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Frequent use of stun gun may be associated with vibration-induced Raynaud’s phenomenon: A case series

Hafez Ghaheri, Mehrdad Karimi, Shervin Assari

ABSTRACT

Introduction: Stun guns are energy-conducting weapons that are becoming more frequently used by law enforcement officials to subdue combative individuals, or civilians for the case of security. Although complications related to stun guns are well known for the victim, less is known about stun guns complications among their users. Case Series: In this case series, we reported vibration-induced Raynaud’s phenomenon in four individuals who were recurrent users of stun guns. Two patients were male and two were female. Symptoms varied from moderate to severe. Although both hands were involved, symptoms were more severe in the right hand. Nifedipine was prescribed for all cases. Two patients received Losartan, as well. All patients reported a decrease in their symptoms in follow-up visits. Conclusion: While further research is needed, clinicians should remain vigilant to stun gun usage as a possible cause of Raynaud’s phenomenon.

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INTRODUCTION

Stun guns have been developed as non-lethal devices for law enforcement officers to control potentially violent subjects. Thus, stun guns are used as alternatives for firearms. Stun guns apply high voltage, pulsatile electric shocks to the subject, which result in involuntary skeletal muscle contraction. As a result, the subject is unable to further resist [1].

The use of stun guns is showing an increase among law enforcement officials because of the increasing demand for non-lethal weapons to subdue combative individuals. In the United States, in the year 2005, stun guns were being used by about 40% of law enforcement agencies, with about 150,000 devices in use by police officers. However, the use of stun guns is not limited to law enforcement officers. Worldwide, 100,000 civilians own a stun gun [2, 3]. In Canada, in only one province, more than 10,000 stun gun uses were recorded during six years [4].

Literature has provided conflicting results regarding the safety profile of stun guns for the victims. Although some studies among healthy volunteers have shown safety of these weapons [5–8], severe injuries have also been reported with its daily usage [9–14]. Main
injuries reported in victims include basilar skull fracture, comminuted nasal fracture, and orbital floor fracture; concussion, facial laceration, and subarachnoid and epidural hemorrhage necessitate craniotomy. Penetration of the outer table and cortex of the cranium by stun guns are also reported [8, 15]. One study in the United States reported an incidence of 0.25% for significant injuries following the use of electrical weapons. Main complications in this study included intracranial injuries and rhabdomyolysis. Mortality has been also reported [16].

Published literature about the safety of stun guns is limited to the victims, not to the users. Herein, we reported four cases of vibration-induced Raynaud’s phenomenon in the persons who fire the weapon, recurrently. In all cases, diagnosis was based on the user’s history of firing the weapon, in addition to a clinical and laboratory examination. Standard clinical tests included cold stimulation test and nail-fold capillaroscopy. Antinuclear antibody (ANA), erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) were also measured for some cases [17]. In all cases, a thorough investigation ruled out diagnosis of vasculitis.

CASE REPORT

Herein, we reported four cases of Raynaud’s phenomenon that may be related to stun gun use. Out of four patients, two individuals were users and the other two were sellers of stun guns. Two patients were men, and other two were women. Age ranged from 22 to 34 years.

Symptoms of Raynaud’s phenomenon had developed between 1 and 48 months after the initial use of stun guns. Stress or cold was the stimulant in three patients. Symptoms were severe in only one patient, other patients reported mild to moderate problems. Although both hands were involved in some cases, symptoms were always more prominent in one hand.

In all cases, treatment included avoiding use of stun guns in addition to medications. Nifedipine (for 18 to 90 days) was prescribed for all cases and two patients received Losartan (for 21 and 38 days), as well. All our patients reported decrease in symptoms in follow-up visits (Table 1).

DISCUSSION

We believe that Raynaud’s phenomenon in our patients may be related to vibration. Thus, we believe the diagnosis in our patients is vibration-induced Raynaud’s phenomenon. This diagnosis should be considered as a new possible complication among the users of stun guns.

As explained before, most previously reported stun gun injuries of eyes, genitalia, and large blood vessels are related to the victims, not the weapon users [18, 19]. The mechanism of vibration-induced Raynaud’s phenomenon is as follows:

Raynaud’s phenomenon has been explained by pathology in the central nervous system, autonomic dysfunction, and vasodysregulation. Symptoms are known to be a result of receptor and nerve ending dysfunction [20]. Stoyneva et al. reported in 2003, that vibration induces endothelial damage, as a result of an increase in shear stresses and elevated plasma level of thrombomodulin and of von Willebrand factor and reduced endothelium-dependent vasodilator responses. In patients with vibration induced Raynaud’s phenomenon, high concentrations of endothelin-1 especially in most advanced stages, and a decreased plasma thiol level have shown an increased production and activity of free radicals, which contributes to vasospastic paroxysms [21].

We used Nifedipine for all our cases in combination with Losartan for two of our patients. Calcium channel blockers such as Nifedipine and angiotensin II receptor antagonist alone or in combination have shown therapeutic effects in Raynaud’s phenomenon [22, 23].

Table 1: Summaries of data of four patients with vibration-induced Raynaud’s phenomenon due to frequent use of stun gun.

<table>
<thead>
<tr>
<th>Code</th>
<th>Gender</th>
<th>Age (years)</th>
<th>Smoking</th>
<th>Exposure nature</th>
<th>Exposure to stun guns (months)</th>
<th>Stress or cold stimulant</th>
<th>Right hand symptoms</th>
<th>Left hand symptoms</th>
<th>Dry gangrene Nifedipine (days)</th>
<th>Losartan (days)</th>
<th>Improved</th>
</tr>
</thead>
<tbody>
<tr>
<td>#1</td>
<td>male</td>
<td>34</td>
<td>Yes</td>
<td>Seller</td>
<td>48</td>
<td>Yes</td>
<td>Moderate</td>
<td>Mild</td>
<td>No</td>
<td>28</td>
<td>-</td>
</tr>
<tr>
<td>#2</td>
<td>female</td>
<td>22</td>
<td>No</td>
<td>User</td>
<td>3</td>
<td>No</td>
<td>Moderate</td>
<td>-</td>
<td>No</td>
<td>60</td>
<td>21</td>
</tr>
<tr>
<td>#3</td>
<td>male</td>
<td>28</td>
<td>Yes</td>
<td>Seller</td>
<td>1</td>
<td>Yes</td>
<td>Moderate</td>
<td>Mild</td>
<td>No</td>
<td>90</td>
<td>-</td>
</tr>
<tr>
<td>#4</td>
<td>female</td>
<td>28</td>
<td>No</td>
<td>User</td>
<td>2</td>
<td>Yes</td>
<td>Severe</td>
<td>Moderate</td>
<td>Yes</td>
<td>18</td>
<td>38</td>
</tr>
</tbody>
</table>
Although vibration-induced Raynaud’s phenomenon is a well-defined clinical entity, it is not very clear how stun gun users may develop this condition. Future research is needed on epidemiology, etiology, and natural history of Raynaud’s phenomenon among individuals who use stun guns.

CONCLUSION

Here we reported vibration-induced Raynaud’s phenomenon in users of stun guns. Stun gun users, including both law enforcement officials and civilians, should be informed of the potential serious complications of recurrent firing. However, any further conclusion needs further studies.

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Author Contributions

Hafez Ghaheri – 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

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

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Primary systemic amyloidosis presenting at an early age: A case report

Arghya Chattopadhyay, Shatarupa Dutta, Amitava Majumder, Jayanta Das

ABSTRACT

Introduction: Primary systemic amyloidosis arises from clonal B cell disorder and may be associated with myeloma or lymphoma. Amyloid deposition can occur in any organ in the body as well as skin lesions. Both sexes are affected by this disease, with onset most commonly in the sixth decade or after. Case Report: A 26-year-old female was presented with gradually changing quality of voice, diffuse pigmentation all over body and lymphadenopathy in upper cervical group. Biopsy of a lymph node showed follicular hyperplasia with a few discrete large cells. Over a period of time, she developed purpuric rash over extremities and periorbital area, respiratory distress and heart failure. Echocardiography revealed cardiac amyloidosis and rectal biopsy with Congo red staining was positive for amyloidosis. Skin biopsy from hyperpigmented lesion of right forearm was also suggestive of amyloidosis and bone marrow examination showed plasma cell dyscrasia with a small M-band in serum protein electrophoresis. Finally, it was diagnosed a case of primary systemic amyloidosis. Conclusion: We report a primary systemic amyloidosis with an early age presentation.

Keywords: Primary systemic amyloidosis, Heart failure, Skin rash, Plasma cell disorder

INTRODUCTION

Primary systemic amyloidosis (AL amyloidosis) arises from clonal B cell disorder and may be associated with myeloma or lymphoma. Plasma-cell clones produced immunoglobulin light chains or fragments of light chains that form extracellular amyloid fibrils. Amyloid deposition can occur in any organ in the body, causing features such as congestive cardiac failure, renal failure and hepatosplenomegaly as well as skin lesions. Both sexes are affected by this disease, with onset most commonly in the sixth decade or after. Although recent advances in therapy are encouraging, the prognosis for primary amyloidosis remains poor.

CASE REPORT

A 26-year-old female admitted in June 2010 with hoarseness of voice, diffuse pigmentation all over body
and lymphadenopathy in upper cervical group of lymph nodes for last one year. It was of insidious in onset and was progressive in nature. Detailed history taking did not reveal any history of fever, joint pain, joint swelling, bone pain or any other constitutional symptoms. Physical examination showed only upper cervical lymphadenopathy, diffuse skin pigmentation with thickening of skin of upper limb. There was no history of Raynaud’s phenomenon and the tongue was thick but of normal size. Differential diagnosis of collagen vascular disease, malignancy with paraneoplastic syndrome, tuberculosis and lipoid proteinosis were made. Routine blood tests and thyroid profile, Antinuclear antibody (ANA), double-stranded deoxyribonucleic acid (ds DNA), Rheumatoid factor (RF), hepatitis B surface antigen (HBsAg), Anti-hepatitis C virus (Anti-HCV) antibody and human immunodeficiency virus (HIV) antigen were within normal limit. Mantoux (with 10 U tuberculin) was positive with 21 mm induration and erythema. Chest X-ray (posterior-anterior view) showed perihilar lymphadenopathy. Contrast enhanced computer tomography of thorax confirmed the X-ray findings but failed to specify the nature of the lesions better than chest X-Ray. Biopsy from the cervical lymph node showed follicular hyperplasia with few discrete large cells and CD 15, CD30 and BCl2 were advised, but the patient sought discharge from hospital for personal reason without undergoing these investigations.

In February 2011, she got readmitted with complaints of respiratory distress, bilateral swelling of feet, and a few purpuric skin rashes over extremities and face. These lesions resolved spontaneously and similar purpuric rash in periorbital region appeared (Figure 1). She also reported pain and tingling sensation in the distal parts of all four limbs and the skin pigmentation has increased remarkably. Physical examination revealed persistence of upper cervical lymphadenopathy, bilateral pitting pedal edema with engorged and pulsatile neck veins and gallop rhythm, orthopnea, decreased vocal resonance and breath sound over lower zone of right lung.

Considering the new features suggestive of heart failure, skin rash and pigmentation a new set of differentials was considered in addition to the previous ones including the possibility of primary systemic amyloidosis.

Laboratory workup revealed leukocytosis with a positive urine culture of *Klebsiella pneumoniae*. Chest X-ray showed persistence of perihilar lymphadenopathy with right sided pleural effusion. Pleural fluid study was transudative. Serological tests encompassing viral markers and autoimmune profile were negative. Abdominal ultrasonography did not reveal any significant findings. Nerve conduction study was suggestive of entrapment neuropathy of right median nerve at wrist joint and bilateral peroneal neuropathy.

Echocardiography (Figure 2) revealed increased ventricular wall thickness with ground glass appearance of myocardium, restrictive left ventricular filling pattern and moderate pericardial effusion posteriorly with borderline left ventricular systolic function and thickened mitral and tricuspid valves. These features were strongly suggestive of cardiac amyloidosis.

Rectal biopsy with Congo red staining revealed orangophilia in amorphous deposits around the blood vessels, suggestive of amyloidosis. Skin biopsy was also consistent with amyloidosis (Figures 3 and 4).

Bone marrow examination revealed 15% plasma cell and suggestive of plasma cell dyscrasia (Figure 5), with M band in serum protein electrophoresis. Twenty-four hour urinary protein was 440 mg without any light chain on electrophoresis.
A final diagnosis of primary systemic amyloidosis was made. The patient was treated with pulse dexamethasone and cyclophosphamide and daily thalidomide. Patient tolerated the drugs well and was discharged with a plan of bone marrow transplant.

**DISCUSSION**

Amyloidosis is the term for diseases caused by the extracellular deposition of insoluble polymeric protein fibrils in tissues and organs. These diseases are a subset of a growing group of disorders attributed to misfolding of proteins. Polypeptide chain gets folded to form a secondary structure of the protein molecule. Helical structure and pleated structure are two important secondary structures of protein molecule. The amyloid protein has a beta-pleated sheet structure, which makes it highly insoluble and resistant to proteolytic digestion and hence difficult to remove from the tissues [1]. About 25 different proteins are known to produce amyloid fibrils in human, most of them are constituents of plasma. These normally soluble precursor proteins, due to some unknown reason, get misfolded and forms a beta-pleated sheet structure and becomes amyloid. Inherited amyloidosis is due to mutation in certain precursor protein, which makes them susceptible to misfolding. In case of primary systemic amyloidosis, the amyloid is derived from monoclonal immunoglobulin light chain and is called AL amyloid where L stands for light chain of immunoglobulin molecule. In case of secondary amyloidosis, which is associated with many chronic inflammatory diseases, amyloid fibrils are derived from cleavage fragment of the circulating acute phase reactant serum amyloid A protein (SAA), hence this type is called AA amyloid. Serum amyloid A protein is synthesized in liver during inflammation [2]. In localized cutaneous amyloidosis, amyloid is derived from keratin released from apoptotic keratinocytes [3]. The possible reason that many diverse conditions are associated with amyloidosis is each of these conditions results in excessive production of proteins that are prone to misfolding [2].

Figure 3: Rectal biopsy showing edematous sub mucosa and deposition of amorphous eosinophilic material predominantly around the blood vessels.

Figure 4: Skin biopsy showing deposition of amorphous material suggestive of primary systemic amyloidosis.

Figure 5: Bone marrow of the patient.
Amyloid deposition can occur in any organ. Primary systemic amyloidosis is known for highly varied clinical manifestations, including organomegaly and organ dysfunction. In tissues leads to distortion of tissue architecture, organ enlargement (organomegaly) and organ dysfunction. Amyloid deposition can occur in any organ. Primary systemic amyloidosis is known for highly varied clinical manifestation.

The mean age of presentation of primary systemic amyloidosis is 50 years, but this case presented at 26 years of age which is really unusual presentation. In literature we found a case report of unusual presentation at 39 years who first developed cutaneous reactions at 18 years age [5]. No other case report of unusual age presentation has been reported.

Cutaneous involvement is seen in 40% patients with AL amyloidosis. Cutaneous manifestation depends upon the site of amyloid deposited. Amyloid deposition in superficial dermis produces shiny waxy translucent papules and common sites for predilection are eyelids, retroauricular areas, neck and axilla. Amyloid deposits around pilosebaceous unit leads to the destruction of hair, producing alopecia. Diffuse infiltration of scalp skin results in the thickening of skin which gets thrown into longitudinal folds resembling cutis verticis gyrata. Diffuse infiltration of large area of skin may simulate scleroderma. Infiltration of nail matrix by amyloid may produce ridging; splitting and brittleness of nail plate [2].

Prognosis in AL amyloidosis is poor and major causes of death are cardiac and renal failure. The amyloid infiltration of vessel wall causes capillary wall fragility, which leads to purpura and ecchymosis after a minor trauma or even spontaneously. Periorbital area is one of the common sites of expression of purpura, as evident in this case. The capillary fragility may be demonstrated by pinching the skin. Purpuric lesions with normal platelet count and normal coagulation profile should suggest the possibility of capillary fragility. Amyloid deposition in tongue leads to macroglossia. Tongue is diffusely enlarged and firm and there may be tooth indentation along its lateral border. Amyloidosis is the most common cause of macroglossia in adults [1]. Macroglossia if severe might lead to dysphagia. Macroglossia was present in our patient. Hepatomegaly occurs in 50% patients and splenomegaly in 10% patients. Cardiac involvement leads to conduction defects, arrhythmias, congestive cardiac failure and may account for 40% of deaths. Cardiac involvement in terms of cardiac amyloidosis with features of congestive heart failure was present in this case. Cardiac involvement may occur in peripheral nerves leading to thickening of nerves and resulting neuropathy often mimicking Hansen’s disease. Renal involvement presenting with proteinuria and renal failure, is one of the bad prognostic indicators.

Our patient had plasma cell dyscrasia and M band in serum protein electrophoresis. The diagnosis was confirmed by demonstration of amyloid in rectal biopsy and skin biopsy. Clinically, it is difficult to distinguish primary, secondary or familial form of amyloidosis. Immunohistochemical staining using commercially available antisera is useful for classifying the type of amyloid deposited in tissues [6]. Biopsy is very important for the diagnosis. Hematoxylin and eosin staining suggests the possibility of amyloidosis but Congo red staining confirms the diagnosis. Congo red staining results in a brick red color of amyloid when seen under ordinary light and under polarized light shows classical green birefringence. Unfortunately, polarized microscopy is not easily available in developing country like India. In systemic amyloidosis, amyloid deposits are seen in dermis, subcutaneous tissue and blood vessels, whereas in localized cutaneous amyloidosis, deposits are seen only in papillary dermis; subcutaneous tissues and blood vessels are not involved. Tuberculosis is definitely a common entity in India, and we also considered it among one of the differentials but in this case it is unlikely to be a case of tuberculosis because: though it is a chronic disease it is unusual to be such a long course of disease as the disease started a year before presentation and it took another 9–10 months to diagnose the case, lymph node biopsy was not suggestive of tuberculosis so cannot be explained under one umbrella. Echocardiography was very much suggestive of amyloidosis, bone marrow aspiration shows 15% plasma cell, serum protein electrophoresis shows M band and rectal and skin biopsy median survival of patients with myeloma-associated amyloidosis is five months and for patients with primary systemic amyloidosis is 2.1 years [1]. Prognosis depends upon the extent of involvement. Treatment of amyloidosis is aimed at reducing the supply of precursor proteins [3]. In AL amyloidosis, the precursor is immunoglobulin light chain produced by B lymphocytes/plasma cells hence treatment with cytotoxic agents like melphalan and prednisolone that reduces plasma cell proliferation is useful [3]. Chemotherapy will be useful only when precursors are supplied by plasma cells like AL amyloidosis.
CONCLUSIONS

We report a primary systemic amyloidosis with an early age presentation.

**********

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

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REFERENCES

Maternal death as a complication of uterine perforation with associated bowel prolapse per vaginum following attempted termination of pregnancy at a peripheral hospital: A case report

Labaran D Aliyu, Makama B Salihu

ABSTRACT

Introduction: Induced abortion is a procedure carried out to terminate a pregnancy for varied indications. It can, however, be complicated by severe complications including bowel injury and in some cases death. Complications are common when the procedure is carried out by the untrained or poorly trained personnel. Case Report: This is a case of uterine perforation associated with bowel prolapsed through the vagina following an attempted termination of a missed abortion at eighth week and six days in a 25-year-old G3P2+0 (with 2 living children). The procedure was carried out by paramedics at a local General Hospital. Misoprostol was first used to induce abortion. When it failed, oxytocin infusion was used and when it also failed, dilatation and curettage was attempted which resulted in the complications mentioned and the patient subsequently died on the fourth postoperative day of an exploratory laparotomy. This case showed how poorly or untrained personnel are still using all means available to terminate pregnancies even in our hospitals resulting in serious complications. Conclusion: Complications of induced abortions in our environment are still common as untrained or poorly trained personnel engage in such procedures with grave consequences. To reduce these complications there is a need for proper training and provision of proper equipment and ensuring proper supervision of medical facilities.

