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Surgical management of an impacted sharp metallic foreign body in esophagus

Sharma NK, Yadav VK, Pokharna P, Devgaraha S, Mathur RM

ABSTRACT

Introduction: Ingestion of foreign bodies is common and the esophagus is the most common site of impaction in the upper gastrointestinal tract. Case Series: We report two cases of impacted metallic foreign bodies in upper esophagus. The presenting symptoms were throat pain and dysphagia. Rigid and flexible esophagoscopy failed to retrieve the foreign body in both cases hence lateral cervical esophagostomy was performed for foreign body removal. Conclusion: Ingestion of foreign bodies is common. Some metallic foreign bodies frequently get impacted in the esophagus because of their large size, rigidity and pointed edges and are difficult to remove by endoscopy. Therefore, surgery is required for retrieval of such impacted, sharp, metallic foreign bodies.

Keywords: Impaction, Lateral cervical esophagostomy, Foreign body

INTRODUCTION

Foreign body ingestion is a common problem encountered in clinical practice. Most of them pass spontaneously but some are really problematic. Endoscopic removal of a foreign body is routinely done but it fails in some cases which require surgical removal of the foreign body. We present two cases of unusual foreign body ingestion, one was a sharp metallic folded tin cover and another was a sharp metallic bottle cap. As these foreign bodies could not be retrieved endoscopically, surgical removal of these foreign bodies was performed successfully. Patients are doing well on follow-up after one year.

CASE SERIES

We report two cases of impacted, sharp, metallic foreign bodies in the upper esophagus. First patient was a 35-year-old male, who was a known case of schizophrenia. He ingested a sharp metallic folded tin cover (Figure 1). The patient presented to us with throat pain and dysphagia. On examination, there was bilateral air entry, equal on both sides, with no added sound. Chest X-ray revealed foreign body in the cervical esophagus (Figure 2). The second patient was a 25-years male. He presented to us with a history of accidental ingestion of sharp metallic bottle cap (Figure 3). At the time of presentation he had throat pain and dysphagia. There was no respiratory distress. On examination chest had bilateral equal air entry without stridor.

In both cases X-ray of the cervical spine and chest (lateral view) were done which revealed foreign body with sharp edges present in the cervical esophagus (Figure 4).

Flexible and rigid esophagoscopy was done for therapeutic removal of the foreign body but failed. Foreign body was impacted in upper esophagus with surrounding inflammation and edema.
Emergency surgery was planned. Longitudinal incision was given on the left side of neck in front of sternocleidomastoid. Carotid artery and thyroid gland were retracted. Esophagus was found to be inflamed and edematous. Longitudinal incision was given on esophagus over impacted foreign body and foreign body was gently disimpacted and removed to prevent further injury to esophagus. Esophagus was repaired using 3.0 vicryl interrupted sutures in two layers, with Ryle’s tube left in situ for three weeks. Postoperative period was

Figure 1: Folded metallic tin cover removed from esophagus (case 1).

Figure 2: Bottle cap removed from esophagus (case 2).

Figure 3: Chest X-ray showing foreign body in the cervical esophagus.

Figure 4: Lateral chest X-ray showing foreign body in the upper esophagus.
uneventful and both the patients are doing well on follow-up.

**DISCUSSION**

It is estimated that up to 90% of all foreign bodies pass spontaneously from the esophagus. Endoscopic management is needed in less than 10% cases, whereas surgery is required for foreign body retrieval or management of complications in approximately 1% patients [1–3]. In both our cases, the patients required lateral cervical esophagostomy, after failed endoscopic retrieval.

Although most of the foreign bodies in the gastrointestinal tract are swallowed accidentally, other conditions such as imprisonment, mental illness, mental retardation, bulimia, alcohol consumption or drug abuse may also be involved, particularly in the Western countries [4]. The majority of foreign body ingestions occur in the pediatric population with a peak incidence between six months and six years of age [2–8]. Edentulous adults are also at greater risk for foreign body ingestion, including that of dental prosthesis [8, 9]. One of our cases had an accidental ingestion of bottle cap while another had a history of mental illness.

Esophageal objects can cause a foreign body sensation, drooling, respiratory distress due to tracheal compression, gagging, dysphonia, vomiting, and dysphagia, depending on the location and nature of the foreign body [10]. Although rare, perforating objects are potentially life threatening because they may lead to the formation of a fistula between the esophagus and the innominate artery thus causing catastrophic bleeding [11, 12]. Young children and those with mental illness may present with choking, refusal to eat, vomiting, drooling, wheezing, bloodstained saliva, or respiratory distress [5, 7, 13, 14]. Our patients had symptoms of throat pain and dysphagia.

The foreign bodies usually lie close to one of the three esophageal anatomical constrictions: the cricopharyngeal ring, the aortic arch narrowing or the esophagogastric junction [15, 16]. The most frequent lodgement site of described in literature is the cricopharyngeus muscle [11, 12].

Plain films (cervical and chest X-rays) are a very important diagnostic tool, especially in defining the location of the foreign body [10]. A barium-swallow X-ray study could be useful in cases of non-radiopaque foreign body, but due to possible barium aspiration and or irritation of the damaged esophageal mucosa, this procedure is no longer used [17–19]. Computed tomography scans can be used to confirm the presence and location of the foreign body (especially in the cases of fish bones), and to evaluate any eventual damage to the neighboring structures [18, 19].

In our cases, it was the clinical history, X-rays and endoscopy that guided us in diagnosing and defining the location of the foreign body.

The treatment of choice for esophageal foreign bodies depends on various parameters such as patient’s age, clinical condition; the type, size, shape, site, and number of foreign bodies. [10, 20] Endoscopy is the preferred method with a reported success rate of 83% [20]. Today, both rigid or flexible endoscopy, performed under general anesthesia or conscious sedation, respectively, are considered to be safe and represent effective methods in experienced hands. For the management of sharp and penetrating foreign bodies, rigid endoscopy is often the treatment of choice. Major risk during esophagoscopy maneuvers include direct instrumental wounds and perforations [16]. Surgical treatment is unavoidable in cases of irretrievable foreign body or esophageal perforation [10].

In our cases, flexible and rigid endoscopies were unsuccessful in the removal maneuvers mainly due to the size and shape of the foreign bodies. Further, the foreign bodies were impacted in esophagus therefore surgical treatment was necessary.

**CONCLUSION**

Ingestion of foreign bodies end their lodgment in the esophagus are common. Some metallic foreign bodies get frequently impacted in the esophagus and dose difficulty in endoscopic retrieval due to their large size, rigidity and pointed edges. Therefore in such cases surgery is required for retrieval of the impacted sharp metallic foreign bodies.

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**Author Contributions**

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

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

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**Guarantor**

The corresponding author is the guarantor of submission.

**Conflict of Interest**

Authors declare no conflict of interest.
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Noncompaction cardiomyopathy: A rare cardiomyopathy

Sachin Kumar Amruthlal Jain, Brijesh Patel, Timothy R Larsen, Yousif Ismail, Shukri David

ABSTRACT

Introduction: Noncompaction cardiomyopathy (NCCM) is a rare form of cardiomyopathy. The American Heart Association classified it as a genetic cardiomyopathy. The pathogenesis of this condition is attributed to the failure of trabecular myocardium to compact and underdeveloped microcoronary circulation. Case Report: We present a case of a 72-year-old African American female who presented with shortness of breath and leg swelling and was found to have severe systolic dysfunction. Stress test showed ischemia. Left heart catheterization which included left ventriculogram showed prominent trabeculations suggestive of noncompaction cardiomyopathy. Conclusion: This condition is often associated with other congenital cardiac defects and arrhythmias. When it is present alone, it is called isolated noncompaction cardiomyopathy (INCCM). The treatment protocol for this cardiomyopathy is the same as other cardiomyopathies, and includes the management of arrhythmias, heart failure and potential risk of embolism.

Keywords: Noncompaction cardiomyopathy, Echocardiography, Heart failure, Congenital heart disease

CASE REPORT

Our patient, was a 72-year-old African-American female who came to the emergency room (ER) complaining of lower extremity edema and shortness of breath for one week. She gave a history of worsening dyspnea and gradual development of orthopnea. She denied paroxysmal nocturnal dyspnea, cough, chest pain, sputum production or palpitation. She was hospitalized for congestive heart failure and lower extremity edema in the past. Her past medical history was significant for severe congestive heart failure, hypertension and chronic obstructive pulmonary disease. She was a smoker. Her
physical examination was significant for shortness of
breath and bilateral lower extremity edema (1+). Her
medication included aspirin, carvedilol, furosemide,
hydralazine, simvastatin and tiotropium. Her electro
cardiogram showed atrial flutter with 2:1 block, bilateral
atrial enlargement, Q-waves in Lead II and III, poor
R-wave progression and prolonged QT interval (499
ms). In the hospital, patient was treated for symptoms
of congestive heart failure, and underwent ablation
procedure for atrial flutter. The patient was in normal
sinus rhythm and with improved symptoms at the time
of discharge.

Image findings and analysis: The patient underwent
echocardiographic study to assess the left ventricular
function and right chamber pressure measurements,
because of her symptoms and the history of severe
congestive heart failure. The study showed enlarged left
ventricle with severe global systolic dysfunction and
depressed ejection fraction of 20%, grade I diastolic
impairment, mildly enlarged left atrium and a trace of
mitral and pulmonary regurgitation. In addition, it showed
trabeculations in the left ventricle (Figure 1). The severe
global systolic dysfunction warranted further evaluation
of her heart. Therefore, she underwent lexiscan cardiolite
nuclear stress test. The stress test showed large partially
reversible inferior and inferolateral defect consistent
with stress induced ischemia and also noted to have
dilated right ventricle. To further evaluate the ischemia,
two days later, she underwent cardiac catheterization/
left ventriculogram that showed ejection fraction of 20%,
severe global impairment in contractility and prominent
left ventricular trabeculation suggesting NCCM (Figure
2), compared to a normal left ventriculogram from another
patient (Figure 3).

**DISCUSSION**

During the first five to eight weeks of embryonic
life, spongy/trabecular myocardium starts to compact.
These trabecular fibers that are separated by ventricular
recesses link myocardium to the ventricular cavity.
The noncompaction cardiomyopathy ensues when the
myocardium fails to compact fully. The coronary
circulation also develops during this period. When the
fibers fail to compact they often have compromised
coronary microcirculation which makes them more
susceptible to ischemia and hypokinesia [2]. Many
times, cases of noncompaction cardiomyopathy also have
concurrent arrhythmias and congenital cardiac defects.
The image studies in our patient did not reveal any

![Figure 1: Transthoracic echocardiogram revealing prominent inferoapical trabeculations (arrow). LV-left ventricle, RV-right ventricle.](image1)

![Figure 2: Left ventriculogram demonstrating prominent apical trabeculae (arrows). (L-left ventricular cavity).](image2)

![Figure 3: Left ventriculogram from another patient demonstrating normal left ventricular myocardium for comparison.](image3)
congenital cardiac defect. Therefore, it is called isolated noncompaction cardiomyopathy.

In 2006, the American Heart Association included noncompaction cardiomyopathy as a genetic cardiomyopathy [1]. However, it may exist in both sporadic and familial forms. In case of familial form, it is associated with X-link inheritance linked to G4.5 gene (named Tafazzin) on Xq28 and a gene located on 11p15 [1]. The actual prevalence of noncompaction cardiomyopathy is unclear. The increasing number of case reports on this condition and various imaging options have helped to approximate the prevalence. Based on echocardiographic studies, 0.014–0.05% of the general population might be affected with this condition [1, 2]. The affected patients may develop systolic or diastolic dysfunction, or both, at the same time [3]. Echocardiography has remained as the main modality for diagnosis of this condition [2]. However, other imaging modalities can offer more information about the structure and function of the heart. The differential diagnosis of isolated noncompaction cardiomyopathy include acquired, ischemic or mixed cardiomyopathy. Currently, it is managed as other cardiomyopathies, and includes appropriate treatment for heart failure, arrhythmias and embolism [2]. Growing literature on noncompaction cardiomyopathy will enhance the understanding of pathogenesis, management and prognosis of this condition.

CONCLUSION

Noncompaction cardiomyopathy is a rare congenital heart condition that presents later in life. Patients with this condition present with heart failure, arrhythmias and/or embolism. In conclusion, patients with noncompaction cardiomyopathy are treated as other heart failure patients.

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Sachin Kumar Amruthal 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
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A young nulliparous woman with right ovarian serous borderline tumor and left ovarian micropapillary serous carcinoma

Vinita Jaggi Kumar, Dhruv Jain, Asit Mridha, Sidharth Nanda, Jyotica Jain, Rajesh Kumar Grover

ABSTRACT

Introduction: Serous borderline tumor (SBT) of low malignant potential (LMP) is heterogeneous group, neither wholly benign nor frankly malignant. It usually involves premenopausal age patients, has good prognosis and may recur even after 20 years. Decades later, transformation to low grade serous carcinoma can occur in 7%, mandating prolonged follow-up. Ten and 20 years survival in stage I is 95% and 80% respectively. Micropapillary type and invasive tumor implants warrant treatment like carcinoma and need adjuvant chemotherapy. Case Report: We present the case of a 25-year-old nullipara, married for four years who had ovarian cystectomy one year back. She reported to our institute with massive ascites and failure of anti-tubercular treatment. Diagnostic tap revealed cells suggestive of metastatic adenocarcinoma. Bilateral complex adnexal masses with ascites were seen on computed tomography (CT) scan. CA125 was 191 U/mL. Thorough staging laparotomy i.e., inspection and palpation of abdomen and pelvic organs, ascitic fluid cytology, bilateral salpingo-oophorectomy, and bilateral pelvic and aortocaval lymph nodes sampling up to inferior mesenteric artery level, along with supra and infradiaphragmatic omentectomy, and multiple peritoneal biopsies was carried out. Uterus was preserved. In our patient while right ovarian tumor was the benign type of SBT, the left ovarian tumor was the aggressive micropapillary type of SBT. Conclusion: This case emphasizes the need of subclassification of serous borderline tumors showing a broad spectrum of clinical and biological behavior from benign to low grade carcinoma. We suggest that in patients of reproductive age with infertility and adnexal masses, despite malignant cells in ascites, serous borderline tumor should be kept as a differential diagnosis and conservative surgery be offered.

Keywords: Micropapillary serous carcinoma, Typical SBT, Malignant ascites, Staging laparotomy, Low malignant potential tumors


INTRODUCTION

Serous borderline tumors (SBT) of ovary are an enigmatic group which are neither wholly benign nor frankly malignant. They occur mostly in premenopausal age group, have favorable prognosis, with 10 years survival rate of nearly 95% for stage 1 tumors. They may recur even after 20 years, hence they are labeled as low malignant potential (LMP) tumors. About half of all LMP tumors are serous tumors [1]. Mean age of borderline serous tumors is 40 years, which is two decades earlier than invasive cancer. [2] According to FIGO and WHO, stromal invasion, defined as destructive infiltrative growth, is the sole criterion used to distinguish SBT from invasive serous carcinomas [2].

