and alkalization by sodium bicarbonate, he responded well to hydration. Patient was not on antibiotics until day 5 of admission when revealed *Salmonella* in stool culture. Ceftriaxone was initiated and one day later switched to oral ciprofloxacin. Good clinical response and complete normalization of most of the laboratory parameters were observed during treatment. On discharge, 7 days after admission, the patient was asymptomatic, enzyme levels had returned to normal, and serum creatinine decreased to 1.3.

DISCUSSION

Rhabdomyolysis is characterized by muscle pain and markedly elevated CPK level [10]. An elevated serum CPK to at least five times the upper limit of normal and usually greater than 5,000 U/L is diagnostic of rhabdomyolysis. Myoglobinuria may be present in rhabdomyolysis. Rhabdomyolysis has been reported in numerous disorders including the common etiologies [10] such as trauma, ischemic disorders, autoimmune diseases, drugs and toxins. It has also been associated with both viral and bacterial infections [11, 12] such as influenza, Coxsackievirus, Epstein-Barr virus, echovirus, HIV, *Legionella*, *Staphylococcus* and *Escherichia coli*. Rhabdomyolysis secondary to *Salmonella* infection has also been described in literature [4, 9, 13]. However, the mechanisms remain not fully elucidated. It was proposed that dehydration, hypoxia, electrolyte disturbances, bacterial invasion of the muscle, and *Salmonella* toxic effects on muscle cell metabolism could be responsible for muscle cell injury [14].

Acute kidney injury (AKI) is a well-known complication of rhabdomyolysis. The reported frequency of acute kidney injury ranges from 13% to approximately 50% [10]. Acute renal failure due to rhabdomyolysis in typhoid was first described as early as 1977 [13]. Later, non-typhoid *Salmonellae*, *i.e. Salmonella enteritidis, Salmonella bonariensis, Salmonella group C and Salmonella typhimurium,* have also been reported with this complication [4, 15, 16]. Therefore, rhabdomyolysis and AKI are potential complications of salmonellosis. Table 2 gives the characteristics of all the reported individual cases of rhabdomyolysis associated with microbiologically proven *Salmonella* infection in English language journals. Janssen et al. [17] reviewed over a two-year period of 44 hospitalized adult patients with *Salmonella* gastroenteritis, and reported 36% of the cases complicated with acute renal failure. However, during the course of *Salmonella* infection, the muscle involvement in some cases may or may not be clinically evident. Therefore, rhabdomyolysis is perhaps under diagnosed and may constitute an extraintestinal manifestation of the infection. In addition, the associated risk of renal failure is not correlated with the peak CPK level. Clinicians should be aware of this possibility of muscular involvement and development of acute renal failure. The occurrence of acute kidney injury in many patients is believed due to dehydration from gastroenteritis, tissue hypoxia from sepsis, rhabdomyolysis, nephrotoxicity of myoglobin [14]. Other rare causes of acute kidney injury in *Salmonella* infection include glomerulonephritis, acute tubular necrosis (ATN) and interstitial nephritis [4, 18].

The overall prognosis of acute kidney injury from *Salmonella* infection based on our review of reported

Table 1: Hematological and blood chemistry laboratory values

Day of admission	1	2	3	4	5	6	7
WBC (10^9/L)	13.3	8.1	6.3	5.3	6.8	8.8	9.5
Hg (g/dL)	16.6	15.0	13.2	12.7	12.7	12.5	12.2
PLT(10^9/L)	218	173	161	171	168	195	206
BUN (mg/dL)	54	68	62	37	18	13	14
Creatinine (mg/dL)	10.5	9.4	5.5	2.1	1.7	1.3	1.3
Serum Na (mEq/L)	127	126	131	131	133	136	137
Serum K (mEq/L)	5.5	4.5	3.6	3.1	3.4	3.3	3.6
Serum HCO_3^- (mEq/L)	16	17	18	27	32	35	25
ALT (U/L)	74	100	110	100	94	97	110
AST (U/L)	467	528	492	356	267	51	58
CPK (U/L)	21 481	25 102	--	11 426	5 660	1 241	849

WBC: white blood cell, Hg: hemoglobin, PLT: platelets, BUN: blood urea nitrogen, CPK: creatinine kinase

Table 2: Details of reported cases of rhabdomyolysis associated with *Salmonella* infection in English literature

Author (Reference)	Age	Sex	*Salmonella* Subtype/ Source	CPK (U/L)	WBC (10^9/L)	Serum Creatinine (mg/dL)	Treatment
Blaauw et al. [3]	27	F	*S. enteritidis*/Urine, stool, blood	45429	9.7	1.37	Ceftriaxone
Abdulla et al. [4]	56	M	*S. enteritidis*/Blood	2801	8	9.09	Ciprofloxacin Hemodialysis
	50	M	*S. enteritidis*/Stool	3200	4.8	7.16	Ciprofloxacin
Retornaz et al. [9]	58	M	*S. enteritidis*/Blood	2104	12	3.05	Tazocillin Ciprofloxacin
Brncic et al. [14]	58	F	*S. infantis*/Blood	>64000	17.1	3.30	Hemodialysis
Sirmatel et al. [15]	21	M	*S. paratyphi B*/Blood	4070	2.5	5.70	Ceftriaxone Ciprofloxacin Deceased
Gingold-Belfer et al. [16]	24	M	*S. group C*/Stool	24073	5.92	1.06	Ofloxacin
Fisk et al. [20]	25	M	*S. entericaserovartyphi*/ Blood	31410	11.8	1.79	Ceftriaxone Ciprofloxacin
Ali et al. [21]	42	M	*S. typhi*/Blood	17000	Normal	12.7	Ciprofloxacin
Jhawar et al. [22]	64	M	*S. typhi*/Stool	9473	3.2	3.7	Cefoperazone, Ciprofloxacin, Imipenemcilastin Hemodialysis
Al-aqeedi et al. [23]	34	M	*S. typhi*/Blood	6341	4.5	2.38	Ceftriaxone
Khan et al. [24]	23	M	*S. typhi*/Blood, urine stool	5350	2.2	6.03	Ceftriaxone
Neau et al. [25]	72	M	*S. enteritidis*/Stool	3008	--	1.22	Ofloxacin
	73	F	*S. enteritidis*/Stool	213	--	1.26	Ofloxacin
	37	M	S. enteritidis/Stool	1124	--	8.05	Ofloxacin
Campistol et al. [26]	43	M	*S. enteritidis*/Stool	1870	14.6	9.9	Ampicillin
	51	M	*S. enteritidis*/Stool	4300	11.8	12	Ampicillin
	38	M	*S. enteritidis*/Stool, blood	2270	11.7	13	Ampicillin

cases, in addition to the current case, seems to have a benign course. Among all 18 reported cases in Table 2, one patient died because of multi-organ failure [15]. Hemodialysis was not required before renal failure resolved in majority of the reported cases. Abdulla et al. [4] described two adult patients with severe rhabdomyolysis due to *S. enteritidis* complicated by ATN requiring hemodialysis in one case. In a retrospective study of 44 adult patients with *Salmonella* infection induced acute renal failure, kidney function recovered in all but 1 patient [17]. However, rhabdomyolysis induced acute kidney injury from *Salmonella* infection in previous reported pediatric cases has largely in the need for dialysis [19]. It is unclear whether there is any difference between adult and pediatric cases in terms of pathogenesis, treatment and prognosis in rhabdomyolysis associated with *Salmonella* infection.

In the current case, the patient's presenting symptoms of fever and diarrhea were typical of *Salmonella* gastroenteritis. We considered his rhabdomyolysis due to *Salmonella* infection since other causes of rhabdomyolysis such as trauma, medications, illicit drugs, ischemic disorders and infection like influenza were all ruled out. Despite having elevated muscle enzymes, he had no muscle weakness on examination, therefore, *Salmonella*-induced myopathy was also ruled out. His kidney function responded well to conservative management and returned to baseline within seven days of admission. Aggressive early recognition of the rhabdomyolysis, extensive fluid replacement and appropriate treatment with antibiotic may have lead to successful management of *Salmonella*-induced rhabdomyolysis and renal failure without progression to ATN in our patient [20–26].