Keywords: Uterine perforation, Induced abortion, Bowel injury, Maternal death

INTRODUCTION

Unsafe abortion is defined as a procedure for termination of unwanted pregnancy either by persons lacking the necessary skills or in an environment lacking the minimal medical standard or both [1]. It is an important cause of maternal morbidity and mortality. It contributes 40% of maternal mortality in Nigeria [2]. Uterine perforation is a recognized complication of unsafe abortion. It occurs in 0.4–15 cases per 1000 induced abortions depending on the study population [3]. Bowel injury is uncommonly reported yet serious complication of induced abortion which is often performed illegally by persons without any medical training in developing countries [4, 5]. This is clearly demonstrated in this case. A sudden increase in cases of bowel injury as a consequence of induced abortion prompted researchers in India to conduct a study into the problem [5]. Many
studies were carried out in Cameroon, Nigeria and Ghana on this derailed complication of induced abortion [4, 6, 7]. The fact that in this particular case the pregnancy was not unwanted, the procedure was done because of a missed abortion in a public hospital, under anesthesia, having failed to terminate the pregnancy with misoprostol and oxytocin infusion make the case an interesting one to report. It clearly demonstrates the pathetic state of our health care delivery system where seeking care in a health facility does not necessarily guaranty that patients are safe from unnecessary and potentially life-threatening complications because such facilities are poorly equipped and manned by ill trained personnel.

CASE SERIES

A 25-year-old G3P2+0 (with 2 living children) patient came into the Gynae emergency room of our hospital with a referral letter from a public hospital in her locality. She had been amenorrheic for about eight weeks prior to presentation and had intermittent episodes of bleeding per vaginum. At presentation at a local hospital a pelvic ultrasound scan was done which revealed a missed abortion at eight week and six days. She was informed of the result and told that the pregnancy has to be removed which she consented to. Initially, misoprostol was used vaginally to induce abortion. When this failed, oxytocin infusion was used which also failed. Two days later she was told that since these two methods did not work she will undergo dilatation curettage (D&C) to remove the pregnancy.

The D&C procedure was carried out and she later started experiencing severe lower abdominal pain and was bleeding through the vagina. The relatives were told also there that the pregnancy could not be removed completely and part of it is protruding through the vagina and the patient will be referred to the Teaching Hospital in the city where the rest will be removed. She presented more than 24 hours after the procedure because they had difficulty getting money and transportation.

At presentation, she was drowsy with offensive smell around her. She was pale, febrile (38.10°C), anicteric and dehydrated. Respiratory rate 26/min, her chest was clinically clear. Pulse rate was 104/minute, blood pressure was 90/60 mmHg. Her abdomen was slightly distended and there was generalized tenderness, bowel sounds were hypoactive. Pelvic examination revealed vulva and vagina smeared with blood. A vaginal pack was removed and dark pink intestines started coming out through the vaginal introitus with slight bleeding.

An intravenous line was secured and two liters of normal saline were rushed and another liter was kept running at slower rate. She was immediately placed on intravenous ceftriazone and metronidazole. Nasogastric tube was inserted and she was catheterized. Samples were taken for complete blood count, random blood sugar, serum electrolytes and urea and 4 pints of blood were cross matched. Her hemoglobin was 8 g/dL and other results were not available before surgery. A surgical team was called to review the patient. A joint decision was taken to carry out an exploratory laparotomy. The patient was counselled and her consent obtained. About 10 hours after presentation she was taken to the theatre for surgery.

Findings at Operation

Offensive hemoperitoneum of about 1000 mL, a bulky uterus of about 16 weeks size, a posterior uterine perforation about 4 cm in diameter with rugged edges with dark pink ileal tissue passing through the perforation about 30 cm long, the tubes and ovaries were enlarged and hyperemic. A linear laceration on the anterior aspect of the upper 1/3 of the rectum with devitalized edges, distal sigmoid colon lacerated with irregular devitalized edges. The uterine perforation was enlarged and placental tissues evacuated gently and the perforation repaired after excising the devitalized edges. Refashioning and anastomosis of the sigmorectoid and ileocecal junctions were done. The peritoneal cavity was copiously lavaged and a corrugated drain passed into pouch of Douglas and fixed to the skin. The abdomen was closed in layers. Two pints of blood were transfused intraoperatively, IV ceftriazone and metronidazole were also given. Intraoperative blood loss was estimated to be 750 mL. Patient was transfused two more pints of blood postoperatively (Figures 1 and 2).

Patient fared fairly well postoperatively with relatively stable vital signs and was making adequate urine. On the fourth postoperative day she started running temperature and her condition suddenly deteriorated and she died the same day.

Findings at Operation

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Patient fared fairly well postoperatively with relatively stable vital signs and was making adequate urine. On the fourth postoperative day she started running temperature and her condition suddenly deteriorated and she died the same day.
Induced abortion has remained a source of serious concern to all gynecologist worldwide especially those in the developing world. This is so because in our environment there are still factors militating against the performance of safe abortion. Most worrisome are untrained quacks whose motive may be financial and their skills negligible [8]. A major barrier to care in many developing countries is the lack of basic equipment and drugs for all gynecological care [9]. Restrictive abortion laws is another important hindrance to safe abortion because women seeking abortion are left with no alternative other than procuring it through the backdoor which make them vulnerable to all complications including death. Legalization of abortion although important is insufficient. India has had legal abortion on the books for several decades as has Zambia, however the devil is in the details [10]. Without skilled providers, adequate facilities and early access, the promise of safe, legal abortion will remain unfulfilled, as in the case of India and Zambia [11]. Bowel injury is a common consequence of uterine perforation either with pointed instruments or grasping forceps. The distal ileum appears to be most vulnerable followed by the sigmoid [12]. The patient in question had similar injuries. Such injuries are associated with peritoneal soilage with fecal material containing virulent intestinal bacteria, which invariably leads to peritonitis and generalized septicemia; this will lead to serious morbidity and in extreme cases death will result as in this case. Early recognition of the injury, adequate resuscitation and experienced surgical intervention may be the only solution if the life of the patient is to be saved. These are usually not possible in developing countries and as a result patients die. Our patient could not come immediately because of financial difficulty and challenges of transportation. This delay must have compounded her clinical condition at presentation.

To arrest this pathetic situation, Governments in partnership with nongovernmental organizations, religious groups and women groups should embark on extensive community awareness campaigns on the issue of abortion and its consequences. Governments should improve the health care delivery system through training, provision of equipment and facilities to be made available and accessible at all times and to most communities no matter how remote they may be. There is also the need to review our abortion laws so that morbidity and mortality attributed to abortion will be drastically reduced.

**CONCLUSION**

Complications of induced abortions in our environment are still common as untrained or poorly trained personnel engage in such procedures with grave consequences. To reduce these complications there is a need for proper training and provision of proper equipment and ensuring proper supervision of medical facilities. There is also a need for public enlightenment on induced abortion and its consequences.

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**Author Contributions**

Labaran D Aliyu – 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

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

**Guarantor**

The corresponding author is the guarantor of submission.

**Conflict of Interest**

Authors declare no conflict of interest.

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Retroperitoneal liposarcoma: A case report

M Sandra Jacob, Shirali Patel, Harvey Sasken, Yomayra Perez, Valerie Katz, Mark Ingram

ABSTRACT

Introduction: We report an interesting case of a 67-year-old female presented with symptomatic cholelithiasis and was found to have an occult retroperitoneal sarcoma on work up. Case Report: A 67-year-old female was referred to the surgery clinic by the gynecology service for symptomatic cholelithiasis. On examination, she was moderately obese with mild right upper quadrant tenderness and a reducible incisional hernia. An abdominal ultrasound revealed cholelithiasis and a left retroperitoneal flank mass. She was referred for abdominal computed tomography scan and magnetic resonance imaging scan which revealed a large left retroperitoneal cystic mass adherent to the left kidney. She underwent en-bloc resection of retroperitoneal tumor, cholecystectomy, and repair of incisional hernia. Her postoperative course was uneventful and she continues to do well without adjuvant chemoradiation. Discussion: One-third of malignant tumors located in the retroperitoneum are sarcomas. The median age of presentation occurs in the sixth decade. As with our patient complete surgical resection is the optimal treatment for patients. The addition of adjuvant radiation therapy to surgical resection is associated with both a reduced risk of local recurrence and a longer recurrence-free interval, but it does not improve overall survival. Conclusion: The review of the literature emphasizes that the management of retroperitoneal sarcomas consists of complete resection of the tumor with adjuvant radiotherapy (if the tumor is high grade) combined with surveillance for early liposarcoma detection of recurrence or metastases.

Keywords: Sarcoma, Liposarcoma, Retroperitoneal tumor

INTRODUCTION

Soft-tissue sarcomas are relatively rare with approximately 8,600 new cases annually and represent less than 1% of all newly diagnosed malignancies in the United States. Retroperitoneal sarcomas are malignant tumors arising from mesenchymal cells, which are usually located in muscle, fat, and connective tissues. One-third of malignant tumors located in the retroperitoneum are sarcomas, and approximately 15% of soft tissue sarcomas arise in the retroperitoneum [1]. According to the World Health Organization (WHO), soft-tissue liposarcomas are categorized into five distinct histological subtypes: well-differentiated, dedifferentiated, myxoid, pleomorphic and mixed type. Retroperitoneal sarcomas have varying clinical courses depending on their histological subtype and grade [1, 2].

The pathologic diagnosis of liposarcoma rests on the identification of lipoblasts in a milieu of supporting...
histomorphologic features. The well-differentiated liposarcoma is a low-grade neoplasm which can present as five histological variants: lipoma-like, sclerosing, inflammatory, spindle cell and liposarcoma with meningotheial whorls. The treatment of choice is complete surgical excision. According to Stoeckle et al., there are no survival benefits of adding adjuvant radiotherapy at this time for a resected well-differentiated retroperitoneal liposarcoma [3].

CASE REPORT

A 67-year-old female was presented to the gynecologist for screening Pap smear. The patient complained at that time of right upper quadrant abdominal pain. Her past medical history was significant for hypertension, asthma, and hyperlipidemia which were well-controlled and there was no significant family history. She had a previous midline scar from a total abdominal hysterectomy with bilateral salpingo-oophorectomy for fibroid. On examination, her abdomen was very obese (BMI 44.8) with right upper quadrant tenderness. No mass was palpated and she had a reducible incisional hernia. Abdominal ultrasound revealed a large heterogeneous left flank mass and cholelithiasis. Origin of the mass was uncertain, computed tomography (CT) scan was recommended for further assessment. She was subsequently referred to surgery for management and imaging studies. The CT scan of abdominal showed a large retroperitoneal mass with displacement of the retroperitoneal organs (Figure 1A–B). The origin and blood supply of the mass could not be determined on the CT scan and magnetic resonance imaging (MRI) scan was recommended, revealing a large complex retroperitoneal cystic mass adherent to the left kidney which extended from the splenic hilum inferiorly to the left lower abdomen (Figure 2).

The case was presented at the multidisciplinary tumor board. The recommendation was to proceed with surgery first. Neoadjuvant chemotherapy was not recommended as there was no tissue diagnosis. After discussion with the patient and her family, she underwent an exploratory laparotomy with en-bloc resection of the retroperitoneal tumor. At surgery, there was a left retroperitoneal mass as per, adherent to the left kidney, but separate from the spleen, pancreas, and colon. The mass was resected en-bloc with the kidney. The gallbladder was removed for chronic cholelithiasis which was symptomatic and her incisional hernia was repaired. Her postoperative course was uneventful.

The mass was well circumscribed and globular composed of fleshy homogeneous yellow tan tissue. The tumor weighed 1670 grams and measured 30x25x15 cm (Figure 3). Routine tissue stain demonstrated a well-differentiated liposarcoma, characterized by its hypocellularity, nuclear pleomorphic atypia and delicate vascularity. Some of these adipocytes have a single vacuole and others demonstrated a floret giant cell configuration (Figure 4). An atypical lipoblast was demonstrated in (Figure 5). This large cell had an enlarged nucleus, irregular in shape, with variably clumped chromatin and the cytoplasm contained numerous vacuoles.

The stroma was variably fibrillar with areas of abundant ground substance. A mild inflammatory infiltrate was present with a significant quantity of plasma cells. Final pathology revealed the tumor to be a low grade, well-differentiated, stage T2bG1N0M0 retroperitoneal liposarcoma. The medical and radiation oncologists suggested observation and interval follow-up for surveillance.
Retroperitoneal tumors are an extremely heterogeneous group of neoplasms, 85% of which are malignant. Liposarcomas constitute between 45–55% of retroperitoneal masses [4]. Age at presentation is younger compared with most other malignancies, with many being diagnosed between 54–65 years of age [5]. There is an equal male/female ratio [1]. The distribution of soft tissue sarcomas by anatomic site can be found in an article by Lawrence et al. [6].

Retroperitoneal sarcomas present 80% of the time as an asymptomatic abdominal mass. Symptoms can also be related to mass effect or local invasion which may lead to pain, gastrointestinal obstruction, feelings of early satiety, and weight loss. In addition, neurologic and muscular skeletal symptoms are referred to the lower extremities [7].

Histopathologic variety is the main prognostic factor. Five histologic types are recognized. Well differentiated liposarcoma represents around 30% like our case and has the best prognosis. The myxoid type is the most frequent liposarcoma, constituting around 50% of all tumors. It has a less favorable progression, as it often recurs early. The pleomorphic, round cell and undifferentiated types display the worst prognosis [4].

After a physical examination CT scan provides an excellent understanding of the relationship between nearby structures and is critical to preoperative planning. A patient presenting with a palpable abdominal mass,
should be have a high-resolution, thin-cut CT scan with intravenous and oral contrast since these images allow for further distinction between intra-abdominal and retroperitoneal structures. This allows a discussion of the need for biopsy if indicated, the operative plan, and the preparedness of the operative team, as well as a discussion with the patient regarding the risks and benefits. The differential diagnosis includes a primary neoplasm arising from a retroperitoneal visceral structure (e.g., pancreas, adrenal glands, kidneys, and duodenum), a retroperitoneal sarcoma, a lymphoma, or a metastatic lesion [5].

The optimal treatment for patients with localized, resectable retroperitoneal sarcomas is surgery with gross and microscopically negative margins. Complete surgical resection frequently requires en-bloc resection of adjacent viscera [8]. The kidney was the most frequently resected organ (36%) followed by segmental resection of the large bowel, spleen, and pancreas [9].

The addition of adjuvant radiation therapy to surgical resection is associated with both a reduced risk of local recurrence and a longer recurrence-free interval. However, it does not improve overall survival. Studies have demonstrated the advantages of preoperative radiotherapy in the management of marginally resectable retroperitoneal sarcomas. The benefits of pre-operative radiation are multiple [3, 10]. It allows for the gross tumor volume to be readily definable for accurate treatment planning. Moreover, the tumor displaces radiosensitive viscera. Thus, no adhesions and tethering of bowel to the tumor bed can occur and the tumor is treated in situ.

Another treatment modality is intra-operative radiotherapy (IORT) which is targeted to a specific region allowing for maximum doses of radiation to the tumor bed. Studies show that IORT improves tumor control in the field. However, it does not influence recurrence-free or overall survival rates [9, 10].

CONCLUSION

The review of the literature emphasizes that the management of retroperitoneal sarcomas consists of complete resection of the tumor followed by adjuvant radiotherapy reduce local recurrence but does not affect overall survival and combined with surveillance for early detection of recurrence or metastases. Imaging studies are essential for proper preoperative planning and allow assessment of resectability prior surgery; preoperative radiotherapy can be considered in patients with questionably resectable tumors. Contrast-enhanced computed tomography scan and magnetic resonance imaging were valuable aids in our case. The patient should be closely followed with regular physical examinations and imaging studies such as chest X-rays and computed tomography scans. Our patient continues to follow-up for surveillance and is doing well.

REFERENCES

Perforation of Meckel’s diverticulum caused by a fish bone: A case report

Hideki Shibata, Koichi Sato, Takashi Tada, Hiroshi Maekawa, Mutsumi Sakurada, Hajime Orita, Tomoaki Ito

ABSTRACT

Introduction: Meckel’s diverticulum is the most common congenital anomaly of the gastrointestinal tract. Perforation of Meckel’s diverticulum caused by a fish bone is a very rare complication. Case Report: We report a 41-year-old male who was admitted to our hospital with deteriorating lower abdominal pain that started two days earlier. An abdominal computed tomography scan showed inflammatory changes thickening the intestinal wall and involving ileocecal fat. Based on a clinical diagnosis of acute appendicitis, we performed an emergency operation. At laparotomy, Meckel’s diverticulum was perforated by a fish bone. Conclusion: In the differential diagnosis as a probable cause of acute abdomen, it is important that computed tomography scan of the abdomen should be checked carefully while considering Meckel’s diverticulum.

Keywords: Meckel’s diverticulum, Perforation, Fish bone

INTRODUCTION

Meckel’s diverticulum is the most common congenital anomaly of the gastrointestinal tract [1]. Meckel’s diverticulum represents a true diverticulum of the ileum, containing all three layers of the bowel wall, and is invariably found on the anti-mesenteric border of the ileum, with 90% located within 90 cm of the ileocecal valve [2]. In general, Meckel’s diverticulum causes no symptoms. However, it can sometimes result in abdominal pain that requires treatment. Patients with perforation of Meckel’s diverticulum may present with right iliac fossa pain, which mimics acute appendicitis. Here, we present an interesting and unusual case of perforation of Meckel’s diverticulum; a very rare complication caused by a fish bone.

CASE SERIES

A 41-year-old male was admitted to our hospital from an outside facility for deteriorating lower abdominal pain that started two days earlier. On admission he showed tenderness in the right lower abdomen without rebound tenderness or defense. His vital signs showed blood pressure 108/62 mmHg, a regular pulse rate 84/min and temperature 36.9°C. His blood test revealed a raised white cell count of 19.1x10³/μL, a high C-reactive protein level 6.2 mg/dL, and high total bilirubin 2.2 mg/dL.
An abdominal computed tomography (CT) scan showed inflammatory changes thickening the intestinal wall and involving ileocecal fat. There was also a 6 mm high density spot in the intestinal wall at the same area (Figure 1). Based on a clinical diagnosis of acute appendicitis, we performed an emergency operation.

At laparotomy, there were a few ascites and an inflamed appendix. Meckel’s diverticulum was perforated by a fish bone about 50 cm from the Bauchini valve and its tip had impaled the cecum (Figure 2). The patient was treated with resection of the perforated ileum, including Meckel’s diverticulum, simple closure of the cecum and appendectomy.

A surgical specimen revealed a 2×3 cm Meckel’s diverticula perforated by a 2.5 cm fish bone. A pathologic examination of the specimen confirmed the presence of inflamed full-thickness mucosa with ectopic gastric mucosa in the diverticulum (Figure 3). The patient made an uneventful recovery postoperatively and was discharged from the hospital on postoperative day-11.

**Figure 1:** Abdominal computed tomography scan showing inflammatory changes thickening the intestinal wall and involving the ileocecal fat. The arrow shows a 6 mm high density spot in the intestinal wall at the same area.

**Figure 2:** Meckel’s diverticulum perforated by a fish bone.

**Figure 3:** The pathologic examination of the specimen confirms the presence of an inflamed full-thickness mucosa with ectopic gastric mucosa in the diverticulum (H&E stain, ×400).

**DISCUSSION**

Meckel’s diverticulum was first described by Fabricus Hildanus in 1598, and was later named by the German anatomist, Johann Friedrich Meckel, who described its embryological origin in 1809 [3]. Meckel’s diverticulum is a remnant of the omphalomesenteric duct, which is normally obliterated by the fifth week of gestation [4]. It is the most common congenital abnormality of the gastrointestinal tract, classically thought to occur in about 2% of the population. Despite the fact that this condition is relatively common, only about 4–16% of cases lead to complications [5]. Bleeding from Meckel’s diverticulum due to ectopic gastric mucosa is the most common clinical presentation, especially in younger patients, but it is rare in the adult population. The complications in adults include obstruction, intussusception, ulceration, hemorrhage, and rarely vesico-diverticular fistula and tumors.

A very small percentage of ingested foreign bodies can cause perforation of the bowel, leading to acute abdomen requiring surgical intervention. Foreign bodies such as dentures, fish bones, chicken bones and cocktail sticks have been known to cause bowel perforation. There are more than 300 cases in literature of bowel perforation caused by foreign bodies [6]. The majority of patients do not recall ingesting the foreign body, it being discovered either on investigation or during an operation. Perforation of Meckel’s diverticulum caused by a fish bone is very rare; only 28 cases have been reported in Japan, including the current patient. Analyses of these 28 cases are given in Table 1.

