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CASE REPORT

An Undifferentiated High-Grade Pleomorphic Sarcoma of Spleen Presenting as a Splenic Abscess

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Abstract

Primary sarcomas are the rarest among primary malignant splenic neoplasms. Undifferentiated pleomorphic sarcoma (UPS), described in the past as malignant fibrous histiocytoma (MFH) of spleen is an extremely rare soft tissue malignant neoplasm. Due to non-specific symptoms, delayed presentation and aggressive nature of the tumour, overall prognosis is poor. We report the management experience of a case of splenic UPS of a 52-year-old Sri Lankan female who presented with epigastric fullness and discomfort. Initial clinical and ultrasonographic impression was of a large complex cystic lesion probably arising from the spleen or distal pancreas. Computed tomography confirmed the lesion to origin from the spleen. Due to spiking temperature and rising inflammatory markers, clinical suspicion was made of a splenic abscess related to the complicated cyst. At the laparotomy, a large predominantly cystic mass with concealed leaks arising from the spleen was noted containing inflammatory material within cyst cavities. The mass was adherent to the stomach and diaphragm. The patient underwent splenectomy with an uncomplicated post-operative period. Histology confirmed the lesion to be a UPS of the spleen and the patient was scheduled for a staging whole-body computed tomography. Unfortunately, she defaulted follow-up after 4 weeks from surgery and got readmitted as an emergency after 3 months with abdominal pain, distention and constitutional symptoms complicated with acute kidney injury. Ultrasonography proved her to have massive early local tumour recurrence and she succumbed due to the illness. This is the first reported case of UPS of the spleen in Sri Lanka.

Keywords Malignant fibrous histiocytoma · Splenic abscess

Introduction

Primary sarcomas are the rarest among primary splenic malignant neoplasms [1]. Undifferentiated pleomorphic sarcoma (UPS), previously known as malignant fibrous histiocytoma (MFH), is a common soft tissue neoplasm in adulthood which usually affect extremities and retroperitoneum than visceral organs [2]. However, primary UPS/MFH of the spleen is extremely rare and less than 25 cases have been reported up to 2020 [1, 3, 4]. Splenic MFH/UPS is an aggressive tumour with poor prognosis which usually shows systemic metastases by the time of the presentation. Current practice is to excise the tumour with a negative histological margin even in recurrent or metastatic disease [5].

Case Report

A 52-year-old Sri Lankan female presented with generalised abdominal distension for 1 month. She complained of dyspepsia but never experienced haematemesis, melena or jaundice. She never had previous attacks of pancreatitis or previous abdominal surgery. She was pale, and on abdominal examination, there was a large tender epigastric mass extending up to the umbilicus. Patient was anaemic (haemoglobin 8.5 mg/dl) and showed marginal neutrophil leucocytosis. Liver biochemistry, serum amylase and bilirubin levels were normal.

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Ultrasonographically, there was an epigastric mass probably arising from the spleen or the tail of the pancreas which predominantly consisted cystic lesions and a few solid areas. Cystic lesions contained echogenic debris with no internal vascularity. Minimal ascites was noted with no focal liver lesions. Computed tomography (CT) confirmed the mass to arise from the spleen and to have an irregular thick walled, well-defined cyst with peripherally solid areas. Furthermore, the mass was displacing the left lobe of the liver and the stomach and there were no calcifications (Fig. 1). Despite the pressure effect on surrounding viscera, there was no local invasion. Few enlarged lymph nodes were seen in para-aortic region. There were no omental, abdominal visceral or bone metastasis. The mass was considered a complex splenic cyst, a splenic abscess or a cystic splenic neoplasm. With the occurrence of elevated temperature and rising inflammatory markers, a splenic abscess in a complex cyst was considered the working diagnosis. The patient underwent an open splenectomy using a rooftop incision. Operative findings were of a large predominantly cystic mass of splenic origin with areas of concealed leaks; displacing stomach, left lobe of the liver and colon; forming adhesions to the diaphragm. Immediate postoperative period was uncomplicated.

