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Case Report

Solitary fibrous tumor of small bowel mesentery with postoperative bowel obstruction: a case report and review of literature

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Abstract: Solitary fibrous tumor (SFT) which is an extremely rare clinical entity has been reported infrequently. Most commonly it is distinguished into pleural and extrapleural forms, with same morphological resemblance. There has been many literatures reported regarding extrapleural form of SFT but few cases of SFT originating from small bowel mesentery have been reported till now. We here report one case of SFT of small bowel mesentery with some eventful postoperative bowel obstruction and literature review.

Keywords: Solitary fibrous tumor, small bowel mesentery, bowel obstruction

Introduction

Solitary fibrous tumor (SFT) (formerly known as hemangiopericytoma) is a rare mesenchymal neoplasm, which was first described in 1931 by Klemperer and Rabin as a distinct mesothelial tumor [1]. It is usually a localized benign neoplasm arising from the submesothelial mesenchymal layer of a soft tissue [2]. SFTs of pleural origin have been reported in many literatures but the extrapleural origin has come in concern in recent literatures. Extrapleural SFTs commonly occur in retroperitoneum, abdominal cavity, deep soft tissue of extremities and head and neck [3] whereas intra-abdominals SFTs includes liver, stomach, retroperitoneum, kidney, prostate and small bowel mesentery. There have been few cases of SFT originating from small bowel mesentery reported in literatures till now. Extrapleural SFTs are difficult to distinguish from other soft tissue tumors as well as its malignant features [4].

We are presenting here, a case of SFT originating from small bowel mesentery. In this case we would like to give more emphasis and concern regarding the postoperative complications and its management that patient had gone through.

Case report

A 41-year-old healthy police officer presented with complains of abdominal mass with a gradual increase in size with recent two months. The patient claimed that the mass was accidentally noticed during his regular exercise two months ago and sometimes uses to be movable. He did not have any symptoms of nausea, vomiting and abdominal distension. There had been no change in bowel habits with irrelevant history of weight loss. He had no relevant medical or family history. During routine health examination in some other hospitals, the mass was presumed to be an intra abdominal “Tumor”. Following this, he was referred to our hospital for a second opinion and further work-up.

During the physical examination, there was a solid, non-tender, palpable mass in left lower quadrant of the abdomen. It was non-pulsatile and mobile during deep palpation. Laboratory finding showed normal blood routine results and unremarkable tumor markers. Routine preoperative evaluation was all in normal limits.

Complete abdominal CT scan was done and which demonstrated well-defined, large mass in the abdominal cavity. It was well-circumscribed and measuring 9.2 cm × 9.0 cm × 7.7 cm in size, with an extensive vascular communication around the tumor margin. Precontrast CT (**Figure 1A**) image showed a mass with unequal density distribution with relative hypo-attenuation. Contrast CT (**Figure 1B**) revealed a spherical inhomogeneous soft tissue mass located in