Furthermore, SBT have been subclassified into benign and malignant types according to their biologic behavior. Micropapillary serous carcinoma (MPSC) is a proliferative, serous, ovarian neoplasm, characterized by a micropapillary pattern. Though it lacks destructive infiltrative growth, yet it behaves as a low-grade invasive carcinoma, hence is more aggressive than a typical ovarian serous borderline tumor. They are also associated with extravascular invasive peritoneal implants and more frequent recurrences than in typical ovarian serous borderline tumors. [3] In absence of invasive implants, MPSC does not imply unfavourable prognosis, and is much closer in its biologic behaviour to SBT than to serous carcinomas, hence it is retained in LMP category [4, 5]. Typical SBT as well as those with noninvasive implants are placed at lower end of proliferative spectrum.

We present the case of a patient who presented with massive ascites and bilateral ovarian tumors who was taken up for laparotomy and on surgical staging was diagnosed as bilateral borderline ovarian tumors in stage IC with typical non-invasive SBT in right ovary and more aggressive non-invasive micropapillary type SBT in left ovary.

CASE REPORT

On 24th June 2010, a nulliparous female, aged 25 years, married for four years, presented with progressive abdominal distension due to massive ascites (Figure 1). There was no history of fertility inducing drugs but one year back, she had undergone ovarian cystectomy in a private hospital, the details of which were not available. There was loss of appetite and weight but no history of hormone intake. She had hypomenorrhea. There were no signs and symptoms of bowel disturbance or any family history of cancer. On examination, she was emaciated with hugely protuberant abdomen due to ascites. There was mild bilateral pedal edema but no lymphadenopathy, or hepatosplennomegaly. Vague mass was palpable in suprapubic area. Per RECTAL examination showed, normal cervix displaced backwards. On bimanual pelvic examination, uterus was not felt separately but incorporated in bilateral adnexal masses, which were felt through both fornices. A large tumor about 6x5 cm, hard to feel was impacted in pouch of douglas. Rectal mucosa was smooth and mobile. Investigations revealed normal hepatic, renal and hematological profile. Preoperative serum Ca125 (marker for ovarian carcinoma) was 191 U/mL; CEA (colon carcinoma), CA19-9 (gallbladder and pancreatic carcinoma) and CA 72.4 (pancreatic and gastric carcinoma), β-hCG (germ cell tumor of ovary), AFP (hepatobiliary malignancy and germ cell tumor of ovary) were normal. Contrast computed tomography (CT) scan revealed huge ascites with bilateral, complex, solid cystic adnexal heterogeneous mass, which measured 8x8 cm on the right side and 7x8 cm on the left side. Uterus, liver, gallbladder, spleen, pancreas, both kidneys, bones and joints were normal. No significant retroperitoneal lymphadenopathy was seen. Computed tomography scan was suggestive of bilateral ovarian tumors with ascites.

First diagnostic ascitic fluid tap revealed clear yellow, serous fluid with atypical cell clusters suspicious of malignancy, while second tap was suggestive of metastatic adenocarcinoma (Figure 2). Adenosine deaminase (ADA) test which is indicative of tubercular etiology was performed in the ascitic fluid. Its value was in the normal range, thus ruling out abdominal tuberculosis.

Upper gastrointestinal tract endoscopy revealed grade 1 esophageal varices, at the lower end at 3’ O clock position. Stomach and duodenum were normal. Colonoscopy could not be done due to improper bowel preparation. Endometrial biopsy revealed secretory endometrium with no evidence of malignancy and PAP smear was normal.

On 25th July 2010, after informed consent the patient underwent staging exploratory laparotomy. Surgery included inspection and palpation of abdomen and pelvic organs, ascitic fluid cytology, bilateral salpingo-oophorectomy, bilateral pelvic and aortocaval lymph nodes sampling up to inferior mesenteric artery level, along with supracolic and infracolic omentectomy, and multiple peritoneal biopsies. Both the fallopian tubes were also removed, however, uterus was preserved.

Per operatively, six liters of clear, straw colored ascitis. On gross appearance uterus was normal, no normal, ovary seen. The ovaries were replaced by ovarian tumors. The ovarian tumors were separate but together they were impacted in the pelvis. Moreover, the left ovarian tumor was adherent to the sigmoid mesentery. The plane of cleavage was maintained. Liver, under surface of diaphragm, abdominal and pelvic peritoneum, ileal mesentery, stomach, small and large intestines and omentum were normal. Left pelvic lymph nodes and para-aortic lymph nodes were palpable and enlarged and were thoroughly dissected, while on the right side they were not enlarged and were palpated to be normal. Hence, right nodes sided were only sampled.

Histopathology revealed right ovary to be 5.4x5.0 cm, multicystic with solid papillary fronds, without any breach of capsule (Figure 3). Left ovary was 6.5x5.8 cm and similar to the right ovary (Figure 3). Right ovarian
tumor did not show any destructive stromal invasion. Mild nuclear atypia with low mitosis and psammoma bodies were seen (Figure 4). It was labeled as non-invasive serous borderline tumor, of typical type following WHO classification. While no stromal invasion was there in left ovarian tumor as well, there were foci of fused solid areas and occasional mitosis, in foci of >5 mm in length (Figure 5). It was labeled as non-invasive SBT, micropapillary type (micropapillary serous carcinoma). Omentum, appendix, peritoneal biopsies, pelvic and para-aortic lymph nodes were devoid of metastasis or implants.

Immediate postoperative recovery was uneventful. Drain was kept until stitch removal on postoperative day-12 as there was substantial amount of fluid from the drain. Postoperatively her CA 125 was 5.89 U/mL. Subsequent to discussion with our clinical oncology team, decision of adjuvant chemotherapy, was taken to give, keeping in view the biologic behavior of micropapillary type of left ovarian tumor. Postoperative recovery was uneventful but after first cycle of chemotherapy she developed parietal wall abscess. The abscess burst through the surgical stitch line and drained massive amount of pus which tested sterile on culture. Tests for tuberculosis performed on the pus were negative. Wound was debrided and left to heal by secondary intention. After this initial delay, she uneventfully completed six cycles of three weekly chemotherapy with carboplatin and paclitaxel. Currently, she is disease free for one and half years with latest report of CA 125 of 4.32 U/mL and normal CT scans of abdomen and pelvis.

DISCUSSION

Micropapillary serous carcinoma (MPSC) is a proliferative, serous ovarian neoplasm which is without destructive infiltrative growth but still behaves as a low-grade invasive carcinoma and hence considered as an in situ form of serous carcinoma [6]. In the absence of invasive implants, MPSC has favorable prognosis with
biologic behavior similar to serous borderline tumors [7, 8].

Twenty-six cases of MPSC were identified in a review of 400 cases of ovarian SBT. None of the stage 1 patients had recurrence, but in higher stages 50% died of the disease. Twenty-four (92%) of MPSC cases were SBT associated. It was concluded that micropapillary type of SBT may progress to invasive carcinoma in some instances [8]. Drescher et al. studied significance of DNA content and nuclear morphology as prognostic factors to predict aggressiveness of borderline ovarian tumors and to guide adjuvant chemotherapy. [9]

Clinical presentation of LMP tumors is with abdominal distension, pain, and pelvic mass which is same as that of malignant tumors. Young patients have associated infertility or pregnancy. History of infertility increases the risk of LMP tumors with odds ratio (OR) of 1.9, while use of infertility drugs has OR of 4. [10] In patients with serous borderline tumors, after ovarian cystectomy, 8–15% patients have recurrence in the ipsilateral or contra lateral ovarian tissue. Bilaterality or synchronous and metachronous tumors are common and seen in about 40% patients [10].

In our patient there was bilateral involvement of ovaries; the tumors in right and left ovary were separate. While right ovary had benign type of typical borderline serous tumor, the left ovary had noninvasive borderline serous tumor of micropapillary type, which as the literature suggests is a more aggressive subtype. Preoperatively ascitic fluid cytology did have malignant cells, however there were no implants detected in the peritoneal lining.

About 80% of low malignant potential (LMP) tumors are diagnosed in stage 1. Some have extravarian tumor implants on peritoneal surface of pelvis or abdomen or both. Though some consider these implants to represent metastasis, yet others consider them to represent synchronous extravarian proliferations arising from surface coelomic epithelium as a result of multifocal field change. Spontaneous regression of extravarian foci has been reported following resection of ovarian primary tumor. [11]

These extravarian tumor implants could be invasive or non-invasive. Serous borderline tumors with invasive implants and micropapillary type of ovarian serous borderline tumors are aggressive subgroups and are classified as carcinomas. They are associated with poor prognosis and are treated with adjuvant chemotherapy. [4, 11]

The LMP or borderline neoplasms have 10 years survival rate of 95% for stage 1 patients. [2] Overall survival is 90–95% at 5 years and 80% at 20 years. In another study, disease-specific survival was >95% for patients with lowstage (stage I) tumors and approximately 65% for patients with highstage (stage II–IV) tumors.

Thus, although both the typical type of borderline serous tumors and micropapillary type of serous borderline tumors appear to be noninvasive histologically, the former tumor is benign, while the latter behaves like a low grade carcinoma. Poor prognosis for MPSC prompts clinicians to use adjunctive chemotherapy [4]. According to FIGO and WHO, stromal invasion defined as destructive infiltrative growth is the sole criterion used to distinguish SBT from invasive serous carcinomas. An aggressive subgroup of proliferative serous lesions with micropapillary projections are named as micropapillary serous carcinoma (MPSC). These tumors and SBT with invasive implants may be associated with malignant behavior [5].

Serous borderline tumors have a very favorable prognosis, but complete surgical staging and prolonged follow-up are advised because pelvic recurrence and occasionally transformation to invasive carcinoma may occur, albeit over delayed period of time. Designation of benign subgroup of SBTs as “atypical proliferative tumors” is not recommended because it discourages complete surgical staging and follow-up. Advanced stage tumors with noninvasive implants are common and behave in a benign fashion, and can be safely treated conservatively.

Although histologically ovarian MPSC lack stromal invasion and therefore are qualified by FIGO and WHO guidelines for inclusion in borderline category, clinically they behave more aggressively than typical serous borderline tumors. Hence, the SBTs associated with invasive implants and SBT-MPSC could behave as low grade carcinoma and their management should be seen in this light.

Various subtypes of serous borderline tumors are summarized in Table 1. [12]
CONCLUSION

We suggest, that in cases in reproductive age group with infertility and adnexal masses, despite presence of malignant cells in ascites we should keep serous borderline tumors in mind and offer conservative surgery. Our case suggests that MPSC and typical type of SBT represent two ends of a broad spectrum of clinical and biologic behaviour seen in low malignant potential ovarian tumors. The MPSC are associated and probably arise from SBT and may account for few cases of SBT which progress to invasive carcinoma. There is the need of subclassification of serous borderline tumors.

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Sigmoid carcinoma: A rare presentation and diagnostic challenge

Bassem Amr, Komal Munir, Natasha Santana-Vaz, Venkateswarlu Velineni

ABSTRACT

Introduction: Adenocarcinoma represents less than 2% of all bladder carcinomas. Metastatic adenocarcinoma is the most common form and usually represents a direct extension from a primary lesion either in the colon, prostate or the female genital organs. Other categories include primary vesical or urachal.

Case Report: We present a case of 79-year-old female presented with microscopic haematuria, frequency, urgency and weight loss. An adenocarcinoma of the urinary bladder of intestinal origin was founded.

Conclusion: Primary adenocarcinoma of the urinary bladder is uncommon neoplasm and the reported incidence of the primary adenocarcinoma is 0.5–2%. It represents a diagnostic challenge raising the inquiry about the site of the lesion. Three major classes were identified: primary vesical adenocarcinoma, urachal adenocarcinoma and extravesical adenocarcinoma involving the bladder. The metastatic adenocarcinoma is the most common category representing a direct extension from a nearby organ or metastatic spread. The most common treatment for urinary bladder adenocarcinoma is surgery. Generally speaking, treatment options include: surgery, radiotherapy, chemotherapy and immunotherapy. Metastatic adenocarcinoma to the urinary bladder especially of colonic origin is of particular interest and need to be ruled out before making a diagnosis of primary adenocarcinoma of urinary bladder.

Keywords: Bowel cancer, Adenocarcinoma, Bladder tumor

INTRODUCTION

Adenocarcinoma represents less than 2% of all bladder carcinomas [1]. Primary adenocarcinoma is an uncommon malignant neoplasm and is a source of a diagnostic challenge. It is less frequently encountered in areas where bilharziasis is endemic whereas the squamous cell carcinoma is more prevalent. This incidence ranges between 5–11.4%. Chronic vesical irritation and infection are predisposing factors for metaplastic changes of the urothelium [2]. Based on the original tumor site, adenocarcinoma of the bladder could be classified into three categories: primary, urachal and metastatic. Primary vesical adenocarcinoma needs to be distinguished from the more common extravesical metastatic adenocarcinoma (direct spread, lymphatic, and hematogenous). The principal primary organs to be considered include prostate, colon, female genital tract, appendix, stomach and breast. Clinical association with bladder exstrophy and schistosomiasis has been well documented. Clinical assessment, imaging, histologic, and immunohistochemical correlation should be done.
while investigating this diagnosis due to histologic and immunohistochemical overlapping between primary bladder adenocarcinoma and metastatic adenocarcinoma [3]. Prognosis and therapeutic options for primary versus metastatic adenocarcinoma vary widely.

