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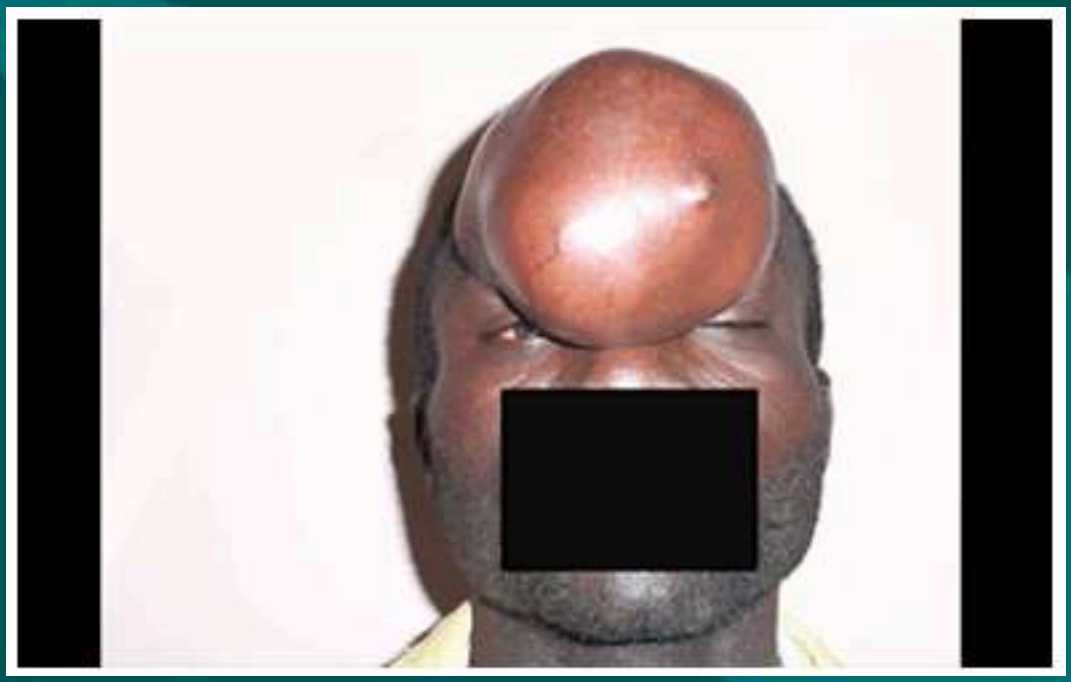

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CASE REPORT OPEN ACCESS

Massive recurrent fibrous histiocytoma of the frontonasal region: A case report

Fomete B, Agbara R, Adeola DS, Idehen OK

ABSTRACT

Introduction: Fibrous histiocytoma is the name for a group of lesions characterized by biphasic cell populations of fibroblast and histiocytes. Both benign and malignant forms have been described. The benign form often shows relapse following treatment and shows poor response to chemotherapy and radiotherapy. Therefore, radical excision or resection remains the therapeutic goal. Case report: Herein, we present a case of recurrent benign fibrous histiocytoma of the head and neck in a 25-year-old Nigerian male that was excised and skin grafted. Conclusion: Fibrous histiocytoma is less common in the head and neck region.

Keywords: Fibrous histiocytoma, Recurrent, Frontonasal

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Fomete B, Agbara R, Adeola DS, Idehen OK. Massive recurrent fibrous histiocytoma of the frontonasal region: A case report. International Journal of Case Reports and Images 2013;4(11):589–592.

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INTRODUCTION

Fibrous histiocytoma is a rare group of tumors with biphasic cell populations of fibroblast and histiocytes and which may exhibit benign or malignant features. Cutaneous and non-cutaneous forms have been described. Cutaneous fibrous histiocytoma refers to all superficial tumor of skin regardless of appearance while similar lesions involving the subcutaneous or deeper tissues are simply referred to as fibrous histiocytoma [1, 2].

In contrast to the malignant form, benign fibrous histiocytoma (BFH) of non-cutaneous tissues has received little attention in literature [1]. Less than 10 cases of non-cutaneous fibrous histiocytoma of the head and neck region have been reported in literature [1–4].

Specific sites of involvement, on the head and neck region, described in literature, include buccal mucosa, submandibular triangle, tongue, larynx, nasal cavity, mandible and supraclavicular fossa [4]. We found no occurrence at the frontal region.

Herein, we present a case of a 25-year-old Nigerian male with recurrent BFH, highlighting the challenges associated with oral surgical practice in a resource-poor environment.

CASE REPORT

A 25-year-old male carpenter was referred to our clinic from the surgical outpatient department on account of recurrent frontonasal mass of two years duration. Swelling was first noticed 10 years ago following trauma to the frontal region and gradually increased in size. The excision of mass was done twice in a peripheral hospital and the last excision was two years prior to the presentation. The forehead was the initial site of involvement but swelling gradually expanded with each recurrence to involve the nasal region. Except for headache, all other medical history was not significant.

Physical examination revealed a frontonasal mass involving both frontal regions but more to the right side, measures about 10 cm in its widest diameter, with some scares and some superficial vessels (Figure 1). Surface was smooth but lobulated, firm in consistency and not tender.

Swelling was attached to the underlying structures with involvement of the overlying skin. But the nasal cavity was free.

Fine needle aspiration cytology (FNAC) revealed a benign lesion, voluntary retroviral screening was negative, full blood count and electrolyte, urea and creatinine were within normal range.

Axial computed tomography (CT) scan revealed an extracranial mass of soft tissue origin with slight erosion of the frontal bone. The mass had a signal characteristics mimicking brain tissue.

Patient had excision of mass under general anaesthesia with delayed split thickness skin grafting two months post excision to allow for adequate granulation tissue formation and possible recurrence (Figure 2). He is currently on follow-up and doing well so far.

Postoperative histology report grossly, a skin covered sessile polyp measuring 13.5x9x9 cm and weighs 434 g. Cut surfaces showed grey solid areas.

Microscopy revealed a circumscribed dermal lesion lined by a focally atrophied, pigmented, keratinized epidermis. The lesion is composed of a mixture of plump spindle cells and histiocytes-like cells growing in a diffuse sheet. The stroma is abundant fibromyxoid to collagenized with foci of lymphocytic infiltrates and congested vascular channels. The deep margins appeared to be involved by the lesion (Figure 3).

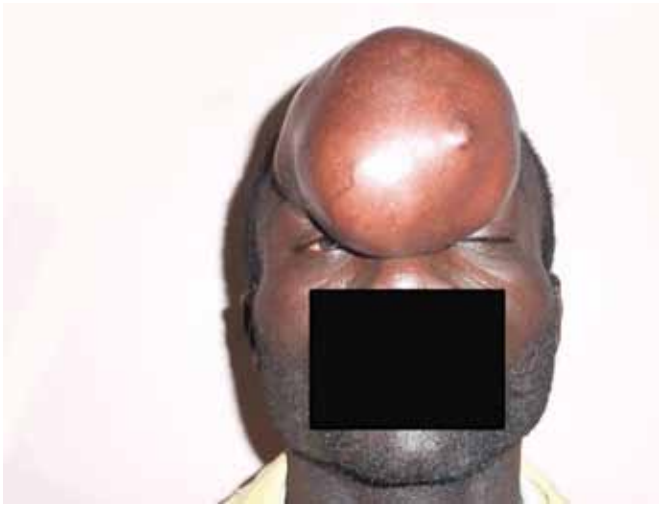

Figure 1: Frontal view of a 25-year-old male with recurrent benign fibrous histiocytoma.

Figure 2: Skin grafted wound.

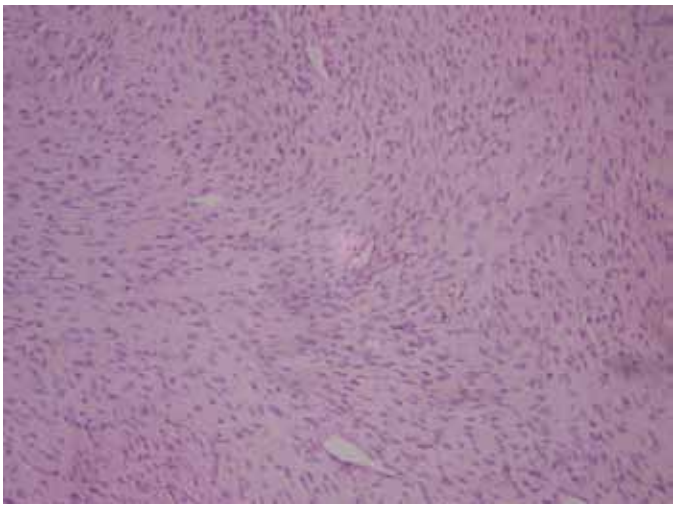

Figure 3: A mixture of plump spindle cells and histiocytes-like cells growing in a diffuse sheet (H&E stain, x100).

DISCUSSION

Benign fibrous histiocytoma was not recognized as a distinct clinical entity until the 1960s as a result of confusion regarding the natural history of fibrohistiocytic lesions. This confusion was settled following the development of immunohistochemical techniques and electronic microscopy [1, 5].

Benign fibrous histiocytoma occurs more in the upper and lower extremities and in the retroperitoneal region. Involvement of the head and neck region have been described and sites involved include buccal mucosa, submandibular triangle, tongue, larynx, nasal cavity, mandible and supraclavicular fossa [1]. This case is unique due to its site of presentation, the frontal region.

The occurrence of fibrous histiocytoma has been associated with trauma, sun exposure and chronic

infection. These have lead to the postulation that it represents a reactive proliferative lesion [4]. The patient in our case had a history of trauma to the frontal region following a fight, and noticed occurrence of initial mass one month later. The average duration of tumor occurrence in literatures ranges from 3–12 months.

The BFH is seen commonly in young or middle age adults with a male predominance. The reported male to female ratio ranges from 1.9:1 to 2.5:1 [6, 7]. The patient in our case was a 25-year-old male. Most patients presented to the hospital as a result of symptoms from interference with normal physiology of the involved area such as dyspnea, dysphagia, visual disturbance and headaches or present for esthetic reasons [1]. The patient in this case, reported a history of headache and was worried about his esthetics.

The diagnosis of fibrous histiocytoma is made primarily from histologic examination of tissue samples in which conventional microscopy shows a mixed population of spindle shaped fibroblast and rounded histiocytes in varying proportions [8, 9]. The lesion in our case was composed of a mixture of plump spindle cells and histiocytes-like cells growing in a diffuse sheet. The stroma was abundant fibromyxoid to collagenized with foci of lymphocytic infiltrates and congested vascular channels. The maximum diameter has been reported to range from 2–12 cm [4]. In our case it was about 13.5 cm in its widest diameter.

Electron microscopic and immunohistochemical studies though not specific, helps in differentiating BFH from aggressive fibrous histiocytic lesions such as malignant fibrous histiocytoma (MFH) and dermatofibrosarcoma protuberans [10]. With immunohistochemical studies, BFH shows a significant population of Xllla-positive cells with scanty CD4 positive cells. The reverse is the case with dermatofibrosarcoma protuberans [10, 11]. Other differentials to be considered are neurofibroma and leiomyosarcoma.

Treatment in most cases is essentially local excision since it is neither radiosensitive or chemosensitive. There is no report of metastases, thus the prognosis is generally good. Recurrence has been reported and incomplete excision is believed to be a reason [4, 12]. About 12 out of 46 cases of benign fibrous histiocytoma followed-up, with a mean follow-up of three years, recurred [6]. The patient in our case report had two recurrences, the second recurrence occurring within one year of excision both in a peripheral hospital. In view of recurrent potential, patient should be promptly refer to the specialist and prolong follow-up, if not for life.

CONCLUSION

Though less common in the head and neck region occurrence of benign fibrous histiocytoma may be associated with functional or aesthetic problems. Understanding its clinical behavior and histological features are vital in differentiating it from other lesions and in its management.

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Author Contributions

Fomete B – 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
Agbara R – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Adeola DS – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Idehen OK – 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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CONSENT

A written consent was obtained from the patient for the use of images and publication.

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CASE REPORT OPEN ACCESS

Tophi gout around the knee joint: An unusual presentation with a soft tissue mass

Feyza Unlu Ozkan, Kerem Bilsel, Ismail Turkmen, Mehmet Erdil, Salih Soylemez, Korhan Ozkan

ABSTRACT

Introduction: Gout is the most common inflammatory arthropathy. It has been reported to affect 2.13% of population in the United States. In this report, we presented an uncommon case of tophaceous gout of the knee presenting as a soft tissue mass. Case report: A 57-year-old male patient with knee pain and localized progressive swelling increasing in time on the medial side of the proximal tibia was seen in our clinic. He did not have any rheumatologic disease known previously except gout arthritis. Excisional biopsy was performed by preserving medial collateral ligament and histopathologic investigations were done next. A tophaceous gouty deposit was identified by low-power photomicrograph. A bluish amorphous material was seen surrounded by bundles of dense collagenized tissue and chronic inflammatory cells. Surrounding the amorphous crystalline deposit is a thin layer of mononuclear and giant cells. Photomicrograph of another section has been stained by de Galantha's method for demonstration of monosodium urate crystals. Conclusion: Especially in patients with extra-articular or subcutaneus mass, tophaceous gout must be considered as differential diagnosis.

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Keywords: Tophi gout, de Galantha's method, Tophaceous gout, Gout arthritis

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INTRODUCTION

Gout is the most common inflammatory arthropathy reported to affect 2.13% of population in the United States [1]. In this report, we presented an uncommon case of tophaceous gout of the knee presenting as a soft tissue mass.

CASE REPORT

A 57-year-old male presented with knee pain and localized progressive swelling increasing in time on the medial side of the proximal tibia. Initially, he was examined by a physiotherapist and treated conservatively. After that he had come to our polyclinic with increased complaints. Physical examination of the knee revealed well circumscribed, solid, measuring approximately 5x5 cm, non-mobile soft tissue mass. The patient has no limitation of the joint motion. The mass was on the proximal tibial metaphysial region, it was located subcutaneously and vicinity of bone. It was hard and fixed with smooth border by palpation. The blood uric acid levels were in normal range. His past history revealed that the patient had been using colchicine for 15 years. A year before, he had traffic accident and had injured his right knee with no fracture. In radiological assessment (X-ray

and magnetic resonance imaging (MRI)) a fusiform shaped mass was determined on pes anserinus bursa which had stretched patellar retinacula (Figure 1A–C). Calcific myonecrosis, pigmented villonodular synovitis, calcinosis, gout tophus and especially synovial sarcoma were considered as differential diagnosis.

An excisional biopsy was planned. A longitudinal incision was made along the length of anteromedial side of proximal tibia. Tumor mass was completely excised with insertion of medial collateral ligament. There was an eroded area underlying the mass on proximal medial tibia but without a lytic area. The tumor and the eroded area of the bone extirpated via a curved osteotome. The remaining (proximal) part of medial collateral ligament was reattached to the tibia by suturing using fiber wire through previously prepared drill holes.

Histopathologic investigations were done next. A tophaceous gouty deposit was identified by low-power photomicrograph. A bluish amorphous material is seen surrounded by bundles of dense collagenized tissue and chronic inflammatory cells. The field examined by polarized light. The birefringence of the crystalline material was evident. Surrounding the amorphous crystalline deposit was a thin layer of mononuclear and giant cells. Photomicrograph of another section had been stained by de Galantha's method for demonstration of monosodium urate crystals (Figure 2).

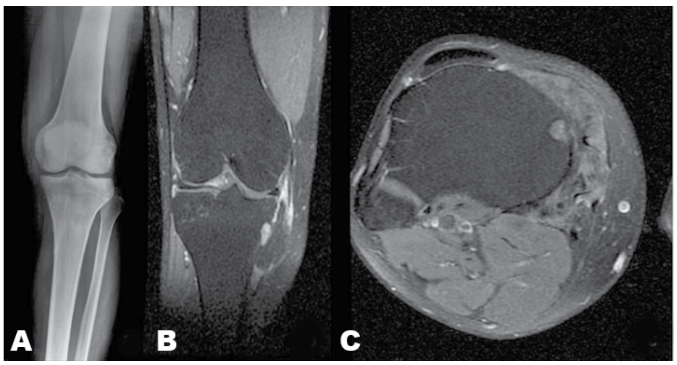

Figure 1: (A) X-ray and (B,C) Magnetic resonance imaging (MRI) scan of the mass. Anteroposterior diameter 54 mm, craniocaudal length 48 mm, mediolateral diameter: 12 mm.

DISCUSSION

Gout is an inborn error of purine metabolism characterized by hyperuricemia and recurrent attacks of acute arthritis. If hyperuricemia persists for a long time, tophaceous deposits may be found in the subcutaneous tissues and the various joints, particularly the first metatarsophalangeal joint, the hand, wrist, or elbow [2–5]. Tophaceous deposition has been reported in various locations such as finger pads, Sacroiliac joint, carpal tunnel, ankle, shoulder, dorsum of the feet, multiple

Figure 2: Macroscopic and microscopic histological images of the mass.

subcutaneous nodules, knee, acromioclavicular joint and axial skeleton [3–14]. A recent study suggested that the frequency of axial involvement may be as high as 14% in patients with clinical or crystal-proven gout [15]. The rate of appearance of gross tophaceous deposits is a consequence of the gout disease and the degree of hyperuricemia [6].

Patients might have such complaints; warmth, pain, swelling and extreme tenderness in a joint, limited motion in the affected joint which mimics septic arthritis symptoms [4]. Chronic tophaceous gout classically occurs after 10 years or more of recurrent polyarticular gout. However, demonstrating tophi as the initial clinical presentation of gout is very rare [8]. Subcutaneous tophi generally occurs as a late clinical outcome and typically located in the peripheral joints of the hand or foot [10]. But subcutaneous tophaceous deposits of monosodium urate, in the absence of arthritis, may occasionally occur as the initial manifestation of gout. In 1996, Iglesias et al. reported a case presented with a 6-year history of multiple subcutaneous nodules and no history of previous articular complaint [16]. They had reviewed literature and found out 28 similar cases that had subcutaneous nodules without any articular complaint and termed this entity of the disease as the 'gout nodulosis'. Bloch et al. in a study review presented 466 patients who had gout arthritis retrospectively [6]. In 84 patients (18%), radiographic findings were positive, but rather suspicious clinically. Thus, it seems that tophi deposition may occur early, even in previously unaffected joints. An earlier correct diagnosis of tophaceous gout could be made incidentally during an arthroscopy or with the help of the radiologist [2, 6].

Tophi are rarely observed in patients without a prior history of gouty arthritis. We describe a patient whose

initial manifestation of gout was tophaceous deposition in an unusual location; medial side of the knee. As far as we know although intra-articular gouty deposits in the knee are common, subcutaneous nodules are rare [10]. Our patient did not have a history of acute gouty arthritis tophi elsewhere. Tophaceous gout without arthritis might be more common than previously recognized. For diagnosis of gouty deposits in and around the knee plain radiographs, MRI and computed tomography (CT) [5, 9, 17] scan can be used. The plain radiographs show asymmetrical soft tissue swelling, calcification and bone erosion [17]. These plain radiographic features generally are normal in early and even chronic gout patients with intra-articular deposits and bone erosions [6]. Tophaceous deposits present as masses on MRI scan with low to intermediate signal intensity on both T1- and T2-weighted images and a characteristic enhancement pattern following intravenous Gd administration. These features relate primarily to internal calcifications, which are most evident on CT scans. Magnetic resonance imaging scan (including Gd administration) supplemented, in some cases, with CT scan allows accurate diagnosis of intra-articular tophaceous deposits [9]. Monosodium urate (MSU) deposits within a tophus can be clearly defined with CT scan. Computed tomography scan discloses round and oval opacities in the tophi [17].

Gout is marked by transient attacks of acute arthritis initiated by crystallization of urates within and about the joints [18–21]. Although peri- and intra-articular structures are involved in the knee visible subcutaneus lesions are extremely unusual [22]. The soft tissue lesions of the tophaceous gout can have similar appearances to calcinosis of chronic renal failure, synovial sarcoma, osteosarcoma, calcific myonecrosis, myositis ossificans and tumoral calcinosis, so medical history of the patient is essential to evaluate the origin of these lesions and to detect the possible etiology.

Synovial sarcomas are most commonly seen in large joints. They originate from bursal and tendineus structures [23]. These were, most frequently, incorrectly diagnosed as bursitis, tendinitis, synovitis or hematoma. In tumoral mass there may be mineralization and also this calcification can be also seen on X-ray.

In some cases the underlying bone is affected and erosions can be seen [24]. Due to this features, in our case there was a diagnostic dilemma between synovial sarcoma and gout tophi. Computed tomography scan should be applied preoperatively to detect bone erosions that could help to plan the surgery.

CONCLUSION

In such cases, especially in patients with extra-articular or subcutaneous mass, tophaceous gout must be considered as differential diagnosis because tophi gout can mimic all kind of mass on extremities.

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Author Contributions

Feyza Unlu Ozkan – 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

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

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

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

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

Korhan Ozkan – 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 OPEN ACCESS

Superior mesenteric artery syndrome: A rare case of bowel obstruction in schizophrenic patient

Ing How Moo, Michael John Clarke, Tiong Thye Goo, Kee Wee Lim

ABSTRACT

Introduction: Superior mesenteric artery syndrome is a rare condition of obstruction to the third part of duodenum. Identification of this syndrome is challenging and is frequently made by a process of exclusion. Case Report: We report the first case of superior mesenteric artery syndrome in a severely cachectic schizophrenic patient with paranoid delusions who presented with features of proximal gastrointestinal obstruction. The diagnosis was not straightforward and was supported by radiologic evidence of obstruction. The patient was treated conservatively with nasogastric tube decompression, fluid and electrolyte correction and enteral nutrition via nasojejunal tube. However, the patient was not cooperative with treatment and subsequently demised. Conclusion: This report aims to strengthen the awareness of this condition to prevent delay in diagnosis and treatment of this life-threatening condition.

Keywords: Superior mesenteric artery (SMA) syndrome, Schizophrenic patient

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INTRODUCTION

Superior mesenteric artery (SMA) syndrome is characterized anatomically by extrinsic compression of the third part of the duodenum by the superior mesenteric artery anteriorly and abdominal aorta posteriorly. It is one of the rarest gastrovascular disorders with incidence of 0.013–0.3% [1]. Only about 400 cases have been reported in medical literature. Most clinicians are unfamiliar with this disease. Delayed diagnosis of SMA syndrome is associated with significant morbidity and mortality [2]. Herein, we report the first case of SMA syndrome in a schizophrenic patient.

CASE REPORT

A 64-year-old schizophrenic elderly male was admitted in Singapore government hospital for epigastric pain of 1 day duration associated with a two-day history of vomiting. The character of the vomitus was uncertain. He had lost 30–40 kg of weight over several months. The patient denied postprandial pain and had no previous similar episode. He had no past history of surgery and was not on any psychiatric medication.

The patient had paranoid schizophrenia but was never compliant with medications. He lived alone and had persecutory thoughts. He was suspicious of people

around him and barred all windows and rooms. He also mentioned cameras watching him and "people" want to harm him. He refused to go out and consumed mainly instant noodles for several years. One month before this admission, he was admitted for hypoglycemia secondary to poor oral intake and incidentally had low calcium and vitamin D deficiency. His vital signs were stable on admission. He appeared comfortable but was severely cachectic with a body mass index of 10.9 kg/m^2 (weight 34.4 kg, height 178 cm). The abdomen was scaphoid, distended and tender over epigastric area with no sign of peritoneal irritation and succussion splash was positive. Per rectal examination was unremarkable. Initial laboratory studies were largely unremarkable besides the elevated total white, low creatinine and urea.

Supine and erect plain abdominal radiograph showed a grossly distended stomach and collapsed bowel distally (Figure 1). In view of the distended abdomen, positive succession splash and dilated stomach on abdominal radiograph, our initial impression was gastric outlet obstruction. Patient was kept nil by mouth and nasogastric tube was inserted to maintain gastric and duodenal decompression. Nasogastric tube immediately aspirated 2.2 liters of bilious fluid. Upper gastrointestinal series showed distended stomach and duodenum to the level of mid third part, where an abrupt cut-off occurred (Figure 2). To determine the aetiology, contrast-enhanced computed tomography (CT) scan of the abdomen was arranged and the third portion of the duodenum was clearly 'pinched' by the abdominal aorta and the superior mesenteric artery. The aorta-SMA angle was 4 degrees and the aortomesenteric distance measured 5.6 mm (Figures 3 and 4). In addition, CT scan also revealed obvious reduction of the intra-abdominal fat. There was no free intraperitoneal air.

Based on the history, examination and imaging findings, we diagnosed SMA syndrome. Endoscopic guided insertion of nasojejunal tube was performed for enteral feeding as patient was unable to tolerate any oral intake due to duodenal obstruction (Figure 5). However, the patient was not cooperative with nasojejunal tube feeding. Subsequently, he developed hypoglycemia, hypokalemia and hypotension that was refractory to resuscitation. Consciousness deteriorated and the patient passed away.

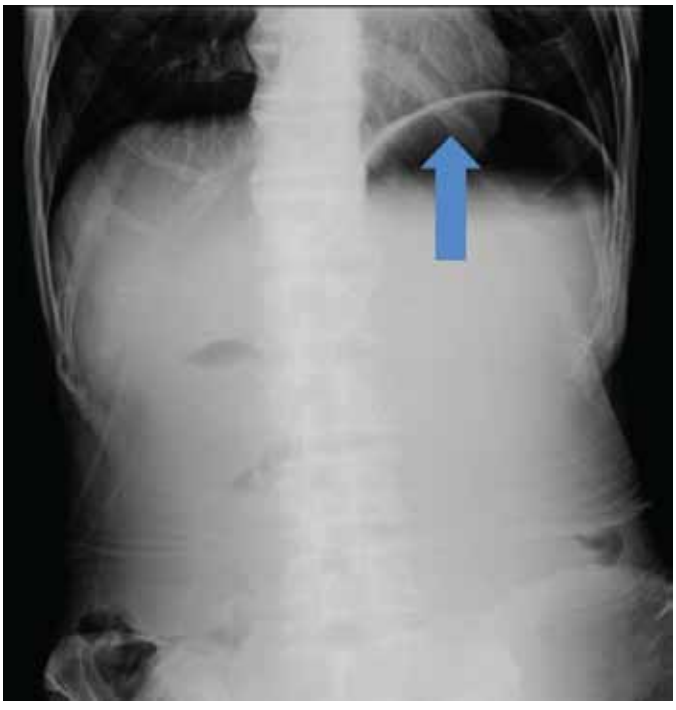

Figure 1: Supine abdominal X-ray showing grossly distended stomach (arrow) and paucity of gas distally in collapsed bowel.

