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CASE REPORT

OPEN ACCESS

Genital tuberculosis mimicking ovarian malignancy: A series of two cases

Rekha Sachan, ML Patel, Malti Maurya, Pushpalata Sachan

ABSTRACT

Introduction: Genital tuberculosis is a common occurrence in developing countries such as India where pulmonary tuberculosis is widespread. **Case Series:** We hereby discuss two interesting case reports. First case was a 34-year-old female who presented with bilateral ovarian mass with high level of serum CA-125. Second case was a 25-year-old female who presented with complaint of infertility, huge lump and pain in lower abdomen in a reproductive age, both mimic like ovarian malignancy but ultimately diagnosed as case of genital tuberculosis. **Conclusion:** Genital tuberculosis should be ruled out before performing any extended surgery. Both hydrosalpinx and ovarian malignancy present as bilateral adnexal masses, transabdominal ultrasonography was also non discriminative, serum CA-125 level was also elevated in both the conditions.

Keywords: Hydrosalpinx, Ovarian malignancy, Tuberculosis, Salpingotomy, Serum CA-125

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INTRODUCTION

Genital tuberculosis is an important cause of infertility in developing countries. It is almost always acquired by hematogenous spread from extragenital sources, primary focus of genital tuberculosis is fallopian tubes causing hydrosalpinx [1]. Pelvic inflammatory disease [2] or ascending infection by Chlamydia or Gonorrhea and sometimes tubercular infection may cause distal tubal occlusion and hydrosalpinx [3]. Hydrosalpinx usually formed due to the blockage of distal portion of fallopian tube which is filled with serous or clear fluid. The blocked tube may get distended and it takes a characteristic sausage or retort like shape [4]. Its true incidence in female is not known because large proportion of patients is asymptomatic. The major presenting symptoms are infertility (45–55%), pelvic pain (50%), poor general health (25%), and menstrual disturbances (20%). The differential diagnosis includes chronic pelvic inflammation, salpingitis and malignancies [4].

CASE SERIES

Case 1: A 34-year-old female (P5+3) with previous five vaginal deliveries followed by three successive induced abortions presented with chief complaints of low grade evening rise fever, anorexia and pain in lower abdomen for the last six months. Her previous history of menstrual cycle was uneventful. From last five to six

months, she was suffering from severe dysmenorrhea. She had her last abortion two years back.

Past history revealed that the woman had undergone laparotomy for the same complain at district hospital about three months back and abdomen was closed as such without any surgical intervention due to presence of dense adhesions between bowel, omentum and peritoneum. On general examination patient was febrile (100°F), and of average built with mild pallor. Abdominal examination revealed a paramedian vertical scar with bilateral adnexal mass without tenderness. On per speculum examination cervix was hypertrophied and circumoral erosion present. Per vaginal examination revealed normal size uterus and bilateral cystic non tender adnexal masses of about 5–6 cm in size attached to uterus with restricted mobility was present. Trans abdominal ultrasonography revealed a normal size uterus with evidence of bilateral heterogenous tubo-ovarian mass of (right 41x69 mm and left 67x69 mm) in size with significant free fluid in cul-de-sac. Culdocentesis was done and aspirated fluid, sent for AFB culture, which was negative for *Mycobacterium tuberculosis*. Mantoux test was 18x16 mm in size on third day and active immunoglobulin for tuberculosis (IgM- TB) was positive. Erythrocyte sedimentation rate (ESR) was raised (34 mm/1st hr) and chest X-ray was normal (Figure 1).

Serum CA-125 was raised to 253.66 IU/mL (normal value <35 IU/mL), which was around eight times higher than the normal value which strongly raised suspicion about ovarian malignancy. Computed tomography (CT) scan of abdomen and PCR for the tuberculosis could not be done due to financial constraints. On the basis of clinical finding and investigation (Mantoux test and IgM-TB), she was consulted to department of pulmonary medicine and they advised category 1 antitubercular treatment as per guidelines of Revised National Tuberculosis Control Programme (RNTCP). After two months of therapy, the patient was re-evaluated and it was found that she was significantly relieved of her symptoms of fever, pain, anorexia and dysmenorrhea. However, ultrasonography did not reveal any regression in the size of the mass but free fluid was disappeared. Repeat serum CA-125 was reported 178.03 IU/mL. Antitubercular treatment was continued for nine months as there was relief in symptoms and serial fall in serum CA-125 levels. The patient was planned for laparotomy for residual tubo-ovarian mass of size (right 36x54 mm and left 52x54 mm) after completing nine months course of antitubercular treatment (four drugs regimen for two months, isoniazid, pyrizinamide, ethambutol and rifampicin and two drugs regimen for four months rifampicin and isoniazid) and at that time serum level of CA-125 was within normal range. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed as patient is willing for the same and not agree for further follow-up. The histopathology of residual mass did not show any evidence of tuberculosis and malignancy. This might be

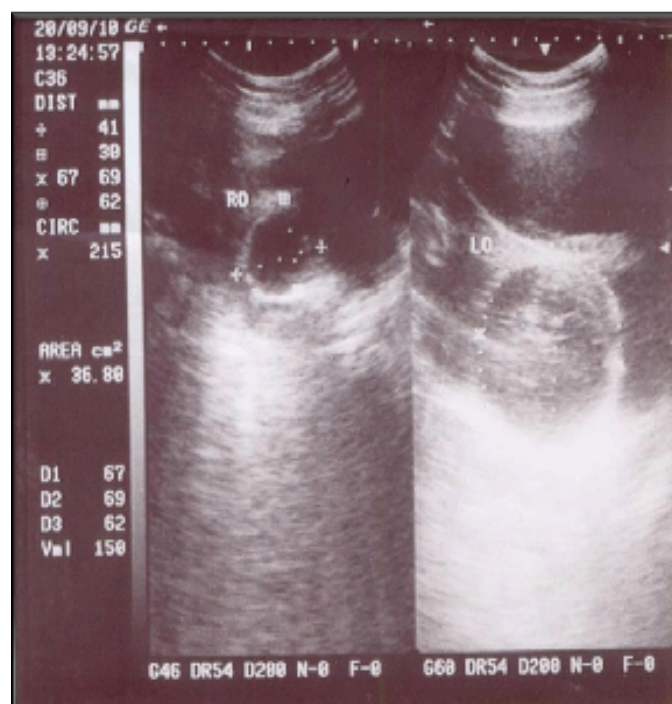


Figure 1: Ultrasonography showing bilateral heterogeneous adnexal mass.

due to patient had completed full course of antitubercular treatment. The postoperative period was uneventful. In follow-up after six weeks patient having no complained and feeling quiet well.

Case 2: A 25-year-old nulliparous infertile women referred to outpatient department with complaints of huge lump in abdomen and pain in lower abdomen since last one month, which was non radiating. She also had a history of generalized weakness. There was no history of fever and vomiting, she was married 12 months back and her menstrual cycles were of 28 to 30 days with average flow for 3 to 4 days and no accompanying dysmenorrhea.

General examination revealed moderate degree of pallor, blood pressure 120/70 mmHg, pulse rate 88 beats/minutes. Patient was a febrile and no lymphadenopathy present. On abdominal examination, there was a large fixed cystic lump present in the lower abdomen slightly deviated towards left measuring about 14x12 cm in size. No abnormality was found on perspeculum examination except cervix was pulled up. On per vaginal examination about 18x12 cm size mass was felt through left fornix which was tense and cystic in consistency and non tender with restricted mobility, uterus could not be felt separately from the mass, another mass of about 8x6 cm, cystic in consistency was felt through right fornix.

Transabdominal ultrasonography revealed a large left adnexal cystic lesion measuring 185x125x75 mm in size with dense internal echoes and mural nodule with anechoic cystic structure within it, another right adnexal cystic lesion measuring 95x92x39 mm in size with septation and dense internal echoes with paraaortic (25 mm and 19 mm) and iliac lymphadenopathy (39 mm and

19 mm size). All laboratory values were within normal limits except serum CA-125 was raised (92.38 U/mL), chest X-ray was unremarkable. A strong suspicion of ovarian malignancy was raised because of raised serum CA-125 along with lymphadenopathy.

An exploratory laparotomy was performed after arrangement of one unit packed red blood cells. Peroperatively a huge hydrosalpinx on left side adhered to omentum with flimsy adhesions to the bowel and peritoneum, on aspiration caseous material was found whereas right sided hydrosalpinx smaller than left was present, uterus and bilateral ovaries were normal (Figures 2–4). Right sided salpingotomy and left sided salpingectomy with adhesiolysis was done. Tissue was sent for histopathological examination. Sections from fallopian tube shows blunt and fused plicae infiltrated with mixed inflammatory infiltrate and well formed granulomas comprising lymphocytes, plasma cells, epithelioid cells and Langhans giant cells were present, suggestive of tubercular salpingitis (Figure 5 and 6). Peritoneal washing and aspirated fluid from cyst was sent for cytological and biochemical analysis. Aspirated fluid biochemical analysis showed protein 2.2 g/dL and sugar 8 mg/dL. So antitubercular treatment was given for six months according to RNTCP (four drug regimen for 2 months and 2 Drug the regimen for four month) along with anti-inflammatory treatment, during follow-up patient was asymptomatic.

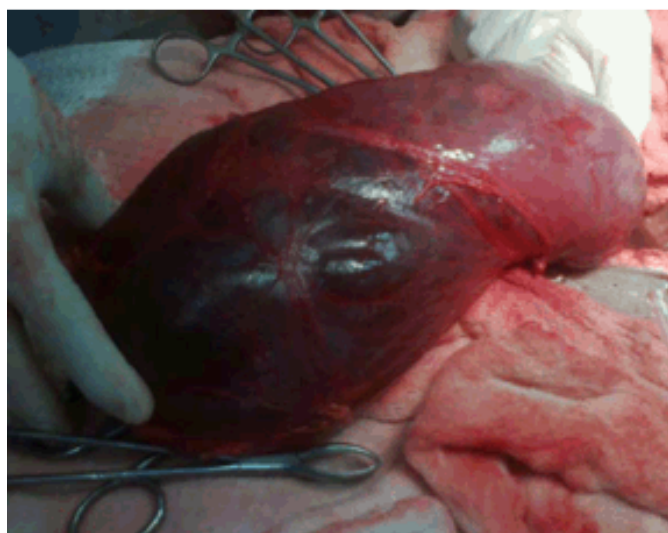


Figure 3: Left sided hydrosalpinx.

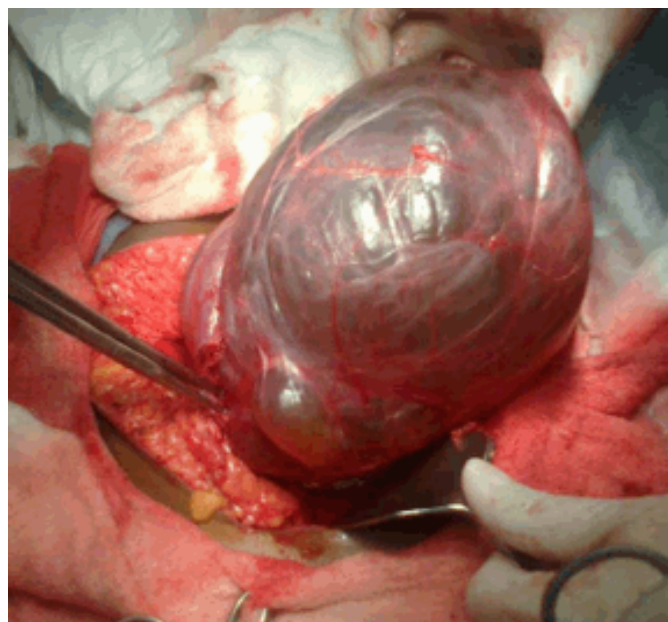


Figure 4: Right sided hydrosalpinx.

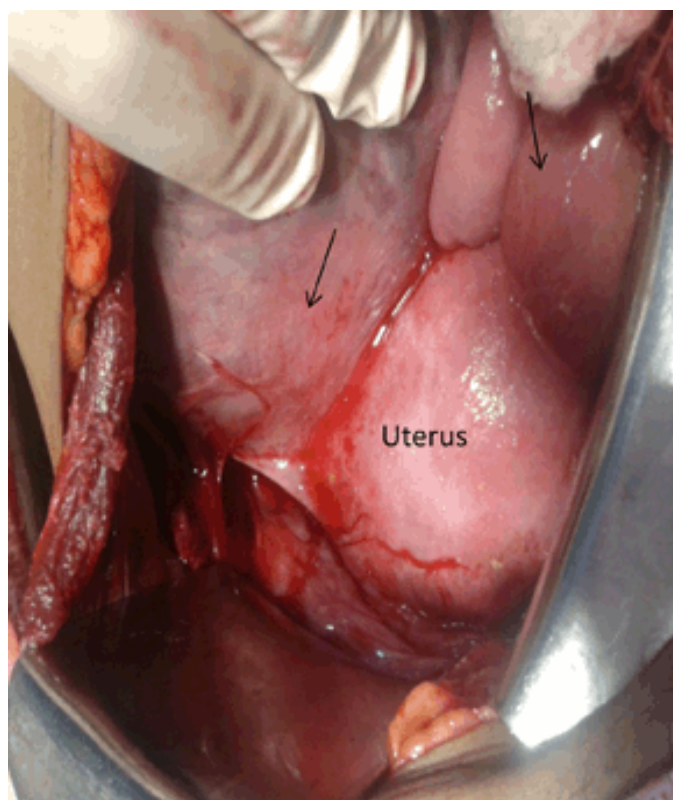


Figure 2: Peroperative finding (arrow) showing bilateral hydrosalpinx with normal size uterus.

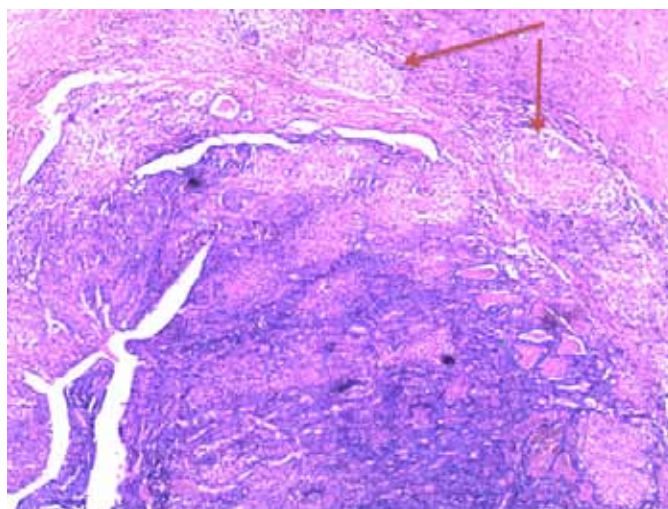


Figure 5: Wall of fallopian tube showing dense chronic inflammatory infiltrate with fused plicae (black arrow) and multiple granulomas (red arrow) (H &E stain, x100).

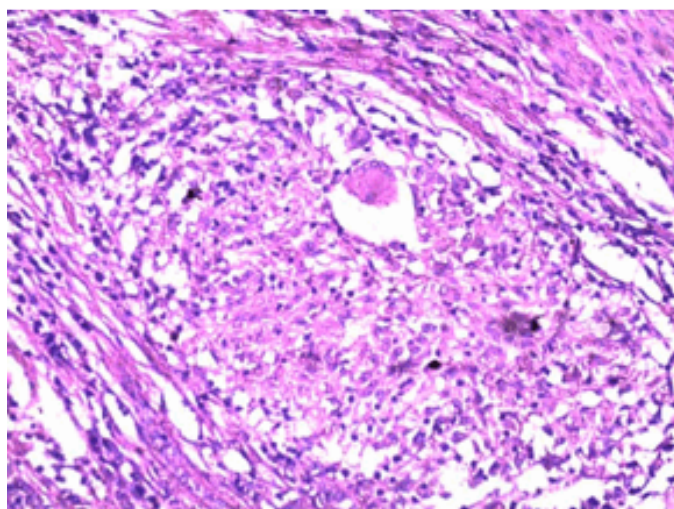


Figure 6: Granuloma showing langhans giant cell epithelioid cells and lymphocytes (H&E stain, x200).

DISCUSSION

Genital tuberculosis may present like ovarian malignancy with elevated serum CA-125 level. For this reason its presence should be ruled out in women presenting with adnexal masses inspite of high levels of serum CA-125 as reported in case one, similar case reported by Manidakis et al. where a case of suspected ovarian malignancy underwent laparotomy, histopathology showed features of typical genital tuberculosis [5]. Ultrasonography did not show any improvements after two months of starting therapy which means that follow-up with ultrasound may not be a very useful tool for evaluation of genital tuberculosis. Serum CA-125 values were falling rapidly after starting antitubercular treatment, which suggests that serum CA-125 may be a useful marker for monitoring and follow-up of genital tuberculosis. Similar conclusion was made by Belgin T et al. in the study who concluded that this marker may be useful for follow-up of patients with genital tuberculosis [6]. Koc et al. also concluded that ascites and high levels of serum CA-125 do not necessarily indicate malignancy in women of reproductive age group [7]. Awareness about peritoneal tuberculosis is still lacking and many women with this disease are initially thought to have ovarian malignancy and undergo unnecessary extended surgery. Similar, past history was present in our case. After the antitubercular treatment total abdominal hysterectomy with bilateral salpingo-oophorectomy was done for residual mass because there was no reduction in the size of mass and no any investigation confirmatory for the tuberculosis, so patient is insisting for the same. Though laparoscopy and guided biopsy to be a sufficient and safe method to provide diagnosis of peritoneal tuberculosis but it was not done as previous record of laparotomy showing dense adhesions [8].

Tubercular hydrosalpinx are almost always bilateral but not symmetrical in size and shape. Tubes are thickened

and show a rough external surface with adhesion, caseous ulceration of mucosa produces ragged contours and diverticular out pouching of both isthmus and ampulla, similarly reported in Case 2.

As tuberculosis heals, the entire tube become encased in heavy connective scar tissue and the lumen develops a beaded rigid stem pipe appearance [9]. The most common site of obstruction is the region of transition between isthmus and ampulla [10]. Hydrosalpinx may be asymptomatic, when it is bilateral, women may be infertile as reported in this case. Other causes of distal tubal occlusion include adhesion formation from surgery and endometriosis. Some patients have recurring lower abdominal pain while others are asymptomatic. Our case also presented with lower abdominal pain.

Generally, treatment for hydrosalpinx is either salpingotomy or salpingectomy so we did left sided salpingectomy and right sided, salpingotomy because our patient was nulliparous [11]. After histopathological examination, it was diagnosed as tubercular salpingitis so antitubercular treatment was given for six months along with anti-inflammatory treatment. Although hydrosalpinx is very uncommon in developed countries but in developing countries still the tuberculosis is important etiological factor responsible for the development of hydrosalpinx [12].

CONCLUSION

This case series demonstrate a high rate of misdiagnosis between advanced ovarian malignancy and genital tuberculosis. Genital tuberculosis should be ruled out before performing any extended surgery. Both hydrosalpinx and ovarian malignancy present as bilateral adnexal masses, transabdominal ultrasonography was also non discriminative, serum CA-125 level was also elevated in both conditions. Whereas abdominal paracentesis is useless in ruling out genital tuberculosis and serum CA-125 levels, may be useful in follow-up of patients.

Author Contributions

Rekha Sachan – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published

ML Patel – Analysis and interpretation of data, Final approval of the version to be published

Malti Maurya – Acquisition of data, Final approval of the version to be published

Pushpalata Sachan – 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

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Right subclavian steal syndrome in a patient with aberrant right subclavian artery, common carotid trunk and hypoplastic left vertebral artery

Boochi Babu Mannuva, Rajasekhar Durgaprasad, Vanajakshamma Velam, Shashanka Chunduri, Vijayalajshmi Devi B

ABSTRACT

Introduction: Subclavian steal syndrome refers to the reversal of vertebral blood flow in the setting of proximal subclavian or innominate artery severe stenosis or occlusion resulting in the symptoms of vertebrobasilar insufficiency related to exercise of the upper extremities. It is more common on the left side. **Case report:** We reported a case of subclavian steal syndrome on the right side in a patient with aberrant right subclavian artery (ARSCA), hypoplastic left vertebral artery and common carotid trunk (CCT) branching in to the left and right common carotid arteries. **Conclusion:** Till now, combination of ARSCA, CCT and left vertebral artery hypoplasia in the same person has not been reported and this combination causes ischemia in large area of brain if the patient develops hemodynamically significant stenosis or occlusion in both CCT and ARSCA proximal to the origin of right vertebral artery.

Keywords: Right subclavian steal syndrome, Aberrant right subclavian artery, Common carotid trunk, Hypoplastic left vertebral artery

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INTRODUCTION

Subclavian steal phenomenon (SSP) is associated with flow reversal in the vertebral artery ipsilateral to a hemodynamically significant stenosis or occlusion of the subclavian artery. Subclavian steal syndrome (SSS) implies the presence of significant symptoms due to arterial insufficiency in the brain (i.e., vertebrobasilar insufficiency) or upper extremity, which are supplied by the subclavian artery. Contorni in 1960, first described retrograde flow in the vertebral artery [1]. In 1961, Reivich first recognized the association between this phenomenon and neurologic symptoms [2]. Fisher coined the term 'subclavian steal syndrome' for this combination of retrograde vertebral flow and neurologic symptoms [3].

Most patients with subclavian artery stenosis are asymptomatic because of the abundant collateral blood supply in the head, neck and shoulder. Symptoms, when they occur, are due mainly to ischemia of the ipsilateral upper extremity. Exercise-induced arm pain, fatigue, coolness, paresthesias or numbness occurs in approximately one-third of patients, but ischemic and trophic changes are rare. Symptoms of vertebrobasilar ischemia (VBI) of brainstem (dizziness, vertigo, ataxia,

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disequilibrium, drop attacks, diplopia, nystagmus, graying of vision, hemianopia, bilateral visual blurring, and syncope) are uncommon and usually occur in patients with concurrent significant stenotic/occlusive disease involving cerebral circulation [4]. In some cases, reduced arterial flow resistance, during upper extremity exercise, can precipitate vertebrobasilar symptoms. Collateral blood supply and the capacity to increase collateral flow determine patients who develop neurologic symptoms.

