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Spontaneous splenic rupture secondary to metastatic malignant spindle cell tumour

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ABSTRACT We report a case of pathological splenic rupture as a manifestation of malignant metastatic spindle cell tumour. To the best of our knowledge, this is the first case report of an atraumatic-pathological rupture of the spleen secondary to metastatic malignant spindle cell tumour. A 63-year-old man with a previous history of right upper limb amputation for an axillary malignant spindle cell tumour was admitted with an acute abdomen. Computed tomography showed a ruptured spleen. The patient subsequently underwent splenectomy. Histopathology confirmed the presence of malignant metastatic spindle cell tumour. Pathological splenic rupture is a rare manifestation of metastatic malignant spindle cell tumour. Background oncological history and thorough examination of the musculoskeletal system may provide important clues to make a prompt diagnosis.

Keywords: atraumatic spleen rupture, malignant spindle cell tumour, splenectomy
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INTRODUCTION

Pathological rupture of the spleen is a rare aetiology of an acute abdomen, and if the diagnosis is delayed, it may be life-threatening. The six major aetiological factors (Table I) that contribute to the pathological rupture of the spleen include neoplastic disorders (haematological, non-haematological, primary, metastatic), infectious disorders (viral, bacterial, protozoal, fungal), inflammatory or non-infectious disorders (pancreatitis, amyloidosis, Wegener's granulomatosis), genetic disorders (storage disease), drug or treatment-related disorders (anticoagulation, thrombolytic therapy, dialysis) and mechanical disorders (congestive splenomegaly, pregnancy).[1] Metastatic deposits from non-haematological cancers such as primary lung cancer, melanoma and choriocarcinoma have already been reported to cause spontaneous pathological rupture of the spleen.[2-4] However, to date, spontaneous pathological rupture of the spleen due to metastatic malignant spindle cell tumour has not been reported in the medical literature, except for a few cases of spindle cell angiosarcoma.[1] Here, we report a patient who underwent an emergency splenectomy following pathological spontaneous rupture of the spleen secondary to metastatic malignant spindle cell tumour.

CASE REPORT

A 63-year-old Caucasian man presented to the emergency department with a four-hour history of sudden onset of central abdominal pain, abdominal distension and episodes of fainting. On examination, he was cold, clammy, tachycardiac and tachypnoenic, with abdominal signs of peritonism. The Focused Assessment with Sonography for Trauma scan, performed in the emergency department, excluded a leaking abdominal aortic aneurysm. Six months prior to presentation, the patient had undergone a right upper limb amputation for an axillary (grade III) malignant spindle cell tumour. Following surgery, he had chemotherapy, the last cycle of which was given two months before this presentation. The patient was known to have multiple neurofibromata-like lumps all over his body (Fig. 1) for many years, but he had never undergone a biopsy to confirm the diagnosis.

After the initial assessment and resuscitation, the patient underwent urgent computed tomography (CT) of the chest, abdomen and pelvis. The CT image demonstrated an irregularly shaped spleen with a large, mixed-density haematoma associated with it (Fig. 2). Free intra-peritoneal fluid and a haematoma surrounding the liver were also seen. There were bilateral enhancing adrenal metastases, along with portal and splenic vein thrombosis. There was also a 10.5 cm heterogeneous and mostly hypodense lesion in the left flank, which was highly suggestive of a large retroperitoneal metastatic deposit, next to the lower pole of the shattered spleen, tail of the pancreas and left kidney. The patient's haemoglobin level at the time of presentation was 1.12 g/L.

The patient subsequently underwent a midline laparotomy and a splenectomy. Operative findings included a large haematoma at the lower pole of the spleen, covering a bleeding fungating tumour (Fig. 3). In addition, free blood in the peritoneal cavity and multiple lumpy deposits over the greater omentum and mesentery were also observed. A large retroperitoneal mass next to the left kidney and tail of the pancreas was not resected due to its close proximity to the surrounding viscera, portal and splenic veins. His postoperative recovery was complicated by the development of bilateral subphrenic collections and required a three-week long course of intravenous antibiotic therapy. Histopathology (Fig. 4) confirmed the presence of metastatic

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Table I. Aetiology of pathological splenic rupture.