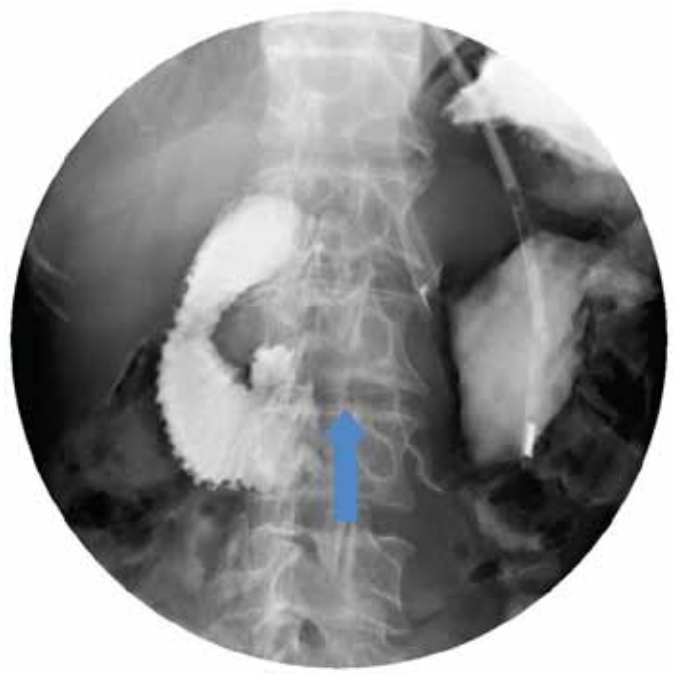

Figure 2: Upper gastrointestinal study with water soluble contrast introduced via a nasogastric tube. This shows a distended stomach and proximal duodenum with a transition point in mid transverse duodenum (arrow).

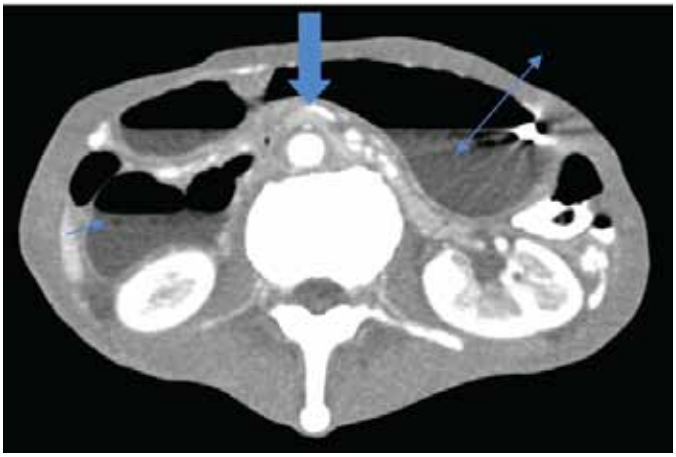

Figure 3: Contrast enhanced axial computed tomography at level of D3. Stomach is distended with nasogastric tube in situ (double headed arrow), D2 and proximal D3 are dilated (short arrow) and the mid D3 is collapsed ventral to the aorta (open arrow). Note also the cachectic body habitus.

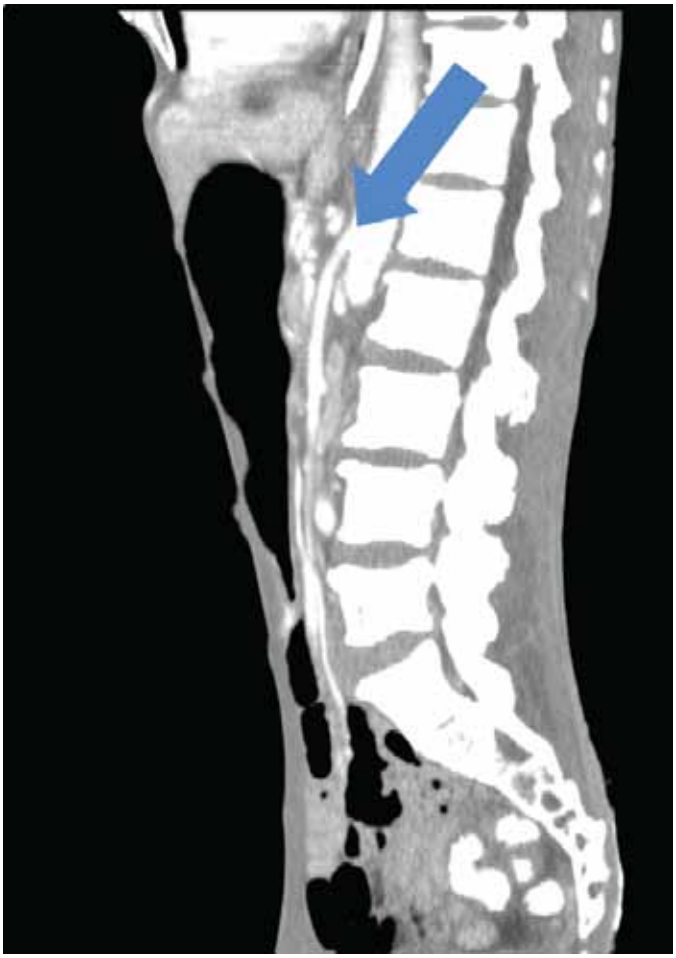

Figure 4: Contrast enhanced CT in sagittal plane. The superior mesenteric artery (arrow) makes an acute angle with the aorta at 4 degrees. Note severe paucity of retroperitoneal fat.

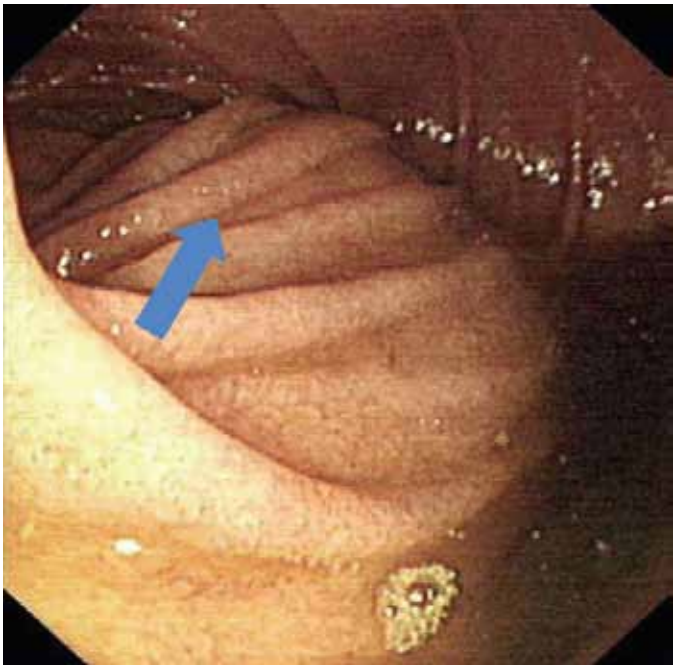

Figure 5: Esophagogastroduodenoscopy demonstrating intraluminal view of the third portion of the duodenum with superior mesenteric artery clearly bulging (arrow) and narrowing the lumen.

DISCUSSION

Superior mesenteric artery syndrome was first described by Austrian Professor Carl Freiherr Von Rokitansky in 1861 [3]. Later Wilkie published the first comprehensive case series of 75 patients with what he initially called "duodenal ileus", 64 of who underwent duodenojejunostomy. Wilkie's detailed anatomical, clinical and pathophysiological description of extrinsic compression of third portion of duodenum by the superior mesenteric artery has become a common eponym for SMA syndrome [4]. Subsequently, a variety of other names have been used such as chronic cast syndrome and arteriomesenteric duodenal compression syndrome [5, 6].

Superior mesenteric artery originates acutely from the abdominal aorta behind the neck of the pancreas at the level of first lumbar vertebra and travels caudally into the root of mesentery. The transverse portion of the duodenum crosses anterior to the third lumbar vertebra and was separated from the superior mesenteric artery by the retroperitoneal fatty tissue. The normal angle between abdominal aorta and the superior mesenteric artery is 25–60° and the aortomesenteric distance is around 10–28 mm [7, 8]. Such relationship correlates with body mass index [2].

Pathophysiological loss of retroperitoneal and paraduodenal fats can result in aortomesenteric distance less than 8 mm and aorto-SMA angle of 22° or less, resulting in duodenal 'clamping'. Such pathophysiology can be seen in chronic wasting disease, catabolic state, anorexia and malabsorption. Surgical interventions can occasionally alter the anatomical relationship, jeopardizing the aortomesenteric angle and resulting in SMA syndrome (e.g., scoliosis surgery, aortic aneurysm repair, bariatric surgery) [1, 5].

The SMA syndrome can present acutely or chronically with signs and symptoms of proximal gastrointestinal tract obstruction (i.e., nausea, vomiting, weight loss, sense of repletion and postprandial abdominal distension). Such presentations, however, are nonspecific to SMA syndrome and diagnosis is frequently delayed. Henceforth, a high index of suspicion is required and a comprehensive investigation is recommended to rule other conditions that are common and has different treatment implication including pancreatitis and peptic ulcer disease [9].

Upper gastrointestinal series, contrast-enhanced computed tomography (CT) scan, magnetic resonance angiography (MRA), ultrasound (US) and endoscopy are modalities that can be utilized to establish diagnosis. Our patient had upper gastrointestinal series that revealed a classical dilated proximal duodenum with abrupt contrast cut-off at the transverse portion of duodenum. Computed tomography scan of abdomen with contrast can clearly demonstrate the obstruction site, determine the aetiology of the obstruction and allows evaluation of aortomesenteric angle and aortomesenteric distance,

both of which are sensitive measures of diagnosis. An aortosuperior mesenteric artery angle of less than 25° and aortomesenteric distance less than 8 mm are highly suggestive of SMA syndrome [9]. In our patients, both parameters were reduced with angle and distance of 4° and 5.6 mm, respectively. Upper endoscopy can be used to rule out stenosing lesions in the duodenum. With the advent of noninvasive radiological studies, conventional arteriography is rarely needed in the workup of SMA syndrome except in cases where diagnosis is not clear.

The treatment of SMA syndrome can be either conservative or surgical. Patients with SMA syndrome initially require nasogastric tube insertion for gastrointestinal decompression, fluid resuscitation, correction of electrolyte abnormalities and early nutritional support. Nutritional support aims to promote body weight gain and restore the retroperitoneal fat tissue with subsequent increase in aortomesenteric angle and reduction in duodenal obstruction. Nasojejunal tube that is placed distal to the obstruction allows enteral administration of nutrition. Parenteral nutrition may be an alternative but it is not without its associated complications. Notably, patient should be monitored for refeeding syndrome during nutritional rehabilitation as these patients are malnourished and are susceptible to electrolyte and fluid shifts. Duration to achieve symptomatic improvement is variable in conservative nutritional treatment and has been documented to range from 2–169 days [1]. Patients with shorter history of SMA syndrome have higher success rate with conservative management. On the other hand, those with more chronic history have a prolonged hospital stay with low success rates of conservative treatment alone and surgery after a period of refeeding and weight gain is indicated [8].

Several surgical options have been proposed to resolve or bypass the duodenal compression including Strong's procedure (caudal mobilization of duodenojejunal flexure by division of ligament of Treitz), gastrojejunostomy and duodenojejunostomy. The advantage of Strong's procedure includes maintaining bowel integrity, easier and quicker to perform and less invasive and is a safer procedure [9]. However, Strong's procedure has high failure rate of 25% and presumably due to difficulty in mobilizing the duodenum with interference from intra-abdominal adhesions and the short vessels of the inferior pancreaticoduodenal artery. Gastrojejunostomy can decompress the stomach adequately but it does not resolve the duodenal obstruction and the patient can have persistent symptoms. Furthermore, such bypass operation can cause blind loop syndrome, gastric bile reflux and peptic ulceration that necessitate second operation (i.e., duodenojejunostomy). Duodenojejunostomy is the surgical treatment of choice and can be performed with or without division of fourth portion of the duodenum. It has a reported a success rate of 90% [10].

CONCLUSION

Diagnosis of superior mesenteric artery syndrome requires a high degree of clinical suspicion supported by radiologic evidence of obstruction. Its nonspecific presentation often creates confusion amongst clinicians and prompts them to investigate for other conditions and lead to delay diagnosis. As demonstrated in our case, unrecognized or inadequately treated cases may progress to dehydration, electrolyte abnormalities, severe malnutrition, bowel perforation, and death. Multidisciplinary team approach comprises surgeon, gastroenterologist, psychiatrist, radiologist, dietician, pharmacist and social worker provides the most beneficial diagnostic and therapeutic result in this often underestimated disease.

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Author Contributions

Ing How Moo – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Michael John Clarke – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Tiong Thye Goo – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Kee Wee Lim – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Müllerian adenosarcoma of cervix in a young nulliparous woman

Sameera Begum Kader Ibrahim, Prasanta Kumar Deka, Tham Seng Woh

ABSTRACT

Introduction: Müllerian adenosarcoma is a rare tumor of the cervix. Typical adenosarcoma presents as a large polypoid mass occupying the endometrial cavity. It occurs mainly in postmenopausal women. It is a very rare occurrence in adolescent girls and young adults. To date, this neoplasm has been reported in only 10 young adult and adolescent girls. The tumor tends to recur locally rather than to disseminate to distant areas. Case Report: A 23-year-old female, single, sexually not active and nulliparous presented to a private hospital for complaints of prolonged menstruation of six months duration. Speculum examination revealed cauliflower like growth arising from the cervix. Polypectomy and hysteroscopic diagnostic dilatation and curettage was done. Histopathology of the specimens showed cervical polyp with features of well-differentiated Müllerian adenosarcoma of low grade with endometrium in secretory phase. A total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and paraaortic lymphadenectomy, appendicectomy were done. Conclusion: Adenosarcoma most commonly arises from the endometrium, but some cases from the endocervix. In postmenarchal adolescents, these polypoid tumors usually arise from the cervix, in contrast to postmenopausal women in whom they usually arise from the endometrium. Cervical Müllerian adenosarcoma usually treated by radical hysterectomy even in younger age group as there is higher chance of recurrence later following conservative surgical management. The presence of malignancy should be always kept in mind while resecting a cervical polyp.

Keywords: Müllerian adenosarcoma, Endometrium, Cervix, Polyp, Vaginal bleeding

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INTRODUCTION

Müllerian adenosarcoma of the uterus is a rare tumor containing benign glandular epithelial and malignant mesenchymal elements. Typical adenosarcoma is a low-grade tumor, presents as a large polypoid mass occupying the endometrial cavity and may protrude into the vaginal cavity [1]. This tumor was first described by Clement and Scully in 1974 as Müllerian adenosarcoma. It occurs mainly in the uterus of postmenopausal women but can occur in adolescents and young adults [2]. Extrauterine

locations such as ovaries, cervix, vagina, peritoneum and Pouch of Douglas also had been reported [3–6]. To date, this neoplasm has been reported in only 10 young adult and adolescent girls. There is a case report of a 10-year-old girl, the youngest female ever reported, who was diagnosed with Müllerian adenosarcoma arising from the endocervix [7]. We report one case of Müllerian adenosarcoma of the cervix because of its rarity.

CASE REPORT

A 23-year-old female, single, sexually not active and nulliparous presented to a private hospital for complaints of prolonged menstruation of six months duration associated with continuous dull suprapubic pain with gradual abdominal distension. There was a history of scanty, thick, yellowish, foul-smelling vaginal discharge with pruritus on and off not responded to medications. She was admitted in a private hospital three times in last six months due to symptomatic anemia and received blood transfusion during each admission. There was no significant past medical or surgical illnesses and no family history of malignancy.

On general examination, severe pallor was present. Other vital signs were normal. Breast and thyroid examinations were normal. On per abdominal examination, no abnormality was detected. Initial trans-abdominal ultrasound scan did not reveal any abnormalities.

During the third admission, a trans-abdominal ultrasound scan was repeated which showed endometrial thickness of 18 mm. Consent was taken for vaginal examination, hysteroscopic diagnostic dilatation and curettage under general anesthesia. Speculum examination revealed a cauliflower like growth measuring about 6x6 cm arising from the cervix and a diagnosis of cervical polyp was made. Bimanual examination revealed normal sized uterus with no adnexal pathology. Polypectomy and hysteroscopic dilatation and curettage was done in view of cervical polyp and thickened endometrium (Figure 1). Histopathological report showed a polypoid fragment with leaf like structures which is composed of hypercellular stroma lined by single layer of benign looking epithelial cells. There are scattered glands within a hypercellular stroma with the cells displaying oval to spindle-shaped nuclei with mild pleomorphic and distinct nucleoli (Figures 2 and 3). Around three mitotic figures were noticed in ten high power fields. Immunohistochemical stains showed the mesenchymal cells are diffusely positive for CD10 (Figure 4), Actin (Figure 5) and Desmin panCK (Figure 6) which highlighted the epithelial component. No tumor necrosis, heterologous components or lymphovascular permeation were identified. The diagnosis of cervical polyp with features of well differentiated Müllerian adenosarcoma of low grade with endometrium in secretory phase was given. She was then referred to our hospital —Government hospital Melaka— for further management. Computed tomography (CT) scan of abdomen and pelvis was done which showed an enlarged uterus with hydrometra and involvement of cervix could not be ruled out as it was bulky. There were bilateral ovarian cysts measuring less than 3x3 cm and no ascites. No evidence of distant metastasis.

The diagnosis of adenosarcoma was revealed to the patient and her parents. After a thorough evaluation of available literature and extensive discussions with the family members, a decision was made to perform radical

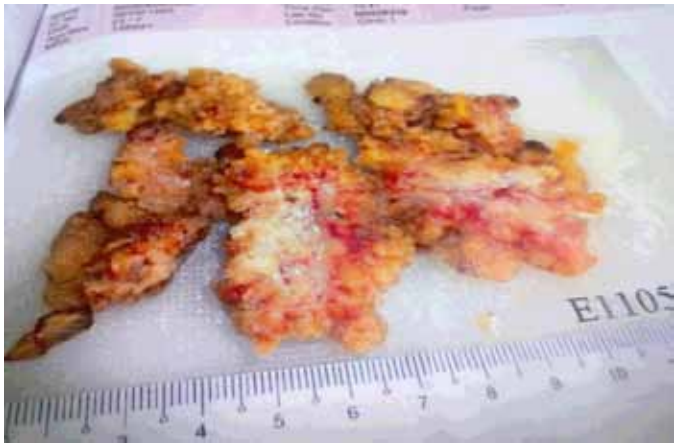

Figure 1: Tissue after polypectomy. It was removed in pieces.

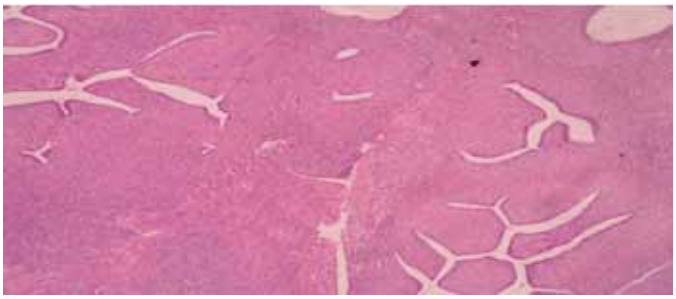

Figure 2: Microscopic picture of the polyp with scattered glands within a hypercellular stroma with the cells displaying oval to spindle-shaped nuclei with mild pleomorphic and distinct nucleoli (H&E stain, x400).

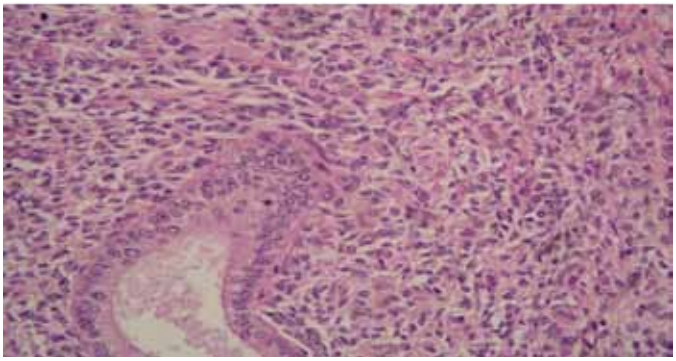

Figure 3: Higher magnification of the tumor showing mild nuclear atypia (H&E stain, x400).

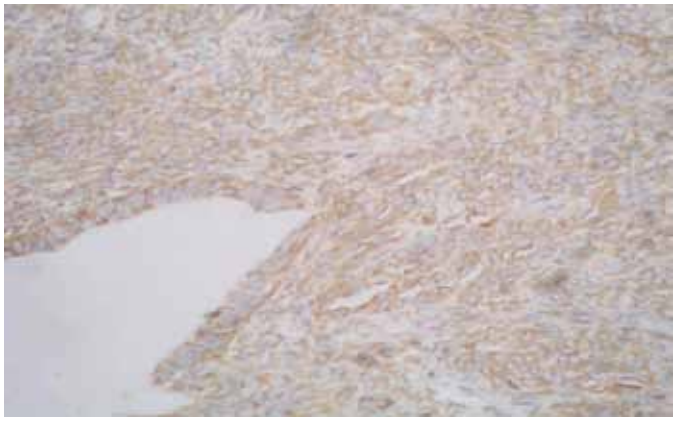

Figure 4: Immunohistochemical stains showing the mesenchymal cells are diffusely positive for CD10 which highlights the epithelial component (CD10, x400).

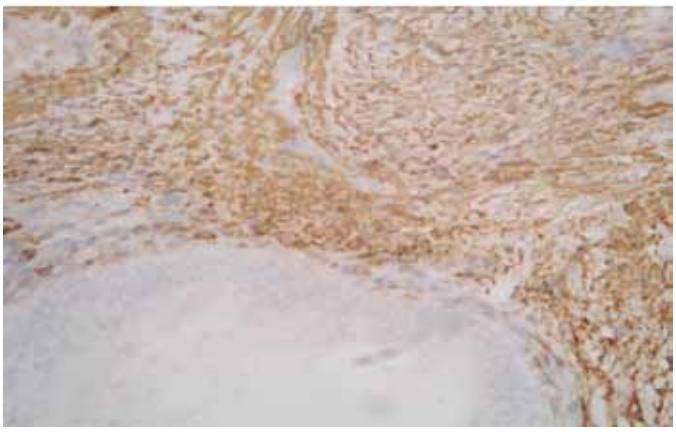

Figure 5: Immunohistochemical stains showing the mesenchymal cells are diffusely positive for Actin which highlights the epithelial component (Actin, x400).

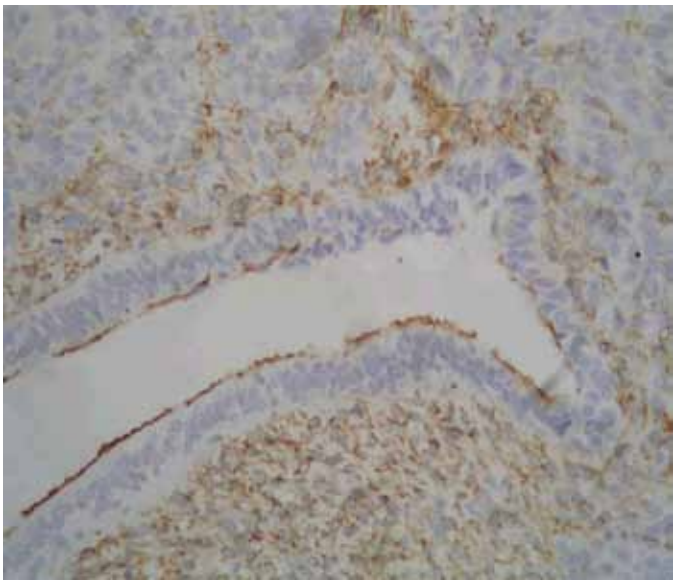

Figure 6: Immunohistochemical stains showing the mesenchymal cells are diffusely positive for Desmin, panCK highlights the epithelial component (panCK, x400).

surgery of the patient. After consented for operation, exploratory laparotomy was carried out. Intraoperatively, a bulky uterus corresponding to 8–10 weeks size of gravid uterus, left endometriotic cyst measuring about 2.6x3.0 cm with normal looking right ovary and a small tumor mass measuring about 3 cm on the surface of small bowel, which is 4 cm from the ileocecal junction were found. Intraabdominal findings were suggestive of an extensive disease/involvement, which warranted our team to proceed with radical surgery in this patient. A total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and paraaortic lymphadenectomy, appendicectomy, a 5-cm resection of small bowel and end-to-end anastomosis were done and peritoneal fluid was sent for cytological study. Her postoperative period was uneventful. She had been clinically free of the disease for 18 months since surgery. Currently, she is on Premarin 0.625 mg once daily and other supplements. Although there are complaints of vague body ache, fatigue and mood-swings, patient is coping well with the treatment given.

Patient is being regularly followed-up once in every 4 months with pelvic examination and ultrasound of abdomen. Histopathological report showed the following findings:

Gross: A brownish mass of 15 mm size at the right endometrial wall with no obvious myometrial invasion, gross involvement of lower uterus, endocervix or parametrium. There are two polyps at the uterine fundus measuring 15x5x5 mm and 10x5x5 mm. Left ovary is slightly enlarged measuring 30x30x25 mm. Other structures were normal.

Microscopy: No residual tumor mass in uterus and cervix. Two benign endometrial polyps, left follicular ovarian cyst, right benign inclusion cyst of ovary and heterotopias of pancreatic tissue over the surface of small bowel. No evidence of malignancy. Peritoneal fluid study is negative for malignancy.

