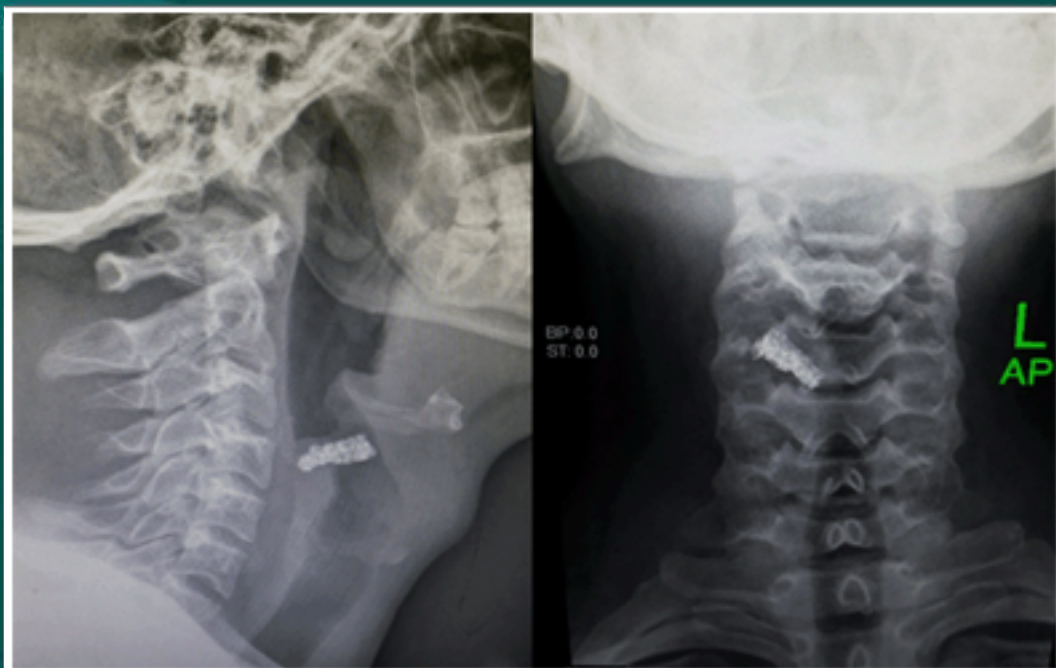


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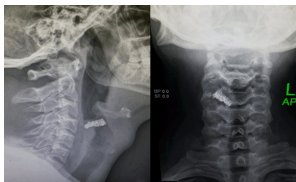
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Cover Image



Lateral plain X-ray of the neck showing radio-opaque steel foreign body in the level of 4th cervical vertebra, and Anteroposterior view of the same patient.

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Neonatal Gastric Perforation: A case series

Aishwarya Venkataraman, Anastasia Vareli, Devesh Misra,
Bhupinder Reel, Kalaimaran Sadasivam

ABSTRACT

Introduction: Gastric perforation in neonates is a rare surgical emergency of uncertain etiology. **Case series:** We report a case series of three neonates diagnosed with gastric perforations in the first week of life and successfully managed by urgent surgical intervention. All three patients had associated gastrointestinal anomalies contributing to the gastric perforation. **Conclusion:** Neonatal gastric perforation is extremely rare and is associated with high mortality. Early diagnosis and prompt surgical intervention are essential to improve the outcomes.

Keywords: Gastric perforation, Pneumoperitoneum, Neonate, Gastrointestinal anomalies

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INTRODUCTION

Neonatal gastric perforation is a rare and catastrophic condition with high mortality. Various factors and theories have been proposed as a possible cause [1–7] but the etiology still remains obscure. Gastric perforations are often large and associated with necrosis of a significant portion of the stomach wall. Early identification and treatment is essential and may improve the outcome. It is, therefore, imperative to highlight the need for increased awareness of NGP so as to optimize outcome through urgent surgical intervention in a timely manner. We hereby describe three cases of NGP, secondary to increased gastric pressure due to distal obstruction, managed successfully with prompt surgical intervention.

CASE SERIES

Case 1

A two-day-old term male neonate, born by elective c-section for breech, presented with a history of vomiting, abdominal distension and grunting. Antenatal period and scans were uneventful apart from polyhydramnios noted at 37 weeks gestation. Feeding was commenced immediately after birth and was well tolerated until 40 hr of life when he developed abdominal distension, vomiting, grunting and remained unsettled. Abdominal X-ray revealed free air in the abdomen (Figure 1). On exploration, undigested milk was found all throughout the peritoneal cavity. A gastric perforation of 5 cm was seen on the anterior wall along the greater curvature higher up. This area was excised coming up to 2 cm of the gastroesophageal junction. No other obvious

gastrointestinal abnormality was seen. Primary repair was done in three layers and abdomen was closed. Histopathology revealed extensive hemorrhagic necrosis of the mucosa and submucosa associated with severe congestion. A contrast study performed postoperatively to evaluate the gastrointestinal tract revealed narrowing at pylorus suggesting pyloric atresia. Hence, a second laparotomy with pyloroplasty was performed to excise the pyloric membrane. Enteral feeding was commenced seven days after the 2nd surgery. He was discharged home after four weeks with anti-reflux medications and full enteral feeds.

Case 2

Twin female neonates (twin 1 and twin 2) were born by elective c-section at 36+3 weeks in good condition. The antenatal period was uneventful and they were monochorionic diamniotic (MCDA) twins. Both of them were discharged at 36 hr of age after enteral feeding was well established. However, they presented on day-4 of life with vomiting, lethargy, distended abdomen and metabolic acidosis. Abdominal X-ray of both babies' revealed free air in the abdomen and Abdominal X-ray of twin 2 showed classical 'football' signs of perforation (Figure 2). On exploratory laparotomy of twin 1, a large volume of a grossly contaminated peritoneal fluid containing undigested foul smelling milk was found. A 5-mm gastric perforation on the greater curvature near the esophagogastric junction and a malrotated bowel with severe narrowing of the root of mesentery was noted. Ladd's procedure and appendectomy were performed. Primary repair was done in three layers and abdomen was closed. Laparotomy findings of twin 2 were similar to twin 1 but she had two gastric perforations on the greater curvature along with severe malrotation. Ladd's procedure along with appendectomy was performed. Primary repair was done in three layers and abdomen was closed. There was no associated volvulus in both the babies and the postoperative period was uneventful. Histology of debrided tissues showed mucosal necrosis and hemorrhage. They received TPN for three weeks and triple antibiotics (metronidazole, amikacin and co-amoxiclav) for five days. Enteral feed was started 14 days after the surgery and slowly increased over one week.

DISCUSSION

Neonatal gastric perforation is extremely rare and associated with poor prognosis [1, 2, 4]. Various theories have been proposed describing the etiology and prognosis but it still remains obscure [1–3, 5]. Congenital absence of gastric musculature [6, 8], high gastric acid production [9], abdominal trauma [7, 10], and other associated gastrointestinal conditions like ischemic bowel, necrotising enterocolitis (NEC), intestinal malrotation, duodenal web, hiatus hernia, Meckel's diverticulum,

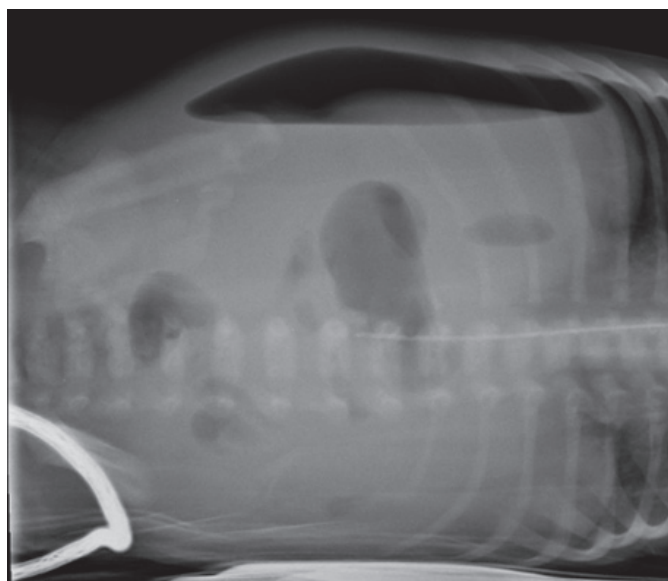


Figure 1: Lateral X-ray of chest/abdomen confirming free gas in abdominal cavity (Case 1).



Figure 2: Abdominal X-ray showing pneumoperitoneum with football sign (Case 2).

and gastroschisis have all been proposed as possible causes of NGP [3, 11–14]. Few authors have reported that gastric perforation was seen in the setting of a distal mechanical obstruction [3, 5, 15]. Shaw et al. [15] through their experiments suggested that gastric perforation was caused by a mechanical rupture of the stomach secondary

to increased gastric pressure. In our series, the presence of pyloric web in one neonate and intestinal malrotation in the twins further supports this hypothesis. All three neonates in this study, as well as those previously reported [1, 3, 16] initially tolerated the feeds well and presented with gastric perforation after few days supporting the theory that raised intragastric pressure may contribute to perforation. Irrespective of the cause, neonatal gastric perforation most commonly occurs in the first week of life [3, 17–20] consistent to that observed in our series. Although predominantly seen in preterm and low birth weight newborns [2, 4, 21], neonatal gastric perforation can occur in healthy term infants [1–3] as seen in our series.

Early diagnosis of neonatal gastric perforation is often difficult due to the fact that the presentation and symptoms are non-specific and can mimic sepsis, respiratory distress, poor feeding, NEC, intestinal obstruction, and pneumoperitoneum without gastrointestinal perforation [3, 22]. The majority of neonates are normal at birth, feeding and passing stools normally until rupture occurs when the baby deteriorates rapidly [15, 22]. Abdominal distension can be striking and infants may also develop rapidly progressive pneumoperitoneum with associated cardiopulmonary compromise. Most of the infants are critically unwell on presentation needing intensive care support both pre and post operatively.

Neonatal gastric perforation is a serious and life-threatening condition, hence prompt and urgent surgical exploration is crucial. The time between symptoms and surgery is also a prognostic factor for survival [17]. Broad-spectrum antibiotics are essential to prevent mortality due to peritonitis and sepsis. Parenteral nutrition during the initial postoperative period is usually required. Despite the availability of the advanced neonatal intensive care facilities and parenteral nutrition, the mortality rate remains high (30–83%) [3, 16, 22–24]. Studies have reported that male sex [1, 16], hyponatremia (serum sodium < 130 mEq/L)[16], metabolic acidosis (pH < 7.3) [16], persistent leucopenia and thrombocytopenia [3], prematurity and low birth weight [1, 2, 4] are associated with poor prognosis. Fortunately, there was no mortality in our study. This could be attributed to early diagnosis and prompt surgical intervention.

CONCLUSION

In summary, neonatal gastric perforation is extremely rare and is associated with high mortality. Early diagnosis and prompt surgical intervention are essential to improve the outcome. Although the etiology of neonatal gastric perforation remains unclear, distal mechanical obstruction leading to increased gastric pressure appears to be a contributing factor as seen in our study.

Author Contributions

Aishwarya Venkataraman – 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

Anastasia Vareli – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

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

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

Kalaimaran Sadasivam – 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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Mesenchymal chondrosarcoma: An unusual lump in posterior maxilla

S. Ravi Raja Kumar, Anuradha A., Namineni Kiran Kumar, Horatti Puneeth Kuberappa, Venkata Raju K., Mahanthi Vijayalakshmi

ABSTRACT

Introduction: Mesenchymal chondrosarcoma is an uncommon, slow growing malignant tumor which is a rare variant of chondrosarcoma, having a predilection for the maxillofacial skeleton; less often involves the soft tissue sites in head and neck. **Case Report:** We report an unusual case of mesenchymal chondrosarcoma on posterior palate in a 45-year-old male, who was previously diagnosed as pleomorphic adenoma in private clinic. **Conclusion:** Mesenchymal chondrosarcoma shows varied clinical and radiographic features, with occurrence of this lesion in unusual locations like posterior region of jaws which may lead to error in clinical diagnosis and early treatment

as it has high affinity for recurrence and delayed metastasis.

Keywords: Cartilage tumor, Mesenchymal chondrosarcoma, Posterior maxilla, Recurrent lesion

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INTRODUCTION

Mesenchymal chondrosarcoma is an uncommon, slow growing malignant tumor which is a rare variant of chondrosarcoma, having a predilection for the maxillofacial skeleton; less often involves the soft tissue sites in head and neck. They usually occur in middle aged individuals, but are rare in young patients, with predilection for anterior portion of maxilla [1]. The tumor is unique due to high tendency for late recurrence and delayed metastasis to lung, bone, and lymph nodes [2]. Histopathologically, it is characterized by a biphasic pattern consisting of hyaline cartilage mixed with undifferentiated pleomorphic mesenchymal cells [1, 3]. To the best of our knowledge, only 17 cases have been reported in English literature affecting posterior maxillary region. Thus we report an unusual case of mesenchymal chondrosarcoma on posterior palate in a 45-year-old male.

CASE REPORT

A 45-year-old male presented with painless swelling in the left palatal region present since four months. Patient gave a history of an excision of lesion in posterior left palatal region nine months ago, which was diagnosed as pleomorphic adenoma in a private clinic. In duration of four months, patient again noticed a swelling in the site of previous excision. On clinical examination a well-defined solitary swelling of size approximately 3x1.5 cm extending from distal aspect of 21 to distal aspect of 26 on the left buccal alveolar ridge anteroposteriorly and mediolaterally extending from palatine gingiva to mid palatine raphe in relation to 22–26. The surface appears to be lobulated and mucosa over the growth appeared normal (Figure 1A–B).

Computed tomography scan and orthopantomogram showed no definite changes, whereas paranasal sinus (PNS) view showed haziness in maxillary sinus area. Magnetic resonance imaging (MRI) scan and positron emission tomography (PET) scan revealed a well-defined mass seen on the left side of the buccal vestibule extending into the maxillary sinus (Figure 2A–B). Based on clinical and radiographic features provisional diagnosis of recurrent pleomorphic adenoma was given.

The patient was advised to undergo surgery, and the tumor was resected by subtotal maxillectomy under general anesthesia. The excised specimen was sent for histopathological examination which revealed focal ulceration of the lining epithelium. The stroma revealed biphasic pattern (Figure 3A) with areas of relatively mature cartilage formation with focal areas showing calcification of mature cartilage and areas of primitive spindle to round shaped mesenchymal cells. Primitive appearing mesenchymal cells had scanty cytoplasm, eccentric placed irregular vesicular nuclei and exhibited moderate pleomorphism with variable mitotic activity (Figure 3B). Focally hemangiopericytoma like vascular pattern was evident (Figure 3C). Transitional zone between the chondroid foci and the mesenchymal component was more plodding but not sharp. Immunohistochemical study was done and the proliferating chondrocytes showed nuclear positivity with S100 protein (Figure 4A) and were negative for pancytokeratin (Figure 4B).

Correlating with the clinical, radiographical, histopathological and immunohistochemical findings the

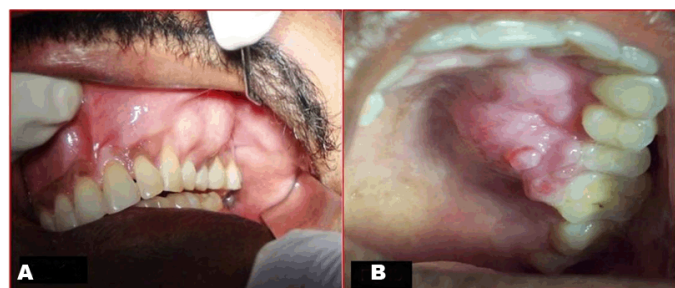


Figure 1: (A, B) Intraoral photomicrograph showing lobulated growth on left buccal and palatal region.

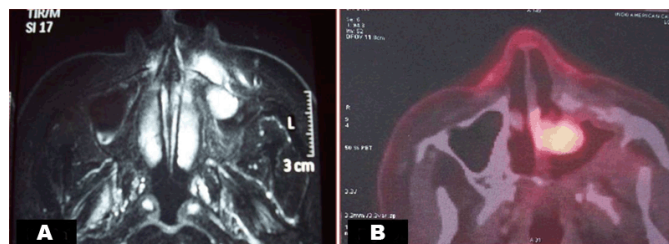


Figure 2: (A, B) Magnetic resonance imaging scan and positron emission tomography scan showing a well-defined radiodense mass on the left side of the buccal vestibule extending into the maxillary sinus.

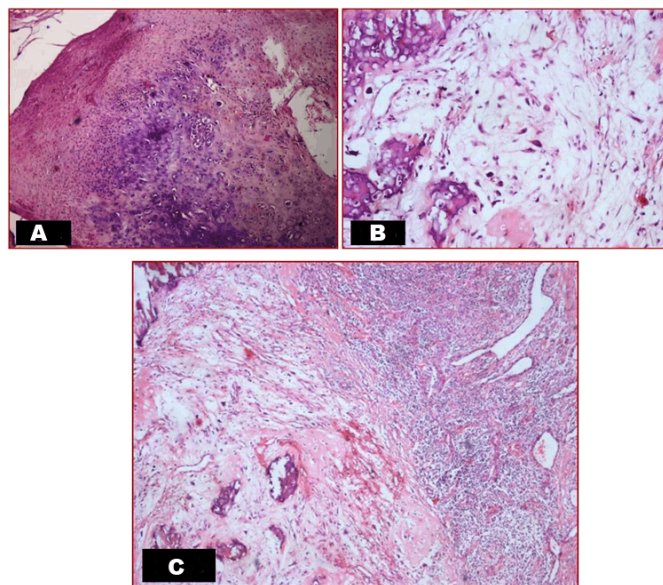


Figure 3: (A) Biomorphic patterns of islands of cartilage surrounded by primitive mesenchymal cellular areas (H&E stain, x200), (B) Primitive spindle to oval shaped pleomorphic mesenchymal cells admixed with calcified chondroid areas with atypical chondrocytes (H&E stain, x200), and (C) Mesenchymal cells arranged around blood vessels (H&E stain, x200).