**CASE REPORT**

A 79-year-old female was presented to her general practitioner in January 2012 with urinary symptoms in the form of frequency, urgency and passing dark urine. She had also noted recent weight loss of 3 kg. Urine dipstick was positive for leukocytes, protein and blood. Antibiotic was prescribed by her doctor for urinary tract infection. The symptoms recurred once the course of treatment was completed; for which she was given three separate courses of antibiotics as her mid-stream urine samples showed *E. coli*. Her ultrasound scan of the urinary tract showed a simple cyst on the right kidney and incidental finding of a bulky uterus; therefore pelvic scan was undertaken. A retroverted bulky fibroid uterus measuring 6.5x2.9x4.6 cm was revealed. A heterogenous myometrium demonstrating echogenic foci throughout anteriorly was founded and a well-defined echogenic area within the left adnexa measuring 2.5 cm with small amount of free fluid was seen on the scan. She was referred to the hematuria clinic on April 2012, where a flexible cystoscopy was performed under local anesthesia. There was an inverted granuloma present at the trigone and an area of inflammation on the right-hand side of the posterior wall. Computed tomography (CT) scan of the abdomen and pelvis showed marked sigmoid colon thickening with associated diverticulosis suggestive of complicated sigmoid diverticulitis with a localized perforation. Also a focal urinary bladder wall thickening was likely secondary to the colonic abnormality with no definitive evidence of a colovesical fistula on this scan (Figure 1). She was admitted to our hospital for cystoscopy and biopsy. Examination under anesthesia revealed a 5-cm pelvic mass extending into the left iliac fossa. Cystoscopy revealed a lesion in the trigone (Figure 2) which was biopsied. Histopathological findings of the cystoscopic biopsy revealed moderately differentiated adenocarcinoma of an intestinal appearance infiltrating bladder mucosa. Her case was discussed at the urology and colorectal multidisciplinary team (MDT) meetings. A staging CT scan showed a sigmoid colon tumor, possibly involving the roof of the bladder with lymphadenopathy and no evidence of distant metastatic disease. Her blood investigations were within normal limits before she was scheduled for anterior resection, partial cystectomy and total abdominal hysterectomy. She was admitted to intensive care unit after a long procedure where she received noradrenaline support for 48 hours and transfused two units of blood before discharging her back to the ward. She made an uneventful recovery before being discharged with a long-term catheter and an outpatient appointment for cystogram and chemotherapy. Patient was followed-up on July 2012 after satisfactory cystogram and again on October 2012 by consultant surgeon. She was doing well and managed to maintain her weight. The final histology confirmed a moderately differentiated adenocarcinoma of colonic origin (pT4 pN0) invading the detrusor muscle of the bladder. The myometrium, both adnexa and cervix were unremarkable.

Figure 1: Thickened wall of sigmoid colon and area of possible perforation with gas bubbles.

Figure 2: Cystoscopic picture demonstrating a lesion at the trigone.
DISCUSSION

Adenocarcinoma of the urinary bladder is uncommon, representing less than 2% of malignant neoplasms at this site. Adenocarcinoma can arise anywhere in the urinary bladder. Nevertheless, in most cases they involve the trigone and posterior bladder wall [1]. About two-thirds of these tumors present as solitary, discrete lesions, unlike the ‘usual’ urothelial carcinomas, which tend to be multifocal [2]. These tumors are more common in men with a male-female ratio of 3:1. It is usually occurring in the fifth to seventh decades of life. Direct extension of adenocarcinoma from adjacent organs such as the prostate, colon or ovary is more common than primary adenocarcinoma of the urethra or urinary bladder [3]. Metastatic lesions to urinary bladder often infiltrate the wall of the bladder rather than ulcerating the mucosa. In some cases, the bladder is the only genitourinary organ involved in metastasis, mainly from the breast. Most of these patients with urinary bladder metastases are asymptomatic. Symptoms produced by these metastases occur only when the bladder mucosa is involved [4]. These tumors show varied histological pictures and degrees of differentiation. Primary adenocarcinoma of the urinary bladder exhibit several histological subtypes: glandular not otherwise specified, mucinous (colloid), colonic (enteric) type, signet ring cell, clear cell (mesonephric) type and mixed type [5, 6]. The majority of the primary adenocarcinoma is of the enteric type and it is morphologically indistinguishable from metastatic adenocarcinoma of colonic origin when evaluated by the cytological, histopathological, histochemical and immunological techniques. However certain features, including the location of the tumor, growth pattern, and clinical history, have been suggested as useful aides for differentiating metastatic colonic adenocarcinoma from primary enteric-type adenocarcinoma of urinary bladder [7, 8]. It is important to discriminate primary adenocarcinoma of the urinary tract from metastatic colonic adenocarcinoma because of their differing treatment protocols as well as its morphological similarity. Clinical and radiological correlation and ultimately histological confirmation are strongly recommended for confirmation of the diagnosis especially in cases of unknown primary [9]. Treatment options include: surgery, radiation therapy, immunotherapy and chemotherapy. For small bladder tumors, partial/segmental cystectomy could be performed. However, for large size tumors radical cystectomy is recommended [10].

CONCLUSION

The diagnosis of urinary bladder adenocarcinoma raises the challenging question of whether the lesion is primary vesical, urachal in origin or metastatic from a distant or an adjacent organ.

Author Contributions
Bassem Amr – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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The corresponding author is the guarantor of submission.

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REFERENCES


ABSTRACT

Introduction: Congenital hypothyroidism is one of the most common endocrinological disorders in newborns. The slow development of obvious clinical symptoms, coupled with the importance of early treatment led to the implementation of widespread newborn screening for this condition. Case Report: This paper presents a case of congenital hypothyroidism that was born at home and was not screened for hypothyroidism. A 17-year-old female patient complained of short stature, mental retardation, delayed puberty and chronic constipation as a result of neglected hypothyroidism. Conclusion: This case discusses the importance of neonatal screening programs for early detection of congenital hypothyroidism cases that are unfortunately still being neglected in some areas of developing countries. It also shows the value of early and adequate replacement therapy for the detected cases as this could protect them against irreversible growth retardation and mental retardation. This is because any diagnostic delay, inadequate treatment or even poor compliance to treatment is always associated with irreversible damage as those manifested in this case.

Keywords: Hypothyroidism, Short stature, Neonatal screening

INTRODUCTION

Congenital hypothyroidism is defined as thyroid hormone deficiency present at birth. Problems with thyroid gland development (dysgenesis) and disorders of thyroid hormone biosynthesis (dyshormonogenesis) are the most common causes of congenital hypothyroidism. Newborn screening for hypothyroidism is not done in many third world countries. Only an estimated one-third of the worldwide birth population is screened. It is, therefore, important that clinicians could be able to recognize and treat the disorder [1]. Also health education with early signs and symptoms of the condition is very important especially in areas where neonatal screening against hypothyroidism is not performed.

This case report describes a neglected case of hypothyroidism that was left untreated for 17 years. The reasons behind this negligence are mainly that the patient was not screened against hypothyroidism at birth. In addition, her parents were not aware of the signs of the condition.

CASE REPORT

A 17-year-old patient was brought by her mother to Alexandria University Hospital, complaining of delayed puberty, short stature, mental retardation and chronic constipation. She was born at home to a family of low socio-economic conditions and was not screened for hypothyroidism at birth. She reported that the patient
was hypoactive and not interested in the surroundings since early childhood, but the case was neglected and the mother never asked for medical advice before.

Upon physical examination the patient was obese. She had very short stature with a height of 129 cm (<3rd percentile). She had large head, short neck, coarse facial features with myxedematous appearance, macroGLOSSia and depressed nasal bridge (Figure 1). Her skin was cold and dry. The patient’s vital signs were normal.

Abdominal examination revealed generalized abdominal distention (pot belly) with umbilical hernia, otherwise abdominal ultrasound revealed normal abdomen. Neurological examination showed generalized hypotonia and hyporeflexia with positive Woltman sign (delayed relaxation phase of elicited deep tendon reflexes). The patient was mentally retarded and had low IQ level. The computed tomography (CT) scan of brain was normal. Pulmonary and cardiac examinations were within normal limits.

Laboratory examination showed high TSH levels. TSH measured 536 IU/mL, with low levels of free T4 and T3 (T4 was 32 ng/mL, T3 was 0.6 ng/mL). Accordingly, diagnosis of primary hypothyroidism was established and replacement therapy with levothyroxine was started to normalize the levels of TSH. Chronic constipation was treated with laxatives.

After two years of treatment, the condition improved slightly where the patient became more oriented and interested in the surroundings, constipation was treated and artificial menstruation was induced at the age of 19 years.

DISCUSSION

The incidence of congenital hypothyroidism based on neonatal laboratory screening is greater than the incidence based on clinical signs and symptoms. The incidence of the condition depending on clinical diagnosis has been found to vary between 1:5800 and 1:6900, whereas based on neonatal screening the incidence is between 1:2900 and 1:3600. This discrepancy may be due to the fact that some children with the disorder go undetected when general laboratory screening is not utilized. On the other hand, neonatal laboratory screening will detect some children with minor disturbance of thyroid function which are clinically insignificant [2].

Thyroid hormones are important for normal growth hormone and IGF functions. Hypothyroidism is associated with a reduction in the pulsatile growth hormone secretion and in growth hormone response to stimulatory tests. In hypothyroidism, serum levels of IGF-I and IGFBP-3 fall dramatically. In a study of 12 patients with hypothyroidism, it was found that long-term replacement therapy in children with hypothyroidism is associated with a physiological increase in IGF-1 and IGFBP-3 [3]. However, studies have also concluded that catch-up growth in hypothyroidism may be incomplete if treatment has been started shortly before or during puberty [4]. So despite treatment, prolonged hypothyroidism may result in compromised adult height in some patients. The contributing factors to this height deficit may include the duration of hypothyroidism, the height deficit at the time of the diagnosis, etiological differences and the diminished potential for catch-up growth in late-diagnosed hypothyroidism [5].

A follow-up study of 30 patients with congenital hypothyroidism concluded that early detection by neonatal screening and treatment enables normal prepubertal and pubertal growth and the achievement of normal adult height, following normal puberty. Adult height in congenital hypothyroidism is significantly correlated with parental height and the mean L-T4 daily dose administered over the first six months of treatment [6].
Another complication of neglected or inadequately treated hypothyroidism is mental retardation. Normal brain development depends on delivery of adequate thyroid hormone for the first 2–3 years of life. Low thyroid levels during this time may result in irreversible damage, whereas hypothyroidism developed after the age of 3 years, it leads to reversible effects if it is early and adequately treated [1].

The New England Congenital Hypothyroidism Collaborative reported that a subgroup of 18 infants who had low serum T4 levels (average T4 8.6 µg/dL) and low L-thyroxine dosing (< 5 µg/kg/day) with a history of poor compliance in the first three years of life, had a mean IQ of 87. The larger, adequately treated group, with a serum T4 in the target range (average T4 11.2 µg/dL), had an IQ score of 105. This study concluded that T4 values in the lower part of the normal range are probably incompatible with maximal intellectual development and every effort should be exerted to maintain serum T4 levels in the upper half of the normal range during the first year of life [7].

It was found that diagnostic delay of hypothyroidism was associated with a steady decline in mean IQ (Wechsler intelligence scale) in a retrospective study of 141 hypothyroid children where the mean IQ was 79.5 for children with congenital hypothyroidity, but was normal in children diagnosed before the age of 6 weeks [8]. And many other studies concluded that IQ levels of early treated congenital hypothyroidism patients detected by neonatal screening programs can be within normal range and they can have normal school attainments [9].

CONCLUSION

In conclusion, growth retardation and mental retardation of children due to hypothyroidism can be totally prevented by three things. First to follow a strict neonatal screening program for hypothyroidism that is unfortunately still being neglected in some areas of the developing countries. Therefore, health education to parents in these countries about the main signs and symptoms of hypothyroidism is very important. Second is the adequate and very early L-thyroxin replacement therapy. Third is very careful follow-up of patients and adjustment of the L-thyroxin dose to maintain serum T4 levels in the upper half of the normal range during the first year of life. This is because any diagnostic delay, inadequate treatment or even poor compliance to treatment is always associated with irreversible damage as those manifested in this case.

Drafting the article, 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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Authors declare no conflict of interest.

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Author Contributions
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Fallaciously elevated glucose level by handheld glucometer in a patient with chronic kidney disease and hypoglycemic encephalopathy

Praveen Pratap Jadhav, Meera Praveen Jadhav

ABSTRACT

Introduction: The handheld glucometers are commonly used for glucose estimation in acutely ill patients, especially in those with neurological deficit. However, in a few conditions like chronic renal failure, it can give fallacious readings. This has serious implications in management of these patients, which can lead to misdiagnosis of the underlying hypoglycemia. Case Report: Here we present a case of a patient with hypoglycemia who had a fallacious normal glucose level when checked by a handheld glucometer (i.e., elevated on glucometer). The underlying pathology giving rise to this fallacy was presence of chronic kidney disease with high uric acid and low hematocrit. Conclusion: Treating physicians should be aware of such possibilities and take suitable steps for appropriate diagnosis of potentially life-threatening hypoglycemia.

Keywords: Hypoglycemia, Glucometer, Chronic kidney disease, Uric acid

INTRODUCTION

Precise diagnosis of acute critically ill unconscious or mentally altered patient is very difficult. Care givers rely on history, examination, imaging and laboratory data for exact diagnosis. However, sometimes laboratory data could be misleading and can lead to hazardous consequences. One such condition, where laboratory results can mislead, is the diagnosis of hypoglycemia in patients with chronic renal failure. Here we present a case where widely used handheld glucometers provided a falsely high capillary blood sugar reading causing diagnostic problems.

CASE REPORT

A 54-year-old lady was presented with a short history, beginning with altered sensorium and drowsiness and leading to unconsciousness in one hour. She was a diabetic with history of chronic kidney disease on regular treatment. During the initial stages of symptoms, the relatives tried to feed her with oral glucose powder, presuming this to be a hypoglycemic attack. However, the patient was unable to ingest it. The blood sugar level checked on Accu-Chek glucometer (Roche Diagnostics) was 187 mg/dL. Since she did not respond to treatment
in a few minutes, she was shifted to the hospital for further treatment. She was brought to the hospital deeply unconscious. Her vital parameters were normal, heart rate 100 per minute, blood pressure 110/70 mmHg and respiratory rate 18 per minute. She was not responding to deep pain, had flaccid paralysis of all four limbs, pupils were constricted but reacting to light, deep tendon reflexes were depressed and plantar reflexes could not be elicited. Respiratory, cardiac and alimentary system examinations were normal. Her capillary sugar checked on a different Accu-Chek glucometer (Roche Diagnostics) was 167 mg/dL. Differential diagnoses of brain stem cerebrovascular accident, metabolic encephalopathy and uremic encephalopathy were considered. Her blood sample was sent for investigations and her magnetic resonance imaging (MRI) scan of the brain was done. The MRI scan revealed bilateral symmetrical hyper intense lesions in the internal capsule, corona radiata and centrum semiovale, with reduced apparent diffusion coefficient. An hour later her blood reports showed hemoglobin 8.2 g/dL, normal blood counts, hematocrit 20%, creatinine 6 mg/dL, blood urea nitrogen 96 mg/dL, sodium 132 mEq/L, potassium 5.2 mEq/L, uric acid 9.2 mg/dL, normal hepatic enzymes and ammonia levels and plasma glucose 24 mg/dL. She was infused with 25% dextrose and then with 500 mL of 10% glucose infusion. She responded dramatically in a few minutes. She became conscious, started moving all the four limbs with complete recovery of all neurological deficit. Temporal event of positive neurological improvement after intravenous glucose suggested the diagnosis of hypoglycemic encephalopathy. Patient was treated with initial intravenous and later oral glucose till plasma levels of sugar were stabilized. She was discharged asymptomatic within 24 hours.

