Primary Malignant Cystic Solitary Fibrous Tumour of the Spleen

Rachel Maria Gomes · Dilip Kothari

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Introduction

Solitary fibrous tumour (SFT) is a rare mesenchymal neoplasm that was first described in 1931 by Klemperer and Rabin. It most frequently affects the pleura and other serosal sites but can arise from virtually any site in the soft tissue. Clinically, SFTs are usually asymptomatic and eventually cause symptoms after reaching a very large size or when vital structures are involved. SFTs are distinguished from other tumours with a similar appearance due to the availability of characteristic immunohistochemical markers. In this manuscript we report the first case of a primary malignant cystic solitary fibrous tumour of the spleen. The small number of patients with extrathoracic SFTs and unreported occurrence within the spleen and its unknown natural behaviour creates the need for reporting and follow-up of all identified cases

Case Presentation

A 62-year-old man presented to us with complains of sudden onset left sided abdominal pain for a few days and noticing of a large lump in abdomen. He had no history of weight loss or loss of appetite. On physical examination, a large mass was palpated that filled the entire left upper quadrant of the abdomen which was tender. All laboratory investigations were within normal limits. Contrast enhanced computed tomography of the abdomen demonstrated a large well defined cystic tumour arising from spleen with

central hypoechoic area suggestive of haemorrhage within and thick irregular wall showing heterogeneous enhancement with intravenous contrast with areas of necrosis and calcification displacing the stomach and left kidney measuring 19×17.5×12.5 cm in diameter (Figs. 1a and b). Laparotomy was performed and revealed a large cystic tumour arising from the spleen and a splenectomy was performed.

On pathology gross examination showed a large tumour arising from spleen measuring 22×19×15 cm that weighed 2010 g. Macroscopically the tumour was well encapsulated and cystic with thick irregular wall with large papillary like projections within (Fig. 2). Microscopically the tumour was composed of neoplastic spindle cells, with uniform, elongated nuclei, separated by few thick bands of collagen (Fig. 3a). A few highly cellular areas with high mitotic rate of 10–13 mitosis/10 HPF were detected. An abundant thin walled vascular network was present with some hemangiopericytoma like vascular spaces (Fig. 3b). Immunohistochemical staining revealed strong expression of CD34, bcl-2 in virtually all tumour cells and focally weak positive for epithelial membrane antigen, S100 and MIC-2. No immunoreactivity was shown with CD-117 and smooth-muscle actin. He was diagnosed as having a solitary fibrous tumor. Postoperative course was uneventful and he is well at a 3 months follow up with no local or distal recurrence.

Discussion

SFTs can be found in any location most commonly in the pleura [1]. Extrapleural SFTs have been reported in the peritoneum, pericardium, lung parenchyma, upper respiratory tract, orbit, thyroid, parotid gland, thymus and liver parenchyma [2]. There have been no reported cases of primary tumours in the spleen.

R. M. Gomes (✉) · D. Kothari
Department of Surgical Gastroenterology,
NUSI Wockhardt Hospital,
Panzorconni, Cuncolim,
Salcette, Goa, India 403 703
e-mail: dr.gomes@rediffmail.com

Fig. 1 Contrast enhanced CT scan demonstrating cystic tumour arising from spleen with enhancement of wall with areas of necrosis and calcification (a) transverse section (b) coronal section

Extrapleural SFTs occur between 20 and 70 years of age and affect both sexes equally. They are often very large tumours with dimensions from 1 to 25 cm and half remain

Fig. 2 Splenectomy specimen. Large cystic tumour arising from spleen opened out displaying interior with irregular papillary like projections

Fig. 3 Micrograph of solitary fibrous tumour (hematoxylin and eosin staining). a Spindle-shaped cells exhibiting nuclear atypicity with collagen deposition. b Thin walled vascular network with some hemangiopericytoma like vascular spaces

asymptomatic at presentation. Large tumors can present with compression symptoms, while rarely with paraneoplastic syndromes (hypoglycemia secondary to insulin-like growth factor). [3]

Tumours occupying at least 40 % of the affected hemithorax have been proposed to be considered as a “giant” solitary fibrous tumour though extrathoracic sites haven’t been defined but the term is used liberally for large tumours. [2, 4, 5]

A positive immunohistochemical staining for CD34 and vimentin with a haemangiopericytoma like appearance on microscopy is considered the hallmark of SFTs [6]. Histologically SFT consist of bland spindle cells with features ranging from hypercellular to myxoid or hyalinised pattern-less hypocellular areas. A hemangiopericytoma-like vascular pattern with abundant branching of thin-walled vessels dissecting through the tumour are typical findings mainly in hypercellular areas of tumors [7]. The focal presence of intense hypercellularity accompanied by increased nuclear atypia, elevated mitotic rate and necrosis are criteria for malignant change. [5, 7]. Some variants described are giant cell SFT and fat forming SFT.

Immunohistochemically a SFT commonly expresses CD34, CD99 and bcl-2. CD34 is highly characteristic of SFTs and is required for diagnosis however it is not entirely

specific expressed by entities such as angiosarcoma and gastrointestinal stromal tumour [8]. Therefore of importance is CD117 negativity, CD 31 negativity, Factor VIII negativity combined with the histomorphology to exclude gastrointestinal stromal tumour and angiosarcoma. [9, 10]. The strong expression of CD99 and bcl-2 in the tumour cells further supports the diagnosis of SFT but alone would not be sufficient. A variety of other entities, such as leiomyosarcoma, fibromatosis, dermatofibrosarcoma protuberans, hemangiopericytoma, malignant fibrous histiocytoma, fibrosarcoma and sarcomatoid mesothelioma which are frequently considered in the differential diagnosis of solitary fibrous tumor, are uniformly bcl-2 negative [11]. Epithelial membrane antigen (EMA), and smooth muscle actin (SMA) may occasionally be expressed. They are usually negative for S-100 protein, desmin and cytokeratins [8, 12].

The treatment of choice for benign and malignant SFTs is complete en bloc surgical resection to achieve tumour-free margins. Local recurrence or onset of metastasis depends mainly on the resectability and in addition correlates with location, histological features (nuclear atypia, increased cellularity, necrosis, mitotic figures $\geq 4/10$ high-power fields), and dimensions.

An intra-thoracic location shows a metastatic rate of up to 15 % while extra-thoracic tumours may demonstrate metastasis in approximately 6 % [13]. In benign SFTs 8 % will recur after first resection with recurrence usually cured with additional surgery and in malignant SFTs approximately 63 % will have recurrence of which more than 50 % will succumb to disease progression within 2 years. A recent study shows that a combination of patient age and tumor size with mitotic index and necrosis better predicted both time to metastasis and disease-specific mortality. While small tumors with low mitotic rates are highly unlikely to metastasize those with necrosis were more likely to metastasize. Large tumors ≥ 15 cm, which occur in patients ≥ 55 years, with mitotic figures $\geq 4/10$ high-power fields have a high risk of both metastasis and death. [14]

They are chemo-resistant tumours like sarcomas and use of chemotherapy and radiotherapy is reserved to metastatic or symptomatic non-resectable SFTs. The most effective drugs are anthracyclines and ifosfamide followed by gemcitabine and dacarbazine commonly used in soft tissue sarcomas. [15] Radiation therapy may be of benefit in combination with chemotherapy. [16] It is recently reported that drug imatinib mesilate seems to exert some activity on

SFT expressing the wild type platelet-derived growth factor. [17]

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