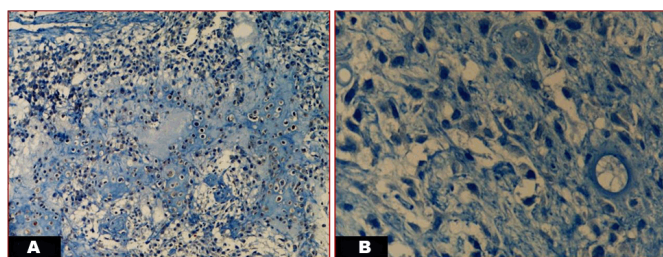


Figure 4: Photomicrograph of IHC stain showing (A) S-100 positive proliferative chondrocytes, and (B) Negative stained for pancytokeratin.

case was diagnosed as mesenchymal chondrosarcoma. The present case was followed for a period of one year with no postoperative recurrence.

DISCUSSION

Chondrosarcoma is the third most common primary malignancy of bone after myeloma and osteosarcoma [4].

Mesenchymal chondrosarcomas are rare biphasic tumor with areas comprising spindle cell mesenchyme interspersed with areas of chondroid differentiation accounting for only 1% of all chondrosarcomas [5].

Mesenchymal chondrosarcomas may develop from pluripotent mesenchymal stem cells and can differentiate into angioblastic, fibroblastic or cartilaginous structures [3], but precise pathogenesis and biological behavior is not fully understood in head and neck mesenchymal chondrosarcoma. But recent studies have shown the identification of new gene HEY1-NCOA2 fusion which appears to be diagnostic, but missing in other subtype of chondrosarcoma [6].

Mesenchymal chondrosarcomas arise from soft tissue or bone in the ratio of 1:2 to 1:6 [3]. In head and neck region commonly involved extra skeletal sites are orbit, meninges and sinonasal tract, whereas intraskeletally anterior maxilla is the most common site [3], where preexisting nasal cartilage is present [6, 7], others sites in descending order of frequency of involvement are body of the mandible, the ramus, the nasal septum and paranasal sinuses [8]. The present case was reported in left posterior maxillary region involving both buccal and palatal areas.

Most of chondrosarcoma occur in 3rd–6th decade of life, where as mesenchymal chondrosarcoma variant occurs in younger age group i.e., 2nd–3rd decade of life in 70% of cases, but few cases have been reported between 5–7th decade [5, 8], where present case also falls in 5th decade. Concerning sex prevalence there some disagreement, as few authors' state male predominance and few state female predominance [1, 8].

Mesenchymal chondrosarcomas show no specific clinical signs and symptoms. The predominant symptom is usually a painless mass or swelling (53%) [5], as in the present case. However, other reported symptoms are nasal obstruction (32%), epistaxis (32%) tooth mobility (24%) [6] and rarely, lymphadenopathy and neurological disturbances such as facial paresthesia and lip paresis can also occur [1, 3].

The radiographic appearance of mesenchymal chondrosarcoma of jaws are not classic, usually exhibit features of a malignancy, consisting of osteolytic process with poorly defined borders. The ill-defined radiolucent area contains scattered foci of neoplastic cartilaginous tissue [3, 8]. The lesion may also cause symmetrical widening of the periodontal ligament space of involved teeth [8, 9], but was not evident in the present case.

Histologically, depending on the cellularity, nuclear staining (hyperchromasia) of the tumor cells and size of the nuclei, chondrosarcomas have been graded into low, intermediate and high grade [5]. Previously, the same case was diagnosed as pleomorphic adenoma due to the presence of chondroid area in the background of myxomatous stroma and also due to inadequate biopsy taken initially might have missed the characteristic areas of mesenchymal chondrosarcoma.

Histopathologically, mesenchymal chondrosarcoma shows characteristic biphasic pattern. Highly cellular undifferentiated/primitive spindle cells is similar to small cell tumor, but presence of islands of chondroid differentiation help in making proper diagnosis, but difficultly arises with small biopsy sample [6, 8]. Most of the mesenchymal chondrosarcomas show rich vascular component, as seen in the present case, often confused with hemangiopericytoma but hemangiopericytoma lacks hyaline cartilage component, additionally, it is positive for CD34.

Histologically, the lesion should be differentiated from small cell osteosarcoma and PNET/ Ewing sarcoma, malignant peripheral neuroectodermal tumor, synovial sarcoma (poorly differentiated type), dedifferentiated chondrosarcoma [10, 11].

Ewing's sarcoma usually contains moderate amount of cytoplasmic glycogen and presence of t(11;22) but lacks cartilaginous component, reticulin meshwork and vascular pattern of mesenchymal chondrosarcoma. Malignant peripheral neuroectodermal tumor contains more pleomorphic small cells than in mesenchymal chondrosarcoma and presence of rosettes. Tumors cells of mesenchymal chondrosarcoma and malignant peripheral neuroectodermal tumors are positive for CD99 and S100 but stromal cells of mesenchymal chondrosarcomas are negative for S100. Small cell osteosarcoma can be differentiated from mesenchymal chondrosarcoma by the presence of osteoid in lace like pattern. Though mesenchymal chondrosarcoma contains bone, it is not produced directly by the stromal cells but formed by endochondral ossification of cartilage islands [3, 11]. Immunohistochemically, the small cell components are positive for vimentin, CD99, and Leu7 but not for S-100 protein; latter is found instead in chondroid areas [12].

Dedifferentiated chondrosarcoma shows large and highly pleomorphic spindle cells with atypical nuclei in contrast to uniform nuclei of small cells of mesenchymal chondrosarcoma. In dedifferentiated chondrosarcoma, there is sharp margin between both components but in mesenchymal chondrosarcoma both are admixed. Monophasic synovial sarcoma (poorly differentiated type) can be separated from mesenchymal chondrosarcoma by the absence of hyaline cartilage. Absences of keratins, especially CK7, and EMA and t(x;18) can differentiate mesenchymal chondrosarcoma from synovial sarcoma [11]. Hemangiopericytoma is distinguished from mesenchymal chondrosarcoma by its lack of cartilage and positive immunohistochemical staining for CD34 [5]. The characteristic expression of type II collagen in matrix of mesenchymal chondrosarcoma helps in differentiation from other small cell sarcomas [6].

In relation to the histological diagnosis, immunohistochemical analysis is of great helpful. Immunohistochemistry is advised for difficult cases. Chondroid areas are positive for S-100 protein and vimentin [3, 5].

Wide surgical excision is the mainstay treatment for chondrosarcoma in the jaw bones [2]. These tumors are radio resistant, so chemotherapy can be used as an adjuvant therapy after wide surgical excision [7]. Our case was treated with wide surgical excision and followed up for three years with no recurrence.

The prognosis of the chondrosarcoma of the jaws is poor as compared to that of long bones. The cause of death is usually by direct extension of tumor into the base of skull, and also through distant metastasis, primarily to lungs and bones. The prognosis is good for low and intermediate grade chondrosarcomas. Especially, the maxillary (0.7% of whole body tumor) and mandibular locations of the tumor have documented inferior prognosis [12].

Fu and Perzin in 1974 described three prognostic factors: location and extent of the lesion, adequacy of surgical therapy, and degree of differentiation of the tumor. Few studies have shown five-year survival rate up to 40–60% and few showed recurrence even after 10–20 years [1]. Hence adequate treatment and a lifelong follow-up after surgery is recommended for patients with mesenchymal chondrosarcomas of the maxillofacial region.

CONCLUSION

Mesenchymal chondrosarcoma shows varied clinical and radiographic features, with occurrence of this lesion in unusual locations like posterior region of jaw which shows ill-defined feature and ossification which may lead to error in clinical and histopathological diagnosis and delay in early treatment. It is an aggressive neoplasm with high penchant for recurrence and delayed metastasis. Hence, patient should be kept under long-term follow-up.

Author Contributions

S. Ravi Raja Kumar – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Anuradha A. – 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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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Recurrent pericarditis as the presenting symptom for diagnosis of systemic lupus erythematosus

Gurneet Matharoo, William Tyler Whitmire, Sam Sirotnikov, Manoj Jagtiani, Niket Sonpal

ABSTRACT

Introduction: Systemic lupus erythematosus (SLE) is a chronic inflammatory autoimmune disease that affects multiple organs. Common findings at presentation are fatigue, fever, weight loss, arthritis/arthralgias, skin manifestations, and renal pathologies. Herein, we focus on pericarditis as a less frequent although significant sign leading to the diagnosis of SLE. **Case Report:** A case of an African-American female with pericarditis at the time of diagnosis. The patient was a 28-year-old female who presented to the emergency department complaining of chest pain and shortness of breath. She reported experiencing similar symptoms on two separate occasions prior to this admission. After appropriate workup an ANA panel was ordered on suspicion which revealed pertinent findings such as an ANA titer of 1:1280, anti-DNA(ds) 10 IU/mL, RNP antibodies >8 AI, Smith antibodies 2.2 AI. **Conclusion:** Diagnosing SLE can be considered challenging, since there are varying degrees of clinical manifestations from patient to patient. The lifetime prevalence of some cardiac manifestation in SLE is estimated

to be 50%, and should be high on the differential of any presentation with chest pain or shortness of breath. It is crucial that physicians consider SLE as a diagnosis when new onset pericarditis occurs in African-American females.

Keywords: Autoimmune, Criteria, Diagnosis, Pericarditis, Systemic lupus erythematosus (SLE)

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INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic inflammatory autoimmune disease that affects multiple organs. Common findings at presentation are fatigue, fever, weight loss, arthritis/arthralgias, skin manifestations, and renal pathologies. Herein, we focus on pericarditis as a less frequent although significant sign leading to the diagnosis of SLE. This disease targets an estimated one and a half million Americans, and over five million worldwide. Sixty-three percent of these patients report being incorrectly diagnosed, with more than half the patients visiting four or more doctors before correct diagnoses [1]. Pericardial involvement many times precedes clinical manifestations of SLE. In SLE specifically, pericardial involvement concurrently

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presenting with pericardial effusion is the most common type of echocardiographic abnormality found in >50% of adult patients, suggesting a significant reason why a patient presenting with the latter should be screened for SLE. In addition, combined autopsy series revealed pericardial involvement in 62% of patients with SLE [2]. It is critical to consider pericarditis as an important presenting symptom of SLE, since early recognition and management will reduce morbidity and mortality.

CASE REPORT

In this report, we describe a case of an African-American female with pericarditis at the time of diagnosis. A 28-year-old female presented to the emergency department complaining of chest pain and shortness of breath. Upon admission to our hospital she described her chest pain as 6–7 out of 10 with radiation to the scapula. The pain was aggravated by movement and inspiration and alleviated by leaning forward. She denied fever, nausea, vomiting, rash or joint pain. Family history was significant for a mother with SLE. On initial examination, the patient had temperature 98.3°F, blood pressure 128/78 mmHg, respiratory rate 20 breaths per minute, and heart rate 95 beats per minute. Normal S1 and S2 heart sounds were heard on auscultation with a regular rate and rhythm. No murmurs, rubs, or gallops were heard. Decreased breath sounds were noted bilaterally at lung bases.

The patient reported experiencing similar symptoms on two separate occasions. The first episode occurred one month prior to the current admission, where she was treated with NSAIDs. Presuming a classic case of pericarditis, the patient was discharged from the hospital. A second episode with identical chest pain occurred three weeks prior to admission, which she sought help from a different hospital than the former. At that time, an echocardiogram was performed revealing a pericardial effusion. NSAIDs were given and a rheumatology follow-up was advised.

Pertinent laboratory findings which lead to the diagnosis in this admission included a comprehensive ANA panel; ANA screen positive with anti-nuclear antibody titer of 1:1280 (<1:80 negative), anti-DNA(ds) 10 IU/mL (≤ 4 IU/mL negative), RNP antibodies >8 AI (<1.0 AI negative), Smith antibodies 2.2 AI (<1.0 AI negative), anti scleroderma-70 0.3 AI (<1.0 AI negative), Sjögren's anti-SSA >8.0 AI (<1.0 AI negative), Sjögren's anti-SSB 3.3 AI (<1.0 AI negative), positive antichromatin >8.0 AI (<1.0 AI negative), anti-Jo1 <0.2 AI (<1.0 AI negative), anticentromere B <0.2 AI (<1.0 AI negative), sedimentation rate-Westergren 79 mm/hr (women under 50 years old: < 20 mm/hr), complement C3 127 mg/dL (75–175 mg/dL), complement C4 13 mg/dL (14–40 mg/dL), elevated CRP cardio 22.052 (≤ 10 mg/L in non-acute phase range). A transthoracic echocardiogram was performed which showed a pericardial effusion.

DISCUSSION

Pericarditis is the most common cardiac manifestation seen in SLE patients. Most cases can be managed with NSAIDs. However some cases may lead to cardiac tamponade. The pathogenesis of SLE is not entirely understood, however, antibodies and immune complexes (IC) are factors contributing to the clinical manifestations seen in these patients. They cause tissue damage via vasculopathy, which is ultimately responsible for many of the clinical manifestations of SLE. Immune complexes cause the activation of complement and cause further damage to the associated organ affected. Immunofluorescence and electron microscopy have identified immune complexes at the dermal-epidermal junction of the pericardium. This deposition may precipitate pericarditis as a presentation in SLE patients. Immune complexes pathogenicity depends on the characteristics of the antibodies. Different factors include the size, charge, affinity, specificity, and the ability to activate inflammatory mediators and/or complement. The rate at which the Fc receptors clear the immune complexes on macrophages also plays a role in pathogenicity [3].

There are two main hypotheses concerning the formation of antibodies in SLE patients. One involves polyclonal B cell activation, which occurs when B lymphocytes are stimulated non-specifically to form antibodies. The other is an antigen driven response with antibodies directed against these specific antigens. Antigens can be either autologous or exogenous. Autologous antigens may be altered through sunlight, infection, or other sources of tissue injury to stimulate an immune response. Exogenous antigens may become immunogenic through molecular mimicry [4].

Diagnosing SLE can be considered challenging, since there are varying degrees of clinical manifestations from patient to patient. Clinically, patients may have symptoms ranging from a rash and joint pain to life-threatening central nervous system, hematological, or renal involvement. Differentiating SLE from other autoimmune diseases proves difficult, however, increasing your index of suspicion and following the guidelines below will be clinically useful in the diagnosis of SLE in future patients. Ultimately, diagnosing SLE is based upon a skilled clinician who recognizes the signs and symptoms consistent with SLE and supports them with serological markers [5].

The current recommended guidelines to follow are the systemic lupus international collaborating clinics (SLICC). The diagnosis of SLE by the SLICC requires 4 out of the 17 criteria listed below, which contain at least one criterion representing the clinical and immunologic sections respectively. Diagnosis can also be made if the patient's biopsy proves Lupus nephritis in the presence of a positive ANA or anti-dsDNA. The following are the clinical criteria: acute cutaneous lupus, chronic cutaneous lupus, non-scarring alopecia, oral or nasal

ulcers, joint disease, serositis, renal involvement, neurological involvement, leukopenia or lymphopenia, and thrombocytopenia. Immunologic criteria are as follows: ANA, anti-dsDNA, anti-Sm, antiphospholipid, low complement, and direct Coombs test positive in the absents of hemolytic anemia [6].

Furthermore, there are varying degrees of probability that the diagnosis is in fact SLE. A definitive diagnosis of SLE consists of the SLICC diagnostic criteria previously stated. Probable SLE is defined as patients having two or three SLICC diagnostic criteria and at least one of the following criteria: optic neuritis, aseptic meningitis, glomerular hematuria, pneumonitis, pulmonary hemorrhage, pulmonary hypertension, interstitial lung disease, myocarditis, verrucous endocarditis, abdominal vasculitis, Raynaud phenomenon, and elevated acute phase reactants. Lastly, possible SLE is considered when only one SLICC criteria is met and at least one of the other features listed above is present [7]. In this case, positive findings included ANA, anti-dsDNA, anti-Sm and pericarditis thus satisfying the SLICC criteria for SLE.

Treatment typically depends on the disease manifestations. When patients become symptomatic with signs of pericarditis, non-steroidal anti-inflammatory drugs (NSAIDs) are typically initiated. Colchicine may be added to NSAID therapy if risk of recurrence is highly likely. In cases where inflammation does not subside with NSAIDs and colchicine, or if the patient is not able to tolerate these medications, then glucocorticoid therapy could be considered an alternative therapy [8].

In acute pericarditis some patients are classified as high-risk, so hospitalization is recommended for close evaluation and treatment. The following criteria suggest such patients; fever ($>38^{\circ}\text{C}$ [100.4°F]) and leukocytosis, evidence suggesting cardiac tamponade, a large pericardial effusion, immunosuppressed state, a history of therapy with vitamin K antagonists, acute trauma, failure to respond within seven days to NSAID therapy, elevated cardiac troponin (myopericarditis). If patients do not have any high-risk criteria, they can be managed as outpatient [9].

Rheumatology Departments of Yaound, Central and General Hospitals, in Cameroon noticed similarity of presenting signs of pericarditis amongst those diagnosed with SLE. Beginning January 2001 a three-year trial occurred observing SLE patients that presented with signs of pericardial involvement. A thorough analysis was done on these patients consisting of 22 females and 1 male with the age range of 13–65. The hospitals detected that 10 out of 23 patients (43.48%) presented with pericarditis. This study is substantial because it shows that these patients had the disease for over two years before it was shed to light. Findings of significance were pericardial rub in seven patients, and dyspnea in six patients. Echocardiography was performed to justify the diagnosis of SLE in every case. These tests also revealed abnormal repolarization in seven patients and a low

voltage QRS complex on three occasions. Physicians on site administered corticosteroids to alleviate symptoms. Serological workup was also obtained and was indicative of SLE in all patients (i.e., anti-nuclear antibody, anti-double stranded antibody). Cardiomegaly was also observed in all patients on X-ray. Thereafter, four patients experienced relapses of pericarditis during ensuing flares of SLE [10].

CONCLUSION

The lifetime prevalence of some cardiac manifestation of systemic lupus erythematosus (SLE) is estimated to be 50%, and should be high on the differential of any presentation with chest pain or shortness of breath. It is crucial that physicians consider SLE as a diagnosis when new onset pericarditis occurs in African-American females of reproductive age. If diagnosed early, patients can receive appropriate treatment while reducing the chance of recurrent episodes and iatrogenic treatment.

Author Contributions

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

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

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

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

Niket Sonpal – 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

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Unusual presentation of adenoid cystic carcinoma: A case report

Bhargavi Dasari, Ravi Kiran A., Kartheeki B., Shaik Izaz,
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ABSTRACT

Adenoid cystic carcinoma is a slow growing, aggressive malignant tumor. It is an uncommon tumor seen in the palate. This case report presents a patient with adenoid cystic carcinoma in the hard palate with a very unique presentation of the neoplasm.