CONCLUSION

In summary, depending upon the extent and severity of muscle damage, the manifestations of rhabdomyolysis secondary to *Salmonella* infection can vary from mild myalgia to severe pain with weakness. The associated risk of renal failure may not correlate with the peak creatinine kinase (CPK) level. Therefore, a high index of suspicion may help to reduce significant co-morbidities associated with the development of rhabdomyolysis in *Salmonella* infection.

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Author Contributions

Ziqiang Zhu – 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
Sina Aghaie – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Andrei Bandarchuk – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Anjula Gandhi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT PEER REVIEWED | OPEN ACCESS

Cardiac rehabilitation for cardiac syndrome X and microvascular angina: A case report

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ABSTRACT

Introduction: Therapeutic strategies for cardiac syndrome X, characterized by the three features of angina, evidence of myocardial ischemia and no obstructive coronary artery disease, are not well known. Recent data indicate that angina secondary to microvascular coronary dysfunction is a common pathogenesis of cardiac syndrome X. While cardiac rehabilitation is well-known to be effective for angina due to obstructive coronary artery disease, less is known in cardiac syndrome X and microvascular angina patients. Case Report: A 34-year-old female with history of pre-eclampsia during three pregnancies, recurrent non-ST-segment-elevation myocardial infarctions, no obstructive coronary artery disease, microvascular coronary dysfunction documented by coronary reactivity testing, ischemic cardiomyopathy, overweight, factor V Leiden mutation, and persistent microvascular angina was referred to cardiac rehabilitation five weeks after a non-ST-segment-elevation myocardial infarction. The patient was taking a beta blocker, ACE, statin and low dose aspirin. She underwent four sessions of supervised cardiac rehabilitation program which progressed to a duration of 45 minutes at a level of 3.3 metabolic equivalents of aerobic exercise. A normal cardiovascular response without arrhythmias was observed. A home program was suggested with moderate intensity of aerobic exercise, 30–45 minutes per day, most days of the week. She experienced decreased angina, improved quality of life and increased functional capacity at the fourth-year of follow-up. Conclusion: Cardiac rehabilitation was beneficial in the patient angina due to microvascular coronary dysfunction. Increased exercise intensity and duration, increased functional capacity, decreased anginal symptoms and improved quality of life were found.

Keywords: Cardiac rehabilitation, Cardiac syndrome X, Exercise training, Microvascular angina, Microvascular coronary dysfunction

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INTRODUCTION

Therapeutic strategies for cardiac syndrome X (CSX), characterized by three features, including angina, evidence of myocardial ischemia on stress testing, and no obstructive coronary artery disease (CAD) are not well known. Data from the National Heart, Lung and Blood Institute-sponsored Women's Ischemia Syndrome Evaluation (NHLBI-WISE) study demonstrates that microvascular coronary dysfunction (MCD) is present in approximately half of women with CSX [1]. In addition, prognosis for patients with CSX may not be as benign as previously thought especially in patients with evidence of microvascular coronary dysfunction. The NHLBI-WISE study demonstrated a five-year cardiovascular event rate of 16% for women with non-obstructive CAD and 7.9% for women with normal coronary arteries, compared to 2.4% for asymptomatic women with no history of heart disease [2]. Nevertheless the contributing pathogenesis of angina in CSX remains controversial. Possible mechanisms for the angina have been reported, including myocardial ischemia due to microvascular and endothelial coronary dysfunction, and abnormal cardiac pain perception [3].

While there is wide acceptance of the benefits of cardiac rehabilitation (CR) for patients with obstructive CAD [4], CR is underutilized with the approximate mean referral rate of 34%. Among CAD patients eligible for CR, participation rates for women are lower than men in most studies [5]. Furthermore, there are few studies evaluating impact of CR on CSX patients and microvascular angina patients. In this report, the case of woman with microvascular angina is examined to highlight the potential benefit of CR in this group of patients.

CASE REPORT

A 34-year-old female was referred to cardiac rehabilitation five weeks after a non-ST-segment-elevation myocardial infarction (NSTEMI) in 2008. She had a prior history of pre-eclampsia with three pregnancies and was heterozygous for factor V Leiden mutation. She had a history of NSTEMI with no obstructive CAD in 2005 and had persistent exertional and non-exertional episodes of angina. She had multiple emergency department visits for angina. She was diagnosed with MCD by invasive coronary reactivity testing using intra-coronary adenosine and acetylcholine [6] in 2007.

MCD was diagnosed by invasive coronary reactivity testing as previously published (Figure 1) [6]. She had normal coronary arteriography without luminal irregularity and left ventricular end-diastolic filling pressure was elevated at 17 mmHg. Coronary flow reserve (CFR) in response to adenosine (Figure 2) was abnormal at 1.9 (normal >2.5), coronary blood flow response to acetylcholine was mildly abnormal at 48% increase (normal is ≥50% increase), coronary artery diameter dilation response to acetylcholine was markedly abnormal at 22% constriction (normal is dilation >0%), and nitroglycerin response was normal at 46% dilation (normal >20%) as depicted in Figures 1 and 2.

Resting echocardiogram in 2005 demonstrated a left ventricular ejection fraction (LVEF) of 55% with basal to mid-septal hypokinesis. From 2005–2010, she had a total of 11 NSTEMI admissions with elevated cardiac troponin I ranging between 0.1 ng/ml in 2008 and 33 ng/ml in 2009. Resting echocardiogram in 2007 demonstrated basal to mid-inferior wall akinesis with hypokinesis of inferolateral wall and interventricular septum with an LVEF of 35–45%. Myocardial biopsy revealed focal fibrosis without vasculitis or inflammation. During the NSTEMI admission prior to CR referral, she had substernal and left sided chest pain which woke her up from sleep. Normal sinus rhythm with inferolateral T wave inversions was shown on ECG, accompanied by an elevated troponin I to 1.72 ng/ml. A diagnosis of ischemic cardiomyopathy and microvascular angina secondary to coronary microvascular dysfunction was made.

At the time of cardiac rehabilitation referral in 2008, cardiovascular risk factor profile revealed a body mass index of 26.3 (overweight), no diabetes, no dyslipidemia, no history of smoking and no family history of premature CAD. She experienced fatigue during ordinary daily

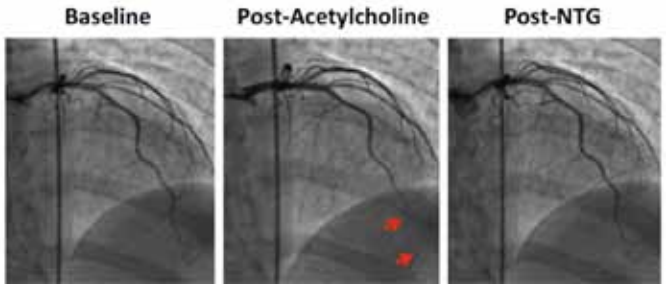

Figure 1: Coronary angiogram and coronary reactivity testing. Coronary reactivity testing is performed with a Doppler wire in the left anterior descending artery. Intracoronary acetylcholine infusion produced vasoconstriction (arrow) of the artery compared to baseline (normal response is dilation), and intracoronary nitroglycerin infusion subsequently dilated the artery with a normal response. Acetylcholine tests endothelial-dependent macrovascular and microvascular function.