Group	Examples
Neoplastic disorders	**Haemotological** • Various types of leukaemia • Hodgkin's lymphoma • Non-Hodgkin's lymphoma • Myeloproliferative disorders • Myelodysplasia **Non-malignant haematological** • Histiocytosis • Idiopathic thrombocytopenic purpura **Primary tumours** • Angiosarcoma • Cystic tumours • Haemangioma **Metastatic tumours** • Choriocarcinoma • Lung cancer • Melanoma
Infectious disorders	**Viral** • Infectious mononucleosis • Cytomegalovirus • HIV **Bacterial** • Tuberculosis • Typhoid • Endocarditis **Fungal** • Aspergillosis **Protozoal** • Malaria tropica • Malaria tertiana
Inflammatory and non-infectious disorders	• Acute and chronic pancreatitis • Primary and secondary amyloidosis • Polyarteritis nodosa • Wegener's granulomatosis
Genetic disorders	• Storage diseases • Rheumatoid arthritis • Systemic lupus erythematosus
Drug induced	• Anticoagulants • Thrombolytic agents
Mechanical disorders	• Pregnancy • Congestive splenomegaly • Spontaneous rupture with and without triggering factors

malignant spindle cell tumour, which was fungating and bleeding at the lower pole of the spleen. Histopathology of the skin lesions confirmed the presence of multiple neurofibromata. Moreover, histopathology of the omental lump also showed the presence of metastatic malignant spindle cell tumour. The patient was referred to the regional tertiary care centre for further treatment.

DISCUSSION

The malignant spindle cell tumour (or malignant peripheral nerve sheath tumour) is a rare variety of soft tissue sarcoma. It has intrigued pathologists and surgeons alike due to the diagnostic challenges associated with it. These difficulties relate to its cell of origin, its relatively aggressive nature (in terms of poor prognosis), the complex surgery for radical resection and the variable recurrence rate, which depends on the tumour grade.[5] Malignant spindle cell tumour is common in adults, but

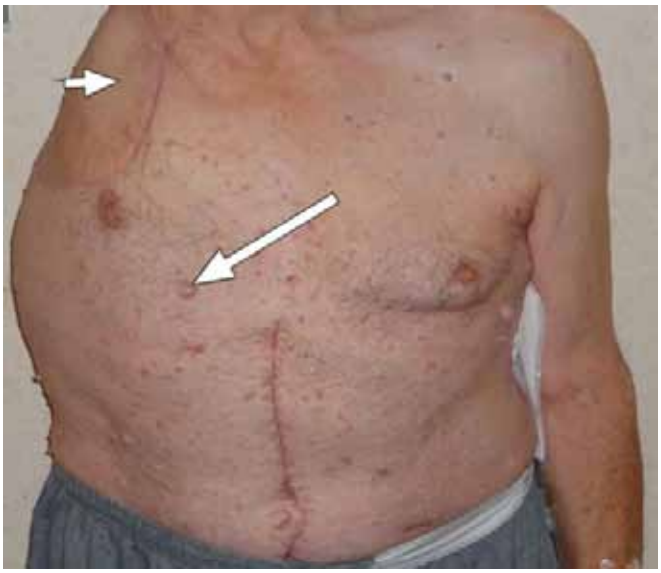

Fig. 1 Photograph shows neurofibromatosis of the skin (arrow) and the previous right upper limp amputation for malignant spindle cell carcinoma (arrowhead).

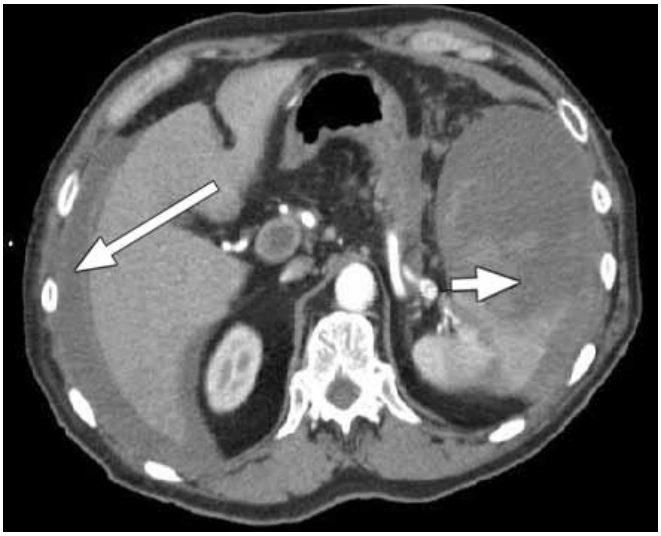

Fig. 2 CT image shows free blood around the liver from spontaneous rupture of the spleen due to the metastatic malignant spindle cell tumour (arrow). A large haematoma is seen at the pole (arrowhead).

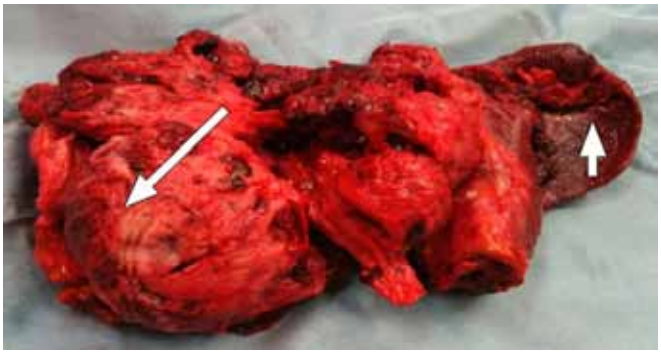

Fig. 3 Photograph of the resected spleen shows fungating metastatic deposits of the malignant spindle cell tumour (arrow) at the lower pole of the spleen and the normal-looking upper pole of the spleen (arrowhead).