Keywords: Adenoid cystic carcinoma, Hard palate

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INTRODUCTION

Adenoid cystic carcinoma is a rare malignant tumor that accounts only 10% of the salivary gland tumors [1]. In addition to the salivary glands, it affects the lacrimal glands, ceruminous glands and occasionally the excretory glands of the female genital tract. It is known for its long clinical course and plodding growth. It is unique for its local recurrence, late distant metastasis encouraging longer survival rate and spread through perineural lymphatics [2]. This is a case report of a 32-year-old male who presented with a seven-months-old swelling in the palate.

CASE REPORT

A 32-year-old male came to the emergency department with chief complaint of a growth in the left side of the palate region since seven months. It was initially smaller, gradually increasing in size and attained present size. Patient gave history of pain initially when it was started. Pain was sudden in onset, moderate, dull aching type, intermittent. No specific aggravating and relieving factors.

On extraoral examination, solitary submandibular lymph nodes palpable bilaterally of size 0.5x1 cm on right side and 1x1 cm on left side. They were of oval shaped, firm in consistency, freely movable and non-tender. On intra oral hard tissue examination, 44 were restored.

On inspection, an ulceroproliferative growth was seen on the left side of the hard palate. It was of oval shaped. It was of size 2x3 cm extending medially from mid-palatal region, laterally up to marginal gingiva, and anteriorly from the rugae region to the junction of hard palate and soft palate posteriorly. Color was whitish at the centre and erythematous at the periphery. On palpation it was tender, rough surfaced. Margins were everted and rolled out. Base was indurated (Figure 1).

Based on the history and clinical examination, a provisional diagnosis was given as mucoepidermoid carcinoma. Differential diagnosis was given as adenoid cystic carcinoma, carcinoma involving hard palate and carcinoma involving maxillary sinus. Blood investigations were done in which there was no abnormality except the raise in erythrocyte sedimentation rate. Panoramic radiograph was not suggestive of any pathology near hard palate (Figure 2). Hence, advanced imaging computed tomography scan was advised which revealed area of altered attenuation showing heterogeneous enhancement on contrast. It also revealed soft tissue attenuation of left maxillary sinus (Figure 3). Based on these findings, the radiographic differential diagnosis was as carcinoma involving the hard palate.

A soft tissue specimen of size 1.5x1 cm was taken and subjected to histology. 4x view revealed parakeratinized stratified squamous surface epithelium with tumor cells in the fibrocellular connective tissue stroma. 20x view showed aggregates of hyperchromatic tumor cells having scanty cytoplasm with central cystic spaces in the fibrocellular connective tissue stroma (Figure 4). Thus histopathological diagnosis confirmed it as adenoid cystic carcinoma.

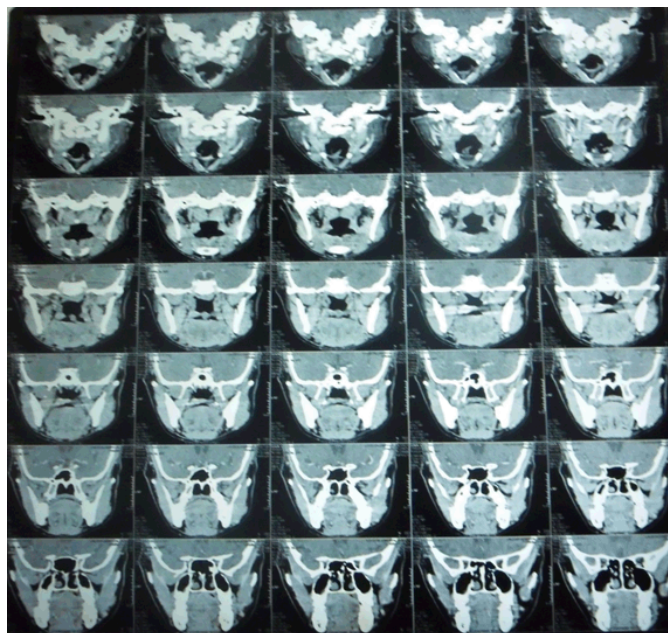


Figure 3: Computed tomography scan revealed area of altered attenuation showing heterogeneous enhancement on contrast and soft tissue attenuation of left maxillary sinus.



Figure 1: An ulceroproliferative growth seen in the left side of the palate.

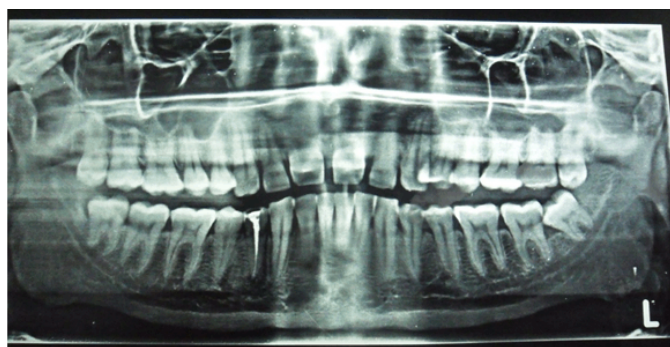


Figure 2: Panoramic radiograph was not suggestive of any pathology near hard palate.

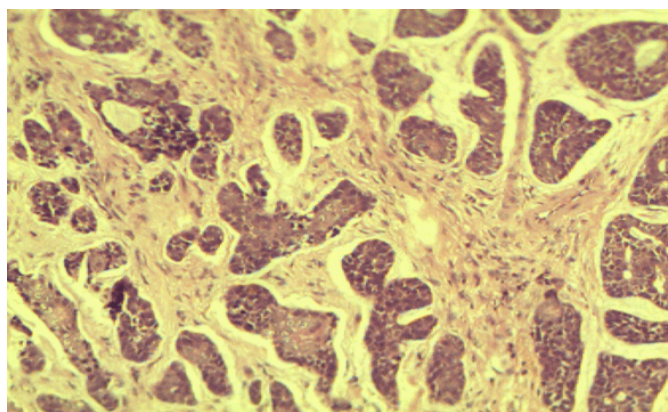


Figure 4: Histological picture showing aggregates of hyperchromatic tumor cells having scanty cytoplasm with central cystic spaces in the fibrocellular connective tissue stroma.

DISCUSSION

Billroth in 1859 was the first person to describe this infrequent malignant salivary gland tumor adenoid cystic carcinoma (ADCC) under the name cylindroma attributing to its cribriform appearance formed by the tumor cells with cylindrical pseudo spaces. The term ‘adenoid cystic carcinoma’ was instigated by Ewing (Foote and Frazell) in 1954. Adenoid cystic carcinoma (ACC) is an aggressive neoplasm with a phenomenal capacity for recurrence. The most common site for adenoid cystic carcinoma was palate as reported in the present case. It affects both genders equally and is mostly encountered in fourth and fifth decades of life which is not a correlating factor in the present case.

Usually, the clinical presentation is asymptomatic growth with a co-presentation of pain and paresthesia at

times. It is also described as an aggressive tumor because of the neural and lymphatic spread in other ways. Adenoid cystic carcinoma has a relatively indolent course and rare lymph node metastases but is familiar for its tendency for neurotropic spread and late local recurrences [3, 4]. In a study of 160 patients of adenoid cystic carcinoma by Fordice et al., disease-specific survival was 89% at five years but only 40% at 15 years. Distant metastasis was the most common type of treatment failure (in 22% patients), lungs being the most common site followed by liver [5].

Treatment of adenoid cystic carcinoma includes a complete excision of the local disease followed by post-operative radiotherapy. Radiation therapy used alone has a high rate of local recurrence but may provide useful palliation in inoperable/disseminated disease [5, 6].

CONCLUSION

As adenoid cystic carcinoma is a slow growing rare malignant tumor, its early detection by the dental specialist aids in favorable prognosis in almost all cases. The role of various diagnostic modalities like biopsy and advanced diagnostic imaging techniques like computed tomography scan has been mentioned in the present case. The therapy involving combination of surgery and radiotherapy remains the modality of choice in most cases.

Author Contributions

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The corresponding author is the guarantor of submission.

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Authors declare no conflict of interest.

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Hypopharyngeal non-penetrating steel shrapnel foreign body: A case report of unusual route of impaction

Ahmad Nasrat Al-juboori, Abdalla Mirghani Hamid

ABSTRACT

Introduction: Foreign body injury is one of the most commonly encountered otorhinolaryngologic emergencies. The diagnosis and management of foreign bodies have mainly been based on the type and location of the foreign body. The workplace is a significant contributor to fatal and non-fatal injuries worldwide and an insufficiently appreciated contributor to the total burden of health care costs. Steel workers sustain a higher occupational hazard of penetrating injuries anywhere in the body, including the head and neck. However, we found no reports in literature about non-penetrating shrapnel foreign body injuries, particularly in the upper aero-digestive tract. **Case Report:** A steelworker presented to the emergency department in Al Wakra hospital with a history of non-penetrating steel foreign body impaction in the throat which had been visualized by GlideScope and removed successfully with the assistance of Macintosh laryngoscope without complications. The purpose of this presentation is to highlight the unusual route taken by a shrapnel non-

penetrating foreign body, through the open mouth to the hypopharynx. **Conclusion:** We concluded that this report could be regarded as the first case report of a non-penetrating steel foreign body with an unusual per oral route of impaction in the hypopharynx. It, also, highlighted its visualization and a comparison between two techniques of its extraction.

Keywords: Foreign body throat, Glide video laryngoscope, Hypopharynx

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INTRODUCTION

Foreign body ingestion is one of the most commonly encountered otorhinolaryngologic disorders, often requiring urgent decision making and management. In particular, it has been reported that children younger than three years exhibit the greatest risk of foreign body swallowing [1, 2]. In adults, foreign body ingestion occurs commonly, the majority of them pass spontaneously but some of them will impact at the hypopharyngeal level [3]. The result may be severe due to possible ulceration or even perforation with consequent life-threatening complications [4, 5]. Many studies of foreign body injuries have focused on the case of young patients [2, 6].

However, it is also important to determine the frequency and characteristics of foreign body injuries in other age groups in addition to young children. Previous studies have demonstrated that some fatal cases occur in elderly patients due to foreign body asphyxia [7].

The diagnosis and management of foreign bodies have mainly been based on the type and location of the foreign body. Therefore, clinical information on the type and location of foreign bodies can expedite the management of these patients [7]. The workplace is a significant contributor to fatal and non-fatal injuries worldwide and an insufficiently appreciated contributor to the total burden of health care costs, like the occupational hazards of steel workers who manifest with penetrating injuries anywhere in the body [8]. However, there are no reported non-penetrating injuries of steel worker foreign body injuries particularly in the upper aerodigestive tract.

The aim of the presentation of this case report is to highlight the route of a non-penetrating steel foreign body in the hypopharynx in a steelworker, and probably this could be the first reported case of this unusual route of entry.

CASE REPORT

A 25-year-old steelworker presented to the emergency department in Al Wakra hospital with a history of foreign body impaction in the throat during work. When one of his colleagues was hammering steel nearby him, he felt something entering through his mouth, when it was open and getting inserted inside the throat. He was complaining of odynophagia, dysphagia with progressive change of voice. On examination, the patient was conscious, afebrile, not in respiratory distress and hemodynamically stable. Complete ear, nose and throat examination was normal, apart from a small wound at the tip of the tongue. Neck examination showed no evidence of external wounds or subcutaneous surgical emphysema but there was mild tenderness over the right side of the neck. flexible nasolaryngoscopy done under local anesthesia showed impacted foreign body (metal) on the right side of the supraglottic area with edema of the surrounding area of the epiglottis, aryepiglottic fold, arytenoids and part of the vocal fold on the right side. Plain X-ray of the neck, anteroposterior and lateral views, showed evidence of a metallic radio-opaque foreign body at the level of the fourth cervical vertebra directed towards the right side (Figure 1).

The patient was prepared for examination under general anesthesia and the removal of the foreign body with the assistance of rigid scopes. Examination under general anesthesia in the supine position with full sedation and assistance of a GlideScope, revealed a clear video picture of the metallic foreign body in the right pyriform sinus (Figure 2A). Trial of removal through the GlideScope guidance failed, unfortunately, because of difficulties in directing the instrument towards the

metallic foreign body. Then with the use of Macintosh laryngoscope the metallic foreign body was removed by forceps (Figure 3). Re-examination of the site of the foreign body was done again with the GlideScope and suction to the site of the foreign body to ensure there was no perforation (Figure 2B).

In the postoperative period, the patient was kept for continuous intravenous fluids for the next 24 hours, with frequent checking of the neck for possible emphysema, then fluid diet was started for the next few hours and then the patient was discharged in good condition on the second postoperative day after psychiatric assessment to make sure that he is of normal mentality.

DISCUSSION

Foreign body ingestion is more common in adults with mental developmental delay, psychiatric and neurological disorders or intoxication and in patients with dentures or dental bridges because of the decrease in tactile sensation during swallowing. The most commonly ingested foreign bodies are fish and chicken bones and there is an apparent predominance of certain types in specific groups of patients [9], e.g., coins and toys in children, razor blades and cutlery in prisoners [10]. Our case report highlights

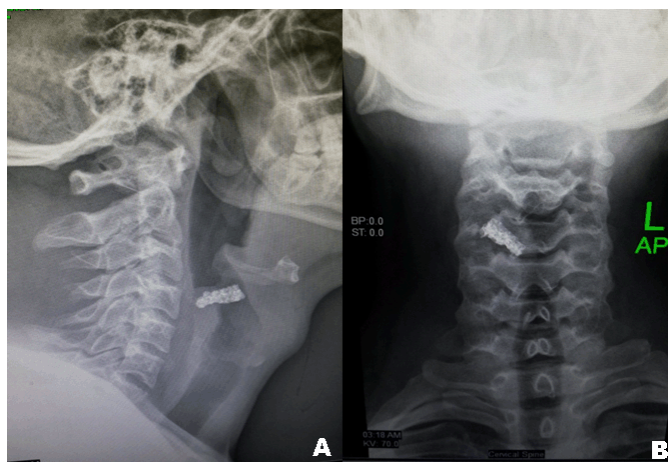


Figure 1: (A) Lateral plain X-ray of the neck showing radio-opaque steel foreign body in the level of 4th cervical vertebra, and (B) Anteroposterior view of the same patient.

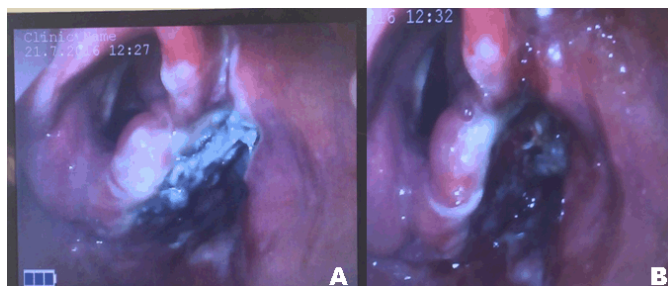


Figure 2: (A) GlideScope video laryngoscope image showing steel foreign body located in the right pyriform sinus, and (B) GlideScope image after removal of foreign body.



Figure 3: Steel foreign body.

the peculiar route taken by the shrapnel foreign body, through the open oral cavity to the hypopharynx, a route we think have not been reported before. Also, it appears that the foreign body has taken a curved projectile rather than a straight pathway. This is suggested by the fact that no injuries were found in the oral cavity or oropharynx, apart from a small laceration at the tip of the tongue. Medline search did not report this kind of foreign body route of entry, so we consider this is the first reported case with such description. GlideScopes represent a recent advancement over the Macintosh laryngoscope. It has increased endotracheal intubation success rate, and it is recommended mainly in the management of potential difficult airways in several patient populations [11]. The use of these devices to remove foreign bodies has been reported previously only in two adult patients affected by a non-impacted partial denture in the hypopharynx [12]. While desirable, it is not reported so far in the pediatric population. Je et al., in a cadaver study, compared Macintosh laryngoscope and the GlideScope for extracting hypopharyngeal foreign bodies. He stated that it is not possible to conclude that videoscopes are inferior to the Macintosh laryngoscope for foreign body extractions, and that the GlideScope might be necessary to aid the foreign body extraction in emergency situations [13]. In our case report, the GlideScope provided good indirect visualization and localization with magnification of the foreign body but it did not provide good access for removal. On the other hand, the Macintosh provided a direct visualization of the foreign body and easy removal and extraction.

CONCLUSION

From previous descriptions, we concluded that this report could be regarded as the first case report of a

non-penetrating steel foreign body with an unusual per oral route of impaction in the hypopharynx. It, also, highlighted its visualization and a comparison between two techniques of its extraction.

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Author Contributions

Ahmad Nasrat Al-juboori – Substantial contribution 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
Abdalla Mirghani Hamid – Substantial contribution 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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The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Lower extremity acute compartment syndrome secondary to inferior vena cava thrombus: A case report

Keegan Devon Bradley, Hao Wang

ABSTRACT

Introduction: Acute compartment syndrome (ACS) occurs when pressure within a muscle compartment builds to dangerous levels that exceed the perfusion pressure supplying blood to that compartment, which can lead to nerve and muscle tissue injury. It is uncommon to have an acute compartment syndrome suspected to be secondary to a clot of the inferior vena cava (IVC). **Case Report:** A 51-year-old female, without recent trauma presented to an outside hospital emergency department with symptoms of acute onset left lower extremity pain and swelling. She was admitted for ACS, and subsequently required emergent fasciotomy. While in a rehabilitation facility, she had worsening leg pain and presented to the hospital, where she was found to have an occlusive thrombus in her IVC that extended down to her lower extremities. It was later noted the anterior compartment of the patients left lower leg had become necrotic, and the patient could no longer dorsiflex her foot. After reviewing the report and outcome of the patient, it was concluded that she had developed compartment syndrome without any obvious instigating cause. **Conclusion:** It is important to have a low threshold and be vigilant for compartment syndrome in any patient presenting with suspicious symptoms, even

patients who seem to have no history or related trauma typically associated with compartment syndrome.

