Solitary fibrous tumor arising from the mesentery of adult patients. Report of two cases and review of the literature

JOSÉ FERNANDO VAL-BERNAL¹, MARTA MAYORGA¹, FIDEL FERNÁNDEZ¹, ALEJANDRO PARRA¹, JUAN CRESPO², MANUEL GARCÍA-POLAVIEJA³

¹Department of Anatomical Pathology, Marqués de Valdecilla University Hospital, Medical Faculty, University of Cantabria and IFIMAV, Santander, Spain
²Department of Diagnostic Radiology, Marqués de Valdecilla University Hospital, Santander, Spain
³Service of General and Digestive Surgery, Marqués de Valdecilla University Hospital, Santander, Spain

Abstract
Solitary fibrous tumors (SFTs) represent an uncommon entity most frequently manifested in the pleura. We describe herein two new cases located in the jejunal and sigmoid mesentery incidentally found in patients aged 61 and 32 years. In a review of the literature, we have compiled 15 mesenteric SFTs including our two cases. The mean age of the patients at presentation was 51.7 years (range, 26–83 years). Most patients were males (males:females 4:1). Although occasionally these tumors were an incidental finding, the majority have been symptomatic. Tumors varied greatly in size (3 to 25 cm), but most of them were large (mean 14.8 cm). Most cases (60%) were located in the small intestine mesentery. The hemangiopericytomatosus (cellular) variant was the most common. All patients were treated by surgery and no other therapeutic approaches (chemo-/radiotherapy) were used. Follow-up data were available in 11 cases and ranged from six days to 21 years, with a mean follow-up period of 36.2 months. None recurred or metastasized. Two (13.3%) of the 15 cases showed atypical histological features concordant with histological, but not clinical malignancy. The main differential diagnosis includes gastrointestinal stromal tumor, synovial sarcoma and reactive nodular fibrous pseudotumor of the mesentery. In one third of the cases, tumor excision did not require intestinal resection. To our knowledge, our Case No. 1 is the first reported that has been removed through laparoscopic surgery. Radical surgery remains the treatment of choice. The unpredictable behavior of SFTs requires a careful, close, long-term follow-up.

Keywords: intestine, laparoscopic surgery, mesentery, solitary fibrous tumor.

Introduction
The term hemangiopericytoma has been gradually abandoned in favor of the term solitary fibrous tumor (SFT), so that most neoplasms that were called conventional (adult-type) hemangiopericytomas 20 years ago tend to be called SFTs now [1]. SFTs are localized, well-circumscribed neoplasms composed of a subset of fibroblast-like cells that can be observed in numerous locations including pleura, peritoneum, pericardium, mediastinum, retroperitoneum, upper respiratory tract, orbit, soft tissues, meninges and visceral organs. SFT is uncommon, but is the most common primary localized neoplasm of the pleura [2]. Nevertheless, these tumors are much less common in the peritoneal than in the pleural cavity [3]. On the other hand, SFTs in mesentery are rare. As far as we are aware, only 13 cases located in the mesentery of the adult have been previously reported [4–16]. We describe herein two new cases related to jejunal and sigmoid mesentery, and review the literature on the subject.

Patients, Methods and Results
Case No. 1
A 61-year-old man was referred to the surgical department for the incidental finding of a mesenteric mass on a control abdominal computed tomography (CT) scan. The patient did not complain of abdominal pain or any other gastrointestinal symptoms and he was in good general condition. Past medical history was significant for the diagnosis of a nevoid malignant melanoma on the left inferior extremity 17 months ago. The melanoma had a thickness of 1 mm and the sentinel node was free of tumor. The abdominal CT scan with contrast revealed a well-defined, solid, heterogeneous mass measuring 3 cm in diameter arising from the small bowel mesentery and situated among the distal jejunal loops (Figure 1A). A diagnosis was suggested of gastrointestinal stromal tumor or other primary soft tissue tumor of the mesentery.
The patient underwent laparoscopic removal of the mesenteric mass with an adjacent 4.5 cm jejunal segment. He had an uneventful recovery and was discharged eight days after surgery. He remained asymptomatic two months after surgical intervention.

The surgical specimen consisted of a 4.5 cm jejunal segment in whose mesenteric portion there was a nodular mass measuring 3 cm in greater diameter. The mass was well-defined, reddish with white (sclerosing) areas, and solid with scattered small cystic structures (Figure 1B). It had no attachment to the intestinal wall. The small intestine portion did not show abnormalities and the margins were clear.

Microscopically, the mass was completely encapsulated and separated from the intestinal muscle layer (Figure 2A). This neoplasm was composed of small, tightly packed, ovoid to spindled cells with an oval nucleus, inconspicuous nucleoli, and ill-defined eosinophilic cytoplasm. Tumor cell nuclei showed open chromatin, giving a vesicular appearance. A cystic change along the neoplasm was prominent (Figure 2B).