Perforation of Meckel’s diverticulum remains a differential diagnosis of right iliac fossa pain. Meckel’s diverticulum is notoriously difficult to diagnose, both clinically and radiologically, as the symptoms and imaging features are non-specific. In our patient, appendicitis was
<table>
<thead>
<tr>
<th>Case</th>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Chief Complaint</th>
<th>Period of illness</th>
<th>Preoperative diagnosis</th>
<th>Operation</th>
<th>Size of fish bone (cm)</th>
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<tr>
<td>25</td>
<td>Oikawa(2007)</td>
<td>31</td>
<td>Male</td>
<td>Lower abdominal pain</td>
<td>4 days</td>
<td>Perforated peritonitis</td>
<td>Partial resection of ileum</td>
<td>2.5</td>
</tr>
<tr>
<td>26</td>
<td>Moriiuchi(2007)</td>
<td>71</td>
<td>Male</td>
<td>Lower abdominal pain</td>
<td>2 days</td>
<td>Strangulated ileus</td>
<td>Diverticulectomy</td>
<td>2.2</td>
</tr>
<tr>
<td>27</td>
<td>Murohashi(2009)</td>
<td>67</td>
<td>Male</td>
<td>Lower abdominal pain</td>
<td>1 day</td>
<td>Perforated peritonitis</td>
<td>Diverticulectomy</td>
<td>4.3</td>
</tr>
<tr>
<td>28</td>
<td>Our case</td>
<td>41</td>
<td>Male</td>
<td>Lower abdominal pain</td>
<td>2 days</td>
<td>Acute appendicitis</td>
<td>Simple closure of the cecum and appendectomy</td>
<td>2.5</td>
</tr>
</tbody>
</table>

Abbreviation: Rt - Right
suspected on CT scan and laboratory findings because of the inflammatory changes. However, at laparotomy, a high density spot at the preoperative CT scan was found to be a fish bone and we ultimately diagnosed Meckel’s diverticulum and a perforated cecum because of this.

The management of symptomatic Meckel’s diverticulum is surgical resection. A wedge resection of Meckel’s diverticulum is generally carried out, and occasionally some ileum is resected by end-to-end anastomosis [7]. Recently, the prevalence of laparoscopic surgery has been increasing. However, a preoperative diagnosis of a complicated Meckel’s diverticulum may be challenging because of the overlapping clinical and imaging features of abdomen. An adequate knowledge of embryological, clinical, pathologic and radiologic characteristics of Meckel’s diverticulum will aid the early and accurate diagnosis of complicated cases [5].

CONCLUSION

Perforation of Meckel’s diverticulum caused by a fish bone is a very rare complication and may lead to a fatal outcome if not recognized early. In the differential diagnosis as a probable cause of acute abdomen, it is important that computed tomography scan of the abdomen should be checked carefully while considering Meckel’s diverticulum.

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Author Contributions

Hideki Shibata – Substantial contributions to conception and design, acquisition of data, and analysis and interpretation of data, Drafting the article, Revising the article critically for important intellectual content, Final approval of the version to be published

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CASE REPORT

OPEN ACCESS

Bilateral synchronous breast cancer in an elderly man: A case report

Unmesh Vidyadhar Takalkar, Shilpa Balaji Asegaonkar, Balaji Narayan Asegaonkar, Pushpa Ravindra Kodlikeri

ABSTRACT

Introduction: Bilateral synchronous breast cancers are seen extremely uncommonly in men. In this report, we describe a case of synchronous bilateral breast cancer in an elderly male without any significant risk factor. Case Report: A 75-year-old male was presented with mass in right breast. After thorough clinical evaluation he underwent modified radical mastectomy for excision of tumor. Histopathologically, it was invasive duct carcinoma of grade II, stage II and positive for estrogen and progesterone receptors. Patient was managed with radiotherapy, adjuvant chemotherapy and oral tamoxifen 20 mg daily. After three months new lump in contra lateral breast was noticed. Again patient underwent modified radical mastectomy with diagnosis of invasive duct carcinoma of grade II and stage II. He received radiotherapy and hormone therapy. We managed the case of synchronous bilateral breast cancer successfully at our center. After bilateral modified radical mastectomy patient received radiotherapy, adjuvant chemotherapy and hormonal therapy with tamoxifen. Conclusion: Proper clinical evaluation, mammography and fine-needle aspiration cytology play important role in diagnosis of male breast cancer. In depth studies focusing on etiopathology of the disease are necessary to optimize the management of male breast cancer patients.

Keywords: Male breast cancer, Bilateral synchronous breast cancers, Invasive duct carcinoma

How to cite this article


INTRODUCTION

Breast cancer is a relatively rare entity in males accounting for less than 1% of all cancer in men and 0.5–0.7% of all breast cancers [1]. Bilateral synchronous breast cancers are extremely uncommon in men. Hence they have not been studied deeply from investigation and management point of view. In this report we describe a rare case of synchronous bilateral breast cancer in an elderly male without any significant risk factors.

CASE REPORT

A 75-year-old male was presented to our center with complaint of mass in the right breast. On acquisition of detailed medical history, there was no history of hormonal therapy, testicular disease and occupational exposure...
to radiation in his lifetime. He was nonsmoker and nonalcoholic. There was no personal and family history of breast cancer. On physical examination, there was a mass in upper outer quadrant of size 4x3 cm under nipple with evidence of bloody nipple discharge. External genitalia examination was normal. Mammography revealed a 3-cm radiopacity with regular margins in retroareolar region. Fine-needle aspiration cytology showed presence of malignant cells. Contra lateral breast was normal on physical examination. The presence of distant metastasis was excluded by computed tomography scan and abdominal ultrasound scan. Laboratory work-up including hematology, liver, kidney and thyroid function tests was within normal limits. Patient underwent modified radical mastectomy with axillary lymph node resection. On gross examination, tumor was of size 4x3x3 cm. Histopathological examination revealed features of invasive duct carcinoma, grade II. Nipple and cut margins of specimen were free of tumor. Also there was no evidence of vascular and perineural invasion. Three of seven axillary lymph nodes were positive for the presence of malignant cells. As per tumor node metastasis (TNM) classification tumor was of stage II. On immunohistochemistry analysis, tumor cells were positive for estrogen receptor (60%) and progesterone receptor (50%).

Patient received six cycles of adjuvant chemotherapy with cyclophosphamide, 5-fluorouracil, epirubicin. After surgical management external radiotherapy was administered to chest wall and axilla with doses of 50 Gy (2 Gy daily with five weekly fractions) for a period of five weeks. Also patient was advised for daily oral tamoxifen 20 mg for 5 years. But after three months during routine follow-up clinician felt a new lump in left breast which was of size 2x2 cm and on fine-needle aspiration cytology showed presence of malignant cells. Patient was managed with modified radical mastectomy with axillary lymph node resection. Histopathologically, tumor was infiltrating duct carcinoma, grade II and 2 of 6 lymph nodes showed presence of malignant cells. Hence, it was stage II carcinoma of breast. Then patient underwent orchiectomy. Again patient received adjuvant chemotherapy and radiotherapy for five weeks and continued with oral tamoxifen 20 mg daily. Now since 18 months patient is disease free.

**DISCUSSION**

Bilateral breast cancer in male is an infrequent condition representing 0.5–0.6% of all breast cancers. Recent studies have shown an increased incidence of male breast cancer from 0.86 to 1.08 per 10,000 men [2]. In India Mitra et al. reported incidence of male breast cancer in a tertiary care hospital 0.6% of all cancers in men and 2.5% of all cases of breast cancer [3]. Risk factors for male breast cancer include family history of cancer of breast, obesity, gynecomastia, exposure to radiation, testicular diseases like undescended testis, orchitis, orchietomy, Klinefelter syndrome (47,XXY) and hormonal therapy [1]. Also hyperestrogenic states due to liver diseases and hypoandrogenic conditions like infertility contribute to breast cancer in men. Gene mutations of BRCA2, BRCA1 and androgen receptor gene predispose to such condition [4, 5]. Our patient was not willing for such genetic studies. No significant risk factors were identified in the present case. Hence this unusual synchronous bilateral breast cancer presentation may be a sporadic event.

Invasive duct carcinoma is the most common type of malignancy of breast observed in male with strong positivity for estrogen and progesterone receptors in 63–90% of cases [6]. In our case also patient had this common type of breast cancer, but within a period of three months he developed same type of tumor on contra lateral side. We managed the case with bilateral modified radical mastectomy, radiotherapy, adjuvant chemotherapy and hormonal therapy with tamoxifen. For hormone receptor positive breast tumor tamoxifen is the first choice of therapy that can lead to improved overall survival and outcome of the patient. Management in male and female breast cancer to some extent is similar.

Because of rarity of bilateral breast cancer in male there is dearth of such reports in literature. Giordano et al. reported only two cases of bilateral breast cancer among 2537 men with breast cancer [4]. As in males no screening strategies are designed usually they are presented in late stage. Kahla et al. reported a case of hormone receptor positive bilateral synchronous breast cancer in a 71-year-old male with 17 months disease free survival managed with bilateral modified radical mastectomy (MRM) and adjuvant chemo-endocrine treatment [7]. Lambley et al. described a 84-year-old male with bilateral infiltrating duct carcinoma managed by bilateral MRM and tamoxifen but not radiotherapy [8]. Small sample of the cases are available for study because of rarity. Shah et al. reported only 32 cases of male breast cancer over 24 years in their retrospective study [9]. Rai et al. retrospectively analyzed 30 cases of male breast cancer treated at a single center in India over 24 years [10]. They observed occurrence of loco regional recurrence in three cases and distant metastasis in nine patients. But there was no evidence of bilateral breast cancer in a single case. Some studies have shown a worse outcome for male breast cancer compared to female breast cancer, but findings are inconsistent. In our patient, there was a latent period of four months between two incidences of breast cancer.

Woo-Young Sun et al. reported a case of a 54-year-old male with simultaneous bilateral breast cancer in Korea with invasive duct carcinoma in right breast and ductal carcinoma in situ in left breast. Out of 24 cases of synchronous bilateral male breast cancer reported in literature hormonal receptor status (ER or PR) was positive in 87.5% cases [11].

Proper clinical evaluation, mammography and fine-needle aspiration cytology play important role in the diagnosis of male breast cancer. Presence of lump,
nipple retraction and discharge, ulceration and palpable axillary or supraclavicular lymph nodes should lead to a suspicion of malignancy of breast in male [12]. Agrawal et al. emphasized the important psychological aspect of male having ‘cancer of female’ which should be managed by clinician and counselor [13]. In male risk of developing second breast cancer after first breast cancer is much high compared to women [14]. Men with breast cancer have 93-fold greater risk of developing contra lateral breast cancer [1]. If investigated in depth and treated, early patients will definitely benefit with optimum outcome.

Purpose of reporting this case from India was to study and compare clinicopathological profile and management of male breast cancer cases reported in literature. This case report also adds to collection of resources for study of male breast cancer because of synchronous presentation of bilateral breast cancer with latent period of four months. Also this will increase awareness of patients and clinicians about this rare condition. Recently, Jonathan White et al. reported rising number of male breast cancer patients from all countries. But there is limited epidemiological and clinical data related to this disease. Most of the information used in understanding and managing male breast cancer comes from data of female breast cancer which may not always provide an accurate picture [15].

CONCLUSION

Although rare, incidence of male breast cancer is increasing and survivors are at risk of development of subsequent primary cancers. Despite absence of major risk factor our patient developed synchronous bilateral breast cancer, but was managed successfully with bilateral modified radical mastectomy, adjuvant chemotherapy, hormonal therapy and radiotherapy. Future in-depth studies focusing on etiopathology of the disease will help to optimize the management of male breast cancer patients.

*********

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

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Guarantor

The corresponding author is the guarantor of submission.

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REFERENCES


Isolated ileal heterotopic pancreas in a child: A clinically undetected cause of ileoileal intussusception

Sarika Verma, Pankaj Bansal, Vivek Manchanda, Ruchika Gupta

ABSTRACT

Introduction: Pancreatic heterotopia has been described most frequently in stomach and duodenum. Isolated occurrence of heterotopic pancreas in ileum is exceptional and is usually discovered incidentally during surgery. In rare cases, heterotopic pancreas has been reported as a lead point of ileoileal intussusception. Case Report: A one-year-old boy was presented with recent onset of constipation and abdominal distention. Radiologic investigation suggested a diagnosis of midgut volvulus with ileoileal intussusception. Resection anastomosis of the intussusception was performed. Histologic examination of the resected specimen showed heterotopic pancreas in the wall of intussusceptum at its tip. No other significant pathology was found. Conclusion: Ileal heterotopic pancreas is a rare cause of intussusception at any age and is usually not evident clinically. Hence, careful examination of the lead point of intussusceptum in resection specimens is mandatory to delineate the underlying etiology of these cases.

Keywords: Heterotopia, Intussusception, Ileoileal

INTRODUCTION

Intussusception is defined as the telescoping of a segment of the gastrointestinal tract into subjacent one. It is the most common cause of intestinal obstruction in children and is often idiopathic [1]. Intussusception with intraluminal heterotopic pancreatic tissue as the lead point is a rare condition [2]. It usually presents in stomach, duodenum and proximal jejunum. Occurrence more distally in the distal jejunum and ileum is exceptionally rare and usually asymptomatic [1, 3]. Intussusception due to heterotopic pancreas has been described in a few cases, including pediatric as well as adult patients [1, 2]. On an extensive review of literature, we found less than 25 cases of isolated heterotopic pancreas of ileum as leading point of intussusception in children. We describe the clinicopathologic features of an infant with ileoileal intussusception due to heterotopic pancreas in ileum. The existing literature is briefly reviewed as well.

CASE REPORT

A one-year-old boy was admitted to our hospital with complaints of fever, constipation, vomiting and distension of abdomen since two days. Physical examination showed marked abdominal distension with venous prominence.
A provisional diagnosis of intestinal obstruction was made. Radiologic investigations showed multiple air fluid levels with features of midgut volvulus and ileoileal intussusception. The child underwent an exploratory laparotomy. Intra-operatively, midgut volvulus with torsion of mesentery and ileoileal intussusception 30 cm proximal to ileocaecal junction was found. The intussusceptum appeared gangrenous. There was no free fluid in the abdomen. Surgical correction of the volvulus with Ladd’s procedure and resection of ileal intussusception was done with end-to-end anastomosis. The resected specimen was submitted for histopathology.

We received a segment of ileum measuring 8.5 cm in length. The segment was gangrenous and showed an intussusception 6 cm in length. Intussusceptum was 5 cm long with a blind end and focal mucosal ulceration. Intussusciens was 7.5 cm in length and showed normal mucosal folds. A congested nodule measuring 0.6x0.6 cm was identified in the lumen of intussusceptum near the tip of the blind end. Multiple sections from the nodule as well as intestinal wall were processed.

Microscopically, the nodule showed a well-circumscribed polyoid tissue with ulcerated mucosa. In the submucosa, pancreatic acini were identified embedded within the gangrenous wall of the nodule (Figure 1). No islet cells were identified. The histological impression was that of heterotopic pancreatic tissue, type 2 in the submucosa of ileum. There was no other significant pathological change such as inflammation or neoplastic transformation in the pancreatic tissue.

Histologically, heterotopic pancreas has been classified into three types by von Heinrich as: Type 1 (all elements of the normal pancreatic tissue are identified), Type 2 (pancreatic tissue with absence of islet cells) and Type 3 (pancreatic ducts only) [14]. Our patient had type 2 heterotopic pancreatic tissue. Although most cases with heterotopic pancreas are asymptomatic, various pathological changes including pancreatitis, pseudo pancreatic cyst, adenoma and carcinoma have been reported in foci of heterotopic pancreas [15]. None of these changes were identified in our case.

On an extensive review of the existing English literature, we found less than 25 cases of isolated heterotopic pancreas of ileum as leading point for intussusception in pediatric age group [2, 4, 6]. The relative frequency of a pathological lesion causing intussusception increases with age. However, the highest frequency of detection of pathologic lesion in cases of intussusception is still the first year of life [6], as in the present case. In a review of 10 children with ileoileal intussusception due to heterotopic pancreas, seven patients were less than two years of age and of these, four were infants [6].

Most cases of intussusception caused by a pathologic lead point are irreducible by barium enema and require...
manual reduction [6]. Hence, it is imperative to palpate for lesions like heterotopic pancreas within the wall of ileum after manual reduction [6]. This is especially essential in patients with submucosal location of heterotopic pancreas since the lesion may not be obvious on visual inspection. Simple excision is considered the treatment of choice for heterotopic pancreas leading to intussusception. This procedure avoids recurrence of intussusception as well as the risk of sequelae of pancreatitis etc [6].

CONCLUSION

In conclusion, isolated pancreatic heterotopia in ileum is rare and intussusception due to such occurrence is an even rarer phenomenon. Pediatric intussusception due to heterotopic pancreas has been described in very few cases. Hence the present case reiterates the need for examination of the tip of intussusceptum or the part of intestine that telescopes for diagnosis of submucosal lesions like heterotopic pancreas as the lead point of intussusception in resection specimens.

**********

Author Contributions
Sarika Verma – Acquisition of data, Drafting the article, Final approval of the version to be published
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REFERENCES
Disseminated *Rhodococcus rhodochrous* infection in an immunocompromised patient

Setu Patolia, Eneh Kennedy, Zahir Mehjabin, Neerja Gulati, Swati Patlia, Dharani Narendra, Rakesh Vadde, Saurav Pokharel, Frances Schmidt, Joseph Quist, Danilo Enriquez

**ABSTRACT**

Introduction: Genus *Rhodococcus* is a rare cause of infection in human. *Rhodococcus equi* has been reported as a cause of majority of these infections. However, *Rhodococcus rhodochrous* has never been reported as an etiologic agent in human diseases. Case report: A 45-year-old female was admitted with cough with yellowish sputum production, fever, chills and shortness of breath for three days. Patient had significantly decline in her functional capacity. Over past three months, patients had recurrent admissions for pneumonia and developed increasing numbers of skin nodules. Blood cultures sent from previous admissions were reported as *Corynebacterium* species. Lung and skin biopsy showed *Rhodococcus rhodochrous* species confirmed by high performance liquid chromatography. Later in the course of disease, patient developed brain abscesses. Conclusion: *Corynebacterium* species in blood should be carefully reviewed in an immunocompromised patient and *Rhodococcus rhodochrous* species infection should be considered as one of the differential diagnosis.

**Keywords:** *Rhodococcus rhodochrous*, Immunocompromised, Disseminated infection, *Corynebacterium, Rhodococcus equi*

**INTRODUCTION**

*Rhodococcus* species is a rare cause of infection in humans. Most of the reported human infections are due to be *Rhodococcus equi* [1–3]. *Rhodococcus rhodochrous* is rarely reported as an etiology in humans [4]. *Rhodococcus* species usually causes disease in immunocompromised individuals [5, 6]. *Rhodococcus equi* infection in an immunocompetent persons have been reported, though such reports are very few [7–9]. We present a case of a 45-year-old HIV patient who had recurrent pneumonia for three months. Blood cultures reports of *Corynebacterium* species were considered as contaminated samples. However, lung and skin biopsy specimen showed *Rhodococcus rhodochrous* species infection.

**CASE SERIES**

A 45-year-old HIV positive female (CD4 cell count- 15 cells/mm³) was brought to the emergency department with complaints of fever, cough, and shortness of breath
of three days duration. Cough was productive of yellowish sputum with no associated hemoptysis or chest pain. Fever was intermittent with chills but no night sweats. Patient’s functional capacity decreased from a prior walking distance of half a block. Review of symptoms was significant for multiple painful skin bumps which were increasing in number. Her past medical history revealed multiple admissions in the last three months, two of which were for pneumonia. Patient was not on antiretroviral therapy and signed out against medical advice from hospital each time she felt better. She was a chronic smoker (1–1.5 packs per day for 25 years) and active cocaine abuser. She denied any history of occupational exposure and never had pets.

On examination, patient was chronically ill looking with low grade fever of 100.4 °F, tachypnea (RR 21/min), tachycardia (HR 128/min). Oxygen saturation was 97% on room air. Other significant findings on examination were poor oral hygiene, oral thrush and right middle lung and basal crackles. Multiple subcutaneous nodules sparsely distributed on the thigh, face, abdomen and upper arm were noted (Figure 1). The nodules were tender, of varying sizes (5–10 mm), with some erythema but non blanching.