Keywords: Atraumatic, Compartment syndrome, Deep venous thrombosis, Inferior vena cava

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INTRODUCTION

The purpose of this case report is to demonstrate a novel case of suspected atraumatic acute compartment syndrome (ACS) secondary to a proximal obstructive venous thrombus. This is a potentially devastating condition when, for multiple reasons, an osseofascial compartment pressure rises to a level that decreases perfusion and may lead to permanent neuromuscular damage in that compartment. Acute compartment syndrome can occur anywhere in the body that muscle is surrounded by fascial tissue, and most commonly follows acute trauma involving long bones fractures [1]. Among all long bone fractures, tibia fractures are the most common fracture type with high ACS risks in adults [2]. In addition, ACS occurs more commonly in patients who sustain comminuted fractures [2]. One study showed that out of 164 patients, 69% of those cases were secondary to fractures in which tibial diaphyseal fractures accounted

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for the majority (36%) and distal radius fractures were the second most common (9.8%) [3]. Similar findings were reported in other study with tibia fractures of diaphysis being the most common one [4]. Acute compartment syndrome, however, can also occur in non-trauma patients. Hynes et al. reported a case of ACS in a female with a presumed DVT after a prolonged episode of drug induced sleep without known trauma [5]. Gutfraynd and Philpott reported a 24-year-old male who developed ACS of the left thigh after being at a concert for the entire day. The patient denied any trauma, but still required fasciotomy [6].

Another study found that in 113 ACS patients, those who sustained no fracture had a significant delay in diagnosis and treatment. An average of 12.4 hour delay from onset of symptoms resulted in muscle necrosis in 20% of patients who eventually required debridement during fasciotomy [3]. Some other non-traumatic causes of ACS include bleeding disorders, vascular disease, nephrotic syndrome (or other conditions that decrease serum osmolarity), certain animal envenomation and bites, extravasation of intravenous fluids, injection of recreational drugs, and prolonged limb compression (e.g., following severe drug or alcohol intoxication or poor positioning during surgery). Postischemic compartment syndrome phenomenon has also been seen commonly following many procedures such as; bypass surgeries, embolectomies, or thrombolysis. Increasing flow into an already swollen compartment puts the patient at high risk for ACS. Dr. Newman et al. demonstrates this with a case report of a 57-year-old male with non-traumatic ACS of three compartments of the left lower extremity [7]. The patient had been recently started on anticoagulation for multiple pulmonary emboli and a deep venous thrombosis of the left posterior tibial and peroneal veins. Patient was found to have 3 of the 4 compartment syndromes with >70 mmHg, which required fasciotomies that resulted in significant blood loss but no deficits postoperatively.

The clinical symptoms of ACS include any combination of: pain out of proportion to the clinical scenario, pain with passive stretch, paresthesias, peripheral swelling, and late findings including paralysis and absent peripheral pulses. However, different patients can present differently with very subtle symptoms. Cohen et al. reported pediatric patients can present more often with paresthesias than pain, and 12% of these patients with ACS will not have any pain at all in their course [8]. Stollsteimer et al. present a case of a young, unconditioned, football player who attempted to run a mile twice under 7.5 minutes. The patient then began to have persistent leg pain, but was able to achieve the run day-3. By this time the patient was diagnosed by the trainer with shin splints, but then was sent to the team physician day-4 due to ongoing pain and new inability to dorsiflex the foot. Patient was sent to hospital and diagnosed in the emergency department with ACS. He underwent fasciotomy, but unfortunately was unable to regain function in that extremity [9]. Another example of a different presentation of atraumatic

ACS is presented by Parisa et al. which demonstrates a patient that develops thigh compartment syndrome with an unknown cause secondary to a hematoma in his leg. The patient presented with 10/10 pain in his right thigh, and was found to have a large spontaneous thigh hematoma that required fasciotomy and evacuation. The only presenting symptom was ongoing pain without numbness or weakness [10].

CASE REPORT

A 51-year-old Caucasian female who presented to study hospital for chief complaint of left anterior leg pain, numbness between first and second toes, and inability to dorsiflex her left foot. Prior to being seen at study hospital the patient had presented to an outside hospital with a chief complaint of acute onset left lower extremity pain and swelling without trauma or an inciting event. She was admitted for ACS monitoring, where she subsequently developed increasing pain, swelling, and numbness in her left foot the following night. Imaging at the outside hospital showed bilateral deep vein thromboses and small distal pulmonary emboli. She was taken to the operating room the same night for emergency medial and lateral incision fasciotomy of her left lower extremity. Patient was monitored in hospital for another week before she was discharged to a rehabilitation facility. While in rehabilitation facility, patient daughter noticed that patient's left leg pain had become worse and she was subsequently diagnosed cellulitis and antibiotics were given. However, patient's leg pain has not improved and patient thus presented to emergency department of the study hospital for further evaluation.

Patient's past medical history included diabetes and hyperlipidemia. Past surgical history included tubal ligation, cesarean section, and recent left lower extremity fasciotomy. No family history of bleeding problems or cancer. Home medications of note include: oral contraceptive pills for a year, as well as Coumadin started at the initial outside hospital.

Physical examination was remarkable for a diffusely swollen left lower extremity with wound vacuum in place over medial and lateral previous fasciotomy sites. Upon removal of the wound vacuum, there was foul smelling serous drainage from both wounds. The medial incision showed viable muscle tissue that contracted with sudden stimulation. The lateral incision revealed a posterior compartment with muscle that contracts on stimulation, but the anterior compartment did not respond to stimulation. Patient was able to plantarflex, invert, and evert her left foot, but unable to dorsiflex. There is a loss of sensation in the deep peroneal nerve distribution, but sensation remained intact in the saphenous, sural, and superficial peroneal nerve distributions. Dorsalis pedis and posterior tibial signals were appreciable on doppler, but unable to be palpated. No pain with palpation of her thigh (Figure 1A–D).

Patient's X-ray of left lower extremity showed no acute fractures, and laboratory examinations were significant C-reactive protein (CRP) 125, lactate 3.5, white blood cell count (WBC) 19, and international normalized ratio (INR) of 2.3.

Ultrasound revealed thrombi extending from distal inferior vena cava to the level of the popliteal veins on left and calf veins on the right. Computed tomography angiography demonstrated occlusive thrombus within femoral and iliac veins extending into the IVC. The extension included the infrarenal, juxtarenal, suprarenal and up to the intrahepatic portion of the IVC. The suprarenal IVC measures 32 mm in diameter, which made it too large from any filter devices to be placed. The patient's subsequent hospital course began with vascular surgery recommending heparin therapy with a hematology consult for hypercoagulability workup, as well as to guide future long-term therapy. The patient was taken to the operating room by orthopedic surgery for fasciotomy and replacement of a wound vacuum. During the hospitalization, the patient also developed heparin induced thrombocytopenia (HIT) and heparin was discontinued. For the remainder of her hospital course her treatment was focused on medical anticoagulation via argatroban bridging to Coumadin with an INR goal of 2–3. Following discharge the patient would require lifelong Coumadin therapy given her extensive clot burden, as well as outpatient follow-up with orthopedics and continued physical therapy. She was most recently seen in the orthopedic clinic approximately six months following discharge from the hospital. She had been continuing outpatient physical therapy, but has not regained any ability to dorsiflex her left foot. (Figure 2A–D).

DISCUSSION

Here, we report a patient who presented initially with inferior vena cava thrombosis, subsequently developed lower extremity compartment syndrome, which eventually resulted in necrosis and fasciotomy. To the best of our knowledge, this case is unique due to few reported ACS cases that have developed without trauma in the setting of an occlusive proximal thrombus. Therefore, it is important to recognize atypical clinical presentations of such ACS and understand its etiologies.

Incidence of reported cases of unilateral ACS so distal to a bilateral occlusive thrombus without reperfusion is very scarce in current literature. Most cases of ACS reported secondary to a thrombus are typically seen when a thrombus is lysed and reperfusion causes increased inflammation and expansion of muscle tissue in the associated compartment as demonstrated by cases such as those presented by Dr. Newman et al. and Dr. Javedani et al. [7, 10]. It is important to note that this patient's underlying pathophysiology leading to her ACS is very different from the well documented post-ischemic compartment syndrome. The etiology of ACS



Figure 1: Patient's leg immediately prior and after fasciotomy was performed and a wound vacuum was applied. Left lower extremity on initial presentation February 2nd, 2015 (A) Left lower extremity after initial fasciotomy at outside hospital February 2nd, 2015, (B) Left lower extremity after initial fasciotomy at outside hospital February 2nd 2015, and (C, D) Left lower extremity taken during initial presentation to our hospital February 12th 2015.



Figure 2: Patient's wounds after the vac was removed and the progress of her healing. (A) Left lower extremity following surgery at our hospital February 17th, 2015, (B) Left lower extremity following surgery at our hospital February 17th, 2015, (C) Left lower extremity while being seen in clinic in April 2015, and (D) Left lower extremity taken at home in April 2015.

case reported here is most likely due to a significant VTE, including a bilateral lower extremity DVT which extends proximally to the IVC and pulmonary vessels. The incidence of bilateral lower extremity DVT is rare, and this should warrant more vigilant considerations of 1) the development of ACS; 2) the proximal large vessel VTE involvements; and 3) the potential systemic coagulopathies (such as anti-phospholipid antibody, protein C, protein S deficiency, etc).

Non-traumatic causes of ACS are not extensively reported, and likely predispose the afflicted patient to risk of prolonged time between presentation and diagnosis. Clinicians must have a lower threshold in diagnosing a patient with ACS who presents without a recent history of trauma. A diagnosis of ACS in a non-traumatic patient should be suspected among patients with lower extremity VTE.

The diagnosis of ACS itself is typically clinical, involving the physician looking for symptoms such as: increasing pain out of proportion to the stimulus, altered sensation, pain with passive stretch, muscle weakness, and palpable tightness of muscle compartments [1]. It is important to note that these symptoms are not, by themselves, particularly specific or sensitive in the diagnosis of ACS. For example, the degree of pain a patient reports can differ dramatically from patient to patient, hypoesthesia and muscle weakness can be secondary to direct nerve damage rather than increased compartment pressure, and compartment stiffness is not as appreciable in deeper compartments that are typically not palpable [2]. Symptoms such as muscle weakness or paralysis are signs that are typically seen at a point in the ACS process when permanent damage has been already occurred [11]. Individually, these tests are not diagnostic, but a constellation of multiple positive symptoms should alert the clinician to a case of potential ACS. Other diagnostic tests such as intercompartment pressure measuring can be applied with many agreeing that a difference between the diastolic blood pressure and the compartment pressure (delta pressure) of 30 mmHg or less be used as the threshold for diagnosing ACS [12]. Unfortunately, many of these advanced methods are not always on hand acutely when concern for ACS arises. It falls on the clinician to assess and use their best diagnostic judgement on whether the patient is suffering from ACS and requires immediate surgical intervention.

This case demonstrates how variable the underlying suspected causes of ACS in a patient can be. This patient's initial presentation was relatively stereotypical for ACS. However, she had a nontraditional underlying cause of an occlusive thrombus in her IVC. The thrombus was found to occlude from her IVC, bilateral femoral veins, and further extension down to the popliteal vein on the left and calf vein on the right. The significance of proximal occlusion is unclear, but further research or cases may demonstrate the possibility that proximal impedance of flow may lead to increased pressure in all distal compartments. Investigation for an occlusive proximal DVT would likely be indicated, if it can be further demonstrated in the future that proximal occlusion is in fact, a risk for development of distal ACS. She had a hypercoagulable workup done while being admitted to our hospital which was negative, except for being initially positive for antiphospholipid antibody which will require confirmatory testing later. Given this patient's potential to be antiphospholipid antibody positive and the potential

that proximal occlusion by a DVT can cause distal ACS, hypercoagulability workup should be considered in a patient with an otherwise non-traumatic unknown cause of ACS and spontaneous DVT formation.

What we learned from this case is that there are still potentially unknown causes of ACS that can be just as devastating. This can also delay a patient's diagnosis and more importantly their intervention. We found that occlusion of venous blood flow can potentially predispose the distal compartments to developing ACS. We suspect the pathophysiology behind the patient's ACS occurring in the setting of a clot without reperfusion could possibly be secondary to: increases in pressure from lack of venous circulation return, an injury the patient does not remember that was significant enough to instigate an already pressurized compartment, or simply idiopathic ACS with a large clot being incidentally identified on investigation for underlying causes.

There remain some limitations of this case report. We are unable to confirm the patient had no trauma in her recent past though she did not recall the event. All pertinent information was obtained from this patient and no formal previous outside hospital health record reviewed. We are also unable to find strong evidence based study in literature to support the hypothesis of a proximal venous occlusion causing distal ACS. Further research should be focused towards assessment of the entire limb distal to the occlusion to measure if all compartments pressures increased or simply that single compartment. It should also be investigated if thrombectomy and return of flow will improve this specific cause of ACS by relieving pressure.

CONCLUSION

It is important for clinicians to suspect non-traumatic acute compartment syndrome (ACS) with atypical signs and symptoms such as bilateral lower leg DVT. More importantly, thorough evaluations including proximal venous images and systemic coagulopathy workups should be performed for potential ACS etiologies among these patients.

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Author Contributions

Keegan Devon Bradley – 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

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

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Percutaneous endoscopic colostomy for management of pseudo-obstruction

Mirza Faraz Saeed, Mobeen Ashfaq,
Othman Yousef Alfrayyan, Amro Salem

ABSTRACT

Introduction: Percutaneous endoscopic colostomy (PEC) is a minimally invasive variation of percutaneous endoscopic gastrostomy that can be offered to patients who may not be ideal candidates for surgery, or in whom surgery may be inappropriate for other reasons. **Case Report:** A 61-year-old male with multiple comorbidities, presented with a one-week history of significant diffuse abdominal pain and distention that has been recurrent, associated with anorexia, nausea and chronic constipation. The patient was admitted with the impression of pseudo-obstruction after meticulous clinical examination and imaging (abdominal X-ray and computed tomography scan). During the course of hospital stay, patient refused to undergo the advised surgical procedure and the rectal tube was put in place which failed to relieve the symptoms. Thus, percutaneous endoscopic colostomy (PEC) was performed as per the method first described by Jeffry Ponski

in 1986. Colonoscopy showed a partial sigmoid volvulus, for which detorsion was performed to relieve the chronic constipation. Subsequently, the patient was able to receive regular colonic enemas to prevent recurrent obstruction with decrease in abdominal girth with significant deflation and clinical improvement. **Conclusion:** Percutaneous endoscopic colostomy (PEC) is a promising technique that has shown good results while reducing the surgery burden in selected patients. This patient shows an excellent prognosis at three months follow-up, however more studies are recommended to evaluate the long-term outcomes.

Keywords: Colostomy, Endoscopic, Percutaneous, Pseudo-obstruction

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INTRODUCTION

Percutaneous endoscopic colostomy (PEC) is a well-established variation of percutaneous endoscopic gastrostomy. Percutaneous endoscopic colostomy offers an alternative for patients who have failed conventional treatment of conditions such as fecal constipation, acute colonic pseudo-obstruction, and recurrent sigmoid

volvulus. It has also been a useful technique for clinicians in cases where surgery may be contraindicated, for example, in the elderly or frail patients [1]. While PEC has shown promise in certain clinical scenarios, there is still doubt and room for debate as to when is the right time for this technique in cases that have vague presentations such as the one presented in this study. This is a case of a 61-year-old male with refractory chronic constipation treated successfully with PEC. A search was conducted through PubMed, Cochrane Library as well as NICE guidelines to look for high quality data to compare the most favorable management plan in similar cases to the one underwent by our patient.

CASE REPORT

A 61-year-old male, known case of hypertension and psychiatric illness, presented with abdominal pain and distension of one week duration. Abdominal pain was generalized and severe. The patient also had a background history of constipation, anorexia and nausea for one year. He had previous similar attacks that were treated conservatively with fleet enemas. The patient has no previous abdominal surgeries. There was no history of weight loss, bleeding per rectum or diarrhea. The patient had no known allergies and no family history of colonic malignancies.

On examination the patient was afebrile, tachycardic, and in pain and distress. Abdominal examination showed massive abdominal distention with minimal tenderness on palpation. The abdomen was also diffusely tympanic on percussion, with high-pitched tinkling bowel sounds on abdominal auscultation. The remainder of systemic examination was unremarkable. Per rectum examination showed massive stool evacuation and release of gases. Initially, the patient was admitted with the impression of Ogilvie syndrome for observation. This was based on the presence of acute pseudo-obstruction and colonic dilatation seen on imaging, and due to the absence of an anatomic lesion to obstruct intestinal flow. On admission the patient was kept nil per os, resuscitative measures done, started on intravenous erythromycin and rectal tube inserted to acutely manage the constipation.

X-ray of the abdomen (erect and supine) showed the appearance of extensive colonic gaseous distention that raised both diaphragmatic leaflets. Colonic liquefied fecal content was seen extending down to the rectum. Overall impression on imaging was of pseudo-obstruction (Figure 1). Computed tomography (CT) scan of abdomen with contrast showed marked diffuse colonic dilatation with no evidence of a mechanically obstructing lesion (Figures 3 and 4).

As part of initial management, a cecostomy tube was inserted that failed to produce contents and the abdominal girth was increasing daily, despite six hourly flushing of the rectal tube. Patient was advised surgery, with all necessary risks and benefits are explained. However,



Figure 1: Supine abdominal X-ray on admission (Cecum approximately 9 cm).



Figure 2: Post-percutaneous cecostomy.

the patient and his family refused the procedure. Plan to perform a percutaneous endoscopic colostomy was implemented. The procedure was done under sedation using midazolam 1 mg intravenous and fentanyl 0.025 mg intravenous. Partial sigmoid volvulus was noted on endoscopy, for which detorsion was performed. Under the guidance of two operators, trans-illumination was performed with the lighted tip of the endoscope pressing outward against the abdominal wall to identify point of insertion. Local anesthetic was injected into the skin and using the percutaneous endoscopic gastrostomy (PEG) kit, a needle was used to enter the colon under direct vision. The wire was grasped by a snare and then the PEC tube was pulled and trailed into the colon and out through the abdominal wall using the Ponsky technique. The PEC tube was inserted at 35 cm from the anal verge, with confirmation of placement via endoscopy (Figure 5).