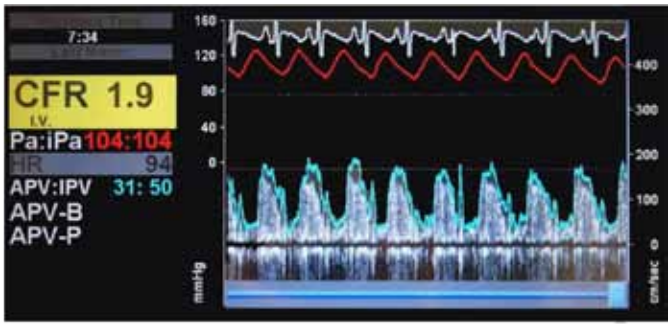

Figure 2: Intracoronary Doppler blood flow velocity waveforms in response to intracoronary adenosine. Coronary flow reserve is the ratio of average peak velocities before and after adenosine administration. Adenosine tests nonendothelial-dependent microvascular function.

activity and shortness of breath during moderate to strenuous physical activity. She did not exercise regularly and had exertional and stress-related angina. No symptoms and signs of heart failure were found. Previously, she was a school teacher and practiced figure skating a few times per week. She was unemployed at the time of CR referral. A Duke Activity Status Index (DASI) questionnaire score was 15.2 corresponding to 4.3 metabolic equivalents (METs) [7]. A general quality of life (QOL) rating was 3 in answer to the question "Overall, how would the patient rate her QOL?" The possible responses fall along an interval scale from 0 (worst) to 10 (best).

During CR, the patient initially had resting blood pressure (BP) of 92/60 mmHg and heart rate (HR) of 86 beats per minute (bpm). Her physical examination was unremarkable. Her CR plan included reconditioning exercises three times a week for 12 weeks, including aerobic and strengthening exercises, as well as nutritional counseling and stress management. She exercised to 2.1 METs for 35 minutes using treadmill and stationary cycling in the first session. Peak HR rose to 111 bpm, BP was 110/62 mmHg and the rating of perceived exertion (RPE) was 11. No arrhythmia on ECG telemetry was noted and no abnormal symptoms were reported. Due to conflicts with childcare, she only attended four CR sessions (Table 1). She increased her exercise tolerance to 45 minutes of treadmill and cycling with the intensity of 3.3 METs. A home CR program with moderate intensity of aerobic exercise such as brisk walk, 30–45 minutes per day for most days of the week was advised because she could not return to a supervised program.

The patient was treated medically with aspirin, dabigatran, metoprolol, ramipril, atorvastatin, nitroglycerin transdermal patch and acetaminophen-codeine as needed for pain. In 2012, she underwent implantable cardioverter-defibrillator placement due to a declined LVEF to 30%. Subsequently, her symptoms were improved, and she continued walking exercise at home. At four-year follow-up, she reported decreased anginal symptoms, improved overall health status, improved functional capacity demonstrated by a DASI questionnaire score of 26.95 (7.7 METs) and a general QOL rating of 5/10.

DISCUSSION

The patient had the general characteristic features of CSX, including angina, evidence of myocardial ischemia and no obstructive CAD by coronary angiography. These findings along with further evaluation and abnormal coronary reactivity testing indicated the specific diagnosis of ischemic cardiomyopathy with microvascular angina secondary to CMD. She was able to increase exercise intensity from 2.1 to 3.3 METs after four sessions of CR program without abnormal chest pain and arrhythmia. While the exercise duration was optimal at 45–50 minutes, low exercise intensity and poor functional capacity were observed. Ideally, a supervised CR program should have been continued but she could not complete a full program of 36 sessions. Home CR program played a key role in this patient, including aerobic exercise, strengthening exercise, stress management technique and healthy heart diet intake.

As a multifaceted structured program that encourages exercise, education, nutrition counseling and psychosocial intervention, CR provides a therapeutic opportunity in CSX and microvascular angina patients (Figure 3) [8]. Eriksson et al revealed that there was improvement in exercise capacity by 34%, delayed onset of chest pain during exercise by 100% and tendency to increased endothelium-dependent blood flow in CSX after eight weeks of aerobic exercise [9]. Moreover, the eight-week CR program was shown to modify cardiovascular risk factors, including lower resting diastolic blood pressure and body mass index, improved physical fitness from Shuttle Walk Test performance, reduced symptom

Table 1: Exercise record.

Mode	Intensity	Duration (min)	METs	HR Range (bpm)	BP Range (mmHg)	RPE
Session 1				Resting HR 86	Resting BP 92/60	
- Treadmill	Speed 1.5 Grade 0%	25	2.1	111	106/60	11
- Bike	Level 1	10	2.1	101	110/62	11
Session 2				Resting HR 95	Resting BP 100/58	
- Treadmill	Speed 3 Grade 0%	40	3.3	114	112/60	10
- Bike	Level 1	10	2.3	108	120/60	11
Session 3				Resting HR 82	Resting BP 94/40	
- Treadmill	Speed 3 Grade 0 %	30	3.3	109	110/58	11
- Elliptical	Level 1	10	5.4	124	116/60	13
Session 4				Resting HR 72	Resting BP 98/60	
- Treadmill	Speed 3 Grade 0%	30	3.3	86	116/60	11
- Bike	Level 2	15	3.3	101	118/58	12

Abbreviations: min=minutes, MET=metabolic equivalent, HR=heart rate, bpm=beats per minute, BP=blood pressure, mmHG=millimeters mercury, RPE=rating of perceived exertion

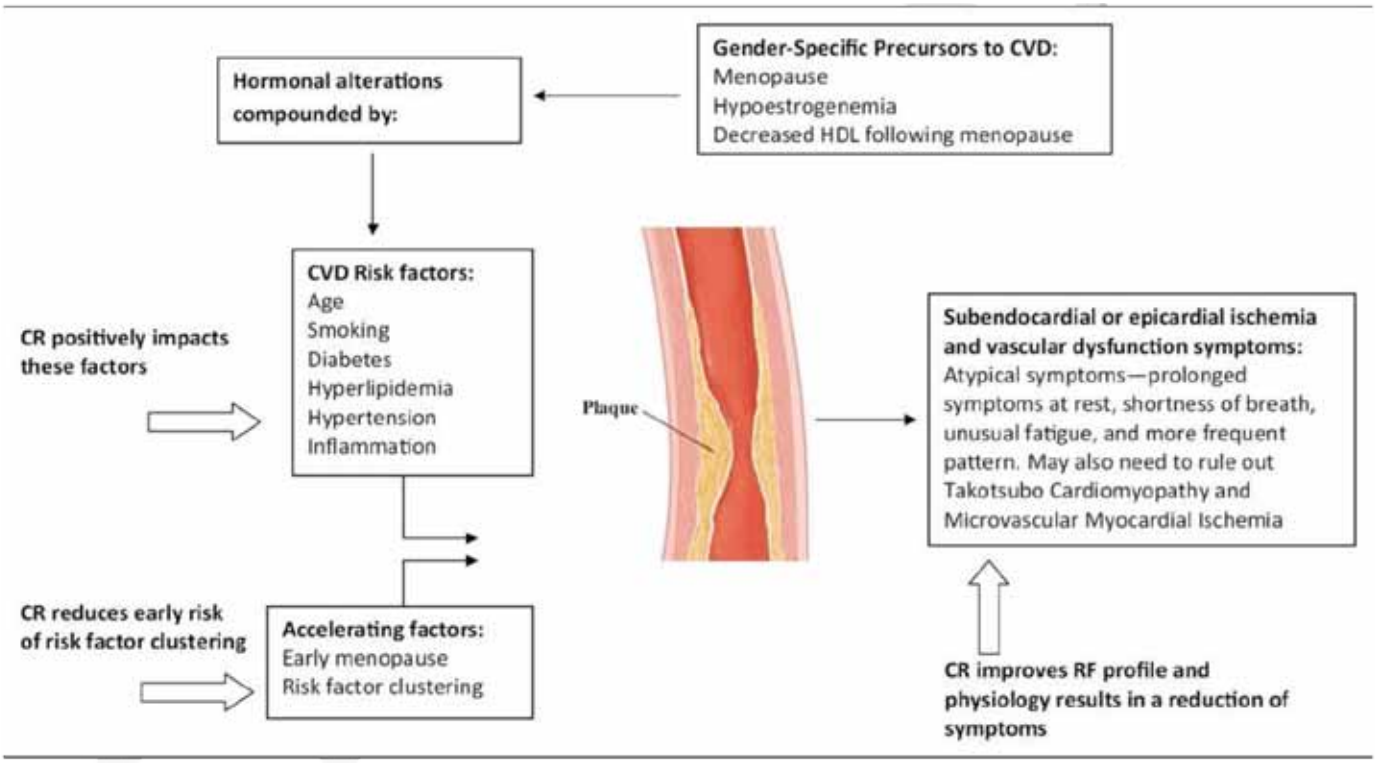

Figure 3: Model of microvascular disease in women. Reprinted with permission from Daniels KM et al. [8].

severity and improved QOL in CSX patients [10]. In microvascular angina patients, Carvalho et al. reported a case with improved endothelium-dependent vasodilator response of brachial artery, reversal of the ischemic myocardial perfusion defect from single-photon emission computed tomography, improved angina symptoms and QOL after the four-month aerobic training [11]. Our report showed the benefits of CR, including increased exercise intensity, exercise duration, improved functional capacity and decreased anginal symptoms in this patient with proven microvascular angina.

