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Fatal outcome in a case of infective endocarditis due to delay in diagnosis and the autopsy findings

Junji Matsuda, Ryo Kojima, Yutaka Matsumura, Junichi Nitta

ABSTRACT

Introduction: Infective endocarditis is an infection of a heart valve or other cardiac structure at a site of endothelial damage. Infective endocarditis is associated with a broad array of complications and has a high fatality rate. Some autopsy case reports regarding causes of death have previously been published. These cases reported that the condition of patients with infective endocarditis worsened drastically, and they suddenly died from fatal complications, such as acute severe heart failure, cerebrovascular major embolism, and hemorrhage or acute coronary embolism. In these cases, early diagnosis did not necessarily have an important role because of the unpredictable and relatively rare situation. Case Report: A 47-year-old male was transferred to our hospital due to fever and severe fatigue. Although he had been experiencing symptoms for several months and visited doctors near his home several times, his condition was not diagnosed and he took antimicrobial drugs for a few days. On admission, he presented with typical clinical findings of infective endocarditis. Although intensive medical treatment was administered, his severe infection was not controlled, and it led to multi-organ failure. On day-15 of hospitalization, the patient succumbed to multi-organ failure. An autopsy also revealed multi-organ failure. Conclusion: This case illustrates the fatal outcome of undiagnosed infective endocarditis in a patient who did not have fatal complications, such as acute heart failure, cerebrovascular major embolism, and hemorrhage or acute coronary embolism, which have been reported in past cases. In this case, early diagnosis could have avoided the unfortunate outcome. We report this case to emphasize the importance of early diagnosis of infective endocarditis.

Keywords: Autopsy, Early diagnosis, Infective endocarditis, Multi-organ failure

INTRODUCTION

Infective endocarditis is an infection of a heart valve or other cardiac structure at a site of endothelial damage. The definition of infective endocarditis has been also expanded to include infected cardiac devices. A variety of
organ systems may be adversely affected in patients with infective endocarditis. Although advances have improved the diagnostic accuracy, infective endocarditis has relatively high morbidity and mortality rates from 16–25% of affected individuals [1–4]. Infective endocarditis is associated with a broad array of complications; cardiac, neurologic, renal, musculoskeletal, and systemic complications related to the infection (embolization, metastatic infection, and aneurysm) [5, 6]. Some autopsy case reports regarding causes of death have previously been published. These cases reported that the condition of patients with infective endocarditis worsened drastically, and they suddenly died from fatal complications, such as acute severe heart failure, cerebrovascular major embolism, and hemorrhage or acute coronary embolism [6–13]. In these cases, early diagnosis did not necessarily have an important role because of the unpredictable and relatively rare situation. We report a case of a 47-year-old male with infective endocarditis; early diagnosis could have avoided his unfortunate death. We report this case to emphasize the importance of early diagnosis of infective endocarditis.

**CASE REPORT**

A 47-year-old male was transferred to our hospital due to fever and severe fatigue. Although he had been experiencing symptoms for several months and visited doctors near his home several times, his condition was not diagnosed and he took antimicrobial drugs for a few days. On admission, he exhibited a systolic murmur and moderate mitral regurgitation with large vegetation on echocardiographic findings (Figure 1). *Streptococcus mitis* was detected in his blood culture and he was diagnosed with infective endocarditis. He also exhibited Janeway lesion, petechial hemorrhage on his conjunctiva and Roth spots on his fundus (Figure 2). His clinical presentation is typical. Computed tomography scan and magnetic resonance imaging scan revealed small multiple embolization to the brain, spleen, and kidney. Cardiac surgery was too risky because he also had severe coagulopathy and his mitral regurgitation was not so severe as needed the surgery. Although intensive medical treatment was administered, his severe infection was not controlled, and it led to multi-organ failure. On day-15 of hospitalization, the patient succumbed to multi-organ failure. Autopsy revealed huge vegetation (30×15×5 mm) in the two-thirds around the mitral valve (Figure 3). Histological analysis demonstrated that the vegetation was composed of necrotic tissue and neutrophils, along with numerous Gram-positive cocci (Figure 4) that destroyed the tissue of the mitral valve and the wall of the left atrium (Figure 5). An autopsy also revealed embolization of kidney and spleen and necrotic intestines, which had caused multi-organ failure.
DISCUSSION

This case illustrates the fatal outcome of undiagnosed infective endocarditis in a patient who did not have fatal complications, such as acute heart failure, cerebrovascular major embolism, and hemorrhage or acute coronary embolism, which have been reported in past cases. Infective endocarditis is associated with a broad array of complications; cardiac, neurologic, renal, musculoskeletal, and systemic complications related to the infection (embolization, metastatic infection, and aneurysm) [5, 6]. Some autopsy case reports regarding causes of death, which are due to fatal complications, have previously been published [7, 10, 11, 14]. In this case, although the patient did not have fatal complications which have been reported in past such cases, he subsequently died as a result of this uncontrollable infection. On admission, intensive medical treatment was administered; antibiotics, renal replacement therapy and blood transfusion. He also had disseminated intravascular coagulation, which is one of the complication of infective endocarditis [15]. Cardiac surgery was too risky because of severe coagulopathy. We think he had best medical treatment in hospital. In present case, delay in diagnosis of infective endocarditis caused his unfortunate death. Clinical presentation on admission was relatively typical. Conservative antimicrobial treatment for undiagnosed fever causes delays in the diagnosis of infective endocarditis. Once infective endocarditis has progressed it is too late to control the infection and recover systemic organ failure even though fatal complications are not a problem. In this case, early diagnosis could have avoided the unfortunate outcome. We report this case to emphasize the importance of early diagnosis of infective endocarditis.

CONCLUSION

Delays in the diagnosis of infective endocarditis resulted in a fatal outcome despite absence of fatal complications such as acute heart failure, cerebrovascular major embolism, and hemorrhage or acute coronary embolism. Early diagnosis of infective endocarditis is crucial.

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Conflict of Interest
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REFERENCES

A case of parastomal pyoderma gangrenosum

Zexi Allan, Alex Wong

ABSTRACT

Parastomal pyoderma gangrenosum is a rare condition where classical skin lesions are found around or near abdominal stomas. Literature reports a 2–4.3% prevalence of parastomal pyoderma gangrenosum in patients with inflammatory bowel disease who undergo stoma surgery. This debilitating skin condition is often missed or wrongly treated due to lack of awareness and understanding. We present a 51-year-old female with parastomal pyoderma gangrenosum on a background of Crohn’s disease. Current literature of diagnosis and management is also discussed.

Keywords: Colorectal surgery, Inflammatory bowel disease, Pyoderma gangrenosum, Stoma

INTRODUCTION

Pyoderma gangrenosum is a type of neutrophilic dermatosis which causes progressive painful cutaneous ulcerations. Half of the cases are found to be associated with underlying systemic diseases, such as inflammatory bowel disease (IBD) [1]. Pyoderma gangrenosum can affect any age, with an average onset between 40–60 years, and females are more common affected than males [2]. Parastomal pyoderma gangrenosum is a rare subset where classical skin lesions are found around or near abdominal stomas. Literature reports a 2–4.3% prevalence of parastomal pyoderma gangrenosum in IBD patients who undergo stoma surgery [3]. This is a debilitating skin condition which is often missed or wrongly treated. Timely recognition and appropriate treatment can significantly improve patients’ quality of life.

CASE REPORT

A 51-year-old female with Crohn’s disease presented electively for reversal of loop ileostomy. She previously underwent subtotal colectomy with ileorectal anastomosis and loop ileostomy six months ago for Crohn’s colitis.

The patient had ongoing issues with her stoma since surgery. She was admitted two weeks postoperatively with suspected parastomal wound infection with severe pain and purulent discharge. Abdominal computed tomography scan showed no collection. She was discharged home with oral antibiotics. Since then, she had constant stoma leakage and pain around stoma site. However, parastomal skin remained intact. Outpatient review five months postoperatively indicated that the parastomal skin started to show signs of disease with circumferential ragged edges, induration and severe...
tenderness to touch. Patient also reported occasional fluid discharge from pinhole breaks of the surrounding skin. Crohn’s associated pyoderma gangrenosum was suspected by stomal therapist, however patient was not referred for medical assessment and was managed with stoma care only.

Pain and local skin changes continued to the time of her reversal procedure. At the time of surgery, her stoma was retracted, with parastomal skin erythema, induration, ulceration and breakdown (Figure 1) and it was very tender to touch around the stoma site. Reversal surgery was performed and skin biopsies were only taken post parastomal skin debridement and curettage. Subsequent histopathology showed chronic inflammation along with reactive epidermal hypertrophy and some fibrosis; features of pyoderma gangrenosum were not seen. Despite the biopsy results, we highly suspect parastomal pyoderma gangrenosum given the clinical appearance and severe pain.

**DISCUSSION**

Pyoderma gangrenosum typically presents as cutaneous ulceration with a well-defined, undermined violaceous border [4]. This can be mistaken for other causes such as infection, stitch abscess, contact dermatitis and irritation from fecal material [1]. Missed diagnosis occur also due to lack of knowledge and familiarity of clinicians to parastomal pyoderma gangrenosum [5]. It is often a diagnosis of exclusion and is particularly difficult to manage owing to its constant pain resulting in poor application of stoma appliance. Correct recognition and appropriate management is essential to improve patients’ quality of life.

There are no absolute diagnostic criteria for pyoderma gangrenosum. While histology from skin biopsy typically show edema and massive neutrophil infiltration confined to the dermis, diagnosis of parastomal pyoderma gangrenosum is mainly via its clinical appearance, a high degree of pain and its rapid progression. The ulcers are very destructive and can expand by 1 to 2 cm from hours to a day [1, 5]. Biopsies are done often to rule out other causes of ulceration rather than to diagnose pyoderma gangrenosum histologically [3]. One study from the Mayo clinic [6] put together a diagnostic guideline; as given in Table 1. The diagnosis is strongly indicated when both major criteria and at least two minor criteria are met.

Once diagnosis is suspected or other causes have been excluded, appropriate management can significantly improve stoma care and patient’s quality of life. A multidisciplinary approach is encouraged involving gastroenterologist, dermatologist, colorectal surgeons and stoma therapist [3]. Medical rather than surgical management is preferred as debridement is not recommended and relocation of stoma is contraindicated unless it is done for other reasons [3]. Given the nature of pathergy, debridement will only exacerbate the problem and relocation often result in recurrence of the disease at the new stoma site [1, 5]. Treatment regimen depends on the severity of the disease and whether there is active systemic disease (i.e., IBD). For mild cases, wound management with moisture-retentive dressings plus topical steroids or intralesional injection of triamcinolone or cyclosporine has been shown to be effective [3]. Systemic therapy is indicated when there is lack of response to topical therapy, if the disease is severe and rapid or if there is active underlying disease.

**CONCLUSION**

Parastomal pyoderma gangrenosum is a rare but debilitating condition and prompt recognition, diagnosis
and management is required to improve stoma care and patients’ quality of life.

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Author Contributions
Zexi Allan – 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
Alex Wong – 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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The corresponding author is the guarantor of submission.

Conflict of Interest
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REFERENCES
Streptococcus pneumoniae induced purpura fulminans in a patient with splenic hypoplasia

Shunsuke Kojima, Eiji Hiraoka, Jun Ehara, Toshihiko Suzuki, Enichi Nakatsuru, Yasuhiro Norisue

ABSTRACT

Introduction: Purpura fulminans (PF) is a rare, life-threatening medical emergency requiring prompt diagnosis and treatment. Common causes include Neisseria meningitidis, Streptococcus pneumoniae (S. pneumoniae), Haemophilus influenzae, Staphylococcus aureus, and fungal or viral infections. It usually occurs in immunocompromised hosts. We experienced a rare case of purpura fulminans due to S. pneumoniae, who had no history of immunosuppressive disease. Case Report: A 57-year-old male was presented to our emergency department in shock state with flu-like symptoms. Empirical broad-spectrum antibiotics and intensive care were started. His condition rapidly deteriorated with multiple organ failure. Blood culture grew up S. pneumoniae. Purpuric skin change developed in all extremities followed by ischemic gangrene, which required amputation. He did not have any history of immunosuppressive disease. His computed tomography scan of abdomen showed small size of spleen. Howell–Jolly bodies were recognized in peripheral blood smear. The patient was finally diagnosed with purpura fulminans with overwhelming pneumococcal sepsis. Although he had no history of immunodeficiency, he had evidence of Howell–Jolly bodies in peripheral blood smear, implying reduced splenic function, possibly due to splenic hypoplasia. To prevent this devastating condition, vaccination against S. pneumoniae may need to be considered for people with splenic hypoplasia. Conclusion: Since delay in therapy would lead to a poor outcome, clinicians should be alert to purpura fulminans in patients in shock state, even lacking typical skin manifestation initially. Splenic hypoplasia may be a risk factor of this condition.