The patient remained stable throughout the procedure and following the procedure, an abdominal X-ray was performed to confirm placement of the PEC. Following

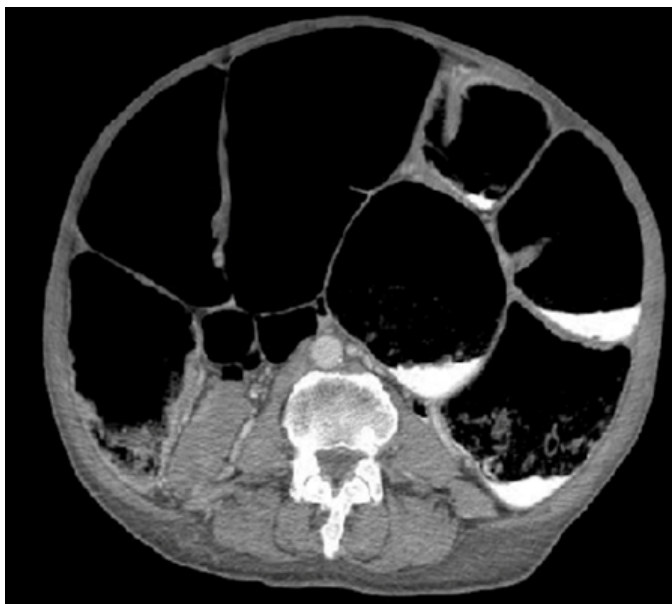


Figure 3: Axial abdominal computed tomography scan showing dilated colon.



Figure 4: Coronal abdominal computed tomography scan view showing dilated colon (ascending colon approximately 10 cm).

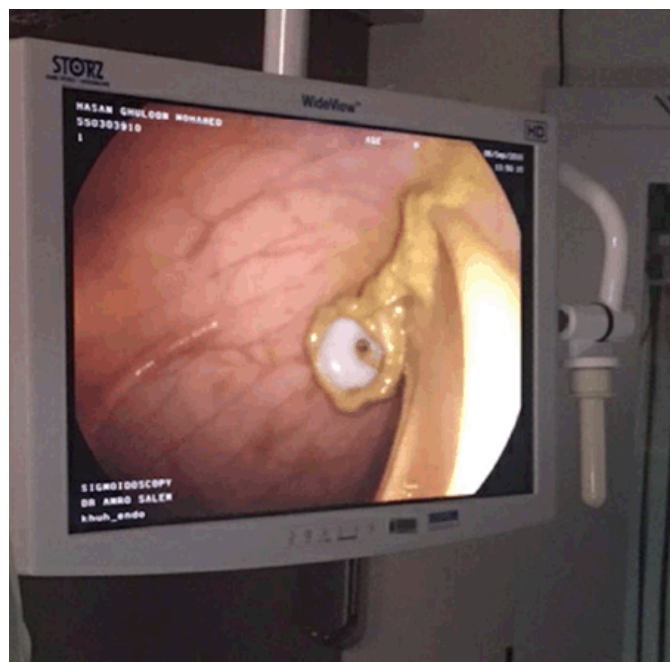


Figure 5: Endoscopic confirmation.

the procedure, abdominal girth began to deflate following daily fleet enema and digital rectal evacuated stool from the colon. The PEC tube was routinely flushed with 20 cc saline to prevent any blockages.

Patient has been followed as outpatient clinic for three months, he was doing well, stoma was functioning well, and has no abdominal distention, pain or vomiting.

Percutaneous endoscopic colostomy tube was removed after three months with no complications.

DISCUSSION

Percutaneous endoscopic colostomy (PEC) was first described in 1986 by Jeffrey Ponsky and it is considered as a safe and minimally invasive procedure [1, 2]. Percutaneous endoscopic colostomy has become a viable alternative to surgical intervention in patients with recurrent sigmoid volvulus, acute colonic pseudo-obstruction and fecal constipation [1], and is also a highly efficient intervention in elderly, with multiple comorbidities who will be under high risk if they go for surgical intervention [3–5].

Complications can include failure of procedure, recurrence of obstruction, tube migration, abdominal wall damage, infection, pain and peritonitis, which is considered the most serious complication among all others [4, 6]. Percutaneous endoscopic colostomy is contraindicated in a number of conditions such as colonic ischemia, anterior abdominal wall infection and mechanical intestinal obstruction [4].

In recent years, there has been an increase in the literature published on this procedure, the largest publication on this procedure reported cases of 15 children

who had PEC indicated due to refractory constipation. At follow-up evaluation 12 months later, all children were socially clean and two were able to have the PEC removed [1]. The pitfalls in studies published about PEC indicated in sigmoid volvulus involve poor sample size and varying techniques for placement of the PEC. Currently, the procedure is not recommended as a routine option and is only performed in cases that a deemed inoperable or unfit for surgery [1].

CONCLUSION

Percutaneous endoscopic colostomy (PEC) is a minimally invasive and highly effective procedure for patients that may be not be fit for surgery, or where high-risk surgical consent cannot be obtained such as in the case presented here. The procedure has shown to be a very durable alternative to surgery in cases presenting with recurrent refractory fecal constipation, recurrent sigmoid volvulus and acute colonic pseudo-obstruction. Our patient was followed for three months' post procedure and he remained stable throughout with no complications. More studies that evaluate the durability of this practice over a longer period of follow-up are recommended.

Author Contributions

Mirza Faraz Saeed – 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

Mobeen Ashfaq – 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

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A unique case of neurological manifestation of hemolytic uremic syndrome which responded to the treatment with intravenous magnesium sulfate

Anza Memon, Salman Rashid, Mitchel T. Williams

ABSTRACT

Introduction: We describe a patient with central nervous system (CNS) manifestation of hemolytic uremic syndrome (HUS) presented with new onset seizure and focal cortical signs with reversible lesions involving the splenium of the corpus callosum (SCC) and evidence of reversible focal cerebral vasospasm (RFCV) on brain magnetic resonance angiogram (MRA) responsive to intravenous $MgSO_4$. This is a rare case of neurological presentation of HUS which improved after treatment with high doses of magnesium sulfate ($MgSO_4$). **Case Report:** A 13-year-old Caucasian female with HUS and positive stool test for Shiga toxin two was being managed with hemodialysis and subsequently with the plasma exchange (PLEX). Following her seventh cycle of PLEX the patient developed left gaze deviation and head version with generalized tonic stiffening. **Conclusion:** $MgSO_4$ is an N-methyl aspartate (NMDA) receptor antagonist. Influx of calcium ions through NMDA receptor can lead to cerebral ischemic injury which can be reversible in the early stages of the disease process. **Improvement of neurological symptoms**

in our patient after treatment with $MgSO_4$ is a novel finding which has not been reported before in HUS.

Keywords: Neuroprotective role of magnesium sulfate, Reversible focal cerebral vasospasm in hemolytic uremic syndrome, Neurology of hemolytic uremic syndrome, Splenium of corpus callosum lesions in hemolytic uremic syndrome

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INTRODUCTION

Neurological manifestation of hemolytic uremic syndrome can be seen in 20–50% of the patients [1]. Common central nervous system presentation of the disease is altered mental status, seizures, visual impairment, brain stem, pyramidal and extrapyramidal symptoms. Neurological findings and abnormal neuroimaging with involvement of basal ganglia, thalami, internal capsule, cerebellum, brainstem, subcortical white matter and SCC have been described in literature [1]. Most of these findings are reversible and basal ganglia are reported to be commonly involved. Thrombotic microangiopathy in HUS leading to endothelial cell damage is the hallmark of renal and extra-renal manifestation of the disease.

Reversible leukoencephalopathy syndrome associated with HUS has also been reported [2], which could be the result of uncontrolled hypertension and renal insufficiency causing subcortical edema without infarction.

In this case report, we describe a patient with CNS manifestation of hemolytic uremic syndrome (HUS) presented with new onset seizure and focal cortical and pyramidal signs. She found to have reversible brain MRI lesions involving splenium of corpus callosum (SCC) with evidence of reversible focal cerebral vasospasm (RFCV) on brain magnetic resonance angiography (MRA) responsive to high doses of intravenous magnesium sulfate ($MgSO_4$).

CASE REPORT

A 13-year-old right-handed Caucasian girl with no prior significant medical history was admitted for abdominal pain, fever, vomiting and bloody diarrhea for one week. She was being managed for HUS with hemodialysis and subsequently with plasma exchange (PLEX). She had positive stool test for Shiga toxin 2. Following her seventh cycle of PLEX, she reported to have a generalized tonic clonic seizure. On examination the patient was drowsy with a persistent left gaze deviation and right sided hemiplegia. She found to have diffuse hyperreflexia and non-sustained ankle clonus. She was afebrile with a temperature of $36.7^{\circ}C$ ($98^{\circ}F$) and normotensive with a blood pressure of 117/77 mmHg. She received 2 doses (2 mg) of intravenous lorazepam and loaded with 20 mg/kg of intravenous phenytoin. She was subsequently started on 100 mg three times daily dose of phenytoin. The patient was intubated due to respiratory insufficiency. She remained normotensive throughout this period. Long-term video EEG recording to evaluate for non-convulsive status revealed a diffusely slow background with significantly attenuated left hemispheric activity. Seizure like activity and epileptiform discharges were not seen (Figure 1). Brain magnetic resonance imaging (MRI) scan demonstrated diffusion restriction in the SCC with low apparent diffusion coefficient (ADC) correlates (Figure 2A–B). Axial FLAIR sequences revealed subtle hyperintensity in the SCC (Figure 2C). Magnetic resonance angiography (MRA) of the head revealed attenuation in the left middle cerebral artery (MCA) territory (Figure 2D). Magnetic resonance venogram (MRV) of the head was unremarkable (Figure 2E).

Spinal fluid analysis indicated normal white blood cell count of 5×10^3 cells/ mm^3 (36% neutrophils, 48% lymphocytes, 16% monocytes), a red blood cell count of 351 million cells/ mm^3 , slightly increased protein of 68 mg/dl, and cerebrospinal fluid glucose of 81 mg/dl. Cerebrospinal fluid (CSF) gram stain and cultures were normal. Cerebrospinal fluid herpes simplex virus (HSV), varicella zoster virus (VZV), Epstein–Barr virus (EBV), cytomegalovirus (CMV), toxoplasma and west Nile virus were negative. The patient's blood, urine and respiratory

cultures were all negative. She had normal white blood cell count with normal comprehensive metabolic panel.

Despite anticonvulsive therapy our patient continued to have left-gaze preference and right hemiparesis, which persisted for three days. It also became evident that despite being intubated she appeared to have a receptive aphasia. Given the MRA showed what appeared to be vasospasm of the left MCA it was elected to give a trial of intravenous $MgSO_4$. She received 3 g (15 mg/kg) of $MgSO_4$ in divided doses with significant improvement of her symptoms within three hours and complete resolution of her gaze preference, aphasia and hemiparesis within 24 hours. Twelve weeks after discharge from the inpatient setting her neurological examination, including cognitive and motor functioning, were completely normal. Repeat MRI scan of brain six weeks after onset of symptoms revealed complete resolution of the signal abnormalities in the SCC on DWI, ADC and axial FLAIR sequences (Figure 3A–C).

Repeat long-term EEG recording eight weeks after her initial seizure was also normal (Figure 4).

DISCUSSION

Neurological manifestation of HUS is considered to be multifactorial including hypertension, microangiopathy and metabolic derangements in the form of electrolyte imbalances secondary to dehydration and renal insufficiency. Most of the studies describe the neurological findings due to verotoxin induced damage to the endothelium of small vessels in the brain causing bleeding, infarction or cerebral edema due to microvascular changes which can be seen on neuroimaging [3, 4]. These changes lead to secondary immune mediated cytokine release injury to the neuron and glial cells. Neurons and glia lack receptors for the verotoxin. Therefore, verotoxin is not directly neurotoxic [5]. Based on the animal studies Shiga toxin II is more neurotropic compared to Shiga toxin I [6]. This finding is also observed in children with HUS with serious

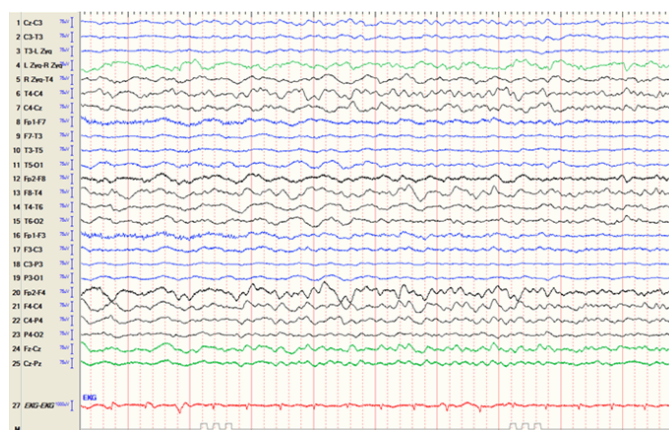


Figure 1: Long-term continuous video EEG showing a diffusely slow background as evidence of diffuse neuronal dysfunction with significantly attenuated left hemispheric activity.

neurological complications seen in patients with Shiga toxin II. The exact reason for comparatively increased virulence of Shiga toxin II is not known. However, it is speculated that since both Shiga toxin I and II compete

for gut absorption through the same receptor so the absence of Shiga toxin I might increase the absorption of Shiga toxin II [6].

Our patient's presentation with a seizure and focal neurological findings of left gaze deviation and right hemiparesis localizes to the left MCA territory which was evident as a possible left MCA vasospasm on MRA head, which correlated with her EEG findings of left hemispheric attenuation. The diffusion restricting lesions within SCC could be related to the disturbances in the intracerebral homeostasis in the absence of apparent metabolic derangements which possibly improved with the administration of intravenous $MgSO_4$.

Therapeutic role of $MgSO_4$ has been extensively studied in cardiovascular and obstetrical practices. It is being used in the treatment of eclampsia and prevention of eclamptic seizures for many years. It is also used in the prophylactic treatment of angiogenic vasospasm in aneurismal subarachnoid hemorrhage. Magnesium sulfate is an NMDA receptor antagonist. It blocks the calcium influx through the voltage gated calcium channels and NMDA receptors; preventing smooth muscle contraction and neuronal damage [7]. It has been implicated that influx of calcium ions through NMDA receptor can lead to cerebral ischemic injury which can be reversible in the early stages of the disease pathogenesis [8]. Improvement of neurological symptoms in our patient after treatment with intravenous $MgSO_4$ is a novel finding which has not been reported before in HUS. The left MCA vasospasm seen on MRA of head in our patient represents possibly early spectrum of the disease course before permanent cerebral ischemia or intracerebral hemorrhage. These changes responded to the treatment with $MgSO_4$. We speculate that $MgSO_4$ is not only involved in restoring the biochemical homeostasis and prevention of calcium influx through NMDA receptors but may also exert the NMDA modulating anti-inflammatory effect. This phenomenon can enhance the vasodilatation of the vessels as well as diminish the degree of immune mediated pathological processes leading to irreversible brain damage and irritation of the brain tissue causing seizures. Another interesting neuroimaging finding seen in our patient is reversible lesion in the SCC. This lesion demonstrated diffusion restriction and low ADC value on initial brain MRI scan represents cytotoxic edema. Reversible splenial lesion syndrome (RESLES) has been described in the literature and could be seen in variety of conditions including infections, seizures, metabolic disorders, malnutrition, high altitude cerebral edema and antiepileptic drug (AED) withdrawal [9]. Reversible splenial lesions are not commonly reported in patients with HUS. In our patient, this could represent an ongoing seizure activity or sequel of an underlying metabolic derangement causing deregulation of the cellular fluid. There are no clear guidelines for the treatment of neurological sequel of HUS. Gitaux et.al. suggested the use of monoclonal antibodies against the terminal complement pathway to treat the early

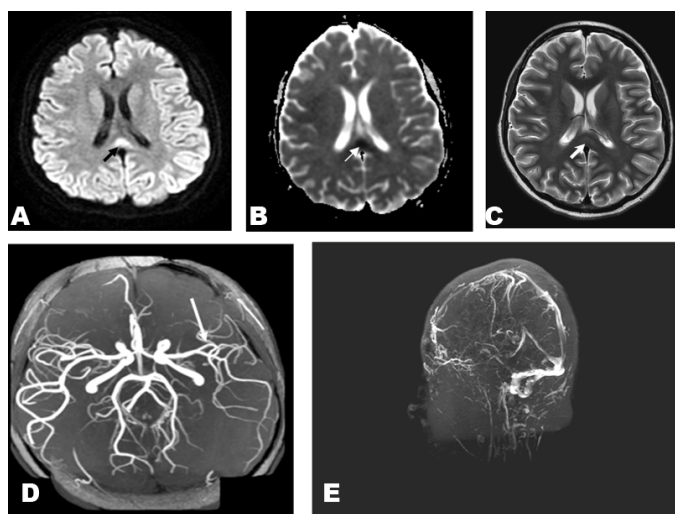


Figure 2: Magnetic resonance imaging (MRI) Brain, Magnetic resonance angiography (MRA) and Magnetic resonance venography (MRV) head without contrast obtained at the onset of symptoms: Splenial lesion on (A) Diffusion weighted imaging (DWI), (B) Apparent diffusion coefficient (ADC) and (C) Axial FLAIR. (D) MRA head shows evidence of diffuse narrowing in the distribution of left MCA territory compared to the right MCA suggestive of vasospasm (long-white arrow), (E) Normal MRV.

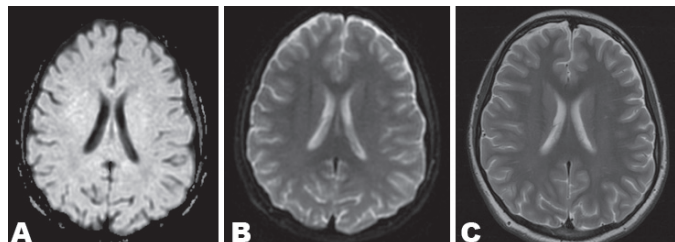


Figure 3: Repeat magnetic resonance imaging scan of brain six weeks later demonstrating complete resolution of the splenial lesion on diffusion weighted imaging (A) Apparent diffusion coefficient (B) and T2 (C).



Figure 4: Repeat long-term video electroencephalogram recording after eight weeks showing return of normal background and resolution of attenuation on the left side.

signs of neurological involvement and prevent further complications in HUS. This study was based on the fact that Shiga toxin II seems to exert its effects through direct activation of the complement system [10]. Other treatments used for cerebral vasospasm include calcium channel blockers. The Magnesium sulfate induced vasodilatation and immune modulation can make it a reasonable treatment option in the early stages of vasospasms to stop the immune mediated pathological sequel as discussed above.