The resected specimen showed a distorted spleen with a large, disrupted complex cyst measuring 10 × 10 cm (Fig. 2).

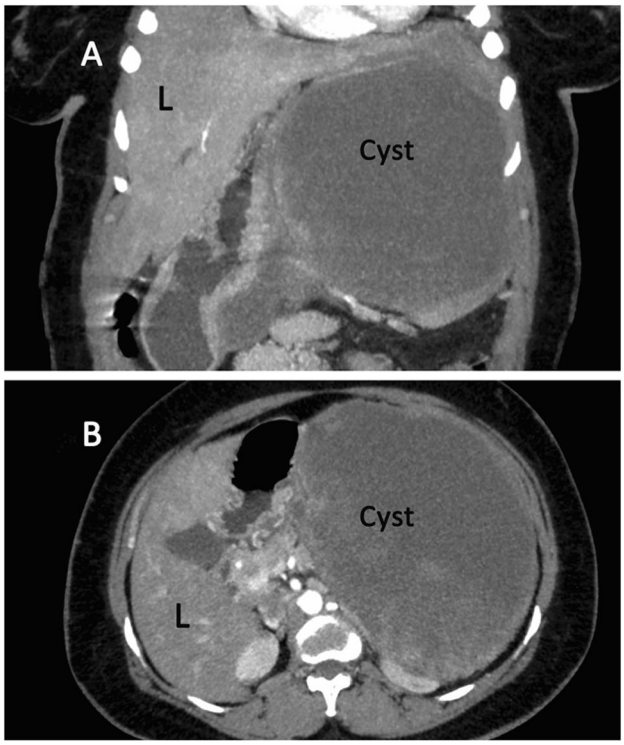

Fig. 1 Computed tomography images of the lesion. 1**A** and 1**B** shows the coronal and axial sections respectively. Also appreciate the displacement of surrounding organs (L, liver)

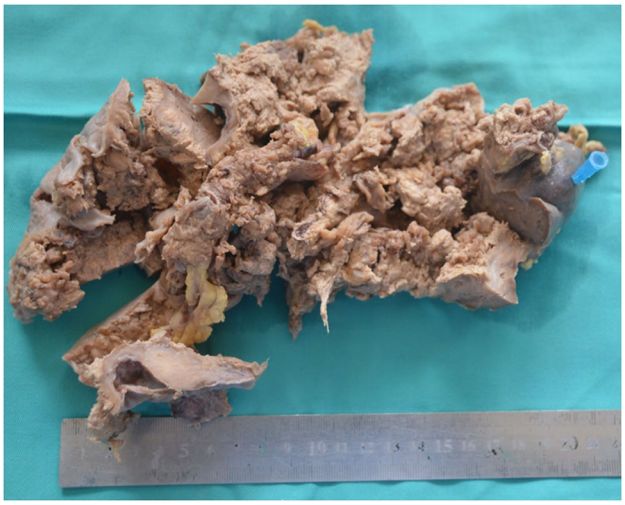

Fig. 2 Macroscopy of the sectioned specimen. Blue needle head shows the normal splenic segment