DISCUSSION

A malignant mixed Müllerian tumor is a malignant neoplasm found in the uterus, ovaries, fallopian tubes and other parts of the body that contains both carcinomatous (epithelial tissue) and sarcomatous (connective tissue) components. It is divided into homologous and a heterologous type. A malignant mixed Müllerian tumor (MMMT) account for 2–5% of all tumors derived from the uterus. It is found predominantly in postmenopausal women. Risk factors are similar to those of adenocarcinomas and include obesity, exogenous estrogen therapies and nulliparity. Uterine adenosarcoma occurs in all age groups, but is most common in women after the menopause [8]. Adenosarcoma most commonly arises from the endometrium, but some cases are situated in the endocervix [9]. In postmenarchal adolescents, these polypoid tumors usually arise from the cervix,

in contrast to postmenopausal women in whom they usually arise from the endometrium. Tumors that arise in the cervix account for 2% of all adenosarcoma of the female genital tract (71% endometrium, 15% ovary and 12% the pelvis) [10]. The common presenting symptom is abnormal vaginal bleeding followed by pelvic pain, an abdominal mass or vaginal discharge. Its association is also described in patients taking tamoxifen and occasional cases have arisen in association with hyperestrogenism or with prior pelvic irradiation [11, 12]. Patient may present with intermenstrual and postcoital bleeding [13]. Clinical diagnosis may be challenging due to the benign gross appearance of the polyps [14]. Pathological diagnosis of low-grade adenosarcoma is often difficult in a tiny histological specimen. Most of the time, the sarcomatous stroma may be of very low grade and may be misdiagnosed as a benign disease such as adenofibroma [15, 16].

The differential diagnosis of adenosarcoma includes benign polyp, adenofibroma, embryonal rhabdomyosarcoma and endometrial stromal sarcoma. Cervical Müllerian adenosarcoma is usually treated by radical hysterectomy even in young age group as there is high recurrence rate following conservative surgical management [7]. Recurrences may occur late. The role of chemotherapy and radiation is limited in the absence of extensive pelvic or residual disease [17]. Since the CT scan finding and intra-abdominal findings during surgery were suggestive of an extensive disease/involvement, which warranted radical surgery in our patient. However, histopathology report revealed no residual tumor mass in the cervix and uterus. So she has not received any adjuvant chemotherapy or radiotherapy. The patient and her family members were informed about the histopathological findings. Poor prognostic factors include sarcomatous overgrowth and high-grade malignant features in the stromal component. The presence of malignancy should be kept in mind always while resecting a cervical polyp.

CONCLUSION

Müllerian adenosarcoma of the endocervix is rare in young women. The appropriate treatment of this tumor is uncertain because of its rarity in this age group, the malignant potential is not clearly defined and the evidence about the management is limited. On literature review, conservative surgical management such as polypectomy, cone biopsy or trachelectomy had more chance of recurrence. Hence, most of the experts recommend definite or radical surgical management irrespective of the age and parity. Chemotherapy and radiation is not recommended in the absence of extensive pelvic and/or residual disease. From literature review, recurrences have been reported 11 years after conservative treatment. Recurrences may occur late and thus long-term follow-up of these patients is recommended.

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Author Contributions

Sameera Begum – 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
Prasanta kumar Deka – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Tham Seng Woh – Substantial contributions to conception and design, Acquisition of data, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Squamous odontogenic tumor: A case report

Mervet Moussa, Marwa Mokbel ElShafei

ABSTRACT

Introduction: Squamous odontogenic tumor is a rare benign locally infiltrative odontogenic tumor of epithelial origin and only a few cases, less than 50, have been reported in literature. The challenge is in diagnosing the tumor is due to its close resemblance to acanthomatous ameloblastoma and well differentiated squamous cell carcinoma. Case report: Herein we report, not previously reported, characteristic case of squamous odontogenic tumor in the anterior maxilla in a teenager female. Conclusion: The diagnosis is predicated on recognition of the histopathologic features of squamous odontogenic tumor to obviate possible misdiagnosis of malignancy or ameloblastoma. The symptoms are modest. Squamous odontogenic tumor has a characteristic pathologic picture, which differs from ameloblastoma.

Keywords: Squamous odontogenic tumor, Odontogenic neoplasms, Anterior maxilla tumor

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INTRODUCTION

Squamous odontogenic tumor (SOT) is a rare benign true odontogenic neoplasm, with less than 50 cases reported in literature [1]. Three main types are identified: intraosseous, mural (mural SOT like-proliferation in a cyst) and extraosseous form, aggressive and multifocal variant have also been reported [2, 3]. Recently, it has been reported to be associated with calcifying epithelial odontogenic tumor [4].

According to Manjunath et al., collection of previous cases in literature, SOT affects a wide age range. The minimum age is eight-year-old and the oldest one is seventy-four [1]. A male predilection is noted and the most frequent sites were the bicuspid-molar region of the mandible and the incisor-cuspid area of the maxilla, with almost same distribution between maxilla and mandible [1, 3, 5]. Radiographically, SOT often exhibits a characteristic unilocular and triangular-shaped radiolucency of the alveolar bone, with the wide base of the radiolucency localized between the diverging apices of the adjacent roots [1].

Histopathologically, SOT consists of islands of well differentiated non-keratinized squamous epithelium in a mature fibrous stroma. The epithelial islands, occasionally, show foci of central cystic degeneration or calcification [1]. Squamous odontogenic tumor is a rare tumor that supposedly develops from the remnants of dental lamina, or of the cell rests of Malassez (ROM), or gingival epithelium [1]. Adebiyi et al. concluded that despite the infiltrative power of SOT, it should be treated conservatively, and this agreed with previous and later

works which stated that the usual treatment has been simple enucleation and recurrence has been rare [1, 4–6]. On the contrary, Ruhin et al. described a case of an aggressive SOT [3]. They recommended a management as a Pseudo-malignant tumor, especially, if located in the maxilla or is invading adjacent cortical bones. However, this kind of treatment depends on its local aggressive recurrence.

A few reports about the malignant transformation of SOT have also been reported by Ide et al. who conveyed its transformation into intraosseous squamous cell carcinoma [7]. The purpose of this article is to report this rare neoplasm as the first case identified as SOT in our dental school in Cairo Egypt and to review literature.

CASE REPORT

A 15-year-old female patient reported to our dental school complaining of mobility of teeth in the right anterior region of the maxilla for last three months. Clinical examination revealed a mild degree of expansion of the facial surface, the overlying mucosa appeared normal, and on palpation the area was slightly compressible but painless. The maxillary right central incisor and canine were mobile.

Based on the clinical and radiographic findings, the initial diagnosis of periodontosis was made and subjected for histologic examination. Histopathologic examination revealed a solid locally infiltrative benign odontogenic epithelial neoplasm composed of variably shaped (ovoid, round, cord-like structure) and sized of cytologically bland-looking mature stratified squamous epithelial islands randomly scattered throughout a dense mature fibrous connective stroma (Figures 1 and 2). These tumor islands did not show any evidence of peripheral columnar cells, reverse nuclear polarity or central stellate reticulum-like cells. The squamous cells are very uniform and exhibit no nuclear hyperchromatism, pleomorphism or mitotic activity, foci of microcystic degeneration was occasionally seen in some islands (Figure 3). Scattered chronic inflammatory cells were present in the stroma; these histopathological findings led us to the diagnosis of SOT.

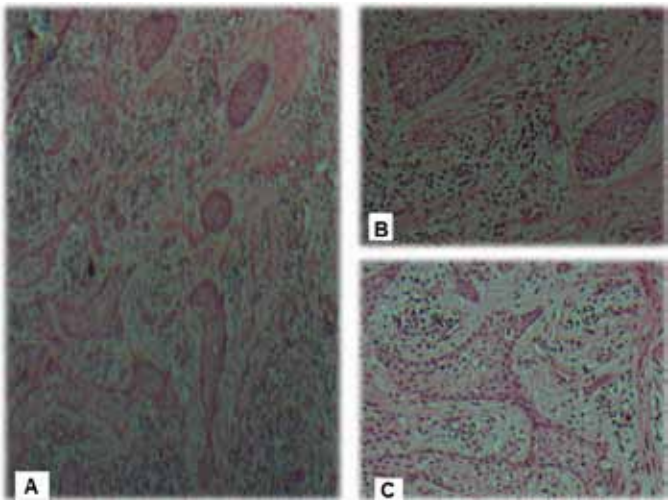

Figure 1: (A) The tumor consists of multiple scattered islands of squamous epithelium set in fibrocollagenous stroma heavily infiltrated with chronic inflammatory cells (H&E x100), (B) The rounded nests of squamous cells (H&E x400) and, (C) The thin epithelial cords anastomosing between the irregularly shaped tumor islands (H&E stain, x400).

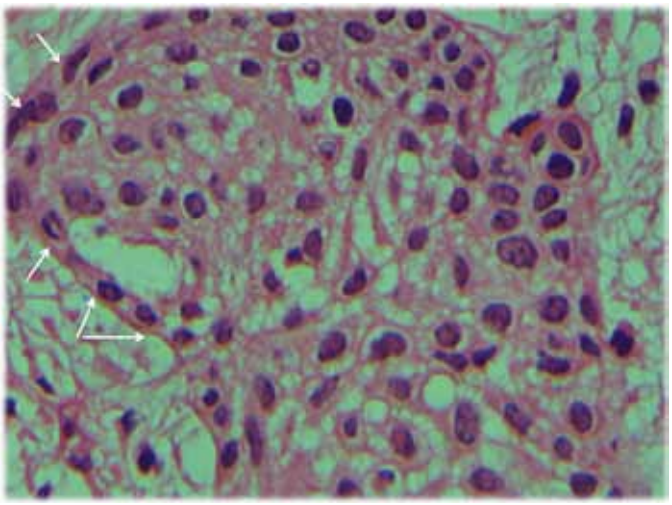

Figure 2: A higher magnification, of an irregular shaped island showing uniform squamous cells exhibiting no pleomorphism nuclear hyperchromatism or mitotic activity. Note the flattened peripheral layer of cells limiting the squamous island (arrows) (H&E stain, x100).

DISCUSSION

To date, about 44 cases of squamous odontogenic tumor have been reported [1]. Due to the paucity of documented cases of SOT; the relative frequency among odontogenic tumors could not be stated.

While our reported case occurred in a 15-year-old female when compared to the age range stated by Manjunath et al. [1]. The SOT occurs in a broad age range, up to the eighth decade, with a mean age range about 38 years. A slight male predominance is also stated in their work. Therefore, the gender is not of value in considering the diagnosis.

Considering the reported cases, the most common location for development of an SOT in the maxilla is anterior region and posterior in case of mandible with almost equal propensity to occur in both jaws. SOTs occurring in maxilla were found to be more aggressive than in mandible. This was mainly due to the anatomy, porous and medullary nature of bone. Our case was in the anterior region of the maxilla, extending posteriorly,

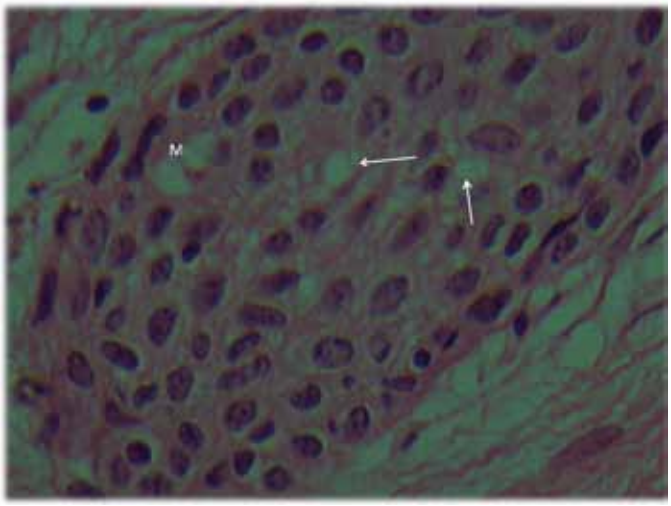

Figure 3: A high magnification photomicrograph of an island of benign squamous epithelium revealing the absence of peripheral columnar cells, palisading nuclei or central stellate reticulum. Note the evidence of microcysts (M) and focal vacuolization (arrows) (H&E stain, x1000).

which matches with the common location of SOT. It also accords with most of the lesions, being in the periodontium of permanent teeth [1, 3, 5].

A typical clinical image of SOT is a slowly growing central lesion that is nearly asymptomatic. Mild pain may coexist with slight swelling of the alveolar process and mobility of the neighboring teeth, an accidental discovery in a routine radiograph is not uncommon [8]. In our case, the patient only complained of mobility of teeth in the right anterior region of the maxilla. Upon examination a mild degree of expansion of the facial surface was revealed.

A well-defined unilocular radiolucency or multilocular pattern may be seen in the radiographic picture, relevant to the size of the lesion [6]. While, when a lesion is peripheral only pressure occurs on the alveolar bone leading to saucerization of the underlying bone [9]. As our case is an intrabony lesion, radiolucency is seen as an ill-defined area, proposing aggressiveness of the lesion, displacement of the canine and lateral incisors medially. The radiographic appearance of the lesion, occurring between the adjacent teeth in the tooth bearing area from the periodontal ligament space and the pattern of bone destruction seeming to emanate relatively uniformly from the areas of tooth roots lend credence to the theory of origin from the periodontal ligament area, i.e., the rests of Malassez [5].

Upon describing the histopathology of a typical SOT, benign squamous epithelial islands are seen, the squamous cells occupy the entire island with no columnar cells at the periphery neither palisading of the nuclei. These features enable the observers to eliminate the diagnosis of an ameloblastoma with squamous metaplasia as a differential one [1]. The epithelial islands might be surrounded with fibrous tissue condensation together with fibroblasts, which are considered a mesenchymal reaction to epithelial proliferation. The presence of keratin pearl, microcysts and calcification is not rare [9]. The precise histological picture, seen in our pathology lab, conforms to the classic picture mentioned above and this led us to the definite diagnosis.

Despite the ambiguous pathogenesis of SOT, still the origin from remnants of odontogenic epithelium is the most unswerving, including epithelial rests of Serres and Malassez. This theory is supported by the fact that most of the lesions are present in the periodontium of permanent teeth [1].

Kim et al. implied the mutation of the ameloblastin gene, which is found to be present in SOT, adenomatoid odontogenic tumor and ameloblastoma, to contribute in non-periodontal lesions [9]. It can also be a factor in the multicentric lesions. While Siar et al. suggested that ameloblastoma differs than SOT in the degree of differentiation of the lesions, depending on the results of their immunohistochemical stain of the two lesions with Notch 1,3 and 4 Jagged 1 and Delta 1 [10].

CONCLUSION

Squamous odontogenic tumor is a benign odontogenic true neoplasm. It is considered a distinct entity, its behavior differs from being well circumscribed and locally invasive, especially in maxilla. It should be treated first conservatively, but upon recurrence a more aggressive line of treatment is to be ensured. Very rare malignant transformation is reported. A proper diagnosis, depending on the histologic features should be ensured.

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Author Contributions

Mervet Moussa – 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

Marwa Mokbel ElShafei – 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 OPEN ACCESS

Idiopathic ovarian vein thrombosis in the postmenopausal age

Ralphe Bou Chebl, Seth Krupp, Kassem Bourgi, Gilbert Abou Dagher

ABSTRACT

Introduction: Ovarian vein thrombosis (OVT) is a rare condition, most often seen in the postpartum period. It has also been associated with pelvic inflammatory disease, hysterectomies and acquired thrombophilia states. Undiagnosed and unrecognized, it can lead to serious complications such as pulmonary embolism, sepsis and death. Case Report: We report a case of idiopathic unprovoked ovarian vein thrombosis in a previously healthy postmenopausal woman presenting with a one week history of right sided back pain. Conclusion: Ovarian vein thrombosis is a very rare condition that carries very serious complications. Clinicians should have a high level of suspicion and should initiate therapy as soon as the diagnosis is confirmed to avoid any catastrophic sequelae.

Keywords: Ovarian vein thrombosis (OVT), Gonadal vein, Postmenopausal

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Bou Chebl R, Krupp S, Bourgi K, Dagher GA. Idiopathic ovarian vein thrombosis in the postmenopausal age. International Journal of Case Reports and Images 2013;4(11):611–614.

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INTRODUCTION

Ovarian vein thrombosis (OVT) is an uncommon but potentially serious condition. It is most commonly seen in the puerperal period with an incidence of 1 in 3000 deliveries. The incidence increases to 1 in 800 with cesarean sections [1]. Ovarian vein thrombosis has also been associated with other hypercoagulable states such as factor V Leiden [2], gynecological malignancies, and it has been reported to occur following pelvic inflammatory disease (PID) and pelvic surgery [3, 4]. The patients usually presented with fever, abdominal pain and a pelvic mass, with a preponderance to involve the right ovary. Though it is a rare disease, it can lead to serious complications such as pulmonary embolism and sepsis, therefore, a higher level of suspicion is required for a prompt diagnosis and rapid initiation of anticoagulation. We describe an unusual case of ovarian vein thrombosis presenting as back pain in a postmenopausal woman.

CASE REPORT

A 79-year-old G5P5 woman with a past medical and surgical history of diabetes, hypertension, appendectomy at the age of five and a hysterectomy at age 30, was presented to the emergency department with a one-week history of right sided lower back pain. She described her pain as sharp, and non-radiating. She denied any fever, vomiting or urinary symptoms. She had no change in her bowel movements but did report some occasional right sided abdominal pain along with decreased appetite.

Vital signs of the patient showed blood pressure 148/68 mmHg, pulse 78 beats/minute, respiratory rate 17 breaths/minute, temperature 38.3°C. Physical examination of the back showed some mild tenderness to palpation in the right lower flank region. There were no focal neurological deficits. She had moderate right lower quadrant tenderness and guarding, without any rebound, and a positive psoas sign.

Laboratory tests including a complete blood count, a metabolic profile, liver function tests, urinalysis and an erythrocyte sedimentation rate (ESR) were within normal limits except for an ESR of 89 mm/hr. Given her presentation and physical examination findings, a computed tomography (CT) scan of her abdomen and pelvis with intravenous (IV) contrast were done and it showed a right ovarian vein thrombosis without any evidence of malignancy (Figures 1 and 2).

The patient was started on heparin and was admitted to the medicine floor for work up of occult malignancy. She underwent an ultrasound of the pelvis and a CT scan of chest which were negative for any malignancies and was discharged on warfarin for 3–6 months. On follow-up with the gynecology service, the patient denied any history of spontaneous abortions, blood clots, or PID. A mammogram was done which showed a suspicious mass in the left breast which was biopsied and was found to be benign.

DISCUSSION

Ovarian vein thrombosis is a rare condition. It is usually seen within the first four weeks postpartum most commonly occurring within the first four days [4]. The right ovarian vein is affected in 80-90% of cases. This has been attributed to the dextrorotation of the uterus during pregnancy compressing that vein, but also to the increased length of the right ovarian vein and the presence of incompetent valves [1]. The etiology is thought to be due to Virchow's triad of stasis, hypercoagulability and endothelial injury. It is associated with pelvic inflammatory disease, malignancy and pelvic surgery [3]. Recognition and treatment of ovarian vein thrombosis is needed to avoid the morbidity and mortality associated with this condition. The complications of OVT include thromboembolic events such as extension of the thrombus into the inferior vena cava or renal vein and lead to ovarian infarction, sepsis and ultimately death [5].

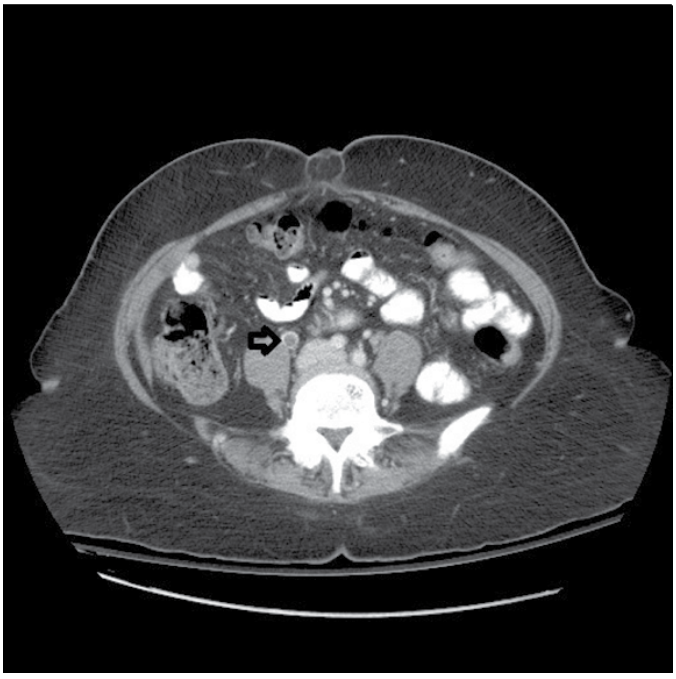

Figure 1: Computed tomography scan of the abdomen showing evidence of a right sided ovarian vein thrombosis (arrow) with enhancement of the ovarian vein wall.

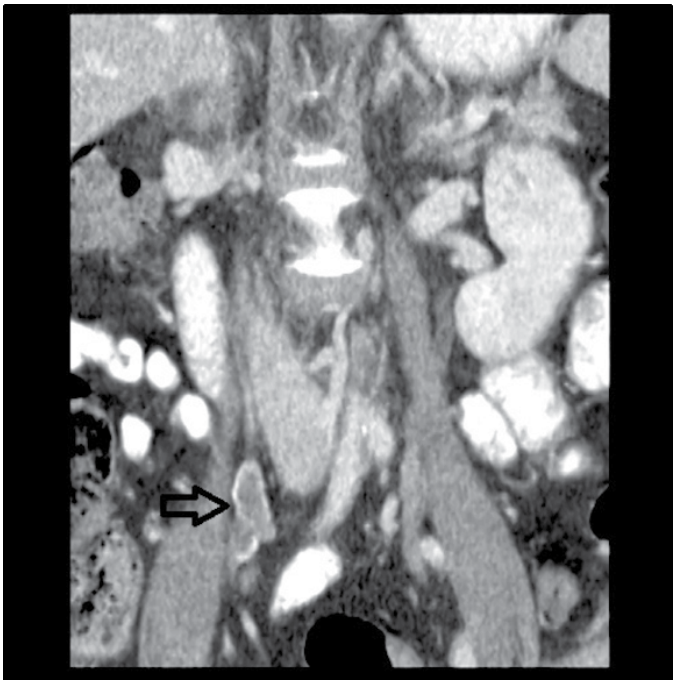

Figure 2: Coronal abdominal computed tomography scan showing evidence of thrombosis and enlargement of the distal end of the right ovarian vein (arrow).

Ultrasound, magnetic resonance imaging (MRI) scan, and CT scan with IV contrast are the best radiologic modalities for diagnosing ovarian vein thrombosis [1, 4]. Ultrasound provides a rapid non-invasive initial examination, free of any radiation to the patient. However, it only has a sensitivity of 56% in diagnosing OVT as it is operator-dependent, and the ovarian vein is often obscured by overlying bowel gas. The CT and MRI scans, with sensitivities greater than 95%, offer a definitive diagnosis of ovarian vein thrombosis and exclude other clinical diagnostic possibilities. The most common findings on CT scan is an enlarged ovarian vein with central hypodensity representing the thrombus, a sharply defined vessel wall, and perivascular inflammatory stranding [1, 4].

There are only two case reports in literature of idiopathic, unprovoked, ovarian vein thrombosis. The cases are from the United States in 2012 where a 67-year-old woman was presented with a one-month history of left sided abdominal pain and from Australia in 2010 where a 42-year-old woman was presented with acute abdominal pain and nausea [5, 6]. Both patients were admitted to the hospital and started on anticoagulation. They were not found to have any predisposing factors for venous

thrombosis. Two other cases were reported in literature as idiopathic but upon examination of the cases, it was found that both patients had risk factors as one of them presented late in the postpartum period and the other had stopped her oral contraceptives two months prior to the development of her OVT [7, 8].

Our case is unusual in that the patient initially presented with back pain. The differential diagnosis of such presentations in elderly women includes abdominal aortic aneurysm, discitis, pathological fractures, urinary tract infection, kidney stones and appendicitis. Usually, ovarian etiologies of abdominal pain are seen more in younger patients as the ovaries undergo a significant decrease in size after menopause [9]. It is unlikely that the hysterectomy caused her venous thrombosis as the surgery was done 40 years ago and there was no evidence of OVT on her previous abdominal CT scan done 4 years prior to this presentation.

The role of anticoagulation and antibiotics is not clear in the treatment of idiopathic ovarian vein thrombosis as the data is limited [1]. Patients are started on heparin and are discharged on warfarin for six months for a documented ovarian vein thrombosis. Antibiotics are usually given in postpartum patients presenting with fever and abdominal pain as an empirical treatment for endometritis [4]. In our review of literature, we found that all cases of idiopathic OVT had complete resolution of their symptoms without receiving any antibiotics [6–8]. Our patient was also not started on antibiotics and had complete resolution of her symptoms [10].

CONCLUSION

This is a rare presentation of unprovoked ovarian vein thrombosis that presented as back pain in a post-menopausal woman. Given the possible complications that could arise and the increased morbidity it carries on the elderly population, clinicians should have a high level of suspicion and should initiate therapy as soon as the diagnosis is confirmed to avoid any catastrophic sequelae.

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Author Contributions

Ralphe Bou Chebl – 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
Seth Krupp – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Kassem Bourgi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Gilbert Abou Dagher – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Uncommon metastasis to thyroid gland presenting as a thyroid nodule

Somnath Gooptu, Surendra Sharma, Gurjit Singh, Iqbal Ali

ABSTRACT

Introduction: Metastatic spread to the thyroid is not common in spite of the fact that it has a high vascularity. It is a rare situation especially in an individual without a prior history of malignancy. Thyroid gland is not the common site of metastasis in case of an esophageal malignancy. Case Report: A 60-year-old female was presented to our hospital with a solitary thyroid nodule involving the left lobe with cervical lymphadenopathy. There were no clinical features of hypothyroidism or hyperthyroidism. However, laboratory investigations revealed hypothyroid state. Patient was started on thyroxine 100 mg daily. Fine needle aspiration cytology (FNAC) revealed the presence of squamous cell deposits. On further investigating by upper gastrointestinal endoscopy, an ulcerative growth was detected 20 cm from the incisors. Biopsy from the growth confirmed the diagnosis of squamous cell carcinoma of the esophagus. Patient was treated with chemoradiation. Conclusion: All atypical neck masses have to be thoroughly investigated as all neck masses may not be related to primary thyroid conditions. This will result in avoiding unnecessary thyroidectomies instead of treating primary pathology.

Keywords: Thyroid metastasis, Esophagus, Fine needle aspiration cytology (FNAC), Squamous cell, Thyroid nodule, Hypothyroidism, Hyperthyroidism

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INTRODUCTION

The thyroid gland is not an usual site for metastasis despite, being second to the adrenal gland in terms of relative vascular perfusion. The most common cause of metastasis to the thyroid gland is the renal cell carcinoma [1]. The thyroid gland is not the most common site of metastasis in case of an esophageal malignancy.