CASE REPORT

A 53-year-old male with hypertension presented with right arm discomfort, dizziness and presyncope that occurred even with minimal exertion of right upper extremity for the last two months. There was no past history of coronary or vascular disease. Physical examination revealed blood pressures 130/70 mmHg in the left arm and 100/60 mmHg in the right arm, with weak brachial, radial and ulnar pulses on the right side. No carotid bruit was auscultated. His 12-lead electrocardiogram was normal. Two dimensional echocardiography and color Doppler sonography showed that three great vessels were originated from aortic arch and their course could not be delineated because of poor window. Color Doppler sonography of vertebral arteries revealed hypoplasia of left one and normal right vertebral artery with retrograde flow on the right side (subclavian steal) and of subclavian arteries showed that severe stenosis in right subclavian artery just proximal to the origin of vertebral artery and dampened flow with prolonged acceleration time was noted in distal right subclavian, axillary, brachial, radial and ulnar arteries. Computed tomography angiography (CTA) of the head and neck revealed that there was a critical stenosis in aberrant right subclavian artery (ARSCA) proximal to the origin of normal right vertebral artery, left and right common carotid arteries arising from the common carotid trunk (CCT), normal left subclavian artery and hypoplastic left vertebral artery (Figure 1 and Figure 2). The patient subsequently underwent coronary and selective ARSCA angiography after informed consent was obtained. His coronary angiogram revealed normal epicardial coronary arteries and selective ARSCA angiogram showed severe stenosis in its proximal part (Figure 3). Later patient underwent successful right subclavian artery angioplasty with stenting.

DISCUSSION

Subclavian steal syndrome defined as the presence of significant symptoms due to ischemia in the brain (i.e., vertebrobasilar insufficiency) or upper extremity, which are supplied by the subclavian artery. Atherosclerosis is the most common cause of SSS, more common on the left side, may be due to a more acute origin of the left subclavian artery leads to increased

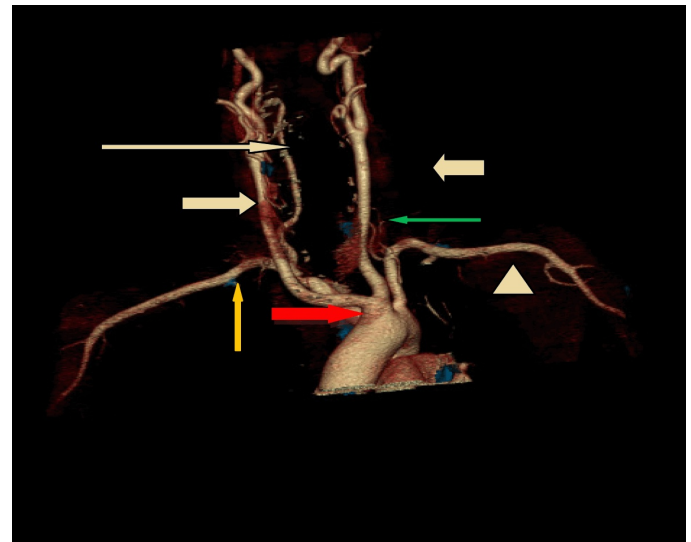


Figure 1: Three dimensionally reconstructed computed tomography angiography in anterior view showing common carotid trunk (red arrow) giving origin to left and right common carotid arteries (white arrows), aberrant right subclavian artery with stenosis proximally (yellow arrow), hypoplastic left vertebral artery (green arrow), normally originated right vertebral artery (long white arrow) and left subclavian artery (white triangle).

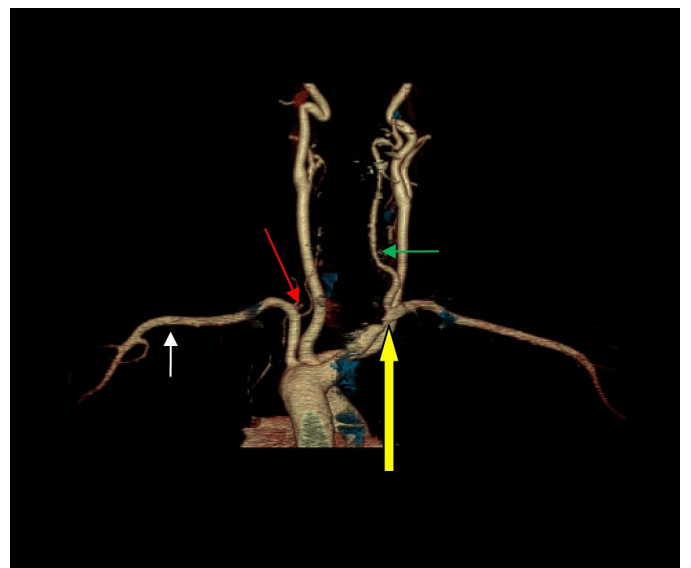


Figure 2: Three dimensionally reformatted computed tomography angiography in posterior view showing aberrant right subclavian artery with severe stenosis proximally (yellow arrow), hypoplastic left vertebral artery (red arrow), normally originated right vertebral artery (green arrow) and left subclavian artery (white arrow).

turbulence resulting in accelerated atherosclerosis [5]. Other conditions causing hemodynamically significant subclavian artery stenosis include Takayasu arteritis, compression of the subclavian artery in the thoracic outlet, following surgical repair of coarctation of the aorta and of tetralogy of Fallot with a Blalock–Taussig shunt and congenital abnormalities such as right aortic arch with isolation of the left subclavian artery and anomalies of the brachiocephalic arteries. Our patient had SSS due to stenosis in ARSCA.

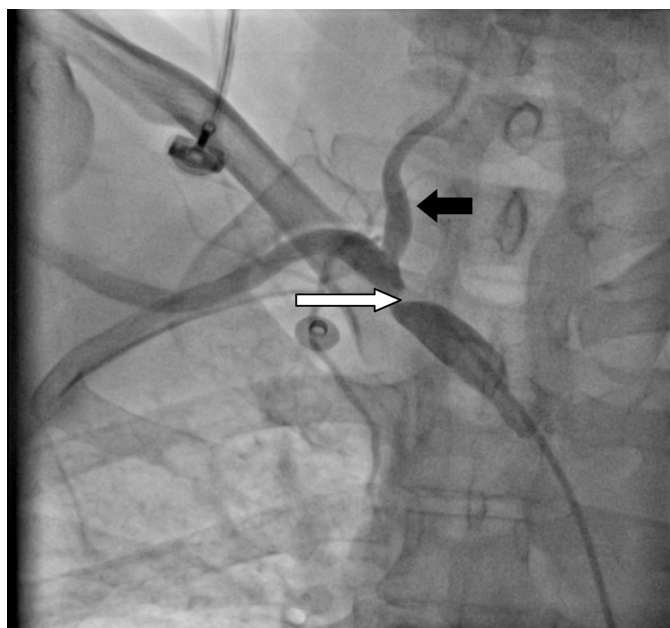


Figure 3: Selective angiogram of the aberrant right subclavian artery in right anterior oblique cranial view showing severe stenosis in its proximal part (white arrow) before the origin of right vertebral artery (black arrow).

The six pairs of aortic arches are a series of vessels that connect on each side the aortic sac with the corresponding dorsal aorta. At a later developmental stage, the aortic arches are both reduced in number and extensively transformed, and finally an asymmetric blood supply system is achieved (Figure 4A). It is understandable that many abnormalities can occur because of many changes involved the transformation of embryonic aortic arches into the adult arterial pattern. Most abnormalities result from the persistence of the parts of aortic arches that normally disappear or from disappearance of parts that normally persistent. Left aortic arch with an aberrant right subclavian artery, last great vessel on the aortic arch, is the most common congenital anomaly of the aortic arch occurring in about 0.5% of the general population [6]. Normally, proximal part of the right subclavian artery arises from the right fourth aortic arch and distal part from the right dorsal aorta present between fourth aortic arch and right seventh intersegmental artery and right seventh intersegmental artery (Figure 4A). The ARSCA develops as a result of the abnormal involution of the right fourth aortic arch, right dorsal aorta present between fourth aortic arch and seventh intersegmental artery and abnormal persistence of right dorsal aorta distal to the right seventh intersegmental artery (Figure 4B) [7]. It has been reported that the ARSCA typically travels in a retroesophageal course in 80% of patients like our patient, between the trachea and esophagus in 10–15% of patients, and anterior to both structures in 5% of patients [8]. Most patients with an ARSCA remain asymptomatic. Only 10% of adult patients with ARSCA have symptoms [9].

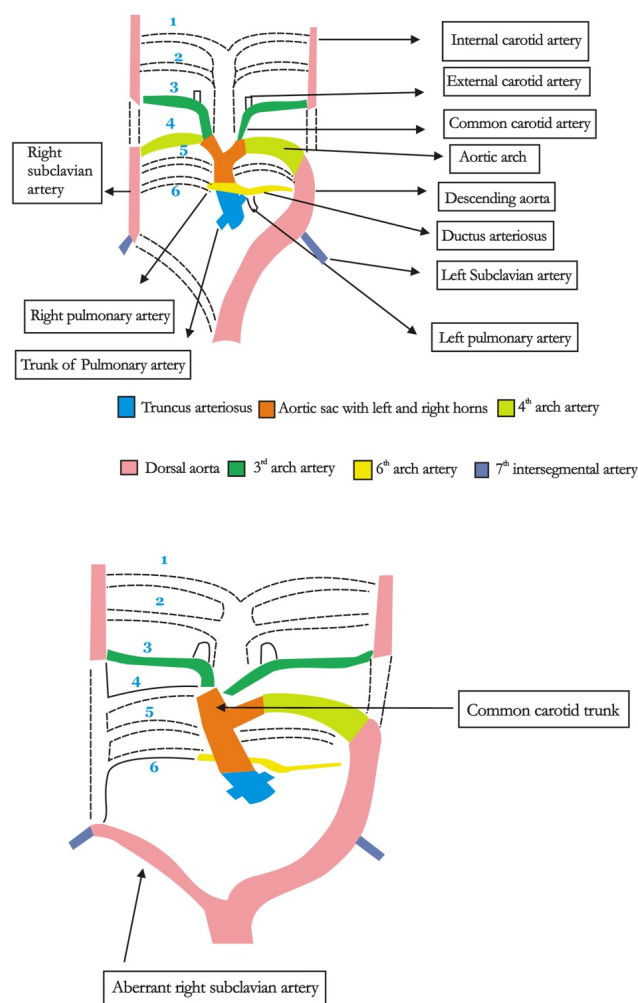


Figure 4: (A) Schematic drawing illustrating the arterial changes that result during transformation of the Truncus arteriosus, aortic sac, pharyngeal arch arteries, and dorsal aortas into the adult pattern. The vessels that are not colored will disappear during development, (B) Schematic drawing illustrating the development of aberrant right subclavian artery and common carotid trunk.

In 1993, Wells et al. reported that CCT is a left aortic arch variant and is present in about 13% of the general population [10]. During development, aortic sac, the most distal part of the truncus arteriosus, bifurcates into right and left horns, which forms the right brachiocephalic trunk and part of arch of aorta, respectively. Normally, third left aortic arch which forms the left common carotid connected to the left horn of the aortic sac. The CCT arises when the proximal part of the third left aortic arch absorbed in to the right horn of the aortic sac. However, incomplete vascular ring caused by an ARSCA associated with CCT like our case is extremely rare and its incidence is about 0.16% [11].

Like our case one vertebral artery is hypoplastic in up to 10–15% of the healthy population and makes little

contribution to basilar artery flow [12, 13]. The left vertebral artery is dominant in approximately 50%; the right in 25% and in the remaining 25% cases the two vertebral arteries are of similar calibre. Three prior studies concluded that individuals with hypoplastic vertebral arteries are at increased risk for posterior circulatory stroke more so if they had risk factors like hypertension, diabetes, smoking, etc. [14–16]. Our patient had hypoplastic left vertebral artery associated with stenosis in aberrant right subclavian artery proximal to the origin of right vertebral artery leading to relatively compromised blood flow to the brainstem and causing the vertebrobasilar symptoms with minimal exertion of the right upper extremity.

CONCLUSION

The combination of common carotid trunk, aberrant right subclavian artery and hypoplastic left vertebral artery in the same person was never reported earlier and this combination causes ischemia in large area of brain if the patient develops hemodynamically significant stenosis or occlusion in both the common carotid trunk and aberrant right subclavian artery proximal to the origin of right vertebral artery. All physicians should be aware of aortic arch variations prior to catheterization so that they can reduce fluoroscopic time and minimize contrast medium use.

Author Contributions

Boochi Babu Mannuva – 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
Rajasekhar Durgaprasad – 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
Vanajakshamma Velam – 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
Shashanka Chunduri – 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
Vijayalajshmi Devi B – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

OPEN ACCESS

Metastatic squamous cell carcinoma of jejunum of unknown primary masquadering as superior mesentric artery syndrome: A case report

Harnam Singh, Pankaj Shivhare, Pankaj Dugg, Sushil Mittal, Ashwani Kumar, Sona Goyal

ABSTRACT

Introduction: Metastatic squamous cell carcinoma of proximal jejunum is rare presentation, in which the diagnosis is made only after biopsy. A rare case of metastatic squamous cell carcinoma of jejunum with unknown primary is reported here. **Case Report:** A 75-year-old female was presented with recurrent episodes of nausea and vomiting. On the basis of radiological investigation, diagnosis of superior mesenteric artery syndrome was made. On exploration growth was present in proximal jejunum. Resection of the growth with adequate margins and primary anastomosis was done. Histopathology showed metastatic squamous cell carcinoma. Whole body positron emission tomography scan shows no evidence of primary. **Conclusion:** Jejunum is rare site for squamous cell carcinoma of metastatic origin of unknown primary and can be confirmed only on biopsy.

Keywords: Squamous cell carcinoma, Intestinal obstruction, Superior mesenteric artery (SMA) syndrome

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INTRODUCTION

Metastatic carcinoma of unknown primary is a common problem, accounting for up to 10–15% of all solid tumors at presentation [1]. Jejunum cancer is rare, and is difficult to diagnose before surgery [2]. Compared to other gastrointestinal malignancies such as gastric cancer and colorectal cancer it is relatively rare [3]. We report a patient in whom preoperative barium meal follow through (BMFT) showed dilatation of the 2nd and 3rd part of the duodenum and contrast-enhanced computed tomography (CECT) whole abdomen reported as superior mesenteric artery (SMA) syndrome. On exploration proximal jejunal mass was found and in biopsy carcinoma, squamous cell, NOS, metastatic was found. Postoperative positron emission tomography (PET) scan showed no evidence of primary.

CASE REPORT

A 75-year-old female, house wife was presented to the outpatient department with complains of nausea and vomiting for last one week. She had been having similar complains in the past on and off for about 4–6 months, which was relieved on its own. There was no history of abdominal distension or weight loss. Bowel/bladder habits were normal. Patient was admitted and work up was done. General physical and

systemic examination which included breast and axilla was within normal limits. Biochemical profile was within normal limits. Barium meal follow through 4:05 min film showed dilated duodenal loop with small amount of contrast in the jejunum (Figure 1). The CECT scan of whole abdomen showed compression of third part of the duodenum in between the aorta and superior mesenteric artery (SMA), with dilatation of 1st and 2nd part of duodenum—SMA syndrome (Figure 2). Considering the diagnosis as SMA syndrome based on CECT report, duodenojejunostomy was planned. On exploration omentum was adherent to proximal jejunum. Growth was felt in proximal jejunum about size 4x5 cm approximately 7–8 cm distal from the duodenojejunal junction. Jejunum was adherent to the root of mesentery. No lymph node was palpable. Resection and primary anastomosis was done. Rest of the viscera grossly appeared normal. Resected specimen showed normal mucosal folds and mass (Figure 3). Histopathological report showed squamous cell carcinoma, NOS type, metastatic. The section showed normal intestinal mucosa which signified that primary tumor was not arising from the intestine and it was metastatic in nature. Figure 4 shows malignant squamous epithelial cells with increased N:C ratio, hyperchromatism, pleomorphism and increased keratin formation. Figure 5 is a high power section showing squamous malignant epithelial cells with increased N:C

ratio, hyperchromatism and pleomorphism. Acute inflammatory infiltrate such as neutrophils, eosinophils, lymphocytes and plasma cells were also seen. In the follow-up period Pap-smear was done which showed reactive hyperplasia followed by cervical biopsy that showed nabothian cyst. IDL was done which was within normal limit. Upper and lower gastrointestinal endoscopy was done and no lesions were detected. The PET scan of whole body was done which reported no evidence of active metabolic disease anywhere in the body with mild (FDG) uptake seen in bilateral tonsils and posterior part of tongue.

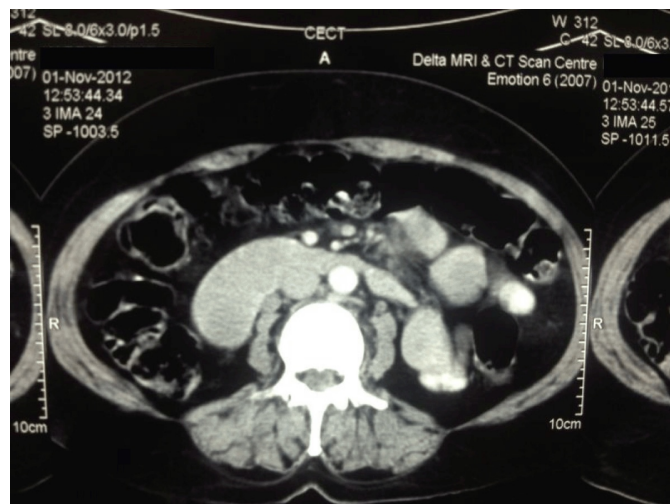


Figure 2: Contrast-enhanced computed tomography whole abdomen compression of third part of the duodenum in between the aorta and superior mesenteric artery, with dilatation of 1st and 2nd part of duodenum.



Figure 1: Barium meal follow through showing dilated duodenal loop with small amount of contrast in the jejunum.



Figure 3: Arrow showing growth in jejunum.

DISCUSSION

Metastatic carcinoma of unknown primary account for up to 10–15% of all solid tumors at presentation [1]. For small intestine, melanoma, lung, breast, colon and

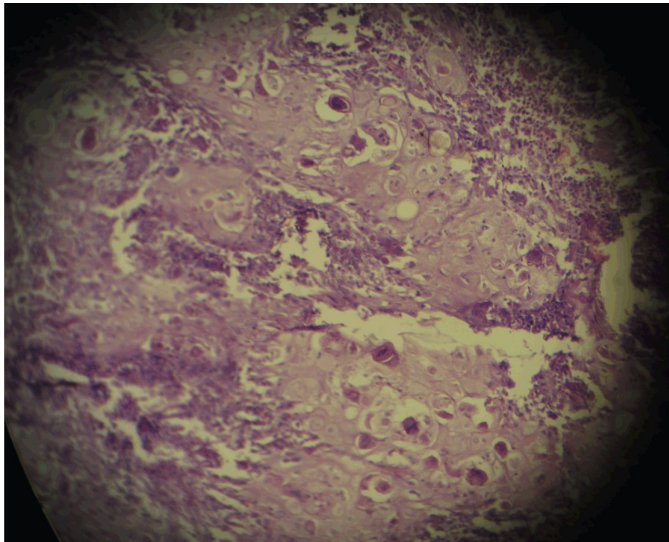


Figure 4: Section showing squamous malignant epithelial cells with increased N:C ratio and hyperchromatism.

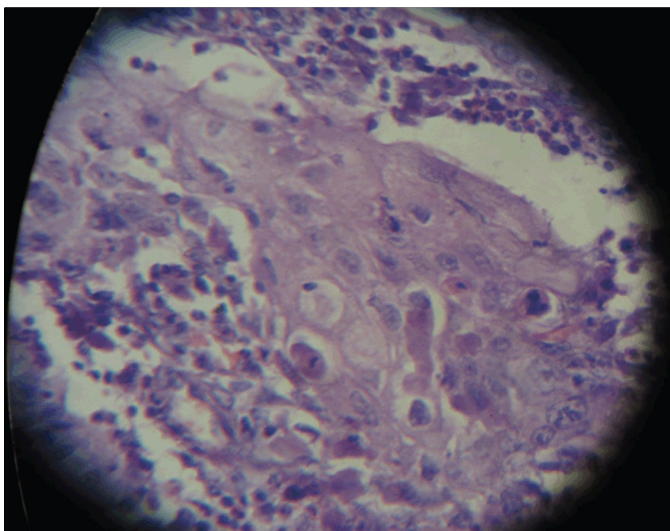


Figure 5: High power resolution show squamous malignant epithelial cells with increased N:C ratio and hyperchromatism.

kidney are the most frequent primary sites. Metastatic spread from primary lung cancer to the small intestine is more frequent than to stomach and colon. Virtually, all primary cancers can occasionally lead to metastases in the small intestine and, because of the low frequency of primary small bowel cancer, a high proportion of small intestinal malignancies are metastatic. The pathogenesis of intestinal metastasis usually involves hematogenous spread of tumor cells [4].

Malignant small intestinal tumors frequently develop in persons aged 40–79 years. The male-to-female ratio is 1.4:1 [5]. Concerning common sites, jejunum cancer develops in the jejunal region 50–60 cm distant from Treitz' ligament in more than 80% of patients [5–7]. Clinical symptoms mainly consist of ileus, hemorrhage of the digestive tract, and phymas on palpation. In

addition, anemia is observed. Occlusive symptoms are frequent in the presence of cancer [7].

For diagnosis, contrast-enhanced radiography of the small intestine, small intestinal endoscopy, abdominal angiography, abdominal echography, and abdominal CT scan are employed. As characteristic findings on contrast-enhanced radiography of the small intestine, Good indicated that circumferential, small shadow defects with a clear border, the disappearance of normal mucosa and ulcer formation, irregular narrowing of the lumen around the lesion, and intestinal dilatation on the orifice side of the cancer lesion were important [6]. It is relatively rare to make a definitive diagnosis before surgery, suggesting difficulty in diagnosis. According to Moriyama et al., a definitive diagnosis was made via endoscopic biopsy in 28.3% of jejunal and 5.7% of ileal cancer patients [8].

According to some studies, the 5-year survival rates of primary small intestinal cancer patients range from 9.1–38.5% [8, 9]. James et al. investigated 144 patients with small intestinal tumors and reported that the 5-year survival rate for small intestinal cancer was 59% [10]. In particular, the 5-year survival rate in patients undergoing total resection was 81%. However, the prognosis of small intestinal cancer is poor, possibly because cancer is advanced at the time of detection in many patients due to a delayed diagnosis.

CONCLUSION

Metastatic squamous cell carcinoma of the jejunum is sufficiently rare to justify a case report. Though the this case was typical with regard to age and symptom of vomiting because of cancer-related occlusion, but atypical with regard to location of the tumor which was present 7–8 cm from duodenojejunal junction whereas the common location is 50–60 cm distant from Treitz' ligament and it was masquading as superior mesenteric artery syndrome with regard to symptoms and contrast-enhanced computed tomography report.

However, metastatic squamous cell carcinoma in jejunum with unknown primary is rare presentation confirmed only in biopsy.