Pertinent laboratory findings were as follows: white blood cell count of 14.5 with neutrophil count of 91.7. Basic metabolic panel was essentially normal. Liver profile showed hypoalbuminemia (1.2 g/dL), mildly elevated alkaline phosphatase (193 IU/L) and lactate dehydrogenase (231 IU/L). Coagulation profile was normal and blood culture was negative. X-ray revealed right middle lobe infiltrate (Figure 2). Computed tomography (CT) scan revealed nodule with cavitation in right middle and lower lobe (Figure 3). Echocardiography showed normal ejection fraction with moderate pericardial effusion but no evidence of vegetation.

Sputum acid fast bacilli test came negative five times; transbrachial biopsy was negative for malignancy, acid fast bacilli stain, and fungal smear. Blood cultures drawn during previous admissions were reported to positive for Corynebacterium spp. on three different occasions. These reports were considered as contamination during past admissions. Culture drug susceptibility reports were requested for previous blood cultures because of patient’s persistent and new symptoms. Patient also agreed for surgical biopsy and specimens were taken from the skin of right buttock, right upper thigh and lung (right lower and upper lobes). Both specimens showed the same findings: acute suppurative and chronic inflammation and fibrohistiocytic granulomatous proliferation with

Figure 1: Subcutaneous nodule on the thigh.

Figure 2: Right middle lobe reticulonodular infiltrate (X-ray of chest).

Figure 3: Nodular infiltrate in right middle lobe with central cavity formation (computed tomography scan of the chest).
no evidence of malignancy. Gomori methenamine silver stain and Gram stains show long filamentous branching, beaded Gram-positive organisms distributed in large aggregates (Figure 4). Morphology was said to be highly suggestive of Nocardia-type organisms. Same specimen was sent out to city department of health where the organism was identified as Genus Rhodococcus and species Rhodochrous by high performance liquid chromatography.

Patient again signed out against medical advice. Patient was given prescriptions for clarithromycin and bactrim, along with antiretroviral therapy. However, the patient was very noncompliant with the treatment. Four weeks later, she presented with headache, vomiting, fever. Computed tomography scan of the brain showed hypodense areas in the left parietal lobe. Magnetic resonance imaging scan with contrast showed multiple ring enhancing lesions scattered throughout both cerebral hemispheres (Figure 5).

DISCUSSION

The genus name Rhodococcus, first used by Zopf in 1891 [10], was revived and redefined in 1977 to accommodate the ‘rhodochrous’ complex which comprised a number of strains that resembled but did not belong to the established genera of Nocardia, Corynebacterium and Mycobacterium [11]. Rhodococci are described as aerobic, Gram positive, non-motile, mycolate-containing, nocardioform actinomycetes [12]. The term ‘nocardioid’ is morphologically descriptive and refers to mycelial growth with fragmentation into rod-shaped or coccoid elements [13]. This morphological similarity may pose a problem in the preliminary differentiation and identification of the organism.

Various species of genus Rhodococcus have been recovered from a variety of sources [14, 15]. Most of the species are saprophytes [16], but occasionally Rhodococcus species have been isolated from humans. In some cases, Rhodococcus has been linked to human infection [17, 18].

It has been suggested that other Rhodococcus species may be of more importance in human disease than previously thought. Osoagbaka described the isolation of a number of Rhodococcus and related bacteria from the sputum of patients with respiratory illnesses [19]. Schaal and Lee also reported the isolation of various rhodococcal species from clinical samples [20]. The identification of other Rhodococcus species in clinical samples is more problematic and it is possible that some cases of non Rhodococcal equi species infection go unrecognized. Rhodococcus rhodochrous was isolated in two patients with pneumonia during autopsy in lung and blood [21]. Rhodococcus rhodochrous species has been isolated from a chronic corneal ulcer [4]. Ventricular peritoneal shunt infection by Rhodococcus rhodochrous was described in a five-month old infant [14]. Most of the cases of Rhodococcus rhodochrous infection described in literature occurred in immunocompromised or debilitated patients [21, 22]. Treatment protocol for the infections caused by Rhodococcus genus is not established. Different studies have noted response to ampicillin, methicillin and intraventricular cephalothin [23]. Our patient was treated based on the available report of the drug susceptibility.
CONCLUSION

*Rhodococcus rhodochrous* is a rare cause of human infection. When cultures are reported as *Corynebacterium* species, *Rhodococcus rhodochrous* should be considered as a differential diagnoses. Primary manifestations of this infection range from pneumonia to skin and brain abscessed. There is no consensus on treatment, but beta lactams and tetracycline derivatives appear to be an effective treatment.

*********

**Author Contributions**

Setu Patolia – 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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**Guarantor**

The corresponding author is the guarantor of submission.

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Authors declare no conflict of interest.

**REFERENCES**

Refractory bilateral scrotal ulcers with numerous IgG4-positive plasma cells: Another skin location for IgG4-related sclerosing disease

Jaclyn L Jerz, Jae Y Ro, Alberto G Ayala

ABSTRACT

Introduction: Immunoglobulin G4-related sclerosing disease (IgG4-RSD) is an emerging diagnosis characterized by mass-forming inflammation which responds to corticosteroids. Cases have been reported in virtually every organ, however, skin cases are rare and tend to involve the head and neck region. Case Report: An 85-year-old male presented with painful, bilateral, non-healing scrotal ulcers refractory to treatment with antibiotics and creams. On examination, he was found to have multiple large, purulent, stage three scrotal ulcers. He was admitted for administration of broad-spectrum antibiotics, but the cultures of the ulcer discharge grew normal, non-pathogenic skin flora. Dermatology was consulted for evaluation of other non-infectious etiologies for the ulcers’ failure to heal. A shave biopsy showed 50% IgG4-positive plasma cells, and light-chain in situ hybridization studies demonstrated polyclonality. The patient’s serum IgG4 was elevated to 115 mg/dL (reference range 7–89 mg/dL), and he had an unclear remote history of Whipple surgery. The patient was discharged home with topical clobetasol (a synthetic corticosteroid) and required no follow-up for the ulcers. Conclusion: IgG4-RSD includes a variety of disease entities and may manifest as diffuse or solitary inflammatory mass lesions. Older males are affected most often overall. The diagnosis is made on histopathology with >40% IgG4-positive plasma cells, fibrosclerosis, and obliteration of phlebitis, regardless of organ. Elevated serum levels of IgG4 are markers of disease, but not required for diagnosis. Given the patient’s histologic findings and laboratory results, this case is likely the first report of IgG4-RSD in scrotal skin.

Keywords: IgG4, Sclerosing disease, Pseudolymphoma, Corticosteroids

INTRODUCTION

Immunoglobulin G4-related sclerosing disease (IgG4-RSD) is a systemic autoimmune inflammatory process. The characteristic tumorous lesions may be solitary or diffuse. The diagnosis is made based on the morphologic findings. The major histologic features are a dense

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lymphoplasmacytic infiltrate, fibrosclerosis (storiform pattern), and obliterator phlebitis; the first two features are seen more frequently. An increased number of IgG4-positive plasma cells is also required, however, the reported numeric cutoffs vary by organ and degree of fibrosis [1].

IgG4-RSD has been reported in virtually every organ, but skin lesions are rare. Since a scrotal presentation has not been reported, our purpose is to describe the clinicopathologic aspects of this case.

CASE REPORT

An 85-year-old male was presented to the emergency department from his nursing home with a two-month history of bilateral, painful scrotal ulcers. His wife reported mild mental status changes during this time, but there were no other associated localized or systemic symptoms. He had been treating the ulcers at home with an unknown antibiotic and topical ointment.

The patient had multiple medical problems including hypertension, diabetes, peripheral vascular disease, and prior cerebrovascular accidents. He had undergone a below-knee amputation for gangrene and recent hip surgery after a fall, which rendered him bedridden. He was incontinent of urine and wore a diaper, which was often wet. He was taking lisinopril 40 mg, aceterminaphen 325 mg, furosemide 80 mg, naproxen 375 mg, amlodipine 10 mg, citalopram 40 mg, aspirin 81 mg, and potassium-chloride 8 mg daily, metoprolol 50 mg twice daily, and pancrelipase three times daily with meals. The patient was also taking doxycycline 100 mg daily for chronic non-healing, left hip surgical wound. He had no known drug allergies. He was a former smoker who had quit in 1942.

The patient’s initial urine culture grew low-titers of Proteus mirabilis and Enterococcus. The culture of the drainage from the patient’s scrotal ulcers grew Proteus mirabilis, Enterococcus, and multi-drug resistant. Acinetobacter baumannii or Acinetobacter calcoaceticus complex management is discussed below. The patient’s initial broad-spectrum antibiotics regimen was tapered, and many of the remaining medicines were switched to of levofloxacin in the emergency department for his presumed urinary tract infection, and he was admitted for more aggressive treatment of his ulcers and further work-up.

The patient began receiving broad-spectrum intravenous antibiotics (vancomycin and cefepime), and the wound care specialty team was consulted for assistance. Dermatology was consulted on hospital day 1 for evaluation of other possible etiologies for the non-healing scrotal ulcers. The dermatologist’s differential diagnosis included infectious (bacterial, viral, fungal, mycobacterial, and sexually-transmitted), malignant (squamous cell carcinoma), and systemic-disease related (pyoderma gangrenosum) causes, however, she felt that the most likely explanation was ongoing skin irritation and maceration from soiled diapers. She sent ulcer tissue for culture (negative), ordered tests for sexually transmitted infections (negative), and performed a shave biopsy to rule-out malignancy (Figure 1). Formalin-fixed, paraffin-embedded tissue sections stained with hematoxylin and eosin were examined and showed full-thickness loss of epidermis, with dermal fibrosis and a band-like inflammatory infiltrate. The inflammation was composed almost entirely of plasma cells (Figure 2). Special periodic acid-Schiff (PAS) staining was negative for micro-organisms. Immunohistochemistry showed numerous IgG4-positive plasma cells (approximately 50% of total plasma cells seen) (Figure 3). In situ hybridization studies for kappa and lambda light-chains demonstrated a polyclonal phenotype.

The patient’s initial urine culture grew low-titers of Proteus mirabilis and Enterococcus. The culture of the drainage from the patient’s scrotal ulcers grew Proteus mirabilis, Enterococcus, and multi-drug resistant. Acinetobacter baumannii or Acinetobacter calcoaceticus complex management is discussed below. The patient’s initial broad-spectrum antibiotics regimen was tapered, and many of the remaining medicines were switched to

![Figure 1: Biopsy of a scrotal skin ulcer with underlying, band-like chronic inflammation (H&E stain, x400).]
oral equivalents. An infectious disease (ID) consultation was requested for assistance with managing the patient’s multiple infections with multiple multidrug-resistant organisms. The specialist saw the patient on hospital day 6. He noted that the Proteus and Acinetobacter in the scrotal ulcers was normal, non-pathogenic skin flora and did not require antibiotics. Based on his experience with similar wounds, the ulcers would require topical wound care and significant time. The consultant also felt that the sample sent for urine culture which grew Proteus and Enterococcus was contaminated. It had been a voided specimen, and there was only a low-titer of organism growth. The ID consultant recommended discontinuation of all antibiotics, except doxycycline for the patient’s chronic MRSA.

The patient’s scrotal ulcers remained stable after the antibiotics were discontinued. Around the time of discharge, the pathologist interpreting the skin biopsy discussed the possibility of IgG4-RSD with the dermatologist, and serum total IgG and IgG1-4 subclass levels were ordered; both were elevated. The total IgG was 1530 mg/dL (reference range 696–1488 mg/dL) and IgG4 was 115 mg/dL (reference range 7–89 mg/dL). The remaining three IgG subclass levels were within normal limits. Based on the combined results, the dermatologist recommended two-weeks of 0.05% Temovate® (clobetasol propionate) ointment, a synthetic corticosteroid with high glucocorticoid activity and some mineralocorticoid activity.

On hospital day 8, the ulcers had no drainage, and were subjectively less painful. The patient was able to be discharged to a skilled nursing facility with wound care instructions and Temovate®. He was told to follow-up with dermatology if the ulcers had not healed within two weeks, and has never made a follow-up appointment.

**DISCUSSION**

The patient’s histologic and immunohistochemical findings, combined with his elevated serum total IgG and IgG4 levels, support a diagnosis of IgG4-related sclerosing disease. To our knowledge, this case is the first report of IgG4-RSD in scrotal skin.

IgG4-RSD was first described by Hamano et al. in 2001, and was initially recognized as a cause of autoimmune pancreatitis in the presence of elevated levels of serum IgG4 [2]. The characteristic lesions of IgG4-RSD encompass a wide range of diagnoses: chronic sclerosing sialadenitis [3], Mikulicz’s disease in the lacrimal gland [4], retroperitoneal fibrosis [5], and tubulointerstitial nephritis [6]. There are far fewer reports of IgG4-RSD in skin, and cases tend to involve the head and neck region [7]. The skin lesions present clinically as plaques or nodules and can be seen in isolation or in the context of systemic disease [8]. Differentiating IgG4-RSD of the skin from other causes of pseudolymphoma-type lesions, such as Rosai Dorfman and angiolymphoid hyperplasia with eosinophilia, is important for treatment reasons, as IgG4-RSD responds dramatically to corticosteroids [1].

The diagnosis of IgG4-RSD requires a dense, IgG4-positive lymphoplasmacytic infiltrate on histology. The inflammation is often accompanied by storiform fibrosis and obliterator phlebitis. The number of IgG4-positive plasma cells specific for IgG4-RSD has been studied for type 1 autoimmune pancreatitis, and ranges from >10/high-power field (HPF) on biopsies to >50/HPF in resection specimens. However, cutoffs are thought to depend on the organ and degree of fibrosis. A more powerful quantitative measure is the IgG4+/ IgG+ plasma cell ratio; >40% has been proposed as a comprehensive cutoff value for any organ. Serum IgG4 levels are often,
but not always, elevated, and abnormal serology is not required for the diagnosis. If the histology shows possible, but not definitive, IgG4-RSD, then additional clinical, serologic, or radiologic evidence is needed to confirm the diagnosis. A serum level of IgG4 >135 mg/dL supports the diagnosis of IgG4-RSD [1].

CONCLUSION

This case represents the first report of IgG4-RSD in scrotal skin, and it is likely that the ulcers are a localized set of IgG4-positive lesions, without underlying systemic disease. Although the reason for the patient’s remote Whipple procedure is not clear, the surgery does not seem to have been performed for autoimmune pancreatitis. The patient’s left hip surgical wound is known to be chronically infected with methicillin-resistant Staphylococcus aureus (MRSA), and his sacral decubitus ulcers are unlikely to be IgG4-positive, given his bedridden condition.

Author Contributions

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

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

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

Guarantor

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Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

Non-traumatic vertebral fractures: An uncommon complication following the first episode of a convulsive seizure

Nalli Ramanathan Uvaraj, Nalli Ramanathan Gopinath, Aju Bosco

ABSTRACT

Introduction: Non-traumatic vertebral fractures that occur solely as a consequence of the muscle forces that develop during a convulsive seizure, has rarely been reported in orthopedic literature. Case Report: Therein, we present a case of non-traumatic vertebral compression fractures in an 18-year-old male, who presented with severe back pain following a convulsive seizure, which occurred while he was sitting in his bed. He had no other reported trauma and no previous history of seizure. A detailed neurological work-up revealed no organic cause for the seizure. His bone mineral density measurements, hormonal and metabolic profiles were normal. Conclusion: Forceful muscle contractions that develop during a single episode of convulsive seizure, occurring for the first time, can result in vertebral compression fractures, even in a normal healthy individual. Vertebral fractures occurring in a healthy young (non-epileptic) male, with normal bone mineral density, presenting with back pain after a convulsive seizure is a rare clinical presentation. These vertebral fractures can appear clinically asymptomatic and can easily be overseen, especially in the absence of overt signs of external trauma and possible postictal consciousness disturbance, which may fail to provide clue to early diagnosis. A high index of clinical suspicion is needed in patients presenting with back pain after a tonic-clonic seizure even in the absence of a fall or a significant trauma. Such patients may be subjected to a systematic musculoskeletal examination and a thorough radiological evaluation to rule out potential bony injuries.

Keywords: Seizure, Epilepsy, Non-traumatic, Vertebral fracture

How to cite this article


INTRODUCTION

Vertebral fractures occur in patients with convulsive seizures. It has been reported in literature that the incidence of vertebral fractures associated with convulsive seizures varies from 0.95–16% [1, 2]. These fractures usually result from trauma due to a fall or accident occurring during the seizure. These patients have an associated osteopenia induced by long-term anticonvulsive medications or other bone mineral density decreasing agents. However, non-traumatic
vertebral fractures that occur from violent contraction of the paraspinal muscles during a convulsive seizure is very rare (0.3%) [3]. Non-traumatic vertebral fractures that occur solely as a consequence of the muscle forces that develop during a convulsive seizure, has rarely been reported in orthopedic literature.

We present a case of non-traumatic vertebral compression fracture in a healthy young male, with normal bone mineral density. The patient is not a known epileptic and had experienced the first episode of a convulsive seizure.

CASE REPORT

An 18-year-old male presented to the medical department with a history of fever with chills and rigor and loss of appetite for one week. In the night after admission, he developed an episode of generalized tonic-clonic seizure for the first-time while sitting on his couch, during the early morning hours. His mother was a witness to the seizure. There was no history of overt trauma as reported by his mother. The patient had no history of previous seizure episodes and a detailed history in this regard also did not throw light as to the cause of the seizure. Next morning, the patient complained of mild back pain and chest pain which were ignored due to the absence of any signs of significant external trauma. His neurologic examination was unremarkable. As the back pain persisted on the third day, the patient was referred to the orthopedic department for further evaluation. A thorough musculoskeletal examination revealed a paraspinal muscle spasm and tenderness in the middorsal region with no obvious deformity. Plain radiographs showed loss of height of D5 and D6 vertebrae (Figure 1A–B). Magnetic resonance imaging (MRI) scan confirmed compression fractures of D5, D6, D7 and D8 vertebrae (Figure 2). His hormonal and metabolic profiles and bone mineral density were normal. A detailed neurological work-up (computed tomography scan of the brain and electroencephalogram) revealed no organic cause for the seizure. The patient was fitted with a thoracolumbar orthotic support. Pain at the fracture site subsided after two weeks when rehabilitation was started, following which the patient resumed normal activities. Radiographs taken at follow-up showed that his spine was stable with no further loss of vertebral body height and no progression of kyphosis (Figure 3A–B). Patient was kept on antiseizure medication and was followed-up periodically. On regular follow-up there had been no further episode of seizures for the last four years.

DISCUSSION

In 1907, Lehndorff was the first to suggest that strong muscle contractions during a convulsive seizure can cause vertebral compression fractures [4]. Violent contractions
of the paraspinal muscles and the muscles of the neck, abdomen and pelvis, occur during convulsive seizures. With increasing muscular tension of the posterior spinous and abdominal muscles, the spine flexes forward and is subjected to axial loading and flexion compressive forces, directed along the anterior and middle columns, which can cause vertebral compression or burst fractures [5, 6]. For biomechanical reasons, seizure induced non-traumatic vertebral fractures have a predilection for the mid-thoracic spine (T3–T8), as in this case [6, 7]. In contrast traumatic vertebral fractures affect the cervical, thoracolumbar or lumbosacral junction. Cervical fracture and lumbosacral dislocation have been documented on rare occasions [8]. Most vertebral fractures induced by seizure are inherently stable with no neurological deficit.

Vertebral fractures are most frequent in patients whose attacks occur during sleep and in patients with a history of convulsive status epilepticus [7]. There has been a previous case report of a vertebral compression fracture which was the only presenting feature of an unwitnessed nocturnal convulsive seizure [4]. But in this case report, the mother witnessed the tonic-clonic seizure which occurred during the early morning hours, which was the only and the most valuable clinical clue to the diagnosis. Most vertebral fractures (80.9%) resulted from recurrent convulsions, due to a repetitive additive mechanism in patients with a history of convulsive status epilepticus [5]. But this is a case of a non-epileptic who was presented with a vertebral compression fracture from the first episode of convulsive seizure.