Keywords: Purpura fulminans, Streptococcus pneumoniae, Erythrocyte inclusions, Splenic hypoplasia

INTRODUCTION

Purpura fulminans is a rare, life-threatening syndrome, requiring early diagnosis and treatment.
Clinical manifestations include four primary features: large purpuric skin lesions, fever, hypotension, and disseminated intravascular coagulation (DIC) [1, 2]. Common causes include Neisseria meningitidis, Streptococcus pneumoniae (S. pneumoniae), Haemophilus influenzae, Staphylococcus aureus, and fungal or viral infections [2, 3]. Purpura fulminans occurs mainly in immunocompromised hosts including those with prior splenectomy, functional asplenia, or other impaired host-immune defense mechanisms [4, 5]. It is rare for immunocompetent adults to have this condition. There have been only two reported cases of immunocompetent adult cases with purpura fulminans [6]. Herein, we report a case of purpura fulminans due to S. pneumoniae infection in an adult with no history of immunsuppressive diseases.

CASE REPORT

A 57-year-old previously healthy male presented to the emergency department with a three-day history of nonspecific symptoms, including fever, malaise, and cough. His vital signs and physical examination results were normal. Blood tests showed no abnormalities except leukocytosis. He was discharged with a presumptive diagnosis of common cold. However, the next day, he was brought to the emergency department after experiencing a syncopal episode. Vital signs on admission were as follow: temperature 38.7°C (101.7°F), blood pressure 73/47 mmHg, pulse rate of 132 beats per min, respiratory rate of 24 breaths per min, and arterial oxygen saturation level of 96% while breathing ambient air. He was alert and oriented. There was no nuchal rigidity. Mottled skin was noted on the arms, chest, abdomen, and pretibial areas. Findings from lung, heart, and abdomen examinations were normal.

Initial laboratory tests revealed thrombocytopenia 43×10^9/L, a normal white blood cell count (7.2×10^9/L), and a normal hemoglobin level (13.6 g/dL). Coagulation studies showed findings consistent with DIC, namely a prothrombin time of 12.3 s, partial thromboplastin time of 53.5 s, fibrin degradation product concentration of 69.7 µg/mL, and D-dimer concentration of 21.4 µg/mL. Liver and renal function tests revealed an aspartate aminotransferase level 150 U/L, alanine aminotransferase level 35 U/L, total bilirubin level 1.58 mg/dL, blood urea nitrogen level 40.7 mg/dL, and creatinine level 3.09 mg/dL, which were indicative of the multiple organ dysfunction syndrome. The serum troponin I level was normal on admission, but was elevated to 6.35 ng/mL within several hours. Arterial blood gas analysis detected an anion gap metabolic acidosis with an elevated lactic acid level of 33.80 mg/dL. Serological test results for HIV, hepatitis B virus, hepatitis C virus, and syphilis were negative. Chest radiography showed no pathological findings, however, computed tomography of the abdomen revealed a small spleen, 61 cm³ in size (Figure 1). Erythrocyte inclusions (Howell–Jolly bodies) were observed on the peripheral blood smear (Figure 2). An electrocardiogram demonstrated sinus tachycardia with no ST-T changes. Echocardiography showed diffuse hypokinesis with an ejection fraction of 18% without vegetation, or valvular disease. Based on the patient’s fever and shock state, as well as his poor cardiac function and elevated troponin, septic shock and cardiogenic shock were considered in the differential diagnosis.

Fluid resuscitation and norepinephrine were administered immediately, followed by endotracheal intubation and mechanical ventilation. Therapy with vancomycin and meropenem was empirically initiated after blood cultures were obtained. Cardiac catheterization confirmed the absence of any critical coronary artery disease. Myocardial biopsy revealed normal findings on microscopy. Intra-aortic balloon pumping was begun for possible cardiogenic shock. Renal failure was managed with continuous renal replacement therapy. Nine hours after admission, blood culture showed the growth for gram-positive cocci and the antibiotic regimen was changed to vancomycin and ceftriaxone. Initial resuscitative management resulted in a reduction of the serum lactate levels and hemodynamic stabilization. Norepinephrine was subsequently discontinued. Intra-aortic balloon pumping was stopped after 48 hours, and continuous renal replacement therapy was replaced with intermittent hemodialysis. Two days after admission, S. pneumoniae was identified in blood culture, and a diagnosis of invasive pneumococcal disease was made. The patient was weaned off the ventilator on day-7.

The patient developed painful ischemic lesions on both his hands and feet during the week after admission, which gradually spread to his forearms and lower legs (Figure 3A–B). By day-5, the peripheral arteries, including the bilateral radial, dorsalis pedis, and posterior tibial arteries, were not palpable. Portions of the skin lesions became vesiculated and edematous, producing hemorrhagic bullae. Gradually, the lesions became more consolidated with dark-colored well-demarcated hemorrhagic necrosis (Figure 3C–D). These findings were consistent with purpura fulminans, due to S. pneumoniae. Seven weeks after admission, below-elbow amputations and below-knee amputations were performed bilaterally. After a four-month hospital stay, the patient was transferred to another hospital for further rehabilitation.

DISCUSSION

Purpura fulminans was first reported by Guelliot in 1884 [7], and is a life-threatening condition characterized by symmetric peripheral gangrene with large purpuric skin lesions, fever, hypotension, and DIC that requires early diagnosis and treatment. Various pathophysiological mechanisms contribute to the formation of the necrotizing inflammatory lesions, and purpura fulminans carries a
The most common causative agents of purpura fulminans are *N. meningitides* infections, followed by varicella, *S. pneumoniae*, and measles infections [8]. Reduced splenic function, asplenism, and protein S or C deficiency can also be risk factors for this condition [2, 4, 5]. The skin lesions usually start as well-demarcated erythematous macules, which worsen rapidly with hemorrhagic necrosis, followed by the formation of dark lesions with vesicles or bullae. The differential diagnosis of purpura fulminans includes idiopathic thrombocytopenic purpura, thrombotic thrombocytopenic purpura, Henoch–Schönlein purpura, and warfarin-induced skin necrosis [9]. Usually, only purpura fulminans and warfarin-induced skin necrosis present with necrotic skin lesions [5].

Currently, there is no standard treatment for purpura fulminans caused by sepsis, and intensive care is the main therapeutic strategy. In addition to aggressive fluid resuscitation, prompt initiation of empirical broad-spectrum antibiotics for underlying sepsis, correction of acid-base imbalance and electrolyte abnormalities, and the early administration of oxygen are also helpful [10, 11]. Heparin may be administered to inhibit further thrombus formation, and may reverse the development of skin necrosis [12]. Because the pathophysiology of purpura fulminans involves intravascular thrombosis, fresh frozen plasma can be used to replete these coagulation factors. Replacement therapy may contribute to arresting the progression of the disease and avoiding amputation of the limbs [5]. Early administration of protein C corrects the deficiency and might contribute to the restoration of peripheral perfusion according to a previous case report [5].

As in our case, the first manifestation of purpura fulminans may be a non-specific flu-like illness, with fever or chills, sore throat, malaise, and occasionally gastroenteritis symptoms, which occur 12–24 hours before the development of purpura fulminans [13, 14]. Therefore, clinicians should be cognizant of purpura fulminans as a differential diagnosis in patients in a shock state, with non-specific flu-like symptoms, and consider empirical antibiotic treatment, even in patients who initially lack the characteristic skin manifestations associated with purpura fulminans.

Patients with asplenism and reduced splenic function are particularly at risk of sepsis. Functional asplenia or hyposplenia can result from splenectomy or various splenic conditions, such as congenital absence, atrophy following repeated infarction (e.g., sickle cell disease), gastrointestinal diseases, hepatic disorders, autoimmune disorders, hematological disorders, and neoplastic disorders [15]. Although scintigraphic methods are most reliable for assessing splenic function, they are not the best options for screening large populations [16]. The presence of Howell-Jolly bodies, which are small round bodies representing nuclear remnants within erythrocytes, indicates splenic dysfunction, although these findings may not be seen in those with only mild impairment of splenic function [5, 16]. Other abnormalities associated with splenic dysfunction that can be seen on peripheral blood smears are acanthocytes (spur cells), target cells, hemoglobin remnants (Heinz bodies), siderocytes, and
iron granulocytes [16]. In our case, Howell–Jolly bodies were observed on peripheral blood smear. The mean splenic length and width in healthy populations are 10.8 cm, and 3.6 cm, respectively [17], and the average volume is 131 cm³ [18]. Our patient’s splenic volume was 61 cm³ (6.2 cm long and 3.3 cm wide), which is small according to previous studies. Although there have been no studies on the association between splenic hypoplasia and dysfunction, there have been case reports of purpura fulminans due to S. pneumoniae associated with splenic hypoplasia [19–21]. It is rare for immunocompetent adults to have purpura fulminans, and only two such cases have been reported in literature [6]. Although our patient was relatively healthy until diagnosis, he was found to have a degree of splenic dysfunction, possibly due to splenic hypoplasia.

The Centers for Disease Control and Prevention recommends the administration of pneumococcal vaccines for asplenic patients; this vaccine protects patients against 73–90% of strains causing post-splenectomy infections [22]. Other guidelines also recommend that patients with asplenia or hyposplenia be immunized against organisms including S. pneumoniae, H. influenzae type b, and N. meningitidis [22, 23]. When a person is incidentally found to have splenic hypoplasia, vaccination against S. pneumoniae to prevent the devastating disease of purpura fulminans may need to be considered.

With deep and extensive skin damage, surgical intervention including fasciotomy, debridement, and limb amputation are possible options. Some reports [8, 24] suggest that prompt surgical consultation for the indications of intervention, debridement, and amputation may reduce the risk of mortality because critical complications, including the compartment syndrome, can occur in up to 7% of purpura fulminans cases, leading to increased morbidity [25]. Conversely, Johansen et al. do not recommend early surgical intervention because the damaged skin area is eventually localized [11]. In our case, it was difficult to determine the extent of necrosis at an early stage because the skin lesion margins were indistinct. After the patient’s general condition stabilized, the margins became apparent. Additional studies are needed to evaluate whether early surgical intervention is necessary to save the patient’s life, or if it is better to wait for a clearer demarcation of the necrotic areas.

**CONCLUSION**

A non-specific flu-like illness may be the first manifestation of purpura fulminans. Clinicians should carefully observe patients to make a timely diagnosis and initiate treatment for purpura fulminans, even in patients lacking the typical signs of purpura fulminans. In patients with splenic hypoplasia, vaccination against S. pneumoniae may need to be considered to prevent this devastating condition.

**REFERENCES**


Percutaneous drainage of delayed traumatic subcapsular hematoma of the spleen following splenic salvage: A case report

Alaa Sedik, Mahmood Makhdoomi, Abrar Hussein, Salwa Elhoushy, Ahmed Morsy

ABSTRACT

Introduction: Percutaneous image-guided splenic procedures are seldom performed due to fear of complications, mainly hemorrhage. The reported cases in literature were due to atraumatic causes. These procedures obviate the need for splenectomy. Thus, preserving the spleen and decrease the pressure to avoid possible rupture. Case Report: We report a 30-year-male with large delayed traumatic subcapsular splenic hematoma after splenic salvage that was managed successfully by percutaneous ultrasound-guided drainage. Conclusion: In stable patients, image-guided percutaneous drainage appears to be another feasible option for large subcapsular traumatic splenic hematomas to prevent splenic rupture and obviate the need for splenectomy.