CONCLUSION

Magnesium sulfate ($MgSO_4$) plays an important role in restoring the biochemical homeostasis and prevention of calcium influx through N-methyl aspartate (NMDA) receptors. It may also exert the NMDA modulating anti-inflammatory effect which could potentially help preventing the reversible neuropathology associated with hemolytic uremic syndrome.

Author Contributions

Anza B. Memon – 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

Salman Rashid – 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

Mitchel T. Williams – 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

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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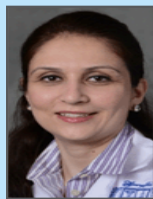
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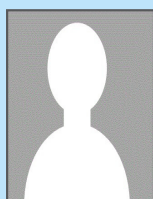
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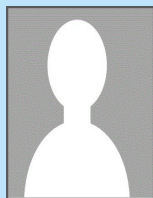
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Pseudo-Meigs syndrome: A case report

Divya Kallarackal, Dharampal Singh

ABSTRACT

Introduction: Meigs syndrome and pseudo-Meigs syndrome both presents with hydrothorax and ascites. Meigs syndrome is characteristically associated with ovarian fibroma whereas pseudo-Meigs syndrome is associated with any ovarian or pelvic tumors, other than ovarian fibroma. **Case Report:** A 48-year-old perimenopausal woman presented with a long history of 8–10 years of abdominal distension. Her examination revealed a right pleural effusion, massive ascites and large heterogeneous pelvic tumor, measuring 42x31 cm. After a preoperative ascitic tapping, the patient underwent an exploratory laparotomy with excision of the tumor, uterus and the right ovary. The tumor was diagnosed histologically as an ovarian mucinous cystadenoma. The postoperative resolution of hydrothorax and ascites confirmed the diagnosis of pseudo-Meigs syndrome. The patient remains in good condition 12 months after surgery. **Conclusion:** Pseudo-Meigs syndrome being a rare syndrome, with a good prognosis should be included in differential diagnosis in women presenting with unexplained hydrothorax and ascites.

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INTRODUCTION

Meigs syndrome is a rare condition, defined as the co-existence of benign ovarian fibroma, pleural effusion and ascites. While, pseudo-Meigs syndrome is characterized by the co-existence of pleural effusion, ascites and other ovarian or pelvic tumors. It was Meigs and Cass who brought out the significance of pleural effusion and ascites in ovarian fibroma. These syndromes should be considered in otherwise healthy postmenopausal women, who present with either hydrothorax or ascites. For both these syndromes, surgical resection of the tumor is the only therapeutic choice, resulting in resolution of fluid accumulations [1].

CASE REPORT

A 48-year-old perimenopausal woman came with history of abdominal distension since last 8–10 years, difficulty in breathing with increasing intensity over the past few months. She became very uncomfortable in supine position. She had no medical or surgical history of note. She is para 3 with uneventful vaginal deliveries. She took no regular medication and had no family medical

history of note (Figure 1). Auscultation revealed absence of breath sounds at the right lower hemithorax and normal heart sounds. On abdominal examination massive ascites was noted. The mass was not palpable because of the tense ascites. Chest X-ray revealed mild right sided pleural effusion (Figure 2). Electrocardiography was within normal limits. On ultrasound of abdomen, a massive multi septate cystic mass with suspected ovarian origin, with massive ascites was noted. Computed tomography scan revealed a huge multiseptate mass with solid and cystic components measuring 42x31 cm arising from the pelvis. Left side ovary was not visualized and uterus was normal sized. Massive ascites was noted. No obvious lymphadenopathy was seen. Her serum CA 125 was 49 U/ml (normal <35U/ml). AFP was within normal limits while β -hCG was not detectable. Her serum proteins were slightly below normal. Routine blood investigations, including LFT's and RFT's were within normal limits. Ascitic tap fluid cytology revealed low cellular fluid comprising of lymphocytes and mesothelial cells. No evidence of malignancy was noted.

A preoperative diagnosis of left ovarian tumor was made and nearly 6 L of ascitic fluid was drained in two settings in ward three days before surgery and the day before surgery. In operation theatre, under epidural anesthesia, a wide bore silicone catheter was inserted and nearly 18 L of ascitic fluid was tapped slowly over a period of 60 minutes in lateral position. Then in supine position, through a midline incision from pubic symphysis to 2 cm above umbilicus, a mass measuring 42x31 cm, weighing 9 kg was removed, originating from left ovary.

There was no any evidence of metastasis or lymphadenopathy. Omental and peritoneal biopsy were taken. Hysterectomy with bilateral salpingo-oophorectomy was done. Grossly uterus with cervix measured 9x6 cm, right ovary measured 2.5x1.3 cm, both unremarkable (Figure 3).

On histopathology report, the mass was diagnosed as an ovarian mucinous cystadenoma. The pleural effusion resolved by postoperative day-10. The patient remains in good condition 12 months after surgery.

DISCUSSION

Meigs syndrome is defined as a triad of benign ovarian tumor (ovarian fibroma), ascites and pleural effusion. Though the association of pleural effusion with benign pelvic tumor was described by Salmon in 1934, it was Meigs and Cass whom brought out the significance of pleural effusion and ascites in ovarian fibroma. It is to be noted that the ascites and effusion resolves completely after resection of tumor. Meigs syndrome is a benign disease with a good prognosis.

Pseudo-Meigs syndrome on the other hand consists of ascites and pleural effusion associated with any pelvic tumor other than fibroma. It is clinically important as it may resemble metastatic pelvic cancer. Cytological

examination of the body cavity effusions is essential to differentiate between reactive process and metastatic tumor spread.



Figure 1: Preoperative picture of patient (written informed consent taken for this picture).

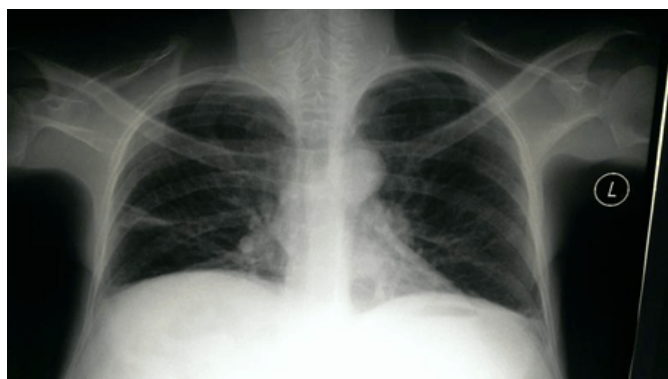


Figure 2: Preoperative chest X-ray of patient.

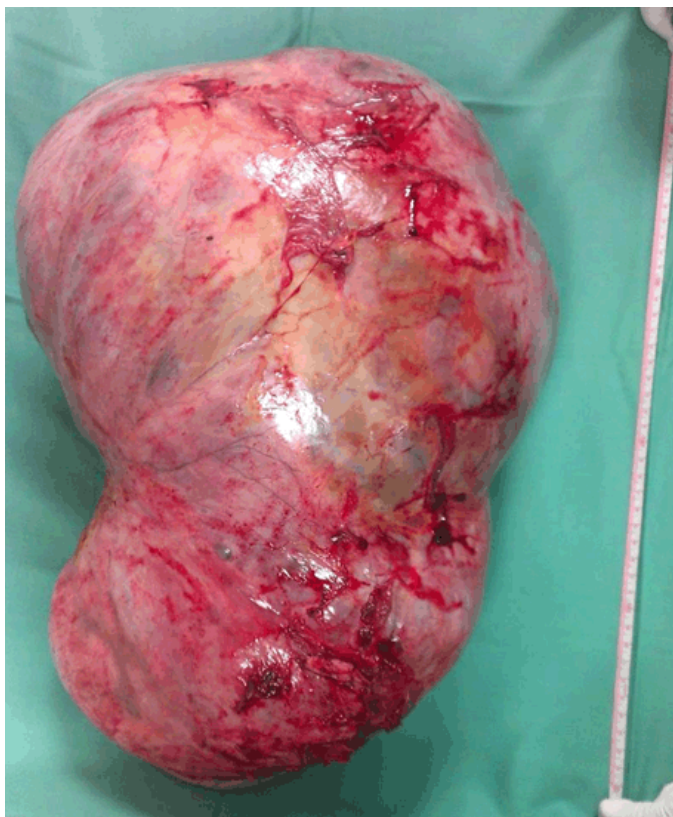


Figure 3: Mass after dissection.

The etiology of the fluid accumulation in Meigs syndrome and pseudo-Meigs syndrome remains unclear, although it appears to be related to lymphatic obstructions. Most likely it is due to filtration of interstitial fluid in the peritoneum through tumor capsule and diffusion to the pleural space through diaphragmatic lymphatic vessels and apertures. The effusion can be moderate or massive and is grossly transudative, but occasionally contains blood cells [2, 3]. It is usually observed with ovarian tumor larger than 6 cm and they completely regress after neoplasm removal.

Malignant tumor with ascites should be differentiated by detection of malignant cells on cellular morphology study and immunochemistry whenever necessary. It is important to understand that an ovarian mass combined with pleural and peritoneal effusion not always represents an advanced stage of malignancy [4]. CA125 values are also not reliable in distinguishing, as some benign pelvic tumors causing pseudo-Meigs syndrome is associated with elevated levels of the tumor marker, like ovarian cystadenomas, struma ovarii, uterine and broad ligament leiomyomas[5–10].

The case presented here is unique with respect to the large amount of ascitic fluid drained, nearly 25 liters. It is to be noted that Meigs syndrome or pseudo-Meigs syndrome can present with such large ascites. Sudden release of this fluid during surgical entry into the abdomen

can lead to sudden drop in blood pressure of the patient and cardio-respiratory collapse. Keeping this thing in mind, we drained some fluid preoperatively. During the surgery, before entering abdomen, we drained the remaining ascitic fluid slowly with a large bore catheter over 60 minutes.

In literature, very few reports have been published on pseudo-Meigs syndrome with such a big mass developing over a period of 8–10 years. It differs in some important respects. Firstly, the age of patient is below fifty, with a history of abdominal distension since 7–8 years back, much younger than when the majority of these tumors present. Secondly, we have not seen a case report with such a big ovarian tumor removed i.e. 42x31 cm size.

CONCLUSION

Pseudo-Meigs syndrome is a rare syndrome, but with a good prognosis. It should be included in differential diagnosis in women presenting with unexplained hydrothorax and ascites. Though a benign disease, prompt diagnosis should be made by ultrasound and subsequent tissue diagnosis. Suspicion of this syndrome will prevent undue delay in diagnosis and management of patients.

Author Contributions

Divya Dayanandan Kallarackal – 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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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What's with all the water? Diuretic use for cerebral edema due to ecstasy

Ruben Hummelen, Laus JMM Mulder, Adriaan Dees

ABSTRACT

Introduction: Ecstasy (XTC) is a widely used synthetic drug, which can cause severe complications. **Case Report:** A 19-year-old male who had visited a rave party and used XTC presented at the hospital with acute hyponatremia and seizures. Initial treatment with hypertonic saline failed to prevent new seizures. When the hyponatremia persisted and we concluded that the patient was also volume overloaded due to polydipsia, we decided to administer a low dose of a loop diuretic. Within hours, more than six litres of urine was produced and the patient made a complete recovery with normalization of the serum sodium level. **Conclusion:** The combination of XTC use and excessive fluid intake can result in hypervolemia, along with treatment refractory hyponatremia. This case report emphasises the potential benefits of diuretics in the management of XTC-related hyponatremia.

Keywords: Ecstasy, Hyponatremia, Syndrome of inappropriate ADH secretion (SIADH)

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INTRODUCTION

Ecstasy (XTC) or 3,4-methylenedioxymethamphetamine (MDMA) is an illicit synthetic drug. It is reported that XTC has been used by approximately 16 million Americans [1]. The drug is (ab)used for its euphoric and psychedelic effects. The desirable influences can be enhanced in electronic dance music or so-called rave party settings. Even though severe side effects of XTC are un-common, potentially lethal adverse events have been reported, including hyperthermia, renal failure, non-traumatic rhabdomyolysis, hyponatremia, cerebral edema, coma, mediastinal emphysema and severe hypoglycaemia [2–4]. Herein, we describe a case of acute, symptomatic hyponatremia due to the combination of XTC and polydipsia.

CASE REPORT

A 19-year-old male presented at the emergency department of the hospital on Saturday morning after a rave party. The patient arrived by ambulance after having had a seizure at the party. Other attendees of the

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party mentioned that the patient had consumed a large amount of water and (diluted) beer that night and had lost consciousness for a short time. On admission he was orientated with a maximum Glasgow Coma Scale (GCS) score of 15. He denied having used illicit drugs and his medical history was unremarkable. On physical examination the patient's blood pressure was 147/117 mmHg and the heart rate regular at 114 beats per minute. His pupils were mildly dilated. The cardiovascular and respiratory examination were normal, no crackles were heard. The patient, however, appeared hypervolemic. His face had an edematous appearance and he visited the toilet several times while awaiting the results of the initial investigations. Shortly after being admitted, he experienced tonic-clonic seizure, for which midazolam was administered. To rule out intracranial pathology, a brain computed tomography (CT) scan was performed which showed evidence of cerebral swelling (Figure 1). Laboratory test findings on admission were as follows: sedimentation rate in the first hour: 2 (normal range <15), serum hemoglobin 8.6 mmol/l (normal range 8.5–11.0), leucocytes $13.0 \times 10^9/l$ (normal range 4–10), sodium 119 mmol/l (normal range 135–145), creatinine 71 $\mu\text{mol/l}$ (normal range 65–110), osmolality 265 mOsmol/kg (normal range 275–300) and CK 1147 U/l (normal range <200). The serum ethanol level was 0.4 (normal range < 0.5‰), while the urine osmolality was 460 mOsmol/kg and the urine sodium level was 113 mmol/l. Urine toxicology screens for XTC and methamphetamine were positive. A diagnosis of acute, symptomatic hyponatremia associated with XTC intoxication was made. The patient was initially treated with hypertonic saline (50 ml of 3% NaCl per hour). The neurologist started the patient on intravenous Depakine to prevent new seizures. Several hours after admission, the patient's sodium level remained stable at 120 mmol/l. Meanwhile the patient experienced two additional seizures. Due to the patient's refractory clinical condition, we decided to treat him with a low dose loop diuretic, so 10 mg of furosemide was administered intravenously. A diuresis of 6900 ml in six hours then followed and the hyponatremia was gradually corrected, reaching 137 mmol/l the next morning (24 hours after admission). The patient was discharged from the hospital the following day, without any sequelae.

DISCUSSION

Acute, symptomatic hyponatremia associated with XTC intoxication, as presented here, is rare. This may in turn lead to cerebral edema and seizures with potential fatal consequences. Mechanisms include acute kidney injury secondary to non-traumatic rhabdomyolysis, and the syndrome of inappropriate anti diuretic hormone secretion (SIADH), which can be triggered by MDMA and its metabolites [4–6]. A study from Amsterdam showed that females are particularly susceptible to developing dilution hyponatremia, which may be related to the effects

of estrogen on anti-diuretic hormone (ADH) release [7]. Recent investigations have also confirmed that MDMA potentiates the effects of water loading on hyponatremia [8]. Since acute symptomatic hyponatremia requires urgent treatment, we treated the patient with hypertonic saline (3% at a rate of 50 ml/hour). The Adrogue–Madias formula was used to estimate the anticipated rise in the patient's serum sodium level following treatment [9]. This regimen was the preferred treatment at the time. In recent years, however, a 100 ml bolus of 3% NaCl has become the standard instead of a continuous infusion [4, 10]. The bolus alternative may facilitate a simpler and more rapid management in the emergency department. The use of isotonic saline solutions is not advised since it may exacerbate volume overload and ADH secretion.

Our patient, however, did not well respond to treatment. We realized that SIADH might not have been the sole mechanism in this case. Ecstasy as well as hyperthermia stimulate feelings of thirst. In addition, the advice and common belief at parties where XTC is consumed is to drink large amounts of fluids to avoid hyperthermia. For our patient, this meant that the consumption of hypotonic liquids could explain the refractory character of the course of the disease. The serum sodium level of 119 mmol/l would have developed on the night of the party. During this time the patient's total body water would have increased by a factor 1.18 (sodium 140/119) or 18%. In males, total body water is approximately 60% of body's weight. Following this logic, with the patient weighing 80



Figure 1: Computed tomography scan of the brain demonstrating diffuse swelling and edema.

kg, he would have had an intake of approximately 8.6 L (18% of 60 L). This estimation does not include the loss of body fluid from perspiration or urination during the time period in question. The patient had an edematous appearance on arrival and experienced an extensive diuresis after receiving a small dose of furosemide. Therefore, we believe it is likely that the patient had been in a hypervolemic state.

Patients with SIADH are usually euvolemic and fluid restrictions are the preferred treatment. In contrast with an euvolemic state, hyponatremia along with a hypervolemic state is usually observed in cirrhosis, heart failure and end-stage renal failure. The patient made a full recovery. The case confirms the clinical course of patients in previous reports. Loop diuretics, and even the osmotic agent mannitol, have been used to manage cerebral edema due to XTC before [2, 11].

CONCLUSION

The case demonstrates that in refractory hyponatremia due to ecstasy, a low dose diuretic can be considered to counter potential volume overload due to polydipsia.

Author Contributions

Ruben Hummelen – 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

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

Adriaan Dees – 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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Multicentric epithelioid hemangioendothelioma: An unusual case report

Liqa Al Mulla, Jawad Al Khalaf, Ayesha Ahmed, Areej Al Nemer,
Yasser El-Ghoniemy, Tarek M El-Sharkawy

ABSTRACT

Epithelioid hemangioendothelioma (EHE) is a rare malignant tumor of vascular origin. Most of the times it affects liver, lung and bones, although this kind of tumor may involve the head and neck area, breast, lymph nodes, mediastinum, brain and meninges, spine, skin, abdomen and many other sites. We report a case of a 59-year-old Jordanian man, who was found to have multiple lesions, in the liver, anterior abdominal wall and the lung. Due to the short duration between the presentation, and as most common reported EHE metastatic cases occur in the bone, we raise up the possibility of primary EHE multi-centric lesions from the beginning rather than metastasis.

Keywords: Epithelioid, Hemangioendothelioma, Multicentric

How to cite this article

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INTRODUCTION

Epithelioid hemangioendothelioma (EHE) is an uncommon low-grade malignant tumor of vascular origin that may develop in the soft tissue, lung, bone, brain, liver, and small intestine. However, the recent (2002) World Health Organization (WHO) classification does not strictly define these lesions as having intermediate behavior, but instead describes them as lesions that fall into the category of locally aggressive tumors and those with metastatic potential [1].