Women with CSX often have impaired functional capacity and/or are deconditioned. The DASI is a 12-item self-reported questionnaire that captures functional capacity. The DASI score is validated to estimate maximal oxygen consumption from exercise stress test. By dividing the DASI score by 3.5, METs are derived. The application of DASI might be used instead of exercise stress test to identify at-risk symptomatic women, predict prognosis and target risk management. Among symptomatic women with suspected myocardial ischemia from the NHLBI-WISE study, functional impairment measured by the exercise stress test and DASI-estimated METs ≤4.7 correlated with an adverse prognosis, including higher death and nonfatal myocardial infarction rates [7]. Notably, even small increases in functional capacity after CR participation could reduce cardiovascular mortality and morbidity. In a retrospective review in women referred for stress testing, for every MET increase in exercise capacity, there was a 25% reduction in the rate of all-cause death [12]. Our patient also reported functional capacity improvement, demonstrated by 3.4 METs increase of DASI score at the fourth-year of follow-up, which suggests better prognosis. In terms of QOL, the eight-week CR program improved QOL scales in women with CSX based on a SF-36 questionnaire [10, 13]. Similarly, a slight increase of a general QOL rating scale and improved overall health status was reported in our patient at the four-year follow-up.

Although CR is recommended for all patients with CAD and chronic angina, participation rate is low especially in women [14]. Women are faced with several barriers to CR attendance, including caregiving responsibilities observed in our patient. Referral of women to CR should be strongly considered by physicians, as it positively affects physical and psychological outcomes in patients with CSX, MCD and microvascular angina.

CONCLUSION

This report suggests that cardiac rehabilitation was beneficial in a patient with cardiac syndrome X, cardiac syndrome X, diagnosed with ischemic cardiomyopathy and microvascular angina secondary to microvascular coronary dysfunction. Even though the patient did not complete the cardiac rehabilitation (CR) session plan, CR increased her exercise intensity and duration, improved functional capacity, decreased anginal symptoms and improved quality of life.

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Author Contributions

Wipawee Laksanakorn – 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
Tanaporn Laprattanagul – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Chrisandra Shufelt – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Margo Minissian – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Puja K. Mehta – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
C. Noel Bairey Merz – Substantial contributions to conception and design, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

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Conflict of Interest

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Suggested Reading

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CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

Pneumothorax in dysferlin myopathy associated with mechanical ventilation

Martín Hunter, Patricia Karina Aruj

CASE REPORT

A 38-year-old female with dysferlin myopathy (and domiciliary use of nocturnal non-invasive positive pressure ventilation and mechanical insufflation-exsufflation respiratory aid) presented with progressive right upper quadrant abdominal discomfort. The vital signs at presentation were: temperature 36.6°C, heart rate 100 beats per minute, blood pressure 140/80 mmHg, respiratory rate 30 breaths per minute, oxygen saturation 96% while breathing ambient air. On examination, breath sounds were absent on the right side. An anteroposterior chest radiograph showed a large collection of gas in the right hemithorax with inversion of the right hemidiaphragm (Figure 1A). Computed tomography (CT) scan of the chest confirmed the diagnosis of large right-sided pneumothorax, with cardiomediastinal shift to the left (Figure 1B). Despite the size of the pneumothorax, the patient was hemodynamically stable and, surprisingly, she did not develop hypoxemia. Pneumothorax was managed with tube thoracostomy. However, since lung expansion was not successfully achieved, a Heimlich valve was placed. After discharge, a new CT scan showed resolution of the pneumothorax with adequate right lung expansion. The patient is still using mechanical ventilation through a tracheostomy at home without further complications.

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DISCUSSION

Mechanical insufflation-exsufflation (MI-E) is a respiratory aid used by patients with weak respiratory muscles to improve cough effectiveness. Machine pressures can range from +60 cm H_2O of insufflation to −60 cm H_2O of exsufflation [1]. A few limb-girdle muscular dystrophies are associated with weakness of respiratory or oropharyngeal muscles and an increased risk of respiratory failure with disease progression [2]. This patient had a severe restrictive pattern in pulmonary function tests and diminished cough peak flows from inspiratory and expiratory muscle weakness. The cause of pneumothorax was probably due to barotrauma related to high insufflation pressures.

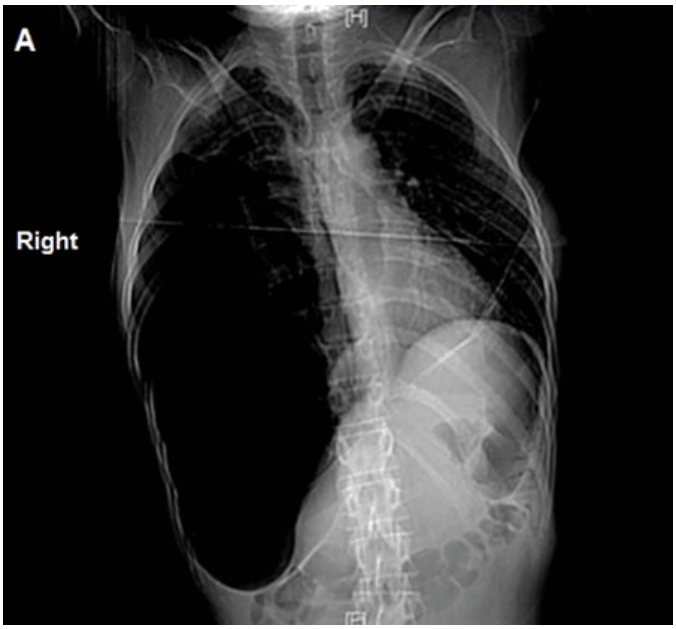

Figure 1A: Anteroposterior chest radiograph showing a large collection of gas in the right hemithorax with inversion of the right hemidiaphragm.

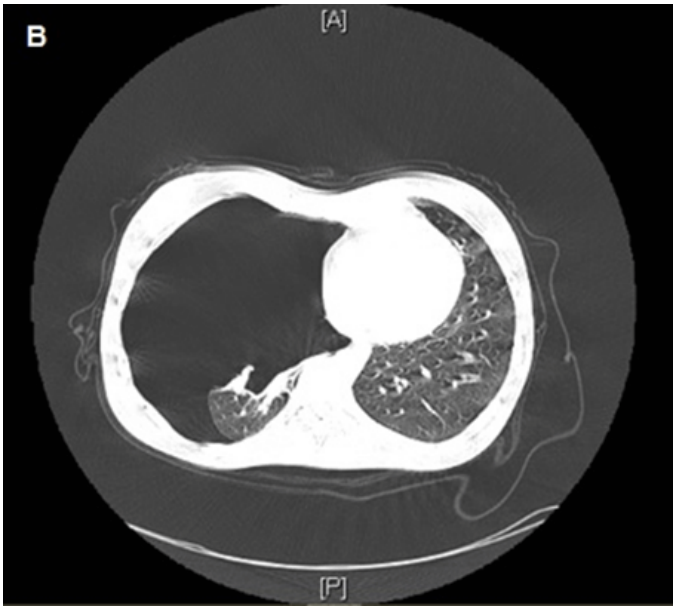

Figure 1B: Computed tomography scan of the chest confirms the diagnosis of large right-sided pneumothorax, with cardiomediastinal shift to the left.