Tumor cells were arranged around numerous thin-walled ramifying blood vessels, which exhibited striking variation in caliber and occasional perivascular hyalinization (Figure 3A). Fibrosis and hyalinization of the stroma were focally present. In a small area of the tumor, the cells were disorganized showing atypia and pleomorphism (Figure 3B). Mitotic count was five mitoses per 50 high-power fields. There were scattered groups of lymphocytes along the neoplasm. Necrosis and hemorrhage were not present.

Figure 2 – (A) The neoplasm is completely encapsulated and separated of the jejunal wall (HE staining, 16×). (B) The neoplasm shows high cellular density and cystic change (HE staining, 25×).

Figure 3 – (A) Cells are randomly arranged, have uniform nuclei with open chromatin, inconspicuous nucleoli, and indistinct cell margins. There are numerous branching, angulated and dilated vessels (HE staining, 100×). (B) Focal presence of large (pleomorphic) atypical cells showing irregular nuclei with clumped chromatin (HE staining, 200×).

Case No. 2

A 32-year-old man was admitted to the hospital for evaluation of microscopic hematuria. An abdominal sonogram discovered a well-defined, large mass with homogeneous medium echotexture in the left iliac fossa. The barium enema was normal. Contrast-enhanced CT scans revealed a 13 cm, well-circumscribed mass with mixed attenuation in the left lower abdomen occupying the mesosigmoid space (Figure 5A). Midline laparotomy disclosed a large, fleshy, solid, well-defined, hypervascular tumor located in the mesosigmoid area not attached to the intestinal wall. A radical excision of the tumor with a 20 cm segment of the sigmoid colon was performed. The postoperative period was uneventful and the patient was discharged five days after surgery. He has remained symptom free for 21 years.

The surgical specimen consisted of a 20 cm sigmoid segment in whose mesenteric portion there was a well-circumscribed mass measuring 13×11.5×9 cm (Figure 5B). The mass was red-brown and solid. Areas of hemorrhage, cystic degeneration or necrosis were not seen. The tumor had no attachment to the intestinal wall (Figure 5C). The small intestine portion did not show abnormalities and the margins of the specimen were free of tumor.

The immunohistochemical study revealed strong diffuse positivity for CD34 (Figure 4A), CD99 (Figure 4B), and calponin in tumor cells. There were focal positivity for bcl2 and EMA; and negativity for CD117, DOG1, alpha-smooth muscle actin, desmin, CD31, D2-40, pancytokeratin, neuron specific enolase, S100 protein, Melan A, HMB45, collagen IV, and CD56. Ki67 (MIB1) labeled 4% of tumor cells.

Figure 4 – (A) Strong and diffuse positivity of tumor cells for CD34 (100×). (B) Strong and diffuse positivity of tumor cells for CD99 (100×).
Microscopically, tumor cells were uniform oval to fusiform with round to oval nuclei and poorly visible cytoplasmic borders. These cells were arranged around prominent anastomosing vessels of varying caliber lined by a single layer of flattened endothelial cells. Focal areas showing storiform and fascicular cellular patterns were seen. The larger vessels showed perivascular hyalinization. Fibrosing and hyaline areas were occasionally present in the neoplasm. Mitotic count was one mitosis per 50 high-power fields. Necrosis, hemorrhage, or cystic changes were not present.

The immunohistochemical study disclosed strong diffuse positivity for CD34, CD99, and bcl2 in tumor cells. There was focal positivity for calponin and alphasmooth muscle actin; and negativity for CD117, DOG1, desmin, CD31, pancytokeratin, and EMA. Isolated CD117+ mast cells were scattered along the neoplasm.

Discussion

The term mesentery refers to that part of the peritoneum that suspends the small intestine from the posterior wall of the abdomen. More loosely, the term mesentery may also refer to the mesocolon. This last unrestrained term is used here to designate a double layer of peritoneum attached to the posterior abdominal wall enclosing in its fold any portion of intestine (small or large), conveying to it its vessels and nerves.

Fibrous tumors and tumor-like conditions of the mesentery are rare and their clinical diagnosis is difficult. They include among other entities mesenteric fibromatosis, sclerosing mesenteritis, inflammatory pseudotumor, and solitary fibrous tumor. Knowledge of this group of diseases is important for accurate diagnosis and appropriate management [17].

In practice, hemangiopericytoma and SFT are the same entities whatever their location. The former is the cellular form of SFT [1]. SFTs have been reported in various extrathoracic sites, including the abdomen.

In our review of the literature, we have collected 15 mesenteric SFTs including our two cases (Table 1).

The mean age of these patients at presentation was 51.7 years (SD, 18; range, 26–83 years). Most patients were males (males:females 4:1). Although occasionally these tumors are an incidental finding, the majority have been symptomatic. Most frequent symptoms were abdominal discomfort or pain, distension and a palpable abdominal mass. No case with hypoglycemia or clubbing of the fingers has been reported. Nine (60%) cases were located in the small intestine mesentery.