CASE REPORT

A 60-year-old Indian female was presented with complaints of swelling in the neck for last one month which increased in size rapidly during this period. Two weeks later, she developed dysphagia for solids. Her voice was normal and she had no difficulty in breathing. There were no clinical features of hypothyroidism or hyperthyroidism. On examination, left lobe of the

thyroid gland was enlarged measuring 6x4 cm, hard in consistency and having restricted mobility. Carotid pulsations were felt on both the sides. Hard single, Level II and III, cervical lymph nodes were palpable on the left side (Figure 1). She was found to be in hypothyroid state on biochemical tests hence started on 100 mg of thyroxine daily.

Ultrasound scan of the thyroid gland revealed heterogeneous thyroid gland with enlarged isthmus and left lobe with a heterogeneous nodule in the left lobe of the gland. Enlarged lymph nodes were present at IB, II, III and IV levels. Fine needle aspiration cytology (FNAC) from the thyroid gland and cervical lymph nodes showed the presence of poorly differentiated squamous cell carcinoma with possibility of metastatic deposits in view of associated features of thyroiditis (Figures 2 and 3). Possible primary sites were considered and in view of history of dysphagia. Barium swallow and upper gastrointestinal endoscopy was done. Barium studies revealed asymmetric, concentric luminal narrowing in mid-esophagus level at the level of carina with mucosal irregularity and apple core appearance (Figure 4A–B). Upper gastrointestinal endoscopy revealed the presence of ulcerative growth involving the full circumference of the mucosa 20 cm from the incisors and biopsy from this site showed infiltrating squamous cell carcinoma (Figure 5). It was decided to treat the patient with chemoradiation.

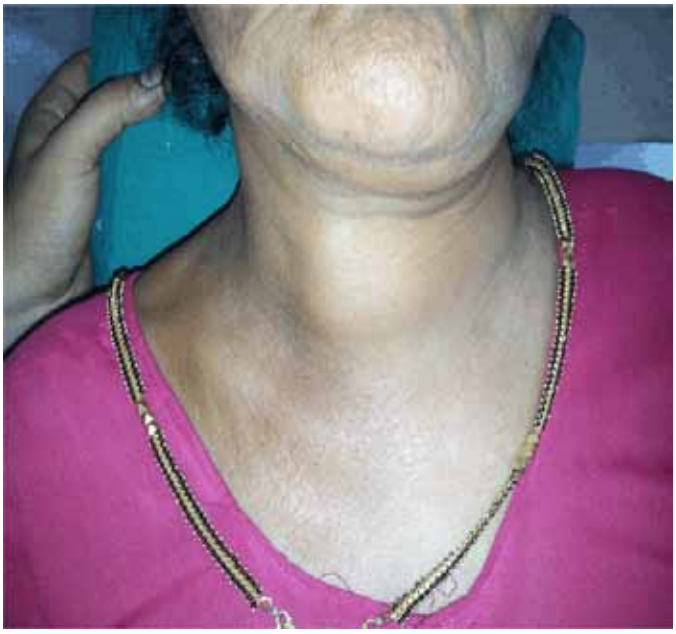

Figure 1: Enlarged left lobe of thyroid gland.

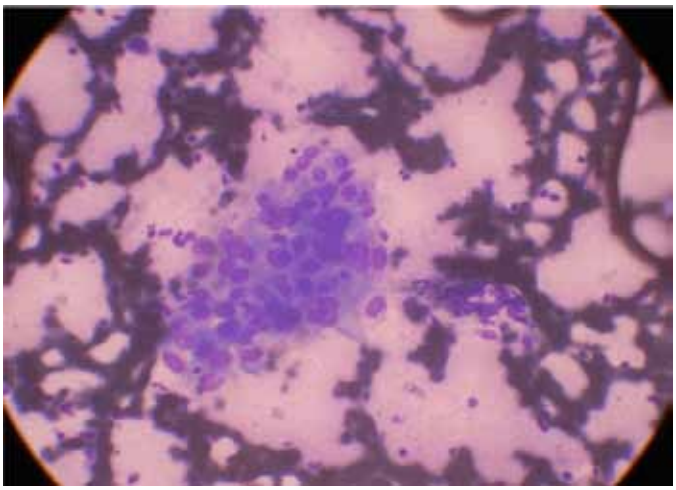

Figure 2: Fine needle aspiration cytology from thyroid gland showing squamous cell carcinoma (H&E stain, x400).

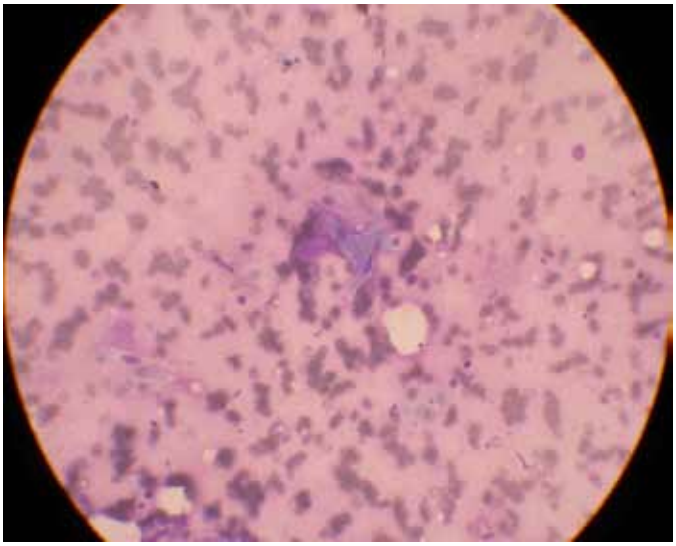

Figure 3: Fine needle aspiration cytology from cervical lymph node depicting squamous cell deposits (H&E stain, x400).

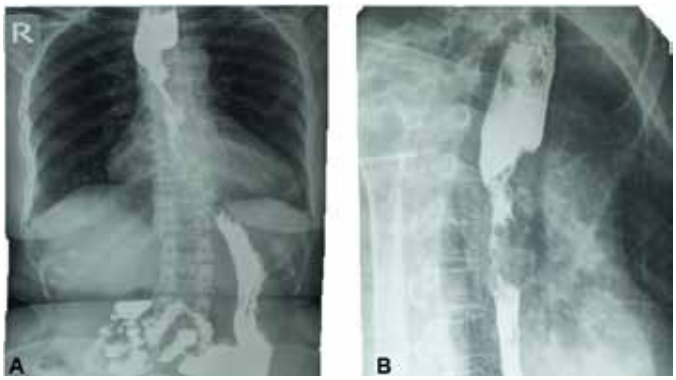

Figure 4: (A) Barium studies revealed asymmetric, concentric luminal narrowing in mid-esophagus level at the level of carina, (B) Mucosal irregularity and apple core appearance.

DISCUSSION

Squamous cell carcinoma of the thyroid is an extremely rare entity with an incidence of 1.4–2.5% of the thyroid malignancies [2]. Normally, in a thyroid gland squamous epithelium is not present but as a result of long standing inflammation intra thyroid squamous cells may be associated with embryonic remnants or result

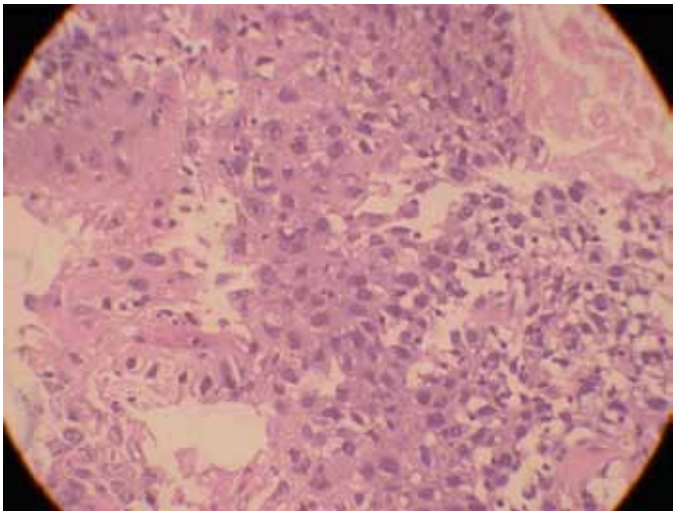

Figure 5: Biopsy from esophageal growth depicting infiltrating squamous cell carcinoma (H&E stain, x400).

from metaplastic transformation of native follicular epithelia [3]. The exact etiology for primary thyroid malignancy is not known. However, the proposed theory includes metaplasia theory (squamous metaplasia of the underlying thyroid disease) and embryonic rest theory (squamous cells originating from the remnant of the ultimobranchial duct or the thyroglossal duct).

Metastatic squamous cell carcinoma of the thyroid is more common. The most common sites of metastasis to the thyroid gland originate from the lungs, breast and kidneys [4]. Most of the series report carcinoma of kidney, colon and melanoma as the frequent primaries from which metastatize to thyroid [5]. Thyroid gland is an unusual site of metastasis from esophageal carcinoma. The etiopathogenesis of secondary metastatic squamous cell carcinoma (SCC) of the thyroid can be divided into three groups:

(i) Direct extension from adjacent primaries like esophageal or laryngeal carcinoma.

(ii) Hematogenous spread from the lungs or other primary sites.

(iii) Retrograde lymphatic spread is rare [6].

Secondary lesions that occur in the thyroid gland are usually solitary [7]. In certain cases, it can also present as a solitary thyroid nodule instead of a diffuse enlargement of the thyroid gland [6]. In our case too, it was solitary enlargement of the left lobe of thyroid. The usual presentation is the rapid enlargement of a neck mass (hard in consistency) with or without cervical lymphadenopathy. It may be associated with other symptoms such as dyspnea, dysphagia and hoarseness of voice. This usually occurs due to secondary infiltration to the adjacent structures. Our patient also presented with dysphagia and a rapidly developing midline neck mass with the presence of cervical lymphadenopathy. Metastatic carcinoma to the thyroid gland from the esophagus is a very rare occurrence [6]. Large autopsy series very rarely demonstrated esophageal metastasis to the thyroid (<1%) [8]. In these patients thyroid function tests are usually within normal limits and hypothyroidism or hyperthyroidism are infrequent findings [9]. Our patient was diagnosed with hypothyroidism and was started on thyroxine 100 mg once daily on empty stomach. Thyroid function tests returned to normal limits within one month of initiation of the therapy and the patient was advised to continue with the same treatment.

The FNAC is usually an important tool for the diagnosis as it helps to know the nature of neoplasm with accuracy in most of the cases [10]. In our case, FNAC from the thyroid gland and cervical lymph nodes showed the presence of poorly differentiated squamous cell carcinoma with possibility of metastatic deposits. However, the limitation of FNAC is that differentiation between anaplastic thyroid carcinoma and metastatic lesions are difficult [4]. SCC of the thyroid immunohistochemically stains for cytokeratin but not for Thyroid Transcription Factor I and thyroglobulin, which are considered to be markers for follicular and papillary carcinoma. The differential diagnosis may also include tumors showing thymic or branchial pouch differentiation as it occurs in the neck in close proximity to the thyroid gland and has a squamous or mucinous differentiation [10]. These have recently been named spindle epithelial tumor with thymus like differentiation (SETTLE) or carcinoma showing thymus like differentiation (CASTLE) [7]. The most unusual feature about the case was that it presented as a thyroid nodule with malignant cervical lymphadenopathy, but finally led to the diagnosis of secondary deposits from esophageal malignancy rather than a primary thyroid malignancy.

CONCLUSION

All uncommon thyroid malignancies and atypical neck masses must be thoroughly investigated as all neck masses are not related to primary thyroid conditions. This is important to avoid unnecessary thyroidectomies in patients having already widespread metastasis and poor prognosis.

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Author Contributions

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

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

Gurjit Singh – Acquisition of data, Analysis and interpretation of data, Drafting the article. Final approval of the version to be published

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

In cholestatic hepatic dysfunction, also consider the kidneys and the heart: A probable case of Stauffer syndrome in renal leiomyosarcoma with cava-atrial extension

Ruth Jones, Les Ala

ABSTRACT

Introduction: Abnormal liver function tests (LFTs) are a common reason for referral to hospital although in some cases, no obvious hepatic pathology is found. Yet, they can indicate other disease processes not immediately obvious. A rare cause of cholestatic hepatic dysfunction is the paraneoplastic manifestation of malignancy. Case Report: Herein, we report a case referred with anicteric cholestatic hepatic dysfunction that was found to have a renal leiomyosarcoma with inferior vena cava (IVC) and right atrium extension. Conclusion: The abnormal LFTs were thought to be a paraneoplastic manifestation of the tumor as these were improving following surgical treatment.

Keywords: Renal leiomyosarcoma, Stauffer syndrome, Inferior vena cava (IVC) right atrial tumor extension, IVC obstruction

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Jones R, Ala L. In cholestatic hepatic dysfunction, also consider the kidneys and the heart: A probable case of Stauffer syndrome in renal leiomyosarcoma with cava-atrial extension. International Journal of Case Reports and Images 2013;4(11):619–622.

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INTRODUCTION

This is a case of a primary renal leiomyosarcoma presenting with hepatic dysfunction and features of intraluminal tumor extension. Following surgical treatment, the patient started to experience improvement in her symptoms as well as her liver function. It highlights the need to consider extra-hepatic causes of hepatic dysfunction including possible paraneoplastic manifestations of malignancies.

CASE REPORT

Clinical features: A 61-year-old female was referred to our ambulatory care unit with a three-month history of worsening dyspnea on exertion. The referral was prompted by the discovery of abnormal liver function tests (LFTs) by her general practitioner. She had a dry cough and ankle swelling, but denied any other respiratory or cardiac symptoms. She did have ongoing right sided flank pain, and pain 'under her ribs' for last two months, described as constant without pleurisy or radiation. On systemic review, she denied weight loss, bowel or urinary symptoms, specifically frank hematuria, but complained of lethargy and intermittent pyrexia over the previous month.

Previously, she had had an oophorectomy for a benign teratoma, and a deep venous thrombosis 10 years before. She denied previous liver or renal problems. She was not on regular medication, did not smoke nor drunk alcohol. Physical examination revealed a slightly pale but comfortable lady with no icterus, clubbing, or obvious lymphadenopathy. Cardiovascular and respiratory examination were normal (blood pressure 135/85 mmHg) but there were prominent 'a' waves in her jugular veins. Her superficial abdominal veins were distended with flow

directed upwards towards the head on re-filling. As well as hepatomegaly (4 cm below the costal margin), she had an indistinct mass in the right flank. Her swollen ankles were mildly pitied.

Investigations: Blood tests showed a normocytic anemia, cholestatic hepatic dysfunction and raised inflammatory markers (Table 1). Thyroid function tests, electrolytes and hematinic profile were normal. Subsequent viral hepatitis and liver autoantibodies screen were negative. Urinalysis showed microscopic hematuria (++++) and proteinuria (++). Her electrocardiogram showed a sinus tachycardia (106 beats per minute).

A chest X-ray showed an elevated right hemi-diaphragm, a new finding as of 5 years previously, but clear lung fields (Figure 1). An ultrasound scan revealed a large right renal mass with a suspicion of IVC tumor extension; the liver was enlarged but there was no obvious metastatic liver disease or biliary tree dilatation.

Following contrast computed tomography (CT) scan of her thorax and abdomen, the large right renal tumor was confirmed, and was seen to extend into the renal vein, IVC and RA (Figure 2). There was no evidence of pulmonary embolism and no liver metastasis but there was a small amount of abdominal free fluid and small bilateral pleural effusion.

On transthoracic echo, a large tumor occupying the entire right atrium was noted and was seen to extend into the inferior vena cava (IVC). There was no tricuspid valve involvement and all other echo findings were within normal parameters (Figure 3).

Table 1: Blood test results at presentation and two months after surgery.

Parameter	At presentation	2 months after surgery	Normal Range and units
Alkaline phosphotase	211	182	30–130 U/L
Gama-glutamyl transferase	201	117	12–43 U/L
Total bilirubin	19	12	0–21 umol/L
Alanine transaminase	38	28	0–52 U/L
C-reactive protein	170	98	<5 mg/L
Ferritin	344	270	15–300 ug/L
Erythrocyte sedimentation rate	N/A	83	1–20 mm/hr
Hemoglobin	10.8	10.6	11.5–16.5 g/dL
Prothrombin time	16	On warfarin	11–14.5 s

Management: Subsequently, the patient was, transferred to a tertiary center under the joint care of the urological and cardiothoracic surgical team, where she had a radical right nephrectomy, atrial and caval tumor thrombectomy and IVC replacement with Dacron graft and femoral vein. After a prolonged post-surgery stay in the intensive care unit, she made a good recover and on

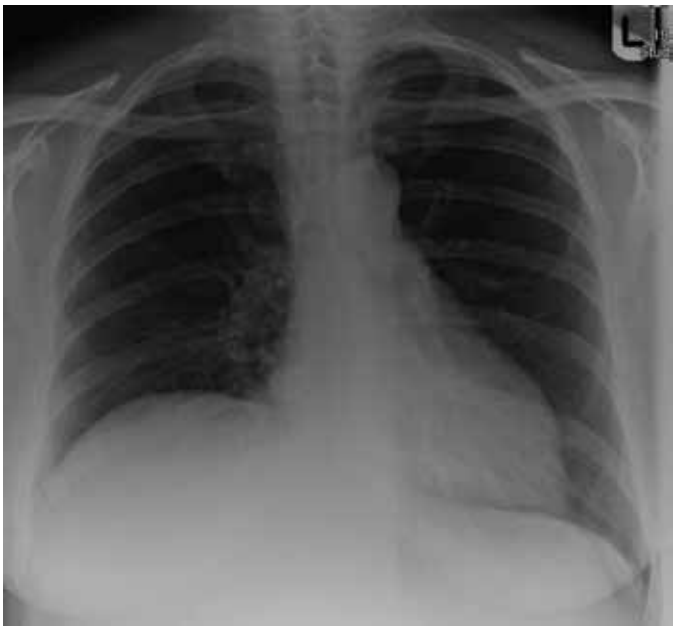

Figure 1: Chest X-ray showing elevated right hemi-diaphragm.

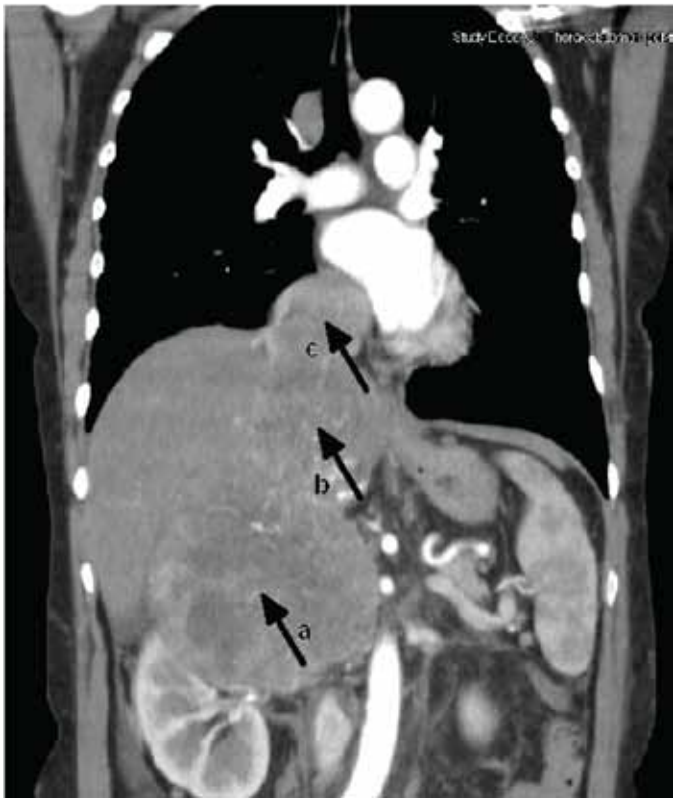

Figure 2: Contrast Computed tomography scan showing renal tumor (arrow a) with extension into inferior vena cava (arrow b) and into right atrium (arrow c).

follow-up two months after the surgery, she remained well with marked improvement in her symptoms.

The tumor was confirmed to be a high grade renal leiomyosarcoma. Further adjuvant treatment is currently being discussed with the oncologists. She remains on long-term warfarin for venous thrombo-embolic prophylaxis.

Following surgical treatment and two months after surgery, her LFTs appear to be improving (Table 1) although they have not as yet returned to normal. A post-operative prothrombin time was not measured as she had been put on warfarin during this period. A post-surgical echo showed a normal sized right atrium with no residual mass in the right atrium or the IVC (Figure 3).

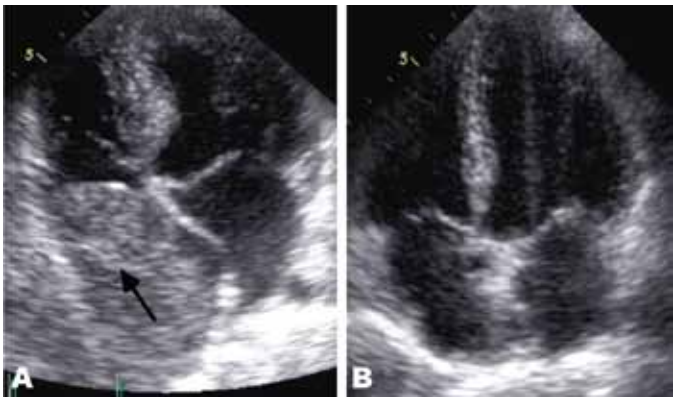

Figure 3: (A) Echo showing right atrium mass before surgery (arrow, left), (B) Echo showing normal right atrium after surgery (right).

DISCUSSION

Primary renal leiomyosarcoma is extremely rare, accounting for only 0.5–1% of all renal invasive tumors [1]. They may arise from the smooth muscle fibers of renal parenchyma, renal capsule, renal pelvis, or renal vessels [2]. These tumors often mimic renal cell carcinoma (RCC) in their presentation namely flank pain, palpable mass, and hematuria all of which are indicators of extensive local disease [1].

Intraluminal extension of the tumor from the kidney into the IVC and beyond is uncommon although some cases have been reported previously [3, 4]. Inferior vena cava involvement may produce lower extremity edema, ascites, hepatic dysfunction and pulmonary emboli.

Other features are more inconspicuous at presentation, particularly the paraneoplastic manifestations. This patient was referred with abnormal LFTs indicating a cholestatic hepatic dysfunction but in the absence of hepatic vein thrombosis, liver metastasis or biliary tree obstruction, such a hepatic profile might indicate Stauffer syndrome, a rare paraneoplastic manifestation of leiomyosarcoma [5].

Stauffer syndrome is well described in renal cell carcinoma but it has been reported in other malignancies such as bronchial carcinoma, prostate adenocarcinoma and there was one previously reported case in literature of Stauffer syndrome in leiomyosarcoma [5–8]. First described in 1961, Stauffer syndrome is characterized by an elevated alkaline phosphatase, gamma-glutamyltransferase (GGT), erythrocyte sedimentation rate (ESR), a prolongation of prothrombin time and hepatosplenomegaly. Histological examination of the liver shows non-specific inflammatory infiltrates [9]. The abnormal blood tests return to normal after surgical resection of the tumor but they reappear if the tumor recurs. The pathogenesis of this paraneoplastic syndrome is not fully understood but it is thought to be related to the release of pro-inflammatory cytokines such as interleukin-6 (IL-6) [10].

Although she had the classic triad of a renal tumor at presentation (flank pain, hematuria and abdominal mass), she also had features of IVC obstruction and possible cardiopulmonary complications. Tumor fragments could even embolize so given her dyspnea and features of possible IVC thrombotic obstruction, pulmonary embolism was in the differential diagnoses. However, with an elevated right hemi-diaphragm on CXR, a mass in the right flank and abnormal LFTs, we felt an ultrasound of abdomen was also warranted at that stage, and this identified a renal mass, confirmed IVC obstruction and excluded liver metastasis. The subsequent contrast CT thorax and abdomen showed the extent of the tumor and excluded pulmonary embolism.

The extension of the leiomyosarcoma into the right atrium may obstruct blood flow and cause signs and symptoms of right heart failure such as fatigue, peripheral edema, hepatomegaly and ascites. This patient did have some of these features but surprisingly, on echocardiography, her cardiac function was not compromised despite the tumor occupying the entire right atrium. The small amount of ascites and her lower limb swelling were therefore most likely due to IVC obstruction rather than right heart dysfunction.

The final diagnosis was only possible from histological examination as the tumor was indistinguishable from an RCC on the radiological images and at surgery.

CONCLUSION

Although renal leiomyosarcoma is extremely rare, it can mimic renal cell carcinoma at presentation. In this patient with a renal leiomyosarcoma, the improving trend of the cholestatic hepatic profile after surgery indicates that she probably had an associated Stauffer syndrome although she would require a longer follow-up period to ensure the liver function tests have returned to normal.

Nevertheless, her case illustrates the wide spectrum of manifestations of renal tumors and their complications, including inferior vena cava and right atrium extension and associated paraneoplastic syndromes.

In such patients, as well abdominal and thoracic radiological imaging, an echocardiogram is also required to exclude tumor embolus and RA mass, especially if they present with dyspnea, features of possible right heart dysfunction or of pulmonary embolism.

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Author Contributions

Ruth Jones – Acquisition of data, Analysis and interpretation of data, Performed echocardiography, Critical revision of the article and helped to write the first draft, Final approval of the version to be published
Les Ala – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Asymptomatic unilateral pulsatile eye: Clinical and therapeutic evaluation of sphenoid bone defect in neurofibromatosis type I

Ka Lung Chong, Syeb Shoeb Ahmad, Fatimah Hussin, Norlina Mohd Ramli, Shuaibah Abdul Ghani

ABSTRACT

Introduction: Neurofibromatosis type I (NF-1) is a common inherited, multisystem, neurocutaneous disorder that predisposes to the development of benign and malignant tumors. Orbital-temporal manifestations of neurofibromatosis are not common. Case Report: We report a rare case of atypical presentation of NF-1 which presented to us with incidental asymptomatic unilateral pulsatile eye. After investigation, the patient was found to have unilateral partial sphenoid bone defect. No other orbital-temporal manifestation of neurofibromatosis was noted. Conclusion: The pathogenesis of unilateral greater wing defect in NF-1 is still unclear. Some authors have reported that the sphenoid bone defect may progress. If the defect does progress, we need to follow-up to monitor sphenoid bone defect manifestation in every orbital-temporal NF-1 patient.