5%–42% of cases are seen in patients of varying ages in the presence of multiple neurofibromatosis or Von Recklinghausen's disease, an association further characterised by an aggressive

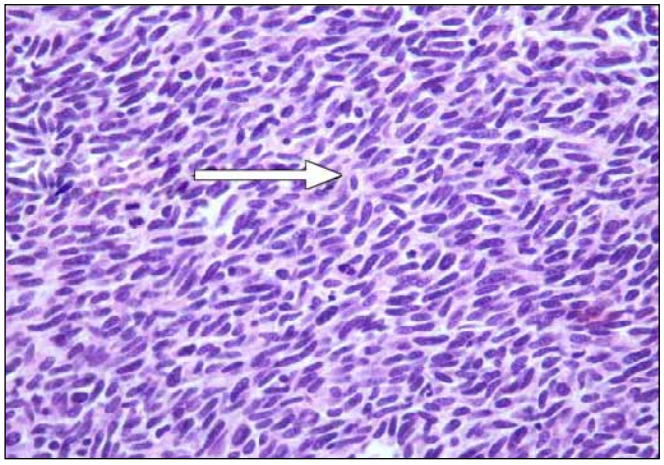

Fig. 4 Photomicrograph of the resected spleen shows malignant spindle cell tumour (arrow) (Haematoxylin & eosin, × 400).

disease course.[6,7] Our patient had been known to have neurofibromatosis for many years, which was proven on biopsy of the skin lesions taken at laparotomy. This certainly explains the aggressiveness of the initial disease, which resulted in splenic recurrence leading to pathological rupture of the spleen within six months of surgical resection of the primary limb's soft tissue tumour and chemotherapy.

In the medical literature, a five-year survival rate of 16%–52% has been reported for patients of malignant spindle cell tumour, but patients presenting with metastasis have a poor prognosis of 33% survival rate.[8,9] Treatment of the metastatic tumour depends on the location of recurrence. During laparotomy, our patient was found to have multiple sites of recurrence (such as the omentum, retroperitoneum, mesentery and spleen). Surgical excision of these multiple metastases was not possible. However, emergency splenectomy for life-threatening pathological rupture of the spleen due to spindle cell metastasis was warranted, and it was the most minimal intervention under the circumstances. To the best of our knowledge, this is the only reported case of pathological splenic rupture as a manifestation of a metastatic malignant spindle cell tumour.

In conclusion, although pathological splenic rupture as a manifestation of metastatic malignant spindle cell tumour is a rare entity, it should be included in the differential diagnosis. In the absence of any history of trauma, the diagnosis may be optimally made, provided that there is sufficient information regarding any possible oncological history. However, if there is no history of soft tissue tumour at the time of presentation of non-traumatic splenic rupture, the musculoskeletal system should be examined to exclude primary malignancy after life-saving measures have been taken.

REFERENCES

1. Renzulli P, Hostettler A, Schoepfer AM, Gloor B, Candinas D. Systematic review of atraumatic splenic rupture. Br J Surg 2009; 96:1114-21.
2. Ghinescu C, Sallami Z, Jackson D. Choriocarcinoma of the spleen–a rare cause of atraumatic rupture. Ann R Coll Surg Engl 2008; 90:W12-4.
3. Romero JI, Oliva RV, Utrera FH, Fernández AB, Jiménez MJ. [Smoker of 59 years with spontaneous splenic rupture]. Rev Clin Esp 2008; 208:163-4. Spanish.
4. Krapohl BD, Kömürcü F, Deutinger M. Spleen rupture to metastasis of thin melanoma (Breslow thickness of 0.75 mm). Melanoma Res 2005; 15:135.
5. Rekhi B, Ingle A, Kumar R, et al. Malignant peripheral nerve sheath tumors: clinicopathological profile of 63 cases diagnosed at a tertiary cancer referral center in Mumbai, India. Indian J Pathol Microbiol 2010; 53:611-8.
6. Brasfield RD, Das Gupta TK. Von Recklinghausen's disease: A clinicopathologic study. Ann Surg 1972; 175:86-104.
7. Evans DG, Baser ME, McGaurharn J, et al. Malignant peripheral nerve sheath tumours in neurofibromatosis 1. J Med Genet 2002; 39:311-4.
8. Wong WW, Hirose T, Scheithauer BW, Schild SE, Gunderson LL. Malignant peripheral nerve sheath tumor: Analysis of treatment outcome. Int J Radiat Oncol Biol Phys 1998; 42:351-60.
9. Cashen DV, Parisien RC, Raskin K, et al. Survival data for patients with malignant schwannoma. Clin Orthop Relat Res 2004; 426:69-73.