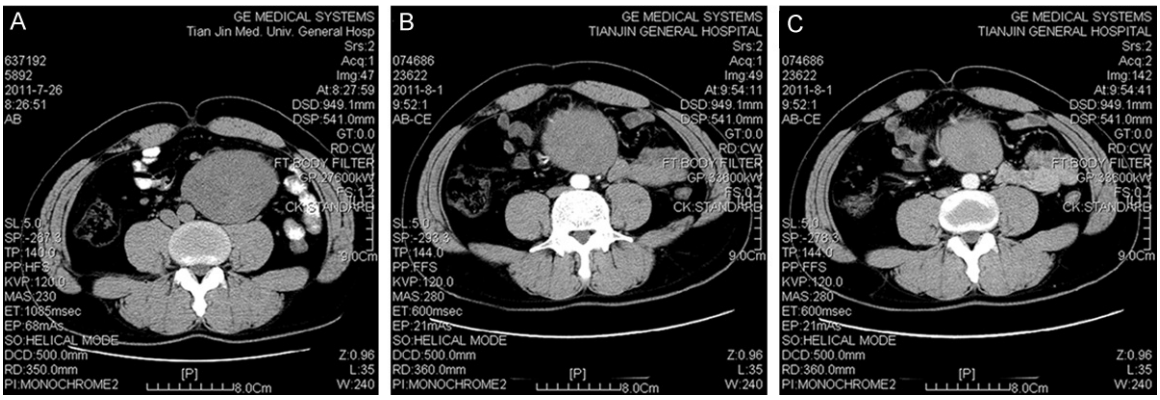

Figure 1. Abdominal CT scan demonstrated well-defined and large mass within the abdominal cavity. Pre-contrast imaging showed an unequal density distribution with relative hypo-attenuation of the mass (A). Arterial Phase showed inhomogeneous well defined mass with extensive vascular communication (B). Venous Phase showed the mass with delayed enhancement (C).