Keywords: Sphenoid bone defect, Orbital-temporal neurofibromatosis, Greater wing defect, Pulsatile eye, Neurofibromatosis type I (NF-1)

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INTRODUCTION

Asymptomatic unilateral pulsatile eye due to greater wing defect of sphenoid bone is rarely the first sign of neurofibromatosis type I (NF-1). We report a rare case of orbital-temporal NF-1 which was found to have only unilateral partial sphenoid bone defect.

CASE REPORT

A 26-year-old young male was admitted to intensive care unit (ICU) in Queen Elizabeth Hospital, Kota Kinabalu, Sabah, Malaysia for severe pneumonia. He was referred to our ophthalmology team after an incidental observation of a pulsatile movement of this left eye. The patient did not have ocular or visual complains and history of ocular or head trauma. Past medical or surgical histories were not significant. There was no similar condition in the family.

On ophthalmology examination his visual acuity was 6/6 in both eyes. Both eyes were symmetrical in position. There was neither proptosis nor enophthalmos (Hertel exophthalmometry measured 18 mm and 17 mm at 103 base reading). The left eye was pulsatile and the pulsatile were synchronous with the carotid pulse (Video 1). No thrill or bruit was detected over the eyeball, forehead or neck. Intraocular pressures (IOP) were 17 mmHg

and 18 mmHg in the right and left eyes, respectively. There were no Lisch nodules in either eye. Fundoscopy was normal in both eyes. Physical examination revealed thoracic scoliosis, axillaries freckling and multiple Café-au-lait spots on his trunk with majority measuring more than 3 cm in size. The rest of the physical examination was unremarkable.

Computed tomography (CT) scans of brain and orbits demonstrated partial defect of the left greater sphenoid wing (Figures 1 and 2). Three-dimensional images were constructed and showed details of a bony defect in the left orbit. A diagnosis of NF-1 was made. Magnetic resonance imaging (MRI) scan of brain was done later which showed diffuse intrinsic pontine glioma in the brainstem (Figure 3A–C). The patient passed away two months later due to complication of recurrent aspiration pneumonia.

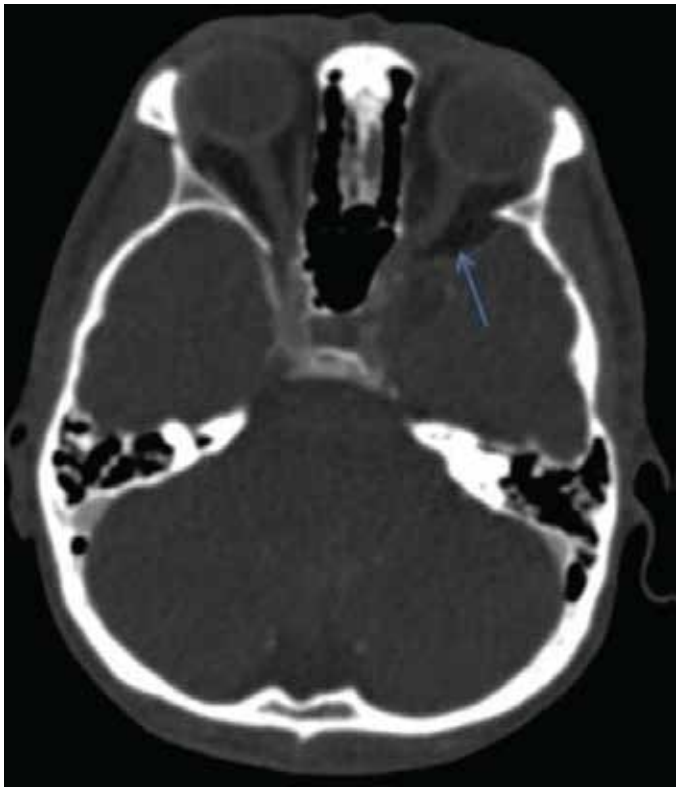

Figure 1: CT orbits in bone setting showing a large bony defect affecting the left sphenoid wing (←).

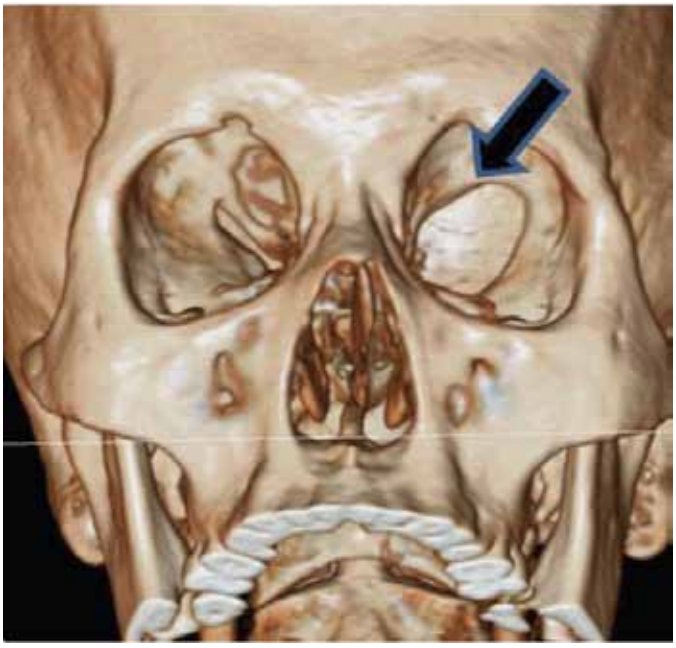

Figure 2: 3D reconstruction of the facial bones showing a large bony defect in the left posterior orbit.

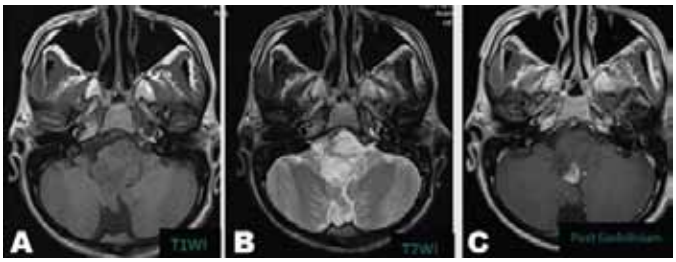

Figure 3: Magnetic resonance imaging (MRI) brain showing an ill defined potine mass which is predominantly isointense to grey matter on T1WI and hyperintense on T2WI with heretogeneous enhancement in post gadolinium with cystic component. This may represent intrinsic pontine glioma commonly found in neurofibromatosis type 1.

DISCUSSION

Neurofibromatosis type I, formerly known as von Recklinghausen disease, which was named after the researcher who first documented the disorder (Friedrich Daniel von Recklinghausen), is the most common inherited disorder caused by a mutation of a gene on the long arm of chromosome 17 [1] . Neurofibromatosis type I gene encodes a protein known as neurofibromin which acts as a tumor suppressor protein. It is an autosomal dominant genetic disorder but up to 50% of NF-1 cases arise due to spontaneous mutation. The incidence of NF-1 is about 1 in 3500 live births [2].

Orbital-temporal manifestations of NF-1 have been found to exist in 1–10% of patients and are characterized by pulsatile exophthalmos, pulsatile enophthalmos, orbital neurofibroma, sphenoid wing dysplasia, expansion of the temporal fossa and herniation of the temporal lobe into the orbit [3]. It can be classified into three groups (group 1: orbital soft–tissue involvement only with a seeing eye, group 2: orbital soft–tissue and significant bone involvement with a seeing eye, and group 3: orbital soft–tissue and significant bone involvement with a blind or absent eye) [4]. Our patient belonged to group 2 and only had a dysplasia of the greater wing of the sphenoid bone. Besides that, he did not have other orbital-temporal manifestations or ocular symptoms which were very different from other reported cases in literature [5].

Unilateral pulsatile eye can be caused by a vascular lesion in the orbit (e.g., orbital varices), carotid-cavernous fistula or defect of the sphenoid bone in the orbit. Greater wing defect of sphenoid bone can be present with clinical features such as pulsatile eye movement, headache, proptosis and diplopia with consecutive herniation of the temporal lobe. Our patient did not have any ocular symptoms except the incidental finding of a pulsatile eye.

Sphenoid dysplasia occurs in less than 1% of NF-1 patients. Further, abnormalities of the sphenoid wings are often considered pathognomonic [6]. According to National Institute of Health (NIH), abnormalities of the sphenoid wings are one of the diagnostic criteria for NF-1 (Table 1). Usually, this manifestation occurs unilaterally. There are still many controversies regarding the origin of this sphenoid bone dysplasia in neurofibromatosis. Although it has been regarded as a developmental anomaly of mesodermal origin, the exact mechanism for the sphenoid bone changes seen in neurofibromatosis is uncertain. An abnormality in the skull and orbital development, altered transmission of cerebrospinal fluid pulsations and interaction between plexiform neurofibroma and sphenoid bone are postulated as possible mechanisms [7]. In our case, there was no plexiform neurofibroma in the brain but the sphenoid bone defect was present. Review of literature has revealed this greater wing defect is not congenital. It may progress and eventually become typical sphenoid bone dysplasia [6].

When the greater wing defect is sufficient to allow the temporal lobe to herniate into the orbital cavity it may appear as an exophthalmos and gross facial deformity. Conversely, the orbital fat may prolapse into the middle cranial fossa and cause enophthalmos.

The greater wing defect is corrected for both cosmetic and functional reasons. Surgical correction can be performed through an intracranial approach or lateral orbital approach but the former method is associated with a high complication rate. Traditional surgical treatment of sphenoid dysplasia involves split bone grafting and repair of the anterior skull base defect. However, the results of this procedure may not be entirely sustainable owing to bone graft resorption and recurrence of proptosis and pulsating exophthalmos. A newer method utilities– a titanium mesh in conjunction with bone graft to act as a barrier between the orbit and the middle cranial fossa [8].

Table 1: Diagnostic criteria for neurofibromatosis type 1. At least two major criteria are required.

- A distinctive osseous lesion such as sphenoid dysplasia or thinning of long bone cortex, with or without pseudoarthrosis
- Two or more Lisch nodules (iris hamartomas)
- Optic glioma
- Axillary or inguinal freckling
- Six or more café-au-lait spots, >0.5 cm in prepubertal children; >1.5 cm in postpubertal individuals
- Two or more cutaneous neurofibromas
- One plexiform neurofibroma
- A first degree relative (parent, sibling, or offspring) with NF-1 by the above criteria

Abbreviation: Neurofibromatosis type I (NF-1)

CONCLUSION

Sphenoid bone dysplasia is one of the diagnostic criteria for neurofibromatosis type I. However, it is very rare for neurofibromatosis type I patient, which first presented with pulsatile eye movement. In our patient, his pulsatile eye was noted incidentally and did not have any ocular or visual symptom despite he had greater wing defect of sphenoid bone. The pathogenesis of unilateral greater wing defect in neurofibromatosis type I was not clear. Some literatures reported that the sphenoid bone defect may progress. We need to follow-up to monitor the defect manifestation in every orbital-temporal neurofibromatosis type I patient.

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Author Contributions

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

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

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

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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Incidentally discovered intrathoracic extra-adrenal pheochromocytoma during preoperative screening

Merces Assumpcao-Morales, Vinuta Mohan, Tasneem Zahra

ABSTRACT

Introduction: We are reporting a case of an incidental mass later diagnosed as an extradural pheochromocytoma at T2-T4 level during a presurgical work up for a gynecological procedure. Only a few cases of extradural paravertebral tumors are described in literature. Case Report: This is a clinical case of an asymptomatic 26-year-old female with an incidental finding of thoracic mass in chest X-ray during preoperative screening for myomectomy. Work up for this mass showed elevated plasma and urinary metanephrine levels which was consistent to pheochromocytoma. Computed tomography, magnetic resonance imaging and metaiodobenzylguanidine scans demonstrate a mass originating from thoracic spine with cord compression, no adrenal mass, multifocal disease or metastasis. Prior to surgery, the patient was treated with phenoxybenzamine and metoprolol to decrease hemodynamic instability during surgical procedure and also embolization to the arteries supplying the mass to decrease intraoperative bleeding. Patient remained stable during and after the removal of the mass. Pathology report confirmed presence of a 6-cm extradural pheochromocytoma at the T2-T4 location. Conclusion: A high index of suspicion for pheochromocytoma was crucial for the diagnosis in this patient. It is well known that the induction of anesthesia, use of certain drugs and manipulation of the tumor can lead to increase of catecholamine release with serious hemodynamics abnormalities and increase rate of mortality, complications that can be decreased with proper medical management. We report a very rare location of a pheochromocytoma. A high index of suspicion was crucial for the diagnosis and proper treatment in this asymptomatic patient.

Keywords: Pheochromocytoma, Extra-adrenal, Paravertebral tumor

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INTRODUCTION

Herein, we report a rare case of an incidental mass later diagnosed as an extradural pheochromocytoma at T2-T4 level during a presurgical work up for a gynecological procedure. A high index of suspicion for pheochromocytoma prompted an evaluation to establish the diagnosis in this asymptomatic patient. It is well known that the induction of anesthesia, use of certain drugs and indirect or direct manipulation of the tumor

can lead to increase of catecholamine release with serious hemodynamics consequences leading to significant complications and increased risk of mortality, all of which can be avoided with appropriate diagnosis and medical management. The supra diaphragmatic location of an extra-adrenal pheochromocytoma is already known to be very rare, and only a few cases of extradural paravertebral tumors are described in literature.

CASE REPORT

An otherwise healthy 26-year-old female with a past medical history of uterine fibroids and cholelithiasis underwent presurgical clearance for resection of uterine fibroids and was found incidentally to have a 5.8-cm left lung apical mass on chest X-ray. Computed tomography (CT) scan of chest revealed an expanding tumor that originates at T2 level with partial destruction of the left pedicle. The patient was admitted for further work up and resection of the mass when she was noticed to be hypertensive along with intermittent tachycardia. Patient did not have any symptoms such as headache, palpitations, diaphoresis or neurologic abnormalities. Clinical examination was remarkable for a systolic murmur in aortic area, lower left and right sternal border grade 1/6. Laboratory evaluation was significant for elevated urine and plasma metanephrine levels. An magnetic resonance imaging (MRI) scan of the thoracic spine demonstrated T2-T3 dumbbell shaped mass suggesting cord compression with flattening (Figure 1). Images studies failed to demonstrate adrenal tumor or metastasis. Metaiodobenzylguanidine scan was requested and patient discharged with follow-up in endocrinology and neurosurgery clinic. Metaiodobenzylguanidine scan demonstrated intense radiotracer accumulation in left upper chest, with no other activity elsewhere in the body cavity which reassured the diagnosis of isolated extra-adrenal pheochromocytoma. Surgical intervention was indicated. Preceding surgery, she was treated with phenoxybenzamine and metoprolol to decrease hemodynamic instability one would expect intraoperatively. An angiogram with embolization to the arteries supplying this chromaffin tissue to prevent excessive intraoperative bleeding was performed two days prior to surgery. T2-T4 laminectomy and decompression of the tumor was performed with no intraoperative complications. Postoperative course was complicated with pneumonia, which responded well with antibiotics. Pathology report confirmed presence of a 6-cm extradural pheochromocytoma at the T2-T4 location.

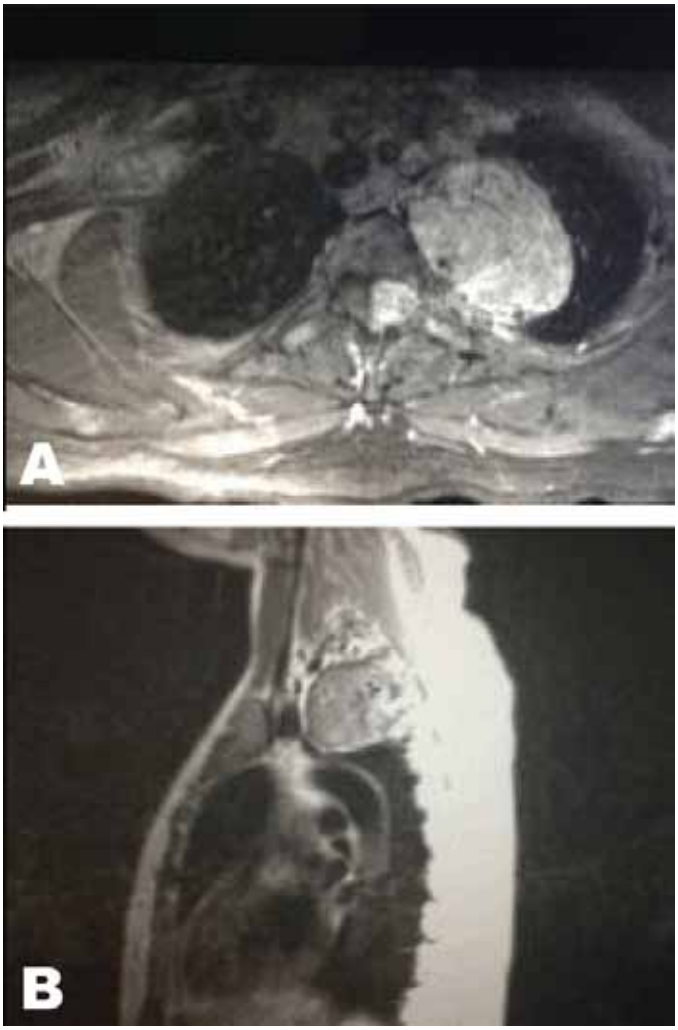

Figure 1: Thoracic spine magnetic resonance imaging scan showing a large dumbbell shaped tumor extending through the left T2-T3 neuroforamina with cord compression (A) Axial view, (B) Sagittal view.

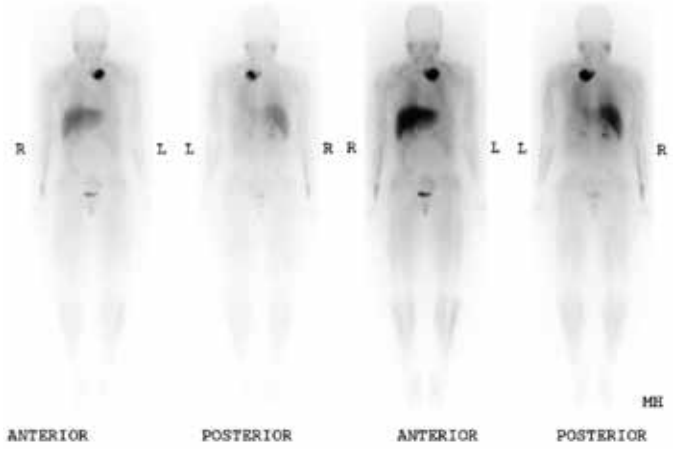

Figure 2: Iodine-123 metaiodobenzylguanidine scan showing large focal region of intense radiotracer accumulation in the left upper chest.

DISCUSSION

Pheochromocytoma is a rare catecholamine-secreting tumor derived from chromaffin cells. They are known to follow the 10% rule: 10% bilateral, 10% malignant,

10% in children, as well as an association that has been described with familial syndromes, known as multiple endocrine neoplasia type II. New studies suggest a higher incidence of extra-adrenal location, 15–20% extra-adrenal location in the adults and even 30% in children [1, 2]. The diagnosis of adrenal pheochromocytoma are most common in the 4–5th decade of life while extra-adrenal sites occur earlier, in the 2nd or 3rd decade [1, 2]. The extra-adrenal locations have been associated with a higher incidence of metastasis and multicentricity. Multicentricity pattern in 15–24% of extra-adrenal tumors was noted in a study conducted by Whalen et al. [2]. Eighty-five percent of extra-adrenal tumors are located in the retroperitoneum, usually arising from the organ of Zuckerkandl–extra-adrenal sympathetic paraganglionic organ located at the origin of the inferior mesenteric artery. Only 1% is located above the diaphragm. The spinal canal can be affected throughout its course, although very uncommon, with the intradural lumbosacral region being the most described location. In a review of literature only nine cases of thoracic extradural pheochromocytoma were reported [3]. Patient symptoms include paroxysmal hypertension, orthostatic hypotension, anxiety, tremors, sweating and palpitations due to increase levels of norepinephrine. Other symptoms are related to compression of adjacent structures by the tumor. The measurements of plasma and/or urinary fractioned metanephrines are the recommended first line tests for the diagnosis of pheochromocytoma. Studies have shown that they have a higher diagnostic sensitivity when compared with urinary and plasma catecholamines as well as urinary vanillylmandelic acid [4]. Imaging is necessary in order to define location, as well as to exclude multicentricity and metastasis. In a prospective study that analyzed 104 patients over 48 years with pheochromocytoma, imaging modalities, such as computed tomography and magnetic resonance imaging scans demonstrated specificity above 97% [5]. Computed tomography scan demonstrated a higher sensitivity, but was not statistically significant. Functional imaging is always required to confirm the diagnosis and rule out metastasis, the most common being scintigraphy with iodine-131 metaiodobenzylguanidine. Positron emission tomography (PET) scan is also is showing promise as a diagnostic modality. Complete surgical resection is the treatment of choice for extra-adrenal as well as adrenal pheochromocytoma. It is important to be aware of risk of hemodynamic instability during surgery that can lead to multiorgan failure as a main complication. Preparation should start two weeks prior to surgery and include use of alpha blockers, such as prazosin or phenoxybenzamine in order to provide full alpha adrenergic blockage and restoration of blood volume [6, 7]. Later consideration in starting beta blockers is also recommended in order to avoid arrhythmia. Following surgery, lifelong follow-up will be required aimed at detecting recurrence of the tumor or metastasis since it can happen anytime after surgery.

CONCLUSION

A high index of suspicion for pheochromocytoma was crucial for the diagnosis in this asymptomatic patient. Since direct or indirect manipulation of the tumor can lead to hemodynamic instability and increase in mortality patients should receive proper medical treatment prior to surgery in order to avoid and reduce those complications. This patient was proper diagnosed and treated prior to surgery remaining stable during per and postoperative course.

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Author Contributions

Merces Assumpcao-Morales – Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Vinuta Mohan – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Tasneem Zahra – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor

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Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Subtle angioedema presented systemic lupus erythematosus: A case report

Shera Irfan Ali, Yousuf Qayser, Rasool Roohi

ABSTRACT

Introduction: Acquired allergic angioneurotic edema secondary to systemic lupus erythematosus (SLE) is rare so herein we present angioedema as the initial presentation that led to the diagnosis of SLE in a young adult female. Case Report: A diagnosis of angioneurotic edema with underlying allergic disorder was made in a young adult female patient presenting with the swelling of dorsum of hands, lips and right periorbital region of the recent origin. The associated clinical and laboratory features finally led to a diagnosis of SLE. Conclusion: The uniqueness of this case suggests that the subtle presentation of angioedema as a secondary cause to various life-threatening medical disorders should not be underestimated.

Keywords: Angioedema, Acquired angioedema (AAE), Systemic lupus erythematosus (SLE), C1-esterase inhibitor protein (C1-INH).

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INTRODUCTION

Angioedema/angioneurotic edema is a well-demarcated localized edema involving deeper layers of the skin, including the subcutaneous tissue [1]. Unlike urticaria, where skin eruptions are distinctly pruritic and can involve any area of body from scalp to the soles of feet, in angioedema the most common sites of involvement are the periorbital tissue and lips [2, 3]. Angioedema may either be acquired due to variety of causes or may be hereditary in nature [4]. Acquired angioedema (AAE) is characterized by acquired deficiency of C1 inhibitor (C1-INH), hyper activation of the classical pathway of human complement and angioedema symptoms mediated by bradykinin released by inappropriate activation of the contact-kinin system [5]. Reports on acquired allergic angioneurotic edema secondary to systemic lupus erythematosus (SLE) have been rare so herein we present angioedema as the initial presentation that led to the diagnosis of SLE in a young adult female.

CASE REPORT

A 32-year-old Kashmiri female was presented with swelling of dorsum of hands, lips and right periorbital region of recent origin. She had a previous history of primary hypothyroidism for last six months. She was taking 75 µg of levothyroxine per day. She denied any recent exposures to drugs, food, insect venom stings and physical factors prior to the onset of facial and hands swelling. She had no previous history of similar attacks,

and no family history of angioedema. Her past medical history was otherwise unremarkable.

On general physical examination, she was tall, thin built and afebrile. She had erythematous swelling of dorsum of hands involving all fingers without tenderness and induration. This was associated with unilateral periorbital swelling and erythema (Figure 1), for which provisional diagnosis of angioneurotic edema with underlying allergic disorder was entertained. There was no pedal or sacral edema, no lymphadenopathy or hepatosplenomegaly on clinical examination. She had no features of vasculitis on physical examination. Laboratory investigation showed bicytopenia with hemoglobin of 11.4 g/dL (normal range 10.7–14.9), a white blood cell count of $2.88x10^3/\mu L$ (normal range $3.30x10^3$–$9.66x10^3$), an absolute lymphocyte count of $0.9x10^3/\mu L$ (normal range $1.04x10^3$–$2.86x10^3/\mu L$) and a platelet count of $46x10^3/\mu L$ (normal range $150x10^3$–$450x10^3/\mu L$). Her ESR measured 30 mm/h (normal range 0-20 mm/h). She was further investigated by estimating levels of C1-esterase inhibitor protein (C1-INH), found low 180 mg/L (normal range 210–390 mg/L). Bone marrow examination and plasma electrophoresis was normal. Imaging techniques namely, ultrasonography, magnetic resonance imaging (MRI) and computed tomography (CT) scans of various parts of body were normal.

The possibility of acquired angioedema with underlying immune-complex disease was made by demonstrating a positive antinuclear antibody and hypocomplementemia. Her C2 complement was 1.3 mg/dL (normal range 1.6–3.5 mg/dL), C4 complement 8.2 mg/dL (normal range 10–50 mg/dL) and C3 complement level 82 mg/dL (normal range 90–180 mg/dL).

In view of bicytopenia with lymphopenia, ANA positive and hypocomplementemia, diagnosis of systemic lupus erythematosus was suspected. This was confirmed when further investigations revealed antibodies to double stranded DNA (dsDNA) 189 IU/L (normal <25 IU/L) and asymptomatic lupus nephritis (24-hour urinary protein of 0.54 g/day). She was asked to undergo kidney biopsy but denied against it. She had raised anti-thyroid peroxidase (TPO) antibodies measuring 71.07 IU/mL (normal <35 IU/mL), further depicted association with autoimmune disorder.