There have been reports of non-traumatic vertebral fractures in convulsive seizures secondary to osteopenia induced by long-term anticonvulsive medications or other bone mineral density decreasing agents, diabetic hypoglycemia, electrolyte imbalance, hypocalcaemia, electroconvulsive therapy or trauma during the convulsive episode [1, 9–11]. Patients with increased muscle mass, anticonvulsant induced osteoporosis, a prolonged seizure or recurrent seizures are at increased risk for fractures [1, 7]. Our patient was not a known epileptic and was not on anticonvulsive medication. His bone mineral density was normal. This is a rare case of a vertebral compression fracture that occurred in a normal healthy individual during the first episode of a convulsive seizure. Hence, the fracture was the sole result of violent muscle contractions alone that occurred during tonic-clonic convulsion. Vertebral fractures occurring in a healthy young (non-epileptic) male, with normal bone mineral density, presenting with back pain after a convulsive seizure is a rare clinical presentation which could easily be missed unless there is a high index of clinical suspicion.

Vasconseles suggested a rate of 15% of primarily asymptomatic fractures caused by seizures [7]. This emphasizes the importance of a critical musculoskeletal examination and a radiographic assessment in patients presenting with back pain after a tonic-clonic seizure even if an event of fall or accident is neglected by the patient. In a young patient, absence of overt signs of trauma, postictal amnesia or an inadequate clinical history provided by the patient or family members may mislead the clinician into the evaluation of a spontaneous non-porotic spine fracture which is often equated with a metabolic disorder or metastasis [4]. This may lead to an unnecessary costly diagnostic work-up.

**CONCLUSION**

Forceful muscle contractions during a single episode of convulsive seizure occurring for the first time, can result in vertebral compression fracture, even in a normal healthy individual. These vertebral fractures can appear clinically asymptomatic and can easily be overseen due to absence of external trauma and also due to possible postictal consciousness disturbance, which may fail to provide clue to early diagnosis. A high degree of clinical suspicion is needed whenever an epileptic patient complains of back pain. Such patients may be subjected to a thorough radiological evaluation to rule out potential bony injury, especially in the presence of symptoms.

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**Author Contributions**

Nalli Ramanathan Uvaraj – 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

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

Aju Bosco – 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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Miliary tuberculosis in an immunocompetent male

Chidozie Charles Agu, Patolia Setu, Hiba Basheer

ABSTRACT

Introduction: Miliary tuberculosis (TB) is a disseminated form of tuberculosis which may involve the lungs and other organs. It is a rare but potentially lethal form of tuberculosis seen mostly in immunocompromised patients. It is, however, not commonly reported in immunocompetent hosts. Case Report: We present a case of a 67-year-old African-American male who presented with persistent non-productive cough for three weeks, diarrhea, fatigue and weight loss. There was no history of active TB contacts, HIV infection, or other predisposing factor for immunosuppression. Physical examination was also unremarkable. Chest X-ray and computed tomography scan revealed bilateral extensive small nodular infiltrates with negative serial acid-fast sputum smears. Acid-fast bacilli were detected in sputum culture at four weeks and were identified by DNA probe as Mycobacterium tuberculosis. Conclusion: Miliary tuberculosis is a potentially lethal form of tuberculosis that mostly affects immunosuppressed patients, although in rare occasions can also affect immunocompetent adults.

Keywords: Miliary tuberculosis, Mycobacterium tuberculosis, HIV/AIDS.

INTRODUCTION

Miliary tuberculosis (TB) is a rare form of TB infection that results from massive lympho-hematogenous dissemination of Mycobacterium tuberculosis bacilli. [1] It involves mostly the lungs but may also affect several other organs in the body. It is more likely to occur in immunocompromised patients due to their depressed cellular immunity and is rarely reported in immunocompetent hosts. In this report, we present a case of Miliary TB in an otherwise immunocompetent patient.

CASE REPORT

A 67-year-old African-American male with a history of hypertension for 10 years, atrial flutter on Coumadin for four years, localized prostate cancer status post radiation therapy in remission came to the emergency room for persistent non-productive cough for 2–3 weeks and watery diarrhea with colicky abdominal pain for about 10 days for which he was prescribed antibiotics and antitussives by his primary medical care provider with no relief. He also complained of fatigue, malaise, poor appetite, and significant weight loss (about 9.07 kg in 2–3 months). He denied fever, chills, night sweats, dyspnea, chest pain, nausea or vomiting. He also denied any history of smoking, alcohol or illicit drug use, recent travel, sick contacts or exposure to industrial dusts. He worked as a...
mailman for the postal service and traveled to Bahamas in 2010. His current medications were metoprolol and Coumadin.

On physical examination, the patient was alert and in no pain or distress. Vital signs were normal except mild tachycardia of 101/minute. Physical examination was unremarkable including respiratory system examination. Laboratory evaluation, normal range is given in parenthesis, revealed mild leukocytosis WBC count of 14,300/µL (4,500–11,000/µL), mild anemia with of hemoglobin of 12.3 g/dL (13.5–17.5 g/dL) and mean corpuscular volume (MCV) of 88.3 (80–100), Erythrocyte sedimentation rate (ESR) 29 (0–15), hypokalemia (serum potassium of 2.9 mmol/L (3.6–5.1 mmol/L), abnormal LFT’s (total bilirubin 0.9 mg/dL (0.3–1.2 mg/dL), aspartate aminotransferase (AST) 77 IU/L (15–41 IU/L), alanine aminotransferase (ALT) 89 IU/L (17–63 IU/L), alkaline phosphatase 103 IU/L (32–91 IU/L), mild elevations of serum amylase 150 U/L (28-100) and lipase levels 59 U/L (22–51 U/L). Coagulation profile was as follows: Prothrombin time (PT) 16.4 s (9.7–11.3 s), international normalized ratio (INR) 2.19 (1.1–1.7) and partial thromboplastin time (PTT) 26.5 s (24.7–34.4 s) as the patient was on warfarin for atrial flutter. Electrocadiography (EKG) showed atrial flutter with variable atrioventricular block at 94 bpm. Chest X-Ray showed bilateral extensive small nodular infiltrates. Computed tomography (CT) scan of the chest, abdomen, and pelvis showed innumerable very small nodules (2–3 mm) seen throughout both lungs, small bilateral pleural effusions, small pericardial effusion and multiple small low attenuation lesions scattered throughout the liver. There were no adrenal or pancreatic lesions or retroperitoneal lymphadenopathy seen. Abdominal sonogram was unremarkable. Differential diagnosis included metastatic lung cancer, hypersensitivity pneumonitis, pneumoconiosis, rheumatoid nodules, vasculitis and fungal pneumonia among others. Three sets of acid-fast bacilli (AFB) sputum smears were negative. Purified Protein Derivative (PPD) skin test, HIV Elisa, urine legionella antigen, hepatitis serology, basic autoimmune work-up (antinuclear, anti-Ds DNA, anti CCP-antibodies, rheumatoid factor), tumor markers (AFP, CEA, CA 19-9, PSA), blood, stool and urine cultures were all unremarkable. Patient was offered bronchoscopy for further diagnostic evaluation but he declined. The diagnostic possibility of miliary TB and possible anti-TB treatment was discussed with the patient owing to the classic radiologic findings despite negative serial AFB smears. He refused treatment at the time because he felt relatively well and had never had any contact with TB. He was then discharged to outpatient clinic for further evaluation.

Acid-fast bacilli were detected in sputum culture at 4 weeks and were identified by DNA probe as Mycobacterium tuberculosis. The patient was readmitted and repeat serial induced sputum acid-fast bacilli smears revealed 1+ AFB. He was started on antituberculous therapy with isoniazid (INH), rifampin, ethambutol, pyrazinamide and pyridoxine. No immediate drug side effects were observed and he was discharged after a week of treatment and negative serial sputum AFB smears. Susceptibility testing revealed pansensitivity to INH, rifampin, ethambutol and pyrazinamide. The patient completed the therapy with two months of INH, rifampin, ethambutol and pyrazinamide and four more months INH and rifampin. At follow-up, the patient was asymptomatic, liver function tests were normal and repeat chest X-rays were normal.
Miliary TB is defined as widespread millet-like (1–5 mm) seeding of *Mycobacterium bacilli* in the lung and possibly in other organs of the body, mostly liver, spleen, lymph nodes, pleura, pericardium, meninges and bone marrow [2]. It is a very rare form of tuberculosis and in literature reviews its frequency is estimated at 2.8% of all TB infections [3]. In the United States 11,182 incident cases of tuberculosis were reported in 2010. Of these, extrapulmonary TB accounted for approximately 22% of cases while miliary TB was reported in only 2.7% [4, 5].

Miliary TB usually occurs in the presence of immunocompromising conditions such as advanced age, cancer, organ transplantation, immunosuppressive and cytotoxic therapy (including biologic agents antitumor necrosis factor), malnutrition, alcoholism, corticosteroids, poorly controlled diabetes, silicosis, end-stage renal disease, and most importantly HIV/AIDS [6].

Among immunocompetent adults, miliary TB accounts for less than 2% of all TB cases and 20% of all extra-pulmonary TB cases in clinical studies [6]. In contrast, in patients with HIV/AIDS, miliary TB accounts for about 10% of all TB cases [3] and more than 50% of all extra-pulmonary TB cases [6]. Although miliary tuberculosis is rare in the immunocompetent population, it is important to recognize that certain genetic defects may predispose immunocompetent individuals to disseminated tuberculosis such as abnormalities in the production or metabolism of interferon-gamma and interleukin-12, which are essential for granuloma formation and protective immunity to *M. tuberculosis*. Unfortunately, quantitative or qualitative tests for these cytokines are not widely available in clinical practice [7].

The clinical presentation of miliary tuberculosis can be acute, subacute or chronic. Acute disease is rare and may occur in advanced HIV/AIDS or other immunocompromised states [6]. It is usually fulminant, including multiorgan system failure, septic shock and acute respiratory distress syndrome (ARDS) [8, 9]. Therefore, miliary tuberculosis should always be considered in patients with ARDS of unknown etiology especially if risk factors are present [10–12]. The subacute or chronic presentations of miliary TB are more common than acute disease and patients may present with failure to thrive, fever of unknown origin, night sweats or dysfunction of one or more organ systems.

The most common laboratory abnormalities include anemia, leukopenia, thrombocytopenia and lymphopenia. Other lab abnormalities may include...
elevated ESR and C-reactive protein, hyponatremia, hypercalcinemia and sterile pyuria [13]. Advanced age (> 60 years), lymphopenia, thrombocytopenia, pancytopenia, hypoalbuminemia, elevated transaminase levels and delayed treatment have been identified as independent predictors of mortality [9, 14].

The classic chest radiograph appearance is a faint, reticulonodular infiltrate distributed fairly uniformly throughout the lungs. This miliary pattern of infiltrates is seen in about 84% of cases [15]. Other chest radiograph abnormalities include pleural effusion, hilar/mediastinal adenopathy, interstitial or alveolar infiltrates, or cavities. Chest CT scan is a more sensitive test for evaluating miliary TB.

Acid-fast microscopy and culture of body fluids, tissue, or drainage from an infected focus establishes the diagnosis especially if organisms or caseating granulomas are seen. The cumulative diagnostic yield of different body fluids and tissues in the diagnosis of miliary TB has been reported as follows: Sputum (41%), Fiberoptic bronchoscopy (47%), urine (33%), cerebrospinal fluid (21%), lymph node biopsy (91%), liver biopsy (89%), and bone marrow aspirate/biopsy (67%) [6]. Fiberoptic bronchoscopy is usually warranted if acid-fast bacilli are not detected at multiple sites (sputum, gastric aspirate, pleural fluid, ascites, urine, etc.) and there is evidence of pulmonary involvement on chest radiography [9].

The tuberculin skin test (PPD) can be a supportive diagnostic tool if positive, but anergy is observed more frequently among patients with miliary TB (up to 68% of cases) than those with pulmonary or isolated extrapulmonary involvement. PPD conversion may often occur following treatment [15].

The approach to antimicrobial therapy for treatment of miliary TB is the same as for pulmonary TB. Early empirical treatment for possible but not yet definitive miliary TB increases the likelihood of survival and should never be withheld while test results are pending.

CONCLUSION

Miliary tuberculosis is a potentially lethal form of tuberculosis arising from hematogenous dissemination of *Mycobacterium tuberculosis* bacilli. It mostly presents in immunosuppressed patients but can also affect immunocompetent adults. Diagnosis of miliary tuberculosis is often difficult due to variable clinical presentations, poorly sensitive smears and diverse radiologic findings. Although positive chest radiographic findings or a positive tuberculin skin test may support the diagnosis, negative results. However, do not exclude extrapulmonary tuberculosis. A high index of clinical suspicion is needed and antmycobacterial therapy should be administered urgently to prevent an otherwise fatal outcome.

Author Contributions

Chidozie Charles Agu – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content and final approval of the version to be published

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

Hiba Basheer – Substantial contributions to conception and design, acquisition of data, Analysis and interpretation of data, drafting the article, Revising it critically for important intellectual content and final approval of the version to be published

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The corresponding author is the guarantor of submission.

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DRESS syndrome associated with allopurinol

Chidozie Charles Agu, Hiba Basheer

ABSTRACT

Introduction: DRESS syndrome (Drug Rash with Eosinophilia and Systemic Symptoms) is a severe drug reaction with an estimated mortality of up to 10% largely due to multi-organ dysfunction. Diagnosis is challenging because the extent of skin involvement does not always correlate with the extent of internal organ involvement and therefore early recognition of symptoms is vital to minimize morbidity and mortality. Management involves prompt cessation of the culprit drug, administration of corticosteroids and supportive treatment. Case report: We report a case of an 87-year-old female with medical history significant for stage five chronic kidney disease, recently started on allopurinol for gout presented with dizziness, skin rash and eosinophilia which rapidly progressed to shock state followed by altered mentation, worsening renal failure, severe metabolic acidosis, deranged liver enzymes and gastrointestinal bleed requiring blood transfusion. Conclusion: Allopurinol is commonly used in clinical practice for the treatment of symptomatic hyperuricemia and gout. It has been associated with DRESS syndrome especially when used indiscriminately.

Keywords: DRESS syndrome, Allopurinol, Multi-organ failure

INTRODUCTION

The drug rash with eosinophilia and systemic symptom (DRESS) is a severe adverse drug reaction characterized by fever, rash, lymphadenopathy, hematologic abnormalities (including eosinophilia and atypical lymphocytosis) and multi-organ dysfunction. Allopurinol, a commonly used drug for the treatment of gout and other complications of hyperuricemia is documented as one of the drugs associated with DRESS syndrome. In this report, we present a patient with recent exposure to allopurinol and multi-organ failure typical of DRESS syndrome.

CASE REPORT

An 87-year-old African-American female with past medical history of hypertension, gout, chronic kidney disease stage five, coronary artery disease, and ischemic stroke with no residual neurological deficit presented with dizziness of one day duration and skin rash (Figure 1) for 1 week. There was no history of allergy or sick contacts. The patient’s medications included aspirin, allopurinol 300 mg/day started about five weeks ago for gout treatment, colchicine, and metoprolol. Her family and social history were noncontributory. Vitals signs were as follows: temperature 96.9°F, pulse rate 96/min, respiratory rate 18/min, blood pressure 104/57 mmHg, SaO2 97%. Physical examination was remarkable for...
diffuse pruritic erythematous papular rash on trunk, bilateral 2+ pitting edema and morbid obesity. The rest of the physical examination including heart, lung, abdomen, nervous system, and rectal examinations were unremarkable. Laboratory examinations were remarkable for (normal range is given in brackets) white blood cell count $7.7 \times 10^3/\mu L$ ($4.5 \times 10^3–1.1 \times 10^4/\mu L$), 23.6% eosinophils (0–7.5%), hematocrit 33.7 (36–46%), platelet count of $106 \times 10^3/\mu L$ ($130 \times 10^3–400 \times 10^3/\mu L$), blood urea nitrogen (BUN) 74 mg/dL (8–20 mg/dL), creatinine 7.4 mg/dL (0.4–1.3 mg/dL), serum bicarbonate 18 mmol/L (22–32 mmol/L) and an anion gap of 8. Liver function tests and coagulation profile were normal at presentation. Urinalysis was also normal. Immunological test results were negative for anti-nuclear antibody, anti-neutrophil cytoplasmic antibodies (cANCA and pANCA). Rapid plasma reagin (RPR), hepatitis B surface antigen, and hepatitis C antibody were negative. Complement (C3) level was low at 37 mg/dL (79–152 mg/dL). X-ray of chest was normal and computed tomography (CT) scan of head revealed remote right cerebellar and right parietal infarcts along with ischemic white matter and intracranial atherosclerosis. Electrocardiography showed normal sinus rhythm at 92 bpm, echocardiogram showed normal left ventricular function with ejection fraction of 55–60%, aortic valve calcification. Carotid Doppler was negative.

Within 48 hours, the patient developed hypotension (89/59 mmHg) tachycardia (105 beats per minute) and tachypnea (26 breaths per minute), drug hypersensitivity was suspected, allopurinol was discontinued, corticosteroids administered (hydrocortisone 50 mg IV every 8 hr) and IV fluids (normal saline) given to stabilize blood pressure. The patient later developed confusion and unresponsiveness (repeat CT scan of head showed no new changes and lumbar puncture was unremarkable). There was worsening of pre-existing renal failure with severe metabolic acidosis (BUN 84 mg/dL (8–20 mg/dL), creatinine 9 mg/dL (0.4–1.3 mg/dL), arterial pH 7.06 (7.35–7.45) and serum bicarbonate 6.5 mmol/L) and hemodialysis was started. Other laboratory abnormalities were as follows: aspartate aminotransferase (AST) 391 IU/L (15–41 IU/L), alanine aminotransferase (ALT) 328 IU/L (17–63 IU/L), serum amylase 244 U/L (28–100 U/L), serum lipase 215 U/L (22–51 U/L) and serum lactate 33 mg/dL (4.5–14 mg/dL). She later developed coffee ground vomitus with a drop in hematocrit to 18.7% (36–46%). Two units of packed red cells were transfused and hematocrit stabilized at about 24–25%. Septic work-up was done and was negative.

During the following week, her mental status improved, vitals stabilized, liver enzymes, amylase/lipase, lactate levels normalized and eosinophilia resolved. The skin rash became hyperpigmented and desquamated with clear, fluid-filled vesicles also noticed in some areas. Skin biopsy showed interface dermatitis with separation of the dermis-epidermal junction (Figure 2A–B). She was later discharged and continued on intermittent hemodialysis for end-stage renal failure.
DISCUSSION

DRESS syndrome a type-IV delayed hypersensitivity reaction is a severe and potentially life-threatening drug induced reaction characterized by severe skin rash, fever, lymph node enlargement, hematologic derangements (including eosinophilia or mononucleosis like atypical lymphocytes) and internal organ involvement—most commonly the liver and, to a lesser extent, the kidneys, lungs, and brain [1].

It typically has long latency period (1–8 weeks) after initiating the culprit drug and symptoms may persist or even worsen after discontinuing the drug as seen in our patient [1, 2]. The rash manifests as a diffuse erythematous eruption (morbilliform rash) on the face, upper trunk, and upper extremities, usually accompanied by fever, facial and periorbital edema. The extent of skin involvement does not always correlate with the severity of internal organ dysfunction. Lesions may later become vesicular or exfoliated [2]. Desquamation/scaling may occur with healing.

It has an estimated incidence of 1 in 1000 to 1 in 10,000 drug exposures and mortality rate of about 10% due to visceral organ compromise especially liver failure [3, 4]. Infectious complications have also been described especially in patients treated with corticosteroids.

Aromatic anticonvulsants (phenytoin, phenobarbital, carbamazepine) and sulfonamides are the most common causes of DRESS syndrome but a number of other drugs including lamotrigine, allopurinol, non-steroidal anti-inflammatory drugs, captopril, calcium channel blockers, mexiletine, fluoxetine, dapson, terbinafine, metronidazole, minocycline and antiretroviral drugs have also been implicated. [4]

Allopurinol, a hypoxanthine analog has been widely used in clinical practice over decades for the treatment of hyperuricemia and gout [5]. It is usually well-tolerated with no adverse effects in most cases, but is by no means a benign drug. About 2% of the treated patients develop a skin rash, and some may experience severe drug hypersensitivity reaction [5]. A multinational study (EuroSCAR) revealed that allopurinol is the drug most commonly associated with Stevens-Johnson syndrome (SJS) or Toxic epidermal necrolysis (TEN) in Europe and Israel [6]. In a Korean study involving 38 patients with DRESS syndrome, allopurinol was found to be responsible for 5.3% of the cases [7].