Author Contributions

Harnam Singh – Substantial contribution to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Pankaj Shivhare – Substantial contribution to conception and design, Acquisition of data, analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Pankaj Dugg – Acquisition of data, analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Sushil Mittal – Substantial contribution to conception and design, Revising it critically for important

intellectual content, Final approval of the version to be published

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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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Recurrent postauricular dermoid cyst: A case report

Rachana Tiwari, Vaishali Sangole

ABSTRACT

Introduction: Dermoid cysts are congenital anomalies that arise from trapped pouches of the ectoderm near the normal fold or from the surface that has failed to separate from the neural tube. Dermoid cysts are very rare in head and neck area and its presence in postauricular region is further exceptionally rare. Only a few cases have been reported. In this study, a rare case of recurrent dermoid cyst located behind the ear with abnormal gradual growth over several years is reported which to the best of our knowledge has not been reported earlier. **Case Report:** A 18-year-old female presented with a swelling in the right postauricular area since six years. This swelling underwent excision six years back. Examination showed a solitary, cystic 4x3x2 cm, smooth surfaced swelling in the right postauricular area with a 2 cm ill-defined scar at the center. Ultrasonography was suggestive of a benign lesion. Fine needle aspiration cytology showed infected epidermal cyst with chronic inflammation. Through the postauricular incision an encapsulated cystic structure with embryonal cells was excised. **Conclusion:** Patient sought medical advice in our case for the cosmetic reason because of the

embarrassing look of the prominent unilateral ear. Her concern to avoid getting a recurrence besides cosmetic correction was successfully taken care of in our case.

Keywords: Dermoid cyst, Postauricular cyst, Congenital dermoid, Recurrent dermoid cyst

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INTRODUCTION

Dermoid cysts are a type of teratoma occurring as a result of the sequestration of the skin along the lines of embryonic closure. Dermoid cysts are very rare in head and neck area, an estimated 7% of all dermoids [1]. In the classic work by New and Erich, 49.5% of head and neck dermoids were located in the orbit and no lesions were specifically identified in the ear [2]. Its presence in the postauricular area is exceptionally rare. Dermoid cysts can recur, if not completely excised. We report a case of recurrent postauricular dermoid cyst in a 18-year-old female.

CASE REPORT

A 18-year-old female presented to the ENT services of our outpatient department at MGM Medical College, Kamothe, Navi Mumbai with complaint of swelling in the right postauricular area since six years (Figure 1). It was gradual in onset, progressed from a peanut size to

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the present size and was painless. There was no history of trauma, discharge from the swelling, discharging ear, decreased hearing or associated congenital anomaly. There were no constitutional symptoms. There was no history of such lesion in her family members. However, there was a similar swelling at the same site noticed since her childhood by her parents. This swelling underwent excision six years back before the recent presentation. The patient did not retain any documents related with the surgical intervention but mentioned about existence of hair tissue in the excised specimen.

On local examination a solitary, cystic, dumb-bell swelling, 4x3x2 cm in size, spherical, non-pulsatile, extending from the highest attachment of the pinna to 2 cm above the mastoid tip with smooth surface, obliterating the retroauricular sulcus with a 2-cm horizontal ill-defined scar mark in the center of the swelling (Figure 2). There was no discharging sinus or pointing abscess. On palpation, the swelling was non tender, cystic in consistency, fluctuant, non-transilluminant, non-reducible with well defined margins. The overlying and surrounding skin was normal with no fistula or signs of infection. Bruit or any pulsation was not present over the swelling. The external auditory canal and tympanic membrane were normal. There was no evidence of facial paresis, facial asymmetry or ocular findings. A clinical differential diagnosis of a sebaceous cyst, lipoma, lymph node, inclusion dermoid, simple cyst was kept.

Ultrasonography revealed a solid, hyperechoic, well defined lesion of size 2.9x2.4x1.5 cm with no vascularity or underlying bone erosion. The features were more in favor of a benign lesion. Fine needle aspiration cytology of the swelling done after seven days coverage of antibiotics was suggestive of infected epidermal cyst. The infection was dominance of the chronic inflammation.



Figure 2: Photograph showing 2 cm horizontal ill-defined scar in the center of the swelling.

Considering the case to be recurrent due to incomplete initial removal at the first surgical attempt and the dumb-bell shaped appearance of the mass a contrast computed tomography scan of the right temporal bone was advised. But in our country due to financial constraints of the patient the benefit of this investigation choice for knowing the extent and bony breach could not be gained.

The patient underwent surgical excision of the lesion via a postauricular incision. Intraoperative findings were that of 3 cm well encapsulated cystic structure containing yellowish putty oily debris with bunch of hairs inside (Figures 3 and 4). There was no connection with the external auditory canal, middle ear, or any



Figure 1: Clinical photograph of a 18-year-old female showing a solitary, cystic swelling, 4x3x2 cm in size, spherical, non-pulsatile, extending from the highest attachment of the pinna to 2 cm above the mastoid tip with smooth surface, obliterating the retroauricular sulcus with a 2 cm vertical scar mark in the center of the swelling was found.



Figure 3: A 3-cm well encapsulated cystic structure containing yellowish putty oily debris with bunch of hairs inside. There was no connection with the external auditory canal, middle ear, or any intracranial extension.

intracranial extension. The cyst was excised whole, and the defect closed primarily. The patient's postoperative recovery was uneventful.

Histopathology of the cyst indicated the diagnosis of dermoid cyst, which was confirmed on microscopic examination by the presence of keratinized stratified squamous epithelium, sebaceous glands and hair follicles (Figure 5).

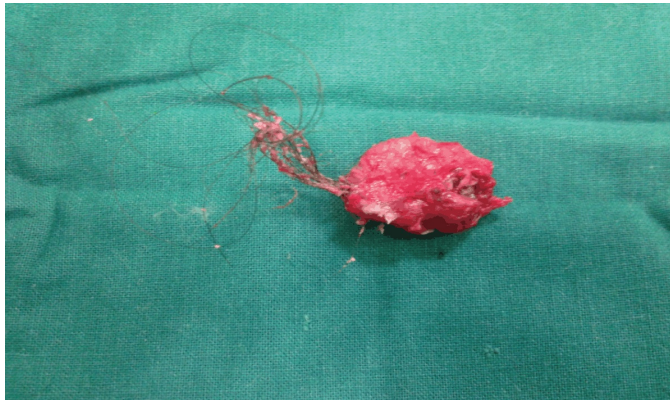


Figure 4: A 3-cm well encapsulated cystic structure containing yellowish putty oily debris with bunch of hairs inside.

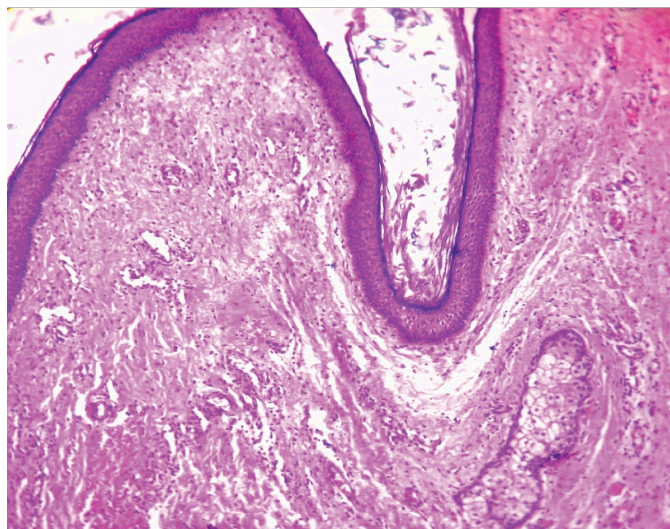


Figure 5: Histopathology showing the presence of keratinized stratified squamous epithelium, sebaceous glands and hair follicles confirming the diagnosis of dermoid cyst (H&E stain, x100).

DISCUSSION

The term dermoid cyst neither appear to be restricted to a single kind of lesion nor it is used in only a single medical discipline. The term dermoid cyst can be found in the vocabulary of dermatologists, otolaryngologists, general pathologists, gynecologists, neurosurgeons, or pediatricians. If asked, all of these clinicians would most probably define and describe dermoid cysts differently. For most dermoid cyst means subcutaneous cysts, which are usually congenital.

In reference to nomenclature, Batsakis has defined a dermoid cyst as “an epithelial lined cavity with variable numbers of skin appendages (hair, follicles, sebaceous glands, etc.)” [3]. Earlier pathology sources had more generally categorized these as being of ectodermal and mesodermal origin.

The parthenogenic theory for its pathogenesis suggests that dermoid cysts take their origin from primordial germ cells. They are postulated to originate from the congenital inclusion of germ layers in the deeper tissues along the lines on embryonic fusion. Meagher et al. suggest the cause for the bilateral prominent ears due to dermoid cyst is multifactorial with certain strong familial predilections [8].

Dermoid cysts can be divided into three types according to their histological characteristics; namely epidermoid, dermoid and teratoid. These congenital hamartomas occur with an incidence of 1 in 4,000 births, 6 times greater in females.

The cyst can exist anywhere but commonly is seen in the midline of the body. The majority of dermoid cysts arise in the ovaries. In a review of 1495 dermoid cysts collected over 25 years at the Mayo clinic, orbital lesions were found in 49.5%, nasal lesions in 12.6%, submental or sublingual in 23.3% and variously placed in the occipital, frontal, cervical, soft palatal and lip regions in 14.6% [2]. Those occurring in the cervicofacial region are uncommon accounting for about 7% of all dermoids [1]. One of the uncommon areas dermoid cysts are seen is the temporal area (postauricular skin, the middle ear or, even rare on the eustachian tube). They have also been reported in the auricle, middle ear and in the auriculotemporal area [4–6].

Dermoid cysts around the auricular region are rare, whereas those located in the postauricular area are extremely rare. To the best of our knowledge, there have been very few cases of postauricular dermoid cyst described in literature [1, 6–9].

In Korea, only three cases of postauricular dermoid cysts have been reported in the past. Moon et al. [10] further described a single case in 2005 and Sung et al. [11] described three cases of dermoid cysts of the auriculotemporal area in Korea in 2009. Pankaj et al. [12] described a case of unilateral postauricular dermoid cyst in an 18-year-old boy in 2007 in India. Ho et al. [13] and Mohammad et al. [14], reported a postauricular lump recently in 2011.

Our case is the rarest, first documented case of a recurrent postauricular dermoid cyst after a thorough Medline search.

CONCLUSION

Patients with postauricular dermoid cysts usually seek medical advice for the cosmetic reasons because of the embarrassing look of the prominent unilateral or bilateral ears. The patient's concern in our case was to avoid getting a recurrence besides cosmetic correction of the postauricular swelling. Sometimes complete excision is not practical if in a dumb-bell configuration

where the cyst extends through a suture line in the skull. We could achieve both the cosmetic and recurrence aspects of concerns of the patient satisfactorily.

Author Contributions

Rachana Tiwari – 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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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Decompressive laparotomy for reduction of incessant increased intracranial pressure in the absence of abdominal compartment syndrome: A case report

Mark Armanious, Louise N Bacon, Johnny Harris, Shayla George, Kristen-Kaye Goulbourne, Omar K Danner, L Ray Matthews, Kenneth L Wilson

ABSTRACT

Introduction: A novel approach for the management of severe traumatic brain injury is to perform a decompressive laparotomy when controlling intracranial hypertension (ICH) appears futile. Bladder pressures measured above 20 mmHg are used to signify the presence of an abdominal compartment syndrome and indicate the need for a decompressive laparotomy. **Case Report:** We are presenting a case study of a 16-year-old male who sustained a gunshot wound to the head that resulted in a severe traumatic brain injury and incessantly elevated intracranial pressures (IVP). The patient did not demonstrate a sustained and significant reduction in his ICH following a decompressive craniectomy. We performed a decompressive laparotomy successfully reducing the patient's ICP below 20 mmHg. The patient survived to discharge and was neurologically improved at follow-up. **Conclusion:** This case highlights the importance of regarding the body a continuous compartment allowing a reduction in intracranial pressure by decompressing the abdomen even in the absence of an abdominal

compartment syndrome measured by elevated bladder pressures above 20 mmHg.

Keywords: Intracranial hypertension (ICH), Cerebral perfusion pressure (CPP), Abdominal compartment syndrome (ACS), Decompressive laparotomy, Brain injury

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INTRODUCTION

One of the most formidable complications of traumatic brain injury (TBI) is increased intracranial pressure (ICP). An increased ICP is a neurologic emergency that if left untreated may cause cerebral ischemia, brain herniation, and possibly death [1]. The primary treatment for patients with TBI is to lower ICP readings below 20 mmHg and to maintain cerebral perfusion pressure (CPP) between 60 mmHg and 90 mmHg in order to provide sufficient cerebral perfusion [1]. Proper head elevation is an effective strategy promoting optimal venous flow for TBI, but more aggressive therapies are needed when elevated ICP accompanies severe TBI. Some of the therapies to lower ICP after TBI include hyperventilation (HV), intravenous mannitol and cerebrospinal fluid drainage from a ventriculostomy (EDV). Decompressive

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craniectomy and barbiturate-induced coma are at the desperate end of the continuum necessary to save a patient suffering from both TBI and increased ICP [2–3].

When all typical and heroic therapeutic treatments have been exhausted for sustained high ICP, an innovative approach termed a decompressive laparotomy can be implemented. This treatment which had been exclusively reserved for abdominal compartment syndrome involves cutting the anterior fascia of the abdomen allowing the abdominal viscera to expand thereby relieving intra-abdominal pressure. The elevation of intra-abdominal pressure displaces the diaphragm cephalad, increasing intrathoracic pressure and central venous pressure. The pressure is then transmitted through the venous system, causing increases in ICP and decreases in CPP [4–6]. We are presenting a case study of a 16-year-old male with a severe TBI and incessant elevated ICP who demonstrated a reduction in his ICP following a decompressive craniectomy without the presence of an abdominal compartment syndrome (ACS).

CASE REPORT

A 16-year-old male presented to our level I trauma center in Atlanta, Georgia after sustaining a single gunshot wound (GSW) to the left occipital region of head. He was confused and combative, requiring intubation in the trauma bay for a glasgow coma score of eight. Significant findings included comminuted fractures of the left thumb and an entry wound to the left temporal bone of the skull. The computed tomography (CT) scan of the head revealed retained bullet fragments and multiple facial fractures, it also revealed a comminuted fractures of the left posterior parietal, occipital and temporal bones. Intracranial damage included a left-sided subdural hematoma; intraparenchymal hemorrhage and diffuse cerebral edema with sulcal effacement of 6 mm with a left to right midline shift (Figure 1).

The patient was taken to the operating room for debridement of the brain parenchyma and a right decompressive craniotomy by the neurosurgery service. Postoperatively, the ICP remained elevated (41–54) and would not respond to propofol sedation, 3% saline infusion and cerebrospinal fluid drainage. The following morning a repeated CT scan of the head was performed which revealed blossoming of the left intraparenchymal hematoma. The patient was transported to the operating room for a left hemicraniectomy and duraplasty. Despite this intervention, the ICP remained in the thirties and a metabolic coma with phenobarbital was induced.

On postoperative day-5, a decompressive laparotomy was performed as an intervention for his refractory intracranial hypertension. The abdominal fascia was left open allowing room for bowel evisceration. ICPs measured immediately following the decompressive laparotomy were lowered ranging

between 11–12 mmHg. On postoperative day-14, the patient was taken to the operating room for an abdominal washout and fascial closure because of ICP measurements that were less than 20 for 9 days since the decompression. By postoperative day-18, the induced pentobarbital coma had been reversed and the external ventricular drain (EVD) had been removed. The patient survived and was discharged to a rehabilitation facility with a glasgow coma score of seven. One year following his gun shot wound to the head, the patient is at home with his parents. He is communicative and writing music. He has gained the ability to ambulate with the assistance of a cane and hopes to walk independently before reaching college.



Figure 1: On admission, computed tomography of the brain demonstrating comminuted fracture of the left occipital bone. A left frontoparietal subdural hematoma is noted along with intraventricular hemorrhage.

DISCUSSION

Approximately, 1.7 million people sustain a TBI annually [7]. In the context of a TBI, the brain has a limited capacity to autoregulate the CPP because of increased intracranial pressure as established by the Monro–Kellie doctrine. The Monro–Kellie doctrine describes the brain as a fixed bone that has decreased compliance when compared to other body compartments. An increase in intracranial volume causes a significant increase in the ICP negatively impacting the CPP of the brain [8]. Systemic vasoactive responses from shock remote to the abdomen can cause “capillary leak” that leading to fluid accumulation inside the abdomen or thorax deleteriously increasing ICP. Intracranial pressure studies have demonstrated that there is a direct correlation between intracranial, intrathoracic and intra-abdominal compartments [9–10]. Multiple-compartment syndrome or

polycompartment syndrome, stresses the importance of increased pressure in closed anatomic spaces threatening the viability of surrounding tissue [11]. Joseph et al. demonstrated a decompressive laparotomy was successful in decreasing ICP, thus supporting the correlation of pressures in the polycompartment syndrome theory. In this study, decompressive laparotomies were utilized for 17 patients and all 17 patients experienced a decrease in the ICP of 10 mmHg or greater [6]. A case report by Dorfman et al. documented the treatment of a 17-year-old female following a motor vehicle collision with TBI that was effectively treated with a decompressive laparotomy in a last ditch effort to control intractable ICPs as a consequence of a massive resuscitation leading to an ACS [12].

When bladder pressures are performed and measured above the normal range (greater than 20 mmHg), a decompressive laparotomy in selective patients can be performed to release an abdominal compartment syndrome. As mentioned by Scalea et al. the performance of decompressive laparotomies for refractory ICP can be associated with an unacceptably high rate of morbidity and mortality, and should be utilized with well-defined criteria [10]. In our study, and with subsequent success with an adult patient, we have expanded our decompressive laparotomies to not only include when medical management, which includes an induced pentobarbital coma, fails to reduce elevations of ICP above 20 mmHg, but also to include when the bladder pressures are normal. Our earlier intervention without bladder pressure elevation may suggest intervening before a measured increase in ACS that would negatively impact perfusion of the injured brain.

Our case outlines the utility of a decompressive laparotomy for an isolated head injury when the abdomen is affected by a massive trauma resuscitation and a polycompartment syndrome can be presumed while the bladder pressures may not yet reflect the volume expansion leading to an abdominal compartment syndrome. The immediate and significant drop of the ICP, which was sustained below 20 mmHg following the decompressive laparotomy highlights regarding the body as being multi-compartmented when treating intractable ICPs. Studies have demonstrated that the pediatric population performs more favorably to decompressive craniectomies than their adult counterparts [13–14].

CONCLUSION

This case study of a 16-year-old male patient and that of a 17-year-old patient by Dorfman et al. perhaps further demonstrates that the pediatric population with TBI with refractory intracranial hypotension can be treated more aggressively with less mortality and a lesser vegetative state when maximal medical therapy fails. The number of published cases and sample sizes are limited in the treatment of incessant intracranial pressures with abdominal compartment syndrome. Our

success with this patient and another young adult demonstrates that a decompressive laparotomy can be safely applied as an alternative to improve cerebral perfusion in the absence of abdominal compartment syndrome when medical therapy fails.

Author Contributions

Mark Armanious – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final approval of the version to be published

Louise Bacon – Substantial contributions to conception and design, Acquisition of data, or analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final approval of the version to be published

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L Ray Matthews – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final approval of the version to be published

Kenneth L Wilson – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article or revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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Concomitant obturator hernia and midgut volvulus in an elderly woman

Yeung Kwok-Wan, Chang Ming-Sung

ABSTRACT

Introduction: Obturator hernia is a rare hernia of the pelvic floor and accounts for less than 1% of all intra-abdominal hernias. Midgut volvulus may be primary without an associated underlying cause, or secondary to a congenital or acquired condition. **Case Report:** A 94-year-old female patient suffered from severe and diffuse abdominal cramping pain and no stool passage for 2 days and vomiting for a day. Blood analysis revealed leukocytosis. A history of constipation and chronic obstructive pulmonary disease was noted and no intra-abdominal operation was performed in the past. Contrast-enhanced computed tomography scan showed distention of the small bowel loop, a whirl sign of the superior mesenteric artery and vein, and a short segment of distal ileum incarcerated between the right external obturator and pectineus muscles. Computed tomography scan of concomitant right obturator hernia and midgut volvulus was made, which was confirmed by surgical exploration. **Conclusion:** Concomitant obturator hernia and midgut volvulus in an elderly woman is rare. Careful and thorough survey of the whole abdomen by using computed tomography prompts

emergent operation to reduce the mortality and morbidity of the patient.

Keywords: Obturator hernia, Midgut volvulus

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INTRODUCTION

Obturator hernia is a rare hernia of the pelvic floor and accounts for 0.05% to less than 1.4% of all intra-abdominal hernias. It is also a rare cause of small bowel obstruction, accounting for 0.2–1.6% of bowel obstruction [1–6]. Obturator hernia may be associated with midgut volvulus [6]. A volvulus represents the most common cause of strangulation with associated bowel ischemia. Midgut volvulus may be primary without an associated underlying cause, or secondary to a congenital or acquired condition [7–10]. We present a case of concomitant obturator hernia and midgut volvulus in a 94-year-old woman with a series of imaging studies.

CASE REPORT

A 94-year-old woman suffered from severe and diffuse abdominal cramping pain and no stool passage for two days, and vomiting for a day. Blood analysis revealed leukocytosis (WBC 16,410/ μ L with elevated band form (7%)) and elevated C-reactive protein level (2 mg/dL). A history of constipation and chronic

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obstructive pulmonary disease (COPD) was noted. No intra-abdominal operation was performed in the past years. Physical examination showed diffuse abdominal tenderness and distention, and increased bowel sound.

Kidney-ureter-bladder (KUB) radiograph revealed diffuse distention of small bowel loop (Figure 1). Multi-detector row computed tomography (MDCT) with intravenous contrast administration showed dilatation of the small bowel loop with intraluminal air-fluid level, and whirl sign of the superior mesenteric artery and vein (Figure 2). A short segment of distal ileum was incarcerated between the right external obturator and pectineus muscles. Therefore, computed tomography (CT) scan of the concomitant midgut volvulus and right incarcerated obturator hernia was made. Emergent exploratory laparotomy by using midline incision showed a 360° counterclockwise volvulus of the small bowel loop, and uncovered a 5-cm distal ileal loop incarcerated in the right obturator hernia (Figure 3). The small bowel loop showed ischemic change but became revascularized again after reduction of the volvulus. The defect of the hernia was simply repaired with the interrupted Dexon suture. No resection of bowel loop was done. However, postoperative pneumonia occurred and extubation was failed. Despite vigorous antibiotics and supportive treatment, the patient died of respiratory failure on the 108th day postoperation.