**DISCUSSION**

Hypoglycemia is a common emergency situation in intensive care units. It is life-threatening, yet easily treatable. The most common way to diagnose this condition is with handheld glucometers. Some conditions, however, have the potential to interfere with glucose measurements by handheld glucometers and provide erroneous readings of blood glucose. This can lead to misdiagnosis and wrong treatment in patients with hypoglycemia. Various factors are attributed to altered glucose measurement on the strips. Some common reasons are given in Table 1.

Though most of these factors cause a minor variation in estimated glucose, sometimes, especially with extreme values of these factors, fallacious readings could be obtained and these could be hazardous. In this patient, though it was obvious that patient had hypoglycemia, it was not detected by glucometer testing.

Glucometers use glucose oxidase (GOD) or glucose dehydrogenase (GDH) enzymes to detect the presence of glucose in blood. In these reactions, glucose is metabolized by GOD or GDH to hydrogen peroxide or reduced nicotinamide adenine dinucleotide, respectively. The amount of hydrogen peroxide or rNAD can then be measured by oxidized dye color change or by electrochemical reactions to calculate the amount of glucose present [1]. The glucose oxidase method makes use of one or both of the following reactions:

<table>
<thead>
<tr>
<th>Factors</th>
<th>Variation</th>
<th>Mechanism</th>
</tr>
</thead>
<tbody>
<tr>
<td>Strip factor</td>
<td>Strip-strip variation</td>
<td>Varying storage conditions. Variation of enzyme quantity in different strips.</td>
</tr>
<tr>
<td>Physical factors</td>
<td>Altitude</td>
<td>Increased sensitivity of GOD biosensor strip to oxygen concentration.</td>
</tr>
<tr>
<td></td>
<td>Temperature</td>
<td>Unpredictable response. Brand specific errors</td>
</tr>
<tr>
<td>Patient factors</td>
<td>Meter-strip miscoding</td>
<td>Altered relation between electrical signal produced by strip and reported blood glucose.</td>
</tr>
<tr>
<td></td>
<td>Hematocrit</td>
<td>Glucose in RBCs is in equilibrium with plasma glucose, but at lower levels. Extreme hematocrit alters the equilibrium.</td>
</tr>
<tr>
<td></td>
<td>Triglycerides</td>
<td>Takes up volume, thereby reducing the amount of glucose in capillary volume.</td>
</tr>
<tr>
<td></td>
<td>Uric acid</td>
<td>Uric acid is oxidized by the electrodes in the tip of the strip.</td>
</tr>
<tr>
<td>Pharmacological factors</td>
<td>L dopa, ascorbic acid, icodextrin in dialysate solutions</td>
<td>These drugs interact with the electrodes at the tip of the strip to give high false values.</td>
</tr>
</tbody>
</table>

GOD – glucose oxidase, RBC – red blood cells
Many conditions can interfere with these chemical reactions happening at the tip of the glucose measuring strip. High levels of uric acid, which is a reducing agent, may result in falsely high glucose readings [1]. Uric acid lowers the value for glucose as determined by the GOD-Perid method. Bleaching of the final color is the source of such interference in this reaction [2]. Reducing agents other than glucose, such as metabolites of icodextrin, may also be detected by these methods. This is a chemical used in the dialysate for patients with end stage renal failure on long-term peritoneal dialysis, and which is partly absorbed in the circulation. High or low hematocrit values can also lead to falsely abnormal glucose readings. Low hematocrit value has also been shown to increase glucose estimations on handheld glucometer [3]. Both, low hematocrit and high uric acid, are common in critically ill patients as well as in patients with renal failure. Hence, it can be seen that patients with chronic kidney disease may have multiple reasons to get falsely high glucose readings when handheld glucometers are used. This patient, though never dialyzed, had high uric acid and low hematocrit values, which could have led to falsely high glucose levels on the reagent strip. Since uric acid is a reducing agent and though it is implicated as an agent to give falsely elevated glucose in handheld glucometer, we did not come across any reference or case reports describing the same. The MRI scan of the patient was also highly suggestive of hypoglycemic encephalopathy [4].

**CONCLUSION**

Hence, we suggest that in patients with chronic kidney disease, especially due to diabetes, effort should be made to diagnose and treat hypoglycemia accurately when they present with altered mental status. This may involve not over-relying on handheld glucometer, using laboratory method to estimate blood glucose for confirmation or empirically treating these patients with intravenous dextrose without laboratory evidence of hypoglycemia.

**Author Contributions**

Praveen Pratap Jadhav – 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

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

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CASE REPORT OPEN ACCESS

Dual causes of multiple myeloma

Zaw Min, Zipporah Krishnasami, William J Cook, J Martin Rodriguez

ABSTRACT

Abstract: Patients infected with human immunodeficiency virus (HIV) infection have been living longer secondary to dramatic improvements in their immune status because of highly active antiretroviral therapy (HAART). Consequently, there is an increasing incidence of non-AIDS defining malignancies and chronic diseases in HIV-infected individuals. The hepatitis C virus (HCV) co-infection is highly prevalent in patients with HIV infection. Case Report: We report a patient with HIV and HCV co-infection who presented with multiple myeloma, and explore literature looking for a plausible causal association between multiple myeloma, HIV and HCV infections. Conclusion: Multiple myeloma is not a commonly associated malignancy with HIV and/or HCV infection although hyperglobulinemia is often associated. Clinicians should be aware that multiple myeloma may occur as a non-AIDS defining cancer in HIV-infected individuals and/or as an extrahepatic manifestation in HCV-infected patients.

Keywords: Multiple myeloma, Human immunodeficiency virus (HIV), Acquired immune deficiency syndrome (AIDS), Hepatitis C infection

INTRODUCTION

Human immunodeficiency virus (HIV)-infected patients have benefited from dramatic improvements in prognosis and life expectancy because of effective highly active antiretroviral therapy (HAART). The incidence of AIDS-defining neoplasms has consequently decreased [1]. However, non-AIDS defining malignancies and chronic diseases are increasingly being reported in HIV-infected populations [1–3]. Chronic hepatitis C virus (HCV) infection is highly prevalent in HIV-infected patients due to similarities in the epidemiology. Hepatitis C virus infected patients frequently develop chronic liver disease, but extrahepatic complications have also been reported including HCV-associated B cell tumors [4, 5]. We describe a patient with HIV and HCV co-infection who developed IgG lambda light chain multiple myeloma. We review literature for a possible causal association between multiple myeloma, HIV and HCV infection.

CASE REPORT

A 47-year-old African-American female co-infected with HIV and HCV presented to our HIV clinic for routine follow-up. She had been diagnosed with HIV and HCV

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infections eight years before. She had been treated with interferon and ribavirin for HCV infection, but she did not complete therapy due to side effects from interferon. Her HIV infection was well-controlled with anti-retroviral therapy (zidovudine, lamivudine and efavirenz) with CD4 count of 1257 cells/µL and HIV-RNA viral load of less than 20 copies/mL. She complained of fatigue, diffuse arthralgias and unintentional weight loss (8 kg in two months). Physical examination was unremarkable. Her laboratory studies showed normochromic and normocytic anemia with a hemoglobin of 10 g/dL (normal range 10.3–15.2 g/dL), elevated serum creatinine at 2.5 mg/dL (normal range 0.4–1.2 mg/dL), and high globulin gap with a serum total protein of 9.6 g/dL (normal range 6.0–7.9 g/dL) and a serum albumin of 3.3 g/dL (normal range 3.5–5.0 g/dL). An extensive work-up to evaluate the etiology for renal dysfunction was undertaken. A renal sonogram revealed normal-sized kidneys. A 24-hour urine collection showed 2.1 g of protein. Anti-nuclear antibodies and complement levels were normal and HCV-RNA viral load was 20 million copies/mL with HCV genotype 1a. Serum cryoglobulin was negative. Serum and urine protein electrophoreses showed a monoclonal spike (4.26 g/dL) and immunofixation electrophoreses demonstrated monoclonal IgG with elevated free lambda light chains. Serum free monoclonal lambda light chain was elevated at 3500 mg/L (normal range 5.7 – 26.3 mg/L). Skeletal survey was negative. The patient underwent a renal biopsy which revealed light chain casts in the renal tubules (Figure 1A–B), confirmed as monoclonal lambda light chain on immunofluorescence (Figure 2). There was no evidence of HIV-associated nephropathy (HIVAN). A bone marrow biopsy showed more than 45% of plasma cell infiltration (Figure 3). After the diagnosis of IgG lambda light chain multiple myeloma was made, she was treated with nine cycles of initial induction chemotherapy (bortezomib and dexamethasone) followed by autologous hematologic stem cell transplant per the institutional treatment protocol. Subsequently, her serum creatinine improved to 1.1 mg/dL. The patient declined anti-HCV therapy because of her previous intolerance to interferon treatment.

**DISCUSSION**

The HIV/AIDS cancer match study, one of the largest epidemiological studies conducted in the United States (US), linked 15 population-based HIV/AIDS and cancer registries in the US. The study analyzed 413,080 HIV-infected persons in 34 US states from 1991 through 2005. During that period, an estimated 79,656 cancers occurred in the AIDS population. It was observed that the incidence of the AIDS-defining malignancies declined markedly over that 14-year period, whereas non-AIDS-defining neoplasms became the predominant type of cancer in HIV-infected persons during the HAART era. Multiple myeloma is one of those non-AIDS-defining
cancers whose incidence has increased recently in HIV-infected individuals [1].

The HIV-infected patients can present with a range of plasma cell disorders, from benign polyclonal hypergammaglobulinemia, or indeterminate monoclonal gammopathy of unknown significance (MGUS) to malignant plasma cell dyscrasias [6]. Multiple myeloma is usually not recognized as a malignancy that is associated with HIV infection. There have been only about 50 reported cases of HIV-infected patients with multiple myeloma in literature since the first case was reported in 1983 [7]. Multiple myeloma has also been reported as the initial clinical manifestation of HIV/AIDS infection [8]. The pathogenesis of multiple myeloma in HIV-infected patients is multifactorial (Figure 4). There are at least two major proposed mechanisms:

(i) a monoclonal paraprotein is specifically directed against the HIV-1 p24 gag antigen [9] and

(ii) continued stimulation by HIV viral antigens alters T-cell regulation of B cells which are transformed to malignant plasma cells [10].

Another postulated factor is interleukin-6 (IL-6), secreted from bone marrow stromal cells, which stimulates the growth of plasma cells as a paracrine mechanism. Human Herpes Virus-8 (HHV-8) has the ability to produce viral IL-6 (vIL-6), a human homolog of growth factor for plasma cells, which may perpetuate the growth of neoplastic plasma cells in HIV-infected individuals who are co-infected with HHV-8 [11].

The association of HCV infection with multiple myeloma is more controversial. Among the hematological disorders, monoclonal gammapathies rarely occur in patients with chronic HCV infection. It is speculated that HCV is lymphotropic and the mechanism which contributes to the pathogenesis of B cell non-Hodgkin lymphomas (NHL) may also play a role in the development of multiple myeloma in HCV-infected populations (Figure 4) [12]. Thus, it is a relatively weak association between HCV infection and multiple myeloma. It is also of interest to note that monoclonal gammopathy, if present, is more prevalent in patients with HCV genotypes 2a and 2c [13].

To the best of our knowledge, this is the first reported case of multiple myeloma in a patient with HIV-HCV co-infection. There are several unique characteristics in our patient. First of all, she is older than the average age (33 years) of HIV-positive patients with multiple myeloma. Secondly, the clinical course of multiple myeloma in our patient was relatively less aggressive than the course usually noted in HIV-infected patients. Thirdly, her HIV infection was well-controlled, and there was no parallel progression of multiple myeloma with HIV infection to AIDS. Lastly, her HCV genotype was 1a, which is not one of the most commonly observed genotypes (2a/2c) in HCV-infected patients with multiple myeloma.

In conclusion, chronic infection with human immunodeficiency virus (HIV) and/or hepatitis C virus (HCV) appears to increase the risk of developing multiple myeloma, although their oncogenic role has been less established. Our case highlights that dual HIV and HCV infections may possibly have an additive effect that drives this oncogenic process leading to multiple myeloma. Further research is warranted to establish a pathogenic causal relationship. Nonetheless, providers should keep in mind that multiple myeloma may occur in HIV-monoinfected or HIV-HCV co-infected patients, especially when serum gammopathy is present.

**CONCLUSION**

In conclusion, chronic infection with human immunodeficiency virus (HIV) and/or hepatitis C virus (HCV) appears to increase the risk of developing multiple myeloma, although their oncogenic role has been less established. Our case highlights that dual HIV and HCV infections may possibly have an additive effect that drives this oncogenic process leading to multiple myeloma. Further research is warranted to establish a pathogenic causal relationship. Nonetheless, providers should keep in mind that multiple myeloma may occur in HIV-monoinfected or HIV-HCV co-infected patients, especially when serum gammopathy is present.
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Authors declare no conflict of interest.

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REFERENCES
Advanced secondary abdominal pregnancy: A complication of induced abortion

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ABSTRACT

Introduction: Secondary abdominal pregnancies usually develop as a result of a tubal abortion, tubal rupture or uterine rupture with intra-abdominal implantation. Case Report: We report a 40-year-old patient gravida IV, para III suffering from abdominal pain. She was in the 29th week of gestation according to a 14th-week normal sonogram. She had unsuccessfully attempted to induce abortion with the assistance of a home midwife in the 14th week. This caused severe abdominal pain, but she chose not to be hospitalized because of fear of prosecution. Ultrasound demonstrated that the fetus was out of the uterus and revealed a heterogeneous mass in the left lower quadrant. Laparotomy revealed an area of thin fibrotic scar behind the uterus with normal tubes and ovaries. The fetus in the amniotic sac was surrounded by intestinal loops. The placenta, implanted behind the left broad ligament at the edge of the scar area was removed with brisk bleeding, which was controlled. No postoperative complications were observed. Conclusion: Presence of little free fluid and a uterine scar with normal fallopian tubes against a history of an unsuccessful induced abortion and severe illness are highly suggestive of a uterine rupture resulting in secondary implantation on the broad ligament. This is probably the first case that shows an abdominal pregnancy can result from unsuccessful induced abortion. In the countries where abortion is not permitted, patients with low socioeconomic status are prone to the dangerous consequences of illegal attempts to induce abortion. Close observation and special follow-up care are necessary in such cases.