Keywords: Hematoma, Percutaneous drainage, Spleen

INTRODUCTION

Splenic salvage is less commonly practiced nowadays. Delayed splenic rupture is mostly due to ruptured hematoma and carries higher mortality (5–15%) than acute (1%) [1–3]. The management of non-traumatic subcapsular splenic hematoma remains controversial. Splenectomy or percutaneous drainage under ultrasound guidance may be done. No reports are available in the literature on percutaneous drainage of traumatic hematoma. We herein reported a 30-year-old male, a victim of road traffic accident (RTA), with large traumatic subcapsular splenic hematoma after splenic salvage that was managed successfully by percutaneous ultrasound-guided drainage.

CASE REPORT

A 30-year-old Indian male presented to our emergency room, a RTA victim, with lower abdominal pain. Resuscitation started according to ATLS protocol and examination was unremarkable except for suprapubic tenderness, tear of glans penis and right side of scrotum. Laboratory workup showed raised renal functions.
Focused assessment with sonography for trauma (FAST) showed mild free intraperitoneal fluid. X-rays showed bilateral lung contusions and pelvic fracture. Computed tomography (CT) scan without contrast was not done due to raised serum creatinine of 145 μmol/L and urea of 11 mmol urea per liter. Scrotal ultrasonography and Doppler were normal. Repeated hemoglobin was 8 g/dl, then ultrasound showed increased intra-abdominal fluid. He was prepared for laparotomy. 1.2 liters of free blood, normal solid organs except for 1 cm lateral capsular splenic tear with minimal bleeding were found. Splenic salvage was done with cautery and 2 pieces of pieces of local hemostats (Surgicel™). Resection anastomosis of 30 cm of ilium was done for transverse large mesenteric tear of the distal ileum. Wash with saline then, the abdomen was closed over drains. On the next day, he developed hematuria. Computed tomography (CT) scan with contrast showed normal solid organs. On day-10, blood workup and ultrasound were unremarkable. Drains removed and antibiotics were discontinued. He remained for wound infection care. On day-18, he had fever and subcostal pain. Clinically, he had mild left subcostal tenderness with no masses. Ultrasonography and abdominal contrast enhanced CT scan showed a subcapsular 12x6.5x3.6 cm splenic hematoma with no free intraperitoneal fluid, left pleural effusion and basal lung consolidation (Figure 1). Conservative treatment continued for 10 days and till complaining of pain. Follow-up CT scan showed no improvement. He was offered ultrasound-guided percutaneous drainage or splenectomy as alternative in case of failure of drain. 40 ml of turbid blood was aspirated and sent for culture. A 16 F pigtail catheter was left inside the cavity. Post procedure chest X-ray showed a left hydro pneumothorax that improved later (Figure 2). The culture of the aspirate was negative. He did well. Regular catheter care with flushing with 5 ml saline. On day-9, the drainage was reduced to 5 ml and serous. The catheter drainage stopped after three days. Ultrasonography showed obliteration of the cavity. The catheter was removed. He was discharged in a good condition and came for outpatient department follow-up three weeks later and was free of complaints. He did not agree to do follow-up CT scan as he had it three times during the course of treatment. CBC and ultrasonography were normal. He has regular follow-up.

**DISCUSSION**

The spleen is one of the most commonly injured intra-abdominal organs mostly due to blunt trauma [1]. The preservation of functional splenic tissue is secondary and in selected patients may be accomplished using non-operative management with or without splenic angioembolization or operative salvage techniques. Splenectomy remains a life-saving measure for many

Figure 1: (A) Abdominal computed tomography scan with contrast, (B) A large splenic subcapsular together with left pleural effusion and basal consolidation, and (C) Follow-up scan, (D) Showed no improvement.

Figure 2: (A) The catheter showed altered aspirated blood, (B) A mild a left hydro pneumothorax, percutaneous catheter drain and contrast in the colon from previous computed tomography scan are shown in the post-procedure chest X-ray, and (C) A discharge chest X-ray.
patients and may be a part of damage control surgery [2]. The American Association for the Surgery of Trauma classified splenic injuries using CT scan [3, 4]. The decision to perform splenectomy versus splenic salvage (i.e., splenorrhaphy, partial splenectomy, electrocautery or topical hemostats or argon beam) is based upon the grade of injury, presence of associated injuries, patient’s overall condition, and experience of the surgeon. Our patient was stable intraoperatively and had a low grade(I), then a salvage with cautery of the bleeding point and topical hemostat done.

Splenectomy was extensively practiced before. Stable patients with lower grades are managed non-operatively. Failure will require splenectomy. All these factors have made splenectomy a rare procedure and the surgeon experience has declined [5, 6]. Percutaneous drainage of splenic subcapsular collections may be a feasible and successful treatment, but there are only few reports available in literature. They discussed the atraumatic and spontaneous hematomas complicating mostly acute or chronic pancreatitis [7–9]. To the best of our knowledge, this is the first case reported in the literature with a traumatic splenic subcapsular hematoma treated successfully with this modality that needs further investigation and work in future.

CONCLUSION

The definitive management of subcapsular splenic hematoma complicating trauma is not yet established. Surgery is the treatment of choice for hemodynamically unstable patients. Follow-up of patients with splenic injury treated non-operatively or after salvage; after discharge may detect a serious complication and may avoid delayed rupture. Image-guided percutaneous drainage appears to be another feasible option for large subcapsular traumatic splenic hematomas to prevent splenic rupture and obviate the need for splenectomy.

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Breast angiosarcoma: A case report

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ABSTRACT

Introduction: Breast angiosarcomas are rare but aggressive malignant endothelial cell tumors. There are two entities: primary angiosarcoma and secondary angiosarcoma to breast irradiation. This neoplasia is characterized by absence of typical features at radiological examination. Case Report: We report a case of a 40-year-old female with a history of invasive ductal breast carcinoma presented with a bilateral breast angiosarcoma. The main treatment is a large surgery with free tumor margins. The role of adjuvant chemotherapy and radiotherapy is still a subject of debate. For inoperable and metastatic disease, chemotherapy is the pillar of treatment. Conclusion: The prognosis of breast angiosarcoma is still poor. Many prospective trials are needed to establish more clear therapeutic strategies.

Keywords: Breast angiosarcoma, Chemotherapy, Post-irradiation, Radiotherapy, Surgery

INTRODUCTION

Angiosarcomas are rare and aggressive blood vessel tumors with high recurrence rates and poor overall survival. Due to the increasing use of radiation therapy in breast cancer, secondary neoplasia is induced. The most common secondary neoplasia of the breast is angiosarcoma. It occurs 4–7 years after radiation therapy. A primary neoplasia can also be observed but less commonly. No evidence-based guidelines exist concerning the ideal treatment of angiosarcoma. Radical surgical resection of the tumor with a sufficient margin of safety is the treatment of choice for angiosarcoma.

CASE REPORT

On September 2007, a 40-year-old woman underwent breast-conservative surgery and axillary dissection for invasive ductal carcinoma of the right breast. Immunohistochemical analysis revealed estrogen and progesterone receptor positivity. The tumor was classified as pT2 pNo M0 SBR 3 according to the UICC–TNM classification. She received four cycles of adjuvant chemotherapy (adriamycin 60 mg/m² and cyclophosphamide 600 mg/m² every 21 days). She had radiotherapy as follows: 50 Gy for the right
breast and chest wall in 25 fractions of 2 Gy/daily with boost of 14 Gy in 7 fractions of 2 Gy/daily. She received adjuvant hormone therapy (tamoxifen 20 mg daily for five years). The patient did not suffer from chronic lymphedema.

On January 2016, we observed a painful, bluish and nodular mass occupying the interne quadrants of the right breast. On examination the size of lesion was 4 cm. An excisional biopsy was performed. Histopathological examination revealed a vascular tumor proliferation. Tumor cells were positive for CD34, CD31, factor VIII and negative for EMA, CK, CD68, ER and PR, indicating an endothelial origin. More than 50% of the cells in the solid component were positive for Ki67. The diagnosis of cutaneous grade-II angiosarcoma of the breast was made. It was probably a radiation–induced angiosarcoma.

No metastases were found at total body computed tomography scan. A right mastectomy was performed. Histopathological examination revealed a grade-II angiosarcoma measuring 4 cm of long axis that ulcerated the skin and dissociated the breast parenchyma. In addition to that, a low grade angiosarcoma measuring 7 mm was identified as a skin lesion. Margins of resection were tumor-free. Adjuvant chemotherapy and radiotherapy were not prescribed.

On October 2016, cutaneous angiosarcoma recurred on the inner end of the right mastectomy’s scar as a 1 cm nodular mass. An excisional biopsy was performed confirming the recurrence.

A body computed tomography scan showed three left axillary lymphadenopathies. Mammography and ultrasound mammary of the left breast did not show any lesion except axillary lymphadenopathies. Left breast magnetic resonance imaging scan showed in addition to lymphadenopathies, a 10 mm nodular lesion isointense on T1 images and hypo-intense on T2 with intense and early contrast enhancement. This lesion was located in the left upper outer breast’s quadrant. The tumor was classified as T1 N1 Mo. An excisional biopsy of the left axillary lymphadenopathy was performed. Histopathological examination revealed an axillary localization of an angiosarcoma.

A left mastectomy and axillary node lymph dissection were performed. Histopathological examination revealed a grade-II angiosarcoma measuring 5 cm. The tumor proliferation invaded the dermis and hypodermis and reached the muscular layer. Margins of resection were tumor-free. Five nodes were metastatic from nine. Four nodes had a capsular rupture. Macroscopic and microscopic aspects of tumor are shown in Figure 1 and Figure 2.

The patient received three cycles of adriamycin and ifosfamide with appearance of liver metastases and deterioration of general condition. The patient passed away on March 2017.
Angiosarcomas are highly aggressive and malignant tumors which arise from endothelial cells lining vascular channels. They usually develop on the head and neck [1]. Breast angiosarcomas are exceedingly rare accounting for less than 1% of malignant breast tumors [2]. Breast angiosarcoma can be either observed as primary tumors or, more commonly, secondary to irradiation for breast carcinoma [2]. In 1929, radiation induced angiosarcoma was first reported in literature [3]. It represents 0.04% of all breast tumors. The diagnostic criteria for radio-induced angiosarcoma include a previous history of radiotherapy, peak incidence between 5 and 10 years, development of sarcoma within a previous irradiated field and histology confirmation [4]. It is characterized by an aggressive nature. The prognosis is poor and local recurrence rates reaches 70% after mastectomy [4].

In a series of 55 cases of breast angiosarcoma, 42% of patients were irradiated. These patients were on average 30-year-old and less likely to present with distant metastatic disease than patients presenting with primary breast angiosarcoma. Radiation-naïve angiosarcoma mainly occurs in 3–4th decade [5]. Twenty-one percent of Bilateral cases are reported. It may be either really primitive bilateral forms or early contralateral metastases [6]. In this report, the patient presented a bilateral breast angiosarcoma. The right breast angiosarcoma is probably secondary to radiation of right breast. The left breast angiosarcoma can be either a metastasis of the right one or secondary to radiation of the chest wall with no protection of left breast.

Clinically, the lesion presents as a rapidly growing, painless breast mass within a previous irradiated field. The overlying skin may be blue or purple [7, 8]. Lymph node involvement is rare. However, the risk increases in locally advanced tumor [9]. The mammography does not suggest a tumor and this is typical of angiosarcoma [8] due to its superficial localization. Ultrasound mammography's findings include ovoid shape, hyperechoic or heterogeneously echoic solid masses associated with architectural distortion. The vascular nature of angiosarcoma can account for the hyperechogenicity feature. Hyperechogenicity is remarkable since most of the breast carcinomas are very rarely hyperechoic [10]. Advanced imaging modalities like MRI scan can bring a profit [11]. Pathologists should keep this diagnosis in mind when dealing with a breast skin lesion in a patient with a previous history of breast cancer and radiation therapy. Microscopically, angiosarcoma composed of vascular channels lined by proliferated endothelial cells with atypical and hyperchromatic nucleus.