Figure 1: Kidney-ureter-bladder revealed diffuse distention of small bowel loop.

DISCUSSION

Obturator hernia occurs through the obturator canal, which is 2–3 cm long and 1 cm wide and contains the obturator nerve and vessels surrounded by fatty tissue [1, 4]. The most common form of obturator hernia is through the pathway between the external obturator and pectineus muscles [3]. It is a rare hernia of the pelvic floor and accounts for 0.05% to less than 1.4% of all intra-abdominal hernias. It is also a rare cause of small bowel obstruction, accounting for 0.2–1.6% of bowel

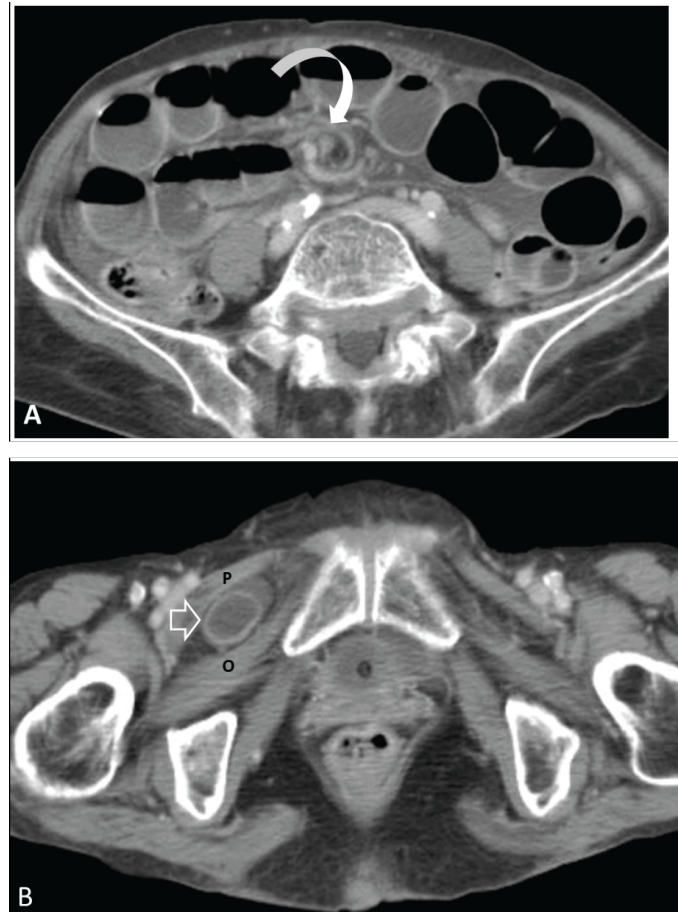


Figure 2: (A) Contrast-enhanced multidetector computed tomography with axial image showed dilatation of the small bowel loop with intraluminal air-fluid level, and whirl sign of the superior mesenteric artery and vein (curved arrow), (B) A short segment of distal ileum (open arrow) was incarcerated between the right pectineus muscle (P) anteriorly and external obturator muscle (O) posteriorly.

obstruction [1–6]. Asians have been shown to have the highest rates while a much lower incidence occurs in the Western countries [1]. The right side is more frequently affected than the left due to protection from the sigmoid colon on the left [2–4]. Bilateral obturator hernias occur in 6% of cases and may be in combination with other types of hernias such as inguinal hernia or femoral hernia [3–4]. Women are affected nine times more frequently than men because the former group has a wider pelvis, a more triangular obturator canal opening and a greater transverse diameter [4, 5]. With the nickname ‘little old lady’s hernia’, obturator hernia most frequently affects the elderly and emaciated women, especially between 70 and 90 years of age, due to loss of preperitoneal fat over the obturator canal, which facilitates the formation of the hernia. Other risk factors may increase the intra-abdominal pressure, relax the peritoneum and predispose the patients to herniation, such as chronic constipation, ascites, chronic obstructive pulmonary disease (COPD), multiparity and kyphoscoliosis. The clinical

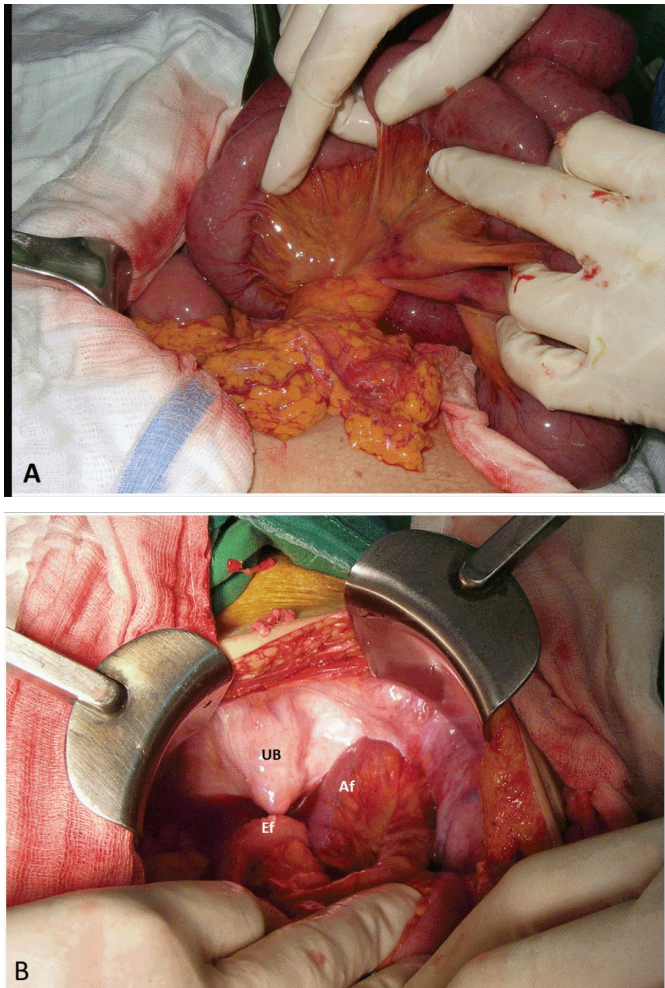


Figure 3: Operative findings. (A) Twisting of the small bowel loop around the mesentery was found, (B) View into the right lower lateral pelvic cavity demonstrated distended and ischemic afferent ileal loop (Af) and collapsed efferent loop converging to the right obturator hernia. UB, foley catheter-indwelled urinary bladder.

presentation is not specific with symptoms of bowel obstruction such as dull, cramping abdominal pain, nausea and vomiting. It may be acute or intermittent if the hernial viscera reduce into the abdominal cavity spontaneously [1, 5]. Partial obstruction is usually encountered and due to a high frequency (41–100%) of Richter's hernia of small bowel into the obturator canal. A palpable mass at the obturator region is uncommon because the herniated mass is concealed beneath the pectineus muscle [5]. Obturator hernia may cause Howship–Romberg sign, which refers to pain along the course of the obturator nerve as a result of compression of the nerve by the hernial sac [1–4]. The pain is prominent in the anteromedial aspect of the thigh and less often in the hip, and is relieved by flexion of the thigh and aggravated by extension, adduction and medial rotation. This sign is only present in 15–50% of cases and often overlooked due to the osteoarthritis of the hip, or may be masked by the more severe symptom of abdominal pain. Hannington–Kiff sign is more

specific but less known than Howship–Romberg sign and refers to an absent adductor reflex in the thigh.

Several imaging modalities have been described to be useful in the diagnosis of obturator hernia, including ultrasonography, herniography, barium enema, and CT scan [1–5]. Among them, CT scan of the abdomen and pelvis has proven its value in definite diagnosis of obturator hernia, which demonstrates entrapment of bowel loop between the external obturator and pectineus muscles, and significantly increases preoperative diagnostic accuracy. Early and definite diagnosis can be made in 100% of cases by using CT scan [1]. Early preoperative CT scan helps to decrease intestinal resection and surgical mortality because the shorter period from the occurrence of symptom to surgical correction can increase viability of the bowel and lower the morbidity and mortality rates. Thus, the rapid evaluation of the patients should be accomplished within hours but not in days [5].

Obturator hernia may be associated with midgut volvulus [6]. Midgut volvulus may be primary without an associated underlying cause, or secondary to a congenital or acquired condition [7–9]. The primary small bowel volvulus is much more common in africa and Asia, and usually occurs in children and young males, in whom no predisposing anatomic abnormality is found during surgery [7–9]. The presence of long mobile mesentery and dietary factors may contribute to the primary volvulus [9].

Secondary midgut volvulus usually occurs in older patients, with a peak incidence of sixth to eighth decades, affecting both the sexes equally, and is more common in western countries [7–9]. The bowel loop is twisted around an underlying point of fixation, causing closed-loop obstruction at two fixed points due to acquired or congenital lesions [10]. The most frequently encountered cause is postoperative adhesions. Other etiologies include internal hernia, external hernia (as noted in our case of obturator hernia), pregnancy, tumors, mesenteric lymph nodes, Meckel's diverticulum, mesenteric lipoma, endometriosis, abscess, aneurysm, hematoma and following gastric surgery. An incarcerated hernia may be complicated by a volvulus of bowel loop through a twist around the fixed small bowel loop at or near the hernial sac [6].

A volvulus represents the most common cause of strangulation with associated bowel ischemia. Characteristics CT findings in closed-loop obstruction include a C-shaped, U-shaped, or 'coffee bean' appearance of the bowel loop, mesenteric vessels converging towards the point of obstruction, fluid-filled bowel loop, a triangular loop, the beak sign and whirl sign at the site of obstruction [10]. Whirl sign represents twisting of the bowel loop with the superior mesenteric artery at the center of the whirl and is a CT sign of midgut volvulus.

The treatment of obturator hernia is surgery, with a variety of operative approaches including abdominal approach, retropubic approach, obturator approach, inguinal approach, and more recently, laparoscopic

approach [1–3, 5]. The abdominal approach is most commonly performed with the advantages of making a diagnosis, avoiding obturator vessels, exposing the obturator ring more easily and facilitating bowel resection, if necessary. Prosthetic plugs or mesh can be used to reinforce the obturator foramen. The laparoscopic surgery is less invasive and reserved for the high-risk patients and the nonstrangulated ischemia with benefits of less postoperative pain, less bowel ileus, shorter hospital stay and fewer complications.

CONCLUSION

Concomitant obturator hernia and midgut volvulus in an elderly and emaciated woman is a rare cause of bowel obstruction. Careful and thorough survey of the whole abdomen by using multidetector computed tomography, and recognition of the computed tomography characteristics can establish definite preoperative diagnosis. Early diagnosis and prompt emergent operation are essential to reduce the mortality and morbidity.

Author Contributions

Yeung Kwok-Wan – Drafting the article, Critical revision of the article, Final approval of the version to be published

Chang Ming-Sung – Conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

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Authors declare no conflict of interest.

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CASE REPORT

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Akinetic-rigid syndrome: An unusual presentation of hypoglycemic encephalopathy

Akhila Kumar Panda, Kushwaha Suman, Aldrin Anthony Dung Dung

ABSTRACT

Introduction: Hypoglycemia is an emergency. If untreated, it can result persistent neuronal injury of the central nervous system. The neurologic manifestations of hypoglycemia ranges from headache, seizure, different neuropsychiatric symptoms to stuporous, coma and stroke like symptoms. Akinetic-rigid syndrome, a severe form of extra pyramidal symptom is rarely described with hypoglycemic brain damage. The magnetic resonance imaging (MRI) findings are variable and reversible which ranges from normal study to diffusion restriction and white matter hyper intensities involving both cortical and sub cortical white matter as well as grey matter. **Case Report:** This case illustrates a 32-year-old male with history of intermittent hypoglycemia of unknown cause for three years duration presenting with prolonged akinetic mutism and extra pyramidal symptoms. His brain MRI revealed bilateral symmetrical white matter hyper intensities involving corona radiata, internal capsule and

middle cerebellar peduncles with 18F-fluorodeoxyglucose positron emission tomography with computed tomography (FDG PET CT) scan of brain showed hypometabolism involving bilateral parietal lobes, basal ganglia, thalamus and cerebellum, which reversed with correction of blood sugar within next four weeks.

Conclusion: Hypoglycemic encephalopathy (HE) may develop reversible extra pyramidal syndromes. Clinico-neuro imaging correlation is not always possible. Besides routine evaluation for hypoglycemia, neuroimaging such as MRI of brain and 18F-FDG PET CT scan may contribute to recognize the disease process and its outcome.

Keywords: Encephalopathy, Hypoglycemia, Cerebellar peduncle, Mutism, Akinetic-rigid syndrome

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INTRODUCTION

Cerebral hypoglycemia causes reversible metabolic brain insults. About 10% of type 2 and 4% in type 1 diabetes mellitus (DM) patients experienced severe hypoglycemia during natural course of the disease [1, 2]. The common neurological manifestations of hypoglycemia are confusion, 'bizarre' behavior, stuporous, coma, seizure, cognitive impairment,

headache, stroke like symptoms and generalized weakness [3, 4]. Extra pyramidal syndromes such as akinetic-rigid syndrome and mutism are the less appreciated cause of hypoglycemic brain damage.

CASE REPORT

A 30-year-old male laborer presented with recurrent episodes of unconsciousness for last three years. These episodes were preceded by perioral and hand numbness followed by diaphoresis, cool extremities. These episodes were persisting for several hours. Such events usually occur in the working field in the late period of daytime and at night. Several times in the early morning he was found unconscious along with severe perspiration. Initially, the frequency was two to three episodes per month which was gradually increased to one every week for last three months. Fifteen days prior to admission, he developed prolonged state of delirium. He was initially misdiagnosed as generalized epilepsy. General physical examinations showed patient was mute with spontaneous reflex eye movements, none communicating to either verbal or gestural commands with normal vital parameters. Cardiovascular, respiratory and gastrointestinal examinations were unremarkable. Nervous system examinations revealed, the patient was mute with spontaneous non purposeful eye blinking and eye movement with normal oculocephalic reflex. Overall cranial nerves examinations including fundus were non contributory. Motor system examinations showed severe generalized lead pipe rigidity involving appendicular as well as axial group of muscles with paucity of spontaneous limb movements, brisk deep tendon reflexes and flexor plantar responses. Detail sensory and cerebellar examinations were not possible. There was no evidence of autonomic instability.

LABORATORY INVESTIGATIONS

The patient had persistent hypoglycemia of random capillary blood glucose ranging from 35–60 mg/dL despite continuous intravenous 5% dextrose solution. Other blood parameters including liver function, kidney function, thyroid profile, complete blood count were unremarkable. His serum for human immune deficiency virus (HIV), hepatitis B and C and syphilis were negative. Cerebrospinal fluid (CSF) study showed acellular background with normal protein and sugar values. CSF polymerase chain reactions (PCR) for herpes simplex virus and antibody analysis for Japanese encephalitis and West Nile virus were negative. Dual phases contrast computed tomography (CT) scan and magnetic resonance imaging (MRI) of abdomen are normal which ruled out Insulinoma as well as other possible abdominal malignancy. The CT scan of chest was normal. Base line morning serum cortisol level was 11.8 µg/dL (normal range 5–25 µg/dL). Serum fasting insulin and C peptide level were 3.8 IU/mL (normal

range 2–25 IU/mL) and 1.94 ng/mL (0.9–7.1 ng/mL). Electroencephalogram showed generalized slow background delta wave activity suggestive of diffuse cerebral dysfunction. Brain MRI (3 Tesla) of T2 and FLAIR sequences showed bilateral symmetrical white matter hyper intensities involving middle cerebellar peduncle (Figure 1A). Diffusion weighted image (DWI) sequences showed hyper intensities in bilateral corona radiata, centrum semiovale and internal capsule (Figure 1B–C). FDG-PET/CT imaging showed decreased FDG uptake in bilateral parietal lobes, basal ganglia, thalamus and cerebellum (Figure 2). He was considered to be hypoglycemic encephalopathy of unknown cause and treated with continuous dextrose solution with frequent high carbohydrate diet. He started improving after two weeks of ICU stay. After four weeks of treatment he started responding and recognizing family members with severe hypo kinetic dysarthria and generalized rigidity. Repeat MRI of brain after four weeks showed resolution of all previous lesions. After two months of admission, he was clinically improved with minimal extra pyramidal symptoms. His both modified rankin scale (MRS) and barthel index scores were five of each at the time of admission which improved to score two and ninety, respectively within a period of two months of hospital stay.

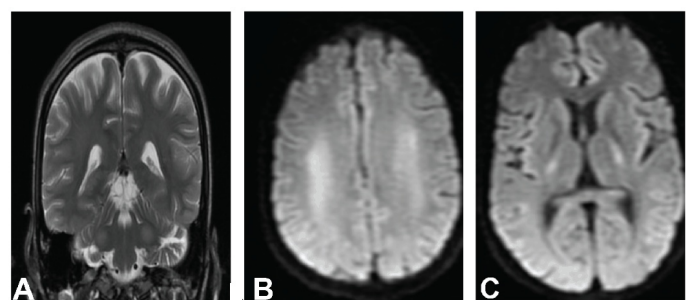


Figure 1: (A) Coronal section of T2-weighted sequence of magnetic resonance imaging of brain showing bilateral symmetrical hyper intensity of middle cerebella peduncle (MCP) (white arrow), (B) Diffusion weighted image showed bilateral symmetrical increased signal intensities in centrum semiovale. (black arrow), (C) Diffusion weighted image showing increased signal intensities in both internal capsules (white triangle).

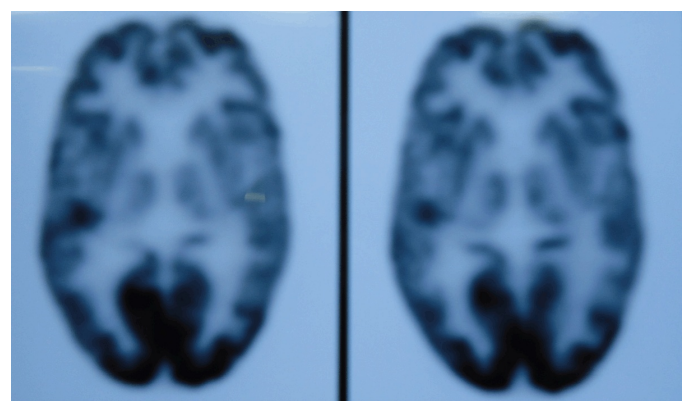


Figure 2: 18F-fluorodeoxyglucose positron emission tomography with computed tomography scan of brain showing decreased fluorodeoxyglucose uptake.

FOLLOW-UP

After three months of hospital discharge, he achieved pre morbid state except minimal rigidity of limbs. His speech became clear on conversation. He was maintaining normal blood glucose with high carbohydrate diet.

DISCUSSION

Though there is no consensus regarding clinical definition of this term, Jens Witsch et al. in their series defined this entity as a clinical state leading to stupor or coma with the blood glucose level of <50 mg/dL and persistence of clinical symptoms for ≥ 24 h despite normalization of blood glucose levels unaccompanied by other systemic complications and possible reasons simulating the same [5]. Most vulnerable area of brain to be affected by hypoglycemia are the subiculum, caudate neurons, hippocampus, the dentate gyrus and superficial cortical layers especially layers 2 and 3 [3].

The exact pathophysiology of hepatic encephalopathy is poorly understood. Hypoglycemic brain produces increase amount of aspartate from oxaloacetate due to lack of acetate in Krebs cycle leading to energy failure of about 25% of normal leading to selective tissue necrosis and brain damage. Hypoglycemic tissue alkalosis and apoptosis may contribute to the above pathogenesis [6]. Hypoglycemic brain damage usually results due to tissue alkalosis because of increased pH, absence of lactic acid and raised ammonia from deamination of amino acids [9]. It preferentially affects neurons and usually spares axons [8, 9]. The mechanism of infarction is quite different from that of hypoglycemic brain damage. Decreased brain pH, elevated lactate and reduced cellular redox system are the main culprit in ischemia [7]. Ischemia affects all parts of neurons as well as glial and endothelial tissues [8, 9].

Hypoglycemia may have variable clinical presentations. Parkinsonism secondary to hypoglycemic encephalopathy is due to degeneration of dopamine neurons in the substantia nigra pars compacta (SNc), secondary to ischemia or severe hypoglycemia. There is hyper polarization of dopamine neurons in acute brain injury during hypoxia and hypoglycemia. However, the role of excitatory synapses in SNc in this condition is poorly understood [10].

Akinetic-rigid syndrome implies difficulty in initiating movements (bradykinesia or akinesia), rigidity, progressive fatiguing, slow mentation (bradyphrenia) with or without tremor at rest and postural instability [11]. It is caused by diverse etiology. It is important to differentiate idiopathic parkinsonism to secondary parkinsonism because of different therapeutic options. The diagnosis is based entirely on clinical profiles.

In this case, the patient suffered from intermittent hypoglycemia for three years. Later on due to sustained unaware hypoglycemic state, he developed prolonged

delirium followed by severe extra pyramidal symptoms in the form of akinesia, rigidity (axial and appendicular), bradyphrenia, and severe hypophonia with postural instability. With replacement of glucose, patient showed clinical improvement and maintained his independent daily activities after three months. We found the following three atypical manifestations in our case:

(1) Akinetic-rigid syndrome secondary to persistent hypoglycemia

(2) Rare neuroimaging findings and

(3) Discordant FDG-PET scans with relation to MRI findings and subsequent recovery of the lesions.

The prognosis of hypoglycemic encephalopathy depends on co morbidities, associated medical complications, early recognition and treatment, extent of neuroimaging findings and its reversibility. The mortality ranges from 23–46% in various series and when the outcome survived is excellent [5].