Keywords: Induced Abortion, Pregnancy, Secondary Abdominal Pregnancy

INTRODUCTION

Abdominal pregnancy is a rare event and is classified as either primary or secondary, with the latter being more common. Secondary abdominal pregnancies usually develop as a result of a tubal abortion, tubal rupture, or uterine rupture with intra-abdominal implantation [1].

CASE REPORT

A 40-year-old gravida IV, para III woman was referred to us in the 29th week of gestation with a complaint of a two-week-long abdominal pain and weakness. The accompanying symptoms were vertigo and loss of appetite. There were signs of anemia. Her vital signs were stable. She had lost 3 kg in two weeks. Pregnancy had been confirmed three months earlier through serum
hCG measurement followed by ultrasound examination showing a normal 14-week pregnancy with antero fundal placenta. She reported no remarkable medical and surgical history and claimed that her early pregnancy was uneventful.

Physical examination upon admission revealed an abdominal mass rising from the pelvis without tenderness, rebound, and guarding. Laboratory results showed hemoglobin level 7.7 g/dL and a hematocrit value 25.7. Other examinations, including BUN, Cr, serum electrolytes, coagulation, and liver function tests, showed were normal. Gestational age was 29 weeks +4 days according to 14 weeks sonogram. However, ultrasonographic scan confirmed a breech-lying 26 weeks +2 days normal fetus out of the uterus with oligohydramnios and a vague heterogeneous 157×79 mm mass in the left lower quadrant with some free fluid in the abdominal cavity. The uterus was empty and normally placed. The MRI scan confirmed the diagnosis (Figure 1).

After the diagnosis was explained to the patient and her family, they revealed a history of an unsuccessful attempt to induce abortion with the help of a home midwife. This attempt caused severe illness, paleness and crampy abdominal pain, which wore off after two weeks. Despite severe illness, she had not visited any physician because of fear of prosecution.

The patient was given four units of packed cell which increased her hemoglobin to 11.7 g/dL. She underwent bowel preparation prior to the surgery. Accessibility of sufficient blood products was confirmed. A team consisting of two obstetricians/gynecologists, a general surgeon, a vascular surgeon and an anesthesiologist prepared for laparotomy under general anesthesia. The parietal peritoneum was opened through a midline incision. The amniotic sac was surrounded by intestinal loops. The baby was delivered. She had no dysmorphic features and weighed 800 g with an Apgar score of 4–0 at 1–20 minutes. The amniotic membranes were loosely clinging to the loops of jejunum and ileum and were easily dissected away. The uterus, fallopian tubes, and ovaries were normal. The placenta was implanted behind the uterus, on the left broad ligament and the cul-de-sac. There was a thin fibrotic scar measuring 3×2 cm behind the uterus near the edge of the placenta and was only covered by uterine serosa. Attempting to remove the placenta resulted in a brisk bleeding, but it was controlled by packing and suturing. The fallopian tubes were ligated, and the uterus was repaired. The patient was transfused with two units of packed cell during operation. No complications were observed after the surgery. The patient was discharged after six days in good health.

DISCUSSION

This case revealed the presence of little free fluid and a uterine scar with normal fallopian tubes. These findings when considered against backdrop of an unsuccessful induced abortion and severe illness are highly suggestive of a uterine rupture resulting in secondary implantation on the broad ligament.

In a review of available literature, we found only a few cases of secondary abdominal pregnancy resulting from uterine rupture. Teng et al. (2007) reported a secondary abdominal pregnancy following rupture of a uterine scar after two cesarean sections [2]. They claimed that their report was the first case of an early scar rupture resulting in a viable intra-abdominal pregnancy. Amritha et al. reported a viable secondary abdominal pregnancy following rupture of a rudimentary horn [3]. We can assume that our case is the first report of secondary abdominal pregnancy as a complication of an induced abortion.

Abdominal pregnancy causes relatively few symptoms, none of which are individually diagnostic [4]. Once she recovered from the acute symptoms of uterine damage, our patient was relatively asymptomatic in the rest of her pregnancy. Thus, a high index of suspicion is warranted for diagnosing abdominal pregnancy [3]. Ultrasonography is the main method for diagnosis [5, 6], but it fails to diagnose in half of the cases [4, 5]. The 29th-week sonogram of our patient revealed an abdominal pregnancy with the following findings: separation of the uterus from the fetus, extraterine placenta, oligohydramnios, abnormal lie of the fetus, some free peritoneal fluid and poor visualization of the placenta.

An MRI scan confirmed all the sonographic findings. Thus, it can be a useful adjunct to the ultrasound and may help identifying the spread of placental implantation. It seems wise to administer both tests to obtain maximal information for surgical planning [3, 4].
We chose to terminate the pregnancy upon diagnosis because a review of previous case reports showed that employing expectant management in the hope of fetal maturity involves a substantial risk of life-threatening hemorrhage [4, 5].

Fortunately, it was possible to separate the whole placenta intra-operatively. Placental management is a highly controversial issue. Retention of the placenta poses substantial risks. Most authors agree that the placenta should be removed provided its blood supply is identified and can be ligated without damaging other organs [3, 5]. Completely removal of the placenta made postoperative methotrexate administration unnecessary. Indeed, postoperative use of methotrexate is controversial in the case of the placenta left in place [4].

CONCLUSION

Abdominal pregnancy is a rare condition and diagnosis requires a high index of suspicion. In the countries where abortion is illegal, patients with a low socioeconomic status are prone to the dangerous consequences of secret attempts to induce abortion. So it is imperative that every primary health care provider closely observe the patients with unwanted pregnancy and provide special follow-up care. Prompt treatment upon discovery of the condition is essential. In managing abdominal pregnancy, maternal morbidity and mortality can be significantly reduced through preoperative diagnosis and evaluation, sufficient blood supply, bowel preparation, availability of a multidisciplinary surgical team and proper operative techniques.

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Author Contributions
Fateme Lalooha – 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
Khadijeh Elmizadeh – Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Fateme Salehi – 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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REFERENCES

Primary cerebellar B cell lymphoma: A case report

Anupam Datta, Arunima Gupta, Krishnangshu Bhanja Choudhury, Aruj Dhyani, Anup Majumdar

ABSTRACT

Introduction: Primary central nervous system lymphoma (PCNSL) is usually a diffuse large B cell non-Hodgkin lymphoma that originates in the brain, spinal cord, leptomeninges or eyes. Primary central nervous system lymphoma is a rare malignant tumor comprising less than 3% of all primary brain tumors. The PCNSL in immunocompetent patients is most commonly supratentorial, found adjacent to the ventricular surfaces and in deep white matter and subcortical structures, such as the basal ganglia, thalamus and corpus callosum. The lesion is single in 60–70% of patients. Lesions are located in the hemispheres (38%), thalamus/basal ganglia (16%), corpus callosum (14%), periventricular region (12%) and cerebellum (9%). Herein, we report an extremely rare case of a primary CNS B cell lymphoma involving cerebellum. Case Report: A 55-year-old female was presented with vomiting, ataxia and disorientation. T2-weighted axial, T1-weighted and gadolinium-enhanced T1-weighted axial magnetic resonance imaging scans show a contrast enhancing mass with peritumoral edema in the right cerebellar hemisphere. Excision biopsy was suggestive of non-Hodgkin lymphoma. Diagnosis was confirmed with immunohistochemistry favoring non-Hodgkin lymphoma of diffuse large B cell type. This patient received cytotoxic therapy with carmustine 200 mg IV 6 weekly 3 cycles followed by external beam radiation therapy 30 Gy. No recurrence was noticed in one year follow-up. Conclusion: Primary B cell lymphoma limited to the CNS is exceedingly rare in the middle aged immunocompetent individual. Little is known regarding etiologic factors, optimal management and prognosis.

Keywords: Non-Hodgkin lymphoma, Carmustine, B-cell lymphoma, Cerebellum

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INTRODUCTION

Primary central nervous system lymphoma (PCNSL) is usually a diffuse large B cell non-Hodgkin lymphoma that originates in the brain, spinal cord, leptomeninges or eyes. The PCNSL is a rare malignant tumor representing 3% of intracranial neoplasms and 4–6% of extra nodal lymphomas with a yearly incidence of 0.5 case per 100,000 people and with a median survival, if untreated, of 1.5–3.3
months. It is rare in immunocompetent patients. Median age at diagnosis is 60–65 years. The lesion is single in 60–70% of patients. Lesions are located in the hemispheres (38%), thalamus/basal ganglia (16%), corpus callosum (14%), periventricular region (12%) and cerebellum (9%). We report unusual primary cerebellar B cell lymphoma in a middle aged immunocompetent female patient.

CASE REPORT

A 55-year-old female was admitted in emergency department with symptoms of ataxia and disorientation of 1 week duration. She was suffering from headache and vomiting for last four months. The headache was diffuse, constant, throbbing and associated with episodic nausea and vomiting. Concurrently, she had a weight loss of 11 kg, daily fever and chills over the same duration. Clinical examination revealed slurred speech, finger to nose ataxia worse on the right side and truncal ataxia, with no other significant neurological deficits. A computed tomography (CT) scan of the brain revealed a solitary, well circumscribed, cystic, ring enhancing mass in the right cerebellar hemisphere with surrounding edema. T2-weighted axial, T1-weighted and gadolinium-enhanced T1-weighted axial magnetic resonance imaging (MRI) scans of brain showed a contrast enhancing mass with peritumoral edema in the right cerebellar hemisphere (Figure 1). Complete hemogram, liver function and renal function tests along with serum electrolytes were normal. Skiagram of chest and ultrasonography of abdomen were normal. She was seronegative for HIV 1 and 2, HBsAg and anti-HCV antibodies. Intravenous dexamethasone was initiated and a subtotal resection of the cerebellar lesion was performed. Hematoxylin and eosin preparation revealed a malignant round cell tumor probably non-Hodgkin lymphoma (Figure 2A–C). Diagnosis was confirmed with immunohistochemistry positivity for CD20 and CD10 (dim) and tumor stained immunonegative for CD3, CD5 and CD23. The Mib-1 labeling index was approximately 60–70%. Therefore, giving the impression of non-Hodgkin lymphoma (NHL) of diffuse large B cell type. With this biopsy report further investigations of CT scan of abdomen and bone marrow examination were done; all studies were normal. After three weeks of surgery cerebrospinal fluid (CSF) analysis showed normal glucose and protein levels, and 2 cells/mm³, all of which were lymphocytes. She was diagnosed as primary CNS non-Hodgkin lymphoma and went on to receive cytotoxic therapy with carbustine 200 mg IV 6 weekly three cycles followed by external beam radiation therapy (EBRT) 30 Gy. No recurrence was noticed in one year follow-up. Post treatment CT scan of the brain revealed no abnormality (Figure 3).

DISCUSSION

The PCNSL is defined as lymphoma limited to the cranial-spinal axis without systemic disease [1]. In the past, PCNSL was considered a rare disorder, accounting for 1–2% of all cases of non-Hodgkin lymphoma and fewer than 5% of all cases of primary intracranial neoplasm [2], just 13% of these arise in the posterior fossa. The PCNSL now represents around 3% of intracranial neoplasms and 4–6% of extranodal lymphomas with a yearly incidence of 0.5 case per 100,000 people.

An increasing incidence of this disease has been seen among patients with acquired immunodeficiency syndrome (AIDS) and among other immunocompromised persons. The incidence of PCNSL in the immunocompetent population has been reported to have increased more than 10-fold from 2.5 cases to 30 cases per 10 million populations [3]. The cause for the increase in incidence of this disease in the immunocompetent population is unknown. Two theories have been proposed. Lymphoma develops within a focus of inflammation in the CNS, as may occur in other organ systems such as the gastrointestinal tract. Another possible explanation is
that the cancerous lymphocytes develop elsewhere in the body, but acquire a receptor on their surface that draws them to a signal expressed only inside the CNS. The CNS is an immunologic sanctuary site rendering the neoplastic lymphocytes which may be systemically eradicated by an intact immune system, find relative protection within the central nervous system. In patients with AIDS, infection with the Epstein–Barr virus, which also causes infectious mononucleosis, is probably an important trigger for PCNSL, although the reason for this is not understood.

The natural history of this disorder differs between patients with AIDS and those without AIDS. The CT scans may show ring enhancement in 50% of AIDS patients while patients without AIDS almost always show only homogeneous enhancement [1]. Both groups do equally poorly without therapy (1–3 months mean survival), but the overall survival for treated patients is much better for patients without AIDS (18.9 months) than for those with AIDS (2.6 months) [1, 4].

Although more than 95% of patients with PCNSL are lymphoma of B cell origin, 45 patients with CNS lymphoma of T cell origin showed no difference in presentation or outcome in a retrospective series with data collected from 12 cancer centers [5]. Almost all are aggressive neoplasms of the diffuse large B-cell type. In a retrospective case series derived from 18 cancer centers in five countries of 40 patients with low-grade primary CNS lymphoma, a better long-term outcome was shown (7-year median survival) than is associated with the usual aggressive CNS lymphoma [6]. Anecdotald cases of primary CNS Hodgkin lymphoma have also been reported [7].

The PCNSL in immunocompetent patients is most commonly found adjacent to the ventricular surfaces and in deep white matter and subcortical structures, such as the basal ganglia, thalamus and corpus callosum. Most of these tumors are supratentorial. In one series, more than 70% of tumors were in a cerebral hemisphere and periventricular location (usually involving the corpus callosum or basal ganglia). Approximately, 50–70% of patients with this disease presented with solitary lesions, whereas the remainder had multifocal disease. These lesions characteristically enhance homogeneously with the administration of contrast agents [8]. The incidence of cerebellar involvement in one study in 2005 (16 of 170 lesions) was similar to the findings of Lanfermann et al. (15 of 174 lesions) [9, 10]. The exact incidence of cerebellar PCNSL in a middle aged individuals remains unclear inspite extensive literature search. The differential diagnosis of PCNSL includes gliomas, metastatic tumors, demyelinating disorders, subacute infarcts, and space-occupying lesions due to an infectious etiology.

The optimal treatment for includes radiation, high dose methotrexate, or more complicated regimen of de Angelis. Surgery other biopsy provides no therapeutic benefit because of the depth and diffuse nature of the tumor and should be reserved for the rare patient with neurological deterioration due to brain herniation. Antimetabolites such as methotrexate (MTX) and cytarabine (ara-C) remain most common chemotherapeutic agents used in PCNSL regimens. The MTX doses in excess of 1 gm/m² result in tumoricidal levels in the brain parenchyma and doses 3 g/m² yield tumoricidal levels in the CSF. However, MTX administration requires intensive monitoring with serum MTX level measurements. Other drugs used are carmustine, temozolomide, procarbazine and vincristine. Whole-brain radiation therapy alone is insufficient for durable tumor control and is associated with a high-risk of neurotoxicity in patients over 60 years of age. The initial response to radiation therapy in immunocompetent patients is excellent, often resulting in complete resolution of radiographic abnormalities. Nevertheless, the duration of response is short and median survival duration with radiation therapy alone averages only 18 months. Relapse in patients with parenchymal disease is usually within the brain, though leptomeningeal, vitreous and, rarely, systemic recurrences are reported.