Immunohistochemical stains for epithelial markers (pancytokeratin), endothelial markers (CD34 and CD31) and other sarcoma markers should help in making the correct diagnosis [12]. The SEER database revealed that the histologic grade is a significant predictor of survival for patients with localized primary breast angiosarcoma (a study about 226 patients) [13]. However, another study interesting 49 cases of primary angiosarcoma suggested that histologic grade is not prognostic [14]. Due to the rarity of the disease, prospective studies concerning adjuvant, neoadjuvant or palliative therapy are limited and no evidence-based guidelines exist. The response to chemotherapy seems to be poor.

The main treatment of breast angiosarcoma is early and complete surgical excision of the mass with tumor-free margins. Although R0 resection is performed, the five-year survival rates are 20–30%.

Unless there are palpable nodes, lymph node dissection is useless [15]. Adjuvant chemotherapy that includes doxorubicin for patients with poorly differentiated breast angiosarcoma results in a higher proportion of patients who are relapse-free compared to patients not receiving adjuvant chemotherapy [16]. For locally advanced inoperable or metastatic disease, chemotherapy is the pillar of treatment [17]. The response rate to doxorubicin, the standard frontline chemotherapy for advanced sarcoma as a single agent or in combination, is reported to the range between 40% and 65% [18]. Taxanes can be active, both as single agents and in combination with gemcitabine or with anthracyclines, with response rates between 20% and 65% [19]. Tolerability of gemcitabine plus docetaxel is acceptable, with less cardiac toxicity compared with anthracyclines but still carrying a significant incidence of neutropenia and thrombocytopenia [20].

The MD Anderson Cancer Center study of 69 patients with breast angiosarcoma found that the response rate to anthracycline-ifosfamide or gemcitabine-taxane combination in the metastatic setting (29 patients) was 48% [21]. Anthracyclines alone or with ifosfamide has led to disease control after several months (between 7 and 24 months) [22]. A control of the disease for five months was observed with weekly paclitaxel as a single agent in the initial treatment of unresectable, radio induced angiosarcoma [23]. Radiotherapy is reserved after lumpectomy, and following total mastectomy if the tumor is larger than 5 cm, the margins are positive, or if the skin or regional nodes are affected [24]. Of all breast cancers, angiosarcoma has the poorest prognosis. The median overall survival is 24 months [25].

CONCLUSION

Breast angiosarcoma is a rare entity characterized by poor prognosis despite optimal surgery and systemic
therapy. Many therapeutic strategies are needed to be explored to improve the outcomes.

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**Author Contributions**

Ayadi Mouna – 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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**REFERENCES**

Meckel–Gruber syndrome in Togo: Prenatal ultrasound and computer tomography diagnosis


ABSTRACT

Meckel–Gruber syndrome (MGS) is a rare autosomal recessive lethal disorder involving multiple systems. It is characterized by occipital encephalocele, polycystic kidneys and post-axial polydactyly. We report a case of MGS with occipital meningocele, bilateral enlarged echogenic kidneys, polydactyly, and severe oligohydramnios. The diagnosis was made by antenatal ultrasound and computer tomography scan. Meckel–Gruber syndrome is a syndrome rarely described in Africa. Its diagnosis is based on classic triad anomaly. None of this triad seems to be constant.

Keywords: Meckel–Gruber syndrome, Ultrasound, Computer tomography, Togo

INTRODUCTION

Meckel-Gruber syndrome (MGS) is a rare and lethal congenital polymorformative syndrome. It is a genetic disease, with autosomal recessive transmission, described for the first time in 1822 by Johan Friedrich Meckel [1]. It is characterized by the triad: encephalocele, cystic dysplasia of the kidneys and polydactyly [2]. Meckel-Gruber syndrome may also cause hepatic developmental defects, and pulmonary hypoplasia. Its global incidence ranges from 1 in 13,250 to 140,000 live births [3]. We report a case of MGS in Togo, whose antenatal diagnosis was made by ultrasound and completed by scanning.

CASE REPORT

A 20-year-old female G1P0 presented to the obstetric department for the first time, with history of 28 weeks amenorrhea for routine antenatal examination. There was history of third-degree consanguineous marriage. She had her antenatal examination at a peripheral hospital. She was taking iron and folic acid (dosages of iron and folic acid were normal). Ultrasound done at our hospital as a routine, revealed a fetus with occipital meningocele (Figure 1), bilateral enlarged echogenic kidneys (Figure 2), polydactyly, and severe oligohydramnios. We found no liver abnormalities. For the precise analysis of polydactyly, a CT scan of the uterine contents was made. It showed a postaxial polydactyly affecting the four extremities, with six fingers at each hand and six toes at each foot (Figure 3). The scanner also found an occipital bone defect from which the encephalon herniated.

The family was counseled and after getting consent, termination of pregnancy was planned immediately at 28
weeks’ gestation. Post-abortion macroscopic examination revealed multiple congenital anomalies including occipital encephalocele, and post-axial polydactyly.

DISCUSSION

The Meckel–Gruber syndrome, also called dysencephalia splanchnocystica [4], is a rare polymalformative syndrome.

The incidence in Belgium and Finland ranges from 1/3,000–1/9,000, respectively [5]. In India, highest incidence is in Gujarati Indians (1 affected birth per 1,300) [6]. Its incidence is not known in Africa and more particularly in Togo. This is a first discovery in our country. Some cases would probably have gone unnoticed. A high risk (25%) of recurrence in subsequent pregnancies is an important point for consideration and stresses the need for prenatal diagnosis in expectant mothers in those families. Meckel–Gruber syndrome affects all races with males and females being equally affected [4, 7]. Early, (11–14th week) prenatal ultrasonography is the best method to diagnose MGS [7]. Subsequent autopsy and molecular studies are confirmatory. In our case, ultrasonography found this affection at 28th week.

Meckel–Gruber syndrome is associated with multiple anomalies. It is characterized by classic triad of polycystic dysplastic kidneys, occipital encephalocele (or other anomalies of the central nervous system) and polydactyly [2]. All these three anomalies (triad) were found in our case. According to Sergi et al. [8], polycystic kidneys were found in all cases (100%), occipital encephalocele in 90% of case and post-axial polydactyly in 83.3% of cases. Salonen et al. [5] found a constant association between cystic dysplasia and liver fibrosis, and concluded that these two abnormalities, together with any other abnormalities of the nervous system, suffice to diagnose MGS. Fraser et al. [9] argue that only cystic renal dysplasia is essential for diagnosis. Wrigth et al. [10] never observed cystic dysplasia. The lack of consensus for the diagnosis of MGS led some authors to conclude that none of the anomalies

Figure 1: Sonographic intrauterine picture showing defect in the cranium and encephalocele.

Figure 2: Sonographic intrauterine picture showing bilateral dysplastic cystic kidneys.

Figure 3: Computed tomography scan intrauterine image with polydactyly (six digits).
of the triad or liver fibrosis is essential for diagnosis. The CT scan was almost never used for the diagnosis of MGS in literature. However, it can allow bone analysis in cases where the ultrasound is insufficient and the MRI scan is not available. In our case, the CT scan allowed to diagnose polydactyly and occipital defect.

Meckel–Gruber syndrome has to be differentiated from other syndromes. The most likely syndrome to be confused with MGS is trisomy 13 [3]. Although the dismal outcome is the same for both, the recurrence rate is different. Trisomy 13 is mostly sporadic with low recurrence rate whereas MGS has 25% recurrence rate. Other syndromes similar to MGS are trisomy 18, Joubert syndrome, Bardet–Biedl syndrome and Smith–Lemli–Opitz syndrome [3, 11].

The post-mortem assessment confirms the diagnosis, while genetic studies contribute to the evaluation of the recurrence risk [2, 11].

Given the lethality of MGS in the perinatal or early infantile period, termination of pregnancy should be discussed if available [3].

CONCLUSION

Meckel-Gruber syndrome is a syndrome rarely described in Africa. Its diagnosis is based on classic triad anomaly. None of this triad seems to be constant. The isolation of the gene responsible for this syndrome would therefore be necessary for a diagnosis of certainty.

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Peripheral emboli in a patient with atrial fibrillation: A case report

Ismail Tabash, Alexa S. Riester, Rebecca Napier, Joe B. Calkins

ABSTRACT

Introduction: Atrial fibrillation is a common arrhythmia that can be associated with a high risk of thromboembolic complications. Ischemic stroke due to involvement of the cerebrovascular circulation is the most common of these complications. Peripheral emboli may not be as easily recognized and the data regarding their risk, incidence and diagnosis is limited. Case Report: A 46-year-old female presented with exertional dyspnea and right leg pain. Her rhythm was atrial fibrillation. Echocardiogram showed a large left atrial mass and computed tomography angiogram demonstrated filling defects in the left atrium and in the superior mesenteric artery. A peripheral angiogram showed occlusion of the right popliteal artery. The patient underwent right popliteal arterial thrombectomy and excision of the left atrial mass, which was found to be an organized thrombus. Anticoagulation was initiated and continued after discharge. Conclusion: Peripheral emboli may account for 10–20% of embolic events related to atrial fibrillation. They are associated with significant morbidity and mortality and may often go unrecognized unless a high index of clinical suspicion is present. These patients are at high risk for recurrent peripheral embolic events and for future thromboembolic stroke. Lifelong anticoagulation is required. The literature concerning the incidence, risk, diagnosis, management and prognosis of peripheral arterial emboli in atrial fibrillation is reviewed.

Keywords: Arterial, Atrial fibrillation, Peripheral, Thromboembolism

INTRODUCTION

Atrial fibrillation (AF) is the most common arrhythmia with a prevalence ranging from 1.2–2.8% in patients 60–69 years of age to 7.3–13% in those 80 years or older [1]. Atrial fibrillation is associated with a high risk of thromboembolic complications, significantly contributing to disease morbidity and mortality [2]. Ischemic stroke is the most common thromboembolic complication and may occur as the initial presentation of AF despite appropriate anticoagulation. Data regarding
the risk of peripheral embolization is more limited and not as well described in the medical literature. We present the rare case of a patient who presented with simultaneous arterial emboli in two arterial distributions in the setting of atrial fibrillation with rapid ventricular rate. We then review the current literature concerning the incidence, risk, diagnosis and prognosis of peripheral arterial emboli.

CASE REPORT

A 66-year-old female with hypertension and hyperlipidemia presented to the emergency department for evaluation of one week of exertional dyspnea and right leg pain. She was a non-smoker and she had no history of peripheral arterial disease, hyperthyroidism or alcohol use. Upon presentation, she was hemodynamically stable. Physical examination was remarkable for irregular tachycardia, hypertension and diminished right pedal arterial pulses. No cardiac murmurs or focal neurological deficits were noted. Telemetry and electrocardiography were consistent with atrial fibrillation with rapid ventricular response. The patient denied a prior history of such arrhythmia.

Rate control was achieved with diltiazem infusion and anticoagulation with intravenous heparin was initiated. Echocardiogram showed a left atrial mass adjacent to the interatrial septum suspicious for atrial myxoma (Figure 1). Computed tomography scan showed a left atrial filling defect that enhanced with contrast and measured 4.5x3.3x4.4 cm (Figure 2) as well as a filling defect in the superior mesenteric artery (Figure 3). Lower extremity angiogram demonstrated abrupt cessation of flow consistent with an embolus to the right popliteal artery (Figure 4) and a patent left popliteal artery. Right popliteal arterial thrombectomy was performed. The patient then underwent excision of the left atrial mass. Maze procedure and left atrial appendage ligation. The examination of excised mass revealed an organized thrombus (Figure 5). The postoperative course was uneventful and the patient was discharged on warfarin and metoprolol.