CONCLUSION

Hypoglycemic encephalopathy is a rare emergency which may develop reversible extra pyramidal symptoms like akinetic-rigid syndrome. Neuro imaging correlation is not always possible to describe the temporal profile of the patients. Besides routine evaluation for hypoglycemia, neuroimaging such as magnetic resonance imaging and ^{18}F -fluorodeoxyglucose positron emission tomography fused with computed tomography scan may contribute to recognize the disease process and its outcome

Author Contributions

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

Suman Kushwaha – Analysis and interpretation of data, drafting the article, Final approval of the version to be published

Aldrin Anthony Dung Dung – Acquisition of data, drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Abbreviations

SNC : Substantial nigra pars compacta

CNS : Central nervous system

MRI : Magnetic resonance imaging

MCP : Middle cerebellar peduncle

FDG PET CT : 18F- fluorodeoxyglucose positron emission tomography fused with computed tomography

CT : Computed tomogram

CSF : Cerebrospinal fluid

DNA : Deoxy ribonucleic acid

FLAIR : Fluid attenuated inversion recovery

HIV : Human immune deficiency virus

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CASE REPORT

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Magnetic resonance imaging features of vaginal endometriosis

Valeria Fiaschetti, Valentina Cama, Laura Greco, Maria Fornari,
Giuseppe Sorrenti, Giovanni Simonetti

ABSTRACT

Introduction: Deeply infiltrating endometriosis (DIE) is defined by the presence of endometrial implants penetrating under the peritoneal surface or under the wall of the pelvic organs to a depth of at least 5 mm. This case report describes a case of DIE involving vaginal wall, periurethral tissue, torus uterinus and sigmoid wall. **Case Report:** A 46-year-old female was presented with pelvic pain and deep dyspareunia which started six years ago, after a spontaneous interruption of pregnancy and uterine curettage. The pelvic magnetic resonance imaging (MRI) showed multiple DIE lesions localized in the vaginal wall mainly in the lower third of the vagina, between the anterior vaginal wall and the urethra, on the contour of the urethra and between the torus uterinus and the sigmoid wall. After MRI, laparoscopic biopsies confirmed the DIE lesions. **Conclusion:** According to the

implantation theory, we assume that in our case the vaginal endometriosis was the result of implantation of endometrial glands into the vaginal tearing during the surgical procedure of curettage. In the current literature no author has so far described a diffuse involvement of the vaginal wall in the presence of uterine curettage after interruption of pregnancy. In our case, the anterior (peri-urethral tissue), middle (vaginal wall) and posterior (torus uterinus and bowel wall) compartments are involved. We can presume that the vaginal DIE is an early lesion, while the others lesions arise from vaginal walls by a contiguous extension. On MRI with endovaginal gel these lesions were more evident rather than without gel. MRI with gel allows us to give essential preoperative mapping for the surgical removal of the DIE lesions.

Keywords: Vaginal endometriosis, Deeply infiltrating endometriosis (DIE), Endovaginal gel

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INTRODUCTION

Endometriosis is a chronic gynecologic disorder affecting women in reproductive age; in the general population the prevalence of endometriosis is 10% [1]. It is characterized by the presence of functional

endometrial glands and stroma outside the uterine cavity. The most common localizations of endometriosis are the ovaries and the pelvic peritoneum, followed by deep lesions of the pelvic subperitoneal space. Deeply infiltrating endometriosis (DIE) is a specific entity, histologically defined by the presence of endometrial implants penetrating under the peritoneal surface or under the wall of the pelvic organs to a depth of at least 5 mm [2]. It involves more frequently fibromuscular structures such as uterosacral ligaments and rectovaginal septum (RVS) (69.2% of cases), rarely the vagina (14.5%) [3]. This case report describes a case of DIE involving vaginal wall, periurethral tissue and torus uterinus with involvement of the sigmoid wall.

CASE REPORT

We reported a case of a 46-year-old female. Her clinical history consisted of exacerbated pelvic pain during her menstrual cycle and deep dyspareunia which started six years ago, after spontaneous interruption of pregnancy and uterine curettage.

The clinical examination detected painful thickening of the vagina but the ultrasonography did not report abnormal findings except for bilateral ovarian cysts and multiple small uterine fibroids. Vaginal bleeding was observed after introduction of the speculum.

The patient was referred by the Department of Gynecology and Obstetrics for clinical suspicion of DIE which was based on the detection of painful thickening of the vagina on physical examination.

We performed a pelvic magnetic resonance imaging (MRI) with a 1.5 T scanner (Achieva, Philips medical systems, BEST, Netherlands) using a pelvic phased-array coil. We used peristaltic inhibitors (10 mg of hyoscine-N-butylbromide, buscopan) diluted in 10 mL saline to reduce gastrointestinal tract peristalsis.

The imaging protocol included a T1-weighted (W) turbo spin-echo (TSE), a T2-W TSE, a T1-W TSE fat-saturation, performed in the transverse plane (perpendicular to axis of cervix). A T2-W TSE sequences and a T1-W TSE fat-saturation were performed on sagittal plane. The transverse and sagittal T2-W TSE and the transverse and sagittal T1-W TSE fat-saturation sequences were performed before and after the introduction of ultrasonographic gel (100 mL) into the vaginal canal. No intravenous contrast medium was administered.

The MRI showed a hypointense concentric thickening of the vaginal wall on T2-W sequences. The thickening was mainly evident in the lower third of the vagina, where it caused pseudostenosis. On the sagittal, T2-W sequence with endovaginal gel this finding was more evident than that without the use of the gel (Figure 1A–B). On T2-W sequence between anterior vaginal wall and urethra, above the perineal body, a hyperintense focus was revealed. It was more evident on the sequence with gel (Figure 1B). On the T1-fat, saturation sequence showed a hyperintense signal

indicating bloody content. Urethral involvement was suspected for hypointense irregular spiculated mass-like thickening on the contour of the urethra involving the anterior vaginal wall (Figure 2). Thin hyperintense lines were revealed in the posterior vaginal wall, clearly visible in the T1 fat-saturation sequence after the introduction of gel into the vagina indicating bloody content (Figure 3). The T2-W sequence showed thin hypointense lines located between the torus uterinus and the sigmoid related to retractile adhesions (Figure 4). After MRI, patient underwent to multiple biopsies to obtain a definitive diagnose. Histopathology tests confirmed islands of endometrial tissues and stroma in the vaginal wall, periurethral tissue and torus uterinus.

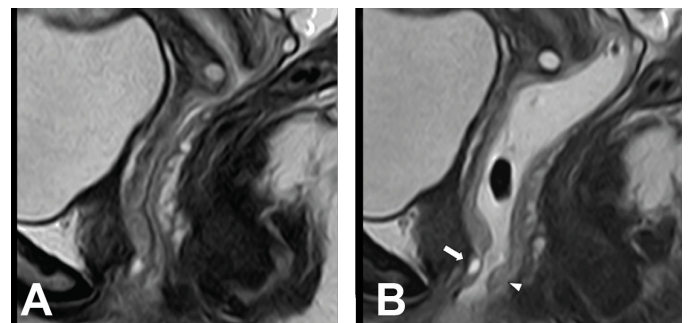


Figure 1: (A, B) Sagittal T2-weighted sequences shows a diffuse thickening vaginal wall and a hyperintense oval lesion localized between the anterior vaginal wall and the urethra, above the perineal body. (A) It is poorly detectable without gel. The vaginal pre-orificial pseudostenosis was not appreciable on the image without gel. (B) The image after gel allows to better evaluate the thickening of the vaginal wall, mainly evident in the lower third where it caused pseudostenosis (arrowhead) and the oval focus (arrow).

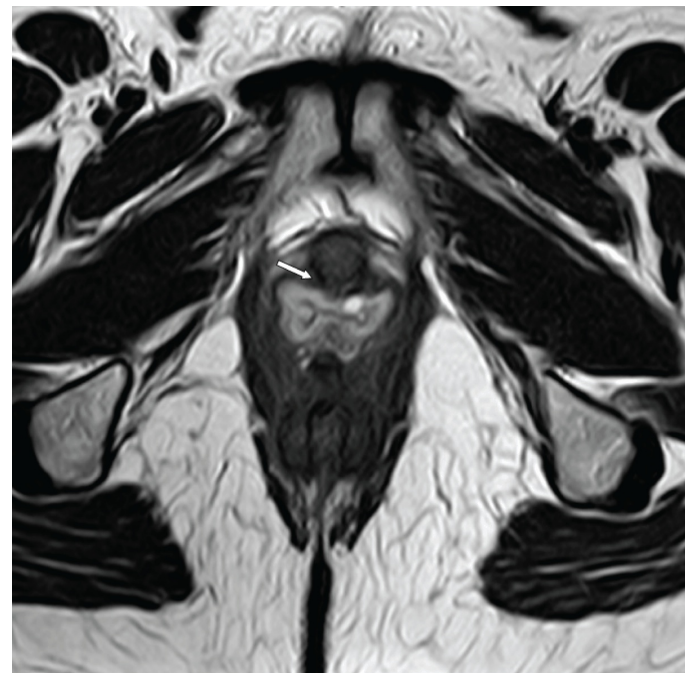


Figure 2: Axial T2-weighted sequence shows an hypointense irregular spiculated mass-like thickening on the contour of the urethra. It involves the anterior vaginal wall (arrow).

The excision of all endometriotic implants is the treatment chosen to control the symptomatology. Following the histopathological response she took pre-operative hormone therapy to reduce the size of the lesions and decrease the risk of postoperative incontinence.

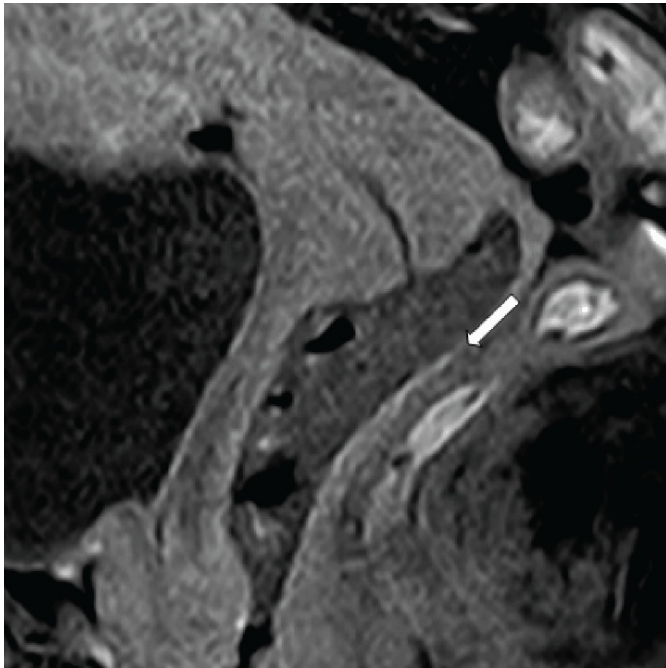


Figure 3: Sagittal T1-weighted sequence shows a thin hyperintense lines of the posterior vaginal wall indicating plaques bloody content (arrow). This finding is visible only after vaginal distension.



Figure 4: Axial T2-weighted sequence shows the hypointense lines located between the torus uterinus and the sigmoid related to retractile adhesions (arrow).

DISCUSSION

The pathogenesis of endometriosis remains controversial and is probably multifactorial. The most widely accepted theory is the implantation theory (Sampson 1927). According to this theory, the lesions are secondary to the implantation and the proliferation of regurgitated endometrial cells in an ectopic position. Angio- and lymphatic theory, impairment of the immune response, heritable tendencies and secreted products of endometriotic lesions may contribute to explain the pathogenesis of this disorder [4, 5].

In this case deep dyspareunia and pelvic pain started immediately after spontaneous interruption of pregnancy and uterine curettage. According to the implantation theory we assume that the vaginal endometriosis was the result of implantation of endometrial glands into vaginal tearing during surgical procedure of curettage. In the current literature some authors have described cases of extraperitoneal locations including vagina, RVS and mostly on episiotomy scars, after obstetrical and surgical trauma, in particular after natural delivery [6]. No author has so far described diffuse involvement of vaginal wall in presence of uterine curettage after pregnancy interruption. Our patient had a history of two cesarean sections as well.

According to a functional and clinical classification in our case the anterior, middle and posterior compartments are involved. In fact, the endometriotic lesions affect periurethral tissue (anterior compartment), vagina (middle compartment) and torus uterinus and bowel (posterior compartment) [7]. We can presume that the vaginal DIE was a previous lesion, while the others lesions arise from vaginal walls through a contiguous extension. Isolated urethral involvement is poorly described in literature. It is more frequently observed as an extension from the others organs such as the bladder [7].

In this case ultrasonography was the first diagnostic tool used. It is recommended for the study of the ovary and bladder but its sensitivity is lower than that of MRI in identifying subperitoneal lesions extensions [8]. Ultrasonography reported the presence of endometrioma in the right adnexal, cyst in the left adnexal and multiple nodules of uterine fibroids.

The MRI showed a hypointense concentric thickening of the vaginal wall on T2-W images. The use of gel allowed us to better evaluate this finding. It was mainly evident in the lower third of the vagina where it caused pseudo-stenosis (Figure 1B). The pseudostenosis was not appreciable on the sequence without gel (Figure 1A).

In another recent report, we demonstrated that the use of intravaginal gel increases the sensitivity of MRI without gel in detecting DIE lesions. Diagnostic difficulties of DIE are related to the inherent features of endometriotic lesions which can have an MRI signal intensity very similar to those of the surrounding fibromuscular anatomic structures. Also DIE involves multiple closely related anatomical structures and with only subtle signal alterations. In this regard the advantage of the gel is its hypersignal on T2-W image

which contrasts the hyposignal of the surrounding fibromuscular anatomical structures, therefore facilitating the diagnosis [9].

Some authors claim that the presence of small foci of high signal within of fibrotic lesions can facilitate the diagnosis of DIE. Our case contradicts this assertion. In fact, the endometriotic blood lesion localized between the anterior vaginal wall and the urethra, above the perineal body, was poorly detected in the sequence without gel (Figure 1A) rather than with gel (Figure 1B). The intravaginal gel relaxing the vaginal walls and the fornices allowed a better view of the anterior deep pelvic area and of the retrocervical area, as confirmed by literature [9].

The thin lines of the posterior vaginal wall, hyperintense on T1-W fat saturation sequences, indicating blood plaques, was detected only after vaginal distension (Figure 3).

An MRI with endovaginal gel allows us to give essential preoperative mapping for the surgical removal of the DIE lesions. Furthermore, this method stands out because higher sensitivity compared to transvaginal ultrasonography and MRI without gel, detecting more lesions [9].

A wide excision of all endometriotic implants remains the chosen treatment and an accurate preoperative assessment of the extension of the endometriosis lesions is necessary for a successful treatment and to control the symptomatology.

In deep extraperitoneal area the radical excision of endometriosis lesions could include partial excision of the sphincter, which that may compromise the urinary continence.

In this case, hormonal therapy was used before of the surgical procedure in order to decrease the risk of postoperative incontinence and to decrease the recurrence rate. In a case series of six patients Liang et al. observed recurrence in patients treated with only surgery, while none of the patients treated with surgical and medical therapy had recurrence of endometriosis [5].

Also the use of preoperative hormonal therapy could reduce the size and facilitate the excision of the lesion reducing damage to surrounding tissue [10].

CONCLUSION

In literature, many authors have described deeply infiltrating endometriosis but none of them has so far described diffuse involvement of the vaginal wall and of the periurethral tissue without involvement of the bladder yet. Furthermore, our study is the first one to describe a case of triple concomitant affected compartments; periurethral tissue (anterior compartment), vagina (middle compartment) and torus uterinus and bowel (posterior compartment) in patient with deep extraperitoneal endometriosis.

Author Contributions

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

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

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

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

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Guarantor

The corresponding author is the guarantor of submission.

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Authors declare no conflict of interest.

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CASE REPORT

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Vascular stromal mass: A unique histological entity mimics malignancy on breast magnetic resonance imaging

Jon Gerry, James Kuo, Kristin Jensen, Irene Wapnir

ABSTRACT

Introduction: Breast magnetic resonance imaging (MRI) is gaining routine acceptance for screening of high-risk patients and is used selectively in the diagnostic work-up of women with undetermined lesions or breast cancer. Malignant masses can be differentiated from benign lesions based on margin characteristics, dynamic enhancement features and size. **Case Report:** A 58-year-old female underwent an excisional biopsy for a non-palpable, MRI-detected mass lesion with irregular borders. The lesion demonstrated marked enhancement on postcontrast images with rapid wash-in and plateau delay phase kinetics. After excision, a 1.5-cm vascular stromal mass (VSM) comprised of admixed fibrous and fatty tissue with a non-organized proliferation of ectatic blood vessels lined by a single layer of cytologically bland endothelial cells was identified. The described pathological entity is histologically distinct from pseudoangiomatous stromal hyperplasia hemangioma or angiolipoma. The MRI enhancement features of the VSM mimicked those of carcinoma, attributable to the characteristic ectatic blood vessels. Moreover, the admixed fibro-fatty stroma comprising the

mass is otherwise radiographically and sonographically similar to adjacent breast tissue, making it undetectable by mammography and ultrasound. **Conclusion:** The VSM is a unique histological entity visualized only by breast MRI. Its appearance is indistinguishable from malignant lesions on MRI and so requires a biopsy.

Keywords: Vascular stromal mass (VSM), Pseudoangiomatous stromal hyperplasia (PASH), Hemangioma, Angiolipoma

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INTRODUCTION

Breast magnetic resonance imaging (MRI) is gaining acceptance for use in screening of patients at high-risk of developing breast cancer and as an additional diagnostic study in the work-up of women with undetermined breast lesions or breast cancer. Mammography is the only prospectively tested breast cancer screening imaging modality that has been validated in the detection of breast cancer. It is also uniquely effective in the discovery of microcalcifications associated with carcinoma in situ. However, it has limited sensitivity in women with dense breasts [1]. Ultrasound has been incorporated more regularly into the routine evaluation of patients with breast complaints, largely used to characterize lesions

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discovered on physical exam or another imaging modality. It can differentiate solid from cystic lesions as well as define the margins of breast masses. Dedicated breast MRI was introduced in 1990s. It employs a gadolinium-based contrast agent and its detection of lesions is dependent on the neovasculature or existing capillaries within lesions [2]. In a retrospective analysis of women 25–89 years of age, dedicated breast MRI had a 92% sensitivity and an 89% specificity when used either as a screening or diagnostic tool [3]. The false-positive rate was 11%, and most false-positive specimens were described as ‘benign breast tissue’. While non-mass lesions tend to account for a majority of false-positive findings on MRI, malignant mass lesions can be differentiated from benign ones based on margin characteristics, dynamic enhancement features, and size [4]. When breast MRI is used in combination with mammography and breast ultrasound, the excisional biopsy rate may double in exchange for a 10% increase in detection of invasive cancer [5].

CASE REPORT

A 58-year-old female was referred from another facility because of an MRI-detected mass in the right breast. The study was prompted after routine screening mammography detected an area of focal asymmetry. Additional diagnostic work-up showed this area to be overlapping glandular tissue without a corresponding sonographic abnormality. The patient had a significant family history for a mother diagnosed with breast cancer in her fifties, which led her to seek a second opinion. Her past medical history was unremarkable aside from early onset menopause at age 30 following a total abdominal hysterectomy and bilateral salpingo-oophorectomy for benign disease. She, subsequently, took hormone replacement therapy for 28 years.

On physical exam, the breasts were large and pendulous with a normal contour. Repeat diagnostic mammography and ultrasound at our institution revealed a focal asymmetry with corresponding acoustic shadowing in the right breast. A core needle biopsy was taken of the abnormality under ultrasound guidance and a marker was placed. This proved to be benign breast tissue. A repeat breast MRI was performed with a 1.5 T scanner. On precontrast images, the mass was both T1 and T2 isointense to the surrounding breast parenchyma. The mass demonstrated marked enhancement on postcontrast images with rapid wash-in and plateau delay phase kinetics with irregular margins (Figure 1A–D). The location of the biopsy marker was discordant, specifically 3.5 cm anterior and lateral to the 2.5x1.7x2.4 cm contrast-enhancing mass lesion.

An MRI-guided wire localization with excisional biopsy was performed because of the suspicious characteristics of this mass lesion and the discordant radiopathological findings on core needle biopsy. A 6.0x5.5x2.5 cm specimen was excised, margins were inked, and the tissue was serially sliced and

radiographed after fixation overnight. No gross or specimen radiograph abnormalities were identified. Microscopic analysis showed proliferative fibrocystic changes. A 1.5-cm mass-like lesion comprised admixed fibrous and fatty tissue was focally identified. Within this area there was a non-organized proliferation of mildly to moderately ectatic blood vessels lined by a single layer of cytologically bland endothelial cells (Figure 2A–B). Immunohistochemical studies using antibodies to CD31, CD34 and calponin highlighted these cells but did not show p63 nuclear staining, suggesting that the lining of the vessels was a true endothelium. No inter-anastomosing channels, cellular atypia, or evidence of rapid cellular proliferation to suggest angiosarcoma was noted.

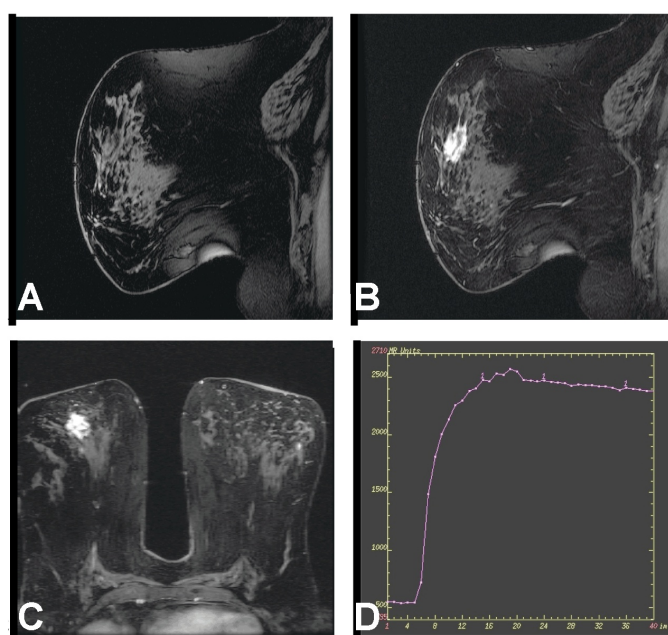


Figure 1: Mass with irregular margins in the right breast, 4.5 cm from nipple. (A) Precontrast sagittal T2-weighted magnetic resonance imaging of the right breast, (B) Postcontrast sagittal T2-weighted magnetic resonance imaging of the lesion in the right breast, (C) Postcontrast T2-weighted axial magnetic resonance imaging of the lesion, (D) Contrast enhancement kinetic profile showing a rapid wash-in and a plateau in the delay phase.

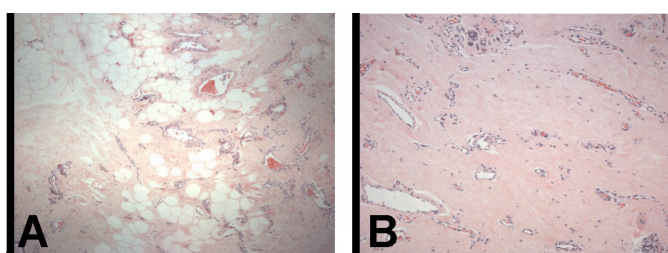


Figure 2: (A) Low-magnification image of the excised mass demonstrates vascular proliferation with admixed fibrous and fatty tissue (H&E stain, x50), (B) Higher magnification image of the mass reveals a bland cytology of the vessels and dense stroma (H&E stain, x100).