On gross examination, these tumors varied greatly in size (3 to 25 cm), but most of them were large (mean 14.8 cm; SD 6.1). They were solid, well-circumscribed, partially or completely encapsulated, usually firm and white to tan or red colored. Tumors may have a variegated appearance with solid areas and areas showing cystic or honeycomb-like changes.

<table>
<thead>
<tr>
<th>No.</th>
<th>Reference</th>
<th>Age [years]/ Gender</th>
<th>Mesenteric location</th>
<th>Greater diameter [cm]</th>
<th>Histological malignancy</th>
<th>Treatment</th>
<th>Follow up [months]/ Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Pérez Cabañas et al., 1990 [4]</td>
<td>45/M</td>
<td>Rectum</td>
<td>9</td>
<td>No</td>
<td>Tumor and rectal resection.</td>
<td>NR/NR</td>
</tr>
<tr>
<td>4.</td>
<td>Prathima et al., 2003 [7]</td>
<td>38/F</td>
<td>Ileum and caecum</td>
<td>20</td>
<td>No</td>
<td>Tumor, ileum and caecum resection.</td>
<td>NR/NR</td>
</tr>
<tr>
<td>5.</td>
<td>West et al., 2004 [8]</td>
<td>61/M</td>
<td>Sigmoid</td>
<td>9.5</td>
<td>No</td>
<td>Tumor and sigmoid resection.</td>
<td>NR/NR</td>
</tr>
<tr>
<td>9.</td>
<td>Medina-Franco et al., 2011 [12]</td>
<td>60/F</td>
<td>Transverse colon</td>
<td>16</td>
<td>Yes</td>
<td>Tumor resection and transverse colectomy.</td>
<td>84/AW</td>
</tr>
</tbody>
</table>
Microscopically, they showed a wide range of morphological features, from predominantly fibrous lesions with marked variation in cellularity and hyalinized zones to monotonously highly cellular ones, which contained numerous thin-walled staghorn branching vessels. Elongated and dilated vessels may show thickened, hyalinized walls. However, in the mesentery the hemangiopericytomatosus (cellular) variant was the most frequent. Immunohistochemically, SFTs commonly showed strong and diffuse staining for CD34, CD99 and vimentin, and variable reactivity for bcl2. No positive staining for cytokeratin, or ALK-1 stains [26]. They are negative for CD34, S100 protein, CD117, alpha-smooth muscle actin or desmin and vimentin. Mononuclear cells can be present. The lesion expresses CD117, muscle-specific actin [27] and DOG1. Synovial sarcoma may occur in two main variants: biphasic and monophasic. The monophasic variant (MSS) commonly shows hemangiopericytoma-like areas throughout the entire neoplasm. However, the MSS frequently has a greater cellularity with a higher nuclear to cytoplasmic ratio, more mitotic figures and straighter cell alignments [25]. On the other hand, the expression of CD34 is absent in MSS and is typically seen in most cases of hemangio- pericytoma. In cases of unusual immunohistochemical results, the molecular detection of the SYT–SSX fusion gene transcripts will help in making a correct diagnosis. RNFPs show low to moderate cellularity and are composed of stellate or spindled fibroblasts arranged haphazardly or in intersecting fascicles. The stroma is rich in wire-like, keloidal or hyalinized collagen. Sparse intralymphoidal and peripheral (arranged in lymphoid aggregates) mononuclear cells can be present. The lesion expresses CD117, alpha-smooth muscle actin or desmin and vimentin. However, they are negative for CD34, S100 protein, cytokeratin, or ALK-1 stains [26].

In 33.3% of the cases, complete surgical excision of tumors did not include intestine (Table 1). As far as we are aware, the present Case No. 1 is the first reported that has been removed through laparoscopic surgery. Complete surgical resection of the SFT of the mesentery remains the only method of curative treatment. As late recurrence can occur in SFT [21] and because some tumors can behave aggressively even in absence of any primary morphologic evidence of malignancy, patients require careful, close, long-term follow-up.

SFT has also been reported in pediatric patients [28, 29]. These cases are not included in the current review.

Conclusions

SFTs arising in the mesentery are very uncommon. They are most commonly seen in male patients with a mean age of 52 years; and usually are symptomatic due to their large size. Most tumors are localized in the small intestine mesentery. Microscopically, the hemangiopericytomatosus (cellular) variant is the most frequent. Thirteen percent of the reported cases show histopathological signs of malignancy. Radical surgery remains the treatment of choice and the only opportunity of cure. Laparoscopic excision is feasible. A long-term follow-up period with clinical and imaging studies is mandatory because of the risk of late recurrence or metastasis.

References

Solitary fibrous tumor arising from the mesentery of adult patients. Report of two cases and review of the literature


Corresponding author
José Fernando Val-Bernal, MD, PhD, Professor of Pathology, Medical Faculty, University of Cantabria, Avda. Cardenal Herrera Oría s/n, ES–39011 Santander, Spain; Phone +34 942 202520 / ext. 73232, Fax +34 942 203492, e-mail: apavbj@humv.es

Received: June 28, 2013
Accepted: January 16, 2014