DISCUSSION

Ischemic stroke accounts for 80–90% of embolic events associated with AF whereas peripheral emboli account for 10–20% [3, 4]. Current guidelines recommend the utilization of the CHADS2 and CHA2DS2-VASc scores to estimate stroke risk. CHADS2 assigns 1 point each for systolic heart failure with LVEF < 40%, hypertension, age ≥ 75 years and diabetes mellitus and 2 points for prior stroke, transient ischemic attack or arterial embolus. CHA2DS2-VASc assigns 2 points for prior stroke, transient ischemic attack or arterial embolus of for age ≥ 75 years and one point each for heart failure with LVEF
<40%, hypertension, diabetes mellitus, vascular disease, age 65–74 years and female gender [2–5]. Although the clinical significance of the CHADS2 and CHA2DS2-VASc scores to predict stroke risk is well established, it is unclear if these scores have similar ability to predict peripheral embolism risk. Yamamoto et al. [6] investigated the utility of the CHADS2 score in the prediction of peripheral emboli in 117 patients with non-cerebral acute arterial emboli. They found the CHADS2 score to be an unreliable predictor of non-cerebral embolism, with a high risk of non-cerebral occlusion even in patients with a low CHADS2 score [6]. The ATRIA study assessed five risk stratification schemes for their ability to predict atrial fibrillation-related thromboembolism (both cerebral and peripheral emboli) in a large community-based cohort [7]. The authors concluded that these risk stratifications schemes had similar and relatively poor ability to predict thromboembolic risk in persons with non-valvular AF [7].

In a Mayo clinic study published in 2008 [8], 72 patients with peripheral emboli associated with non-valvular AF were randomly compared to 100 patients with stroke in the setting of AF to assess clinical and echocardiographic features determining thromboembolic site. In this study, age >75 years, hypertension and heart failure were associated with a higher incidence of peripheral emboli. Echocardiographic features associated with higher risk included severe left ventricular systolic dysfunction, severe left atrial enlargement, spontaneous echocardiographic contrast and left atrial thrombus [8].

Bekwelem et al. [9] examined the data from four large randomized clinical trials of anticoagulation in AF. Among 37,973 patients and 91,746 patient-years of follow-up, 221 systemic embolic events occurred in 219 subjects. In comparison with stroke patients, those with systemic embolic events were more often smokers, were female, had peripheral arterial disease and had a previous systemic embolic event [9].

In Bekwelem’s analysis, systemic embolic events comprised 11.5% of all clinically recognized thromboembolic events, with an incidence of 0.24 of 100 patient-years compared to an incidence of 1.92 of patient-years for stroke [9]. The most common locations of peripheral emboli were the lower extremity (58%), visceral-mesenteric (31%) and the upper extremity arteries (10%) [9].

An earlier Danish trial published in 2001 [10] analyzed the risk of peripheral emboli in the setting of AF. In this study, 14,917 men and 14,945 women aged 50–89 years diagnosed with AF in the Danish National Hospital Discharge Register from January, 1980 through December, 1993 were followed from the diagnosis of AF until the first diagnosis of a thromboembolic event, death or the end of the study period. In this study, both men (relative risk 4.0, 95% CI: 3.5–4.6) and women (relative risk 5.7, 95% CI: 5.1–6.3) were at increased risk for the occurrence of a peripheral thromboembolic event [10]. The extremities were the most common extracranial location of emboli (61%), followed by the mesenteric arteries (29%), pelvic arteries (9%), aorta (7%) and renal arteries (2%) [10].

Simultaneous emboli in different arterial distributions, as occurred in our patient, are encountered less frequently. In Bekwelem’s analysis [9], simultaneous emboli occurred in only 2 of 219 patients with peripheral emboli. Lee et al. [11] report the case of a patient who

![Figure 4: Lower extremity arteriogram showing abrupt cessation of flow in the right popliteal artery (arrow).](image_url)

Figure 4: Lower extremity arteriogram showing abrupt cessation of flow in the right popliteal artery (arrow).

![Figure 5: Thrombus excised from the left atrium.](image_url)

Figure 5: Thrombus excised from the left atrium.
presented with simultaneous cerebral and brachial arterial emboli in whom the pain and weakness in his right arm was initially believed to be the result of an acute thromboembolic stroke.

Interpretation of the clinical situation may be difficult as a result of atypical symptoms and more subtle signs and because the sequelae of an embolus to a particular arterial distribution may overshadow those occurring as a result of occlusion of another territory. Additionally, because of redundant circulation, the minor clinical consequences of smaller hepatic or splenic infarcts or the lack of acute symptoms with renal arterial emboli, peripheral emboli may be silent or unrecognized [9, 12]. As a result, the incidence of peripheral emboli and multiple acute emboli is probably higher than reported whereas the ratio of intracranial to extracranial emboli is likely lower than reported.

The symptoms and signs of the more commonly encountered emboli to the arteries of the extremities are more specific than those due to emboli to the visceral organs. The clinical presentation of arterial occlusion involving the extremities includes paresthesia, pain, pallor, pulselessness, poikilothermia and paralysis (the six P’s). Vascular imaging with duplex ultrasonography, computed tomographic angiography (CTA), magnetic resonance angiography (MRA) or standard angiography will confirm the diagnosis. However, anticoagulation should be initiated promptly based on the clinical history and physical exam findings and should not be delayed while awaiting vascular imaging results. Definitive treatment includes catheter-based thrombolytic therapy and/or mechanical embolectomy depending on the duration of ischemia and embolus location and length [13].

The clinical manifestations of emboli to visceral organs can be quite variable and nonspecific depending on the location of the organ affected and the presence or absence of collateral circulation. Early diagnosis is vital to prevent organ infarction and requires a high index of suspicion. Emboli to the mesenteric arteries should be suspected in patients with AF presenting with sudden severe abdominal pain that is out of proportion to the physical exam signs. Diagnosis can be confirmed by vascular imaging including CTA or standard angiography of the mesenteric vasculature. Emergent treatment with anticoagulation and surgical embolectomy are necessary to prevent bowel infarction. Clinical presentation of acute occlusion of the renal arteries is nonspecific and symptoms can be attributed to nephrolithiasis or pyelonephritis. However, acute renal artery occlusion should be suspected in patients with AF and acute unilateral flank pain associated with hematuria and hypertension. Computed tomographic angiography (CTA) or magnetic resonance angiography (MRA) is necessary for the diagnosis. Treatment includes anticoagulation and percutaneous embolectomy. Splenic artery occlusion due to AF is rare. Symptoms are also nonspecific and include left upper quadrant abdominal pain, nausea and vomiting. Diagnosis is confirmed angiographically [14].

Morbidity and mortality increases following a peripheral embolic event. Gökş-Bierska [15] and Sundboll [16] demonstrated a higher rate of thromboembolism among patients with peripheral emboli in the setting of AF compared to those with a stroke and AF. This risk persisted over a follow-up period of 10 years [16]. Patients with a visceral-mesenteric embolus had a significantly higher mortality rate as compared to those with stroke alone [9] over a mean follow-up period of 2.4 years. Mortality risk was also increased to a lesser degree in those with lower extremity and upper extremity emboli over the same period [9].

**CONCLUSION**

Although ischemic stroke remains the most common embolic complication of atrial fibrillation (AF), peripheral emboli may also account for 10–20% of embolic events related to this arrhythmia. Peripheral emboli are also associated with significant morbidity and mortality and may go unrecognized unless a high index of clinical suspicion is present. Early diagnosis and therapy for a wide array of embolic events associated with AF is essential to reduce complications and improve clinical outcomes. All patients with AF and peripheral emboli remain at high risk for recurrent embolic events and should receive lifelong anticoagulation not only to prevent recurrence but also to decrease the risk of future stroke. Although the CHADS2 and CHA2DS2-VASc scores are not reliable predictors of the risk of peripheral emboli, it should be emphasized that an arterial thromboembolic event immediately increases these scores to at least 2. These patients are therefore at significantly higher risk for a future thromboembolic stroke and lifelong anticoagulation is required.

**Author Contributions**

Ismail Tabash – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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A case of subareolar abscess in the male breast: Usefulness of magnetic resonance imaging scan for diagnosis

Takaaki Fujii, Keiko Yanai, Shoko Tokuda, Yuko Nakazawa, Sasagu Kurozumi, Sayaka Obayashi, Reina Yajima, Tomoko Hirakata, Hiroyuki Kuwano

ABSTRACT

Introduction: Subareolar abscess in the male breast is extremely rare. The main concern with this condition is performing the differential diagnosis, since clinically it often resembles breast cancer. Case Report: A 30-year-old Japanese male presented with pain and swelling of the left breast. Physical examination revealed a 3.0x3.0 cm palpable lump with redness of the skin in the left subareolar region. Magnetic resonance imaging (MRI) scan showed a ring-enhancing mass without wash out with central necrosis below the left nipple, suggesting subareolar abscess. A core needle biopsy was performed, and histological findings revealed granulation tissue with the infiltration of numerous neutrophils, a finding that was compatible with subareolar abscess. Conclusion: Male subareolar abscess is quite a rare disease, however, awareness of how to perform differential diagnosis of subareolar abscess and the use of MRI scan may help in making a definitive diagnosis of subareolar abscess in the male breast and might aid in the proper management of the condition.

Keywords: Breast cancer, Male breast, Pain and swelling of present, Subareolar abscess

INTRODUCTION

Benign nonneoplastic entities may occur in the male breast, including intramammary lymph node, sebaceous cyst, diabetic mastopathy hematoma, fat necrosis, and subareolar abscess [1, 2]. However, subareolar abscess in known to occur more often in women than men [3], and in males it is extremely rare [1–5]. We herein report a rare case of subareolar abscess in male breast. The main concern with subareolar abscess in male breast is performing the differential diagnosis, since clinically it often resembles breast cancer. In our case, magnetic resonance imaging (MRI) scan was useful for the differential diagnosis between subareolar abscess and malignancy.
relevant medical history. Physical examination revealed a 3.0x3.0 cm palpable lump with redness of the skin in the left subareolar region (Figure 1). There was no abnormal nipple discharge. Sonography revealed a complex, hypoechoic subareolar mass with a heterogeneous hypoechoic internal echo containing complex fluid and internal vascularity (Figure 2). Magnetic resonance imaging showed a ring-enhancing mass without wash out with central necrosis below the left nipple (Figure 3), suggesting subareolar abscess. A core needle biopsy was performed, and histological findings revealed granulation tissue with the infiltration of numerous neutrophils, a finding that was compatible subareolar abscess (Figure 4). Ultrasound-guided drainage and antibiotic therapy (cefcapene pivoxil) were prescribed. Culture showed the growth of *Propionibacterium acnes*. The patients required incisional drainage through subareolar incision. No recurrence has been seen for sixteen months.

DISCUSSION

We report herein an extremely rare case of subareolar abscess in the male breast. Male subareolar abscess is a localized infection secondary to ductal ectasia, chronic obstruction, and inflammation [1, 6]. Smoking is recognized as an aggravation factor, and almost 70% of patients smoked more than 10 cigarettes a day [6,
In this case, the patient was a smoker. Common clinical features include pain, nipple swelling, and nipple discharge [1]. On sonography, the abscess usually appears as an incompletely circumscribed mass containing complex fluid (Figure 1). Doppler imaging often shows accentuation of peripheral vascularity with sparse-to-absent internal flow. However, occasionally abscesses may have accentuated internal flow, making the diagnosis of infection less certain and therefore requiring tissue sampling. In our case, core needle biopsy was performed [1]. However, MRI scan may be useful for reaching a differential diagnosis of subareolar abscess. MRI scan revealed a ring-enhancing mass without wash out below the left nipple, which is a characteristic finding in cases of abscess. This case indicates the importance of MRI in differentiating between subareolar abscess and malignancy in the male breast. Familiarity with the features of the male subareolar abscess including MRI findings allows for accurate imaging and make it possible to avoid unnecessary and often invasive treatment.

CONCLUSION

In conclusion, we report here a rare case of subareolar abscess in the male breast. Male subareolar abscess is quite rare; however, awareness of how to perform the differential diagnosis of subareolar abscess and the use of magnetic resonance imaging scan may help in making a definitive diagnosis of subareolar abscess in the male breast and might aid in the proper management of the condition.