DISCUSSION

Herein, we describe a non-palpable, suspicious mass identified on breast MRI, called a vascular stromal mass (VSM), which is histologically distinct from pseudoangiomatous stromal hyperplasia (PASH), hemangiomas, and angioliipomas, although these benign breast lesions bear the closest histological resemblance to the VSM. The VSM has a tortuous yet bland vasculature contained within a discrete fibro-fatty stromal mass. The PASH is characterized by a dense collagenous stroma arranged around anastomosing cleft spaces lined with spindle-shaped myofibroblasts and not actual vascular endothelium [6]. In contrast, the VSM consists of endothelium-lined ectatic blood vessels that were confirmed on immunohistochemical stains. Hemangiomas tend to contain capillary or cavernous vascular components with bland-appearing endothelial cells, and they are divided into lobules by fibrous septations [7]. The VSM vascular component is not organized into lobules. Also, the VSM has a more prominent stromal component than a hemangioma. Hemangiomas may also contain tiny calcified phleboliths, presumably caused by hemostasis within the hemangioma and thrombus formation. Phleboliths were not observed in the VSM. Angioliipomas are extremely rare in the breast. They are characterized by a predominance of adipocytes and the presence of hyaline thrombi [8]. The VSM has a predominant fibrous stromal component and there were no hyaline thrombi noted within the mass.

An MRI characteristics of the VSM are different from PASH, hemangiomas and angioliipomas, but indistinguishable from carcinoma based on margin characteristics and contrast enhancement kinetics. Importantly, the VSM is T1 and T2 isointense and is only visualized on MRI after administration of intravenous contrast with rapid wash-in and plateau delay phase kinetics. The PASH lesions demonstrate variable MRI signal intensity with a slower wash-in that continues during the delay phase [9]. Breast hemangiomas are T2 hyperintense and have increased contrast enhancement on the delay phase similar to PASH [10]. Angioliipomas of the breast are rare, and MRI characteristics have not been reported. Angioliipomas of spine have variable T1 signal intensity, are typically T2 hyperintense, and display variable contrast enhancement. Finally, the irregular margins of the VSM distinguish it from PASH, hemangiomas, and angioliipomas.

CONCLUSION

We characterize vascular stromal mass as a new benign histological entity which exhibits features on breast magnetic resonance imaging reminiscent of malignant lesions. If discovered on a core needle biopsy, complete surgical excision would be advised given its rarity.

Author Contributions

Jon Gerry – 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

James Kuo – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article

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Irene Wapnir – 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

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Solitary plasmacytoma of clavicle: A case report

Aruj Dhyani, Abhash Shankar, Arunima Gupta, Rupesh Kumar,
Koushik Chatterjee, Anup Majumdar

ABSTRACT

Introduction: Plasmacytoma is a localized collection of malignant plasma cells. The disease can be divided into solitary plasmacytoma of the bone (SPB) and extramedullary plasmacytoma (EMP), both of which are distinct entities. An EMP, the rarer of the two, has soft tissue infiltration by clonal plasma cells without any occult or systemic myeloma. Whereas, SPB has a solitary lytic bone lesion with infiltration of monoclonal plasma cells, with absence of the same on a random marrow sampling or any evidence of systemic myeloma. Though SPB may involve any bone in the body but involvement of medial end of clavicle, though reported, is extremely rare. **Case Report:** Herein, we report a case of a 65-year-old farmer presented with pain and swelling of the medial end of the left clavicle, which was postoperatively diagnosed to be a solitary plasmacytoma by virtue of histopathology and IHC, with no other evidence

of osteopenic lesions on radiographs, a negative bone scan, absence of marrow involvement by monoclonal plasma cells, presence of M-band on serum protein electrophoresis, in the IgG region and no evidence of anemia, hypercalcemia, hyperuricemia, hypercreatininemia, nephropathy, respiratory and urinary tract infections that can suggest the presence of any systemic myeloma. **Conclusion:** The case of solitary plasmacytoma of the clavicle reported here is a very rare as a disease entity, and rarest because of the site of involvement. The patient was treated with surgery and radiotherapy and had excellent disease control.

Keywords: Solitary plasmacytoma bone (SPB), Multiple myeloma, Clavicle, Extramedullary plasmacytoma

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INTRODUCTION

Solitary plasmacytoma of the bone (SPB) comprises only 3–5% of all plasma cell neoplasms [1]. The new world health organization (WHO) criteria define SPB as a solitary lytic bone lesion with infiltration of monoclonal plasma cells, with absence of plasmacytosis on a random marrow sampling, with absence of any evidence of systemic myeloma (renal insufficiency/anemia/hypercalcemia) and a negative bone scan; along

with a monoclonal gammopathy on serum protein electrophoresis.

The SPB more commonly involves the axial skeleton, with an active marrow, as in vertebra, ribs, skull, pelvis, femur, clavicle and scapula relatively sparing the appendicular skeleton [1].

Primary bone tumors and tumor-like lesions of clavicle are uncommon, comprising only 0.45% of the total, and their occurrence is more likely to be malignant, than benign [2, 3]. We hereby present a case of SPB involving the clavicle, which is rare as per the disease entity and rarer because of the site involved.

CASE REPORT

A 65-year-old male farmer was presented with a two-month history of swelling and a month old pain, increasing on exertion, around the left shoulder, with loss of appetite for the same duration. The lump was initially the size of a lemon, before it progressed. Clinical examination revealed a hard non-tender lump on the medial end of the left clavicle 4x5 cm with restricted shoulder mobility. There were no palpable nodes. Radiographs of chest showed a lytic expansile lesion involving medial one-third of left clavicle associated with a soft tissue mass in the apical region of the chest (Figure 1).

A plain non-contrast computed tomography (CT) scan of the neck and thorax is shown in Figure 2.

- (i) Normal CT scan of the neck,
- (ii) Soft tissue density lesion with bony expansion and erosion of medial end of left clavicle with adjacent soft tissue density lesion and no evidence of rib crowding- bone biopsy suggested,
- (iii) Fibrotic strands in both upper and right lower lobes.
- (iv) Simple liver cyst.

Ultrasonography of the whole abdomen showed a simple cyst in right lobe of liver. Blood counts, liver and kidney function tests were normal except for serum LDH 725 U/L. Bone marrow examination revealed anemia of chronic disease, with no evidence of plasmacytosis or secondary deposits.

Fine needle aspiration of the clavicular lump revealed a malignant mesenchymal neoplasm, possibly spindle cell sarcoma with lymphoplasmacytic infiltration. Independent review of the slides too, did not reveal the presence of any abnormal or immature plasma cells, probably because of sampling error or error in terms of slide preparation or preservation (Figure 3). The patient then underwent an excision of the left clavicular lump. The postoperative histopathology showed the presence of a plasma cell neoplasm, comprising sheets of immature plasmacytoid cells, admixed with a few mature plasma cells (Figure 4). Immunohistochemistry (IHC) showed expression of CD 138 and CD 38, and are immunonegativity for cytokeratin, CD 56 and CD 20, confirming the diagnosis to be of plasma cell neoplasm. The IHC for immunoglobulin light chain was not done.

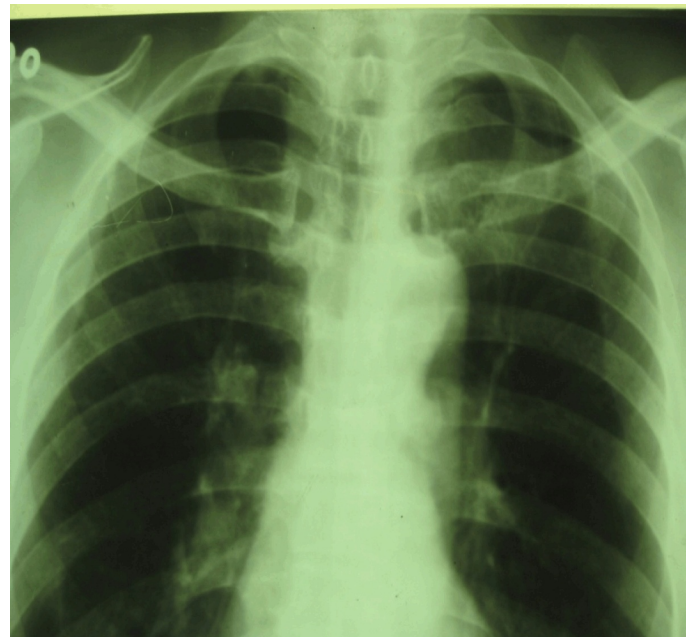


Figure 1: Radiograph of the left clavicle showing lytic expansile lesion involving medial end of the left clavicle associated with a soft tissue mass in the apical region of the chest.

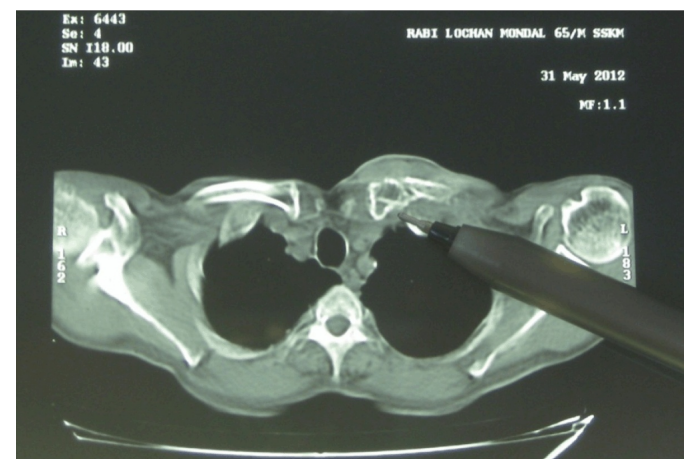


Figure 2: Computed tomography scan of upper thorax showing an expansile lytic lesion in the medial end of the left clavicle.

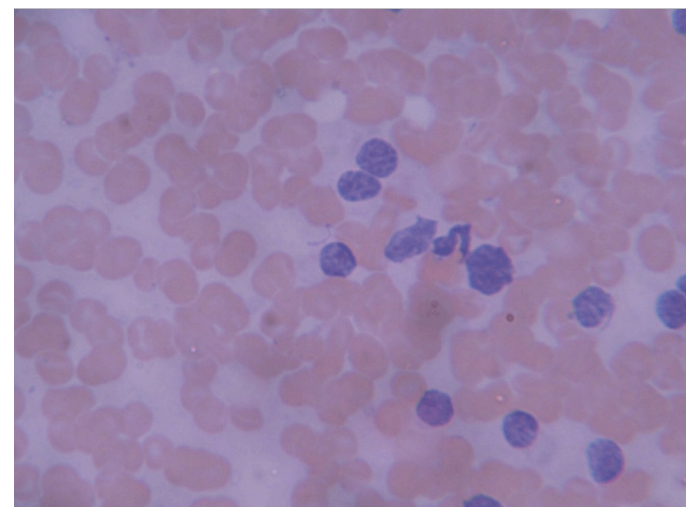


Figure 3: Features suggestive of malignant mesenchymal neoplasm, possibly spindle cell sarcoma with lymphoplasmacytic infiltration.

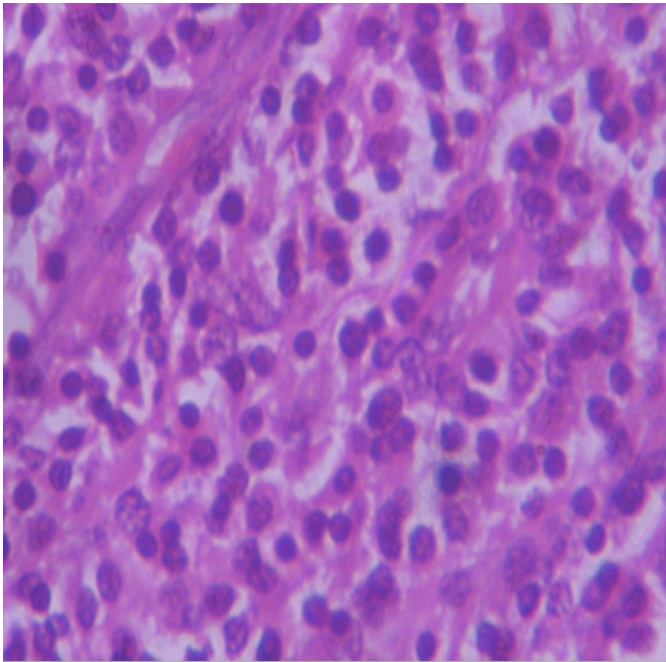


Figure 4: Microscopic examination showing tumor composed of sheets of immature plasmacytoid cells with mature plasma cell also present.

Serum protein electrophoresis showed the evidence of monoclonal gammopathy in the IgG region. There was no Bence Jones protein in urine. Serum calcium was 9.2 mg/dL with albumin 3.8 g/L, X-rays revealed no evidence of any other lytic bone lesions, the whole body bone scan was negative, and no evidence of urinary tract or respiratory tract infections.

This patient was then treated with postoperative radiotherapy. At the third month of follow-up the patient is doing well with almost full range of movement at left shoulder joint and normal daily activities.

DISCUSSION

Primary bone tumors and tumor-like lesions of clavicle are uncommon. Klein et al. presented a series of around 1300 primary bone tumors and found that only 0.45% of them involved the clavicle [2]. In a review of 35 primary bone tumors of the clavicle that have been treated at the Memorial Sloan-Ketterin cancer centre, Smith et al. reported only five benign lesions [3]. Primary bone tumors of the clavicle are therefore more likely to be malignant than benign. Solitary plasmacytoma of the bone accounts for 10–30% of primary chest wall malignancies. They are more common in male patients and usually occur at mean age of 60 years or later. The most common chest wall location is ribs, clavicle, sternum [4]. Considerable debate exists regarding the relationship of SPB and EMP to multiple myeloma. Most authors believe that SPB is simply an early presentation of multiple myeloma whereas others consider that SPB is a clinically distinct entity [5].

As far as treatment is concerned excision remains the mainstay of treatment, where ever possible. Radiotherapy has also shown to be successful for local control of solitary plasmacytoma [6]. However, chemotherapy is recommended to patients with disseminated disease and evidence of progression following the primary modes of treatment. This case is extremely rare, and the patient had an excellent disease control with surgery and radiotherapy. Long-term follow-up will be necessary for our patient as 35–55% of patients of SPB, might end up developing multiple myeloma over a period of 10–12 years from initial diagnosis [7].

CONCLUSION

We reported a case of solitary plasmacytoma of clavicle which is a rare disease, presenting in an extremely rare site. The diagnosis was confirmed by clinical suspicion, postoperative biopsy, immunohistochemistry, and other investigations relevant to rule out the presence of systemic myeloma. The patient was treated by surgery followed by radiotherapy. As per world literature this is the standard of care. The patient had excellent disease control, and is currently undergoing follow-up.

Author Contributions

Aruj Dhyani – Conception and design, Drafting the article, Final approval of the version to be published
Abhash Shankar – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Arunima Gupta – Acquisition of data, Critical revision of the article, Final approval of the version to be published
Rupesh Kumar – Acquisition of data, Critical revision of the article, Final approval of the version to be published
Koushik Chatterjee – Conception and design, Critical revision of the article, Final approval of the version to be published

Anup Majumdar – 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

Authors declare no conflict of interest.

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CASE REPORT

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Giant chylous cyst of liver: A case report

Mayank Baid, Manoranjan Kar, Sayak Roy, Someshubhra Datta Roy

ABSTRACT

Introduction: We present here a case of chylous cyst of liver with multiple cysts in mesentery, in a 14-year-old. **Case report:** A 14-year-old female presented in surgical outpatient unit with gradual swelling of abdomen for five months without any history of fever, jaundice, trauma or previous operation. Ultrasound and contrast enhanced computed tomography scan suggested a large multiseptate space occupying hypodense lesion. On laparotomy, a huge lobulated cyst containing clear fluid was found arising from left lobe of liver extending up to pelvis. Multiple small cysts were present in mesentery, mesoappendix and serosa of gallbladder. The giant cyst was excised completely with cystic left lobe of liver. Histopathology suggested chylous cyst of liver. There is no recurrence after two years of follow-up. **Conclusion:** We conclude that patient had chylous cysts at multiple places with largest from liver.

Keywords: Chylous cyst, Cyst of liver, Multiple cysts

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INTRODUCTION

In 1842, von Rokitsansky described a chylous mesenteric cyst. Since then, a few reports of chylous cysts are reported. We present here a case of chylous cyst of liver with multiple cysts in mesentery, in a 14-year-old female. As per our review, this may be a first reported giant chylous cyst of liver.

CASE REPORT

A 14-year-old female was presented at our surgical outpatient unit with complaint of gradual swelling of the upper abdomen and pain at the same site for five months. There was no history of fever, nausea, vomiting or jaundice. Bladder and bowel habits were normal. There was no history of loss of weight but slight decrease in appetite. There was no history of trauma to abdomen or any previous operative intervention in the abdomen.

On examination patient had mild pallor, no jaundice, pulse 84 beats/minute, blood pressure 110/70 mmHg, respiratory rate 18/minute, no fever, and had no lymphadenopathy. There was a huge swelling in epigastrium extending to right hypochondrium laterally and extending beyond the umbilicus inferiorly (Figure 1). On palpation swelling was cystic, non-tender, intra abdominal, margin was ill defined and local temperature was not raised. Liver and spleen were not separately palpable. There was no ascites. Air entry on both sides of chest, and spine were normal.

Laboratory examination showed hemoglobin 10.4 g/dL, total count of white blood cell 6000/μL, Total

bilirubin 1.1 mg/dL, SGPT 196 U/L, SGOT 173 U/L, and alkaline phosphate 386 U/L (Normal <98 U/L), serum amylase level 67 IU/dL. Ultrasonography and contrast enhanced computed tomography (CT) scan suggested a fairly large multiseptate space occupying lesion of size 20x12 cm in relation to left lobe of liver, abutting the liver and pancreas, with impression of resolving liver abscess or pancreatic pseudocyst (Figure 2).

During exploratory laparotomy, on opening the peritoneum a huge lobulated cyst with clear content was found arising from the left lobe of liver extending up to pelvis (Figure 3). Multiple very small cystic lesions were also seen to be arising from mesentery of the intestine (Figure 4), mesoappendix and serosa of the gallbladder. Right lobe of liver, kidney, spleen, intestinal wall and stomach appeared normal. After aspiration of content, the cyst completely collapsed and was excised in toto including the involved part of left lobe of liver. The nature of contained fluid was clear. Patient underwent cholecystectomy and appendicectomy. Cysts arising from mesentery were left intact. Postoperative period was uneventful. Histopathological report was cystic lesion lined by flattened endothelium, the cyst wall composed of fibrocollagenous tissue infiltrated with lymphocytes with impression of chylous cyst of liver, gallbladder wall infiltrated by chronic inflammatory cells and appendix showed lymphoid hyperplasia.



Figure 1: Clinical photograph of abdomen of the patient.

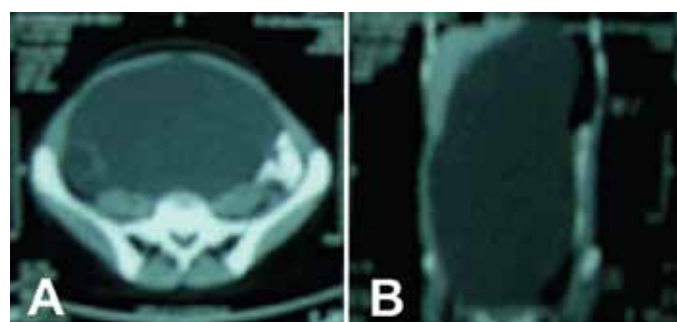


Figure 2: Contrast enhanced computed tomography scan of abdomen showing hypodense multiseptate lesion involving left lobe of liver extending up to pelvis.



Figure 3: Peroperative photograph of the cyst.



Figure 4: Multiple small cysts arising from mesentery.

After the operation the patient remained asymptomatic and there was no development of any detectable cyst on ultrasonography or recurrence in liver after two years on follow-up.

DISCUSSION

Classically, cystic disease of the liver is divided into nonparasitic and parasitic cyst. Nonparasitic cyst is more prevalent worldwide, although prevalence may vary significantly by geographic region. Generally, most hepatic cysts, regardless of type remain asymptomatic. Hepatic function is rarely affected by their presence [1].

Chylous cysts are rare pathological entities and relatively few cases have been reported in literature. These cysts are thought to represent benign proliferation of ectopic lymphatic that lack communication with the

normal lymphatic system [1]. They are usually associated with mesentery of intestine. But have also been reported in colon [2], retroperitoneum [3] and liver (in this case). Chylous cyst are often congenital but may be related to previous abdominal surgery, pelvic disease and trauma [4]. Mesenteric chylous cyst most commonly occur in second decade of life, but they have also been seen in first decade. Symptoms are usually poor and unspecific. Rarely, clinical presentation may be dramatic with acute abdominal pain and symptoms of intestinal obstruction [5] or simulating the rupture of aortic aneurysm [6]. The preoperative diagnosis requires all the common abdominal imaging techniques. Ultrasonography usually demonstrates, a cystic lesion, whose content may form a fluid-fluid level. A fat fluid interface on computed tomography is indicative of a chylous cyst [7]. But the correct diagnosis is usually, made in the operation theater or during histological examination [4]. Histologically, cysts are lined by a flattened endothelium with a fibrous wall; in which dilated lymphatics may be observed. Presence of cholesterol clefts further supports the pathological diagnosis of a chylous cyst [8]. Apart from being cause of an acute abdomen, these cysts may undergo rupture, hemorrhage or infection [9]. Malignant transformation is rare but not unknown [10]. Treatment of choice is complete surgical excision and is usually curative.

This patient was asymptomatic apart from the swelling in the abdomen. On examination, she had an intra-abdominal, cystic, swelling in epigastrium. Preoperative diagnosis was inconclusive even after contrast enhanced computed tomography scan diagnosis was made during laparotomy which was confirmed by histopathological examination. Complete excision of the liver cyst was possible and the multiple cysts in the mesentery which were left intact. There is no evidence of development of symptomatic large cysts or recurrence in liver after two years on follow-up.

CONCLUSION

We conclude that patient had chylous cysts at multiple places with largest from liver. Most likely they are of separate origins but we cannot say conclusively. We have planned to follow-up this patient every six months with clinical examination and ultrasonography to look for development of any large symptomatic cyst.