CONCLUSION

Although the use of mechanical insufflation-exsufflation (MI-E) alone may or may not increase the risk of barotrauma, clinicians should be aware that the use of MI-E may add to other risk factors to increase the risk of pneumothorax. Although rare, pneumothorax associated with mechanical ventilation has been reported previously in this patient population.

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Author Contributions

Martín Hunter – Substantial contributions to conception and designn, 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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

Total urethrovesical anastomotic disruption

Han-Kuang Chen, Alicia Helena Mackowski

CASE REPORT

A 68-year-old was male presented to the hospital on day-8 post robot-assisted laparoscopic radical prostatectomy (RALRP) for routine catheter removal and trial of void. He had no other significant medical history and the procedure was performed for Gleason 7 biopsy-proven prostate adenocarcinoma. On admission, he reported feeling unwell for the last two days and passing dark blood-stained urine with debris, but no symptoms of infection were experienced. These symptoms were preceded by an episode of "coughing fits". On examination, the patient was afebrile and his vital signs were within normal range. Cardiopulmonary examination was normal and abdomen was soft and non-tender. The laparoscopic surgical wounds appeared clean and healthy. Pelvic computed tomography scan showed disruption of the anastomosis with the catheter lying within the contrast-filled prostatic cavity (Figure 1).

The patient was returned to operating theatre for Da Vinci Si HD (Intuitive Surgical Inc, USA) robot-assisted reconstruction of urethrovesical anastomosis. Intraoperatively, complete urethrovesical anastomosis disruption was evident (Figure 2), and there appeared to be an anastomotic suture breakage and splitting of the bladder neck. The repair process was difficult due to tissue fragility. The anastomosis was repaired with monofilament suture by van Velthoven style; reinforcements with simple interrupted suture were made at 2 o'clock, 5 o'clock, 7 o'clock and 10 o'clock. Patient recovered well postoperatively but anastomotic leak was persistent. The urinary catheter was kept in for

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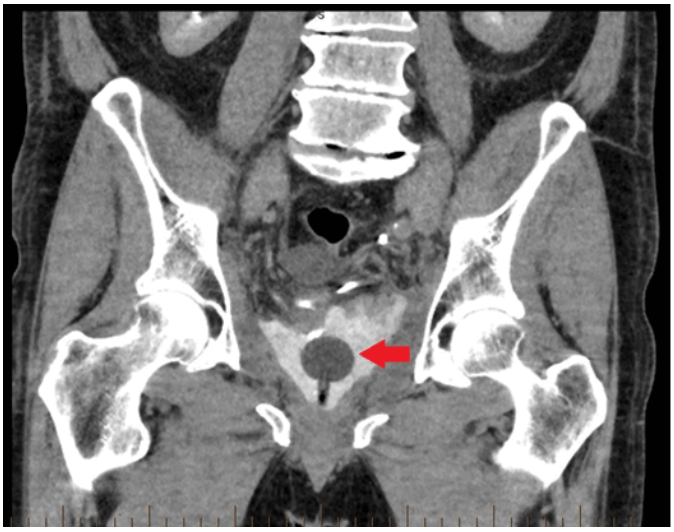

Figure 1: Pelvic computed tomography scan showing disruption of the anastomosis with the catheter lying within the contrast-filled prostatic cavity.

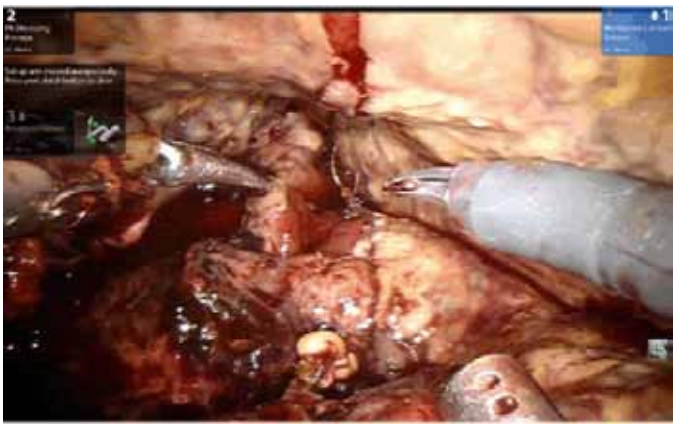

Figure 2: Total urethrovesical anastomotic disruption seen on robotic camera.

four weeks total after which cystogram showed water-tight healing.

DISCUSSION

Urethrovesical anastomotic leak is one of the most common complications after radical prostatectomy with an incidence of 0.3–15.4% [1]. Many factors have been associated with urethrovesical anastomotic leak, such as patient characteristics, surgeon experience and the technical details of the procedure [1]. It is reasonable to assume that disruption of the anastomosis is associated with the same factors.

The RALRP in this case was performed by an experienced surgeon (more than 100 RALRPs performed) on an otherwise healthy 68-year-old male who had no other significant co-morbidity and a healthy BMI of 25. Therefore, it was unlikely that patient characteristics or surgeon experience contributed to this adverse event. In terms of technical details of the anastomosis, we performed posterior reconstruction of the rhabdosphincter with a dissolvable monofilament suture as per Rocco et al. [2] technique after resection of the prostate. The urethrovesical anastomosis was performed using a modified van Velthoven [3] technique with two 20-cm 3-0 Monocryl sutures tied together at the free ends. There was no urethrovesical leak when inflating the bladder with 120 mL of normal saline. Operative time was 140 minutes. Estimated blood loss was 300 mL. There was no immediate complication post-operatively and length of hospital stay was two days.

The coughing fits: reported by the patient was a suspicious factor in this case as the patient's symptoms started after the coughing episode. It is known that coughing increases urethral pressure and it is greater than that generated by voluntary pelvic floor contraction [4]. The intraoperative findings of suture breakage and splitting of the bladder neck also suggested that force was involved in causing the urethrovesical disruption. Therefore, in our opinion, this is the most likely cause for the adverse event in this case. To our knowledge, no other similar cases have been reported.

CONCLUSION

Urethrovesical anastomotic disruption, as oppose to anastomotic leak, is likely associated with the same factors, such as patient characteristics, surgeon experience and the technical details of the procedure. In this case, however, presenting history and intraoperative findings suggested that force was involved in causing the urethrovesical disruption. Therefore, we concluded that the most likely cause for the urethrovesical disruption in this case was the sudden change in urethral pressure related to coughing.

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Author Contributions

Han-Kuang Chen – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Alicia Helena Mackowski – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

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Han-Kuang Chen – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
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Guarantor

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Conflict of Interest

Authors declare no conflict of interest.