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Takaaki Fujii – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
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REFERENCES

Lemierre’s syndrome: A forgotten entity with a very rare presentation

Sailesh Kumar Bansiwal, Prabal Rajvanshi, Harshita Sharma, Rajesh Manocha

ABSTRACT

Lemierre’s syndrome is a potentially lethal condition, which originates as a complication of an oropharyngeal infection which causes suppuration of lateral pharyngeal space, bacteremia, and thrombophlebitis of internal jugular vein (IJV) leading to distant septic emboli. This is a forgotten disease in modern era of antibiotic therapy though it has been reported with increasing frequency in 21st century. We present a case of 34-year-old female presented with fever, headache, neck pain, odynophagia, and hemoptysis, found to have thrombosis of IJV and internal carotid artery (ICA) on imaging which responded to antibiotics and anticoagulant therapy. A high index of suspicion is necessary for diagnosis and early treatment is life saving.

Keywords: Forgotten disease, ICA thrombosis, Lemierre’s syndrome, Septic thrombophlebitis of internal jugular vein

INTRODUCTION

Lemierre’s syndrome, also known as postanginal septicemia or necrobacillosis was first reported in 1890. It was named after André Lemierre who explained the disease in 1936 [1]. It is usually caused by gram negative anaerobic bacillus *Fusobacterium necrophorum* [2]. However *Peptostreptococcus*, *Staphylococcus*, *Streptococcus* and Proteus have also been isolated. The disease progresses in several steps following a primary infection, which is usually pharyngitis (87.1%). Further invasion of deep neck spaces occurs, leading to internal jugular vein (IJV) thrombophlebitis (71.5%), and infective metastasis to distant sites most commonly to lungs (79.8%) [3]. Imaging and other investigations helps in the diagnosis. Surgery is seldom required as prolonged IV antibiotics have been documented as the mainstay of treatment.

CASE REPORT

A 34-year-old housewife presented with a short history of fever, and headache for 20 days; odynophagia, neck pain, right sided facial swelling for 10 days; and hemoptysis for five days. The patient denied any history...
of seizures, altered sensorium, photophobia, and arthralgias. The past history was unremarkable.

On examination, the patient was ill looking, tachypneic, and febrile to touch. However, her vitals were normal with pulse rate 104/min, and blood pressure 130/80. General examination showed right sided facial swelling and tenderness along the right sided sternocleidomastoid. Oral examination revealed inflammation of right tonsillar fossa. In the systemic examination, there were fine infraaxillary crepitations in bilateral lungs; and neurological examination showed terminal neck rigidity; while the examination of other systems was unrewarding (Figure 1).

Laboratory data showed total leukocyte count 18,650 (normal range 4,500–11,000 per µl) with 86% neutrophils. C-reactive protein (CRP) with value 31 mg/L (normal range 0–10 mg/L) and erythrocyte sedimentation rate (ESR) with value 62 mm/hr (normal range 0–20 mm/hr) were significantly raised; antinuclear antibody (ANA), anti-phospholipid antibody (APLA), protein C, protein S, Factor 5 Leiden, antithrombin, homocysteine were normal and HIV was non-reactive. Cerebrospinal fluid analysis revealed 40 cells with 98% lymphocytes while adenosine deaminase (ADA), proteins and sugar were insignificant.

Ultrasoundography of abdomen and echocardiography were normal. However, electrocardiography showed sinus tachycardia. The chest radiograph showed bilateral patchy shadows with mild bilateral pleural effusion (Figure 2).

Magnetic resonance imaging scan of head and neck showed right peritonsillar abscess, thrombosis of bilateral proximal IJV along with the thrombosis of right sigmoid sinus and left retromandibular vein. It also suggested bilateral ethmoid and sphenoid sinusitis/abscess, with extension of abscess to bilateral masticator space caudally and cranially to dura leading to subdural abscess and leptomeningitis.

A computed tomography angiography chest (Figure 3) and neck (Figure 4) was done which revealed thrombosed left internal carotid artery (ICA), partially thrombosed left IJV, sphenoid sinusitis and cavitation in bilateral lung fields suggestive of septic emboli. Color doppler bilateral neck was done to assess the extension of thrombus in the vessels. Surprisingly, it showed involvement of both IJV (partially occluded) and ICA (80% occluded) of left side.

Blood (anaerobic and aerobic), urine, sputum, and throat cultures were negative as the patient had received antibiotics prior to admission in our hospital.

The patient was given intravenously ceftriaxone (1 g q12 h), vancomycin (1 g q12 h), and clindamycin (600 mg q8 h) for 4 weeks. She also received anticoagulation with LMW heparin (6 mg SC q12 h) and acenocoumarol (2 mg q24h) and INR was monitored subsequently.
The patient responded to the therapy as evident on radiographic resolution of the thrombosis both in internal jugular vein and internal carotid artery.

**DISCUSSION**

Lemierre’s syndrome, being a rare entity (incidence 1 in a million) [4] has been drawing our attention because of its increasing incidence and dreaded complications; reemergence of this disease is most likely due to the antibiotic resistance and discouragement of the use of antibiotics for sore throat.

This patient had been worked up to rule out the closely mimicking differentials which were disseminated tuberculosis, cavernous sinus thrombosis, autoimmune vasculitis and thrombophilies.

Lemierre’s syndrome can have a spectrum of presentation ranging from mild pharyngitis to peritonsillar abscess. However, sinusitis, otitis media, mastoiditis, and odontogenic infections have also been described rarely. Sinusitis/sinus abscess (sphenoid and ethmoid) was probably the primary source of infection in our case (as per the history given by the patient), which involved the pharynx (peritonsillar abscess), lateral pharyngeal space, masticator space and the meninges. Meningitis in this case could be either due to direct spread from the sinus (sphenoid and ethmoid), or indirect spread through septic emboli (via ICA and IJV), or retrograde involvement of the sigmoid sinus (via IJV). There was involvement of the carotid sheath which was evident on CT angiography and color Doppler neck, leading to thrombosis of ICA and IJV. In our case, ICA thrombosis could be either due to its proximity to IJV or due to septic emboli. Though involvement of ICA is rarely reported in literature but in our case, this finding was very much evident on the imaging. Considering the endovascular nature of the disease, metastatic spread most commonly involves the lungs (79–100%) [3]. However, joints, bones, skin, liver, and kidney may also be involved.

A prolonged antibiotic course (2–6 weeks) is needed in order to eradicate the microorganisms within the thrombus. The role of anticoagulation is controversial, however, some authors consider it if there is extension of thrombosis. Surgical intervention is seldom required in the present era.

Untreated cases may have mortality of up to 90% [5]. The prognosis of Lemierre’s syndrome is favorable if there is early diagnosis and treatment. Rare presentation of Lemierre’s syndrome, ICA thrombosis along with IJV thrombosis, meningitis and sinusitis are being highlighted in our case report.

**CONCLUSION**

Lemierre’s syndrome occurs primarily in young, otherwise healthy individuals and is characterized by a history of recent oropharyngeal infection, clinical or radiological evidence of internal jugular venous thrombosis and anaerobic bacteremia caused primarily by *F. necrophorum*. This is a rare illness in the modern era of antibiotic therapy, though it has been reported
with increasing frequency in the 21st century. Lemierre’s syndrome should be suspected in young, healthy patients with prolonged symptoms of pharyngitis followed by symptoms of septicemia or pneumonia, or an atypical lateral neck pain. Diagnosis is often confirmed by the identification of internal jugular vein thrombophlebitis by an imaging study and growth of anaerobic bacteria on blood culture. Prolonged antibiotic therapy is the cornerstone of the treatment, occasionally combined with anticoagulation.

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Sailesh Kumar Bansiwal – 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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Spontaneous expulsion and migration of a bronchial foreign body: A flusterling rare dental accident

Koken Ameku, Mariko Higa

ABSTRACT

Introduction: A bronchial foreign body is a dangerous medical emergency that is potentially life-threatening. Spontaneous expulsion should neither be expected nor experienced. Thus, early removal with a bronchoscope should be performed to prevent complications. Spontaneous expulsion of a bronchial foreign body is rare, with few cases reported. Additionally, its occurrence and associated clinical complications are not studied and unclear. We describe a patient in whom a bronchial foreign body was expectorated spontaneously and swallowed into the digestive tract. Case Report: An 80-year-old male aspirated a tooth in the left lower airway during a dental procedure. Flexible bronchoscopy performed on the next day found no foreign body in either bronchial tree. However, it was found on the abdominal radiograph. It was considered to have been expectorated and then swallowed before bronchoscopy. One week later, the patient had passed it. Conclusion: This case demonstrated that a bronchial foreign body can rarely be expelled spontaneously. Besides, an expelled foreign body can be swallowed, or it can migrate to another location. Regarding spontaneous expectoration and migration, injury to the airway and digestive tract can occur depending on its shape. However, such dangers have not been addressed, because spontaneous expectoration is rare and unrecognized. Recognizing these dangers and warning patients of them could avoid additional complications.

Keywords: Dental care, Foreign bodies, Foreign body migration, Spontaneous remission

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INTRODUCTION

A bronchial foreign body is a dangerous medical emergency that is potentially life-threatening. Spontaneous expulsion should neither be expected nor be experienced. Thus, early removal with a flexible or rigid bronchoscope should be performed to avoid serious respiratory complications, such as suffocation, obstructive pneumonia, hemoptysis, atelectasis, and lung abscess. In large case series of adults, the success rate of removal by flexible bronchoscopy is almost 90% with few complications [1]. In case of flexible bronchoscopy failure, rigid bronchoscopy or thoracotomy is performed.
Spontaneous expulsion of a bronchial foreign body is rare with few cases reported [2]. Additionally, its occurrence and associated clinical complications are not studied and unclear. We describe a patient who spontaneously expectorated an accidentally aspirated tooth during a dental procedure, which was expectorated and migrated to the digestive tract before bronchoscopy could be performed.

CASE REPORT

An 80-year-old male was referred to our hospital after he aspirated a tooth into his airway during a dental procedure. An 8-mm radiopaque foreign body was detected in the left lower bronchus on a chest radiograph and computed tomography scan (Figures 1 and 2). Removal by flexible bronchoscopy was attempted on the next day, but the foreign body was not found in either bronchial tree. The radiograph showed the tooth in the abdomen (Figures 3). The tooth was considered to have been expectorated and swallowed into the digestive tract. He was discharged, and one week later, we confirmed that the patient passed it.

DISCUSSION

This case demonstrated several important issues. First, a bronchial foreign body can rarely be expelled spontaneously. However, physicians may have infrequently experienced this occurrence previously. Second, an expelled foreign body can be swallowed, or it can migrate to another location. Third, accidental aspiration of foreign bodies during dental procedure does occur in extreme rare occasions. In addition to its physical strain, the event is confusing for both patients and dentists. Moreover, in referred emergency units, the migration of a foreign body, as in the present case, can be an additive danger and cause distress for both the patient and emergency physician.

A bronchial foreign body can rarely be expelled spontaneously. However, physicians may have infrequently experienced this occurrence previously. Foreign body aspiration is an uncommon clinical entity in adults, with an incidence of about only 1 in 400 bronchoscopies [1]. As a result, this disease is not frequently experienced by physicians. Spontaneous expulsion of a foreign body occurs even more rarely in approximately 1–2% of cases. Thus, it is not recognized by physicians [1–3]. Although few cases of spontaneous expulsion have been described in literature, it is generally not focused on or studied; this may be why it is not reported [2, 4–6]. Among airway foreign bodies, inorganic materials, such as metallic or denture-like ones, as in the present case, cause little mucosal

Figure 1: An 8-mm radiopaque foreign body is seen in the left lower bronchus on a chest radiograph.

Figure 2: A foreign body is seen in the left lower bronchus on a computed tomography scan.