In the outpatient setting, she had received moderate-dose of 0.5 mg/kg oral corticosteroids for six weeks followed by maintenance dose of 0.07 mg/kg per day. She is currently well; with no proteinuria, normal full blood counts and improved outlook. Angioneurotic edema subsided at sixth day of her treatment. Her C1-INH levels and C4 complement had normalized on the first follow-up visit to 224 mg/L and 13 mg/dL, respectively. These further increased subsequently to 310 mg/L and 48 mg/dL at last follow-up. Her latest anti-TPO antibody levels have considerably decreased to the normal limits. She was regularly following our allergy/immunology clinics till date.

Figure 1: Allergic angioneurotic edema in a young adult female.

DISCUSSION

Acquired angioedema is first suspected in patients aged 40 or above who presented with recurrent cutaneous and/or mucosal angioedema without urticaria, without an evident triggering factor, and without family history of angioedema. The measurement of C1-INH and C4 antigen in such patients is the first step [4]. In our case, the levels of C1-esterase inhibitor protein (C1-INH) was found low, 180 mg/L (normal range 210–390 mg/L), her C3 complement levels were 82 mg/dL (normal range 90–180 mg/dL) and her C4 complement was 8.2 mg/dL (normal range 10–50 mg/dL) indicating the provisional diagnosis of acquired angioneurotic edema with underlying allergic disorder. Acquired angioedema in lupus has been found associated with classical pathway mediated hypocomplementemia (low C3 and C4) and transient low C1-INH antigenic and functional levels. Further, AAE in lupus has also been found associated with absence of lymphoproliferative disease and absence of clinical SLE activity during acute angioedema [6]. As our patient presented with the acute angioedema, our focus was more on any underlying allergic disorder rather than on SLE. In order to establish the diagnosis, additional investigations for lymphoproliferative and autoimmune disease were carried out including complete

blood count (CBC), erythrocyte sedimentation rate (ESR), antinuclear antibodies, anti-thyroid peroxidase (TPO) antibodies, 24-hour urinary protein, bone marrow examination and whole body scan. There was least possibility of orbital cellulites and pseudotumor as patient was afebrile and had normal MRI of orbits. The patient did not show any lymphoproliferative diseases like non-Hodgkin lymphoma of orbit and hairy cell leukemia that was ruled out by the imaging techniques and bone marrow examination. Although lymphoproliferative diseases represent the main group encountered in AAE and a direct pathogenetic relationship between the two conditions cannot be questioned, SLE, different neoplasia and infections have also been described in association with AAE [7-17]. Benign forms of lymphoproliferation disorders like monoclonal gammopathy of uncertain significance (MUGS) have been reported with high frequency in association with AAE [5]. The patient's angioneurotic edema subsided at sixth day of oral corticosteroid treatment. Resolution of angioedema with immunosuppressive therapy is associated with normalization of C3, C4 and C1-INH levels [18].

CONCLUSION

Allergic angioneurotic edema secondary to systemic lupus erythematosus occurs due to the acquired deficiency of C1-esterase inhibitor protein (C1-INH), positive antinuclear antibody and hypocomplementemia. The uniqueness of this case suggests that one should not underestimate the subtle presentation of angioedema as a secondary cause to various life-threatening medical disorders.

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Author Contributions

Shera Irfan Ali – Conception and design, Acquisition of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Yousuf Qayser – Acquisition of data, Drafting the article, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Rasool Roohi – Acquisition of data, Drafting the article, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

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CASE REPORT OPEN ACCESS

Chronic urticaria and angioedema associated with Hashimoto's thyroiditis in a child: A case report

Ashutosh Kumar, Sandeepkumar Kuril, Sasikumar Kilaikode, Paul Saenger

ABSTRACT

Introduction: The association between chronic urticaria and Hashimoto's thyroiditis has been rarely reported in children. Case Report: We are reporting a case of an eight-year-old girl with chronic urticaria unresponsive to antihistaminic therapy, who was subsequently diagnosed to have Hashimoto's thyroiditis. Patient's urticarial lesion remitted after treatment with levothyroxine. Conclusion: We emphasize to consider testing for Hashimoto's thyroiditis by thyroid autoantibodies and thyroid profile in cases of chronic urticaria, and starting the patient on levothyroxine for the symptomatic improvement of both chronic urticaria and Hashimoto's thyroiditis.

Keywords: Angioedema, Chronic urticaria, Hashimoto's thyroiditis, Levothyroxine

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INTRODUCTION

Hashimoto's thyroiditis is an organ specific autoimmune disease characterized histologically by lymphocytic infiltration of the thyroid gland. This disorder is 2–4 times more frequent in girls than in boys. Most of the affected children are euthyroid and asymptomatic. The most common clinical manifestations are goiter and growth retardation secondary to hypothyroidism. Thyroid anti peroxidase antibody (TPO Abs) and antithyroglobulin antibodies are present in 90% of the affected children [1].

Childhood chronic urticaria is a common disorder characterized by the appearance of hives for more than six weeks [2, 3]. The pathogenesis of chronic urticaria is poorly understood and the cause is unknown in majority of cases [4]. A subset of patients with chronic urticaria has been classified as autoimmune on the basis of association with thyroid autoimmunity and anti-IgE and/or anti-IgE receptor antibodies [5].

As per literature review, an association has been found between Hashimoto's thyroiditis and chronic urticaria in adults [6], but there are only a few cases reported in children [7]. Hence, we are presenting a case of an eight-year-old girl with chronic urticaria and Hashimoto's thyroiditis who became asymptomatic after treatment with levothyroxine.

CASE REPORT

An eight-year-old Hispanic girl referred by immunologist to our pediatric endocrinology clinic in month of September 2010 with abnormal thyroid function test and chronic urticaria. On August 2010,

patient was presented to immunology clinic with a two-month history of recurrent urticarial lesions over face, trunk and extremities accompanied by swelling of lips and tongue which were non-responsive to antihistamine treatment. Patient also complained of weakness, fatigue and significant weight gain over the period of six months. In the immunology clinic routine laboratory analysis, allergy tests and thyroid function tests including thyroid autoantibodies were performed. Patient was found to have high TSH 9.8 mIU/mL (< 4.6 mIU/mL), free T4 0.5 ng/dL (normal limit 0.7–1.5 ng/dL), high anti-TPO antibodies 352 IU/mL and positive anti-Fc epsilon Receptor (anti-FCER) antibody. Routine laboratory analysis and total serum IgE was within normal limits. Allergic testing to common food allergens was negative.

On examination in pediatric endocrinology clinic, patient's vital signs were normal, weight 46.03 kg (>95 percentile), height 133.5 cm (75–90 percentile) and BMI- 25.83 (>97 percentile). Her physical examination showed multiple urticarial lesions over the face, trunk and extremities, thyroid gland was not palpable. Systemic examination was unremarkable. Patient was started on levothyroxine 100 μg/day and antihistamine as needed for urticaria and angioedema. Patient was followed-up in February 2011; urticarial lesions were completely remitted during the visit while on levothyroxine with normal thyroid function. Patient was continued to follow-up at regular interval in our endocrinology clinic and remained asymptomatic after one year on hormone replacement therapy with normal thyroid function test (Table 1).

Table 1: Changes in thyroid function test with treatment

Thyroid Function Tests	Prior to levothyroxine treatment	4 months after treatment	12 months after treatment
TSH (mIU/mL)	9.8	2.16	4.15
Free T4 (ng/dL)	0.5	1.7	1.6
Anti-TPO antibodies (IU/mL)	352	-	-

DISCUSSION

Chronic urticaria is a common clinical condition whose etiology in about 75% of cases is unknown. A link between chronic urticaria and autoimmune thyroid diseases such as Hashimoto's thyroiditis has been proposed and studied [8]. Approximately one-fourth of chronic urticaria patients have serological evidence of thyroid autoimmunity suggesting association between them. There are various hypotheses on how these two entities might be related:

(a) Immune complexes produced in the course of Hashimoto's thyroiditis are trapped in the skin and may cause urticaria.
(b) Inflammatory cells activated in the thyroid are directed toward a cross-reactive antigen existing in the skin.
(c) There is no direct relationship between the inflammation in the thyroid and the skin, but autoimmunity directed toward FcER coexists with autoimmunity directed toward the thyroid gland in susceptible patients.
(d) There is no relationship between chronic urticaria and Hashimoto's thyroiditis, but they are both common disorders coexisting in a small percentage of patients [9].

Hashimoto's thyroiditis with chronic urticaria in children is rare with only a few reported cases. A study by Levy et al. described eight female patients in the age group of 7–17 years with chronic urticaria and positive thyroid autoantibodies. Two of the eight patients were hypothyroid and were started on thyroxine without any improvement in urticaria [7]. In our patient, the common causes of chronic urticaria like allergy to external agents, hereditary angioedema and occult infections were excluded and the patient was diagnosed to have Hashimoto's thyroiditis based on high TPO antibodies titers and abnormal thyroid profile. The patient was started on levothyroxine therapy. After four months of treatment with levothyroxine, urticarial lesions disappeared without any recurrences noticed over the one year follow-up. This was in contrast with the two hypothyroid patients studied by Levy et al. whose urticaria did not respond to thyroxine treatment.

Therefore, we assume an association between chronic urticaria and Hashimoto's thyroiditis with clinical remission of resistant urticarial lesions after levothyroxine treatment.

CONCLUSION

We recommend considering testing for Hashimoto's thyroiditis by thyroid autoantibodies and thyroid profile in cases of chronic urticaria and starting levothyroxine in a hypothyroid patient with Hashimoto's thyroiditis for its symptomatic improvement as well as for ameliorating chronic urticaria.

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Author Contributions

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

Sandeepkumar Kuril – Conception and design, Acquisition of data, Analysis and interpretation of data,

Drafting the article, Critical revision of the article, Final approval of the version to be published
Sasikumar Kilaikode – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Paul Saenger – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

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CASE REPORT OPEN ACCESS

Giant cell arteritis presenting as tongue necrosis

Ritesh Kohli, Eleni Tiniakou, Joao M Nascimento, Gbonjubola Oyefeso

ABSTRACT

Introduction: Giant cell arteritis (GCA) is a large and medium-vessel systemic vasculitis affecting predominantly the internal and external carotid arteries with particular affinity for its extra-cranial branches (especially, the superficial temporal artery). The most frequent complication of this disorder is visual loss. We report the case of a patient who suffered tongue necrosis (a rare complication) secondary to giant cell arteritis. Case Report: A 71-year-old female who after having her dentures refitted developed severe pain, edema of the tongue, odynophagia, dysphagia and resultant anorexia. She was admitted with the diagnosis of glossitis and initially treated with antifungals and later antiviral medications without improvement. She subsequently developed tongue ulcerations, severe pain in temporomandibular joint (TMJ) with right sided headache. Temporal arteritis was suspected at this point, re-examination revealed palpable and tender temporal arteries (R>L). Tongue examination demonstrated dark greyish plaques in the dorsum with areas of ulceration without palatal involvement. An elevated ESR and the right sided temporal artery biopsy that followed demonstrating chronic inflammation with the presence of giant cells confirmed the diagnosis of GCA. Treatment with oral prednisone (1 mg/kg/day) produced a rapid improvement of her symptoms and ulcers with complete resolution at three months. Conclusion: The presence of glossitis in GCA represents a severity marker of this disease and is associated with a higher risk of vision loss and mortality. As such a high level of suspicion is necessary for the early recognition of this rare presentation and prompt institution of treatment to prevent the aforementioned complications.

Keywords: Giant cell arteritis (GCA), Scalp necrosis, Temporal artery biopsy

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INTRODUCTION

Giant cell arteritis (GCA) is a systemic vasculitis affecting large and medium sized arteries. It chiefly involves the extra cranial branches of carotid artery, particularly the posterior ciliary arteries that supply the optic nerve and the superficial temporal artery. The most frequent complication of this disorder is visual loss. We report a case of a patient who suffered rare complication, tongue necrosis, as a result of GCA. Early diagnosis and treatment of this can limit tissue injury and the progression of disease.

CASE REPORT

A 71-year-old female who after having her dentures refitted developed severe pain and edema of the tongue. She was admitted with the diagnosis of glossitis and initially started on antifungal therapy. She later developed tongue ulcerations and was started on antiviral medications with no significant improvement. Subsequently, she developed severe pain in temporomandibular joint (TMJ) and right sided headache to the extent that she had marked difficulty swallowing and decreased oral intake. Her past medical history was positive for well controlled hypertension and hyperlipidemia alone. Her social and occupational history was non-contributory as were the review of systems. Temporal arteritis was suspected at that point and re-examination revealed palpable and tender temporal arteries especially on the right side. The oropharyngeal examination revealed the presence of dark-greyish plaques and focal areas of ulceration on the dorsum of the tongue. Laboratory examination was remarkable for a grossly elevated sedimentation rate at 95. A right sided temporal artery biopsy was performed and demonstrated chronic inflammation and the presence of giant cells confirmatory for the diagnosis of GCA.

Treatment with oral prednisone (1 mg/kg/day) produced a rapid improvement of her symptoms and a gradual improvement in the appearance of the ulcerated areas. The lesions were completely resolved at a three-month follow-up.

DISCUSSION

Giant cell arteritis is a chronic granulomatous vasculitis, of unknown etiology, that affects individuals older than 50, especially females. Aging is the single greatest risk factor for the disease. The GCA never occurs before the age of 50 years. Although the cause of GCA is unknown, the disease appears to be T cell dependent and antigen driven [1].

The classic manifestations of GCA are headache, jaw claudication, polymyalgia rheumatica and visual symptoms including diplopia and loss of vision. Atypical manifestations include dry cough, pinna or parotid gland pain due to involvement of posterior auricular artery, fever of unknown origin and otolaryngeal manifestations including tongue pain, glossitis, dental pain, tongue ulceration and gangrene. Necrosis of the tongue occurs rarely owing to the extensive collateral blood supply. There is usually associated dysphagia, excessive salivation, tongue pain and masticatory claudication prior to a diagnosis of arteritis [2]. These symptoms are important warning signs of impending lingual infarction.

The diagnosis of GCA is mostly clinical. Recently, the American College of Rheumatology, established diagnostic criteria, which states that to be classified as having a GCA, a patient must meet three of the following five criteria.

(i) Aged 50 years or older.

(ii) The presence of new onset localized headache.

(iii) Temporal abnormalities like temporal tenderness or decreased temporal pulse.

(iv) ESR of 50 mm or higher.

(v) Abnormal temporal artery biopsy findings demonstrating mononuclear infiltration or granulomatous inflammation [3].

The presence of three or more of these five criteria is associated with a sensitivity of 93.55% and a specificity of 91.2%.

In clinical practice, establishing the diagnosis of GCA requires a biopsy of the temporal artery. The temporal artery biopsy must be carried out in all patients with a suspicion of GCA, even if the ESR has been normal. If the biopsy is negative, and the clinical suspicion is high, contralateral biopsy should be performed. Temporal artery biopsy is positive in only half of the patients who manifest the disease, as skip lesions occur commonly so a negative biopsy does not exclude GCA. The administration of corticosteroid given for less than two weeks does not reduce the yield of temporal artery biopsy [4]. In cases of lingual necrosis, tongue biopsy is usually nonspecific and is not indicated.

The most dreaded consequence of GCA is visual loss, which is usually irreversible. The early treatment of patients with GCA is crucial to avoid visual loss and the therapy should be started based on clinical suspicion, and not delayed for the biopsy results. Once the diagnosis of GCA is suspected, early treatment with steroids is recommended, to prevent complications and promote the complete resolution of this self-limiting condition. Typically, the treatment begins with prednisone 1 mg/kg body weight in adults [5]. It must be maintained until the symptoms have disappeared and the ESR is back to normal.

In the absence of characteristic signs and symptoms, diagnosing GCA can pose a challenge. This case reinforces the importance of suspecting GCA as cause of tongue necrosis especially in the absence of typical symptoms like headache and vision loss. Tongue necrosis is a rare occurrence in GCA and its presence represents a severity marker of the disease associated with an elevated risk of vision loss as well as overall higher mortality rates, in comparison to patients not presenting with this manifestation.

CONCLUSION

The aim of this article is to alert the physicians to the atypical manifestations seen in this disease like tongue claudication and tongue infarction which can all serve as a warning sign, as such a high level of suspicion must be held for these clinical findings, in order to initiate prompt and proper treatment and avoid blindness.

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Author Contributions

Ritesh Kohli – 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
Eleni Tiniakou – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Joao M Nascimento – 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 OPEN ACCESS

Upper limb deep venous thrombosis following a simple clavicle fracture

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ABSTRACT

Introduction: Fracture of the clavicle is a common injury, accounting for 5–12% of all fractures and up to 44% of injuries of the shoulder girdle. About 70–80% of these fractures are in the middle third of the clavicle [1]. Damage to neurovascular structures associated with closed fractures of the clavicle due to blunt trauma is rare and more frequently related to penetrating injuries. The usual mechanisms of injury include fall on an outstretched hand or on the point of the shoulder and direct or indirect trauma associated with contact sports [2]. Literature about the incidence of deep venous thrombosis in upper limbs in orthopedic practice is limited [3]. This case report presents a case of upper-extremity deep venous thrombosis following conservative treatment of an acute clavicular fracture. Case Report: A 50-year-old female was presented with a fractured left clavicle after a high-velocity motorcycle accident. At presentation, there were no associated injuries and she was intact neurovascularly. X-ray showed a middle third left clavicle fracture which was treated conservatively in a sling. After 17 days of injury, the patient was again presented with acute discomfort and swelling of the left upper extremity. The patient had no risk factors for venous thromboembolism. However, a deep venous thrombosis was clinically suspected. Doppler ultrasonography confirmed thrombotic occlusion of both axillary and basilic veins. Anticoagulation therapy was initiated and continued for a total of three months. At three months follow-up, the fracture healed successfully and the swelling completely subsided. Conclusion: We conclude that a high index of suspicion is necessary to rule out possible vascular lesions in cases of high-energy blunt trauma to the shoulder associated with clavicular fracture. Treating it would prevent a potentially fatal pulmonary embolism.

Keywords: Deep venous thrombosis (DVT), Clavicle, Axillary vein thrombosis

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INTRODUCTION

Fracture of the clavicle is a common injury, accounting for 5–12% of all fractures and up to 44% of injuries of the shoulder girdle. In sports related fractures, it is even the most frequent fracture. About 70–80% of these fractures are in the middle third of the clavicle [1]. Damage to neurovascular structures associated with closed fractures of the clavicle due to blunt trauma is rare and more

frequently related to penetrating injuries. The usual mechanisms of injury include fall on an outstretched hand or on the point of the shoulder and direct or indirect trauma associated with contact sports [2]. Little evidence exists on the incidence of axillary vein thrombosis in orthopedic practice. It is considered an uncommon clinical event with considerable potential for morbidity [3]. This article presents a case of upper-extremity deep venous thrombosis following conservative treatment of an acute clavicular fracture.

CASE REPORT

A 50-year-old fit and active female was presented to our emergency department with isolated left shoulder pain after falling from motorcycle at a speed of approximately 50 mph. There was tenderness over the left clavicle and she had difficulty in moving her left shoulder. There was no sensory or motor deficit in the upper limb. Ipsilateral peripheral pulses were palpable, with good capillary filling. X-ray of the left shoulder showed a simple middle third clavicle fracture (Figure 1). The arm was supported in a broad-arm sling for conservative treatment of the fracture.

After a week follow-up, the patient remained intact neurovascularly and a check X-ray was done which was satisfactory. As a routine practice with conservatively treated clavicular fractures, the patient was allowed to mobilize the elbow actively. After 10 days, she presented to accident and emergency department with acute discomfort and swelling of the left upper extremity.

According to the thromboprophylaxis assessment protocol done for all orthopaedic admissions in our hospital, the patient had no risk factors for venous thromboembolism (age <60, body mass index <30 kg/m^2, no medical co-morbidity, not on thrombogenic medication, no personal history of thromboembolism and no anticipated significantly reduced mobility). Deep venous thrombosis was suspected clinically. Doppler ultrasonography showed thrombotic occlusion of the axillary and basilic veins with no flow in that segment (Figure 2).

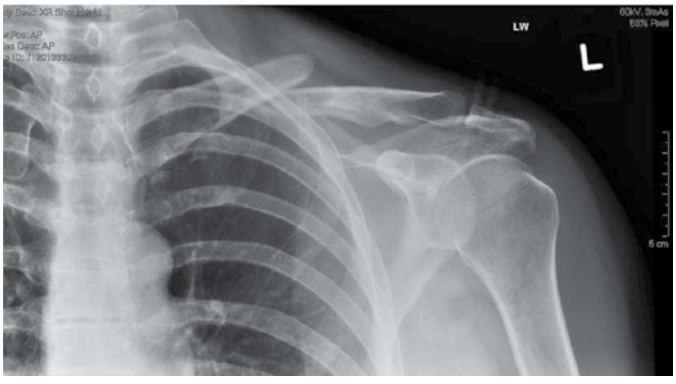

Figure 1: X-ray showing fracture of left clavicle.

Anticoagulation therapy was started with low molecular weight heparin then warfarin for a total period of three months. After six weeks follow-up the patient showed clinical union at the fracture site and regained range of motion of the left shoulder. Upper limb swelling has noticeably improved and radiography showed satisfactory union (Figure 3). Considering the uncommon presentation of upper limb deep venous thrombosis, the patient was finally reviewed and discharged after a period of three months.

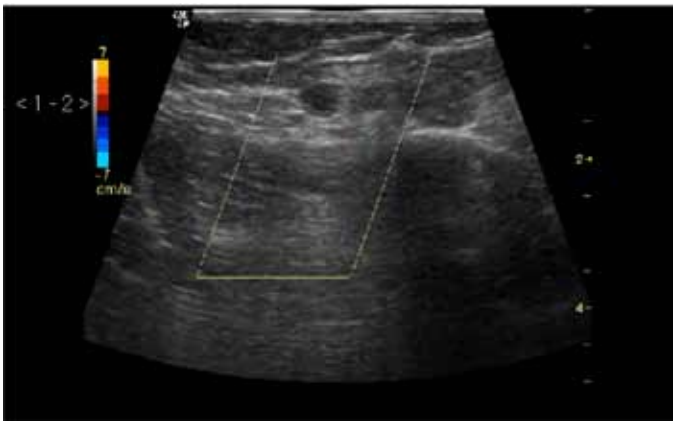

Figure 2: Doppler ultrasonography showing axillary vein thrombosis.

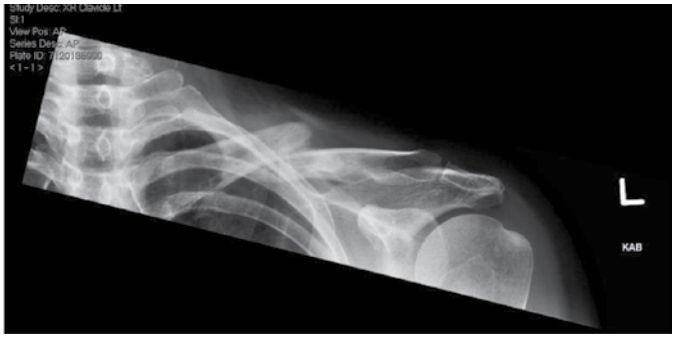

Figure 3: X-ray showing radiological union of the left clavicle.

DISCUSSION

Middle third fractures of the clavicle account for 70–80% of clavicular fractures [1]. The usual mechanisms of injury include fall on an outstretched hand or on the point of the shoulder and direct or indirect trauma associated with contact sports [2]. Approximately, 10% of all cases of deep venous thrombosis involve the upper extremities, resulting in an annual incidence of 0.4 to 1 case per 10,000 people.

The classic presentation of upper extremity deep venous thrombosis is acute discomfort, arm swelling, and risk factors such as vigorous arm exercise, an implanted

central venous catheter or pacemaker, or a history of deep venous thrombosis. Oedema, discoloration, and visible venous collaterals are also typical signs [4]. A high degree of clinical suspicion is necessary to detect deep venous thrombosis from blunt trauma. A Doppler ultrasound can throw light on the degree of vascular damage or occlusion, the level of injury, the involvement of the artery and vein and any external compression due to hematoma [5]. In this case, there were no risk factors for deep venous thrombosis. She sustained a direct trauma to the left clavicle. The fracture was treated in a broad-arm sling and antithrombotic medications were given for three months and regular follow-up appointments were arranged. We postulate that high-velocity trauma, which can occur in a road traffic accident, may include direct blunt injury to the clavicle resulting in fracture of the middle third. In these circumstances, displaced sharp bone fragments can cause intimal damage [5–7]. Early recognition of these rare lesions can guide the physician in planning the appropriate treatment that could prevent a fatal pulmonary embolism [8]. There is little evidence in literature about the incidence of deep venous thrombosis in upper limbs in orthopedic practice [8].

CONCLUSION

We conclude that a high index of suspicion is necessary to rule out possible vascular lesions in cases of high-energy blunt trauma to the shoulder associated with clavicular fracture. Treating it would prevent a potentially fatal complication.

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Author Contributions

Andy Tanagho – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Conflict of Interest

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CASE REPORT OPEN ACCESS

Solitary ileal bezoar: A rare cause of acute ileal obstruction in a teenager

Sefu Juma Uledi, Fauzia Ayubu Masumai

ABSTRACT

Introduction: Bezoars may be described as conglomerates of partially digested or indigestible organic substances in the gastrointestinal tract. Bezoars are classified based on their composition. There are four main categories—phytobezoars, trichobezoars, pharmacobezoars, and lactobezoars. Typically, bezoars are ingested and primarily conglomerate in the stomach over time. Occasionally, they may migrate distally and cause obstruction of small bowel. Bezoar formation mainly occurs in patients with predisposing factors like altered gastrointestinal anatomy due previous surgery or impaired gastric motility. The presence of isolated bezoar in the small bowel without synchronous existence of primary gastric bezoar or any apparent predisposing factors for bezoar formation remains an exceedingly a rare presentation. Case Report: We hereby, report a case of isolated ileal bezoar causing acute small bowel obstruction in rather a healthy 15-year-old girl. Conclusion: Solitary bezoar induced ileal obstruction in a healthy teenager is a rarity occurrence. This report is, therefore, aimed at highlighting this atypical cause of small bowel obstruction in this age group and cautions clinicians in our environment that bezoars form an essential part of the differential diagnosis when assessing patients with small bowel obstruction.