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Author Contributions

Mayank Baid – 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

Manoranjan Kar – 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

Sayak Roy – 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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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Carotid body tumor with thyroid involvement

Hlatshwayo B, Panicker A, Tsatsi LDR

ABSTRACT

Introduction: The carotid body tumor (paraganglioma) is a tumor that originates from paraganglion cells. The tumor is usually benign and non-functional, and can expand early but rarely metastasizes. These tumors are rare and make up 65% of all head and neck tumors. **Case Report:** Herein we discuss a case of a 66-year-old female patient who presented with a right side neck mass measuring 2x3 cm, soft and non-tender. The mass was gradually increasing in size. Blood pressure 142/182 mmHg, pulse 120 beats/min. Blood workup: Hemoglobin 10.5 g/dL (11.0–15.0 g/dL), platelets count $479 \times 10^3/\mu\text{L}$ ($150\text{--}400 \times 10^3/\mu\text{L}$), thyroid stimulating hormone 0.26 mIU/L (0.34–5.6 mIU/L), thyroxine 13.5 g/dL, Retro viral disease exposed, and liver function test was normal. **Conclusion:** Carotid body tumors are rare entity; they account for 65% of all head and neck tumors and should be considered in evaluating all lateral neck masses. They are diagnosed in the 4–5th decade of life. Treatment of choice is surgical excision, but their differential diagnosis is important for all neck masses and the definite diagnosis is needed before surgery.

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INTRODUCTION

The carotid body is a small oval structure, with an irregular surface and is pink in color measuring 3–5 mm in diameter [1]. Ten percent of tumors are bilaterally located in the bifurcations of common carotid arteries [2]. The paraganglion are tumors that originate from paraganglion cells of the neural crest and develop in the paravertebral region associated with blood vessels, skull nerves and the venous system. This tumor is usually benign and nonfunctional; it expands early and rarely metastasizes. They are classified according to its locations: carotid body, jugular vein, vagal body, orbital and laryngeal. They sometimes arise within the abdominal cavity usually in the retroperitoneal space. They may originate from little chemoreceptor organs which are found in the adventitia of common carotid arteries bifurcation. Paraganglion are rare and make up 65% of all head and neck tumors.

CASE REPORT

We are presenting a case of a 66-year-old female patient who complained of right side neck mass measuring 2x3 cm, soft and non-tender. The mass was gradually increasing in size. Clinical examination showed blood pressure 142/182 mmHg, pulse 120 beats/min. Laboratory examination showed blood workup:

hemoglobin 10.5 g/dL (11.0–15.0 g/dL), platelets count $479 \times 10^3/\mu\text{L}$ ($150\text{--}400 \times 10^3/\mu\text{L}$), thyroid stimulating hormone 0.26 mIU/L (0.34–5.6 mIU/L), thyroxin 13.5 g/dL, HIV-1,2 and liver function test were normal.

Radiological features: Characteristic of carotid body tumors is the splaying of both internal and external carotid arteries described as the lyre sign. The computed tomography (CT) scan showed soft tissue density on non-enhanced contrast as well as bright and rapid enhancement post contrast (Figures 1 and 2).

Magnetic resonance imaging (MRI): T1WI the tumor was iso to hypo tense compared to the muscle. There was a salt and pepper appearance, representing a combination of punctuate region of the hemorrhage or slow flow (salt) and flow voids (pepper). There was intense enhancement following gadolinium. T2WI the tumor was high signal intensity compared to the muscle (Figure 3). The salt and pepper appearance could be seen on T2WI (Figure 4).

Angiogram: The splaying of carotid vessels lyre sign was identified by an intense blush in the tumors with early vein filling due to arterovenous shunting. Malignant transformations were encountered in 2–36% with metastasis common in bones, lungs and livers [4].

Differential diagnoses were as follows:

- (i) Vagal schwannoma (tends to displace both vessels rather than splaying).
- (ii) Vagal neurofibroma (tends to displace both vessels rather than splaying).
- (iii) Lymph node mass (may look similar to carotid body tumor if hyper-vascular).
- (iv) Glomus vagale tumor has the same pathology as a carotid body tumor but is located more rostrally.

DISCUSSION

In 1743, Van Haller first described carotid body tumors. It is a round reddish brown to tan structure found in the adventitia at the bifurcation of common carotid

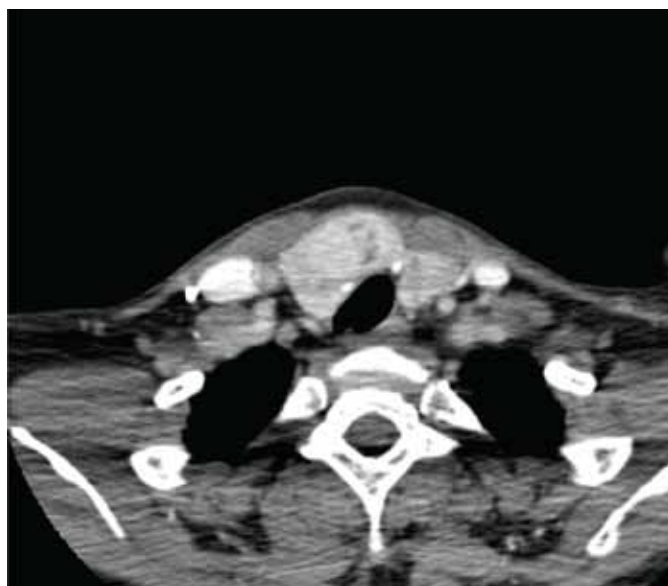


Figure 2: Axial contrast-enhanced computed tomography showing markedly enhanced mass on the right lobe of the thyroid gland compressing and displacing the airway to the contra-lateral side; the enhancement pattern is the same as the carotid body tumor.

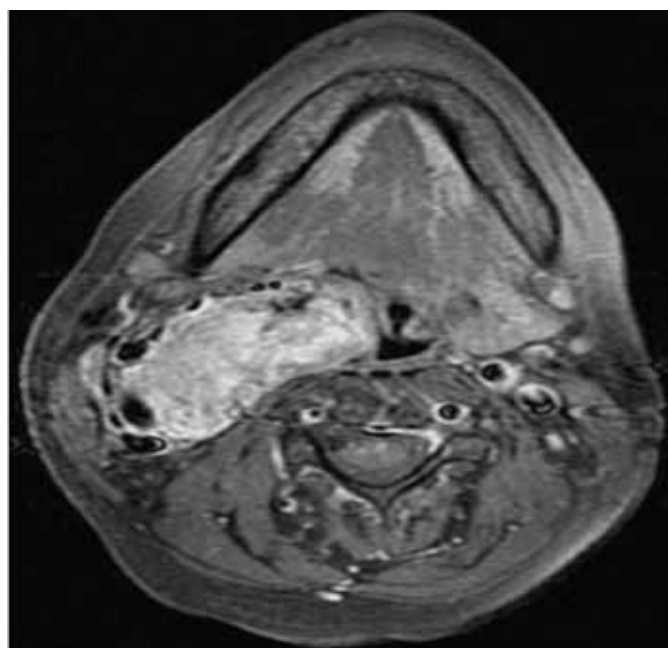


Figure 3: Axial magnetic resonance imaging T1WI post gadolinium showing high signal intensity of the mass.

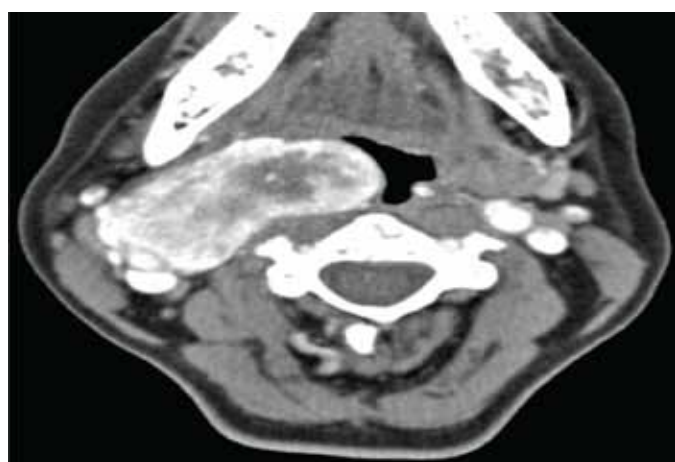


Figure 1: Axial contrast-enhanced computed tomography showing markedly enhanced mass extending from the right carotid space to the nasopharynx space and partially obstructing the nasopharynx. The mass measures 5.96x2.67 cm in size.

arteries, on the posteromedial wall of the vessels [1, 2]. They originate from paraganglion cells of neural crest [2]. They may also originate from little chemoreceptor organs [2]. Carotid body tumors are rare entities; they account for 65% of all head and neck tumors, and should be considered in evaluating every lateral neck mass [2, 3]. Malignant transformation is encountered in 2–36% cases, with metastasis common in bones, lungs, livers and regional lymph nodes [2]. They are diagnosed in the 4–5th decade

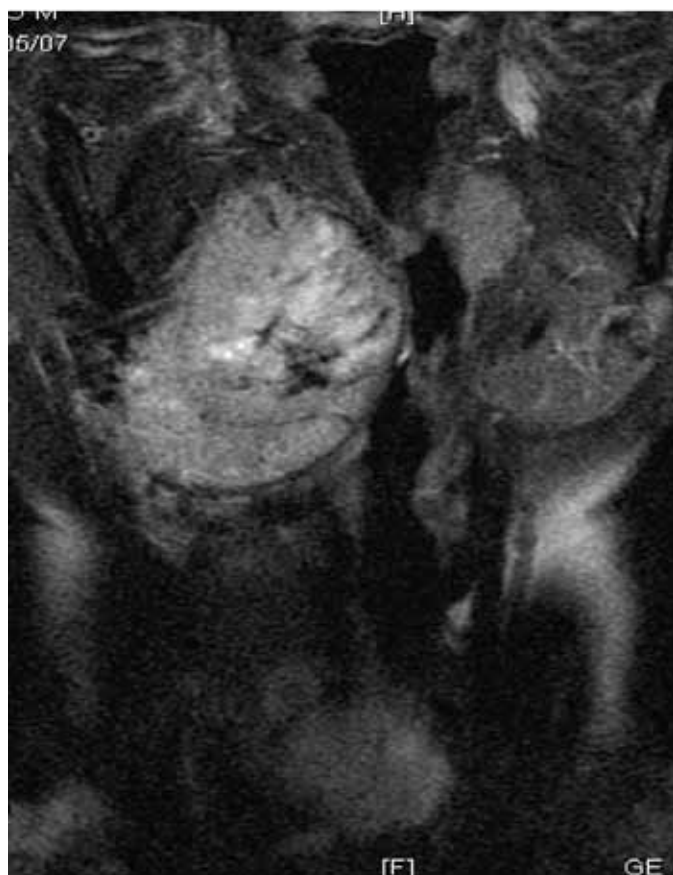


Figure 4: Coronal view shows the mass narrowing the airway and displacing the carotid artery laterally and right parathyroid space medially. It shows the areas of salt and pepper appearance within the mass. The other high signal intensity mass is seen on the right thyroid lobe.

of life and have a higher female prevalence [4]. When familial, they are usually autosomal dominant inheritance and associated with multiple endocrine neoplasia (MEN IIA, MEN IIB), tuberous sclerosis, neurofibromatosis NF1 and Von Hippel–Lindau disease [4].

The following are different types of carotid body tumors:

- Familial (10–50%) is more common in young patients.
- Sporadic
- Hyperplastic

Sporadic form is the most common type, representing approximately 85% of carotid body tumors. Hypoxia, which includes those patients living at a high altitude. Hyperplastic form is also seen in patients who has chronic obstructive pulmonary disease or cyanotic heart disease. Shamblin's classification of carotid body tumors are [4]:

Type 1: Tumors without encasement of the vessel wall, tumor size is less than 5 cm (easily resectable).

Type 2: Tumors that are attached to the walls of the arteries without encasing them (hard resection).

Type 3: Tumors that are located inside the blood

vessel with encasement of arterial wall, tumor size is larger than 5 cm, with wide carotid bifurcation (risky resection).

Tumor size is important because those greater than 5 cm in diameter have a markedly higher incidence of complications. [2].

Management: Surgical excision is the treatment of choice for carotid body tumors. However, tumor size and other patient co-morbidities determine whether embolization is necessary; vascular surgeons generally perform transcatheter embolization with plastic particles, ethanol or glue [5]. For small tumors that are less than 3 cm surgery is recommended; for larger tumors in patients who are not surgical candidates, radiations with or without radiation embolization is the treatment of choice [6].

Embolization may also have a role in palliative therapy.

Our patient was referred to a vascular surgeon for embolization and surgery but she refused surgery and opted to consult the traditional healer. We contacted her and she told us that she is feeling much better after consultation with traditional healer.

Complication: One possible complication of resection of carotid body tumors is stroke.

CONCLUSION

Carotid body tumors are rare tumors and treatment is surgical; the need to know the differential diagnosis and establish the definitive diagnosis is important. In our setting most patients are referred at later stages with large tumors. The treatment of choice for most carotid body tumors is surgical excision, but their location is in close approximation to important vessels and nerves. Tumor size is important because those greater than 5 cm in diameter have a higher incidence of complication.

Author Contributions

Hlatshwayo 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

Panicker A – Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Tsatsi LDR – Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT

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Mid-ventricular takotsubo: A case report

Sachin Kumar Amruthlal Jain, Hrishabh Modi, Timothy R Larsen,
Shukri David

ABSTRACT

Introduction: Is mid-ventricular takotsubo really a variant? Takotsubo cardiomyopathy (TCM) most commonly presents with both mid-ventricular and apical segment hypokinesia. Less frequently, the apical segment is spared and this variant has been termed mid-ventricular takotsubo. **Case Report:** We present a case of a 66-year-old female with chest pressure and abnormal electrocardiography and positive troponin. Recently her mother was placed in hospice. Echocardiogram showed severe systolic dysfunction. She underwent left heart catheterization which included left ventriculogram that showed normal coronaries and basilar and apical hyperkinesis and mid-ventricular hypokinesia suggestive of mid-ventricular takotsubo. **Conclusion:** We suggest that Takotsubo cardiomyopathy (TCM) can be thought of as a continuum in which the mid-ventricular segment is hypokinetic first and then the apical portion becomes hypokinetic later. Moreover, mid-ventricular hypokinesia is a key to a diagnosis of TCM; regardless of the presence of apical dyskinesia. The TCM merely presents in different ways and that the mid-ventricular variant is not a different disease entity from typical TCM.

Keywords: Takotsubo cardiomyopathy (TCM), Mid-ventricular Takotsubo

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INTRODUCTION

Takotsubo cardiomyopathy (TCM), also known as stress-induced cardiomyopathy, broken heart syndrome and apical ballooning syndrome, is a syndrome that mimics myocardial infarction in the absence of obstructive coronary artery disease [1]. A TCM is usually triggered by acute emotional stress in postmenopausal women and it is reversible within several weeks to as little as under 24 hours [1, 2]. Takotsubo is characterized by transient systolic dysfunction with wall motion abnormalities of both hyperkinesis and hypokinesia. Takotsubo most commonly involves both mid-ventricular and apical hypokinesia with simultaneous basal segment hyperkinesis. Mid-ventricular presentation of takotsubo, which is not frequently described in literature, involves only mid-ventricular hypokinesia, with hyperkinesis of apical and basal ventricular segments. Here we describe a case of takotsubo cardiomyopathy that involved only the mid-ventricular segment.

CASE REPORT

A 66-year-old female was presented to the emergency room complaining of chest pressure after receiving news that her mother was placed in hospice. The discomfort was central in her chest, described as pressure-like,

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and present for the past 24 hours. The chest pain was not pleuritic, exertional, or radiating. There were no alleviating or exacerbating factors. She was mildly nauseated with no vomiting. She denied diaphoresis, shortness of breath, fevers, chills, orthopnea, paroxysmal dyspnea, or lightheadedness. Her medical history was significant for hypertension, dyslipidemia and diabetes mellitus. Her family history was positive for coronary artery disease with a brother who suffered a myocardial infarction in his late fifties. She did not use tobacco.

Electrocardiography (EKG) of the patient showed normal sinus rhythm, no acute ST segment or T-wave changes. Ventricular rate was 83 beats per minute, PR interval was 162 milliseconds, and QRS duration was 72 milliseconds. Her baseline EKG showed anteroseptal Q-wave pattern with some nonspecific ST and T-wave changes. Chest X-ray was normal with no evidence of pulmonary infiltrate, pneumothorax or pleural effusion. Serial troponin assays six hours apart were 0.20, 0.21, and 0.18 (normal <0.02). Thus the diagnosis of non-ST segment elevation myocardial infarction (NSTEMI) was made. The patient received aspirin, morphine, nitroglycerin, and intravenous heparin. Subsequent echocardiogram showed left ventricle normal in size with severe global systolic dysfunction with hypokinesia of the anterior and anteroseptal walls and left ventricular ejection fraction (LVEF) of 25% (Figures 1 and 2). Mild mitral, aortic and tricuspid regurgitations were also present. The right heart chambers are normal in size with normal right ventricular function. There was no evidence of pericardial effusion.

It was decided to proceed with left heart cardiac catheterization with coronary angiogram, which showed normal coronaries, left ventriculogram showed basilar

and apical hyperkinesis and mid-ventricular hypokinesia, LVEF was 25–30%, consistent with the echocardiogram (Figures 3 and 4). She was started on appropriate medications for heart failure including an ACE inhibitor and beta blocker.

Repeated echocardiogram two months later showed LVEF of 50% without any wall motion abnormality (Figures 5 and 6).

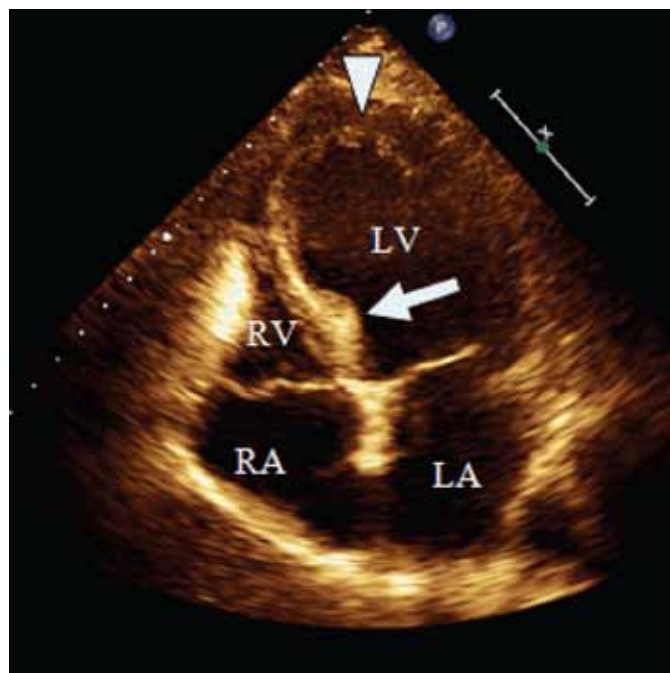


Figure 2: 2D-echocardiogram, apical 4 chamber view, during systole, demonstrating contraction of the apex (white arrow head) and basal segments (white arrow), right ventricular contraction is normal. (LV= left ventricle, LA= left atrium, RV= right ventricle, RA= right atrium).

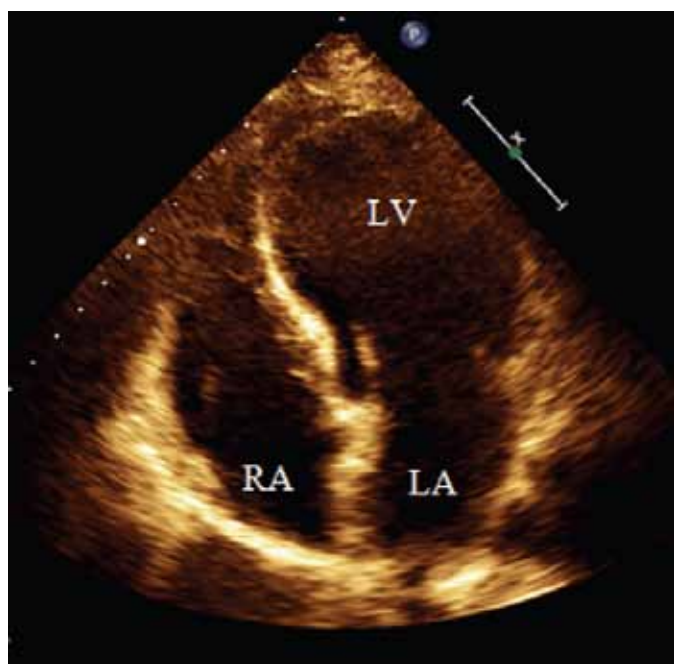


Figure 1: 2D-echocardiogram, apical 4 chamber view, during diastole, demonstrating global dilatation of the left ventricle (LV= left ventricle, LA= left atrium, RA= right atrium).

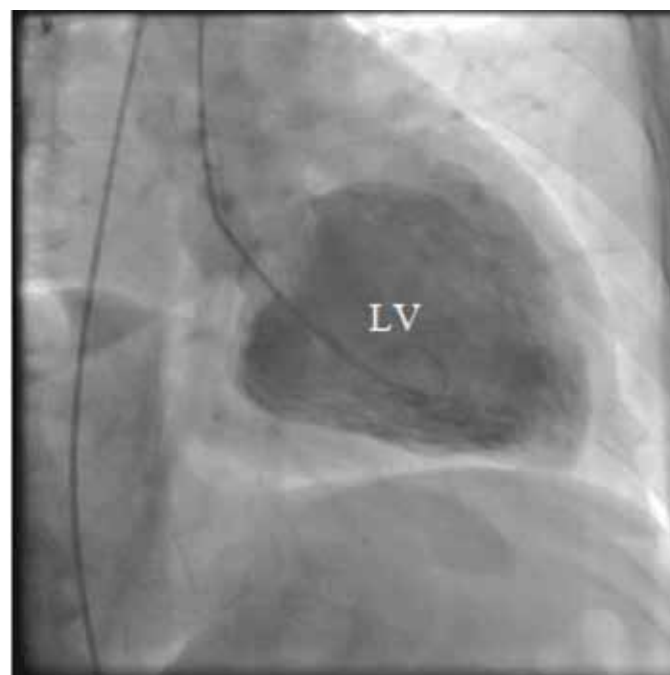


Figure 3: Left ventriculogram during diastole demonstrating left ventricular dilatation. (LV= left ventricle).