Figures 3: (A, B) During bronchoscopy on the next day after admission, the foreign body is not seen on a chest radiograph. The radiopaque foreign body appears in the abdomen. It passed through the patient’s digestive tract one week later.
inflammation and granulation; therefore, they are more likely to mobilize than organic ones. Reported cases of spontaneous expectoration mostly involved inorganic, straight, and sharp materials [2, 4–8]. Although our patient did not have any complications, when such sharp foreign bodies are not fixed in the lower airway, and then they are expectorated, damage to the airway can occur, and suffocation can even occur depending on how it is expelled. If it is probable that the foreign bodies will be expelled spontaneously, warming and preparation should be taken to avoid dangers. However, such dangers have not been addressed, because spontaneous expectoration is rare and unrecognized. The migration of a foreign body lodged in the bronchium, which was almost surgically resected, has also been reported [9]. As spontaneous expulsion of a foreign body can lead to serious clinical accidents and its occurrence may be underreported, it should be studied and addressed more. Besides, an expelled foreign body can be swallowed or migrate to another location. Several cases of patients who have swallowed a foreign body into their digestive tract after expectoration have been reported [7, 8]. Although most ingested foreign bodies pass through the gastrointestinal tract uneventfully over a period of seven to ten days, large, sharp, or pointed objects can cause other complications, such as perforation, obstruction, or hemorrhage [10]. Endoscopic removal or even operation is required in almost 10% and 1% of cases, respectively. Additional complications by ingestion after expectoration should be recognized by emergency physicians, and these should be avoided by giving precautions to the patients. Accidental aspiration of foreign bodies during dental procedure does occur in extreme rare occasions [3]. In addition to its physical strain, the event is confusing for both patients and dentists. Moreover, in referred emergency units, the migration of a foreign body, as in the present case, can be an additive danger and cause distress for both the patient and emergency physician. The incidences (cases/patients) of accidental ingestion or aspiration during dental procedures have been reported as 0.0041 and 0.0044% per year. The occurrence (cases/dentists) per year was reported as 0.018 [10]. Dental materials are mostly inorganic and likely to be expelled spontaneously, depending on its shape. When it occurs, the expelled objects are teeth, dentures, dental prostheses, dental instruments, and broken orthodontic appliances, which are sharp, large, and long, metallic materials; thus, they can be more hazardous [3]. This aspect of a dental accident must be addressed so dentists and emergency physicians can avoid additional complications. Furthermore, more studies should be performed on this topic.

CONCLUSION

This case demonstrated that a bronchial foreign body can rarely be expelled spontaneously. Besides, an expelled foreign body can be swallowed, or it can migrate to another location. Regarding spontaneous expectoration and migration, injury to the airway and digestive tract can occur depending on its shape. Recognizing these dangers and warning patients of them could avoid additional complications.

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REFERENCES


ABSTRACT

Introduction: A hydrocele of the canal of Nuck is an exceedingly rare condition. Reports describing its treatment have almost exclusively involved open surgery. Advancements in minimally-invasive techniques have allowed us to employ laparoscopic hernia repair methods as a novel way of treating this condition. Case Report: We present a 50-year-old obese female who was diagnosed with a 4.2x3.2 cm encysted hydrocele of the canal of Nuck on computed tomography. This arose from the peritoneal cavity in association with the left uterine round ligament and extended towards the left labia majora. The hydrocele was approached using a standard transabdominal preperitoneal technique. After dissecting the hydrocele from the canal, it was further parietalized along with its associated round ligament, which was then ligated and divided. A prolene mesh was used to cover the resultant defect in the internal inguinal ring. The peritoneal flap was repositioned and the peritoneal defect traversed by the round ligament was closed intracorporeally with an absorbable suture. The patient made an uneventful recovery and was discharged the next day. Conclusion: Transabdominal preperitoneal approach allows for adequate assessment and complete excision of the canal of Nuck cyst, while combining the benefits of minimally-invasive tension-free mesh repair.

Keywords: Canal of Nuck, Hydrocele, Transabdominal preperitoneal mesh repair

INTRODUCTION

In 1691, Dutch anatomist Anton Nuck (1650–1692) described an abnormality found in the groins of females the canal of Nuck. The canal is said to occur when there is failure of obliteration of the peritoneal fold, as it accompanies the round ligament of the uterus, when it descends into the labium majora through the inguinal canal. This obliteration is said to occur usually by eight months of gestation [1], failure of which results in a communication with the peritoneal cavity, which can manifest as an inguinal hernia or a hydrocele.

There have been case reports of hydroceles within the canal of Nuck and discussions regarding the management, but the ideal treatment for this rare condition have yet to be established. While earlier reports focussed on open surgery, advancements in minimally-invasive techniques have allowed us to employ laparoscopic hernia repair methods in the treatment of these patients. We present the successful management of such a case.
CASE REPORT

A 50-year-old female with a body mass index of 31.4 presented with complaints of pain and a fullness in her left groin which had been increasing in size over the past five months. She denied any history of trauma, fever, bowel or urinary dysfunction. On examination, there was a fullness in her left groin that was mildly tender on palpation, but no overlying skin changes, evidence of infection, or regional lymphadenopathy. There was no expansile cough impulse, and no audible bowel sounds.

A computed tomography scan of the abdomen and pelvis showed a 4.3x3.2 cm ovoid structure arising from the peritoneal cavity, associated with the round ligament of the uterus and extending to the left labia majora (Figure 1). Hematological investigations were essentially unremarkable.

The patient underwent a transabdominal laparoscopic excision of the left canal of Nuck cyst with ligation of the left round ligament, and pre-peritoneal mesh placement. Three ports were utilized for the procedure. A 10 mm camera port was inserted in the infraumbilical region, followed by two 5 mm ports in the mid-clavicular line on either side of the camera port. The pre-peritoneal plane was entered and developed to expose the cyst (Figure 2). The cyst was then dissected out and parietalized with its associated round ligament. The left round ligament was ligated intraperitoneally using polydioxanone (PDS) loop suture before division, excising it en bloc with the cyst. The resultant internal inguinal ring defect was then covered with a 10x15 cm Ultrapro mesh and anchored with SecureStrap® (Ethicon). The peritoneum was re-approximated with SecureStrap.

Postoperative recovery was uneventful and the patient was discharged the following day. Histopathological examination of the specimen confirmed a benign cyst.

DISCUSSION

A hydrocele in the canal of Nuck is a rare developmental disorder. The age at presentation ranges from 18 months to 51 years, with the majority of diagnoses being made between 30–40 years of age. Huang et al. reported that the incidence in children is approximately 1%, but the incidence in adults has yet to be studied [2]. In 2003, Stickel et al. reported that only 400 cases exist in literature [3]. Interestingly, the incidence of a patent processus vaginalis in adults is about 30% at autopsy, although it is not fully understood why only some individuals develop hernias or hydroceles [4].

Conventional open approach would have entailed a similar incision to that of an open inguinal hernia repair. Following excision of the hydrocele, the fibrous connection towards the deep inguinal ring would be sutured and ligated and the rest of the wound closed in layers. Ensuring closure of the internal ring defect would be a crucial step in obliterating the canal and preventing hernia formation. In the case of a patent canal of Nuck, simple ligation without mesh placement may fail to achieve this, resulting in a higher rate of hernia occurrence.

An understanding of the anatomical basis behind this condition allows us to employ minimally-invasive techniques in its treatment, thereby providing the same benefits that laparoscopy has afforded to open inguinal hernia repair. In view of her high body mass index (BMI) and the vagueness of the swelling, open surgery would have required a more extensive dissection in our patient. A minimally-invasive approach was clearly preferred.

Common approaches to inguinal hernia repair include the totally extraperitoneal (TEP) approach and the transabdominal preperitoneal (TAPP) approach. Matsumoto et al. wrote that patients who underwent TEP reported shorter recovery time and were at less risk of bowel adhesion formation [5]. They also argued that the TEP approach could provide surgeons with information on the origin and type of hydrocele in the canal of Nuck.
In our patient, the intraperitoneal component of the cyst would not have been adequately accessed using an extraperitoneal approach. The TAPP approach was, therefore, utilized to allow assessment of the attachment of the cyst to the round ligament, followed by complete dissection and removal. Through the transabdominal approach, we were also able to exclude a concurrent contralateral hernia, reported in 13% of patients presenting with suspected unilateral inguinal hernias [6].

CONCLUSION

In summary, the transabdominal preperitoneal approach allows for adequate assessment and complete excision of the canal of Nuck cyst, while combining the benefits of minimally-invasive tension-free mesh repair.

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REFERENCES
Appendicitis in the setting of undiagnosed non-rotation of the midgut: A case report

Ashraf Tokhi, Eliza Muir

CASE REPORT

A 32-year-old male presented with a history of three-day left sided abdominal pain. The pain was initially generalized, then later localized to the left iliac fossa with associated fevers, vomiting, anorexia and constipation. Examination elicited generalized peritonism, with rebound tenderness worst in the left iliac fossa, and positive psoas sign.

Variation in locality of tenderness and uncertain diagnosis required further investigation; a computed tomography scan of abdomen and pelvis. Imaging demonstrated marked dilatation of the appendix and periappendiceal fat stranding, confirming acute appendicitis (Figure 1). Non-rotation of the midgut was also noted, with the appendix and cecum located in the left lower quadrant, and the majority of small bowel seen on the right side of abdomen (Figure 2). No bowel obstruction was present. The patient was commenced on intravenous antibiotics and proceeded to theatre for emergency appendicectomy.

Laparoscopy showed four quadrant peritonitis with a left-sided cecum. The appendix was grossly inflamed with a perforation at the base. The procedure was converted to laparotomy, the appendix was resected and the peritoneal cavity was washed out copiously (Figure 3). No evidence of bowel obstruction was noted.

The patient had an uncomplicated postoperative course, and was discharged at postoperative day-10. He remained well at second week follow-up.

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Figure 1: Coronal computed tomography slices showing a left sided appendix, acutely inflamed with two fecaliths present in its lumen.

Figure 2: Axial computed tomography slice showing the superior mesenteric artery placed to the right of the superior mesenteric vein.
DISCUSSION

Intestinal non-rotation is a congenital abnormality resulting from a lack of midgut rotation during the first trimester, and is one of a number of malrotation variations. Malrotation is estimated to occur in one in 500 live births. However, incidental non-rotation has been noted in 0.5% of autopsies [1].

During the fourth to eighth week of embryonic growth, the midgut exits the coelom into the yolk sac where its rapid growth can be accommodated. The midgut rotates 90 degrees counterclockwise in the yolk sac. Non-rotation occurs when, after re-entering the abdomen during the eighth to tenth week, the midgut does not continue any additional rotation. This results in a left sided colon and predominantly right sided small bowel. It is hypothesized that this is due to laxity of the umbilical ring, resulting in an early reduction of the colon and cecum [2].

Generally, there are three different presentations of non-rotation pathology; acute obstruction, chronic intermittent obstruction, and as an incidental finding [3]. Almost 60% of individuals with any gut malrotation will present with an acute obstruction secondary to volvulus by their first birthday [4]. In the adulthood only 12% of presentations relating to malrotation are secondary to volvulus [5]. The more common presentation is chronic obstruction, with symptoms of non-specific abdominal pain, intermittent nausea and vomiting. Contrary to traditional teaching, nearly half of patients with midgut non-rotation present symptomatically in adulthood [5, 6]. Infrequently, such as in our case, midgut non-rotation is observed as an incidental finding for investigation of other conditions [3, 7].

The presence of non-rotation is important when considering surgical intervention, due to the variant anatomy. In our situation, laparoscopic access was obtained with port insertion into the suprapubic area and the right low quadrant over McBurney’s point. This provided adequate access to the appendix and cecum, now located in the left lower quadrant.

Whilst surgical intervention in the form of a Ladd’s procedure has been suggested for symptomatic non-rotation, major guidelines recommend that in asymptomatic patients, a conservative approach is followed, with an emphasis to the patient to present early if any symptoms occur in the future [8].

CONCLUSION

As was noted in this case, patients with midgut non-rotation can present a diagnostic dilemma, due to the disparity in location of abdominal viscera, and thus variation in symptomatology. Delay in diagnosis and treatment can have severe consequences, and thus it is important to consider early radiographic investigation for further evaluation. In line with guidelines, we do not recommend surgical intervention in asymptomatic patients, but encourage early presentation if any abdominal symptoms occur in the future.