The prognosis of nonselected patients with PCNSL is comparable to that of glioblastoma multiforme patients, with a 5-year survival rate after conventional chemotherapy and radiotherapy (RT) usually at less than 10% [11]. Unfavorable prognostic factors for survival are more than 60 years of age, WHO performance status greater than 2, poor neurologic function, elevated serum lactate dehydrogenase, extensive tumor spread, CSF protein level greater than 0.6 g/L, corticosteroid dependence, and absence of chemotherapy administration [12–14].

**CONCLUSION**

Primary B cell lymphoma is limited to the central nervous system, more so to cerebellum. It is rare in middle aged, immunocompetent individual. Little is known regarding etiologic factors, optimal management, and prognosis.

**Author Contributions**

Anupam Datta –Conception and design, Drafting the article, Critical revision of the article, Final approval of the version to be published

Arunita Gupta – 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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REFERENCES
Recurrent intraosseous ganglia in the proximal tibia following anterior cruciate ligament reconstruction using patella tendon bone autograft with a titanium interference screw: A case report

Aysha Rajeev, James Henry, Nanjappa Kashyap

ABSTRACT

Introduction: Intraosseous ganglia are benign cystic lesions and often multiloculated lesions located in the subchondral bone. These lesions are often asymptomatic but in cases located close to neurovascular structures or articular surfaces, they can be symptomatic, causing pain, neurologic dysfunction or articular fractures. We report a rare case of recurrent intraosseous ganglion following an anterior cruciate ligament (ACL) reconstruction in the proximal tibia. The ganglion developed after ten years of ACL reconstruction using patella tendon bone autograft fixed with a titanium interference screw. Initially the swelling mimicked the clinical and radiological features of a bone tumor. Case Report: A 40-year-old Caucasian male underwent ACL reconstruction using patella tendon bone graft and a titanium interference screw fixation in the tibia. After ten years of surgery he noticed a lump in the proximal tibia at the site of tibial screw. The radiological features were unremarkable. He had excision of the swelling along with screw removal. The swelling recurred again within six months. The lump was excised again and tibial tunnel bone grafted. After 4 weeks the lump reappeared, magnetic resonance imaging scan showed a large cystic lesion in the proximal tibia. After taking the advice from the oncologist, the lesion was curetted and bone grafted. The cyst went on to heal completely in six months time. Conclusion: The occurrence of intraosseous ganglion with absorbable interference screw has been reported. In our case report the intraosseous ganglion formed after using a titanium interference screw which has not been reported previously. It is also unique in the sense that the swelling recurring thrice and mimicked characteristics of a bone tumor.

Keywords: Recurrent, Intraosseous, Titanium interference screw, Mimic bone tumour

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INTRODUCTION

The occurrence of intraosseous ganglions is not that common. Ganglions are commonly soft tissue benign tumors usually arise in close proximity to joints, tendon or its sheaths [1]. The intraosseous penetration of a periosteal ganglion-like structures with subsequent formation of a bone cyst has been described in the past.
Intraosseous ganglia occur subchondrally, with or without an intact articular cartilage [3]. Sometimes the lesions occur in the metaphysis with connections to the joint surface or to the adjacent ligamentous structures [4]. Various theories have been proposed as to the pathogenesis of these ganglions. These include degeneration of connective tissue and migration of synovial fluid from an adjacent joint [1]. The possibility of acute or chronic trauma as one of the causative factors in these type of ganglions have also been postulated in a small percentage of patients [5]. The presence of serosanguinous mucin in the aspirate of these ganglions support this theory. Trauma sometimes cause to myxoid or mucinous degeneration which in turn leads to the pathogenesis of the ganglion [6].

In this case report, the patient developed an intraosseous ganglion almost ten years following an anterior cruciate ligament (ACL) reconstruction using patella tendon bone graft. The ganglion was located in the site of the tibial titanium interference screw. We are reporting this case as a unique and rare cause of recurrent and resistant intraosseous ganglion which has never been cited in English literature.

CASE REPORT

A 40-year-old Caucasian male underwent ACL reconstruction of the right knee using patella-tendon bone graft in 1999. He was doing extremely well for 10 years. In 2009 while playing football he had a knock on the medial aspect of the tibia. He noticed a lump just below the medial side of the knee which became very painful. There were no symptoms of instability. He had pain radiating from the lump on to the front of the upper leg. He was active and continued with his job.

On examination, there was no deformity of the knee. There was lump on the medial aspect of the tibial tuberosity, at the site of the tibial screw insertion for ACL reconstruction. The swelling was about 4x4 cm, tender with on redness or signs of inflammation. The movements were full range and tests for instability were negative. There was no distal neurovascular deficit. The radiological examination of the knee showed early degenerative changes in all three compartments. The position of the tibial screw was satisfactory with no bony changes (Figure 1). He was listed for planned procedure to take the tibial screw out. At the time of surgery the surgeon noticed a large soft tissue ganglion over the screw head. This was excised en-bloc and screw removed. The patient was reviewed back in the clinic two weeks following the surgery. He had mild ooze from the wound and swab was send for culture and sensitivity. The patient was again reviewed back in the clinic after one week which showed a jelly like material oozing from the wound but the culture reports were negative. He continued to receive wound dressings over the next few weeks and the wound healed satisfactorily, and the lump reappeared.

The patient returned six months after the operation with increased pain and swelling in the medial aspect of the knee. Physical examination showed a tender lump about 5x5 cm under the scar tissue. A decision was taken to excise the lump and bone graft the tibial tunnel. The scar was opened and the ganglion was excised and the tibial tunnel was grafted using freeze dried bone. Postoperatively the patient did well, the wound healed satisfactorily and the swelling disappeared.

The patient was then re-referred within four weeks by his general practitioner with increasing pain and swelling at the operative site. On examination, the wound has healed well and there was only a small tender lump palpable about 2x2 cm. The X-ray revealed that graft is incorporating well (Figure 2). In view of the continuing symptoms he was referred for an MRI scan urgently. The scan revealed a large cystic lesion in the proximal tibia (Figure 3). The patient was referred to the regional tumor unit for expert opinion. The tumor surgeon reported that the patient had got a recurrent intraosseous ganglion. A decision was taken to curette and bone graft lesion. The patient underwent excision of the ganglion and bone grafting using fresh frozen femoral head and tissue samples were send for histopathology (Figure 4). The patient was followed-up second week, sixth week and sixth month following the operation. The histopathology showed mature bone with fibro-collagenous tissue showing myxoid change and chronic inflammation. The myxoid material extends into bone suggesting an intraosseus ganglion. The cyst healed (Figure 5) and patient regained full knee movements. He was discharged from the outpatient clinic after 1 year.

Figure 1: The radiograph showing tibial screw in-situ with no bony changes.
The diagnosis of intraosseous ganglion should be considered in the absence of previous inflammatory and degenerative joint lesions. Most of these cases occur between the ages of 14–73. There are basically two types of intraosseous ganglia—one is idiopathic and the other one caused by penetration of an extraosseous ganglion into the underlying bone [1]. The juxta-articular, intraosseous ganglion reported in our case belongs to the second type, in which a soft tissue ganglion developed first and later penetrated into the underlying proximal tibia.

There are several theories proposed about the pathogenesis of intraosseous ganglia. The best possible theory would be a combination of intramedullary metaplasia followed by fibroblast proliferation. The fibroblasts secrete both hyaluronic acid and mucin which tend to accumulate and cause pressure necrosis. This leads to intramedullary trabecular degeneration and the formation of intraosseous cyst formation [3].

Most of the intraosseous ganglia lie subchondrally, with a normal articular cartilage and rarely communicate with joint or tendon sheath. The intact cyst often appears relatively smooth, round to oval shaped with an ochre-yellow color similar to a soft tissue ganglia, containing a thick gelatinous material. Microscopically, the walls are lined by poorly vascularized fibrous tissue and the cavity with flattened connective tissue cells which sometimes

**DISCUSSION**

Figure 2: X-ray showing the graph incorporating well after the first surgery.

Figure 3: Magnetic resonance imaging scan showing a large cystic lesion in the proximal tibia.

Figure 4: Histopathology showing bony tissue mixed with myxoid fibrous tissue consistent with an intraosseous ganglion.

Figure 5: Final radiograph showing the cyst well healed after curettage and bone grafting.
resemble synovial histiocytes. The secretion is high in hyaluronic acid and other mucopolysaccharides along with glucosamine, albumin and globulin.

The radiological features of an intraosseous ganglion include a scalloped cortical defect with a sclerotic margin. They appear thick and well defined which indicates a slow growing lesion [7]. These lesions very rarely shows calcification [8].

An MRI scan shows features similar to degenerative cysts with the overlying joint showing a normal articulation. The intraosseous ganglion may or may not communicate with the joint. The lesions show a fluid like appearance with low intensity on T1-weighted images, moderately low intensity on proton density weighted images and high homogenous intensity on T2-images.

The differential diagnosis of intraosseous ganglion is mainly giant cell tumor and chondroblastoma. Giant cell tumors are locally aggressive composed of giant cells, connective tissue and stromal cells. They usually occur between 3rd and 4th decade and predominantly epiphyseal location after growth plate closure. Chondroblastoma are commonly seen before skeletal maturity. Radiologically, they tend to be well defined lucent lesions with a sclerotic margins with calcification seen in 50% of cases.

The formation of cyst in the tibial tunnel after the use of an absorbable interference screw have been described [9]. Majority of the interference screws being used now-a-days are made of biodegradable material. The polylactide material causes hydrolysis and phaocytois [10]. This causes inflammatory foreign-body reactions responsible for as osteolytic reaction around the screw. Malhan et al. postulated that the gradual accumulation of breakdown products in a closed tibial tunnel ultimately lead to a cyst formation [11]. In our case report the interference screw was made of titanium.

The occurrence of pretibial cyst formation in the proximal tibia after ACL reconstruction has been reported with patella tendon autograft with the use of bioabsorbable interference screws [12, 13]. The actual mechanism by which these cyst forms has not been established. A mismatch of graft tunnel diameter, eccentric tibial tunnel, bone necrosis may slow down the incorporation of the graft into the tibia. This may lead to a water-channel between the knee joint and the proximal tibia there by leading to the formation of a cyst [14]. There has been no reports of any cyst formation using a titanium interference screw.

**CONCLUSION**

This is a rare case of intraosseous ganglion following anterior cruciate ligament reconstruction. The intra-articular communication after anterior cruciate ligament reconstruction of proximal tibia thereby causing the synovial fluid to flow to the proximal tibia under pressure may have attributed to the development of intraosseous ganglion. The clinician should be aware that it can mimic a bone tumor. The pathologic lesion can be successfully treated with curettage and bone grafting.

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**Author Contributions**

Aysha Rajeev – 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

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

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**Guarantor**

The corresponding author is the guarantor of submission.

**Conflict of Interest**

Authors declare no conflict of interest.

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**REFERENCES**


Missed retained knife blade injury: A potentially lethal trap for the unwary

Victor Kong, John Bruce, Leah Naidoo, George Oosthuizen, Grant Laing, Damian Clarke

ABSTRACT

Introduction: Retained knife blade is an uncommon injury and they often present in a spectacular fashion. Concealed retained knife blade, however, is difficult to diagnose without a detailed clinical and radiological assessment. Extraction requires careful planning in a controlled environment, preferably in the operating theater. Case Report: We present the highly unusual case of a 25-year-old male in whom a retained knife blade following a stab to the left anterior leg was missed on three separate visits to a rural hospital. Conclusion: Whilst diagnosis and management continues to be challenging, clinicians must always remain vigilant to the possibility of such injury. When the presentation is unclear, a high level of suspicion, careful clinical assessment and judicious use of radiography is of paramount importance. Early referral to a trauma center for definitive management is crucial.

Keywords: Missed injury, Retained knife blade, Extraction

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INTRODUCTION

Knife injuries are common and can present with a full spectrum of pathology, ranging from minor cuts to severe life-threatening injuries. Retained knife blade injury logically falls within the scope of this injury spectrum [1]. Delayed presentation due to missed injuries is associated with significant morbidity [2]. We report a highly unusual case of a 25-year-old male who presented to our trauma unit some three weeks after the initial injury, with a concealed, retained knife blade in the left leg. It emerged that the actual injury was missed and he had been sent home by the doctor on multiple occasions.

CASE REPORT

Mr. B was 25-year-old male who presented to a rural hospital following a single stab injury to the left anterior leg. He was moderately intoxicated at the time. He was assessed by a locum doctor at the hospital and was thought to be an uncomplicated superficial stab wound.
He was discharged home after being given a prescription for paracetamol. One week after his initial injury, the patient presented again complaining of increasing pain from the wound. Apart from some localized tenderness, there was no discharge or surrounding cellulitis. He was given a course of oral flucloxacllin, 250 mg, four times daily for presumable wound sepsis. He represented again the following week with no improvement and complained of a sense of fullness and increasing pain in the anterior leg extending to the calf. It was decided that antibiotics dose was to be increase and he was again discharged with a further course of oral flucloxacllin, this time at 500 mg, four times daily. A week later, he presented for the third time complaining of worsening of his symptoms, and at the patient’s insistence, he was referred to our trauma unit for a second opinion.

On arrival, the patient was in significant discomfort. His baseline vitals were: heart rate: 90/min, blood pressure: 135/70 mmHg, temperature 37.5°C. A small puncture wound (approximately 1.5x1.5 cm) was noted in the anterior aspect of the left leg (Figures 1 and 2) and approximately 10 cm inferior to the tibial tuberosity. The wound edges appear sloughy, with no surrounding cellulitis. Tenderness was noted that extended from anterior leg to the calf but no foreign body was palpable. His pulses in the lower limbs were normal and equal, with normal Doppler flow signal. Suspicion was aroused about the retention of a foreign body and an urgent X-ray was arranged. A large retained knife blade was seen, with the trajectory in the superior direction, lodged between the interosseus membrane (Figure 3). He was commenced on intravenous co-amoxiclav, 1.2 g, and was taken immediately to the operating theatre for a planned removal. Intraoperatively, a longitudinal incision was made extending from the wound edge, and the knife blade was immediately visible. The knife blade was easily extracted with minimal hemorrhage (Figure 4). There was no evidence of osteomyelitis. An on-table angiography was performed, which showed no vessel injuries. The wound was thoroughly irrigated and was left to close by secondary intension. He had an uneventful recovery and was discharged on fifth day.