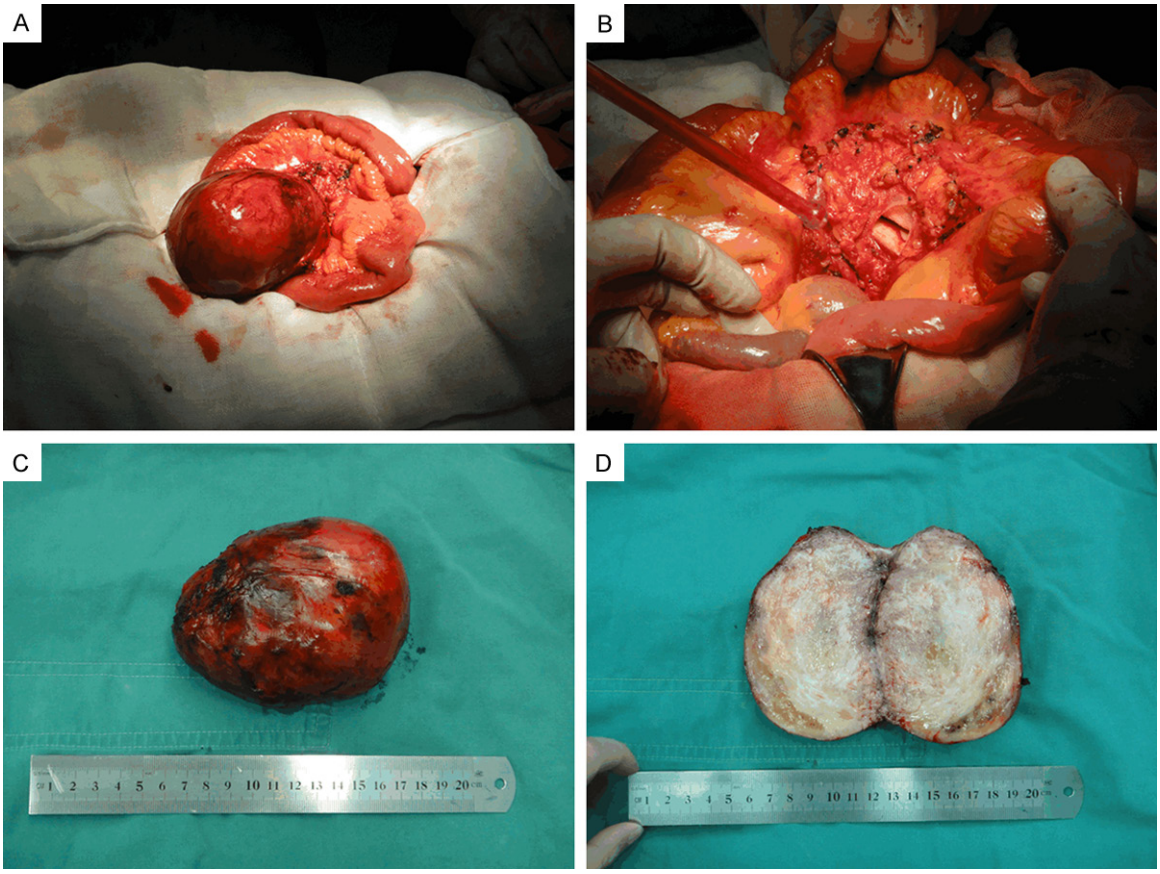

Figure 2. Intraoperative findings (A) with a gross appearance of a firm, well demarcated, and lobulated mass originated form the small bowel mesentery. After excision of the mass from the small bowel mesentery, we checked the viability of the small bowel (B-D). Gross appearance of cut surface showed yellowish white solid mass.

left abdominal region with involvement of the small bowel mesentery. In the venous phase (**Figure 1C**) mass showed a characteristic of delayed enhancement.

Exploratory laparotomy was done with intraoperative finding of a large rounded and well demarcated mass arising from the mesentery of the distal ileum, approximately 3.5 meters

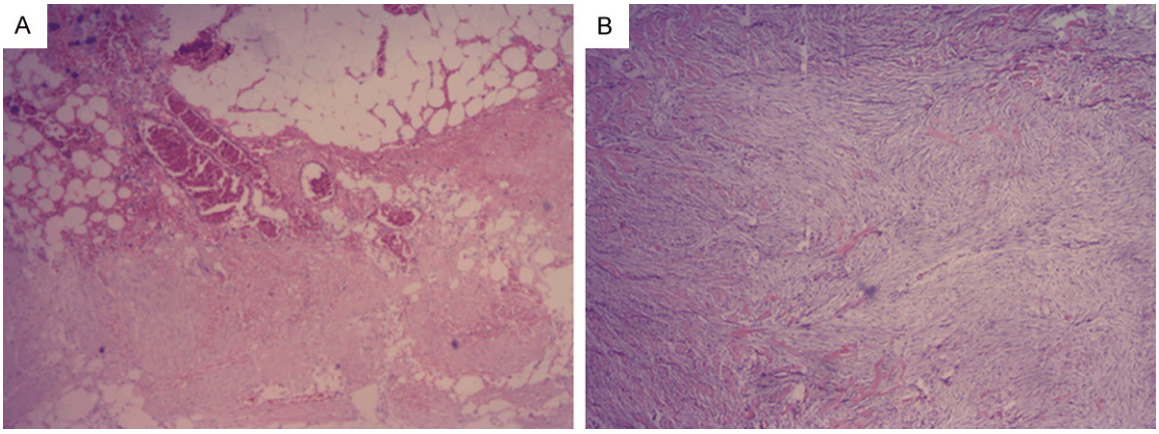

Figure 3. Histological finding (H&E staining): Microscopy showing a tumor composed fibrous lesions containing large collagenized area with hypercellular and hypocellular/fibrous area (×40) (A). Spindle cells with patternless pattern (×100) (B).