Keywords: Isolated bezoar, Small intestinal obstruction, Enterotomy, Laparotomy

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INTRODUCTION

Bezoar refers to impacted concretions of partially digested or indigestible organic substances in the gastrointestinal tract. Various types of bezoars have been described. However, bezoars are classified based on their composition. There are four main categories. These are phytobezoars, trichobezoars, pharmacobezoars, and lactobezoars [1].

Phytobezoars, being the most common, consist of non-digestible fibers of fruits and vegetables and mainly occurs in patients with predisposing factors like altered gastrointestinal anatomy due previous surgery or impaired gastric motility. Trichobezoars, these consist of hair fibers and seen mostly in patients with behavioral or psychiatric illness. Pharmacobezoars are composed of medicines such as antacids, ferrous sulfate and cholestyramine. Less common category is lactobezoars which are composed of milk, curd and occasionally encountered in premature infants on formula feeds [1–3].

Typically, bezoars are ingested and primarily conglomerate in the stomach over time. Occasionally,

they may migrate distally and cause obstruction of small bowel. Occurrence of isolated bezoar in the small bowel without synchronous existence of primary gastric bezoar or any apparent predisposing factors for bezoar formation has been barely reported and remains exceedingly a rare presentation [3, 4].

We hereby, report a case of isolated ileal bezoar causing acute small bowel obstruction in a healthy 15-year-old girl. This case report is, principally, aimed at documenting an extraordinary aetiology of small bowel obstruction in this age group and concurrently alerts clinicians that bezoars form an essential part of the differential diagnosis when evaluating patients with small bowel obstruction.

CASE REPORT

A 15-year-old girl presented to our hospital with a history of severe colicky abdominal pains, abdominal distension, and bilious vomiting for two days. Patient gave a history that she had been relatively well until about two days prior to admission when she started experiencing severe abdominal pains. Pains were, colicky in nature, non-radiating and more marked on the periumbilical region. Patient sited no aggravating or relieving factors for the pain.

Associated with abdominal pains patient gave history of nausea and non-projectile bilious vomiting, she reported to have vomited five times prior to admission. Vomitous was foul smelling, and mainly consisted of watery feculent fluid which was heavily bile stained. There was no history of hematemesis or fever.

In the same duration, patient gave a history of progressive abdominal distension and absolute constipation. Patient was a primary school girl who had been experiencing normal menses and had no history of previous surgery or any history suggestive mental illness. Also, she had no history of similar complaints in the past and generally gave no history suggestive of impaired mastication, abnormal deglutition or chronic diseases such as diabetes mellitus.

On general examination we saw a young girl in good nutritional status, afebrile, not pale, and moderately dehydrated. She was well oriented but looked rather anxious and distressed.

She had a pulse rate of 116 beats per minute, regular with good volume, her blood pressure was 110/60 mmHg, with obvious tachypnea. Per abdomen examination revealed moderately and uniformly distended abdomen which was moving with respiration. There was visible peristalsis on the epigastric region.

The abdomen was rather tense with slightly generalized tenderness. There was no obvious palpable mass or organomegaly. Tympanic note was elicited on percussion. Bowel sounds were present with a high-pitched metallic tinkling tone. Digital rectal examination revealed an empty rectum. The rest of systemic examination was essentially normal.

She had complete blood count (CBC), serum electrolytes, blood grouping and cross matching, abdominal ultrasonography as well as plain abdominal X-rays. Complete blood count revealed moderate leucocytosis where as serum electrolytes showed mild hypokalemia. Abdominal ultrasonography showed moderate amount of fluid in peritoneal cavity and dilated loops of small intestine.

Erect view of the plain abdominal radiograph showed multiple air fluid levels in a step ladder pattern, there was no free gas under the diaphragm. Supine view revealed diffuse gaseous distension of small bowel loops (Figure 1A–B).

In view of the history, physical examination and investigations a diagnosis of small bowel obstruction was made, though at this juncture we could not precisely ascertain the exact underlying aetiology. After adequate resuscitation and obtaining informed consent from the

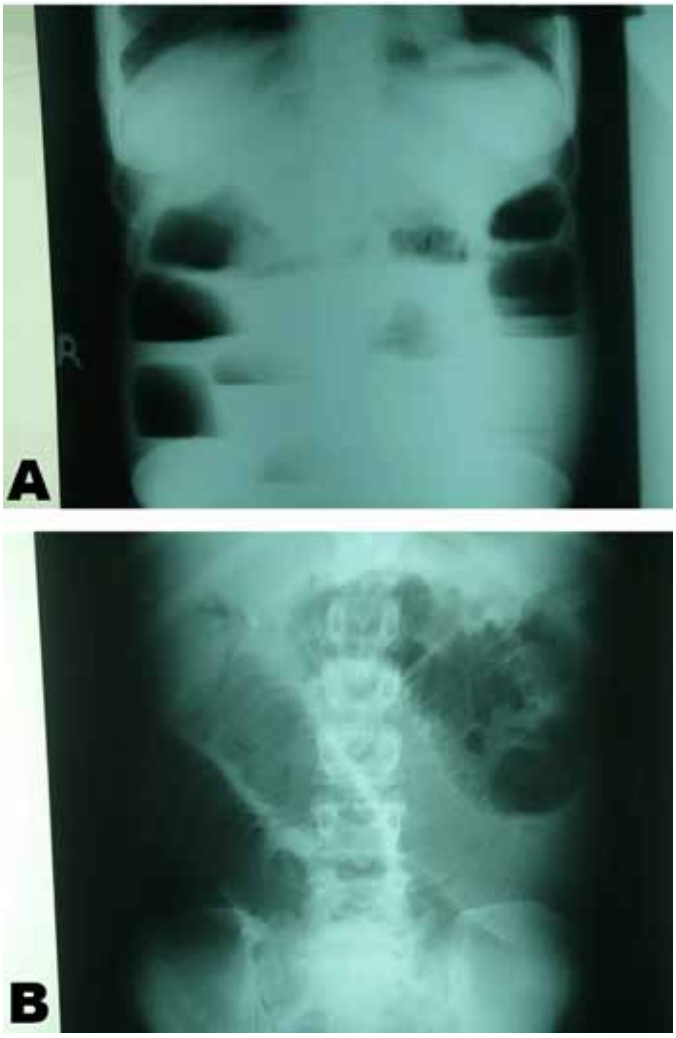

Figure 1: (A) Erect view of the plain abdominal radiograph showing multiple air fluid levels in a step ladder pattern with no free gas under the diaphragm, (B) Supine view of the plain abdominal radiograph showing diffuse gaseous distension of small bowel loops.

patient's parents, she was planned for emergency surgery whereby laparotomy was performed, through a midline incision which revealed significantly distended proximal ileal loops with an impacted palpable intraluminal hard mass which was well circumscribed and situated at about 55 cm from the Ileocecal junction, small bowel distal to the mass was collapsed (Figure 2).

Endeavors to carry out extramural fragmentation of this firm mass by using fingers or atraumatic babcock forceps were futile due to its large size and consistency. Therefore, we had to resort to enterotomy.

Enterotomy was performed and a darkish unclean foul smelling bezoar was found. The ileal bezoar measured 6×4×4 cm, was completely delivered out with minimal difficulties (Figures 3 and 4). There were no synchronous existence of primary bezoars noted in the stomach, duodenum or proximal jejunum. The

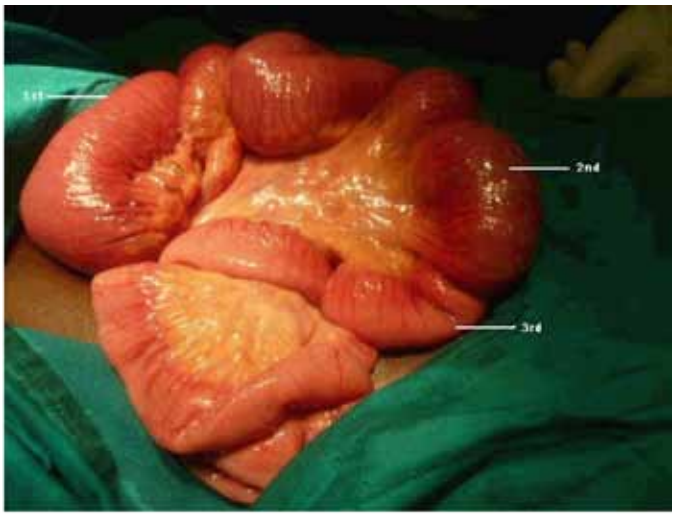

Figure 2: Laparotomy revealed significantly distended proximal ileal loops (1st arrow) with impacted mass in the ileum situated at about 55 cm from the ileocecal junction (2nd arrow). Note, collapsed bowel distal to the impacted mass (3rd arrow).

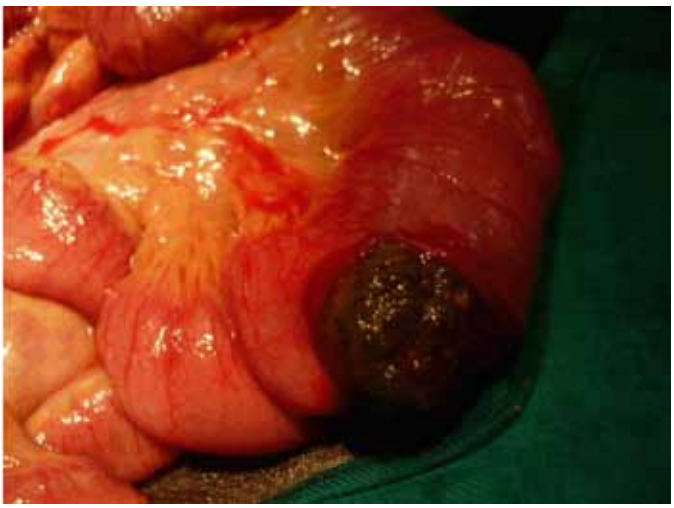

Figure 3: Enterotomy showed well defined impacted isolated bezoar which measured 6x4x4 cm

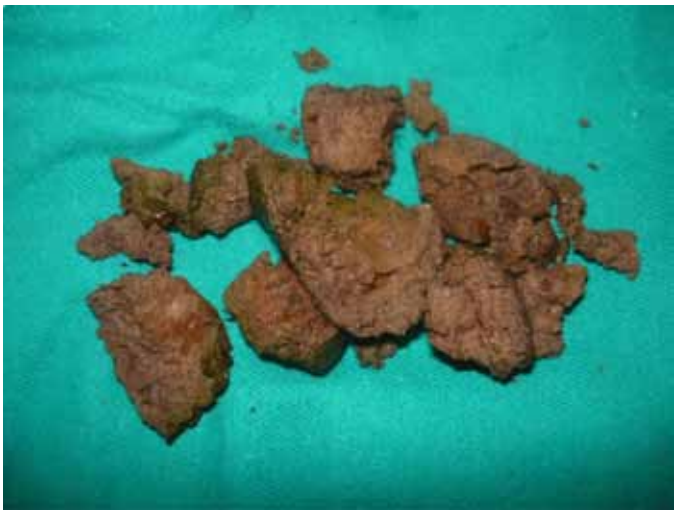

Figure 4: Macroscopic appearance of the fragmented bezoar which was successfully removed after enterotomy.

postoperative period was uneventful and the patient was well and had no complaints at last follow-up six months postsurgery.

DISCUSSION

Our reported patient had no obvious predisposing factors which lead to small bowel bezoar formation and subsequently bowel obstruction. Bezoar formation in rather healthy subjects has been previously reported though still remains an extremely infrequent clinical entity [2–4].

Bezoars mostly stem from the stomach and occasionally drift into small bowel. Primary small bowel bezoar development may be foreseeable in patients with underlying small bowel disease such as stricture due to previous surgery, tuberculosis or Crohn's disease. Other predisposing factors include poor mastication, bolus intakes of indigestible vegetables, diverticuli formation, and small bowel tumors. It is hypothesized that these conditions provide areas of sufficient stagnation within a dilated bowel segment for a bezoar to conglomerate over time [2–4].

Often patients exhibit diverse clinical presentation depending on the size, type and location of the bezoar. The most common clinical features comprises abdominal pains, poor appetite, vomiting, weight loss and anemia. Also patients may present with a wide range of gastrointestinal tract complications such as ulcer formation, bleeding, pressure necrosis, perforation and intestinal obstruction [1–4].

The most frequent site of obstruction is at the level of the gastric outlet or duodenum. Obstruction of distal parts of the small bowel or large bowel remains a rarity occurrence. Features suggestive of psychiatric ailment

and other chronic disease like diabetes mellitus or hyperthyroidism may as well be encountered [1–5].

The diagnosis of bezoar as an underlying aetiology of gastrointestinal obstruction has always been arduous. In most instances, a definitive diagnosis has been made during surgery. Usually apart from hematological and biochemical work up, a battery of radiological investigations have been recommended for detection of bezoars. These include the following plain abdominal X-ray, barium studies, ultrasonography, magnetic resonance imaging (MRI) scan, and computed tomography (CT) scan [1–6].

A CT scan is very useful diagnostic tool in assessing patients with bezoars, it clearly delineate the site, size and nature of a bezoar. Characteristically, bezoar is revealed as a well-defined intraluminal mass with mottled gas on CT scan. When available, MRI is equally good in evaluating patients with suspected bezoar induced small bowel obstruction [1–3, 6].

Though not to the same magnitude such as CT scan or MRI, barium studies may as well be employed to confirm the presence of bezoar in the gastrointestinal tract and at the same time aid to detect other complications such as ulcers [1–4]. Abdominal ultrasonography is another reliable modality in the diagnosis of gastrointestinal bezoars. Characteristically, sonographic visualization of an intraluminal mass with a hyper echoic arc-like surface and a marked acoustic shadow is greatly indicative of a bezoar [6].

Unlike the latter imaging modalities, plain abdominal X-ray mostly show features of bowel obstruction and occasionally depict an opaque intraluminal soft tissue air-containing mass which may be highly suggestive of bezoar [1–3].Generally, plain abdominal X-ray lacks fine details and remains rather limited in evaluation of patients with bezoars.

Where accessible, use of endoscopy is extremely crucial. Endoscopy could be utilized for both diagnostic and therapeutic purposes. Endoscopic examination allows visualization of both gastric and small bowel bezoars and where possible bezoar may be retrieved endoscopically [1–4].

Often treatment of bezoars entails a wide range of options contingent upon the size, site and nature of the bezoar. However, retrieval of bezoar remains the core and primary treatment goal. In some cases surgical removal by gastrostomy or enterotomy is adequate but in others segmental bowel resection may be warranted due to bowel necrosis or denuded serosa [1–5].

The use of endoscopy and laparoscopic gadgets is becoming increasingly popular treatment modality for bezoar removal despite the fact that they may be of limited use when a bulky or rigid bezoar is involved [2, 3, 7]. Bezoars dissolution therapy has been mentioned in some series, however, by far there has been no tangible success with this treatment modality [2]. By and large, a timely intervention carries an excellent prognosis.

CONCLUSION

Small bowel obstruction resulting from solitary bezoar formation without synchronous existence of primary gastric bezoar or any apparent predisposing factor remains an exceedingly rare presentation. Due to its rarity usually pose a significant diagnostic challenge and may subsequently lead to treatment delay and dreadful ramifications. Therefore, good clinical acumen, high index of suspicion, early detection of bezoar and prompt intervention are central in reducing morbidity and mortality.

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Author Contributions

Sefu Juma Uledi – 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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Retained fecaliths after laparoscopic appendectomy disappearing spontaneously with non-operative management

Hideki Katagiri, Mai Ishitani, Takashi Sakamoto, Yasuo Yoshinaga, Tadao Kubota, Akira Miyabe

ABSTRACT

Introduction: Intra-abdominal abscess after laparoscopic appendectomy is a well-known complication. In cases of perforated appendicitis, the frequency of postoperative intra-abdominal abscess formation can be up to 20%. However, intra-abdominal abscess due to retained fecaliths has rarely been reported. A retained fecalith following appendectomy is a rare complication and it has been reported that retained fecaliths should be removed immediately after their diagnosis because of its potential to cause abscess. We present a rare case of retained fecaliths after laparoscopic appendectomy which disappeared spontaneously with non-operative management.

Keywords: Retained fecaliths, Laparoscopic appendectomy, Intra-abdominal abscess

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Katagiri H, Ishitani M, Sakamoto T, Yoshinaga Y, Kubota T, Miyabe A. Retained fecaliths after laparoscopic appendectomy disappearing spontaneously with non-operative management. International Journal of Case Reports and Images 2013;4(11):650–653.

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INTRODUCTION

A fecalith is often detected in cases of acute appendicitis. It can drop pre- or intraoperatively into the peritoneal cavity [1]. The frequency of retained fecaliths after appendectomy is unknown and only a few case reports have been published [2]. Postoperative abscess after appendectomy is a well-known complication and, in cases of perforated appendicitis, the frequency can be up to 20% [3]. A retained fecalith can cause intra-abdominal abscess and the abscess often relapses despite adequate drainage [4]. Previous reports recommended the removal of complicated fecaliths after diagnosis. We present a very rare case of retained fecaliths after laparoscopic appendectomy which disappeared spontaneously with non-operative management.

CASE REPORT

A 70-year-old male was brought to emergency department complaining of abdominal pain for last three days. He had a past history of myocardial infarction, paroxysmal atrial fibrillation, hypertension, chronic renal failure and benign prostate hypertrophy. On physical examination, he showed severe tenderness of the right lower quadrant and generalized peritonitis. Computed tomography (CT) scan revealed a swollen appendix with several high density areas, which were compatible with fecaliths, and pelvic fluid was detected (Figure 1). We diagnosed him with perforated appendicitis and performed emergency laparoscopic appendectomy. On exploring the peritoneal cavity, a small amount of dirty peritoneal fluid was detected and the appendix had perforated. During the operation, some fecaliths had

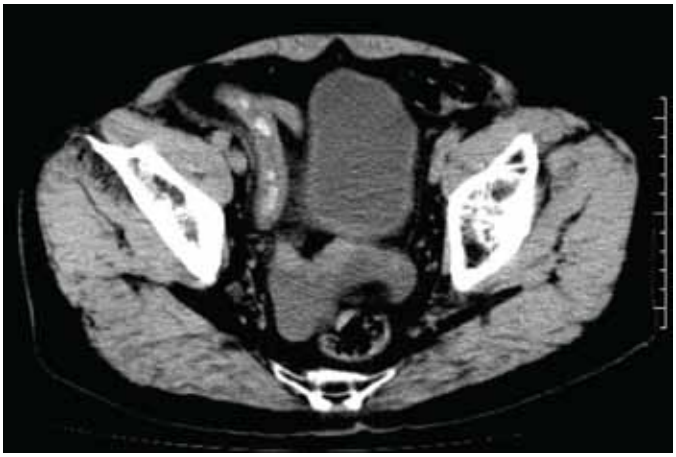

Figure 1: Computed tomography scan of the abdomen demonstrating a swelling appendix and fecaliths inside with a small amount of peritoneal fluid.

dropped into the peritoneal cavity. We retrieved the fecaliths, followed by removing the appendix.

After surgery, a slight fever and paralytic ileus prolonged for several days. We suspected an intra-abdominal abscess, and CT scan of abdomen was done. The CT scan revealed small, high-density areas in the peritoneal cavity, which were compatible with retained fecaliths, and small abscesses around them (Figure 2A–B).

As the patient refused further surgery, we administered antibiotics and decompression with a nasogastric tube. Fortunately, his symptom improved, and he was discharged after 22 days of surgery.

Three months later, a check-up abdominal CT scan was done. The CT scan revealed no intra-abdominal abscess and the fecaliths had disappeared (Figure 3).

DISCUSSION

Acute appendicitis is one of the most common surgical emergencies in daily practice. It affects approximately 7% of the population over the lifetime [2]. There are several complications after appendectomy and the most common is infection. It occurs typically in patients with a perforated appendicitis. In cases of perforated appendicitis, the risk of a dropped fecalith is high. It can also drop at the time of resection of the appendix, during forceful extraction through the port, or when the appendix perforates [1]. Fecaliths are composed of inspissated fecal material, mucus with trapped calcium phosphate and inorganic salts [1]. To avoid spilled fecaliths, gentle manipulation of the acutely inflamed or gangrenous appendix and an endoscopic bag or pouch to facilitate removal should be employed [5].

A dropped gallstone during laparoscopic cholecystectomy is a relatively common complication. However, a dropped and retained fecalith after appendectomy is rare and its frequency is unknown.

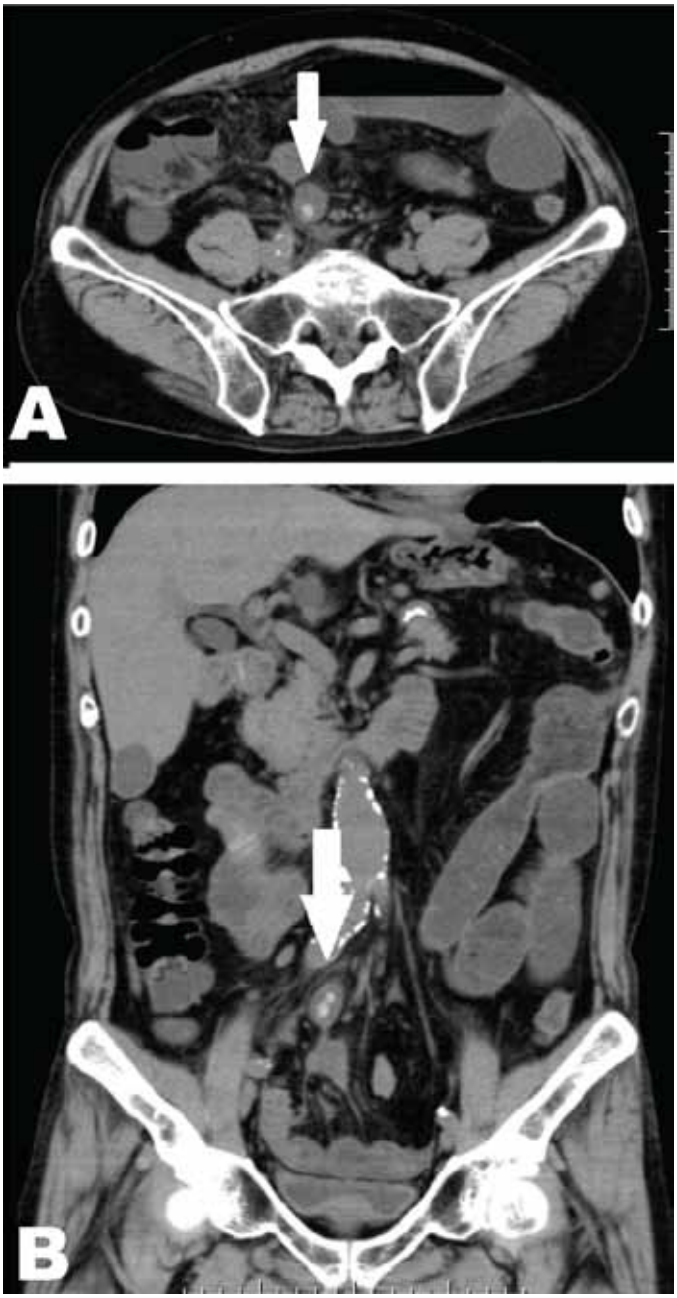

Figure 2: (A, B) Computed tomography scans of axial and coronal views of the abdomen. Retained fecaliths and surrounding abscesses were detected (arrow).

Compared with a gallstone, a retained fecalith has the potential risk of causing an intra-abdominal abscess [5]. Days to years can elapse between an appendectomy and clinical manifestations of a retained fecalith [2, 6]. Patients with retained fecaliths present abdominal pain, fever, elevated white blood cell count, or a combination of these findings [2, 4]. In this patient, slight fever has persisted but he did not complain of abdominal pain.

An intra-abdominal abscess due to a retained fecalith often relapses despite adequate drainage [4], so treatment should not only involve drainage but also removal of the fecalith. The management of a complicated

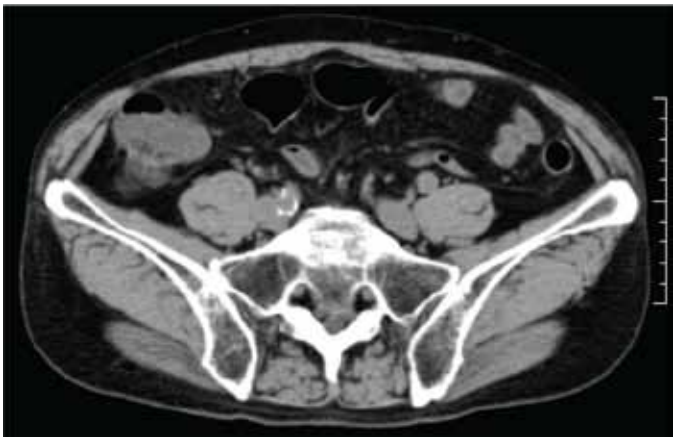

Figure 3: Computed tomography scan of the abdomen, three months after surgery. There was no evidence of a retained fecalith or abscess.

fecalith most commonly described in literature is open or laparoscopic surgery [1, 2, 4, 5]. Percutaneous extraction of the fecalith has also been reported [6]. Preoperative and intraoperative localization of fecalith using guidewire or intraoperative ultrasound as majority of cases locating a fecalith can be difficult in laparoscopic surgery. Prevention of this complication should focus on avoidance of dropping fecaliths.

In this patient, the retained fecaliths were enclosed by an intra-abdominal abscess and they caused postoperative ileus. We considered surgical removal. However, the patient refused surgery, and the location of abscess was hard to approach percutaneously, so we continued nonoperative management with antibiotics. Fortunately, his symptoms improved and the retained fecaliths diminished. To our knowledge, this is the first reported case of retained fecaliths disappearing without surgical procedures.

At present, the diagnosis of a fecalith is mainly based on CT scan. The CT scan can effectively detect calcification, so the feces, which are not calcified completely, might be revealed as fecaliths. Such fecaliths can possibly be treated as tiny abscesses.

CONCLUSION

A retained fecalith after appendectomy is a rare complication. However, surgeons have to be aware of this rare complication. We have experienced a very rare case of retained fecaliths after laparoscopic appendectomy which disappeared spontaneously with nonoperative management. In some patients with retained fecaliths, there might be cases not requiring surgical procedures.