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CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

Clavicle chronic recurrent multifocal osteomyelitis: Surgical excision and neo-clavicle

Jorge H. Costa, Tiago P. Marques, Miguel Pádua

CASE REPORT

This is a case of an 11-year-old boy with left clavicle pain with two months of evolution presented to emergency department. No history of trauma, fever or systemic signs, recent illness and no relevant personal or familiar medical history. No neurological or vascular abnormalities were found in the upper arms. Left shoulder X-ray revealed a hyperdense lesion and increased thickness of left clavicle (Figure 1). In this context a computed tomography (CT) scan and after a magnetic resonance imaging (MRI) scan were ordered. Both scans demonstrated morphological changes of the median half of the clavicle, cortical destruction and periosteal reaction. Also changes in the surrounding soft and muscular tissue and bulky and numerous locoregional ganglion formations (Figures 2 and 3). Blood analysis, blood cultures and bone biopsy were negative for infection and neoplastic disorder. In this context, by exclusion, and in the presence of two Jansson major diagnostic criteria (radiologically proven osteolytic/osteosclerotic bone lesions and sterile bone biopsy with signs of inflammation) [1], we reached the diagnosis of chronic recurrent multifocal osteomyelitis (CMRO) (Table 1). Patient was treated with NSAIDs for six months and a short course of corticoids without any response. By keeping complaints of marked and recalcitrant pain in clavicle region, we opted for the surgical treatment: resection of the lesion en bloc, about 7 cm of clavicle (Figure 4), maintaining the integrity of periosteum to allow neo-osteogenesis of the clavicle. No bone grafting or another material was interposed in the dead space left by the resection of the clavicle. Post operatory X-ray, showing a radiopaque area corresponding to the excision of the medial portion of the clavicle (Figure 5).

At third month after surgery, there is an image compatible with a neo-clavicle and patient is asymptomatic (Figure 6).

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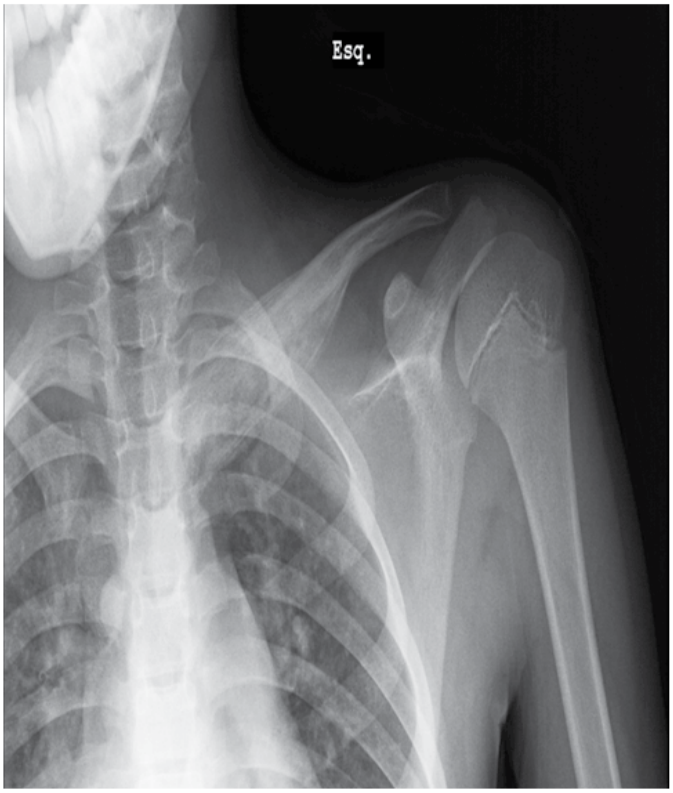

Figure 1: X-ray revealed a hyperdense lesion and increased thickness of left clavicle.

Figure 2: Computed tomography scan morphological changes of the median half of the clavicle (increased thickness) and cortical destruction.

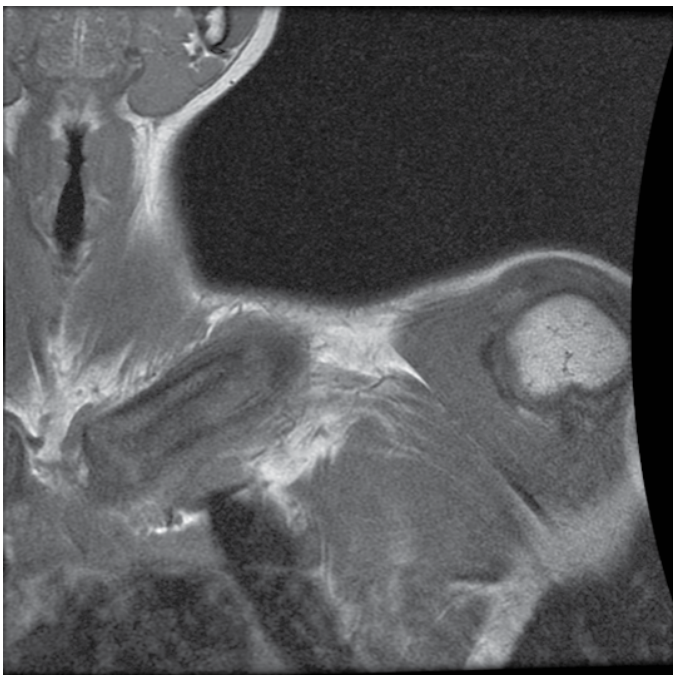

Figure 3: A coronal T1-weighted magnetic resonance imaging (MRI) scan showing an increased thickness of left clavicle and changes in the surrounding soft and muscular tissue and bulky with numerous locoregional ganglion formations.

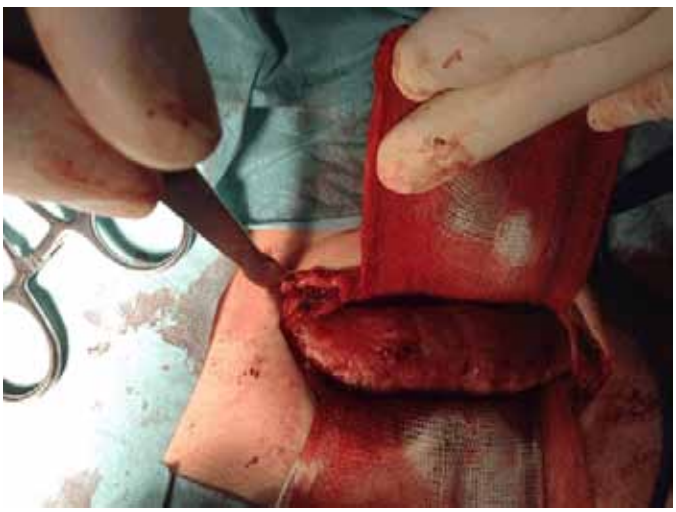

Figure 4: En bloc resection of 7 cm of clavicle.

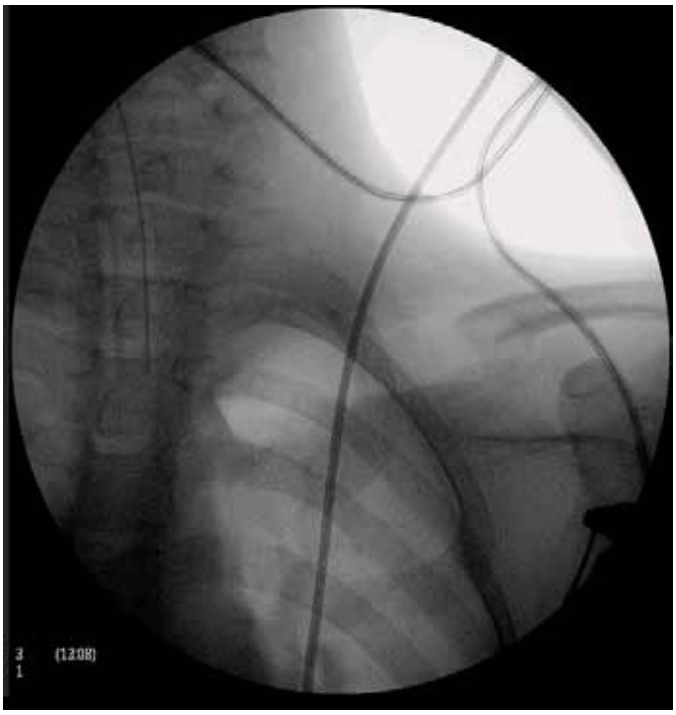

Figure 5: Postoperative X-ray showing a radiopaque area corresponding to the excision of the medial portion of the clavicle