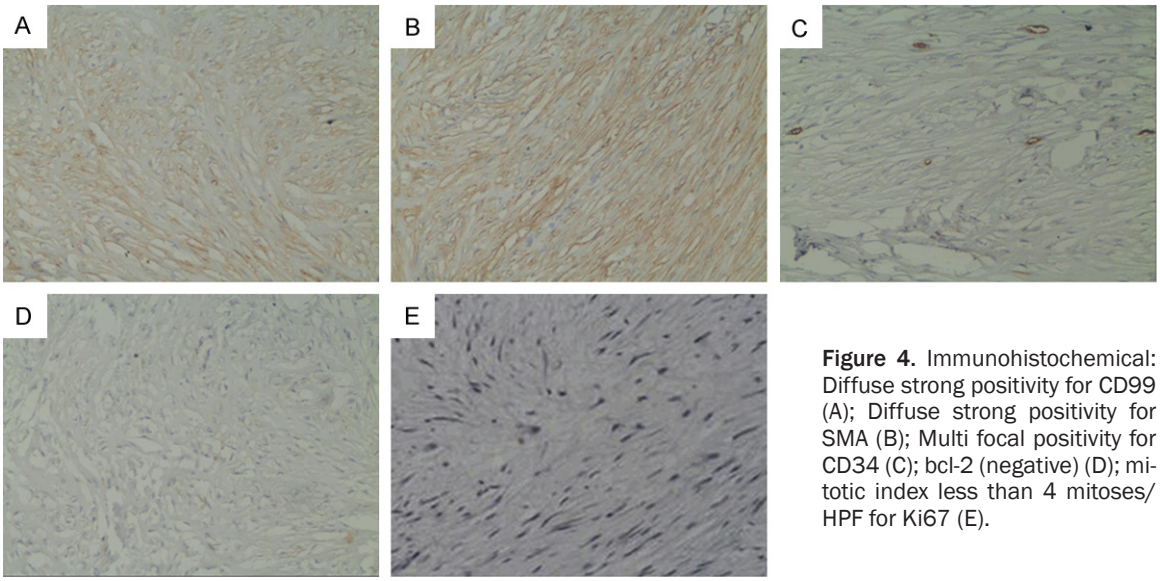

Figure 4. Immunohistochemical: Diffuse strong positivity for CD99 (A); Diffuse strong positivity for SMA (B); Multi focal positivity for CD34 (C); bcl-2 (negative) (D); mitotic index less than 4 mitoses/HPF for Ki67 (E).

distal from the Treitz Ligament. Successful surgical excision of the mass was done (**Figure 2A**). After excision of the mass from the mesentery (**Figure 2B**), viability of small bowel was checked and was found to be normal. The gross surgically resected specimen showed a large mass of size 10 cm × 8 cm × 8 cm, which was hard in consistency, smooth surface and completely encapsulated (**Figure 2C**, **2D**).

Histologically, the tumor was revealed with spindle cell proliferation (**Figure 3A**, **3B**). Immunohistochemical staining showed diffuse strong positivity for CD99 (**Figure 4A**) and SMA (**Figure 4B**) as well as multi focal positivity for CD34 (**Figure 4C**) where as Bcl-2 (**Figure 4D**) was negative. The mitotic index for Ki67 (**Figure 4E**) was less than 4 mitoses/HPF.

First three postoperative days was quite uneventful but during the fourth and fifth postoperative days the patient had features of (subacute) incomplete bowel obstruction. Conservative treatment was carried out but in the tenth postoperative day the patient had a feature of acute bowel obstruction. The radiological findings were suggestive of acute bowel obstruction with increased density of mesentery in the operated site (**Figure 5**).

Emergency exploratory laparotomy was done on the tenth postoperative day with a finding of

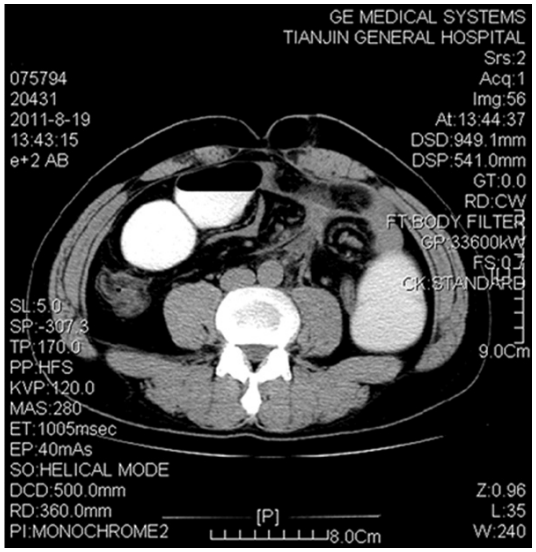

Figure 5. Emergency Plain abdominal CT scan on day 10 Post-operation showed the features of acute bowel obstruction with dilated bowel loops and increased density of mesentery, may be previous surgical area? (White arrow).