The pathogenesis of DRESS syndrome is not fully known but different factors have been postulated including immunological factors, genetic factors (risk as high as 25% if first degree relative had the syndrome) and drug detoxification pathways [8]. In a prospective study involving 40 DRESS patients, Epstein-Barr virus, human herpesvirus 6 and 7 reactivations were found in 76% of the cases. The culprit drugs are able to trigger these viral reactivations that in turn induce a pathogenic antiviral CD8 immune response [9].

A possible mechanism may be allopurinol or oxyipurinol (major metabolite of allopurinol) hypersensitivity and immune complex formation resulting in vasculitis [10]. The accumulation of oxyipurinol especially with reduced renal clearance leads to a greater risk of developing DRESS syndrome. Multiple studies have shown that advanced age, underlying renal impairment, higher doses, and concomitant use of thiazide diuretics are potential risk factors for developing allopurinol-induced DRESS syndrome [11].

Published data from the French pharmacovigilance center showed that allopurinol was associated with more severe forms of DRESS syndrome and mortality rate much higher than cases due to other drugs [12].

Diagnosis is based on clinical and laboratory findings and although skin biopsy may support diagnosis, it is usually non-specific. It may show a lymphocytic infiltrate in the papillary layer of the dermis, which may also contain eosinophils (generally denser than in other drug reactions) [2]. The mainstay of therapy in DRESS syndrome involves the immediate cessation of the culprit drug. In cases where the culprit drug is not obvious, clinical judgment must be used to select which medication to discontinue. Patch testing or lymphocyte transformation tests used to detect delayed hypersensitivity may be helpful in identifying the drug [13].

Corticosteroids are currently being used with some success, but their role remains controversial due to lack of controlled clinical trials supporting their use [14].

At present, the only indications for allopurinol use are symptomatic hyperuricemia (i.e., gouty arthritis, urate nephropathy and nephrolithiasis) and prophylaxis of urate nephropathy during chemotherapy in neoplastic diseases. However, a Medline literature review of 101 reported cases of allopurinol hypersensitivity syndrome revealed that in 80% of cases allopurinol was administered primarily for asymptomatic hyperuricemia and most patients received excessive doses [14]. Therefore, it is imperative that allopurinol be administered only when indicated and in the appropriate dose.

The patient described in this case report is very old lady with multiple comorbidities started about five weeks prior to presentation for the treatment of gout and presented with rash, eosinophilia and multi-organ failure. Differential diagnoses included drug hypersensitivity reaction (DRESS syndrome), Churg–Strauss syndrome (CSS), idiopathic hypereosinophilic syndrome and eosinophilic leukemia [14]. Churg–Strauss syndrome was excluded as the patient had no history of asthma, allergic rhinitis, or sinus abnormalities and P-ANCA was negative. Idiopathic hypereosinophilic syndrome was ruled out as it is a diagnosis of exclusion, typically affects men aged 20–50 years, and requires persistence of eosinophilia for at least six months. Eosinophilic leukemia was also excluded with the absence of immature eosinophils in peripheral smear. Of all the medications taken by the patient, the only potential trigger documented.
in literature was allopurinol. This medication was immediately discontinued and IV corticosteroids started along with other supportive measures. The patient showed progressive improvement afterwards.

CONCLUSION

Allopurinol is a drug commonly used in medical practice but its indiscriminate use can lead to severe consequences. DRESS syndrome is a severe drug reaction that has been associated with allopurinol. Recent use of this drug coupled with the presence of skin rash, eosinophilia and multi-organ dysfunction should raise clinical suspicion of DRESS syndrome. Prompt cessation of the drug, use of corticosteroids and other supportive measures has shown to improve outcomes.

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Chidozie Charles Agu – Substantial contributions to conception and design, Acquisition of data, analysis and interpretation of data, drafting the article, Revising it critically for important intellectual content and final approval of the version to be published
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Authors declare no conflict of interest.

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CASE REPORT

Wilkie’s syndrome: A rare cause of vomiting and weight loss

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ABSTRACT

Introduction: Superior mesenteric artery (SMA) syndrome, also known as Wilkie’s syndrome, is extremely rare and is characterized by postprandial epigastric pain, nausea, vomiting and loss of appetite, with subsequent weight loss, which aggravates the condition of the patients. The syndrome is caused by compression of the third part of the duodenum in the angle between the aorta and the superior mesenteric artery. Herein, we presented a patient with Wilkie’s syndrome and discussed the diagnostic difficulties and surgical treatment options.

Case report: We report a case of a 13-year-old female who was diagnosed with SMA syndrome. There was no history of recent trauma, surgery, prolonged immobilisation or neurological illness. Her weight loss was gradual. The patient was initially diagnosed as anorexia nervosa due to her symptoms of vomiting and anorexia without any clinical cause, but later on, she underwent computed tomography scan and was diagnosed with SMA syndrome. Her SMA syndrome, resolved after successful nonoperative management based on accepted guidelines for nutritional therapy, thus avoiding the need for operation. One year follow-up was uneventful.

Conclusion: Superior mesenteric artery syndrome is a rare cause of intestinal obstruction and its first-line treatment is usually conservative with jejunal or parenteral nutrition for restoration of the aortomesenteric fatty tissue. If conservative management fails, surgical options, open or laparoscopic duodenojejunostomy or duodenal mobilization. Its recognition is important because early diagnosis of a partial obstruction may allow for medical rather than surgical intervention, as exemplified by our case.

Keywords: Superior mesenteric artery (SMA) syndrome, Wilkie’s syndrome, Weight loss, Vomiting, Enteral feeding

How to cite this article


INTRODUCTION

Superior mesenteric artery (SMA) syndrome which is also known as Wilkie’s syndrome, is a rare cause of upper gastrointestinal obstruction [1–3]. Superior mesenteric artery syndrome was first described in 1861 by von Rokitansky, who proposed that its cause was obstruction of the third part of the duodenum as a result of arteriomesenteric compression [4]. Later, Wilkie provided a more detailed clinical and pathophysiologic description in a series of 64 patients and suggested treatment approaches. This is usually associated with conditions that cause significant weight loss, such as anorexia nervosa, malabsorption, or hypercatabolic states such as burns, major surgery, severe injuries, or malignancies as fat loss causes direct compression of third part of duodenum with superior mesenteric
artery anteriorly [3]. Despite diagnostic confusion with intestinal dysmotility syndrome, conservative therapy with nutritional supplementation is the initial approach and duodenojejunostomy is favored, if non-surgical treatment fails.

**CASE REPORT**

A 13-year-old female who was initially diagnosed as anorexia nervosa due to her symptoms of vomiting, anorexia and weight loss for 12 months, admitted to our emergency service. There was no history of recent trauma, surgery, prolonged immobilisation or neurological illness. Her weight loss was gradual. Abdominal examination revealed a distended abdomen, mild epigastric tenderness, and hyperactive bowel sounds. There was no palpable organomegaly. Her hemoglobin was 10 g/dL, leucocyte count was $8.0 \times 10^3$/mm$^3$. Serum albumin was 2.5 g/dL and rest of the investigations were normal. Abdominal radiograph revealed a dilated stomach with a prominent air fluid level. Subsequent abdominal computed tomography (CT) scan revealed dilatation of stomach, first and second part of duodenum. Third part of duodenum was compressed between superior mesenteric artery (SMA) and superior mesenteric vein (SMV) anteriorly and aorta and vertebrae posteriorly. On prone position, the narrowing of fourth part opens up but dilatation of third part persists. These features were suggestive for SMA syndrome (Figures 1 and 2). The small bowel distal to the SMA was decompressed. Endoscopic examination of the upper gastrointestinal tract revealed mild esophagitis, dilated stomach and proximal duodenum, and narrowing of the third part of the duodenum due to a pulsating extrinsic compression. She was hospitalized and total parenteral nutrition was administered with Daily 950 kcal (35 kcal/kg/d) of olicinomel N7 (Baxter, Eczacıbası, Turkey). After 15 day of admission, she tolerated oral nutrition. She gained 4 kg weight before discharge and she has been following without any complication and complain for one year.

**DISCUSSION**

Superior mesenteric artery syndrome is an uncommon but well recognized clinical entity characterized by compression of the third, or transverse, portion of the duodenum against the aorta by the SMA, resulting in chronic, intermittent, or acute, complete or partial duodenal obstruction [1]. The precise incidence of this entity is unknown. In a review of literature, approximately 0.013–0.3% of the findings from upper gastrointestinal tract barium studies support a diagnosis of SMA syndrome [3]. More females are affected by SMA syndrome. The SMA usually forms an angle of approximately 45° (range 38–56°) with the abdominal aorta, and the third part of the duodenum crosses posteroinferiorly to the origin of the SMA, coursing between the SMA and aorta [1, 4, 5]. Any factor that sharply narrows the aortomesenteric angle to approximately 6–25° can cause entrapment and compression of the third part of the duodenum as it passes between the SMA and aorta, resulting in SMA syndrome. In addition, the aortomesenteric distance in SMA syndrome is decreased to 2–8 mm (normal is 10–20 mm). Important aetiologic factors that may precipitate narrowing of the aortomesenteric angle and recurrent mechanical obstruction include, thin body build, exaggerated lumbar lordosis, visceroptosis and abdominal wall laxity, depletion of the mesenteric fat caused by rapid severe weight loss due to catabolic states such as cancer, surgery, burns, or psychiatric problems. Severe injuries, such as head trauma, spinal disease, deformity, or trauma leading to prolonged bedrest, dietary disorders such as anorexia nervosa and malabsorption may cause loss of fat [4, 6]. Our patient was having symptoms of anorexia nervosa and was treated for it and later on due to severe weight loss she developed SMA syndrome.

Patients with SMA syndrome may present symptoms of gastrointestinal obstruction, such as upper abdominal distension and epigastric tenderness, usually relieved
by posture changing. The acute presentation is usually characterized by signs and symptoms of duodenal obstruction. Chronic cases may present with long-standing vague abdominal symptoms, early satiety and anorexia, or recurrent episodes of abdominal pain, associated with vomiting. Delay in the diagnosis of SMA syndrome can result in malnutrition, dehydration, electrolyte abnormalities, and even death. Our patient was having chronic symptoms.

The differential diagnosis includes anorexia nervosa and bulimia. In addition, SMA syndrome should be differentiated from other causes of megaduodenum, including diabetes mellitus, collagen vascular conditions, and chronic idiopathic intestinal pseudoobstruction. The diagnosis of SMA syndrome is difficult. The diagnosis of SMA syndrome should be considered in patients with rapid weight loss who develop atypical, recurrent obstructive symptoms not to other common causes.. Confirmation usually requires radiographic studies, such as an upper gastrointestinal endoscopy, hypotonic duodenography, and CT scan. Contrast-enhanced CT scan and magnetic resonance angiography (MRA) is useful in the diagnosis of SMA syndrome and can provide diagnostic information, including aorta-SMA distances and duodenal distension [4]. Also, it can be used to assess intra-abdominal and retroperitoneal fat. Both these procedures are noninvasive and are probably equivalent to angiography, which has previously been suggested as the reference standard for establishing the diagnosis [5]. Computed tomography scan was the diagnostic in our patient as it revealed the narrow aortomesenteric angle and compression of third part of duodenum. Upper gastrointestinal study with barium revealed characteristic dilatation of the first and second parts of the duodenum, with an abrupt vertical or linear cut-off in the third part with normal mucosal folds. Once radiologic studies established diagnosis, first-line treatment is usually conservative with jejunal or parenteral nutrition for restoration of the aortomesenteric fatty tissue. Conservative initial treatment is recommended in all patients with SMA syndrome. This includes adequate nutrition, nasogastric decompression, and proper positioning of the patient after eating (i.e., left lateral decubitus, prone, or knee-to-chest position) [2, 5, 7, 8]. Enteral feeding using a double lumen nasojejunal tube passed distal to the obstruction under fluoroscopic assistance is an effective adjunct in treatment of patients with rapid severe weight loss and also eliminates the need for intravenous fluids and the risks associated with total parenteral nutrition [2]. Our patient’s symptoms had been resolved by nutritional support without any surgical treatment. Total parenteral nutrition (TPN) had revealed all the symptoms and we did not need any surgical interventions. Total parenteral nutrition is not definitive treatment for Wilkie’s syndrome.

Surgical intervention is indicated when conservative measures are ineffective, particularly in patients with a long history of progressive weight loss, pronounced duodenal dilatation with stasis,and complicating peptic ulcer disease [1, 2, 9]. Duodenojejunostomy is the most frequently used procedure, and it is successful in about 90% of cases. The use of laparoscopic surgery that involves lysis of the ligament of Treitz and mobilization of the duodenum has been reported [7]. Our patient was responded to the conservative management. The only medical treatment was administered TPN without any surgical treatment. She gained 4 kg before discharge and she has been following without any complication and complain for one year.

CONCLUSION

The treatment of Wilkie’s syndrome is aimed at the precipitating factor, which usually is related to weight loss. Therefore, conservative therapy with nutritional supplementation is the initial approach, and surgery is reserved for those who do not respond to hyperalimentation. Its recognition is important because early diagnosis of a partial obstruction may allow for medical rather than surgical intervention, as exemplified by our case. The diagnostic challenges of identifying vascular constriction between the aorta and superior mesenteric artery have been answered by advances in the field of computed tomography. Despite diagnostic
confusion with intestinal dysmotility syndrome, conservative therapy with nutritional supplementation is the initial approach and duodenojejunostomy is favored, if non-surgical treatment fails.

**********

Author Contributions
Ali Coşkun – 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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Bipartite patella separation with quadriceps tendon avulsion: A rare surgical case

Hasan Raza Mohammad, S Bitar, I Mc Laughlin-Symon, A Henry, G Batra

ABSTRACT

Introduction: The occurrence of bipartite patellar disruption simultaneously with quadriceps tendon avulsion is an extremely rare injury. A literature review found three documented cases in which only two repaired both the patella and tendon. We present a case report outlining this unusual injury and propose a method of surgical treatment. Case Report: A 45-year-old male presented following a fall directly onto his knee, with no significant medical history or previous injury. He was diagnosed with both separation of a type III bipartite patella and a clinical suspicion of a quadriceps tendon rupture. Intra-operatively, we found a complete tear of the quadriceps tendon at its insertion to the patella with disruption of the synchondrosis and significant separation of the bipartite patella. The displaced fragment was reduced and held using two cannulated screws, the quadriceps tendon was reattached to the superior pole of the patella using two anchor sutures and the retinaculum repaired.

Conclusion: We could find no documented cases of using both cannulated screws and anchor sutures as a fixation method for this injury. We recommend this technique as we found that it gave a stronger fixation and was associated with a good functional outcome for our patient.

Keywords: Bipartite patella, Quadriceps avulsion, Surgical repair.

INRODUCTION

The occurrence of bipartite patellar disruption simultaneously with quadriceps tendon avulsion is an extremely rare injury which is often misdiagnosed as a patellar avulsion fracture. We present our case of bipartite patellar disruption and quadriceps avulsion which we treated in our institution with open reduction and internal fixation of the bipartite component and anchor repair of the tendon component.

CASE REPORT

A 45-year-old male, fence erector (weight: 108 kg, height: 178 cm) presented to Accident and Emergency Department after slipping on a mat and impacting his left knee on a concrete step. He had no significant medical history. His main complaints were anterior knee pain, swelling and inability to weight bear. He scored 5/10
for both subjective and objective pain scores. It was an isolated injury and on examination, his knee injury was closed. The knee was significantly swollen and tender, more so over the pre-patellar and supra-patellar regions. The patient was able to actively flex his knee but could not demonstrate straight leg raise. However, any movement was very painful, (scored 3 on the baseline score admission on moving). Left hip and ankle examination were normal and no neurological abnormalities were detected. The patient received 60 mg codeine phosphate, 1 g paracetamol and 400 mg ibuprofen orally as pain relief.

Anterior posterior and lateral radiographs of his left knee were obtained and these showed a superolateral patellar bipartite fracture (Figure 1A–B). The patient denied having any injury to his patella prior to the incident. The patient was diagnosed with quadriceps tendon avulsion and bipartite patella through clinical examination and radiographic evidence.

Operative treatment was discussed with the patient and informed consent was obtained. The patient was treated surgically under general anaesthetic in the supine position. Tourniquet was used for 90 minutes during the operation. A longitudinal midline incision approach was used. Operative findings included a complete tear of quadriceps tendon at its insertion to the patella with disruption of the bipartite patella which had displaced a result of the injury (Figure 2). The displaced fragment of the bipartite patella was judged to be significant in size therefore reduced to its anatomical position and internally fixed to the main patellar fragment using two 4.0 mm ASNIS screws under direct intraoperative X-ray control (Figure 3). The distal quadriceps tendon then was repaired into the superior pole of patella using two 5.0 mm Miteck anchors. Repair of the retinaculum on the sides was completed using 2.0 Vicryl. The wound was closed in layers with Vicryl and Monocryl with the knee in slight flexion. The knee was then placed into a cylinder splint and he was scheduled to a follow-up clinic. Meanwhile he was encouraged to do range of motion ankle exercises.

Figure 1: (A) Antero-posterior radiograph of left knee. Indicates superolateral patellar bipartite separation, (B) Lateral radiograph of left knee showing the bipartite separation.

Figure 2: Intraoperative photo of left knee showing the complete tear of quadriceps tendon at its insertion to the patella along with disrupted, displaced bipartite patella.

Figure 3: Intraoperative photo of left knee showing reduction of displaced fragment of bipartite patella to its anatomical position.
Follow-up at nine month revealed that the patient was back at work which involves heavy lifting. The patient described painless walking on flat surfaces but described slight pain on walking quickly. On examination patient appeared to walk normally. There were no palpable gaps around the suprapatellar region or along the extensor mechanism and no bony tenderness over the patella or over the tendon. He could fully straighten his knee and do active straight leg raise comparable to the contra lateral side. Fully flexion of his knee was possible without discomfort and there was no joint effusion or joint line tenderness. Anterior drawer test and Lachman test were negative and the posterior cruciate ligament and collaterals were clinically stable. McMurray test was negative for both menisci. Check X-rays were also normal (Figure 4A–B). There was also no evidence of distal neurovascular deficit. However, the patient did have an area the size of a penny over the lateral aspect of the scar that was hypersensitive. This is likely to be due to the midline knee approach.

DISCUSSION

The occurrence of bipartite patella simultaneously with quadriceps avulsion is an uncommon injury which is often misdiagnosed as a patellar avulsion fracture [1]. Bipartite patella occurs when the secondary ossification centre of the patella fails to fuse with the primary center [1]. It has an incidence of 2-6%, with males having a higher predisposition rate (8:1) [2]. It occurs bilaterally in 43% of patients with bipartite patella [2]. However, usually it is asymptomatic (98% of cases) [3–4]. Bipartite patella is often diagnosed incidentally by plain radiographs [3, 5].

Direct trauma may disrupt the synchondroses, causing irritation and inflammation which manifests with fracture like symptoms. Onset either occurs gradually or immediately after the injury [6].

Common symptoms of inflamed/irritated bipartite due to trauma include; anterior knee pain, swelling of the synchondrosis and painful range of motion of the knee [5].

In 1943, Saupe developed a classification system describing the three types of bipartite patella (Table 1) [7, 8].

For fractured bipartite patella, if there is an extra articular fragment it can be excised, however if there is an intra articular fragment open reduction and internal fixation is advised [2, 9–11]. Different methods of internal fixation are described in literature including tension band wiring, using parallel interfragmentary lag screws and the combination of tension band wiring with cannulated lag screws. Biomechanical evaluation of different approaches has reported that patellar fractures stabilized with screws were significantly less likely to displace than the tension band technique [12]. The preferred method for this patient by the operating surgeon was fixation with interfragmentary screws.

Figure 4: (A) Anterior-posterior radiograph of left knee postoperatively showing satisfactory reduction and screw fixation of the separation as well as anchors to reattach the quadriceps tendon. (B) Lateral radiograph of the left knee postoperatively.
Table 1: Saupe’s classification of bipartite patella. Adapted from [1, 7, 8]

<table>
<thead>
<tr>
<th>Type</th>
<th>Part of patella affected</th>
<th>Relative prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Inferior pole</td>
<td>5%</td>
</tr>
<tr>
<td>II</td>
<td>Lateral margin type</td>
<td>20%</td>
</tr>
<tr>
<td>III</td>
<td>Superolateral nature</td>
<td>75%</td>
</tr>
</tbody>
</table>

A diagnosis of quadriceps avulsion is obtained through a full history and examination. It characteristically presents with a triad of pain, supra patellar gap and a loss of extensor mechanism of the affected leg [13].