CASE REPORT

A 59-year-old Jordanian male who sought medical advice complaining of chronic fatigue, abdominal discomfort and weight loss. Abdominal examination revealed a huge nodular liver and moderate splenomegaly. The positive lab findings were leukocytosis, thrombocytosis, low Hg level with low MCV and MCH, high ESR, elevated alkaline phosphatase and GGTP. Computed tomography scan of chest and abdomen showed multiple bilateral hypodense nodules in the lungs, liver and spleen. He had been followed-up by cardiothoracic and oncology departments for many years. One month back, he presented to the surgical clinic with an anterior abdominal wall hyperpigmented lesion. Physical examination showed a dark grey firm nodule on anterior abdominal wall, measuring 1.5x0.5x0.5 cm. Histopathological findings reveal a dermal neoplasm formed of nests and cords of epithelioid and spindle cells embedded in fibro collagenous stroma (Figure 1A). The cells have abundant cytoplasm with occasional vacuoles and lumena, some of them have red blood cells (Figure

1B). There is low mitotic activity and mild to moderate pleomorphism. Some of the nuclei are grooved. Scattered eosinophils are present within tumor cells and in stroma.

Immunohistochemical stains show positivity for Cluster of differentiation (CD31) (Figure 2) CD34, factor VIII and focal positivity for S100 protein in neoplastic cells and negativity for cytokeratin (CK), carcinoembryonic antigen (CEA) and tumor protein 63 (p63).

A diagnosis of epithelioid hemangioendothelioma was made. Onemonthlater, when the liver biopsy was taken from an outside hospital, the slides were received and showed the same histopathological and immunohistochemical findings. In the same month, the right lung nodule was discovered, with similar findings histopathologically and immunohistochemically, consistent with diagnosis of epithelioid hemangioendothelioma. The patient was doing well after receiving imatinib that was discontinued later on because he was intolerant to it as he developed gastric upset. On October 2016, he developed severe jaundice, tense ascites, melena and due to respiratory failure he passed away (Figure 3).

DISCUSSION

This case of a 59-year-old male with epithelioid hemangioendothelioma presented with multiple lesions found sequentially in anterior abdominal wall, liver and lung.

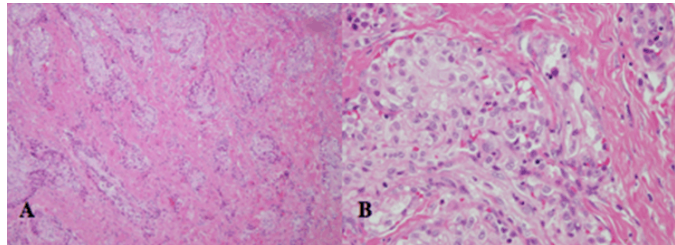


Figure 1: (A) Anterior abdominal wall skin nodule in the low power view showing nests and cords of epithelioid and spindle cells embedded in fibro collagenous stroma (H&E stain, x100). (B) The high power view of the abdominal skin nodule showing cells have abundant cytoplasm with occasional vacuoles, some of them have red blood cells (H&E stain, x400).

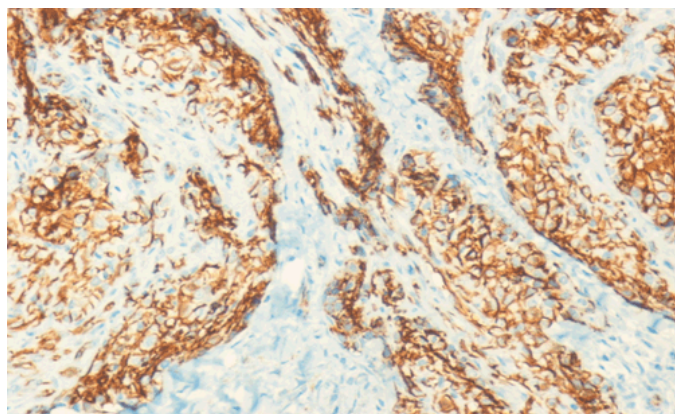


Figure 2: The tumor cells showing diffuse positive staining for CD31.

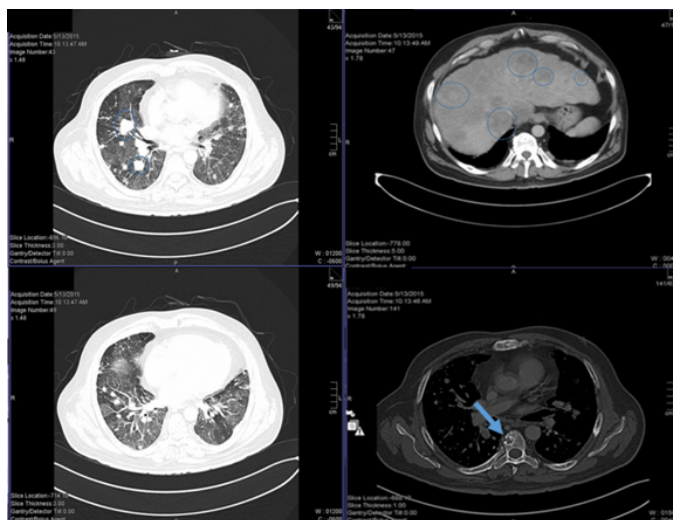


Figure 3: (A, B) CT chest with multiple bilateral pulmonary nodules. (C) CT abdomen showing multiple hypodense lesions in the liver. (D) CT chest showing osteolytic lesions in the vertebral body.

Most of reported cases of EHE in literature have single organ involvement. However, EHE can arise from many organs, including lungs, liver, bone, and soft tissue, simultaneously or sequentially. When this occurs, it may be difficult to determine if the tumor is multicentric from the beginning or if there is a primary lesion with metastases to the other organ tissue.

Hua Zhang et al. reported a case of a 20-year-old male, who presented with a right knee pain for eight months and diagnosed as EHE [2]. Lucas Rios Torres et al. reported a case of a 28-year-old female presented with a hypoechogenic hepatic nodule incidentally found at routine ultrasonography (US), discovered to be an EHE after hematoxylin and eosin and immune histochemical staining [3]. Muna M. Dahabreh et al. in 2011 reported a case of a 12-year-old with epithelioid hemangioendothelioma presented with simultaneously found multiple lesions in the lungs, trachea, liver and abdominal rectal muscle [4].

Jinghong et al. reported one case of a 20-year-old female with pulmonary epithelioid hemangioendothelioma accompanied by bilateral multiple calcified nodules in lung [5]. In 2010, Madhusudhan et al. reported a case of an 11-year-old boy with hemoptysis who was diagnosed with EHE simultaneously involving lung and liver [6]. Kalra et al. reported a case of a 70-year-old female with coexistent hepatic and pulmonary epithelioid hemangioendothelioma [7]. Al-Shraim et al. reported a case of a 51-year-old man with primary pleural epithelioid hemangioendothelioma with metastases to the skin [8]. Adher et al. in 2005 reported a case of a child with syncopal episodes who was found to have generalized multifocal EHE lesions in bones, lung, kidney and liver [9]. Besides, Kasteren et al. reported a single case of EHE which was misdiagnosed initially as lung histiocytosis but was later found to have multi-organ involvement at autopsy [10] (Table 1).

Table 1: Summary of multicentric epithelioid hemangioendothelioma cases reported in current literature.

Author	Year	Age	Sex	Organs involved	Histopathological findings
Hua Zhang et al. [2]	2015	20 year-old	Male	Bones and lung	Solid nests and anastomosing cords of epithelioid cells with a moderate quantity of eosinophilic cytoplasm. The cells were variably spindle-shaped and epithelioid, with a number of intracytoplasmic vacuoles.
Lucas Rios Torres et al. [3]	2012	28 year-old	Female	Liver, lung and bone	Neoplastic spindle and epithelioid cells infiltrating the sinusoidal structures, residual hepatocytes intermingled with neoplastic cells
Muna M. Dahabreh et al. [4]	2011	12 year-old	Female	Lungs, trachea, liver and skeletal muscle	Epithelioid to spindle shaped cells having large nuclei, prominent nucleoli and intracytoplasmic bubbly lumina. The cells were present in individual forms and in very small clusters embedded in a dense hyalinized stroma.
Jinghong et al. [5]	2011	42 year-old	Female	Solitary pulmonary and multiple calcified lung nodules	Small nests, cords and strands of epithelioid cells embedded in a highly eosinophilic stroma. Extensive tumor cell necrosis and stromal sclerosis were present. The epithelioid cells were mildly atypical, with polygonal to plump, abundant eosinophilic cytoplasm, irregular round nuclei, coarsely granular chromatin, and occasional nucleoli.
Madhusudhan et al. [6]	2010	11 year-old	Male	Lung and liver	—
Kalra et al. [7]	2009	70 year-old	Female	Liver and lung	Fibrotic tumor with variable sized vascular channels with high-grade mitotic figures. Tumor clusters were present within smaller branches of portal and central veins with sinusoidal extension.
H Al-Shraim et al. [8]	2005	51 year-old	Male	Skin and lung	Strips of dense fibrous tissue, with focal infiltration by nests and trabeculae of epithelioid cells. These cells showed irregularly round and rather uniform appearing nuclei, with open chromatin and a single prominent nucleolus. Many tumour cells showed cytoplasmic vacuoles or lumens .
Adher et al. [9]	2005	16 year-old	Male	Bone, lung, liver and kidneys	—
Kasteren et al. [10]	1995	10 year-old	Male	Lung, left kidney, liver, pancreas, adrenal gland, spine and paraaortic lymph nodes	Small foci of epithelioid cells with oval nuclei and abundant eosinophilic cytoplasm. The nucleus had a coarse chromatin pattern and one or more nucleoli. Anisokaryosis was prominent. The cells were localized around vessels and bronchioles. There was a sparse lymphoplasmocytic infiltrate, with a few eosinophilic granulocytes and many iron-containing macrophages.

Our case was initially presented with anterior abdominal wall hyperpigmented nodule, where skin biopsy was obtained and was confirmed to be EHE lesion. In less than one month, a lung nodule was discovered incidentally, and a tissue biopsy taken, in the same time where we received an outside slides of a liver biopsy, all consistent with the diagnosis of EHE.

CONCLUSION

Epithelioid hemangioendothelioma (EHE) is a rare tumor of vascular origin, where patients present with multi-nodular lesions involving more than one organ. Due to the short duration between the presentation, and as most common reported EHE metastatic cases occur

in the bone, we raise up the possibility of primary EHE multi-centric lesions from the beginning rather than metastasis.

Author Contributions

Liqā Al Mulla – 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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Authors declare no conflict of interest.

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Complete urogenital and colonic duplication: An extremely rare developmental anomaly in an adult female

Aarti Deenadayal Tolani, Deenadayal Mamta, Kadambari,
Nori Vijay Bhasker

ABSTRACT

Complete urogenital duplication is an extremely rare congenital syndrome, where the etiology is unambiguously not explained. Misexpression of certain genes and teratogenic factors are presumed to have a prominent effect on duplications and malformations at various stages during embryogenesis. Several cases have been reported on gastrointestinal and genitourinary duplication. However, no two cases described in literature are alike and they differ with the level of duplication and the associated anomalies. Interestingly, most of the cases reported are at infancy and adult caudal duplication syndrome (CDS) cases are exceptional. Herein, we report a case of a 36-year-old unmarried female with duplication of external genitalia associated with duplication of the genitourinary and colonic system. Imaging revealed associated skeletal and spinal anomalies. Patient did not

have any reconstruction surgeries previously and consulted us regarding the possibility of intercourse and fertility. The extent of urogenital duplication and associated anomalies was delineated with help of X-ray, abdominal and trans-perennial ultrasound and magnetic resonance imaging (MRI).

Keywords: Caudal duplication syndrome (CDS), Developmental anomalies, Duplication of external genitalia, Genitourinary and colonic duplication

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INTRODUCTION

Caudal duplication syndrome (CDS) is a very rare congenital abnormality that is associated with duplication and malformation of caudal structures comprising of the spine, the spinal cord, gastrointestinal and urogenital systems [1]. The etiology of CDS is not completely understood. However, it is assumed that misexpression of HOX genes encoding for the transcription factors that regulate the developmental process may have a prominent role in it [2]. The prevalence of this syndrome is less than 1 in 100,000 births and adulthood CDS reports are extremely rare. In this article, we present an adult female with caudal duplication syndrome.

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CASE REPORT

A 36-year old unmarried woman approached our clinic for evaluation of external genital malformation and possibility of intercourse in the future. A detailed physical examination revealed a double vulva with a 15-cm intervening area comprising of a pad of fat covered by hairless skin. Each vulva had pubic hair, a separate clitoris, urethra, vagina and anal orifice (Figure 1). The patient gave a history of passing two separate streams of urine during micturition, dual passage of feces from both anal orifices and menstruating through both the vaginas. She had an absent pubic symphysis and a non-surgical irregular scar on her lower abdomen (Figure 1) and there was no umbilicus.

Trans-abdominal pelvic sonography showed the presence of two separate bladders, two uteri and multiple follicles in both the ovaries. Two kidneys and two ureters along with colonic duplication with two separate rectums were observed.

Magnetic resonance imaging (MRI) scan (Wipro GE OPTIMA 360 advance 1.5 TESLA) results demonstrated normal kidneys, liver, gallbladder, biliary tract, pancreas, spleen, aorta, inferior vena cava, adrenals, mesentery and omentum. A single esophagus, stomach, duodenum and proximal small bowel with evidence of congenital duplication of the colon from the ileocecal junction ending in 2 separate ani were observed in MRI scan. It also showed duplicated bladders, vagina, uteri with double cervix and ovaries (Figure 2). Fibroids were noted in both the uteri, of which the one in the left uterus is larger (Figure 3). The distal sacrum showed partial agenesis. Ascites or abdominal lymphadenopathy was not observed. We also observed a clear tethering of the spinal cord with low insertion, dural ectasia in the lumbosacral spine and a split dural sac at sacral level and segmentation abnormalities of the lower dorsal vertebra.

Normal levels of FSH, TSH and prolactin were noted during the blood workup. Renal function test was normal.

DISCUSSION

Caudal duplication syndrome typically consists of a combination of several rare malformations and duplications of the distal organs derived from the hindgut, neural tube and caudal mesoderm. Although the specific reasons for caudal duplication are not well-known, it is presumed that an incomplete separation of monovular twins would be the one of the reasons. It is also believed that the impact of genetic, environmental and teratogenic factors influence the duplication of the embryonic cloaca and notochord at various levels during embryogenesis.

Even though caudal duplication syndrome is a rare condition, majority of the cases reported are soon after the birth. Reports on asymptomatic urogenital duplication in adulthood are uncommon. Early detection of malformation and duplication of organs may increase



Figure 1: External genitalia duplication. Double vulva with separate urethra, vagina and anal orifice.

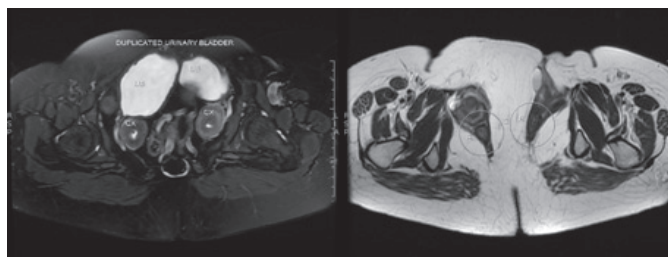


Figure 2: Magnetic resonance imaging scan (coronal plane) images showing duplication of urinary bladder, cervix and rectum. UB: Urinary bladder; CX: Cervix, R: Rectum.



Figure 3: Magnetic resonance imaging scan showing the duplicated uterus with fibroids in both the uteri. Left uterus has larger fibroid. UT: Uterus, FIB: Fibroid.

the chances of successful corrections, especially in cases of urogenital duplications. A study by Greenberg et al. (1997) [3] showed successful vaginal delivery of a patient where the cloacal malformation was repaired at infant stage. Very few cases of complete urogenital duplication in an adult are reported all around the world till date [4]. In this report, we presented a 36-year-old unmarried female with non-symptomatic complete external and internal duplication of the urogenital system with fibroids developing in both the uterus. We delineated the extent of the colonic, genitourinary and skeletal anomalies by using multimodal imaging. Duplication of the external genitalia in this particular case is highly unique with internal urogenital doubling and skeletal anomalies. While reconstructive genitourinary surgeries are performed to prevent complication and address the fertility. In this particular case, although the reproductive physiology, menstruation and gonadotropin levels are normal, due to the abnormal urogenital anatomy it is difficult to anticipate normal coitus. However, there has been case report published of successful cesarean delivery of an adult woman with duplication of the urogenital system [5, 6]. As the fibroids are asymptomatic patient was not advised any treatment. Since both the uterus has fibroids, this case raises the interesting possibilities of an association of the development of fibroids in anomalous uterus.

CONCLUSION

It is beneficial to diagnose of malformations and duplications of caudal structures in infancy to allow possible early surgical correction. Improved non-invasive imaging as seen here has opened an era of new possibilities for diagnosing, delineating and monitoring such complex anomalies both in childhood and adulthood for better treatment strategies in the future.

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Author Contributions

Aarthi Deenadayal Tolani – 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

Dr. Mamta Deenadayal – Substantial contributions to conception and design, Acquisition of data, Analysis and

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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Pediatric ovarian torsion in a nine-year-old girl: A twisted tale of pain

Mutiso Steve Kyende, Oindi Felix Mwembi

ABSTRACT

Introduction: Ovarian torsion is a rare gynecological emergency in children with a non-specific clinical presentation. It poses a diagnostic dilemma and quick diagnosis and management are key to good outcomes. We present a case of pediatric ovarian torsion and subsequently conduct a literature review outlining its salient features. **Case Report:** A nine-year-old girl presented with acute lower abdominal pain to the emergency department. She was worked up and tests were suggestive of ovarian torsion. She had surgical detorsion via laparoscopy and recovered well from surgery. **Conclusion:** Pediatric ovarian torsion is a rare disorder that offers a diagnosis and management challenge to gynecologists. The case presented outlines one such case and this may help in adding to the body of knowledge about this condition. It should always be considered in cases of acute unilateral lower abdominal pain in a girl and its quick diagnosis and conservative approach to management is associated with good outcomes.