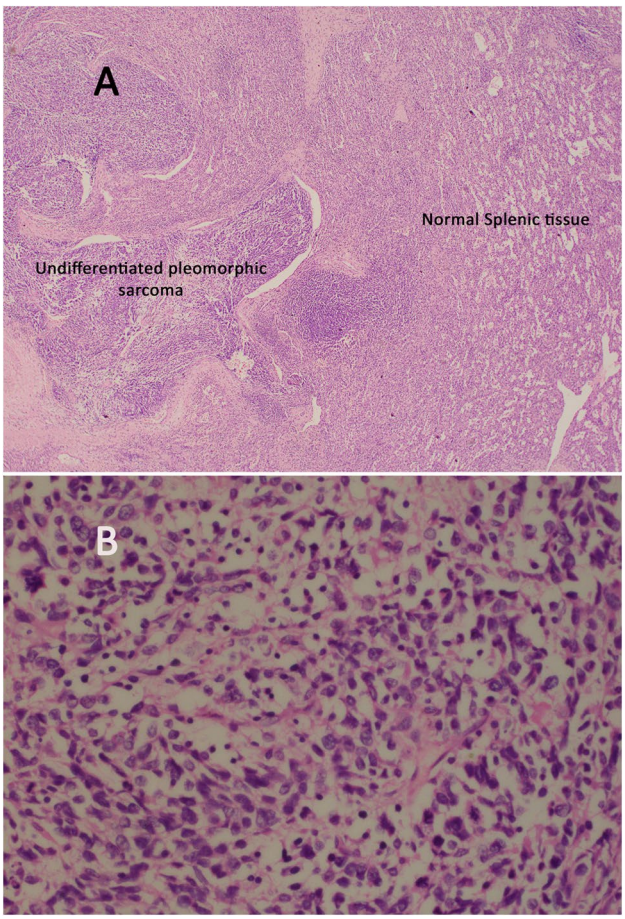

Fig. 3 Photomicrographs of the resected specimen. 3**A**, high-grade sarcoma within splenic parenchyma (haematoxylin and eosin × 10). 3**B**, undifferentiated sarcoma comprising fascicles of pleomorphic cells that lack specific immunohistochemical differentiation (haematoxylin and eosin × 40)

Table 1 Summary of previously reported cases of MFH/UPS of spleen

Author (year) country	Age/gender	Case summary	Management	Survival
Govoni et al. (1982) Italy	51/F	Abdominal pain, weight loss and splenomegaly	Splenectomy	Alive well at 7 months
Wick et al. (1982) USA	48/M	Fever, weight loss and splenomegaly	Splenectomy, chemotherapy	Alive with liver metastases at 18 months
	51/F	Fever, abdominal pain and splenomegaly	Splenectomy	Alive well at 17 months
	54/M	Abdominal pain and splenomegaly	Splenectomy	Alive well at 3 months
Bruneton et al. (1988) France	54/M	Abdominal pain and splenomegaly	Partial excision	Died at 3 months due to metastasis
Seiber et al. (1990) USA	41/M	Night sweats, weight loss and splenomegaly	Splenectomy, chemotherapy and radiotherapy	Died at 6 months due to metastasis
Lieu et al. (1993) Singapore	71/M	Abdominal pain, weight loss and splenomegaly	Splenectomy	Died following surgery in presence of liver metastasis
Bonilla et al. (1994) Spleen	42/F	Abdominal pain, weight loss and splenomegaly	Splenectomy, chemotherapy and radiotherapy	Died at 8 months due to peritoneal metastasis
Delgaudio et al. (1994) Italy	22/F	Acute abdomen and splenomegaly	Splenectomy	Alive well at 36 months
Mallipudi et al. (1998) UK	73/F	Abdominal pain, fever, weight loss and splenomegaly	Splenectomy	Died at 19 months due to peritoneal metastasis
Colovic et al. (2001) Yugoslavia	45/F	Abdominal pain, fever, weight loss and splenomegaly	Splenectomy, liver biopsy and chemotherapy	Died at 15 months due to recurrent liver metastasis
Ozaras et al. (2003) Turkey	51/F	Abdominal pain, fever, weight loss and splenomegaly	Splenectomy	Not available
Katsuura et al. (2006) Japan	82/M	Abdominal pain, fever, weight loss and splenomegaly	Splenectomy	Alive well at 18 months
Mantas et al. (2010) Greece	66/F	Abdominal pain and splenomegaly	Splenectomy	Alive well at 46 months
Hashmi et al. (2010) USA	76/M	Abdominal pain	Laparoscopic splenectomy	Not available
He et al. (2011) China	35/M	Acute abdomen and splenic rupture	Splenectomy	Died at 7 months due to metastasis
Feng et al. (2011) China	48/F	Abdominal pain and weight loss	Laparoscopic splenectomy	Alive well at 13 months
Amatya et al. (2011) Japan	77/M	Abdominal pain	Imaging studies	Died of tumour rupture
Dawson et al. (2012) India	30/M	Abdominal pain and splenomegaly	Splenectomy	Not available
Das et al. (2013) India	30/M	Abdominal pain and splenomegaly	Splenectomy and deroofing of the liver cyst	Died 3 weeks after surgery
Rakic et al. (2013) Croatia	57/M	Abdominal pain and weight loss 8 years after splenomegaly	Gastrectomy, distal pancreatectomy and left hemicolectomy for recurrent disease	Died at 4 months
Makis et al. (2017) Canada	63/M	Abdominal pain and splenomegaly	Core biopsy	Died prior to commencing treatment
Ashmore et al. (2020) UK	56/F	Abdominal pain and anaemia	Splenectomy	Alive with metastasis few months after surgery
Current study (2021) Sri Lanka	52/F	Abdominal pain and splenomegaly	Splenectomy	Died at 3 months due to metastasis