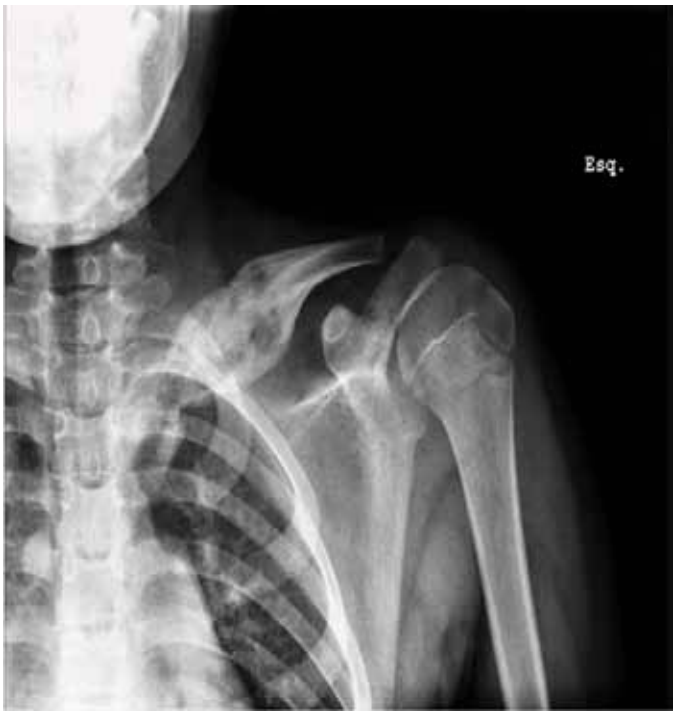

Figure 6: Image compatible with a neo clavicle, three months after surgery.

DISCUSSION

Chronic recurrent multifocal osteomyelitis is a non-bacterial bone inflammation (osteitis), that has symptoms similar to the conventional osteomyelitis, but without infection. It is characterized by multifocal bone

Table 1: Proposed major and minor diagnostic criteria of CRMO*

Major diagnostic criteria	Minor diagnostic criteria
Radiologically proven osteolytic/sclerotic bone lesion	Normal blood count and good general state of health
Multifocal bone lesions	CRP and ESR mildly-to-moderately elevated
Pustulosis palmoplantaris or psoriasis	Observation time longer than 6 months
Sterile bone biopsy with signs of inflammation and/or fibrosis, sclerosis	Hyperostosis
	Associated with other autoimmune diseases apart from pustulosispalmoplantaris or psoriasis
	Grade I or II relatives with autoimmune or autoinflammatory disease or non-bacterial osteitis

*CRMO is confirmed by two major criteria or one major and three minor criteria

lesions with pain and swelling recurring over months to years, accounting of 2 to 5% of all cases of osteomyelitis [2–3] mostly children and adolescents. The differential diagnosis includes bacterial osteomyelitis, Ewing sarcoma, leukemia, lymphoma, rhabdomyosarcoma, neuroblastoma metastasis, eosinophilic granuloma or Langerhans cell histiocytosis. The tubular long bones, pelvis, hip, sternum and mandible are the most affected bones [4–5]. The CRMO, in pediatric patients , may be associated with various skin disorders: Pustulosis palmoplantar is syndrome [6–8], diffuse pustulosis, psoriasis vulgaris, acne [9–11], Sweet's syndrome and pyoderma gangrenosum [9]. Multiple sites of apparent osteomyelitis with negative pathology and cultures and no response to antibiotherapy characterize it [12]. NSAIDs as naproxen, indomethacin or aspirin are the initial therapy. A short course of corticosteroids can be used in refractory cases. Other treatments such as sulfasalazine or methotrexate [6], bisphosphonates [13] and hyperbaric chamber [14] are also indicated in refractory cases. Antibiotics do not seem to be helpful. Although surgical treatment has been used, its role is not yet clearly defined. We opted for a surgical excision of the clavicle, because of the constant and intense pain with no response to six months of conservative treatment. We think that the good response to surgery makes this case exceptionally unusual. The prognosis for these patients is however good. In one study, 17 of 23 patients had complete resolution of the clinical findings, at an average of 5.6 years after diagnosis. Six patients continue to have active disease, and the other six had intermittent relapses or chronic pain. 78% had no physical impairment [2], but further prospective studies are needed to determine the optimum outcome measures and treatment strategy [15],

CONCLUSION

The absence of positive microbiologic results, the clinical course and the presence of reactivating and remitting lesions over time suggest the diagnosis of chronic recurrent multifocal osteomyelitis. As the disease is self-limiting, knowledge of its characteristics appearance can lead to the most appropriated treatment, and can prevent overly aggressive medical and surgical evaluation and treatment.

How to cite this article

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Author Contributions

Jorge H. Costa – 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
Tiago P. Marques – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Miguel Pádua – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

Advanced hepatocellular carcinoma extending to the inferior vena cava and right atrium

S. M. Habib, L. C. W. De Jonge, R. A. Carels, C. Verveer, A. A. M. Zandbergen

CASE REPORT

A 65-year-old Surinamese male visited our outpatient clinic due to fatigue and weight loss of five kilograms in the last three months. The patient was diagnosed with chronic hepatitis B five years ago for which he did not receive treatment. Furthermore, he was a moderate alcohol drinker (1 drink/day) and had a 40 pack-year history of smoking. At physical examination, a cachectic man was seen with a blood pressure of 109/67 mmHg and a pulse rate of 84 beats per minute. He had no signs of jaundice. Physical examination of the abdomen revealed distended superficial abdominal veins and an enlarged liver, but no signs of ascites. Laboratory testing revealed a normal hemoglobin level of 9.9 mmol/L, an elevated bilirubin level of 95 µmol/L, an elevated alpha-fetoprotein level of 36 µg/L, and elevated liver enzymes (alanine aminotransferase 91 U/L, gamma glutamyl transferase 576 U/L, and alkaline phosphatase 245 U/L).

Subsequently, a computed tomography (CT) scan was performed, showing a large mass in the right lobe of the liver extending to the caudal lobe with occluded portal vein branches. Importantly, the mass was found to be invasive, compressing the vena cava inferior, and extending into the right atrium (Figure 1). No sign of liver cirrhosis were seen on computed tomography (CT) scan.

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A magnetic resonance imaging (MRI) scan was performed to establish a final diagnosis. MRI scan confirmed the presence of a 13.5x8.4 cm mass in segment 6 and 7 of the liver with a tumor thrombus invading the inferior vena cava (Figure 2) and right atrium.

DISCUSSION

Hepatocellular carcinoma (HCC) is frequently a fatal malignancy and accounts for the majority of cases

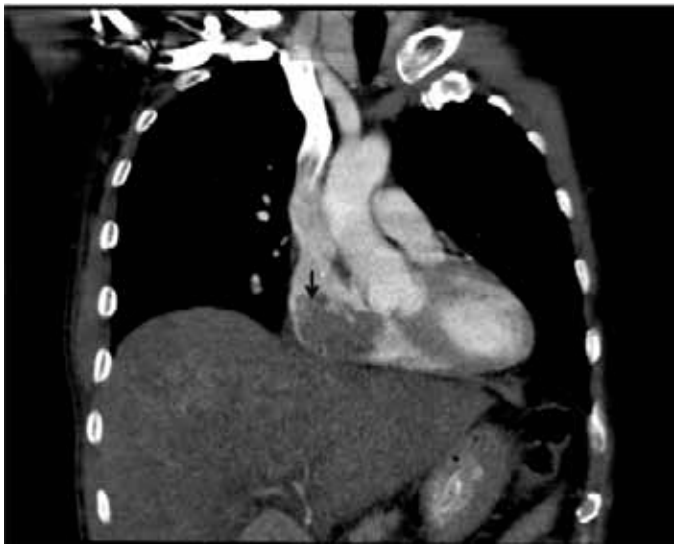

Figure 1: Computed tomography scan showing a mass extending into the right atrium.