*********

Keywords: Appendicitis, Malrotation, Non-rotation

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REFERENCES
Intestinal perforation due to fish bone diagnosed preoperatively by computed tomography

Yoshimasa Kishi, Atsuyoshi Iida, Kohei Tsukahara, Atsunori Nakao

CASE REPORT

A 73-year-old female was presented to emergency department with a one-day history of increasing lower abdominal pain. The patient had attended a wedding party of her grandson and ate baked red snapper. Her past medical history was unremarkable and she was taking medication for hypertension. Her vital signs included blood pressure 119/66 mmHg, pulse rate 80 beats/min, and body temperature 36.2°C. On examination, the patient had a slightly distended abdomen with significant right iliac fossa guarding and tenderness. Her white cell count and C-reactive protein levels were 11900/mm³ and 1.24 mg/dL, respectively, indicating systemic inflammation. Abdominal computed tomography demonstrated pneumoperitoneum and fluid within the abdominal cavity, as well as dilated intestine, suggesting diffuse peritonitis due to alimentary tract perforation (Figures 1 and 2). Under general anesthesia, the patient underwent diagnostic/therapeutic laparoscopy, which showed acutely inflamed ileum and purulent ascites. A foreign body, assumed to be a fish bone, was observed piercing through the small bowel wall at the ileum. As the site of perforation was not clearly determined via laparoscope, a lower median laparotomy was performed. Lavage of the abdominal cavity enabled us to detect a 2-mm ileal perforation by foreign body by confirming bubble from the perforation. Diffuse purulent peritonitis was evident in an area with adhesions. The foreign body was removed, primary suture of the intestinal perforation was performed, and the abdominal cavity was drained. Intravenous antibiotics were administered. The patient was discharged on day-5 after admission without complications.

Figure 1: Plain abdominal computed tomography showing thickened intestinal segment, localized pneumoperitoneum. Linear density crossing the intestinal wall was noted (black arrow).

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DISCUSSION

Ingestion of foreign bodies is a common clinical problem encountered in emergency departments. Meat boluses are the most common foreign bodies ingested in Western countries, while fish bones are the most common in Oriental countries where unfilleted fish is a culinary delicacy [1, 2]. Although most fish bones pass through the gastrointestinal tract without complications, patients who accidentally ingest a fish bone are occasionally asymptomatic after ingestion initially, but may at a later date present remotely with serious complications such as perforation, obstruction, and abscess formation in the gastrointestinal tract. In fact, fish bones are the most common objects ingested and the most common foreign body to perforate the gastrointestinal tract [3]. Fish bones can perforate all segments of the alimentary tract. However, perforation tends to occur in areas of acute angulation such as the rectosigmoid and ileocecal junctions [4]. Early laparoscopic or surgical removal of the fish bone and abdominal lavage is recommended [5]. Eventually, as with our patient, determining the perforation site on laparotomy is difficult. Careful observation and administration of antibiotics are absolutely required for the treatment of intestinal perforation by fish bone.

Diagnosis of foreign body perforation of the gastrointestinal tract can be challenging and is rarely correctly diagnosed preoperatively. Radiography is unreliable in the diagnosis of fish bone perforation. Computed tomography (CT) scan has been helpful in the detection of nonmetallic foreign body perforation. Fish bone perforation typically appears as a linear calcified lesion surrounded by an inflamed area on CT scan [6]. Evidence of pneumoperitoneum is only seen in approximately 30% of patients with intestinal perforation caused by fish bone ingestion [1]. Computed tomography scan sensitivity for detection of fish bone peritonitis is known to be relatively high (71–100%). Since the main reason for missed diagnosis is the observer’s lack of awareness, a high degree of clinical suspicion should be maintained in order to make a correct diagnosis.

CONCLUSION

We described a case of acute peritonitis after perforation of the ileum by a fish bone that was detected by computed tomography. We emphasize the value of CT in diagnosing fish bone peritonitis; familiarity with its appearance on CT scan can help to detect fish bone perforation along with any associated complications and help guide further management.

Keywords: Foreign body, Peritonitis, Sea food

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REFERENCES
CASE REPORT

A 59-year-old female presented with weight loss, night sweats, and diarrhea since four months. Stool examination, serologic testing, esophagogastroduodenoscopy (EGD), colonoscopy (C-scope), and computed tomography (CT) scan of chest and abdomen were non-contributory. Diagnosed with celiac disease at 51, she had had no prior digestive symptoms despite poor adherence to a gluten free diet and persistently positive anti-tissue transglutaminase antibodies. Physical examination revealed cachexia, widespread erythematous maculopapular skin lesions, and no palpable lymphadenopathy. Laboratory examination showed hemoglobin 9.8 g/dL, mean cell volume 92 $\mu$m$^3$, white blood cell count 5500/$\mu$L (lymphopenia at 300/$\mu$L), and serum albumin 2.2 g/dL.

The PET scan showed intense FDG uptake in a left tonsillar mass with multiple FDG-avid subcutaneous lesions, and bowel and mesenteric involvement (Figure 1). The left tonsil and one of the cutaneous lesions were biopsied, revealing a CD8+ T-lymphoid cell infiltrate, most consistent with T cell lymphoma showing a CD4 negative, CD8 positive, and CD 56 negative cytotoxic phenotype. Balloon enteroscopy revealed a mosaiform small bowel mucosa with multiple clean-based ulcers throughout the jejunum. Biopsies were consistent with the tonsillar findings. A few days following admission the patient underwent a laparotomy for small bowel perforation, and was subsequently transferred to the intensive care unit where her course was complicated by recurrent gastrointestinal bleeding, septic shock and respiratory failure, leading to death.

DISCUSSION

Celiac disease (CD) is an immune-mediated enteropathy triggered by ingestion of gluten in genetically
susceptible individuals. It is a common disorder affecting approximately 0.5–1% of Caucasians [1]. Overall, 2–10% of patients with CD develop (RCD) [2]. Refractory celiac disease can be divided into types I (RCD I) and II (RCD II). About half of RCD II patients develop enteropathy-associated T cell lymphoma (EATL) [3]. RCD II is therefore considered an EATL precursor lesion. Although rare, EATL is one of the main causes of death in patients with symptomatic CD diagnosed as adults. In patients with CD presenting with fever, night sweats, pruritus, significant unexplained weight loss, prolonged diarrhea, anorexia, overt or occult gastrointestinal bleeding, abdominal pain, and bowel obstruction, EATL should be sought [1, 3].

Multiple endoscopic or imaging techniques are available to visualize the entire small bowel such as capsule endoscopy, CT or magnetic resonance imaging enterography, deep enteroscopy, and 18F-FDG PET scan. The latter two techniques are of great utility to exclude malignancy in CD patients presenting with alarm features. Abdominal CT scan has shown limited effectiveness in detecting EATL and distinguishing it from uncomplicated refractory celiac disease. Findings of bowel wall thickening, lymphadenopathy, intussusception and hyposplenism raise suspicion for RCD II and EATL. Apart from evidence of metastatic disease there are no well-described CT findings that will distinguish EATL from RCD II. Moreover, the role of radiologic imaging may be limited in diagnosing EATL as the neoplastic changes may be restricted to the epithelial layer of the small bowel, even when the lymphoma affects the whole small intestine [4].

The PET scanning displays higher sensitivity and specificity for the detection of EATL when compared with CT scan [5], but yields false positive results due to increased uptake in inflammatory tissues as is seen in non-refractory CD and RCD. However, the standard uptake value is lower than in EATL (0.0–4.6 vs 6.4–8.0 SUV), making PET a reliable method for identifying EATL [6]. Although PET scanning cannot be a substitute for histologic examination in diagnosing EATL, it may guide histological sampling.

Endoscopic examination of the small bowel in patients with celiac disease permits direct visualization and biopsy. Using EGD yields a limited examination of the small bowel, whereas only the distal ileum can be reached with colonoscopy [7]. Deep enteroscopy can provide full-length examination of the small bowel, and the ability to both perform biopsies and deliver therapy [7]. Video capsule endoscopy (VCE) allows full examination of the small bowel and assessment of the extent of involvement. Mucosal flattening and ulcerations can be found in EATL, but are also seen in ulcerative jejunitis. Video capsule endoscopy may also yield false negative results, as EATL can present as a sub-mucosal mass. Thus, reliable detection using VCE may be difficult. Most importantly, detected lesions cannot be sampled at VCE for histological examination [8].

Once a diagnosis of EATL is confirmed, most patients undergo surgical debulking followed by chemotherapy that usually includes the administration of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP). If feasible, surgery aids in diagnosis, while also decreasing the risk of potentially lethal perforation following chemotherapy and tumor necrosis. Stem cell transplantation (SCT) may be considered in patients presenting with an overall good clinical state, although no randomized trials have been performed. Unfortunately, as 64% of patients are diagnosed with stage IV disease, the prognosis remains very poor with one and five-year survival rates of 31–39% and 8–20% respectively [1].

CONCLUSION

We present a 59-year-old female with celiac disease of eight years duration who developed weight loss, night sweats, and diarrhea since four months. Biopsies at enteroscopy confirmed an enteropathy-associated T cell lymphoma, noted in 50% of patients with refractory celiac disease type II. As most, she had advanced stage IV disease; she died of small bowel perforation.

Keywords: Celiac disease, Enteropathy-associated T cell lymphoma, Enteroscopy, Positron emission tomography scanning

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REFERENCES
Secondary obstructive giant megaureter leading to massive pyogenic urinary infection

João Fonseca, Maria Amparo Castellano, Manuel Veríssimo, Armando Carvalho

CASE REPORT

A 70-year-old female presented with right back pain and fever. Six months earlier was submitted to hysterecmy and bilateral salpingo-oophorectomy for an ovarian epithelial cancer (stage IA). On physical examination, she had chills, fever, and tenderness at the right costovertebral angle. Blood tests indicated acute renal failure and marked elevation of C-reactive protein. In renal ultrasound, a mild right hydronephrosis was identified, and the respective ureter proved to be impossible to visualize due to the presence of a large cystic-like structure (19 cm of longitudinal size). As there was a suspicion of obstructive pyelonephritis, a renal scintigraphy was performed indicating the presence of a significant unilateral obstruction. Non-contrast computed tomography (CT) scan showed a markedly distended right ureter (up to 9.5 cm in diameter) with tortuosity (Figures 1 and 2), not evident in previous follow-up CT scan. No calculus or other anatomical urologic abnormalities were found. The patient underwent percutaneous nephrostomy, which drained 1500 cc of purulent fluid. *Escherichia coli* were isolated and adequate antibiotic therapy was instituted. The clinical condition of the patient declined progressively and had passed away after three days.

DISCUSSION

Giant megaureter is the name given to a massively dilated ureter. This pathological finding is rarely seen in clinical practice, especially in the geriatric age. The mechanism should be classified as obstructive, refluxing or non-obstructive non-refluxing [1]. We theorize that this case of obstructive megaureter was a late complication of a prior gynecological surgery, considering its absence in previous follow-up imaging exams. Almost half of the ureteral iatrogenic complications result from gynecological surgeries, of which hysterectomy is the main cause [2, 3]. Its early diagnosis is crucial, as it can lead to kidney damage and urinary tract infections [2].

In these patients with impaired renal function, non-enhanced CT scan can be of great value. This characterizes the extent of urologic changes, and identifies the presence

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Figure 1: A cystic-like structure (9.5 cm in diameter) is easily identifiable in the right pararenal location, on the computed tomography scan axial cuts, corresponding to the right ureter.
of calculi, compressive masses, and other anatomical abnormalities [4]. Magnetic resonance urography may also play an important role because it allows optimal noninvasive evaluation of many abnormalities of the urinary tract, including urinary tract obstruction [5]. Additionally, new gadolinium-based contrast agents associated with a few/unconfirmed cases of nephrogenic systemic fibrosis (like Gadobenate dimeglumine - MultiHance®), may be used safely even with severe renal dysfunction [6].

Due to the severe clinical condition of the patient, who met criteria for severe sepsis, after discussion of the case with the urology department, percutaneous nephrostomy was considered the safest option at the time. Other procedures, such as nephrectomy and resection of the ureter, were not considered because the patient and her family refused more invasive surgical interventions. This case is interesting because it presents a rare image of an extremely aberrant dilation of ureteral architecture.

CONCLUSION

We presented a patient who exhibited a secondary giant megaureter complicated by a severe urinary tract infection. The obstructive lesion resulted probably from gynaecological surgery, which is the main cause of iatrogenic ureteral injury. Sometimes these complications are only detected after serious consequences.