Keywords: Conservative, Ovarian, Pediatric, Torsion

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INTRODUCTION

Ovarian torsion is an uncommon gynecological emergency in both children and adolescents. It occurs when the ovary twists on its ligamentous support with possible impediment of blood flow [1, 2]. Delay in diagnosis and treatment may lead to necrosis and loss of ovarian function. In addition, some patients may end up with infection, peritonitis and even death [3, 4]. The clinical presentation is largely non-specific, especially in the pediatric group of patients, posing a diagnostic challenge. Moreover, the clinical presentation mimics other acute abdominal conditions such as acute appendicitis and the various diagnostic imaging via Doppler ultrasound, computed tomography (CT) scan, and magnetic resonance imaging (MRI) scan may be equivocal [2]. Rare as it may be, ovarian torsion should be considered in any female child presenting with acute abdominal pain. We present a case of ovarian torsion management and subsequently conduct a literature review to discuss its features.

CASE REPORT

A nine-year-old African girl presented to the emergency department with a one day history of severe

left sided abdominal pain not relieved by analgesics. She had associated postprandial vomiting but no diarrhea, constipation, fever or urinary symptoms. Significant in her past medical and surgical history was an appendectomy done one year earlier. Besides being in pain, she was alert and not dehydrated. Her initial vital signs were pulse 117 beats/min, respiratory rate 24 breaths/minute, axillary temperature 36°C, and oxygen saturation 99% on room air. The abdomen was tender to palpation in the left lower quadrant with voluntary guarding. A urinalysis, full hemogram and renal function tests that were done were essentially normal.

An abdominal-pelvic ultrasound revealed an enlarged left ovary, 20.9 cm³ volume with no evidence of blood flow on Doppler studies favoring a diagnosis of ovarian torsion (Figure 1). She underwent an emergency laparoscopy where the left adnexa were found to be twisted and dark (Figure 2). The right ovary was examined during the surgery and found to be normal. The left adnexa was detorted (figure 3) but oophoropexy was not performed. However, on checking of the perfusion to the left ovary, it was found to be intact with fresh bleeding visible and change of color of the adnexal tissue to pink. A small incision on the ovary did not reveal any cyst or mass. The surgery was completed with no complications. Postoperative symptoms were pain on the port sites with mild nausea that was managed well with oral analgesics and antiemetics. The patient recovered well from the surgery and was allowed home on the second postoperative day. On follow-up after two weeks the patients had no symptoms and reverted fully.

DISCUSSION

Ovarian torsion accounts for approximately 3% of acute abdominal pain in women over 20 years of age [1] and 2.7% among those 1–20 years of age. Among this young populous, the estimated incidence of ovarian torsion is 4.9 per 100,000 females [5]. Occurrence is also more likely in the presence of ovarian masses, whether benign or malignant. The incidence is higher in the presence cysts greater than 5 cm in diameter. The normal pre-pubertal ovarian volume is 1–2 cm³ [4]. The patient had an estimated volume of 20.1 cm³ possibly predisposing her to torsion. However, not all cases of ovarian torsion occur in the presence of ovarian cysts. Indeed, the length of the suspensory ligaments and fallopian tubes can influence ovarian mobility and subsequent vascular compromise without any ovarian lesions as in this case.

Ovarian torsion majorly occurs on the right side (up to 60%) [1, 4] presumably because the sigmoid colon leaves little space for left adnexal movement and the fact that the right ovarian ligament is longer. The right preponderance of ovarian torsion makes differentiation from appendicitis difficult. However, the patient had left adnexal torsion and had previously had an appendectomy.

The clinical presentation of pediatric ovarian torsion is usually nonspecific [6]. The most common symptom is usually abdominal or pelvic pain that is variable in nature and usually unilateral [6]. This may have a previous history of occurrence and the pain may have disappeared only to reappear at a later date [7]. The pain may be common on the right side than the left due to the left adnexa having limited mobility due to the presence of the sigmoid colon [8]. The patient may also have constitutional symptoms such as nausea and vomiting due to peritoneal irritation [9]. Pelvic examination may be difficult to perform in the pediatric population and will usually be avoided so the only positive sign may be tenderness in the lower abdomen [10]. The current patient presented with abdominal pain and had nausea as a constitutional symptom and we did not perform a pelvic examination of the patient.

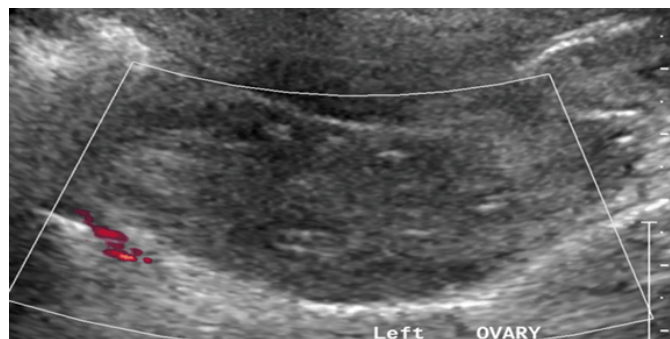


Figure 1: Ultrasound of the left ovary showing diminished Doppler flows.



Figure 2: Intraoperative view of the adnexal torsion before detorsion.

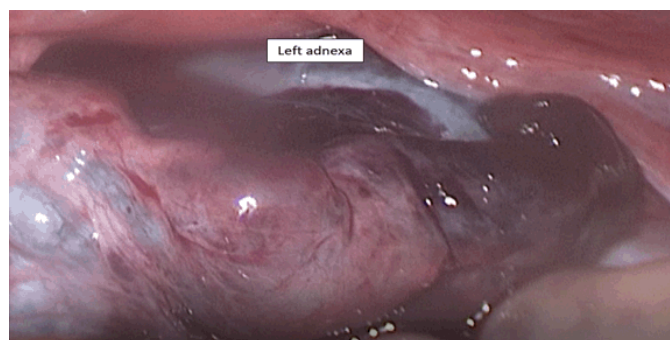


Figure 3: Left adnexa after detorsion.

Although laboratory tests may not be diagnostic of pediatric ovarian torsion, tests like a full hemogram, pregnancy test and kidney function tests may be useful as initial tests [6]. They will in most instances be normal but are useful in ruling out differential diagnoses such as pelvic inflammatory disease and ectopic pregnancy [7]. The laboratory tests conducted in the current patient were all normal and did not point to a certain differential. The best imaging test to diagnose pediatric ovarian torsion is usually a pelvic ultrasound with Doppler evaluation of ovarian blood flow [11]. The ultrasound examination may show a unilaterally enlarged ovary with a heterogeneous appearance due to edema and diminished or absent Doppler flows [12]. There may also be a whirlpool sign due to the twisting of ovarian pedicle and its vessels with peripherally displaced follicles due to ovarian edema [11]. The patient discussed had an enlarged ovary on ultrasound with diminished blood flow on Doppler which is in keeping with expected features. Adjuvant imaging such as computed tomography and magnetic imaging resonance at times may be needed to delineate ovarian anatomy further and rule out differentials although this methods are not commonly employed [6].

The management of pediatric ovarian torsion usually involves exploratory surgery as a first step [8]. This may be either through open surgery or laparoscopy, although laparoscopy is usually preferred where available [13]. Surgery may serve to confirm the diagnosis and as treatment in the same procedure. Although traditionally it was thought that oophorectomy was usually invariably needed, a more conservative approach is advocated for in recent literature [14]. The decision is usually based on intraoperative findings with oophorectomy done for tormented ovaries that have signs of necrosis. This is usually the case in just over half of the cases of pediatric ovarian torsion [15]. Conservative approach to management involves detorsion, ovarian cystectomy and preservation of the adnexa [9]. Oophoropexy which involves fixing of the ovary to the peritoneum, uterosacral ligaments or even the round ligaments may be considered in cases of recurrent torsion. This procedure is usually controversial but may aid in cases of re-torsion [16]. The patient was managed with simple detorsion and no oophoropexy was performed since it was the first episode.

The prognosis of pediatric ovarian torsion is good with ovarian function being reported to near normal at six week after conservative management [17]. Follow-up should be done at six week with ultrasound to check for ovarian function and presence of follicles. Further follow up may be needed at third month and then after every six months [6].

CONCLUSION

Pediatric ovarian torsion is a rare disorder that offers a diagnosis and management challenge to gynecologists. The case presented outlines one such case and this may

help in adding to the body of knowledge about this condition. It should always be considered in cases of acute unilateral lower abdominal pain in a girl and its quick diagnosis and conservative approach to management is associated with good outcomes.

Author Contributions

Mutiso Steve Kyende – 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

Oindi Felix Mwembi – 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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Unusual root canal anatomy in a maxillary second molar

Toshiko Inoue, Makoto Saito, Fumio Nishimura, Takashi Miyazaki

CASE REPORT

The micro-computed tomographic images represent unusual root canal anatomy in a maxillary second molar with two palatal roots and two buccal roots in a 53-year-old female (Figure 1A–F). The patient had no history of systemic disease; the maxillary second molar was extracted because of periodontal disease. Scanning was performed with an X-ray micro-computed tomography (micro-CT) system (SMX-90; Shimadzu, Kyoto, Japan). The tooth was imaged to reconstruct its structure.

The common root anatomy of maxillary second molars has been described as three roots with three canals [1]. However, the number of root canals and roots among teeth can vary. The prevalence of maxillary second molars with two palatal roots was only 0.4% in a radiographical survey of 1,200 teeth [2]. Although the incidence of maxillary molars with four roots is extremely low, this possibility should be taken into consideration during treatment.

DISCUSSION

Radiographs are one of the most important tools for detecting anatomical variations in clinical dentistry [3].

However, radiographs produce only two-dimensional images of a three-dimensional object, resulting in superposition of structures. Radiographic interpretation was confusing in this case because of the overlap of the buccal and secondary palatal roots, indicated by a white arrow in Figure 1D. Superposition of anatomical structures on X-ray images could result in failure to diagnose a distal palatal root canal, which, if left untreated, could result in failure of root canal treatment.

In recent years, significant noninvasive technological advances in dental imaging have been introduced, including digital radiography, densitometry, magnetic resonance imaging, ultrasound, and computed tomography [4]. In particular, micro-CT scan has been used to evaluate root canal anatomy because of its high resolution and non-destructive nature. The development of micro-CT scan is increasingly important in endodontic research because it offers a reproducible technique that can be applied quantitatively as well as qualitatively for the 3D assessment of the root canal system.

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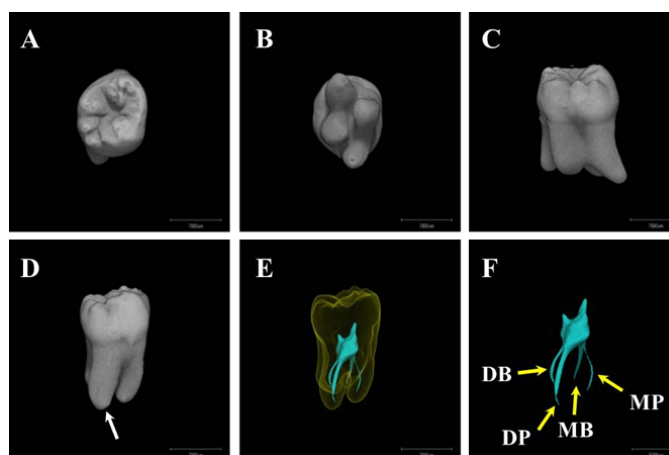


Figure 1: Three-dimensional reconstruction of a four-rooted maxillary second molar (A) Occlusal view, (B) Apical view, (C) Distal view, (D) Palatal view, (E) The external and internal structure, (F) Root canal morphology showing mesiobuccal (MB), mesiopalatal (MP), distobuccal (DB), and distopalatal (DP) roots.

The existence of extra roots in maxillary molars has clinical implications in endodontic treatment [5]. Its posterior location and the radiographic superimposition of anatomic structures are two important reasons for failure to diagnose a second palatal root canal [6]. Endodontic treatment can fail because of the presence of microorganisms remaining after insufficient canal obturation or the presence of untreated canals [7]. The main goal of endodontic therapy is to obtain 3D obturation of the root canal system after a sequence of cleaning, shaping, and filling procedures [8].

CONCLUSION

A thorough knowledge of root and root canal morphology and accurate anticipation of a tooth's possible morphological variations are essential for reducing endodontic failure caused by incomplete root canal preparation and obturation. Variations in the root and root canal morphology are a constant challenge for dentists. Dentists need to be familiar with the variations in root canal configurations for successful endodontic therapy. Micro-Computed tomography scan could be a useful tool for assessing root canal system anatomy in experimental endodontic studies.

Keywords: Micro-computed tomography, Molar, Root, Teeth

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Toshiko Inoue – 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

Makoto Saito – Substantial contributions to conception

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Acute thyromegaly in Hashimoto's thyroiditis mimicking lymphoma

Derick Adams

CASE REPORT

A 32-year-old female with a history of Hodgkin's lymphoma presented with a two week history of thyromegaly. Hodgkin's lymphoma was diagnosed at the age of 29 years and was treated with two cycles of ABVD (doxorubicin, bleomycin, vinblastine, and dacarbazine) chemotherapy and radiation therapy which was 2000 cGy in 10 fractions to the left lower cervical and supraclavicular nodal regions. One year after her diagnosis of lymphoma, she was considered to be in remission. She was also diagnosed with hypothyroidism with elevated anti-thyroid peroxidase antibody levels at the age of 29 years and treated with levothyroxine. At the age of 32 years, she developed thyromegaly causing dysphagia and hoarseness over a two-week interval. Physical examination revealed thyromegaly but no cervical lymphadenopathy. Due to her history of lymphoma positron emission tomography (PET) imaging was performed and demonstrated increased, diffuse fluorodeoxyglucose (FDG) uptake in the thyroid with right lobe being larger than the left (Figure 1). Ultrasound was also performed and showed thyromegaly especially on the right side but no thyroid nodules (Figure 2). Fine needle aspiration (FNA) of the right lobe was performed with flow cytometry of the needle washings (Figure 3). Flow cytometry of the needle washings did not show any clonal or aberrant populations of lymphocytes making lymphoma unlikely. Cytologic examination showed

a background of lymphocytes and lymphoid stroma consistent with Hashimoto's thyroiditis. Over the next two months the patient's thyromegaly, dysphagia, and hoarseness gradually resolved. She continued to be treated with levothyroxine for her hypothyroidism related to Hashimoto's thyroiditis.



Figure 1: Coronal view of positron emission tomography imaging showing increased, diffuse fluorodeoxyglucose uptake within the thyroid. The thyroid has been circled.

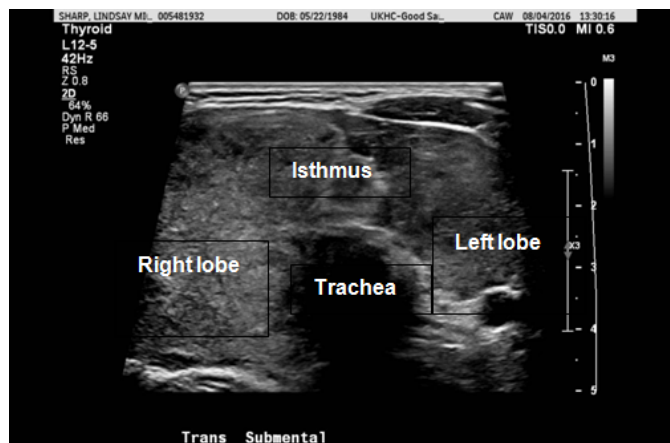


Figure 2: Transverse ultrasound image demonstrating thyromegaly surrounding the trachea in the center of the image. The right lobe, left lobe, isthmus, and trachea have been labeled.

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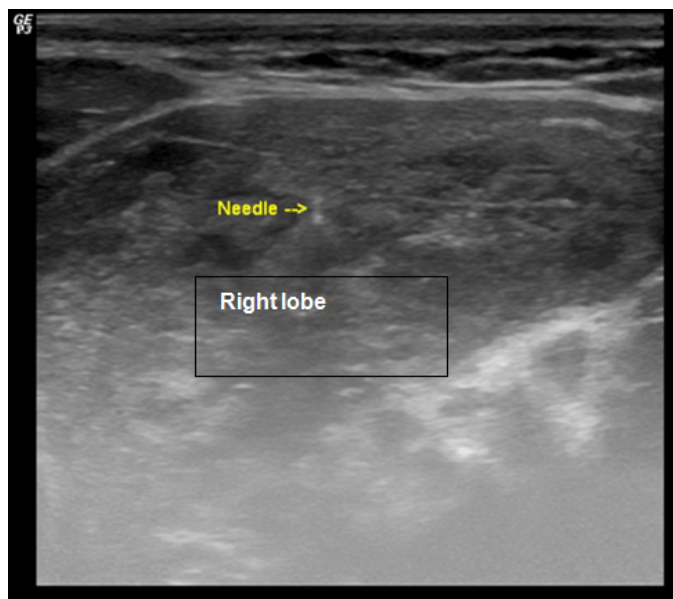


Figure 3: Transverse ultrasound image demonstrating needle during fine needle aspiration of thyroid. The needle and right lobe have been labeled.

DISCUSSION

This case illustrates how an atypical presentation of Hashimoto's thyroiditis can mimic thyroid lymphoma. Lymphoma of the thyroid classically presents as the acute onset of significant thyromegaly often with dysphagia or hoarseness. The risk of lymphoma of the thyroid is also increased by a factor of 67 in patients with Hashimoto's thyroiditis [1]. Clinicians should be aware that Hashimoto's thyroiditis may also present as acute thyromegaly. Due to this patients past history of lymphoma PET imaging was performed. Given the widespread use of PET imaging in some countries, clinicians should also be aware that up to 9% of patients with Hashimoto's thyroiditis can also have diffuse FDG uptake of the thyroid [2]. Therefore, FDG uptake in the thyroid must be interpreted with caution because this finding can represent both a benign and malignant process.

CONCLUSION

Hashimoto's thyroiditis may result in acute thyromegaly and increased fluorodeoxyglucose uptake on positron emission tomography imaging which may mimic the presentation of lymphoma of the thyroid. Fine needle aspiration of the thyroid with flow cytometry of needle washings can be used to distinguish lymphoma of the thyroid from Hashimoto's thyroiditis.

Keywords: Acute thyromegaly, Hashimoto's thyroiditis, Hypothyroidism, Lymphoma of the thyroid

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