There are several diagnostic imaging mediums such as plain radiographs, magnetic resonance imaging (MRI) and ultrasound. Plain radiographs should be the initial tool used as they demonstrate a good overall view, show loss of quadriceps and supra patellar mass etc. MRI scan is the most effective in establishing injury pattern [14].

The treatment for incomplete quadriceps avulsion is non operative, immobilization in extension followed by a physiotherapy program [14, 15].

Complete rupture/avulsion indicates prompt surgical management within 72 hours, which yields best results according to literature [15–17]. Most patients obtain a good range of motion but some may have persistent weakness preventing them from carrying out strenuous exercise [17].

Delay in surgery of more than 72hrs gives suboptimal results due to quadriceps tendon retraction and patella bafa [18].

CONCLUSION

Although bipartite patella and quadriceps avulsion is rare, repairing the avulsed tendon with fixation of the bipartite fragment in our case resulted in a good outcome. Therefore we can recommend this method of treatment in such an injury.

**********

Author Contributions

Hasan Raza Mohammad – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Writing the article and revising the content, Final approval of the version to be published

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Malignant triton tumor of maxilla: A case report

Rajjyoti Das, Anupam Sarma, Partha Sarathi Chakraborty, Jagannath Dev Sharma, Amal Chandra Kataki

ABSTRACT

Introduction: Malignant triton tumor (or malignant Schwannoma with Rhabdomyoblastic differentiation) is a very rare entity with poor prognosis. It is considered a high grade malignant neoplasm with poor outcome. In 70% of cases the neoplasm is associated with von Recklinghausen neurofibromatosis and in the remaining 30% it is the sole morbid finding. Local recurrence is frequent and distant metastasis preferentially situated in lung and brain. Surgery is the treatment of choice and post operative radiotherapy is always indicated. Histopathology and immunohistochemistry are helpful in diagnosis. We report clinical course, therapeutic approach, histopathology and immunohistochemistry of such a case. Case Report: A 19-year-old female presented in Dr B Borooah Cancer Institute, Guwahati with a complaint of swelling in the right side of the cheek with ulceration in the oral cavity. On clinical examination there was a round swelling in left cheek of 4x4 cm size with overlying normal skin. Computed tomography scan revealed a hypo dense lesion in the right maxillary sinus with erosion of the floor. Punch biopsy was taken from palatal growth and sent for histopathological examination which revealed a malignant neoplasm with sarcomatoid features composed of spindle cells. Immunohistochemical stains showed tumor cells positive for Vimentin, S-100 protein and, Desmin. Patient was taken up for surgery followed by radiotherapy. Conclusion: Malignant triton tumor of maxilla is extremely rare. The diagnosis must be based upon imaging study, histopathology and immunohistochemical features. Considering the aggressive nature of disease radical surgery should be followed by adjuvant chemoradiation.

Keywords: Malignant triton tumor (MTT), Surgery, Histopathology and immunohistochemistry

INTRODUCTION

Malignant peripheral nerve sheath tumor (MPNST) accounts for about 5–15% of all soft tissue sarcomas [1]. Malignant Triton Tumor (MTT) constitutes about 5% of all MPNSTs [2]. Common sites for MTT include head, neck, extremities and trunk [1–3]. This rare tumor with fewer cases reported in literature, generally affects adult patients but can also occur in children [4]. Morphology
and immunohistochemistry are helpful in diagnosis. A 5-year survival is low up to 10–20% indicating the high malignant nature of the disease [5]. Surgical resection of the tumor is thought to be best but due to its aggressive nature adjunct chemotherapy and radiotherapy may increase the survival [6, 7].

CASE REPORT

A 19-year-old female reported in the head and neck oncology out patient department with a complaint of swelling in the right side of the cheek since one and half months, with ulceration in the oral cavity since 25 days and pain in the right upper teeth with occasional bleeding from the mouth. On general examination she was average built. Respiratory and cardiovascular examination was normal. On ear, nose and throat clinical examination there was a round swelling in right cheek of 4x4 cm size with overlying normal skin. On palpation tenderness was present over the swelling with normal sensory finding. Intra oral examination revealed an irregular surface ulcer in the right side of hard palate with loose premolar and molar tooth. Neck examination revealed no cervical lymphadenopathy. Indirect laryngoscopic examination was normal. All other hematological examination and chest radiogram were normal.

Computed tomography (CT) scan revealed a hypo dense lesion in the right maxillary sinus with erosion of the floor, there was enhancement of the lesion in contrast study (Figure 1). Punch biopsy was taken from palatal growth and sent for histopathological examination (HPE). The HPE revealed a malignant neoplasm with sarcomatoid features composed of spindle cells (Figure 2). The malignant cells showed hyperchromatic nuclei and pale cytoplasm. Differential diagnosis for spindle cell sarcoma includes malignant peripheral nerve sheath tumor (MPNST), spindle cell carcinoma, monophasic synovial sarcoma and spindle cell form of malignant melanoma. Immunohistochemical stains showed tumor cells positive for Vimentin, subsets of tumor cells positive for S-100 protein and another subset of tumor cells was positive for Desmin (Figures 3, 4). Immunohistochemistry was negative for CK, HMB-45 and chromogranin.

Patient was taken up for surgery. With a Weber-Ferguson incision right maxilla was approached and it was found that growth eroded the anterior maxillary wall and whole tumor was removed enblock including the involved palatal part. Tumor free margins was obtained at the time of surgical resection macroscopically and temporary palatal prosthesis was applied. Three weeks following surgery a final prosthesis was applied.

Specimen consisted of multiple fragments of bone, fibrous tissue and tumor mass which weighed 16.3 gm measuring 5x4x3 cm. On cut surface it was composed of grey firm tissue with cystic spaces. The tissue was sent for histopathological examination. HPE confirmed Malignant Triton tumor. Bone and cut margins were

Figure 1: Computed tomography (CT) scan showing disease extension of the patient.

Figure 2: Photomicrograph showing sarcomatoid spindle shaped tumor cells with variably pleomorphic and hyperchromatic nuclei and mitosis. (H&E, 100X).

Figure 3: Immunohistochemical staining showing subsets of tumor cells are positive for S-100. (S-100, 100X).
free of tumor. Three weeks following surgery patient was subjected to radiotherapy (66 Gy in 33 fractions for 6 weeks) over maxillae including upper neck.

DISCUSSION

Masson first described a lesion that consisted of a malignant schwannoma with rhabdomyoblastic differentiation [8]. Subsequently, Woodruff et al. coined the term “Malignant triton tumor” to indicate such a neoplasm [9]. There are two types of tumor, sporadic or in association with NF-1. Those with von Recklinghausen neurofibromatosis constitute over 70% and displayed a marked male predominance, young age with a common presentation in head and neck. On the other hand, those without von Recklinghausen neurofibromatosis are mostly common in older age, female predominance and frequently located on trunk [10].

MPNST can be diagnosed on histopathology supported by positivity for S-100 protein. In morphology most areas show appearance of an extremely cellular spindle cell neoplasm with abundant mitoses. Although most tumors are quite monomorphic but heterologous elements like rhabdomyoblasts, cartilage and bone may be present in some cases. Such tumors show positivity for S-100 protein in 50-90% of cases, suggesting a nerve sheath origin. Rhabdomyoblasts are positive for immunostains such as desmin, myogenin and myo-D1 [11].

Though MPNST is non-radio-sensitive, like other sarcomas, wide local surgical excision followed by adjuvant radical radiotherapy is mostly adequate. Role of chemotherapy is still not been clearly defined. But chemotherapy might play some role in adjuvant setting in eradicating micro metastasis [6, 7].

MTT has poor prognosis with 5 years survival rate of only 5–15%, where as MPNST has a good survival of about 50-60% [12]. Poor prognosis of MTT depends on the location, grade and total radical resection [13]. Prognosis is good in head, neck, and extremities but very poor in buttock and other sites.

In spite of adequate treatment, local recurrence and distant metastasis rates approximate 25 and 48%, respectively. Survival of patients with head and neck MTT ranges between four months and 22 years [11]. Cytogenetic study has revealed some karyotypic changes associated with this tumor. Cytogenetic analysis has revealed some breakpoints which are considered as region for myogenic differentiation and probably responsible for rhabdomyoblastic differentiation [14]. Aggressive biologic behavior is may be due to amplification of c-myc oncogene [15].

CONCLUSION

Malignant triton tumor of maxilla is extremely rare. The clinical presentation may be misleading. The diagnosis must be based upon imaging study, histopathology and immunohistochemical features as it was in our case. Additional physical signs of NF-1 should also be excluded. Considering the aggressive nature of disease radical treatment of surgery should be followed by adjuvant chemoradiation.

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Ascaris intestinal perforation after trivial trauma

Kuldip Singh Ahi, Anand Munghate, Mahak Chauhan, Harnam Singh, Ashwani Kumar

ABSTRACT

Introduction: Isolated gastrointestinal perforation after blunt abdominal trauma and perforation seen from parasitic infestation are infrequent case presentations. Ascaris lumbricoides (round worm) is a common parasitic infestation in underdeveloped as well as developing countries. Case Report: Herein, we report a case of a 42-year-old male patient who was presented to the emergency department with history of trivial trauma and complain of abdominal pain, investigations lead to the diagnosis of peritonitis. The emergency laparotomy was done and unexpectedly, a live round worm was found to be the cause of a single jejunal perforation. It is suggested that the trivial trauma might have exacerbated the impending ascariasis perforation. Conclusion: Ascaris lumbricoides, an intestinal roundworm, is one of the most common helminthic human infestations worldwide. Infestation with this can result in a wide range of clinical presentations ranging from asymptomatic worm infestation to potentially fatal complications. Thus ascariasis should be investigated in patients with non-specific abdominal pain or intestinal perforation especially in tropical countries. It is saddening that in spite of worldwide improvement in public awareness of hygiene and good sanitation, there are still some parts of the world where prevalence of helminthiasis and their complications are rising. A jejunal perforation with single ascaris after trivial trauma is a rare entity. This unique case has highlighted the probability of blunt trauma intensifying an impending perforation by roundworm in this patient.

Keywords: Ascaris lumbricoides, Peritonitis, Trauma, Helminthiasis

INTRODUCTION

Worldwide Ascaris lumbricoides is one of the most common human helminthic infestations [1]. The durability of eggs, high number of eggs produced per parasite, poor socioeconomic conditions lead to its high prevalence. Ascaris transmission is increased as asymptomatically infested individuals continued to shed eggs for years [2]. In tropical countries where warm and wet climate provides suitable environmental conditions for its high prevalence, contrast to dry areas, where transmission occurs mainly in rainy season [3]. Jejunum or ileum is usual sites of habitat of an adult
Ascaris lumbricoides, an intestinal roundworm, is one of the most common helminthic human infestation worldwide [1]. Perhaps as much as one quarter of the world's population is infested, with a prevalence of 45% in Latin America and 95% in parts of Africa [5]. The faeco-oral route being common mode of Ascaris transmission by ingestion of raw vegetables and fruits containing embryonated eggs. Adult worms may be found in gastrointestinal system, hepatobiliary system or peritoneal cavity, resulting in a wide range of clinical manifestation like volvulus, gastrointestinal obstruction, intussusceptions, cholangiohepatitis, liver abscess, peritonitis, pancreatitis, cholecystitis and Loeffler’s pneumonitis [6]. The presence of dead adult worm in peritoneal cavity or reaction to ascaris eggs may result in granulomatous peritonitis, so one should suspect helminthiasis infestation in patient presenting with non-specific complain of abdominal pain or gastrointestinal perforation to avoid further complications [7]. The perforation due to helminthiasis is rare. Normal worms may result in two types of gastrointestinal perforation, primary and the secondary. The perforation through healthy intestine occur in primary type, where in secondary type it usually occurs in association with presence of a predisposing factor e.g., trauma, typhoid, tuberculosis, amebiasis or a weakness in intestinal wall [6, 8]. Trauma to the intra-abdominal structures can be classified into two primary mechanisms of injury—

**CASE REPORT**

A 42-year-old male patient was admitted to our emergency department with complains of abdominal pain in periumbilical region with history of blunt trauma to abdomen due to fall from motor cycle while taking the turn at minimum speed. He was not a frequent traveler and resides in small village near Patiala, India. Patient had history of intermittent abdominal pain for the last three months which use to subside by taking oral anti-acids and anti-spasmodic. There was no history of vomiting, fever or obstruction. On examination, tenderness and guarding were present in epigastric and periumbilical region. All routine blood investigations were performed and found to be within normal limits. The abdomen X-ray (erect position) showed free air under both domes of diaphragm. Peritonitis was diagnosed and patient was taken up for exploratory laparotomy. Intraoperative findings were as follows: A single perforation was present in the jejunum, about 14–16 cm distal to duodenojejunal junction, measuring 1x0.5 cm in size, on the anti-mesenteric border. One live round worm measuring about 12 cm in length was found protruding out of the perforation site (Figure 1), rest of the small intestine was examined for presence of any other worm. The biopsy was taken along the margin of perforation and the perforation site was closed in double layer. Peritoneal cavity was washed with warm normal saline. The abdomen was closed after placing proper drains. Postoperatively, the patient was given broad spectrum intravenous antibiotics and anti-helminthic therapy. Postoperative period was uneventful and patient was discharged under satisfactory condition on tenth postoperative day after removal of skin sutures. Biopsy showed focal acute non specific inflammatory reaction which implies that the inflammation was secondary to Ascaris infestation.

**DISCUSSION**

Ascaris lumbricoides, an intestinal roundworm, is one of the most common helminthic human infestation worldwide [1]. Perhaps as much as one quarter of the
compression forces and deceleration forces. Compression or concussive forces rupture the intestine by transiently increasing intraluminal pressure [9]. As seen in our case where trivial trauma increased the intraluminal pressure leading to rupture of an impending perforation because of presence of ascaris. Literature also discusses the synergistic action between typhoid and taenia causing intestinal perforation [10].

It is saddening that in spite of worldwide improvement in public awareness of hygiene and good sanitation, there are still some parts of the world where prevalence of helminthiasis and their complications are rising. The provision of clean drinking water, safe disposal of sewage, legislation to ensure high standards of food hygiene and programs to detect and monitor chronic carriers are advocated. These efforts should be complemented by mass anti-helminthic chemoprophylaxis which may further ameliorate the risk of early intestinal perforation [6].

CONCLUSION

A jejunal perforation with single *Ascaris* after trivial trauma is a rare entity. Infestation with *Ascaris* is common in underdeveloped and developing countries and should be evaluated. As a delay in management, abdominal complications can have a fatal outcome. In our case, the trivial trauma precipitated the impending perforation leading to puncture of the small intestine and symptoms of peritonitis. Thus, this unique case has highlighted the probability of blunt trauma intensifying an impending perforation by roundworm in this patient.

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CASE REPORT

Bleeding from aberrantly originating left gastric artery diagnosed by computed tomography scan

Mahibul Islam, Sonia Sandip, Md. Abu Masud Ansari, Raj Kapur

ABSTRACT

Introduction: Variations of branching pattern of the celiac trunk are well documented. Left gastric artery takes origin directly from the aorta in 0.5–15% of cases. Bleeding from such artery is rarely reported in literature. This type of rare variation has significant importance in surgical and radiological procedures. Herein, we describe a case of bleeding from a variant left gastric artery. Case Report: A 56-year-old male was admitted to our hospital with weight loss for endoscopic fine-needle aspiration of splenic abscess. Following fine-needle aspiration the patient had gastric bleed which was diagnosed by contrast-enhanced computed tomography (CECT) scan and abdominal angiography. The CECT scan of abdomen also revealed the variant left gastric artery which was source of bleeding. Upper gastrointestinal endoscopic was done and hemostasis was achieved by applying endoclip. Conclusion: Anatomical knowledge of variation is important for management of bleeding from such aberrant artery.

Keywords: Left gastric artery; Aberrant origin, Bleeding

INTRODUCTION

Left gastric artery is the largest artery supplying the stomach. It takes origin from celiac trunk as trifurcation with common hepatic and splenic artery. It may have a direct origin from the aorta [1]. This variation is significantly important in angiographic treatment of gastrointestinal hemorrhage. Angiographic embolization of such aberrant vessel is easier than normal anatomical one.

CASE REPORT

A 56-year-old male was referred from gastrointestinal centre for endoscopic ultrasound (EUS) and fine needle aspiration (FNA) of splenic abscess for evaluation of weight loss. The patient was hemodynamically stable and no evidence of hematemesis or melena. Endoscopic ultrasound guided FNA was done from splenic abscess along the greater curvature of stomach and no other intervention was done. During observation period, patient had tachycardia and pain abdomen and one episode of hematemesis. Computed tomography (CT) angiography was done which showed intragastric bleeding from left gastric artery (Figure 1). Endoscopy
was repeated and a spurting vessel in lesser curve was found and three endo-clips were applied, bleeding was stopped. In this particular case left gastric artery was arising from aorta directly (Figures 2–4). Patient was admitted with future plan of endovascular intervention but with no evidence of bleeding after 48 hours he was discharged home.

**Computed tomography angiography finding:**
Left gastric artery was found directly arising from anterior surface of abdominal aorta cranial to hepatolienal trunk. Hepatolienal trunk (Figure 1) was arising from anterior aspect of aorta just caudal to the origin of left gastric artery (Figures 2–4). Diameter of left gastric artery is 2.9 mm and of hepatolienal trunk was 6.3 mm. High attenuation (50–70 HU) material was seen in the stomach which was suggestive of hemorrhagic content (Figure 1). In addition, endoclips were seen along lesser curvature of stomach.

**DISCUSSION**

The classic description of normal celiac trunk anatomy is that the main trunk trifurcates into the left gastric, splenic and common hepatic arteries, which was found in 86%[1]. Reported cases of left gastric artery origin directly from aorta widely vary from 0.5–5% [1].

In 1928, Adachi first classified anatomical variations of the coeliac trunk. Adachi and Michels have classified the coeliac trunk into the following six different types [2, 3]:
- Type-1: normal branching
- Type-2: hepatosplenic trunk and left gastric artery from aorta
- Type 3: hepato-spleno-mesentric trunk and left gastric from aorta

![Figure 1: Contrast-enhanced computed tomography scan in cross section of same patient shows extravasation (longer arrow) of contrast and endoclip (shorter arrow).](image1)

![Figure 2: Contrast-enhanced computer tomography scan of abdomen in sagittal section in maximum intensity projected showing left gastric artery arises from abdominal aorta. Abbreviation: HST: hepatosplenic trunk, LGA: left gastric artery.](image2)

![Figure 3: Contrast enhanced computed tomography scan in sagittal section in volume rendering images shows left gastric artery arises from abdominal aorta. Abbreviation: LGA: left gastric artery.](image3)
• Type-4: hepatogastric trunk and splenic artery from superior mesenteric artery
• Type-5: splenogastric type- splenic and left gastric from the coeliac trunk and common hepatic artery from superior mesenteric artery; and
• Type-6: Celiacomesenteric trunk- splenic, left gastric, common hepatic and superior mesenteric arteries arise from a common trunk.

Variable fusion of right and left primitive yolk arteries when they localize in the dorsalis meso root give rise to these anatomical variations. Probably the hepatogastric trunk originates from right yolk artery, and the splenomesenteric trunk from the left one [4].

Sound knowledge of anatomical variation of celiac trunk is important for liver transplant surgeon and laparoscopic surgeon. With technical improvement, application of radiological intervention increases in gastroenteric diseases and penetrating abdominal trauma. Radiological anatomy of celiac axis and variation is basic requirement for such intervention [5].

It is worth mentioning that the reported findings are interesting given that this variation is quite rare and is not responsible for any hemodynamic problems [6].

There are level I evidences in favor of early endoscopy (within the first 24 hours of an acute upper gastrointestinal bleeding (UGIB) episode) in upper gastrointestinal bleeding. Early endoscopy significantly reduces rates of recurrent bleeding, the need for emergent surgery, and mortality in patients with acute non-variceal upper gastrointestinal bleeding [7]. Early endoscopy is appropriate for most patients and reduces mortality by 50% [8].

Hemostatic clips and endoclips: Clip immediately closes the defect in the vessel wall and stops the bleeding. Modification of the delivery system has made clip placement much easier. Four models of hemoclips are available: QuickClip, Resolution Clip; TriClip and InScope Clip. Out of all these, Resolution Clip is the clip of choice. Present best evidence shows the efficacy of hemoclips is similar to that of thermal coagulation in regards to control of initial bleeding, rebleeding rates and procedure duration [9, 10].