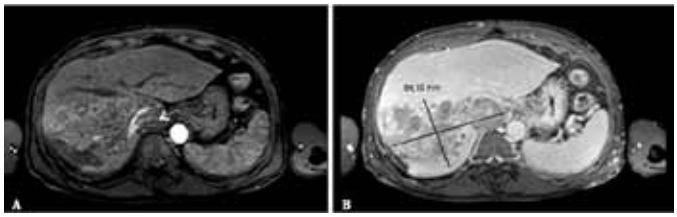

Figure 2: Magnetic resonance imaging scan showing (A) Early arterial enhancement of the tumor mass in the liver invading the vena cava inferior (arrow), and (B) Fast washout of the enhancing lesion.

with primary liver cancer worldwide. The incidence of hepatocellular carcinoma (HCC) is low, however, seems to be as high as 11–20 cases per 100,000 male inhabitants in some countries [1].

Significant risk factors of HCC include chronic hepatitis B virus infection and other chronic liver diseases, usually in combination with cirrhosis [1, 2]. The most common locations of extrahepatic metastases of HCC include the lung, abdominal lymphnodes, and bones [3]. An initial presentation of HCC with tumor thrombus invasion in the vena cava inferior and right atrium is extremely rare (1–4%) and hazardous. An atrial thrombus can cause right heart failure and pulmonary embolism, and most patients die within the first year after diagnosis [4].

Establishing the diagnosis of HCC is a challenge, but preferably must be done based on non-invasive techniques, such as imaging (CT and/or MRI), in most cases without histological confirmation [2]. HCC in our patient was diagnosed based on the presence of the typical radiological criteria including the irregular tumor surface, the early arterial enhancement and the fast washout of tumor areas on the dynamic contrast-enhanced MRI scan.

The management of HCC requires a multidisciplinary approach and is closely linked to the stage of disease. Treatment options include radiofrequency ablation, partial liver resection, liver transplantation, systemic medical treatment (e.g., Sorafenib), and transcatheter arterial chemoembolization [2]. Given the limited treatment options and incurability of HCC with an atrial tumor, our patient was discharged from the hospital after one week with supportive care.

CONCLUSION

In conclusion, this case illustrates an uncommon presentation of advanced hepatocellular carcinoma (stage IIIC), which could be successfully diagnosed non-invasively by radiological imaging.

How to cite this article

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Author Contributions

S. M. Habib – 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

L. C. W. De Jonge – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

R. A. Carels – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

Extensive pericardial thickening without constriction

Salahaddin Ubaid

CASE REPORT

A 73-year-old male presented with atypical chest pain. He had no significant past medical history of note and he was otherwise fit and well and not on prescribed medications. He was an ex-smoker and had no family history of cardiovascular diseases. Physical examination was unremarkable and revealed a normal blood pressure, non-elevated jugular venous pressure, normal heart sounds and no other significant abnormal findings. Laboratory investigations showed normal blood count and normal kidney and liver function tests.

In view of his presentation, age and history of smoking he underwent a coronary angiography procedure to exclude coronary artery disease. Unexpectedly, it showed a grey shadow around the heart mainly on the right side (Figure 1). Pericardial thickening and possible constrictive pericarditis was suspected, subsequently, a right heart catheterization was performed and intracardiac pressures were assessed. Intracardiac hemodynamics were entirely normal (right atrial pressure 3 mmHg, right ventricular and pulmonary artery systolic pressures 20 mmHg, mean pulmonary capillary wedge pressure 8 mmHg and left ventricular end diastolic pressure was 12 mmHg).

Transthoracic echocardiography suspected the presence of pericardial thickening. Otherwise there was no evidence of other structural heart abnormalities. A computed tomography scan of the thorax confirmed it to be pericardial thickening. No particular cause for the pericardial thickening was identified (idiopathic). He remained well and is under regular twelve monthly cardiology follow-up.

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DISCUSSION

Pericardial constriction happen when a thickened, and frequently calcified pericardium impairs cardiac filling, limiting the total cardiac volume [1, 2, 3].

The most common causes include mediastinal radiation such as for Hodgkin's lymphoma, chronic

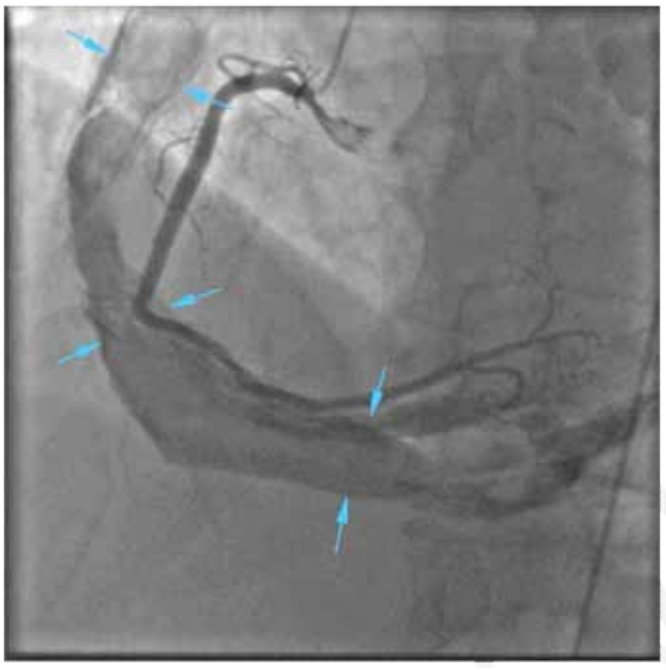

Figure 1: A coronary angiogram in left anterior oblique view during cardiac catheterization showing extensive pericardial thickening. Arrows pointing to the pericardial thickening. Cardiac hemodynamics were completely normal. The right coronary artery is also shown.

idiopathic pericarditis, post-cardiac surgery and tuberculous pericarditis [1, 4–6]. These conditions tend to cause chronic pericardial inflammation causing pericardial scarring with thickening, fibrosis, and calcification [3].

The pathophysiological hallmark of pericardial constriction is equalization of the end-diastolic pressures in all four cardiac chambers; as a result systemic congestion is much more marked than pulmonary congestion. Typical presentations are due to elevated systemic venous pressures and low cardiac output [5].

The increased systemic venous pressure lead to marked jugular venous distension, hepatic congestion, ascites, and peripheral oedema, while the lungs remain clear. Low cardiac output causes exercise intolerance that could progress to cardiac cachexia and muscle wasting.

Transthoracic echocardiography is an essential diagnostic test in patients being evaluated for constrictive pericarditis.

Computed tomography scan of the heart is useful in the diagnosis of constrictive pericarditis and can provide additional data to guide perioperative management decisions.

Gated cardiac magnetic resonance imaging provides direct visualization of the normal pericardium, which is composed of fibrous tissue and has a low MRI signal intensity [7]. CMR is advocated by some as the diagnostic procedure of choice for the detection of certain pericardial diseases, including constrictive pericarditis [8–10]. Characteristic CMR features in patients with constrictive pericarditis include increased pericardial thickening and dilatation of the inferior vena cava, an indirect sign of impaired right ventricular diastolic filling.

Pericardiectomy is the only definitive treatment option for patients with chronic symptomatic constrictive pericarditis.

Most patients with pericardial constriction have a thickened pericardium (>2 mm) [1, 3, 11]. It is important to recognize, however, that pericardial constriction can be present without pericardial calcium and, in some cases, even without pericardial thickening. In a series of 143 patients from the Mayo Clinic with surgically proven pericardial constriction, 26 (18%) had a normal pericardial thickness (<2 mm) [12].

So it is possible to have constrictive pericarditis without pericardial thickening, but it is not common to have pericardial thickening without constriction.

Our case demonstrates that in spite of extensive pericardial thickening, our patient was asymptomatic with no hemodynamic evidence of constriction during cardiac catheterization which is an unusual occurrence

CONCLUSION

Not all patients with pericardial constriction have pericardial thickening and not all patients with pericardial thickening have pericardial constriction.

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Conflict of Interest

Authors declare no conflict of interest.

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