**DISCUSSION**

Retained knife blade remains a rather uncommon injury [1]. Most of these injuries usually present in spectacular fashion and the protruding objects are usually obviously visible [3, 4]. Heroic attempts at extraction in the emergency department may result in massive torrential hemorrhage [5]. Most experience reported in literatures has been from isolated case reports and several small case series [1, 6, 7]. Being an uncommon injury, most centers have limited experience in its management [1]. Concealed retained knife blade injury is much more
unusual. It becomes even more problematic if the initial retained blade is not recognized. Delayed presentation is not uncommon and one series reported a delay of up to eight weeks after the initial injury [7]. Delay can be associated with significant morbidity relating to sepsis, even more so with injuries to the extremities.

This highly unusual case highlighted several important issues worth discussing. Our patient initially presented to a rural hospital and based on an unclear history related to intoxication, the injuries was missed. However, with the subsequent multiple representations, the possibility of a retained blade injury was never considered, leading to significant further delay in diagnosis. No radiography was even taken on the suspected areas as the injury was never considered, which would have easily revealed the retained blade. In this case, the old adage of ‘what the mind does not know, the eyes do not see’ stood the test of time.

It is equally important to recognize that the appearance of a stab wound can be deceptive. It is also obvious that it bears no relationship to the magnitude of the potential underlying injury. In our case, it was also rather peculiar having noted that the wound was only approximately 1.5 cm, but the width of the retained blade was over 2 cm. The most likely explanation of this disparity was probably related to a degree of wound contraction after a long delay. However, it is crucial to understand that this type of presentation could potentially be misleading.

This case, however, is not an isolated incidence related to suboptimal management for these patients who present to rural hospitals in South Africa [8]. Rural hospitals in South Africa and other developing countries, facilities and resources are often severely limited [8]. This situation is usually further compounded by a severe and on-going shortage of medical staff and poorly maintained infrastructures. The few who provide these essential health services often lack the training and experience in managing trauma patients [8]. Furthermore, even in well-staffed urban trauma centers, diagnosis and management of these patients is still difficult.

Once the retained knife blade was identified, the issue with definitive management can be difficult. In most situations, a spiral computed tomography (CT) scan can usually help defining the relation between the retained blade and major significant anatomical structures [5]. This will generally facilitate a far safer extraction plan [1]. Angiography is generally recommended if initial CT scan is equivocal, especially if a major vascular injury is suspected [1]. In our case, we elected to pursue a combined exploration and extraction in the operating theater due to the anatomical location of the injury. It was anticipated that major hemorrhage in the vessels inferior to the arterial trifurcation of the knee could be safely dealt with and adequate vascular control achieved. Furthermore, if there were injuries to one of the small branches from the trifurcation, it was highly unlikely this would have significant impact on the lower limb perfusion, and a simple ligation would have been a safe option [9]. The subsequent on table angiogram performed proved to be reassuring. Despite the patient’s protracted delayed to missed diagnosis on multiple occasions, he fortunately made a full recovery.

**CONCLUSION**

Retained knife blade is an uncommon injury. Concealed, retained knife blade is notoriously to diagnose. Therefore, a high index of suspicion must be maintained, coupled with judicious use of imagining, especially if the history is unclear. Early and carefully planned extraction in the operating theatre by a suitably trained trauma specialist remains the cornerstone of good management.

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**Author Contributions**

Victor Kong – 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

John Bruce – Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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Guarantor
The corresponding author is the guarantor of submission.

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REFERENCES
Novel surgical extrication of penile constricting metal ring: A case report

Abimbola Olaniyi Olajide, Amogu Kalu Eziyi, Folakemi Olajumoke Olajide, Babatunde Kazeem Beyioku

ABSTRACT

Introduction: Penile entrapment with strangulation is a rare clinical entity requiring urgent decompression [1]. Entrapping object is usually applied when penis is flaccid or semi-erect. It becomes impossible to remove when penis becomes erect leading to swelling of the penis distal to the object [2]. The management of this condition poses unique challenges to the treating physician through variable presentation as well as lack of specific treatment options. No standard treatment modality is described in literature; every case needs an individual approach depending upon the circumstances and facilities available [3]. Therefore, penile entrapment remains a challenge occasionally faced by urologists; it is tasking and requires ingenuity for successful treatment [4].

Methods described in literature include aspiration of corpora, use of saws, grinders, bolt cutters, and dental drills. Others include denudation of penile skin with postoperative skin grafting, amputation or use of myofascial flap for reconstruction of penis [3].

We report this case to illustrate a surgical technique used in the management of a penile entrapment with successful outcome.
CASE REPORT

A 16-year-old boy presented in accident and emergency department with 4 days history of entrapment of his penile shaft in a steel ball bearing which he has used for erotic purpose. The ring had slipped in easily and entrapped after penile erection with failure of all attempts to remove the metal ring. He did not seek any help until four days. Later when he reported to relatives who made several attempts at remove the metal ring including use of metal cutting saws with no success. Urine stream has reduced progressively with increasing straining at micturition.

On examination, stainless steel ring of about 1.5 cm width impacted at the base of the penile shaft with marked swelling of the penile shaft distal to the metal ring and two areas of dark discolorations were noted on the penile skin (Figure 1). Sensory sensations were preserved on the penile shaft but arterial pulsation was not palpable. Initial attempts by lubrication and gentle traction on ring were unsuccessful following which he was transferred to the operating room. After failure of Gigli, orthopedic and metal cutting saws, he was given general anesthesia through a face mask, a transverse circumcising incision was made just proximal to the coronal sulcus, and this was extended vertically on the midline of the ventral surface of the penis. This incision was deepened to the sub-dartos plane, skin and dartos layer were subsequently raised off the Buck’s fascia. Fluid with blood oozed out of the subcutaneous tissue with swelling of skin and subcutaneous tissue observed to reduce immediately. The skin and subcutaneous tissue then slid easily under the metal ring (Figure 2), followed by the erectile tissue contained within the Buck’s fascia (Figure 3). Following removal of the metal ring, the skin was closed in single layer using nylon 3-0 suture (Figure 4). Size 14 Foley’s catheter was passed and left in place for 48 hours.

Postoperative period was uneventful and he was discharged and sutures were removed on 4th and 8th postoperative days, respectively. The skin recovered completely and there was no complaint with urination and erection thereafter (Figure 5). He attended clinic for five months and lost to follow-up thereafter.

Figure 1: Preoperative picture showing entrapment and strangulation of penile shaft in the metal ring.

Figure 2: Reduction of swelling with sliding of skin and subcutaneous tissue under metal ring after penile degloving.

Figure 3: Metal ring on the erectile tissue after releasing the skin and subcutaneous tissue from under the metal ring.
DISCUSSION

Though rare, penile entrapment can be associated with serious complications. It causes blockage of the venous return resulting in swelling of the penile skin and subcutaneous tissue distal to the object. Delay in treatment results in penile strangulation with ischemic necrosis and gangrene of the tissue [5].

Treatment requires urgent removal of the offending device and decompression of edema to prevent ischemic injury to the affected tissues. Various methods for removal of the various constricting devices have been documented in literature, these often involves consultation of various services, including emergency services to obtain special instruments to remove the constricting object [3, 6–9]. Most authors have canvassed for a multi-disciplinary approach including the assistance of jewelers and locksmiths and fire department to procure appropriate tools for removal [1]. A few surgical techniques have also been described including winding of silk from the glans, string techniques and aspiration of corpora tissue [5, 10]. None of these methods is considered a standard modality of treatment; each case needs an individual approach depending upon the encircling material, assessment of severity and available facilities and tools [3].

The method presented in this report is applicable and successful because the swelling in penile entrapment is due mainly to venous engorgement resulting in swelling mostly in the skin and the subcutaneous fascia (dartos) which is loosely attached to the underlying Buck’s fascia. In addition, thickness of Buck’s fascia and corporeal tissue resist pressure on the deep vessels, avoiding gangrene of the erectile bodies [3]. These facts allow for easy elevation of the penile covering over Buck’s fascia and its content (degloving). As soon as degloving started, fluid and blood was observed to egress from the subcutaneous tissues with immediate reduction of the swelling of the penile skin and subcutaneous tissue. This gave space for skin and subcutaneous tissue to slide under the constricting ring first and the penile shaft thereafter.

This procedure is simple, purely surgical and not new to most urologists because it is part of some forms of urethroplasty for hypospadias and urethral stricture. It obviates need for any special instrument and further delay in waiting for fire fighters, jewelers or locksmiths.

CONCLUSION

Penile entrapment and strangulation remains a rare challenge to urologists worldwide with need for emergency decompression to prevent grave complications. Simple degloving of the penis reduces the swelling and allows easy removal of the constricting object.

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Author Contributions
Abimbola Olaniyi Olajide – Conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Amogu Kalu Eziyi – Conception and design, Critical revision of the article, Final approval of the version to be published
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Beta-hemolytic group B Streptococcus meningitis in a young healthy woman

Li Han, Rohit Gosain, Maria Plataki, Daniel Horowitz, Daniel Bordea

ABSTRACT

Introduction: Group B Streptococcus (GBS) is the leading cause of bacterial meningitis and sepsis in neonates but is a rare cause of meningitis in adults. The GBS infections in adults include bloodstream infections, pneumonia, skin and soft-tissue infections, and bone and joint infections. Case Report: A 23-year-old woman with a past medical history of stroke at the age of four and Staphylococcus aureus toxic shock syndrome at age 10 was presented to the emergency department with three days of worsening frontal headache, fatigue and mild confusion. Patient denied fever, photophobia, neck stiffness, nausea and vomiting. Brudzinski’s sign and Kernig’s sign were negative. Laboratory data showed initially elevated WBC (13.5 x 10^3/µL) and normal metabolic panel. Cerebrospinal fluid (CSF) analysis was normal with two white cells, all lymphocytes. After two days of hospitalization, she was back to her normal state without any headache or confusion and was discharged home. One day after discharge, both urine and CSF culture grew β-hemolytic GBS but with negative blood cultures. She was readmitted and placed on ceftriaxone 2g IV twice daily for 14 days. Conclusion: This case illustrates an unusual presentation of group B Streptococcus meningitis. Internists should be aware of atypical group B Streptococcus meningitis and consider treating patients with empiric antibiotics on clinical suspicion.

Keywords: Group B Streptococcus, β-hemolytic

INTRODUCTION

Group B Streptococcus (GBS) is gram-positive β-hemolytic coccus characterized by the presence of group B Lancefield antigen and the main virulence factor is the polysaccharide anti-phagocytic capsule. The GBS is the leading cause of bacterial meningitis and sepsis in neonates but it is a very rare cause of meningitis in adults. The GBS infections in adults include bloodstream infections, pneumonia, skin and soft-tissue infections, and bone and joint infections. This organism has been recognized with increasing frequency as a substantial cause of morbidity and mortality among non-pregnant adults.

CASE REPORT

A 23-year-old woman with a past medical history of stroke at the age of four and Staphylococcus aureus...
toxic shock syndrome at age 10 was presented to the emergency department with three days of worsening frontal headache, fatigue and mild confusion. She complained of continuous throbbing-like headache in the left frontal lobe, which she rated as 8 on a severity scale of 10. Patient denied fever, photophobia, neck stiffness, nausea and vomiting. In the emergency department, she was found to be confused with behavioral changes such as trying to scan one of her cards on the Purell hand sanitizer machine and clicking her car remote control to turn on the TV. Physical examination was normal except for a 2/6 systolic murmur at the apex with no radiation. No nuchal rigidity or focal neurological deficit was observed. Brudzinski’s and Kernig’s signs were negative. Laboratory examination showed initially elevated WBC 13.5x10^3/µL that went down to 9.5x10^3/µL the next day and normal metabolic panel. Cerebrospinal fluid (CSF) analysis was normal with two white cells, both lymphocytes (Table 1). Computed tomography (CT) scan of the head showed no acute intracranial abnormality. Magnetic resonance imaging (MRI) of the brain revealed encephalomalacia in the left frontal lobe but no acute infarction (Figure 1). Electroencephalogram (EEG) was slightly abnormal with occasionally increased amount of intermittent delta activity, seen mostly in the frontal central areas bilaterally. This finding could correlate with the patient’s clinical confusion. Transthoracic echocardiogram showed mild concentric left ventricular hypertrophy with ejection fraction 60–65%, trace mitral and tricuspid regurgitation. After two days of hospitalization without using any antibiotics, she was back to her normal state without any headache or confusion and was discharged home. One day after discharge, both urine and CSF culture grew β-hemolytic GBS. Blood cultures were negative. Urinalysis showed specific gravity 1.03, leukocyte esterase negative, nitrate negative, white blood cell count <5/Hpf, glucose negative, protein negative, ketones negative. CSF culture showed 4 colonies of β-hemolytic GBS and urine culture showed 50,000 organisms/mL. She was readmitted and placed on ceftriaxone 2 g IV twice daily for 14 days. Further laboratory work-up included HIV testing, and immunoglobulin level measurement both were negative. Computed tomography of the abdomen and pelvis was performed and abscess was excluded. She was discharged home to complete 14 days of intravenous antibiotic treatment.

DISCUSSION

The GBS is a gram-positive β-hemolytic streptococci and bacitracin resistant organism. It is a part of the normal flora of gastrointestinal tract, upper respiratory tract and genital tract in both men and 15–45% women [1]. However, GBS is capable of causing serious infections, primarily in neonates, pregnant women and immunocompromised individuals. Neonates acquire GBS infection in utero or during passage through vagina. Physiological changes in pregnant women make them more susceptible to GBS infections, mainly in the urinary tract where the bacteria are found in high numbers. However, rarely it may also affect non-pregnant or immunocompetent adults, such as those diagnosed with diabetes mellitus, malignancy, HIV infection, and advanced hepatic renal disease [2]. Incidence of invasive GBS infections has increased by 2–4 folds in the past two decades and approximately two-thirds of these cases are observed in adults [3].

Classical cases of bacterial meningitis presents with diffuse headaches, fever, nuchal rigidity, and change in mental status [4, 5]. Some patients have temperatures ranging above 38°C, and some present with hypothermia, but it is important to note that no one presents with normal temperature[6]. However, our case was unique such that the patient denied fever and presented with headache, confusion and normal CSF analysis.

CONCLUSION

We report a very unique case of group B Streptococcus meningitis in a 23-year-old non-pregnant immunocompetent female. The patient did not present with classical triad of typical bacterial meningitis, rather with headache, confusion and normal cerebrospinal fluid analysis. This patient acquired the infection probably from urinary tract because both the urine and cerebrospinal fluid cultures are positive for β-hemolytic streptococci.
group B Streptococcus. Internists should be aware of atypical group B Streptococcus meningitis and consider treating patients with empiric antibiotics.