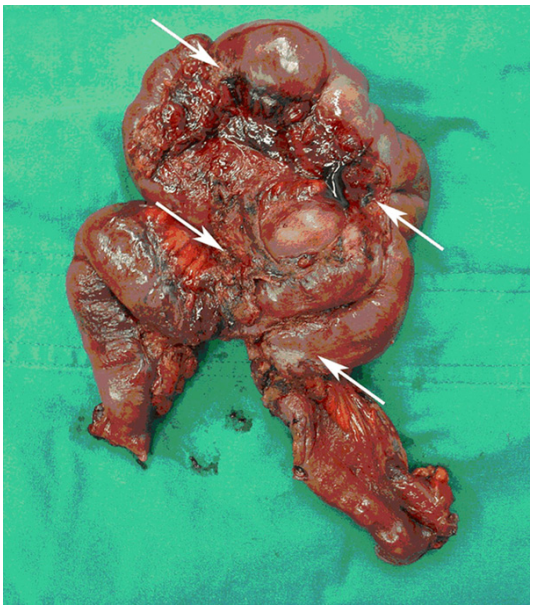

Figure 6. Second post operative Specimen showed multiple strictures in the small bowel mainly aggregated around the mesenteric segment (white arrow).

aggregated bowel mass, 1.8 meters proximal to ileocecal junction (**Figure 6**), with four to five stricture sites at the aggregated bowel mass and mesentery. Resection of the aggregated bowel mass and side to side anastomosis of the small bowel was done. The resected specimen was hard in consistency (**Figure 6**).

The postoperative period was still quite uneventful after the second surgery. The patient was started with liquid diet but whenever his diet was changed to semisolid food then he had complain of abdominal distension, nausea and sometimes vomiting. So he was again observed for some days with nasogastric decompression and parenteral nutrition. This treatment cycle went on for two times in one month of postoperative period. Finally after the treatment with corticosteroid in a tapering dose the patient did not have any abdominal complains. He was slowly able to have his normal diet.

Discussion

SFTs are rare mesenchymal neoplasms, which were described as a distinct pathologic entity in 1931 [5]. Originally SFTs were thought to be of mesothelium or submesothelium origin normally involving the pleura, but nowadays it has been reported in other extrapleural locations such as the lung, mediastinum, pericardium, mesentery, peritoneum, extraperitoneal spaces, nose, and paranasal sinuses as well [6-8]. Approximately around 800 cases of SFTs of pleural and extrapleural origin have been reported in literatures recently [2]. Around 160 cases appeared to be of intra-abdominals, which include liver, stomach, retroperitoneum, kidney, prostate and small bowel mesentery [9]. Approximately 30% of SFT arise in extrapleural locations [1]. To our knowledge, this would be another new case of SFT originating from small bowel mesentery reported till now, but might be a first case with eventful postoperative bowel obstruction [10-13].

SFT seems to be an asymptomatic mass which grows slowly, normally found in middle aged adults equally influencing men and women [14, 15] but it is also reported that most of the patients having peritoneal SFTs are males with mean age of 54 years [11]. The presence of clinical symptoms solely depends on the size and location of the tumor itself with systemic symptoms including hypoglycemia, arthralgia, osteoarthropathy and clubbing [16, 17]. In our case, the patient did not display symptoms of abdominal distension, nausea, vomiting or any other systemic symptoms.