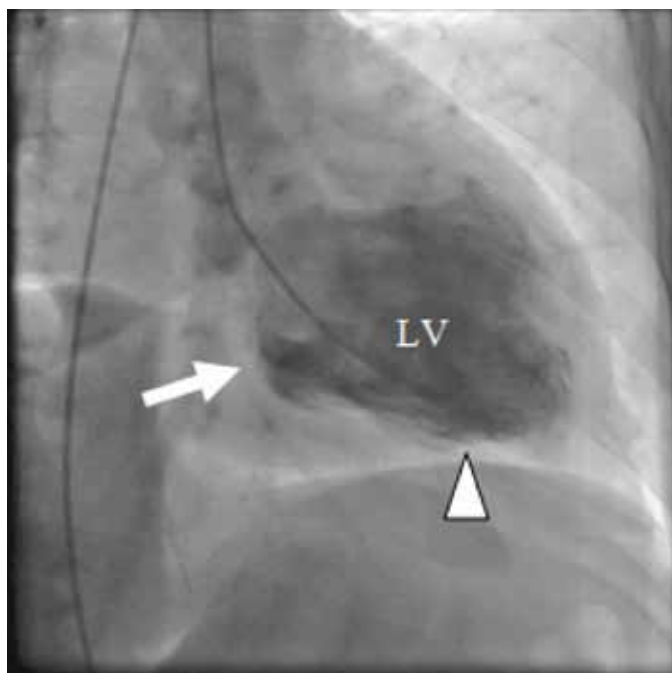


Figure 4: Left ventriculogram during systole demonstrating apical (arrow head) and basal (arrow) hyperkinesis with mid-ventricular hypokinesis. (LV= left ventricle).

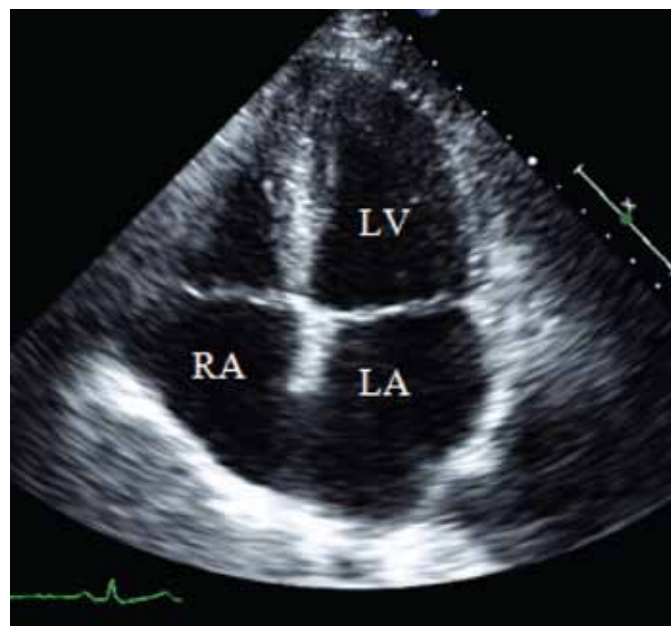


Figure 6: 2D echocardiogram, apical 4 chamber view, during systole, demonstrating improvement in left ventricular contraction, no areas of dyskinesia, ejection fraction of 50%. (LV= left ventricle, LA= left atrium, RA= right atrium).

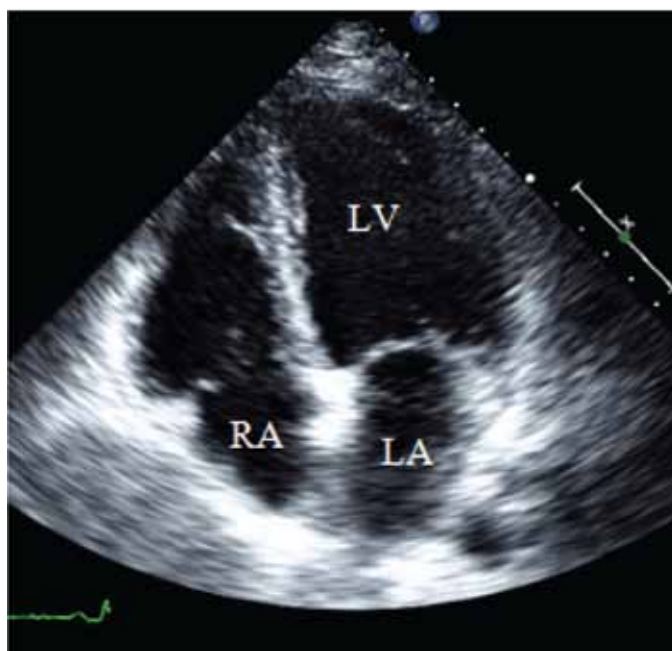


Figure 5: 2D echocardiogram, apical 4 chamber view, during end diastole, demonstrating improvement in left ventricular dilatation two months after initial presentation. (LV= left ventricle, LA= left atrium, RA= right atrium).

DISCUSSION

Takotsubo cardiomyopathy often occurs in postmenopausal women, with 82–100% of cases occurring in this demographic [3]. The triggering mechanism is very frequently attributed to emotional stress (e.g., death of close relative, receiving bad news, severe argument, and

even a surprise party [4]). It is thought that this stress leads to excessive catecholamine stimulation, metabolic disturbances, and dysfunction of microcirculation [5]. Excessive catecholamine stimulation leads to calcium overload resulting in ventricular dysfunction [6]. In another case control study of 24,701 patients with takotsubo, it was found that patients were more likely to have a history of drug abuse and cerebrovascular accidents [7]. However, a definitive mechanism is still uncertain.

It is likely takotsubo cardiomyopathy is under diagnosed, making the true incidence difficult to determine. One estimate reports takotsubo to be present in more than 1% of patients presenting with acute coronary syndromes [8]. Another report by Prasad et al. suggested that there are between 7,000 and 14,000 cases of takotsubo in the United States annually [4].

Patients typically present after an inciting emotional event, with substernal chest pain, dyspnea, syncope, and occasionally cardiogenic shock. A review from 14 studies found chest pain to be present in 67.8% of patients presenting with takotsubo [9]. The ST segment elevation, elevated cardiac biomarkers (troponins) and elevated norepinephrine levels also commonly found in 81.6%, 86.2%, and 74.3% of patients, respectively [9]. Most patients had LVEFs ranging from 20 to 49% which improved over a period of several weeks [9]. Our patient presented with pressure-like chest pain after an emotional event, had elevated troponins, and reduced ejection fraction that recovered within several weeks, following the usual course as described in literature.

Researchers at the Mayo Clinic have proposed criteria for the diagnosis of takotsubo cardiomyopathy (Table 1) [4]. All four criteria must be met to diagnose TCM. These criteria, however, require knowledge of patient's

Table 1: Mayo clinic criteria

A	Transient dyskinesia of left mid-ventricular segment with or without apical involvement
B	Absence of obstructive coronary artery disease
C	New EKG abnormalities (ST segment elevation or T wave inversion) or mild troponin elevation
D	Absence of pheochromocytoma and myocarditis

coronary anatomy, which is typically obtained via left heart catheterization and coronary angiogram. This is problematic as patients presenting to institutions lacking cardiac catheterization capability would likely be unable to utilize these criteria. This presents a therapeutic dilemma as takotsubo cardiomyopathy mimics acute coronary syndrome (including ST-segment elevation myocardial infarction) but patients would not benefit from fibrinolytic agents. An accurate diagnostic paradigm that does not require knowledge of a patient's coronary anatomy would be most helpful for making crucial decisions, such as whether or not to administer fibrinolytics.

The management of takotsubo largely consists of supportive measures as ventricular function is likely to improve over time. Initial management should follow current guidelines for treatment of acute coronary syndrome, including early administration of aspirin, EKG, cardiac biomarkers, and cardiac angiograms. The systolic dysfunction of takotsubo cardiomyopathy is treated similarly to systolic heart failure of any etiology and includes ACE inhibitors, beta blockers, and diuretics. If takotsubo occurs in the absence of heart failure, IV fluids and beta blockers may improve basilar hypercontractility [4]. Further research is needed in order to identify possible disease specific therapies, ideal therapeutic duration, and possible prophylactic measures.

In our patient, TCM involved only the mid-ventricular segment and the literature has reported that this mid-ventricular presentation of TCM occurs in up to 40% of cases [8, 10]. However, there is no difference between the clinical presentation of mid-ventricular TCM and typical TCM [10]. Differentiating the two forms of TCM requires heart catheterization and coronary angiogram. We suggest that these differences found in coronary angiogram can be thought of as a continuum in which the mid-ventricular segment is always affected and that the typical variant is an extension of cardiac involvement reaching the apical segment. In this TCM continuum, we also believe that the mid-ventricular segment becomes hypokinetic prior to the apical segment becoming

hypokinetic. The reason for this pattern may be due to different locations of cardiac adrenergic receptors, the extent of sympathetic stimulation, and the susceptibility to adrenergic stimulation [10]. Since there is no clinical difference between the two forms and by grouping them into a continuum, it can be reasoned that the mid-ventricular TCM presentation is really not a distinct disease variant.

CONCLUSION

Takotsubo cardiomyopathy is an important disease that needs to be properly recognized, diagnosed and managed. In our case, the clinical course and presentation were identical to 'mid-ventricular variant' takotsubo cardiomyopathy. The mid-ventricular presentation of takotsubo needs to be further understood because most of the results from literature (pubmed.gov searches) are on the typical presentation, but a significant number of Takotsubo cardiomyopathy cases present in this unique way. Furthermore, we proposed the idea of viewing takotsubo cardiomyopathy as a continuum in which the typical presentation involves two segments of the heart while the mid-ventricular presentation affects only one segment. However, a concrete pathophysiologic mechanism of takotsubo cardiomyopathy and the reasons for different presentations have not yet been ascertained and this remains a topic of further research.

Author Contributions

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

Hrishabh Modi – 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

Timothy Larsen – Acquisition of data, Analysis and interpretations, Critical revision of the article, Final approval of the version to be published

Shukri David – Acquisition of data, Analysis and interpretations, Critical revision of the article, Final approval of the version to be published

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The corresponding author is the guarantor of submission.

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CASE REPORT

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Coexistent of giant left atrial myxoma and severe coronary artery disease presenting with paroxysmal atrial fibrillation and syncope

Emine Gazi, Bahadır Kirilmaz, Ahmet Temiz, Hicran Yildiz Şimşek, Mustafa Saçar, Tolga Kurt

ABSTRACT

Introduction: Large intracardiac mass is a rare condition but it brings extremely high-risk of hemodynamic and embolic complications [1]. Presentation is usually heart failure, syncope or embolic events, but occasionally it may have atypical symptoms even may be asymptomatic. **Case Report:** Herein we are discussing a case of a 69-year-old male with syncope due to giant left atrial myxoma and coronary artery disease. **Conclusion:** Although atrial myxomas are the most common type of benign cardiac tumors coexistent with significant coronary artery stenosis is very rare and only a few cases available in literature. Surgery performed safely despite patient age, mass size and coronary artery disease coexistence and ultimate prognosis is good. Myxomas may be symptom free despite the coexistent of severe coronary artery disease, older age and large size of the mass until a contributing factor, such as atrial fibrillation episode as in our case, bring out the symptoms. Once the myxomas are detected before any complication surgery

performed safely despite patient age, mass size and coronary artery disease coexistence and ultimate prognosis is good.

Keywords: Giant myxoma, Syncope, Atrial fibrillation, Coronary heart disease, Echocardiography

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INTRODUCTION

Large intracardiac mass is a rare condition but it brings extremely high-risk of hemodynamic and embolic complications [1]. Presentation is usually heart failure, syncope or embolic events, but occasionally it may have atypical symptoms even may be asymptomatic. Herein, a case is reported with syncope and paroxysmal atrial fibrillation due to giant left atrial myxoma.

CASE REPORT

A 69-year-old male was admitted to our clinic suffering from palpitation just started a few hours ago with ensuing syncope. History revealed mild to moderate exercise dyspnea in last two years. In physical examination, auscultation revealed mild diastolic murmur at apex. Rhythm was atrial fibrillation in electrocardiography. Subsequently, the patient underwent transthoracic

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echocardiography, which revealed normal systolic function, significantly enlarged left atrium with a giant, ball-shaped mass in its cavity. Transesophageal echocardiography (TEE) was performed to determine the presence of additional cardiac anomaly and to confirm the diagnosis of myxoma. The TEE demonstrated the mass that is 60x39 mm in diameter, hyperechoic, immobile and attached to the fossa ovalis territory of the interatrial septum. Surgical treatment was planned and coronary angiography performed due to the older age of the patient and in order to detect vascularity of the mass. Coronary angiography demonstrated two vessel disease including left anterior descending artery and circumflex artery, and no vascularity of mass. The patient undergone mass excision and two vessel coronary by-pass graft operation (Figure 1). Pathologic assessment ascertained the diagnosis of benign atrial myxoma. Patient was discharged home with no complication in hospital.

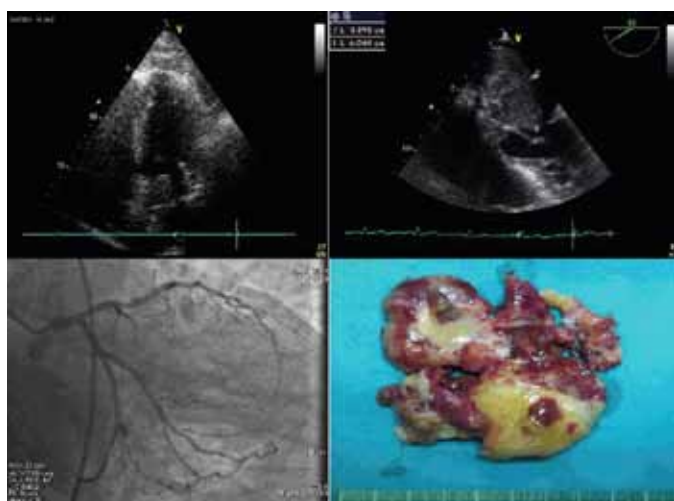


Figure 1: Left to right; apical four-chamber view in transthoracic echocardiography, transesophageal echocardiography view, coronary angiography view, postoperative material.

DISCUSSION

Atrial myxomas are the most common type of benign cardiac tumors whereas primary cardiac tumors are rare [2]. Giant intracardiac mass is rarely found and it may be diagnosed incidentally during an echocardiographic examination and usually located in left atrium and an attached to the interatrial septum, at the fossa ovalis region. Thrombus formation may superimpose on tumoral mass and, although rarely, embolic events may occur by the pieces of the mass. Clinical features are different, such as cerebral or peripheral embolic events, symptoms of heart failure due to impaired left ventricle filling or mitral regurgitation due to failure of coaptation of the mitral valve leaflets because of the mass and syncope due to obstruction of the mitral valve orifice, but occasionally

can be asymptomatic or with atypical symptoms [1] and in a case report, it is demonstrated that myxoma may cause severe pulmonary hypertension [3]. In our case, there was no embolic event although the mass was giant and despite the episode of atrial fibrillation. Syncope probably occurred due to obstruction of the mitral valve orifice and ensuing decreasing of cardiac output. Coexistence of giant myxoma and significant coronary artery stenosis is very rare and only a few cases available in literature. Our patient had two vessel coronary artery disease which required surgery. Generally, transthoracic echocardiography is the first imaging technique for detecting myxomas but TEE is extremely useful in the assessment of intracardiac structures, thrombus detection and interatrial septum evaluation so that TEE has been shown to be superior method for defining the characteristics of a mass in the left atrium [4]. Recently, three-dimensional transthoracic and transesophageal echocardiography serve a useful technic for detection and differential diagnosis of cardiac masses [5]. Surgery should be performed promptly when the possibility of embolic complications and even acute myocardial infarction can be caused by coronary emboli [6] and diagnosis has been established.

CONCLUSION

It is possible that myxomas may be symptom free despite the coexistent of severe coronary artery disease, older age and large size of the mass until a contributing factor, such as atrial fibrillation episode as in our case, bring out the symptoms. Once the myxomas are detected before any complication surgery performed safely despite patient age, mass size and coronary artery disease coexistence and ultimate prognosis is good.

Author Contributions

Emine Gazi – 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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Tolga Kurt – 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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Conflict of Interest

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CLINICAL IMAGE

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Severe pneumomediastinum after voice training

Hiroshi Ono, Naoki Nishimura, Katsunori Oikado, Kouyu Suzuki

CASE REPORT

A 76-year-old male was brought to our emergency department with a 10-day history of shortness of breath on exertion despite long-term oxygen therapy (1.0 L/min continuously). He complained of mild precordial discomfort, which was aggravated by movement. He had emphysema and a past medical history of pneumothorax, tuberculosis, gallstone surgery, and surgery for stomach cancer in 1987. He was in chronic distress and appeared emaciated. This laboratory examinations were as follows blood pressure 102/56 mmHg, heart rate 92 bpm, respiratory rate 28 bpm, body temperature 37.2°C. Oxygen saturation was 100% at a flow rate of 1.0 L/min. There were no audible rales in the chest. Blood tests showed a low concentration of serum albumin (3.1 g/dL). There were no electrocardiogram changes. Chest radiography demonstrated residual scarring from tuberculosis and mild pneumomediastinum. Computed tomography (CT) scan of chest demonstrated that the pneumomediastinum was limited mainly to the mediastinal tissues to the left of the trachea.

The day after admission we confirmed that the pneumomediastinum was improving by chest X-ray and

the patient started to receive training in swallowing and voice production by a speech therapist. He complained that after the voice training his shortness of breath had been exacerbated and he had developed acute, severe chest pain, by which time he was having difficulty in breathing and speaking. Marked subcutaneous emphysema was found in the neck and supraclavicular area. Furthermore, the breathing sounds were weakened on both sides of the chest. X-ray and CT scan of chest demonstrated severe exacerbation of the pneumomediastinum associated with subcutaneous emphysema, subpleural bullae and the formation of adhesions between the lungs and the chest wall (Figure 1). We immediately inserted a chest tube for drainage. However, the chest drainage did not work and bubbles

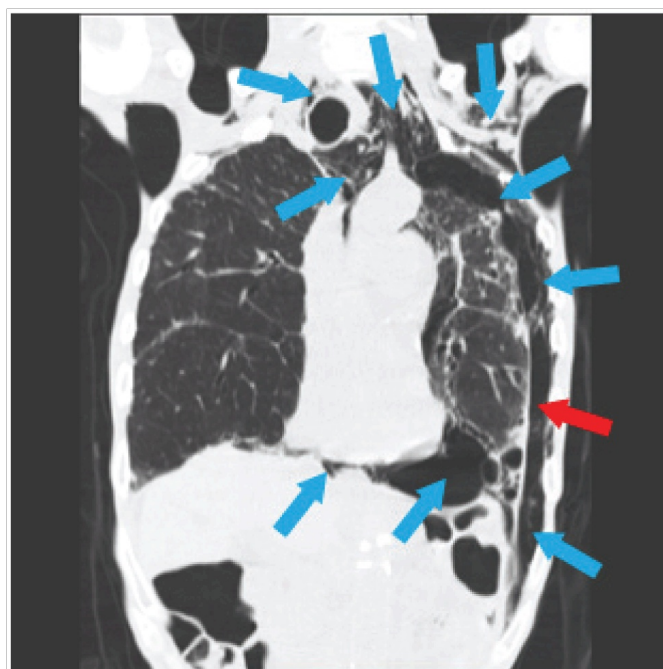


Figure 1: Computed tomography scan of chest (coronal section image). Marked pneumomediastinum and interstitial emphysema were observed (blue arrows). We inserted chest tube into the space marked by red arrow. But the chest drainage did not work. Dissection revealed that the tip of chest tube was located between chest wall and parietal pleura.

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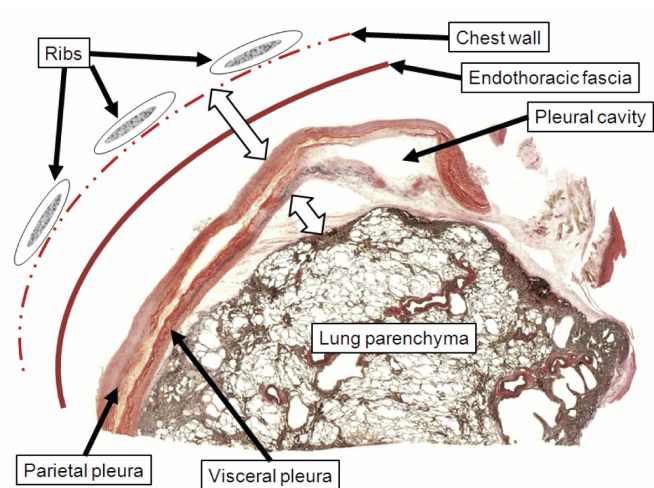


Figure 2: Histological findings of the left lower lobe of the lung and the pleura. Elastica van Gieson stain. Ribs, chest wall, and endothoracic fascia were confirmed macroscopically during dissection, and drawn by computer drawing program. Open and both ends-arrows indicate pneumomediastinum.

formation through the water valve was not noted. We were unable to relieve the patient's symptoms. The patient became weaker because the patient was put on complete bed rest and oral intake was restricted for the treatment of severe pneumomediastinum.

On day-23 after admission, the patient died of acute bacteremia due to *Serratia* infection. After obtaining consent from his family, we conducted an autopsy. On dissection, it was noted that air leakage had spread hierarchically into the clefts between the chest wall and the endothoracic fascia (at gross dissection), the endothoracic fascia and the parietal pleura (at gross dissection), the visceral pleura, and the lung itself. There were multiple bullous or cystic formations on the surface of the lungs. These findings were consistent with those of chest computed tomography. Although we filled the chest cavity with water and infiltrated the lungs with air via the mouth and nose, we were unable to identify the primary lesion causing air leakage. The diagnosis of severe pneumomediastinum was confirmed pathologically (Figure 2). The tip of chest drainage tube was located between chest wall and parietal pleura.

DISCUSSION

Pneumomediastinum is a rare disorder involving the presence of air within the mediastinum. It is classified by the site of origin of the air [1], namely the upper respiratory tract, intrathoracic airways, lung parenchyma, gastrointestinal tract, or an external source. The causes of pneumomediastinum include diabetic ketoacidosis, asthma, delivery, sports trauma, and anorexia nervosa. In our patient, the initial pneumomediastinum was considered spontaneous, but the vocal exercise and state of malnutrition unfortunately triggered a series of events resulting in severe

pneumomediastinum. This is the first case report about a pneumomediastinum exacerbated by vocal exercise in English literature. We confirmed histologically that the presence of air was limited to the lung parenchyma and did not extend to the pleural cavity.

Pneumomediastinum usually follows a benign and self-limiting course and usually occurs in younger patients [2]. The treatment required is bed rest, oxygen therapy, reassurance, and analgesia [3]. This case shows that the patient's nutritional status and past history of similar disease should be considered before vocal exercise is initiated; such exercises can impose a strain that is potentially life-threatening.

CONCLUSION

In pneumomediastinum, the presence of air is limited to the lung parenchyma and does not extend to the pleural cavity. Emaciation is one of the risk factors for this condition. Care should be taken in initiating vocal exercises in such patients.

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Author Contributions

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Conflict of Interest

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