*********

Author Contributions
Li Han – Substantial contributions to conception and design, Analysis and interpretation of data; Drafting the article, Final approval of the version to be published
Rohit Gosain – 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
Maria Plataki – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published
Daniel Horowitz – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published
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Acardiac twin: Conservative management

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ABSTRACT

Introduction: Acardiac twin is a rare anomaly occurring in 1 in 35,000 pregnancies. The outcome is fatal for the acardiac twin with a 50–75% mortality of the normal pump twin. Management options include conservative management, medical therapy, occlusion of connecting vessels and selective termination of the acardiac twin. The management of a twin pregnancy complicated by an acardiac twin is a challenge because the continuous growth of the acardiac fetus is deleterious to the healthy pump twin leading to cardiac insufficiency, polyhydramnios, prematurity and even death of the normal twin. Case Report: We report three cases of acardiac twinning which presented with different manifestations, acardius acephalus, acardius myelocephalus and acardius amorphous. All the three cases were managed conservatively with good outcome of the pump twin in all the cases. We present the cases with review of literature. Conclusion: Conservative non-intervention may be appropriate in developing countries with low resources.

Keywords: Acardiac twins, Twin pregnancy, Conservative management

INTRODUCTION

Acardiac twinning or Twin Reversed Arterial Perfusion (TRAP) is a rare anomaly occurring in monozygotic multiple pregnancies with an incidence of 1% and in 1 in 35,000 pregnancies [1]. The mortality in recipient acardiac twin is 100% whereas the normal donor twin is also at an increased risk of morbidity and mortality. This is a report of three cases of acardiac twins with different presentation. One was acardius acephalus, another was acardius myelocephalus and the third one was acardius amorphous. All cases resulted in live birth of the normal twin.

CASE REPORT

Case 1: A 30-year-old third gravida with previous two uneventful vaginal deliveries came to us for the first time at 34 weeks of gestation with twin pregnancy in active labor. She delivered a normal female baby of
1.7 kg followed by a malformed, dead acardiac fetus of 900 g (Figure 1). The acardiac fetus had a grossly malformed upper body without upper limbs head and neck (acardius acephalus) (Figure 2A). The trunk was partially developed. The lower limbs were developed but had bilateral congenital Talipes equinovarus deformity and syndactyly (Figure 2B). External genitalia resembled female (Figure 2C). There was a single placenta with two cords, one small and thin belonging to the acardiac fetus. (Figure 3). Autopsy showed the absence of neural tissue, (Figure 4) absence of lungs, incomplete diaphragm, rudimentary cardia, liver tissue, uterus, fallopian tube, ovary, kidney and ureters. (Figure 5). The normal twin was fine at the time of discharge three days later.

**Case 2:** A 26-year-old second gravida with previous normal delivery came with 24 weeks pregnancy. On examination, the uterus was of 28 weeks size. Ultrasound revealed twin pregnancy with one normally formed fetus of 24 weeks and another fetus with a partially formed head with hydrocephalus and enlarged lateral ventricle (Figure 6A–B). The spine was partially seen and the lower limbs
were recognizable. No other anatomical structure was made out. Gross abdominal edema and ascites was noted (Figure 7). The placenta was a single large mass. Color Doppler of umbilical artery of the abnormal fetus showed reversal of flow thus confirming the diagnosis of acardiac pregnancy. The parents were counselled regarding the prognosis of the surviving twin, options of conservative and invasive management and non availability of invasive management in our center. After obtaining their consent, the lady was managed conservatively with regular follow-up and serial ultrasound examination. At 36th week of gestation, she developed polyhydramnios and pre-eclampsia and an emergency cesarean section was done. A borderline term male fetus of 2 kg weight with normal morphology was delivered by breech extraction. The other twin was acardiac weighing 750 grams with a partially developed head and face. Lower limbs could be made out. (acardius myelocephalus) (Figure 8A–B). External genitalia were poorly developed. Autopsy was not done for the acardiac fetus. The normal twin fared well and was discharged on seventh postoperative day.

**Case 3:** A second gravida of 21 years of age with a previous cesarean section presented at 24th week of gestation with twin pregnancy. Ultrasonography showed the first twin to be an acardiac amorphous mass without any recognizable structures. Only the spine was made out throughout its length (Figure 9). The second twin was normally developed. There was a single placenta (Figure 10). After explaining about the treatment options, the lady opted for conservative management in view of the cost and non-availability of facilities for invasive management at our hospital. She was followed-up by serial ultrasound examinations. The growth of the normal fetus was satisfactory without any complications. The patient had regular antenatal check ups and was treated for anemia. She went into labor at 36th week of gestation. An emergency cesarean section was done in view of previous cesarean section. The first twin was an amorphous soft, globular mass without any recognizable structure and was weighing 700 g (acardius amorphous) (Figure 11). The second twin was a normal female of weight 2.1 kg. The postoperative period was uneventful. The mother and the normal twin were discharged on the eighth postoperative day.
Multiple pregnancy accounts for 1.5% of all pregnancies with a perinatal morbidity and mortality of 10% [2]. Acardiac twinning or TRAP sequence is a rare congenital anomaly of monozygotic multiple pregnancy due to abnormal placental anastomosis characterized by formation of a malformed fetus with an absent or rudimentary heart (acardius) and other structures. There is usually a normally formed donor twin who may have features of heart failure. Acardiac fetuses were first described by Benedetti in 1533 [3]. Acardiac acephalic variety is the most common type of acardiac fetus. It has been hypothesized that the TRAP sequence is caused by a large artery to artery placental shunt often accompanied by a vein to vein shunt. Within the single shared placenta, arterial perfusion pressure of the donor twin exceeds that of the recipient twin who thus receives reverse blood flow of deoxygenated arterial blood from its co-twin. This used blood reaches the recipient twin through its umbilical arteries and preferentially goes to its iliac vessels. Thus only the lower body is perfused and disrupted growth and development of the upper body results [4].

An acardiac twin should be suspected in all monochorionic, malformed fetuses with cystic hygroma, generalized edema and an absent cardiac pulsation with a non-functioning heart. Also, an ultrasonography finding of twins revealing discordant or grotesque malformation along with reverse flow in the umbilical artery is usually diagnostic of an acardiac twin [5]. This can be diagnosed in first trimester by vaginal scanning and color Doppler sonography. Serial ultrasonography is indicated to assess such twin pregnancies.

Failure of the head growth is called acardius acephalus, a partially developed head with identifiable limbs is acardius myelocephalus and failure of any recognizable structure to form is acardius amorphous [6]. Acardiac twins can also be classified as follows:
- Acardius anceps – when head is poorly formed,
- Acardius acephalus – if the head is absent
- Acardius acormus – when only head is present.

Based on the development of heart, acardiac twins can be classified as hemiacardius when heart is incompletely formed and holoacardius if the heart is absent [7]. The prominent features of the recipient twin are total or partial absence of cranial vault, holoprosencephaly, absent facial structures, anophthalmia, microphthalmia, cleft lip, cleft palate, absent or rudimentary limbs, lungs, heart, liver and gallbladder, diaphragmatic defects, esophageal atresia, ventral wall defects, ascites, edema of skin and single umbilical artery.

Management of twin pregnancy with an acardiac fetus is a challenge as the continuous growth of the acardiac fetus is deleterious to the healthy pump twin. It can lead to cardiac insufficiency, polyhydramnios, prematurity and even death of the structurally intact twin in up to 50% of cases [8].

Moore et al. reviewed 49 cases of acardiac twins and reported that perinatal outcome was related to the ratio of weight of the acardiac twin to the weight of the normal twin. They stated that when twin weight ratio which is the acardiac twin weight divided by the normal twin weight was above 70%, preterm labor, hydramnios and congestive heart failure in the pump twin were found and when the weight of the acardiac fetus was less than 25% compared to the pump twin, the prognosis was better.
CONCLUSION

Expectant management with close antepartum surveillance deserves consideration in cases of monozygotic twins with Twin Reversed Arterial Perfusion sequence. Neonatal mortality of the pump twin diagnosed antenatally may be considerably less than reported. The interruption of vascular communication between the twins is difficult to accomplish. It needs expensive equipment and trained personnel for the procedures. Hence invasive treatment should be considered only in cases with poor prognostic factors. Such pregnancies should be managed by fetal medicine specialists familiar with invasive procedures. Intra fetal ablation procedures are better than cord occlusion techniques. In our cases, all the three pump twins survived with expectant management.

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[8]. The weight ratio in this study was derived from post delivery weights. They also proposed a second order regression equation to predict weight of the acardiac twins antenatally: Weight (g) = 1.2 x (longest dimension (cm)) 2 – 1.7 x longest dimension (cm). Also the use of abdominal circumference ratios could provide a better approach to establish the differences in body weights [8].

Brassard et al. reported that low pulsatility indices in the umbilical artery perfusing the acardiac twin compared with the pump twin correlated with poor prognosis [9].

Optimal management is controversial. Expectant management versus prenatal intervention is to be debated. Many methods of management have been proposed including termination of pregnancy, serial ultrasound scans to monitor for signs of decompensation, medical management of polyhydramnios or by serial amniocentesis, hysterotomy to remove anomalous twin and invasive procedures. Goal of prenatal treatment is to stop blood flow to the acardiac twin without affecting the pump twin in order to improve its outcome. Platt et al. in 1983 were the first to suggest occlusion of the circulation to the acardiac twin as the definitive treatment to interrupt blood supply to it [10]. Minimally, invasive intervention methods are through cord occlusion techniques or intrafetal ablation. Cord occlusion has been attempted by embolization, cord ligation, laser coagulation, bipolar diathermy and monopolar diathermy while intrafetal ablation is performed with alcohol, monopolar diathermy, interstitial laser and radiofrequency [11].

Tan and Sepulveda recommended intrafetal ablation as the treatment of choice than cord occlusion [11]. They claimed ultrasound guided intrafetal approach to be easier, less invasive and with a higher rate of success than ultrasound and fetoscopy guided cord occlusion procedures.

Invasive treatment should be restricted to those pregnancies which would benefit from prenatal intervention like those where the pump twin is at a significant risk of prematurity, cardiac insufficiency or death and should be considered in presence of poor prognostic factors like polyhydramnios, ultrasound markers of cardiac insufficiency, large acardiac twin and rapid growth of or evidence of substantial blood flow perfusion through the umbilical vessel supplying the parasitic mass [12].

Sullivan et al. advocated expectant management in all cases [13]. They reported 90% survival in pump twin in 10 pregnancies with an acardiac twin managed expectantly. They cautioned against aggressive intervention and recommended expectant management with close fetal surveillance. Stamatan et al. gave an opinion that conservative management is indicated in cases where the acardiac twin is small and when there are no signs of cardiovascular impairment in the pump twin [14]. Serial ultrasound surveillance is important for detecting any worsening of the condition, which may suggest the need for interventions to optimize the pump-twin’s chance for survival [15].
Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES

Primary cavernous hemangioma of the thyroid gland

Devika Gupta, Sunita Kakkar, Pooja Gupta, Vandana Rana

To the Editor,

Hemangiomas are benign vascular neoplasms that have a characteristic clinical course marked by early proliferation and spontaneous involution. They occur in a number of organs including skin, lips, liver, colon, brain, etc. [1, 2].

Primary thyroid hemangiomas are extremely uncommon with only a few cases have been reported [3]. We report a case of cavernous hemangioma presenting as solitary nodule thyroid.

A 49-year-old male presented with one year history of slowly enlarging swelling on left side of the neck. There was no history of any associated pain, voice change or dyspnea. There was no history of any trauma or previous fine needle aspiration cytology (FNAC). There was no family history of thyroid disease.

Local examination revealed an approximately 3x2.5 cm nontender swelling in front and left side of cricoid cartilage. The swelling was firm to hard with well defined margins and it moved with deglutition.

Serum thyroid stimulating hormone was 3.8 mIU/L (normal range 0.4–4.0 mIU/L) and free T4 was 9 pmol/L (normal range 10.3–24.5 pmol/L). These hormonal levels were normal and no antithyroid antibodies were detected.

Thyroid ultrasonography revealed a 2.5x2.8 cm isoechoic lesion in the left lobe of thyroid with a hypoechoic rim and a prominent vessel having arterial flow in its superior aspect (Figure 1). Ultrasonography did not show any areas of calcification or cervical lymph nodes. Right lobe of thyroid was normal.

The FNAC of the mass was attempted thrice but was inconclusive as only blood was aspirated. Patient underwent left hemithyroidectomy. Gross examination of the cut surface of the specimen revealed a well circumscribed nodular lesion measuring 2.5x2.8 cm. It appeared cystic and hemorrhagic in appearance.

Histological examination of the tumor confirmed a cavernous hemangioma. There were multiple, dilated, irregular anastomosing vascular channels of varying sizes. These vascular spaces were lined by bland endothelial cells. Amidst these were noted an occasional entrapped thyroid follicle (Figure 2A–B). The patient was discharged from the hospital after five days with no signs of complication.

In most cases, a cavernous hemangioma of the thyroid gland represents a secondary hemangioma occurring due to previous FNAC. These are formed due to vascular proliferation in organized hematoma following FNAC [4, 5]. Organization of the hematoma generally results in complete resolution, but it can give rise to vascular and fibroblastic proliferative changes that resemble a cavernous hemangioma. This is defined as secondary hemangioma.

Primary hemangioma is a developmental anomaly resulting from inability of angioblastic mesenchyma to form canals [6]. Preoperative diagnosis of hemangioma of the thyroid is difficult since there are no pathognomic features on FNAC, ultrasonography or computed tomography scans. The presence of heterogenous signal intensity and serpentine pattern on magnetic resonance imaging (MRI) scan is considered highly suggestive of cavernous hemangioma [7]. In patients with a thyroid swelling who have a cold nodule on thyroid scan and only blood is aspirated on repeated FNACs, Technetium-99m erythrocyte blood pool imaging may be performed to diagnose hemangioma. Little or no increased activity is
seen soon after injecting the label, and this appearance of poor perfusion and slow filling of the tumor is characteristic of cavernous hemangioma. Hemangioma should be considered in the diagnosis of any pulsatile mass involving the thyroid gland. Diagnosis before surgery is difficult and the definite diagnosis relies on histological findings of surgical specimen.

Figure 1: Ultrasonographic image of the left lobe of thyroid showing 2.4x2.6 cm well defined isoechoic lesion with hypoechoic rim and a large prominent vessel within having arterial flow (marked with arrow).

Figure 2: (A) The thyroid nodule shows dilated, congested varying sized anastomosing vascular channels (H&E stain, x20), (B) A few scattered colloid filled thyroid follicles amidst the vascular channels (H&E stain, x40).

REFERENCES


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

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Conflict of Interest
Authors declare no conflict of interest.

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