According to Chun HJ and Kinoshita T, CT of SFTs appear as well-circumscribed, hypervas-

cular masses that may displace or exert pressure effects on neighboring organs such as the bowel, urinary bladder, vessels, and ureter. Central hypoenhancing or nonenhancing areas may be seen in the tumor, which represents necrosis or cystic changes. Calcification is rare and can be seen in large benign or malignant tumors [9, 18]. The CT appearance of our case was well-circumscribed mass measuring 9.2 cm × 9.0 cm × 7.7 cm in size with characteristic of delayed enhancement with unequal density distribution and hypo-attenuation on the pre-contrast. There was even extensive vascular communication around the tumor margin. CT impression was gastrointestinal stromal tumor (GIST). It is quite difficult for the radiologist to differentiate SFT and GIST [19]. Only the imaging is not sufficient to differentiate SFT from other intra-peritoneal benign and malignant pathologies [10]. Hence histology and immunohistochemistry are the best current diagnostic markers for SFT.

SFT was initially thought to be rare mesenchymal neoplasms of fibroblastic origin with controversial histogenesis [20]. Nevertheless recently pathologists have defined SFT as fibrous tumors originating from ubiquitous dendritic interstitial cells with a histological characteristic so-called “patternless pattern”, hemangiopericytoma like appearance of the spindle cells and with high vascularity [21, 22]. In our case the histological finding was revealed with spindle cell proliferation.

Immunohistochemically, it is seen that SFTs commonly show strong and diffuse staining for CD34, bcl2 and vimentin, while epithelial membrane antigen (EMA) and smooth muscle antibody (SMA) are occasionally expressed [14, 17, 23]. SFTs are rarely positive for S100 proteins, desmin, actin and cytokeratins on IHC analysis [14]. In our case Immunohistochemical staining showed diffuse strong reaction for CD99 (**Figure 4A**) and SMA (**Figure 4B**) as well as focal positivity for CD 34 (**Figure 4C**) whereas CD117, Desmin, S-100, Bcl-2 (**Figure 4D**) showed negative expression. Hasegawa et al. and Chilosi et al. reported that a majority of SFT at various sites showed strong expression of bcl-2 [24, 25]; however, the tumor in this case revealed negative expression. On the other hand, we were able to detect a reaction for CD99, which is consistent with Renshaw’s study [26]. As its pathological behavior is unpredictable it’s hard to correlate between morphology and outcome of SFTs [3]. Most of the SFTs are histopathologically benign with 20% rate of malignancy [27]. The malignant variant generally consists of a large tumor (>5 cm diameter) that is hypercellular and invasive, with nuclear pleomorphism and tissue necrosis with high mitotic index (more than 4 mitoses per 10 HPF) [22, 27].

Positive immunohistochemical staining mainly for CD34 has an important role in the differential diagnosis of SFT with other soft tissue tumors [13]. Recent advances in pathology have led to better understanding of the histogenesis, tumor distribution, and remarkable histological heterogeneity of SFTs [28]. Hence, according to Nomura et al. in order to exclude other $CD34^-$ positive tumors, the histological and immunohistochemical features are helpful, where the presence of immunoreactivity for CD34 and absence of immunoreactivity for CD 117, desmin, and S-100, strongly favors a definite diagnosis of SFT. Therefore surgery can be considered the treatment of choice for SFTs whenever possible. Regarding chemotherapy and radiotherapy, there has been no recent literature.

Michael Lau et al. and Sarah Bouhabel et al. [13] have presented with the similar case. They did an en-block resection of the related mesentery and bowel, but in our case only excision of the mass was done without intestinal resection which might be the cause of postoperative bowel obstruction with aggregated bowel and mesentery due to excessive ongoing proliferation of fibrotic tissues. So we might come in conclusion that, resection of the affected mesentery and related bowel may be the treatment of choice in large SFT of intestinal mesentery with malignant in nature. Regarding the conservative treatment after second operation, using steroid in a tapering dose may also be a choice [1].

In conclusion, we presented a rare case of SFT arising from the small bowel mesentery with eventful postoperative complication of acute and subacute intestinal obstruction. This lesion showed typical imaging features of GIST, but immunohistochemical analysis confirmed the diagnosis of SFT. The surgical en-block resection of the affected mesentery including the bowel might be the surgical indication for large SFT in small bowel mesentery.

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Disclosure of conflict of interest

None.

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