Microscopy showed a necrotic, cystic tumour of splenic origin. There were viable foci of tumour within splenic parenchyma surrounding the cyst space (Fig. 3A and B). The tumour showed diffuse sheets of spindle to epithelioid cells arranged in storiform pattern in a loose myxoid stroma. The constituent cells contained oval to round hyperchromatic nuclei and short ill-defined eosinophilic cytoplasmic processes. Mitoses were abundant. Vascular emboli are present. The tumour was limited to the spleen. Pleomorphic cells were positive for Vimentin and negative for LCA, AE1/AE3, CD68, MPO, Desmin, CD34, CD31, CD99, S-100, HMB45

and Myogenin. Therefore, the tumour was concluded as an undifferentiated high-grade pleomorphic sarcoma.

The patient defaulted follow-up at 4 weeks from surgery and got readmitted after 3 months with generalised abdominal distention and ill health. Ultrasonography showed tumour recurrence on the surgical bed with liver, pelvic and peritoneal deposits. Patient deteriorated drastically and succumbed due to early tumour recurrence, 3 months after splenectomy.

Discussion

Limited number of cases of splenic MFHs/UPSs has been reported since the first description in 1982 (Table 1). Majority have presented with upper abdominal discomfort/pain and splenomegaly, with or without constitutional symptoms. There have been exceptional presentations with acute abdomen due to tumour rupture or haemorrhage [1, 6]. CT has been the main preoperative imaging method for assessment of cystic/solid lesions of the spleen in presence of MFH/UPS. Misleading working diagnosis of splenic abscesses and Echinococcosis/hydatidosis has been made due to the rarity of this entity [1, 4, 6].

Since Theodor Billroth performed the first splenectomy for a soft tissue sarcoma in 1881, surgical resection with a histologically negative margin has been the mainstay of treatment for splenic sarcomas [5]. Despite achieving tumour-free margins, local recurrences have been common leading to poor survival outcomes (Table 1) [5, 6]. Adjuvant chemotherapy and radiotherapy have been used in splenic UPS/MFHs without much long-term survival [5]. Prognostic data is limited in these extremely rare malignancies. Within available data, the histological grade of the tumour seems to have higher influence on survival rather than tumour staging [1, 5, 6]. In poorly differentiated tumours, survival has been less than 1 year as in our case.

From a surgical point of view, this case highlights the need for the surgeons to be considerate of rare entities like splenic UPS as differential diagnosis, when patients present with splenomegaly and constitutional symptoms. Multidisciplinary approach would be essential to get the birds' eye view of the clinical picture with the complexities in interpretation of imaging modalities in the presence of elevated inflammatory markers.

Declarations

Competing Interests The authors declare no competing interests.

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