

## Solitary Fibrous Tumour of the Rectosigmoid Wall: Report of a Case and Review of the Literature

### Dear Editor,

Solitary fibrous tumours (SFTs) are rare, usually benign, mesenchymal tumours most commonly found in the pleura, where they were first described in 1931.<sup>1</sup> Subsequently, SFTs have been described in many other sites. To the best of our knowledge, SFTs arising from the colonic wall have not previously been reported. We report a case of an SFT arising within the wall of the rectosigmoid colon in a 53-year-old woman and discuss its differential diagnosis and clinical implications.

### Case Report

A 53-year-old female with a history of uterine fibroids was referred to the hospital after a pelvic ultrasound showed a mass in the posterior cul-de-sac extending toward the right adnexa. The mass was predominantly solid with cystic areas and demonstrated internal blood flow with low resistance, highly suspicious for an ovarian malignancy. However, the CA-125 was normal. A computed tomography (CT) scan demonstrated a complex mass associated with the right ovary with no extra-ovarian disease (Fig. 1A).



Fig. 1A. CT scan demonstrating the tumour, initially thought to be ovarian in origin.

The patient underwent an exploratory laparoscopy. Intraoperatively, both ovaries appeared normal. However, a well-encapsulated mass arising from the serosal surface of the rectosigmoid colon was noted (Fig. 1B). The mass filled the entire pelvis and was in close apposition to the right ovary; however, no attachment was noted between the mass and the right adnexa. Colonoscopy was only significant for a hyperplastic polyp. The Carcinoembryonic Antigen

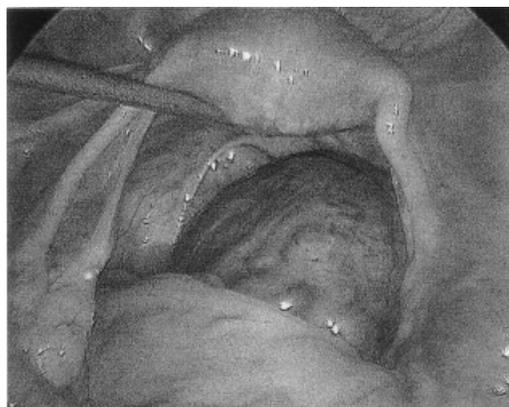


Fig. 1B. Intraoperative view of the tumour.

(CEA) level was 1.8. At laparotomy, a mass arising from the sigmoid colon was observed. No other disease in the gastrointestinal tract or liver was observed. As such, the intraoperative diagnosis was that of a soft tissue tumour of indeterminate nature and a low anterior resection was performed.

Grossly, the tumour was red and bullous, measuring 11.5 x 11.5 x 3.5 cm with a fleshy, tan cut surface with multiple, loculated, smooth-walled cysts. The mass was deep to the muscularis propria, but did not involve the mucosa or the serosa. Microscopic examination revealed a spindle cell neoplasm with a staghorn vessel pattern (Fig. 1C). The neoplastic cells were reactive with BCL2, CD34 and CD99; and they were not reactive with C-kit, epithelial membrane antigen and cytokeratin AE1/AE3. The mass was diagnosed as a solitary fibrous tumour (SFT). The tumour displayed necrosis (Fig. 1D) and marked cellularity (Fig. 1E) but brisk mitotic activity and pleomorphism were not noted.

### Discussion

SFTs are rare spindle cell neoplasms of mesenchymal origin most often found in the visceral pleura of adults between the fourth and seventh decades of life. First reported by Klemperer and Rabin<sup>1</sup> in 1931 as a localised fibrous mesothelioma in the lung pleura, SFTs have now been recognised in other serosal and non-serosal sites. In the pleura, SFTs classically present as well-defined, pedunculated and circumscribed masses.<sup>2</sup> SFTs occurring at extrapleural sites have similar histologic, immunohistochemical and ultrastructural characteristics

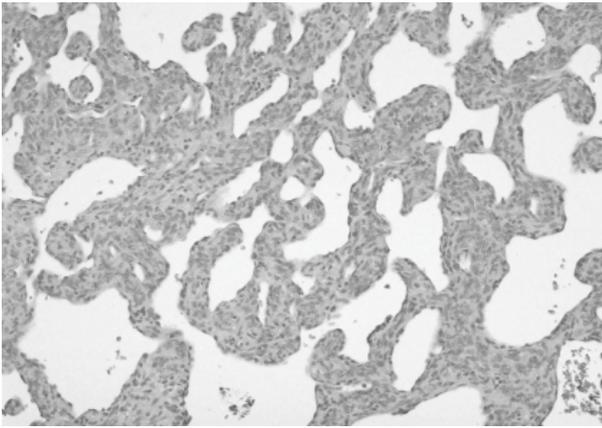


Fig. 1C. Staghorn vessel pattern seen in the SFT.

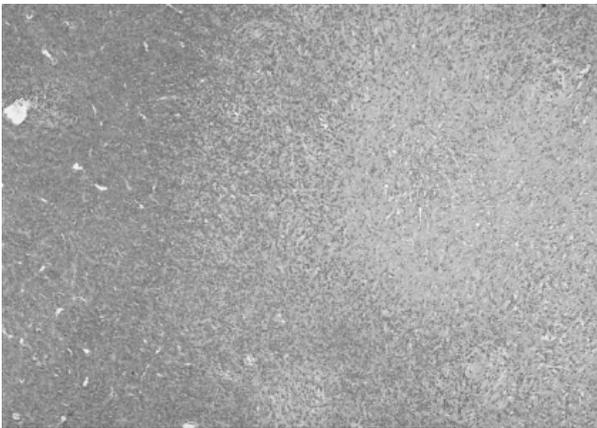


Fig. 1D. Areas of necrosis in the SFT.