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Author Contributions

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

Mai Ishitani – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Takashi Sakamoto – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Yasuo Yoshinaga – Conception and design, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

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

Akira Miyabe – Conception and design, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

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CASE REPORT OPEN ACCESS

Intravenous leiomyomatosis diagnosed by catheter-based contrast venography

Huabin He, Qun Chen

ABSTRACT

Introduction: Intravenous leiomyomatosis (IVL) is a rare histologically benign smooth muscle tumor that often originates form uterus and common iliac vein, sometimes extends to the right heart. Case Report: A rare case diagnosed in catheter-based contrast venography with a pathological finding is described, together with a comparative analysis of all the previous published cases. Conclusion: Intravenous leiomyomatosis has some distinctive features on catheter-based contrast venography. The imaging technique can better detect the intravenous leiomyomatosis consecutive from uterus. Based on the result of the study, it is recommend that catheter-based contrast venography should be considered as an investigative tool in the presence of cardiac or inferior vena cava tumor.

Keywords: Intravenous leiomyomatosis, Catheterization, Contrast venography, Angiography images and videos

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INTRODUCTION

Intravenous leiomyomatosis (IVL) is a rare histologically benign smooth muscle tumor that usually affects premenopausal women. Fewer than 100 cases of intracardiac leiomyomatosis have been described so far in literature [1]. Its cardiac extension is often overlooked or misdiagnosed because of its low morbidity. The imaging appearance of IVL is non-specific, but the major role of imaging examination is to define the extent of the lesion [2]. Therefore, it is necessary to find a more sensitive diagnostic method for IVL.

Herein, we report our experience with the use of catheter-based contrast venography in a patient with suspected IVL to directly determine its characteristics, the extent of the lesion.

CASE REPORT

A 44-year-old female was admitted to our department with a five-day history of mild to moderate acute pain and edema of the left lower extremity. Total abdominal and pelvic ultrasound demonstrated a left-sided abnormal uterine adnexal tumor and an intrauterine device (IUD) before admission. She had no relevant past medical history. Physical examination demonstrated a regular sinus rhythm with heart rate 82 beats/min and a grade 3/6 systolic murmur was heard at the apical region. Laboratory findings were generally non-specific. Deep vein thrombosis (DVT) was suspected on the basis of clinical practice.

During peripheral vascular catheterization, a 5F pigtail catheter was introduced through the common femoral vein to perform angiography in different parts of the body, which showed a dilated left iliac vein containing a mass extending into the right heart chambers. Pelvic angiogram revealed the left common iliac vein was mildly dilated and obstructed by the mass compared to the right common iliac vein (Figure 1). Images showed an elongated mass (150.03×16.24 mm) in the lumen of the IVC extending from inferior vena cava (IVC) to the right atrium (Figure 2). When the catheter was shown passing through superior vena cava, intracardiac angiography demonstrated a mobile oval-shaped mass (62.50×35.85 mm) in the right atrium (Figure 3). The tumor of IVC was dissected and isolated under low temperature and cardiopulmonary bypass through partial sternotomy and median laparotomy. Uterus had not been touched. The removed mass was smooth and encapsulated. Pathology, subsequently, warranted the diagnosis of intravenous leiomyomatosis of the IVC (Figure 4). The histopathology examination reported proliferation of smooth muscle bundles without any atypical cellular signs excluding leiomyosarcoma and angiomyolipoma. After 12 months from the surgery, there was no recurrence of intravenous leiomyomatosis.

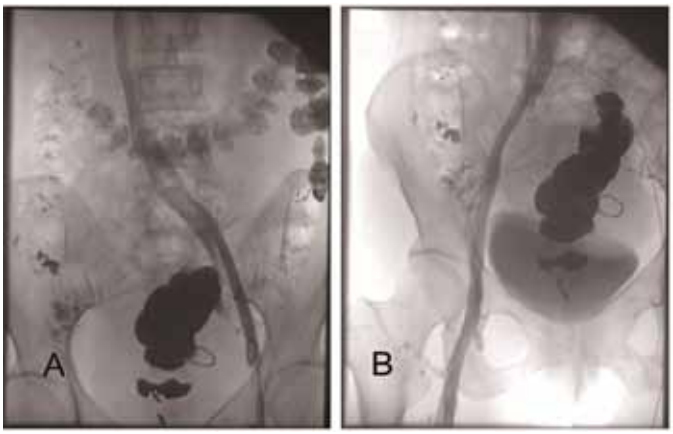

Figure 1: (A) The left pelvic angiogram revealed that the left common iliac vein was mildly dilated obstructed by the mass compared to the right common iliac vein, (B) Right pelvic angiogram revealed a normal right common iliac vein.

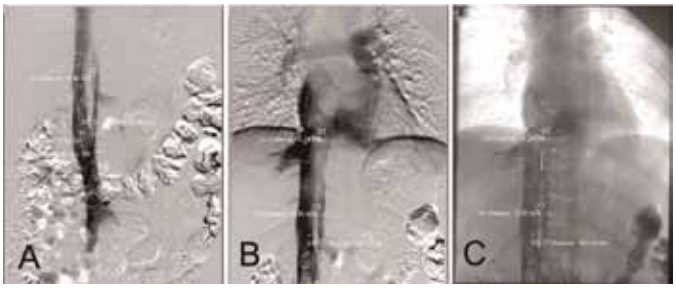

Figure 2: (A, B) Images in the lumen of inferior vena cava showed two cord-like filling defect with a surrounding of contrast extending from inferior vena cava to the right atrium, (C) The two cord-like filling defects overlapped each other in the upper segment of the inferior vena cava.

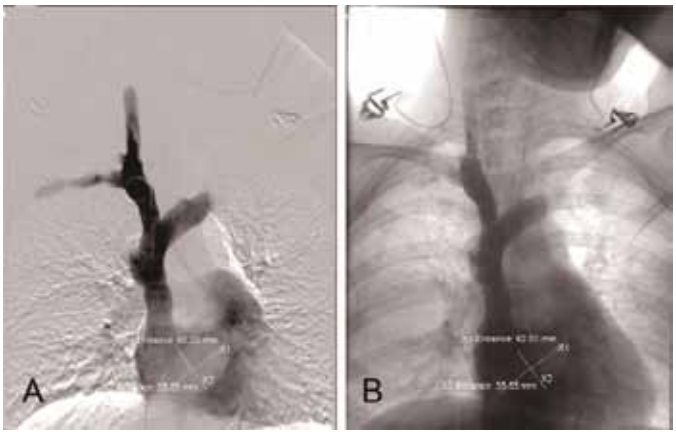

Figure 3: (A, B) Intracardiac images revealed a mobile oval-shaped mass (62.50×35.85 mm) in the right atrium.

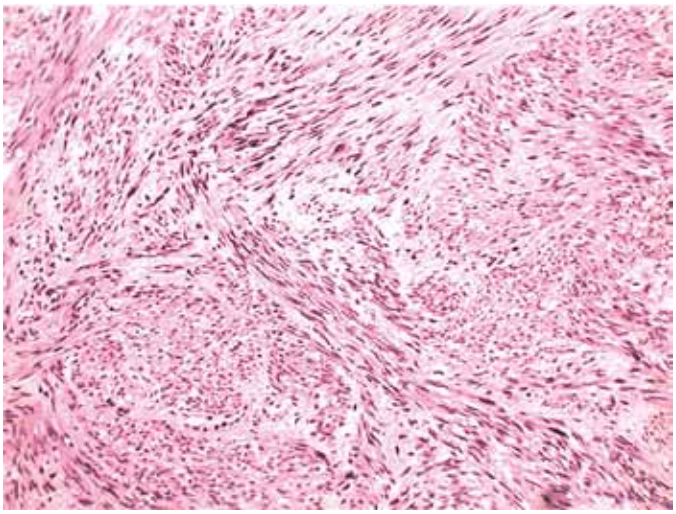

Figure 4: Pathology confirmed the diagnosis of intravenous leiomyomatosis of the IVC (H&E stain, x200).

DISCUSSION

The procedure in our case was performed in catheter-based contrast venography, which had not been reported in previous literature. Angiographic images can provide important information unavailable by other imaging techniques because of its superb-quality intravascular digital subtraction angiography (DSA) images and video, direct multi-angle imaging capability and unique ability to assess intraluminal filling defect by the direct injection of contrast agents in various lesions. Moreover, for some patients with deep vein thrombosis, catheter-based contrast venography plays a therapeutic role by the direct injection of anti-thrombotic drugs.

Radiologically, the differential diagnosis of IVL mainly includes malignant tumor and atrial myxoma. All the tumor emboli of the malignant tumors from kidney and liver have the same imaging features as the primary tumors and are often directly connected with them [3]. The majority of atrial myxomas are found in the left

atrium, and the right atrium is a less common site [4]. The right atrial myxoma has a pedicle attached to the wall of the atrium and generally does not spread to the IVC. Although tumors involving IVC and right atrium have their own imaging features different from those seen on IVL with cardiac extension, the final diagnosis depends on histopathology.

CONCLUSION

In conclusion, an early diagnosis of intravenous leiomyomatosis relies on a high index of suspicion. Intravenous leiomyomatosis has some distinctive features on catheter-based contrast venography, which can clearly show characteristics of the tumor, the path of extension, and anti-thrombotic therapy. All of these are very important to the diagnosis, surgical operation plan, prognosis, and the follow-up of tumor. Catheter-based contrast venography could have unique advantages for diagnosing intravenous leiomyomatosis. With our experience, the imaging technique can better detect the intravenous leiomyomatosis consecutive from uterus. Based on the result of the study, it is recommend that catheter-based contrast venography should be considered as an investigative tool in the presence of cardiac or inferior vena cava tumor.

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Author Contributions

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

Qun Chen – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

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CLINICAL IMAGE OPEN ACCESS

A giant fecaloma in a seven-year-old healthy boy

Yuji Koike, Yasutomi Kuroki

CASE REPORT

A seven-year-old Japanese boy was presented with severe abdominal pain and vomiting. The patient had otherwise been well until he developed constipation at three years of age. One month before the current visit, he took a one-week domestic trip with his family. Since then, he had been constipated and visited physicians because of abdominal pain and was treated with glycerine enemas and laxatives. Thereafter, he only demonstrated watery stool. The patient had no history of Hirschsprung disease, abdominal surgery or the frequent use of anticholinergics or narcotics.

On examination, his abdomen was flat and bowel sounds were present. There was a large hard mass in his lower abdomen with tenderness, although there was no guarding or rebound tenderness. The edge of the mass was also smooth and palpable on a rectal digital examination. A plain X-ray of the abdomen obtained showed a large, round mass with laminar contents in the pelvis (Figure 1). Flexible coloscopy revealed a large fecal ball (fecaloma) in the upper rectum with a normal mucosal membrane (Figure 2). The next day, the patient was admitted to our hospital to remove the fecaloma under general anesthesia. It was too hard to crush colposcopically, and therefore, we had to press and crush it externally and finally remove it manually. The restored fecaloma was a cannonball-like shape measuring 6×10 cm in diameter and weighing 260 g. Thereafter, the patient showed a good recovery and had normal bowel movements every day. He had been in good condition for more than one year.

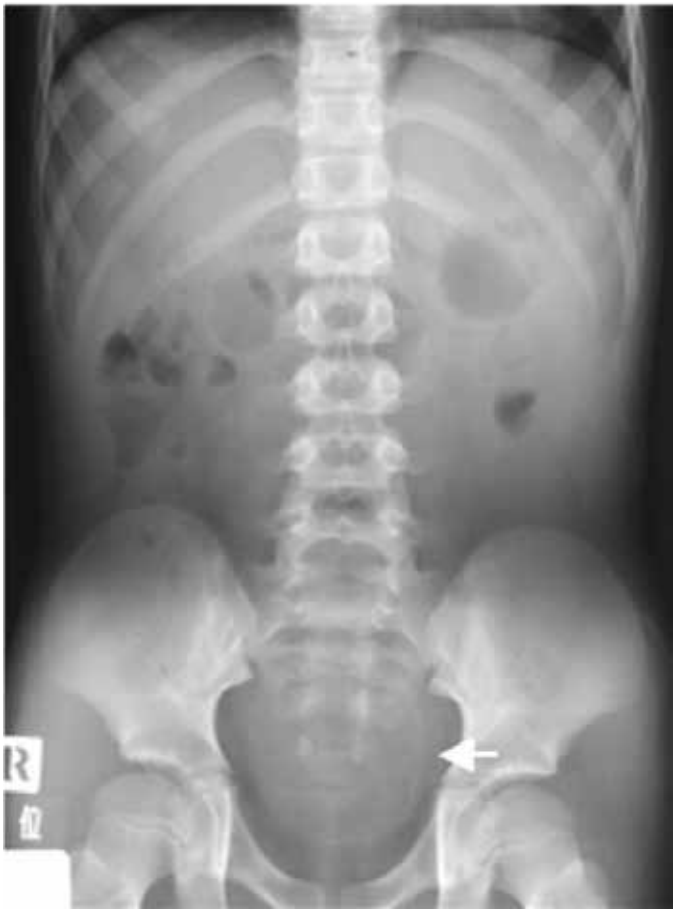

Figure 1: A plain X-ray of the abdomen showing a large, laminated mass in the pelvis (white arrow).

DISCUSSION

Fecal impaction sometimes grows into a hard and laminated large mass known as a fecaloma which may present features of acute gastrointestinal tract pain or mimic a carcinoma. Patients with fecaloma tend to have underlying conditions such as being elderly, having

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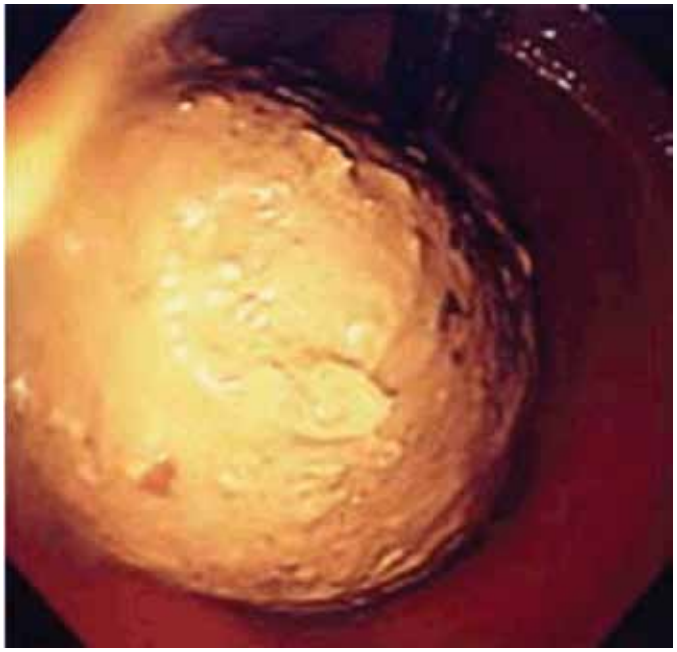

Figure 2: Flexible coloscopy showing a fecal ball in the upper rectum.

experienced previous abdominal surgery or having abused drugs (e.g., anticholinergics or narcotics). Although there have been a few reports concerning fecaloma in children complicated with encopresis, cerebral palsy, or Hirschsprung disease, reports of children with giant fecaloma with no underlying diseases are rare [1, 2].

Fecal impaction in the rectum often becomes difficult and painful to evacuate in a constipated child. Therefore, more retention occurs and a vicious cycle is accomplished [3]. Patients with fecaloma often present with abdominal pain and discomfort, vomiting, and weight loss. Interestingly, overflow diarrhea tends to sometimes be observed in such patients because the liquid stool is evacuated by the movement of the colon through a void between the fecaloma and the colonic wall, as was observed in our patient. As the large hard mass is usually palpable in the abdomen in almost all patients, fecaloma may sometimes be confused with a colonic malignancy [4, 5]. It is not so difficult to accurately diagnose fecaloma, however, if one notes the characteristic features of plain abdominal X-rays which normally suggest laminar components with fecal impaction.

Finally, balanced diet that includes fruits, vegetables and liquids is recommended as a fundamental management of constipation in children. Low fiber intake has been shown to be a risk factor for chronic constipation. Carbohydrates in prune, pear and apple juices may increase water content in stools [6]. Caretakers should be aware of the dietary modification for their children who have hard bowel movements.

CONCLUSION

Clinicians should therefore be aware that fecal impaction can grow into a giant fecaloma even in a healthy child. Furthermore, a detailed history and a plain X-ray can provide important diagnostic clues in such patients.

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Koike Y, Kuroki Y. A giant fecaloma in a seven-year-old healthy boy. International Journal of Case Reports and Images 2013;4(11):657–659.

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Yuji Koike – 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

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

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CLINICAL IMAGE OPEN ACCESS

Transverse and sigmoid sinus thrombosis after traumatic brain injury

Andrew Coker Wiggins, David P Nguyen, Nawaar Al-Rawas, Karen Laauwe, Latha Ganti

CASE REPORT

A 40-year-old male was presented to the emergency department with complaints of diffuse frontal headache with photophobia, nausea and sinus pressure that developed gradually over three days. His medical history was significant for a basilar skull fracture from a motor vehicle collision. He was cleared of his cervical collar two weeks prior. Remaining review of systems was negative. No focal neurologic deficits present. The computed tomography (CT) scan showed an area of increased density in left transverse sinus which was concerning for thrombosis (Figure 1) so a magnetic resonance imaging (MRI) scan was done which confirmed the diagnosis. Magnetic resonance venography demonstrating non-visualization of the left transverse and sigmoid sinuses, and proximal left jugular vein, consistent with thrombus in the vein. He was treated with anticoagulation (Figure 2).

DISCUSSION

Cerebral venous thrombosis (CVT) as an etiology of headache is not always easy to diagnose unless suspected. The clinical manifestations are non specific, and overall, it is still not a common diagnosis, especially in North America [1].

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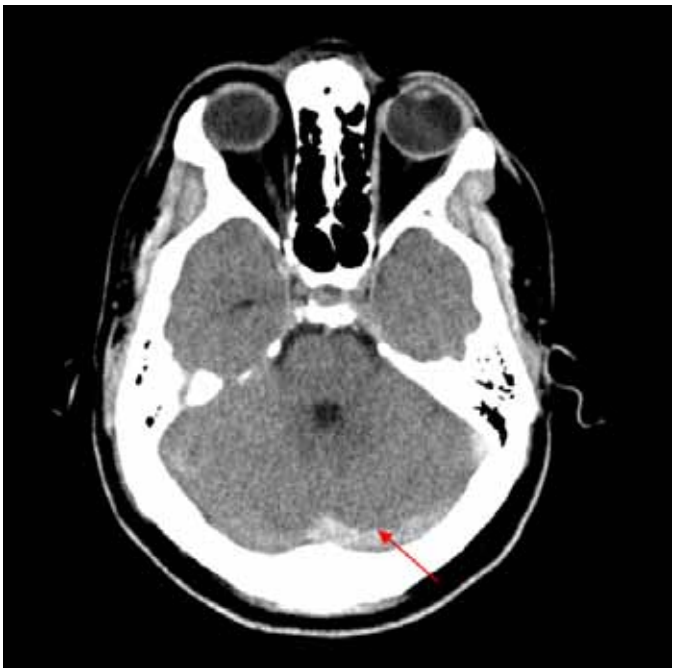

Figure 1: Noncontrast head computed tomography. Arrow points to area of increased density in left transverse sinus, (dense clot sign) which is asymmetric when compared to right side.

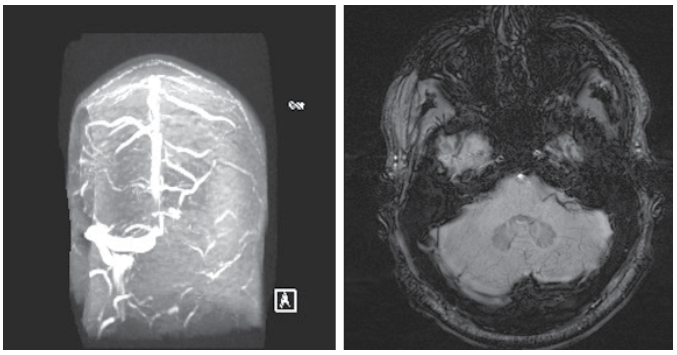

Figure 2: Magnetic resonance venography demonstrating non-visualization of the left transverse and sigmoid sinuses, and proximal left jugular vein, consistent with thrombus in the vein.

Sinus thrombosis is not classically thought of as being associated with closed head injury, even though trauma is certainly one of the etiologies. Rather, the

classic etiologies are neoplasms or hypercoagulable states such as pregnancy. However, with the increasing prevalence of traumatic brain injuries, and imaging for these injuries, it is being recognized more as a risk factor, both in adults and children. The initial imaging study of choice is unenhanced CT scan, which can demonstrate a hyperdensity in the sinus, referred to as a 'dense clot sign', in about 20–50% of cases. The follow-up study is MRI scan and magnetic resonance venography, or CT venography. A retrospective review found a 40% thrombosis rate, and concluded that CT venography should be performed in patients with fractures extending to a dural venous sinus or jugular bulb. In all imaging techniques, a hypoplastic or aplastic dural sinus can mimic thrombosis [2].

CONCLUSION

Given the increasing prevalence of traumatic brain injury, it is important to recognize that several sequelae, perhaps some not commonly recognized, are possible. Maintaining a high index of suspicion and following-up on potentially abnormal initial imaging findings is thus imperative. Untreated, cerebral venous thrombosis can lead to hemorrhagic infarction and death.

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Parvaiz MA, Isgar B, Aluwihare N. 'Wallpaper paste sign' of mucinous breast carcinoma. International Journal of Case Reports and Images 2013;4(11):660–662.

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Conflict of Interest

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LETTER TO EDITOR OPEN ACCESS

Novel oral anticoagulants after gastric bypass surgery: Caveat emptor

Daniel J Lachant, Imran Uraizee, Rohit Gupta, Angelo J Pedulla

To the Editor,

The Food and Drug Administration recently approved two new classes of oral anticoagulants: direct thrombin inhibitors, dabigatran etexilate and factor Xa inhibitors, apixaban and rivaroxaban [1]. All these three drugs are for the treatment of nonvalvular atrial fibrillation with rivaroxaban also gaining approval for venous thromboembolism. Absorption of each drug is regulated by P-glycoprotein efflux transporters and is variable. The bioavailability of dabigatran etexilate is 7% with peak effect within two hours after administration. Apixaban has a 50% bioavailability with peak effect occurring 3–4 hours after administration and rivaroxaban has a bioavailability greater than 80% with peak effect occurring 2–4 hours after administration [1].

A P-glycoprotein is located on the apical membrane of enterocytes, act by pumping substrates back into the intestinal lumen, thereby limiting drug absorption. Its concentration gradually increases from the stomach to the distal part of the intestine. Above a certain concentration, efflux outpaces influx preventing absorption. All three drugs have warnings against co-administration with medications that induce P-glycoprotein expression, resulting in increased P-glycoprotein concentration earlier in the gastrointestinal tract leading to decreased drug absorption [1, 2].

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With the current obesity epidemic, an estimated 180,000 bariatric procedures are performed annually in the United States. The most common intestinal diversion procedure is the Roux-en-Y gastric bypass. The consequences of this procedure are a 95% reduction in gastric capacity as well as a reduction in the functional length of the gastrointestinal tract from bypass of the duodenum and proximal jejunum. These changes augment the effect of P-glycoprotein on limiting drug absorption, similar to the effect medications have on inducing P-glycoprotein expression. The surgery has been linked to nutritional deficiencies but has not been extensively studied for reductions in drug absorption [2, 3].

We report a 41-year-old male admitted to the cardiac care unit with chest pain while taking dabigatran etexilate 150 mg twice daily and dofetilide 500 μg twice daily for atrial fibrillation. Computed tomography (CT) scan revealed a saddle pulmonary embolus. His activated partial thromboplastin time (aPTT) was 25.2 seconds (normal 22.3–35.3 seconds), and he had no previous PTT results while on dabigatran etexilate. Electrocardiogram showed normal sinus rhythm. He started dofetilide for uncontrolled atrial fibrillation about 18 months ago and reported compliance with both dofetilide and dabigatran etexilate, taking them earlier that day. He underwent a Roux-en-Y gastric bypass for morbid obesity 21 years ago. After the pulmonary embolus was diagnosed, a heparin drip was started and was safely discharged home on enoxaparin.

This case raises an important issue related to the potential for impaired absorption of the novel oral anticoagulants (NOAC) after gastric bypass. In our case, the normal aPTT suggests no active dabigatran in his plasma since there should have been enough time from when he took his last dose to be reflected by an elevated aPTT [4]. We hypothesize that the normal aPTT while on dabigatran etexilate is explained by the loss of critical absorption necessary to achieve therapeutic plasma levels that normally occurs in the stomach and duodenum, a low P-glycoprotein environment. There is the possibility

that medication noncompliance led to the normal aPTT, and due to safety concerns, he was not re-challenged with dabigatran etexilate and aPTT monitoring to prove whether gastric bypass prevented his absorption. However, the patient mentioned that he was regularly taking his medications, a claim supported by the fact that he was in sinus rhythm on admission.

Despite the known effects of gastric bypass leading to malabsorption in the proximal small bowel, there are no warnings about the potential for decreased absorption of NOAC following gastric bypass surgery. There is a single case report of a bariatric surgery patient noted to achieve therapeutic anti-Xa levels after receiving rivaroxaban [5], although it is not clear, if this patient had a classic Roux-en-Y. This may suggest enough absorption can still occur with rivaroxaban and apixaban as they are not inhibited by the higher concentrations of P-glycoprotein that found earlier in the altered intestinal tract after gastric bypass surgery because absorption normally occurs more distally in a higher P-glycoprotein concentration compared to dabigatran etexilate.

Currently, there are no warnings or recommendations regarding the use of NOAC following gastric bypass surgery. This is a large group at risk for developing venous thromboembolism and atrial fibrillation and subsequently starting one of these NOAC, especially, if they have had difficulties on warfarin with its variable absorption after gastric bypass [4]. Unfortunately, we are unable either to prove or disprove whether the normal aPTT was from impaired absorption or noncompliance. With the lack of evidence at this time further studies are needed to assess whether post bypass patients are capable of obtaining a therapeutic level with the current dosing recommendations with the NOAC. Until assays are available to ensure adequate plasma levels are achieved, there remains the potential for sub-therapeutic dosing.

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

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

Angelo J Pedulla – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article. Final approval of the version to be published

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