Endoclips may be preferred over other hemostatic methods in treatment of ulcers with coagulopathy or who require ongoing anticoagulation. Endoclips may also be preferable in the retreatment of lesions that rebleed after initial thermal hemostasis and active bleeding from larger vessel [10].

Over the Scope Clip Device (OTSC) is a shape-memory Nitinol alloy clip. As it is made of Nitinol, when the open clip released from the applicator it returns to its initial closed shape and close the defect in the vessel wall. Application is similar to the band ligator device. An endoscopy is performed before application of clip. The OTSC provides a safe alternative to endoclip in management of bleeding [11].

CONCLUSION

Bleeding from aberrant artery may be diagnostic problem. Knowledge of anatomical variation and endovascular procedure makes management of such bleeding much easier.

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Sonia Sandip – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
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Rapid regrowth of a large hepatic cyst following spontaneous rupture

Tomoki Nakajima, Manabu Okajima, Akiko Shibuya, Junko Yamaoka, Toshiaki Nakashima, Yoshito Itoh

ABSTRACT

Introduction: Spontaneous rupture of a simple hepatic cyst is rare, and the clinical course after rupture is not fully known. We report a case of a huge hepatic cyst which rapidly regrew after spontaneous rupture. Case Report: A 74-year-old male underwent further examination and follow-up of a huge hepatic cyst which was first detected at annual health check. On his first visit, the cyst was 10×9 cm in size. During the first 7 years of follow-up, the cyst gradually grew to 18×14 cm. Eight years after his first visit, ultrasonography showed that the huge hepatic cyst was without intracystic echogenic content. However, nine days after that, a computed tomography (CT) scan revealed that the cyst had nearly disappeared with remnant minimal cystic fluid and ascites although the patient was asymptomatic. Subsequently, within one month, the patient complained of back pain, and another CT scan showed that the cyst rapidly regrew to 13×10 cm in size. The cyst gradually regressed after sequential intracystic injection of absolute ethanol and minocycline. Conclusion: The rapid disappearance of the cyst was considered to be due to intraperitoneal rupture. The patient was asymptomatic just after rupture, but complained of back pain possibly because the regrowth was rapid. In some cases of huge hepatic cyst, rupture may be dismissed because it can be asymptomatic and the cystic fluid may have possibly already accumulated by the time symptoms appear.

Keywords: Spontaneous rupture, Hepatic cyst, Absolute ethanol, Minocycline

INTRODUCTION

Simple hepatic cysts are considered to be congenital. They are generally stable in size over time and require no treatment [1]. But some cysts may slowly enlarge and occasionally become symptomatic due to mass effect, rupture, hemorrhage, or infection [1]. Spontaneous rupture of a simple hepatic cyst is a rare occurrence [1, 2], and the clinical course after rupture is not fully known. We report a case of a huge hepatic cyst which rapidly regrew after spontaneous rupture. To our knowledge, this is the first case report which demonstrated such a rapid change in the size and morphology of the hepatic cyst during a short period after spontaneous rupture.
in the anterior lobe of the liver which was detected at annual health check. Magnetic resonance imaging (MRI) scan suggested that the lesion was a simple cyst of 10×9 cm in size and had no sign of infection, hemorrhage or malignancy (Figure 1). On April 30, 2008, he revisited our clinic for follow-up. A computed tomography (CT) scan demonstrated that the cyst had increased to 18×14 cm in size, but there was no evidence of complication or malignancy (Figure 2). On June 10, 2009, he visited our clinic again for follow-up of this change in the lesion. Ultrasonography showed that the huge hepatic cyst was without intracystic echogenic content (Figure 3). However, a CT scan performed on June 19, 2009 revealed that the huge cyst had nearly disappeared during this nine-day period, and only a small amount of cystic fluid remained (Figure 4A–B). Intestinal loops were seen in the region previously occupied by the large cyst. A small amount of ascites was found in contact with the anterior surface of the liver. Since the remnant cyst fluid in the cyst cavity and ascites were both minimal, the communication between these fluids was not directly shown on CT images. However, it was considered that the cyst had ruptured into the peritoneal space first, that the extravasated fluid had been mostly absorbed, and that the remnant fluid was detected as minimal ascites. There was no finding of communication between cyst cavity and biliary trees. On July 15, 2009 the patient complained of back pain, and another CT scan showed reappearance of the cyst which had grown to 13×10 cm in size (Figure 5). It was speculated that the ruptured lesion was repaired, and cystic fluid had accumulated in less than one month. The cystic fluid was macroscopically serous, and the cytology showed no sign of hemorrhage, malignancy or parasitic infection. After complete aspiration of the cystic fluid, we intracystically injected contrast medium and ensured there was no communication with the biliary tree or extravasation into the peritoneal cavity. Then, 70 mL of absolute ethanol was injected into the cyst. The ethanol was left in place for 20 minutes and all was withdrawn, as reported previously [3, 4]. One week after the injection, the cyst reappeared, thus, the therapeutic intervention was not sufficient. After complete aspiration of cystic fluid once again, 200 mg of minocycline hydrochloride dissolved in 20 mL of saline was injected into the cyst. The solution was left in place for 15 minutes and it was withdrawn [4]. Because the cyst remained for 2 months, 100 mL of absolute ethanol followed by 200 mg of minocycline hydrochloride dissolved in 20 mL of saline were sequentially injected with an interval of one week. All the intracystic injections were given under ultrasonic guidance and percutaneously. Thereafter, the cyst gradually regressed; the size of the cyst was 9.0×7.3 cm on December 2, 2009, 6.7×4.8 cm on April 7, 2010, 5.9×4.3 cm on August 4, 2010, 5.9×3.0 cm on January, 2011, 3.5×2.3 cm on December 9, 2011, and 3.0×2.3 cm in the last follow-up on March 22, 2013. The liver function tests were within normal range and stable throughout this clinical course.

**DISCUSSION**

There are some reports on the disappearance of non-parasitic hepatic cysts caused by intraperitoneal rupture after blunt trauma to the abdomen or coughing episodes.
However, since our patient had no previous history of trauma or other episodes leading to mechanical injury to the cyst, the rupture was spontaneous [2, 7, 8]. This complication of hepatic cysts is a rare occurrence [1, 2]. In one report, spontaneous rupture was attributed to necrosis of the secretory cells by increased intracystic pressure, or ischemic necrosis due to pericystic scar formation caused by local inflammation [7]. Some researchers observed intracystic hemorrhage prior to the rupture and speculated that the injured epithelial lining cells resulted in rupture [8]. In our case, there was no sonographic sign of intracystic hemorrhage on June 10, 2009, and the CT density of the remaining cystic fluid and ascites was not suggestive of hemorrhage. However, compared with the MR images in 2001 and the CT images in 2008, the cyst definitely increased in size and possibly became vulnerable to faint mechanical injury or insufficient blood supply to the lining cells which presumably primed the cyst for rupture.

There are a small number of reports on the sequential changes in the size of hepatic cysts during the obliteration process. In the case of cyst regression possibly through the disruption of the blood supply to the lining cells without rupture, a reduction in cyst diameter from 7.7 cm to 1.0 cm was observed during eight years of follow-up. Thus, the regression process seems to require a long-time [9]. However, cyst obliteration through rupture is reported to be sudden-onset and to rapidly progress. In our case, we speculated that the accumulation of a large amount of ascites just after rupture, its absorption by the peritoneal membrane and its clearance occurred before cyst disappearance [6]. This case suggests that the entire process can be completed only in nine days or less.

There are a few reports on the clinical course after the disappearance of hepatic cyst following rupture. In our case, cystic fluid accumulated again in less than one month, which suggested that the ruptured point was small and underwent spontaneous healing and closure.

Since re-rupture might cause bleeding inside the cyst or into the peritoneal cavity, prophylactic treatment was considered. Treatment options generally include radiological intervention by needle aspiration combined with injection of sclerosing agents or surgical approaches such as internal drainage with cystojejunostomy, wide deroofing and liver resection. Recent trends in the management of symptomatic hepatic cysts have shifted to minimally invasive procedures. Radiological intervention is safe, relatively noninvasive and considered as a first-line treatment especially for patients with high surgical risk or polycystic liver disease [1].

As a sclerosing agent, ethanol has been conventionally used, but it can cause pain, fever and intoxication. Some authors mentioned that that the ideal quantity of ethanol is 25% of the total cystic aspirate and recommended that not greater than 100 mL of ethanol be injected at one time [4]. Although there was no leakage of intracystically injected contrast medium prior to ethanol injection, absolute ethanol may cause re-rupture of the repaired cystic wall during injection. Therefore, in the first session, only 70 mL of ethanol was injected. Since no problems arose following the first session, 100 mL of ethanol was injected in the second session. To compensate for the volume reduction of ethanol and to bring about the cooperative effect, we also used minocycline hydrochloride expecting its strong acidity to degenerate or kill secretory cells of hepatic cysts. 

Figure 4: The computed tomography scan on June 19, 2009 revealed that the huge cyst disappeared during this nine-day period, and only a small amount of cystic fluid remained (arrowhead). Intestinal loops were observed in the space which had been previously occupied by the huge cyst. A small amount of ascites was found in contact with the anterior surface of the liver (arrow). (A) Plain images, (B) Postcontrast images.

Figure 5: On July 15, 2009 the patient complained of back pain, and another computed tomography scan showing re-appearance of the cyst, which grew to 13×10 cm in size.
hepatic cyst [4, 10].

In our case, from April 30, 2008 to June 10, 2009, the patient did not complain of pain or discomfort probably because cyst enlargement gradually progressed. On the other hand, on July 15, 2009, although the volume of cystic fluid was less than that on April 30, 2008, he complained of back pain possibly because the regrowth process after rupture was rapid. Thus, if the hepatic cyst is not accompanied by bacterial or parasitic infection or intracystic hemorrhage, rupture can be asymptomatic, as seen in our case.

CONCLUSION

Simple hepatic cysts are generally stable in size over time. Spontaneous rupture of a simple hepatic cyst is a rare occurrence and the clinical course after rupture is not fully known. We radiologically demonstrated that the huge hepatic cyst was rapidly obliterated by spontaneous rupture and fast regrew during a one-month period. To our knowledge, this is the first case report which demonstrated such a rapid change in the morphology of the hepatic cyst during a short period after spontaneous rupture. In our case the patient was asymptomatic before and just after rupture and he complained of back pain only after the cyst regrew rapidly. Therefore, in some cases of huge hepatic cysts, transient episodes of rupture may be dismissed because it can be asymptomatic and the cystic fluid may have possibly already accumulated by the time symptoms of abdominal or back pain appear.

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REFERENCES

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Metastatic melanoma causing multiple small bowel intussusceptions

Mark Halls, James Williamson, Mike Williamson

CASE REPORT

A 69-year-old female with known metastatic malignant melanoma presented with a three-week history of vomiting, altered bowel habit, abdominal distension and passing altered blood per rectum. An eighteen-month history of generalized abdominal pain, weight loss and anorexia were also noted. Prior to these three months, she had a similar obstructive episode. Computed tomography (CT) scan demonstrated metastatic deposits within the liver and on the serosa of the small bowel. This was managed conservatively with resolution of symptoms.

Clinical findings on the second presentation were: a distended, tympanic abdomen with no signs of peritonism. Initial bloods tests revealed a microcytic anemia and an elevated inflammatory response, but no signs of renal impairment or electrolyte imbalance.

Repeat CT scan revealed disease progression and a ‘target shaped lesion’ consistent with an intussuscepting small bowel loop within the right iliac fossa (Figures 1A–B). Laparotomy revealed four metastatic deposits within the small bowel. One lesion had caused obstruction due to complete intussusception, one had caused a partial intussusception and the remaining two were non-obstructing serosal lesions (Figure 2). The segment of small bowel containing all four lesions was excised by a wedge resection and continuity was restored with an end-to-end anastomoses.

Figure 1: (A) Transverse image from computed tomography scan showing classical target lesion of bowel intussusception (with oral contrast), (B) Coronal image from computed tomography scan showing intussusception of proximal bowel in the distal segment (with oral contrast).
In pediatric patients intussusception is the second most common cause of abdominal emergencies and is idiopathic in 95% of cases [1]. In contrast, it is rare in the adult population, accounting for only 1% of bowel obstruction and is frequently attributed to neoplasia [2]. Malignant melanoma is a locally invasive disease with a high capacity for metastasis. Metastatic spread is initially through the lymphatic system with distant metastases as a late feature [3]. The gastrointestinal tract represents one of the most common sites for metastatic spread of melanoma. The jejunum and ileum are particularly vulnerable to deposition [4]. Metastatic deposits are either submucosal, causing small bowel obstruction and potential ulceration; or polypoid, which can become a lead point for intussusception [3, 5].

CONCLUSION

In any patient with a history of malignant melanoma and non-specific gastrointestinal symptoms, including small bowel obstruction, the possibility of a small bowel metastases should be considered. This case illustrates the varying progression of metastatic lesions within the small bowel from serosal deposition, through to partial and complete intussusception.

REFERENCES


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How to cite this article

Moyamoya by magnetic resonance imaging scan

Caroline Edward Ayad, Ahmed Alamin Alnoor, Aymen El-Mesallamy

CASE REPORT

A 15-year-old Sudanese male was referred to the Magnetic Resonance Imaging (MRI) department with severe headache. Brain MRI axial T1, T2, diffusion, post contrast series, fluid attenuation inversion recovery (FLAIR) and time-of-flight magnetic resonance angiography (TOF-MRA) techniques were obtained. Images showed tiny punctuate signal void in all pulse sequence in both basal ganglia. Also there are a number of collaterals seen surrounding the mid brain and within the supra sellar cistern.

In postcontrast enhancement images there are obvious enhanced collaterals within the sulci of both cerebral hemispheres, the posterior fossa structures including the brain stem appeared intact. The obtained diffusion weighted images showed no evidence of recent ischemic insult. There was no obvious intra axial mass lesion, signal of blood degradation or extra axial fluid collection.

Magnetic Resonance Angiography (MRA) describes described both internal carotid arteries; they showed obvious occlusion of their supraclinoid portions without well formed circle of Willis, besides tortuous collaterals seen surrounding cistern forming A net-like appearance, the vertebral arteries as well as the basilar artery were intact.

An MRA revealed bilateral supraclinoid internal carotid arteries occlusion without well formed circle of Willis and collateralization as Moyamoya disease (Figure 1).

DISCUSSION

Moyamoya is a rare cerebrovascular disease among Japanese [1]. Ethnicity-incidence ratios for Whites as compared to Asian-Americans were 1:4.6, and as compared to African-Americans was 1:2.2. To the best
of our knowledge, no similar cases have been reported in Sudanese population in existing literature [2]. The pathogenesis of Moyamoya is idiopathic progressive arteriopathy of childhood; where occlusion of the circle of Willis, narrowing of distal internal carotid artery (ICA) and proximal circle of Willis vessels with secondary collateralization are detected the disease is poorly understood and may be due to genetic and environmental factors [3, 4]. The diagnosis of Moyamoya disease can be diagnosed by different imaging modalities and radiographic evaluation and it primarily depends on angiographic results, including occlusion of the supraclinoid ICA and formation of extensive collateral vessels [1, 3]. Several previous studies have revealed that Moyamoya disease can be identified on contrast-enhanced computed tomography (CT) or MRI, owing to their sensitivity to ischemic changes [5–7].

Three-dimensional CT angiography have limitations in diagnosis of Moyamoya disease because of the limited spatial resolution, difficulty in covering the whole intracranial vasculature network of leptomeningeal anastomotic channels [1]. The assessment of cerebral ischemia associated with Moyamoya by diffusion-weighted MRI has value in disease evolution [8]. Infarctions are better delineated with T₁ and T₂ imaging [9, 10]. The MRI findings in T₁ are multiple dot-like flow voids in basal ganglia, T₂-weighted images are of high signal small vessel cortical and white matter infarcts, collateral vessels appear as “net-like” cisternal filling defects, FLAIR shows Bright sulci at leptomeningeal “ivy sign” [3]. Another MR protocols are useful in Moyamoya; T₂ star gradient recall echo is useful prior hemorrhage, T₁ with contrast shows Lenticulostriate collaterals with enhancing “dots” in basal ganglia and “net-like” thin vessels in cisterns, Leptomeningeal enhancement gives “ivy sign”, MR spectroscop show Lactate in acutely infarcted tissues and NAA/Cr and Cho/Cr ratios frontal white matter increase after revascularization, Perfusion-weighted imaging (PWI) shows low perfusion deep hemispheric white matter, is relatively high perfusion posterior circulation, PWI may be abnormal when MRI still normal [3].

Moyamoya on MRI is characterized by diminishing of flow voids in the internal carotid and middle and anterior cerebral arteries together with collateral flow voids in the basal ganglia and thalamus [11–13].

Magnetic Resonance Angiography (MRA) has been used to demonstrate the intracranial vessels, where it is non invasive, no contrast media, and ionizing radiation is used. MRA is useful in the diagnosis of Moyamoya disease as mentioned by Yamada et al. [14]. MRI has been proposed to be used instead of conventional cerebral angiography [12].

CONCLUSION

Regardless of the excellent diagnostic value, broad imaging protocols and noninvasive nature, it has been proposed that magnetic resonance angiography should be used as a diagnostic imaging modality for Moyamoya disease.

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REFERENCES
Differentiation of pulmonary artery sarcoma from pulmonary embolus utilizing 128-slice dual source prospective cardiac-gated computed tomography scan

Antonino Germana, Nathan C Hawkes, Scott A Alexander, Gilbert E Boswell

CASE REPORT

A 54-year-old female with no significant past medical history presented with progressive dyspnea with exertion. Transthoracic echocardiogram was significant for a mass lesion and a pressure step-up from 6–55 mmHg at the origin of the right pulmonary artery (PA). Sarcoma was diagnosed on subsequent cardiac magnetic resonance imaging (MRI) based upon contrast enhancement of the mass (Figure 1) [1]. Thin section Flash® non-contrast, first pass post-contrast, and delayed adaptive sequential images were obtained to stage the disease and determine extent of involvement (Somatom Definition Flash®, Siemens Healthcare, Forchheim, Germany). Contiguously soft tissue-filled PAs were found to enhance with contrast (Figure 1). These findings are consistent with sarcoma and not embolus, which guided the patient toward neoadjuvant chemotherapy with adriamycin/ifosfamide and attempt at curative resection [2]. Subsequent pathologic examination confirmed a sarcoma with leiomyosarcomatous differentiation (Figure 2).

DISCUSSION

Primary pulmonary artery sarcoma is a rare malignancy with poor prognosis that frequently presents with symptoms of PA obstruction, pulmonary arterial hypertension, and right ventricular failure [3]. More frequently seen extensive thromboembolic disease presents similarly, which can delay definitive imaging diagnosis and management. Magnetic resonance imaging (MRI) is highly specific for identifying PA sarcomas because the tumor enhances with gadolinium contrast more than bland thrombus [1]. Chest computed tomography (CT) scan findings such as a low-attenuation filling defect occupying the entire luminal diameter of the main or proximal PA, an expansion of any segment of the pulmonary artery with an extensive intraluminal filling defect, or extraluminal extension can also help
differentiate pulmonary artery sarcomas from pulmonary artery thromboembolism [4]. As much as delayed phase contrast enhancement has helped in differentiating between benign and malignant mass lesions, visualization of a low attenuation filling defect within a pulmonary artery on contrast-enhanced cardiac CT studies can further suggest malignancy, such as pulmonary artery sarcoma, if the lesion demonstrates enhancement throughout the delayed phase.

CONCLUSION

New computed tomography imaging techniques such as 128-slice dual source computed tomography with prospective technique can optimize evaluation of a pulmonary artery mass and provide concomitant thin-section evaluation of the intra- and extravascular spaces, which can be critical in pre-planning for neoadjuvant chemotherapy and attempt at curative resection. To the best of our knowledge, we are first to present a case of pulmonary artery sarcoma that demonstrates enhancement throughout the delayed phase on prospective gated cardiac computed tomography. This finding may further differentiate pulmonary artery sarcoma instead of chronic thromboembolism.

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Figure 2: (A) Pulmonary artery sarcoma with leiomyosarcomatous differentiation and treatment effect (H&E stain, x100), (B) Pulmonary artery mass lesion in cross-section demonstrating complete occlusion of the vessel (H&E stain, x400).