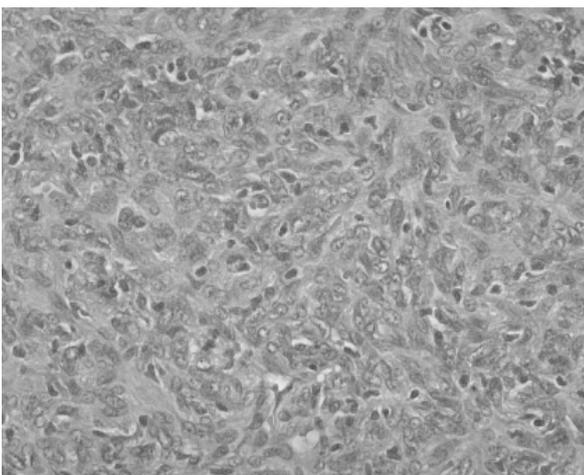


Fig. 1E. Areas of marked cellularity in the SFT.

as compared to their pleural counterparts.<sup>3</sup>

SFTs are diagnosed based on gross and histologic features, with immunohistochemistry used to support the diagnosis. Grossly, SFTs tend to be circumscribed tumours.<sup>2</sup> England et al<sup>4</sup> observed that 51% of tumours were gray-white. Only 13% of tumours had a cystic appearance like the case we report.<sup>4</sup> SFTs arising from serosal surfaces are usually exophytic masses, as in this case.

Histologically, SFTs are composed primarily of spindle cells with scant cytoplasm.<sup>3</sup> SFTs usually display admixtures of hypercellular and hypocellular areas. The cellular areas, made up of plump to spindle cells, may be arranged in a variety of patterns even within the same tumour.<sup>3</sup> The various patterns include palisading, fascicular, herringbone, diffuse sclerosing, storiform and hemangiopericytic patterns and the so-called patternless pattern.

A review by England et al<sup>4</sup> of 223 cases showed that amongst those with a single architectural pattern, the patternless pattern was the most frequent. The inconsistent histologic pattern of SFTs has led to much difficulty in diagnosis as they can resemble many other proliferative processes. Consequently, various types of benign and malignant spindle cell neoplasms should be considered in the differential diagnosis of an SFT.

Immunohistochemical analyses serve as an important aid in differentiating SFTs from other spindle cell neoplasms, aiding treatment decisions and prognostication. SFTs generally express CD34, BCL-2, CD99, vimentin and sometimes actin,<sup>3</sup> but they do not express cytokeratin and desmin.

For SFTs arising from the bowel wall, the main differential diagnosis includes gastrointestinal stromal tumours (GISTs), desmoid tumours and nerve sheath tumours.

As GISTs respond to the tyrosine kinase inhibitor imatinib mesylate, differentiating them from SFTs is clinically important. Although GISTs are stroma-poor tumours and SFTs tend to have more stroma, some tumours may have an overlapping histology. Both SFTs and GISTs are immunoreactive for CD34. However, immunoreactivity for CD 117 (c-kit) has been shown to be a sensitive and specific marker for GIST.<sup>5</sup>

Solitary desmoid tumours are frequently abdominal and may affect the bowel wall<sup>6</sup> but are more likely to show an infiltrative growth pattern and generally do not express CD34, CD99 or BCL-2.<sup>7</sup> More importantly, they are more likely to recur after surgical resection as compared to SFTs.

Peripheral nerve sheath tumours and smooth muscle tumours should be considered in the differential diagnosis of gastrointestinal SFTs. Schwannomas may express CD34 and BCL-2, but they are often encapsulated, associated with peripheral nerves and S100 protein positive, in contrast

to SFTs.<sup>3</sup> Malignant peripheral nerve sheath tumours may also be CD34 positive and may mimic malignant SFTs histologically, but they are S100 positive. CD34 immunoreactivity may also be seen in smooth muscle tumours; however, negativity for markers such as actin and desmin may be used to support the diagnosis of an SFT.<sup>3</sup>

Previously, SFTs were distinguished from soft tissue hemangiopericytomas. However, recent evidence suggests that the two are in fact the same tumour. They share similar morphology, ultrastructure and immunophenotype. In addition, immunohistochemical studies have failed to show pericytic differentiation in soft tissue hemangiopericytomas.<sup>8</sup>

SFTs are sporadic and usually asymptomatic but may become symptomatic when they become large or involve vital structures. However, there have been reports of solitary fibrous tumours causing hypoglycaemia secondary to secretion of high levels of insulin-like growth factor (IGF) II.<sup>9</sup> Although SFTs are usually benign, a small number of recurrent and malignant cases have been reported. There have also been rare reports of multifocal lesions.<sup>10</sup> Unfortunately, there is currently no good way to predict the clinical behaviour of SFTs.

The main treatment of SFT is surgery. Chemotherapy and radiotherapy have been used after incomplete resections or for recurrence. The role of adjuvant therapy is uncertain in complete resections. Because of the risk of recurrence decades after surgery and the difficulty in assessing malignant potential, long-term follow-up is obligatory.

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