

Case Report

Solitary fibrous tumour of mesentery: a rare tumour-case report and a comprehensive review of literature

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Received: 12 June 2024

Revised: 10 July 2024

Accepted: 12 July 2024

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ABSTRACT

Solitary fibrous tumours (SFTs) represent an infrequent clinical entity distinguished into pleural and extrapleural forms, with morphological resemblance. Literature exists on extrapleural forms of SFTs but few cases originating from small bowel mesentery have been reported. We report here one such case of a 64-year-old male presenting with abdominal pain and mass in the right lower quadrant of abdomen. Radiology suggested a neoplastic etiology arising from the jejunum which was surgically resected. Histopathology confirmed a spindle cell tumour originating from small bowel. Patient had an uneventful clinical course and was discharged after 1 week of surgery. We also reviewed the literature on SFTs arising from mesentery and found that only 24 cases have been reported worldwide in the past 35 years.

Keywords: Solitary fibrous tumour, Mesentery, Spindle cell tumour, Small bowel, Abdominal mass, Extrapleural

INTRODUCTION

Solitary fibrous tumours (SFTs) are rare spindle cell mesenchymal tumours that primarily arise from the thoracic pleura. Formerly known as hemangiopericytoma, they were first reported in 1931 by Klemperer and Rabin. These tumours are usually benign, localized, well-circumscribed, neoplasms consisting of fibroblast-like cells.¹ Although extrapleural SFTs are uncommon and have been reported arising from various sites, SFTs arising from mesentery are extremely rare. Such SFTs may be asymptomatic or may present late as painless palpable abdominal mass or pain in the abdomen if the tumour is pressing on adjacent viscera. CT and MRI can identify the tumour but cannot differentiate well between SFTs and other soft tissue tumours. The diagnosis of such extrapleural abdominal presentation of SFTs becomes challenging due to the aforementioned reasons. Here, we report a rare case of spindle cell tumour arising from jejunum and review literature on this subject.

CASE REPORT

A 64-year-old male, hypertensive and chronic alcoholic, presented to the outpatient clinic with an insidious, dull-aching, non-radiating pain in the right lower quadrant of the abdomen, and generalized weakness for 8-9 months which was associated with intermittent episodes of fever, loss of appetite and constipation. Abdominal examination revealed a lump (~6×6 cm) in right lumbar and right iliac region, which was mobile with irregular borders, mildly tender, not fixed to abdominal wall, non-pulsatile and overlying skin was normal. He was anemic with stable vitals and normal other systemic examinations. On CECT abdomen, ill-defined circumferential thickening of bowel was noted forming lobulated mass in right hemi-abdomen with air-fluid level and heterogenous post-contrast enhancement with fat stranding indicating a neoplastic etiology (Figure 1 A-C). Further investigation with PET-CT divulged ill-defined, hypermetabolic, circumferential mural thickening of jejunal loops of size 7.7×12.8×14.1 cm, not associated with any lymphadenopathy.

The patient underwent a diagnostic laparoscopy (Figure 2 A and B) followed by a midline laparotomy and a 25×22 cm soft, solid, proximal jejunal mesenteric tumour, densely adherent to the right-side colon, ileal loops and omentum was identified (Figure 3 A and B). Tumour was resected from root of mesentery after adhesiolysis, and 30 cm of jejunal segment, 10 cm from duodeno-jejunal flexure, was resected along with tumour and anastomosed side-to-side (Figure 4 A-C). Frozen section sent intra-op showed spindle cell neoplasm. Histopathology report suggested spindled to epithelioid cells arranged in fascicles and bundles, with vesicular chromatin and moderate eosinophilic cytoplasm, with focal areas of necrosis, cystic degeneration and neutrophilic infiltrate, not associated with mitosis/nuclear atypia.

Patient had uneventful clinical course and got discharged 1 week after the surgery. Patient was followed up for 6 months of duration in which he was clinically stable.

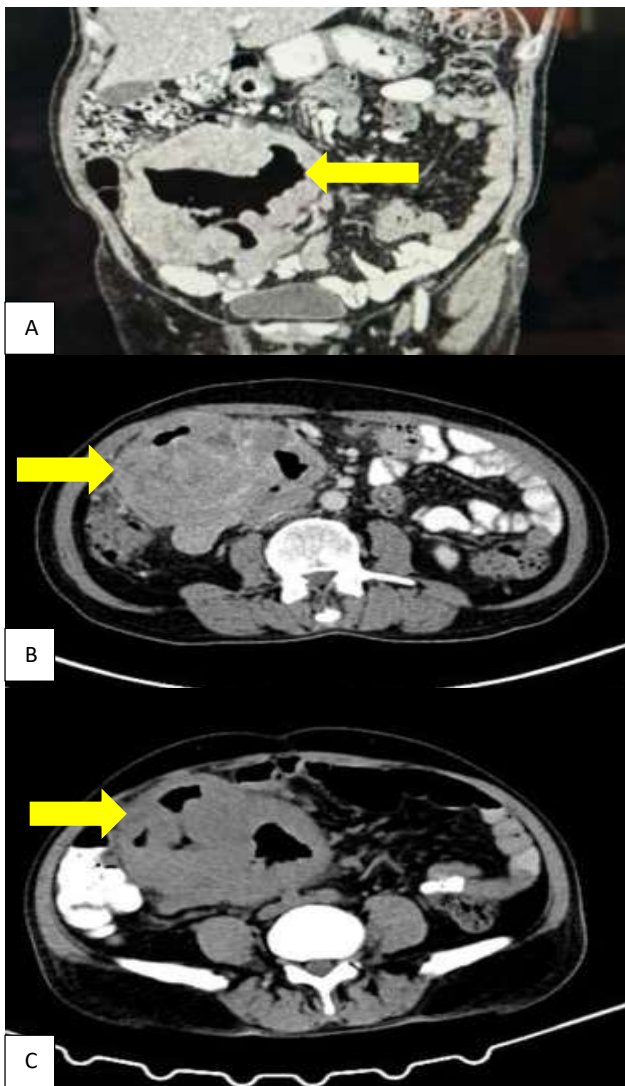


Figure1 (A-C): Abdominal CT scans revealing heterogeneously enhancing lobulated mass in right hemi-abdomen with circumferential wall thickening containing air fluid level.

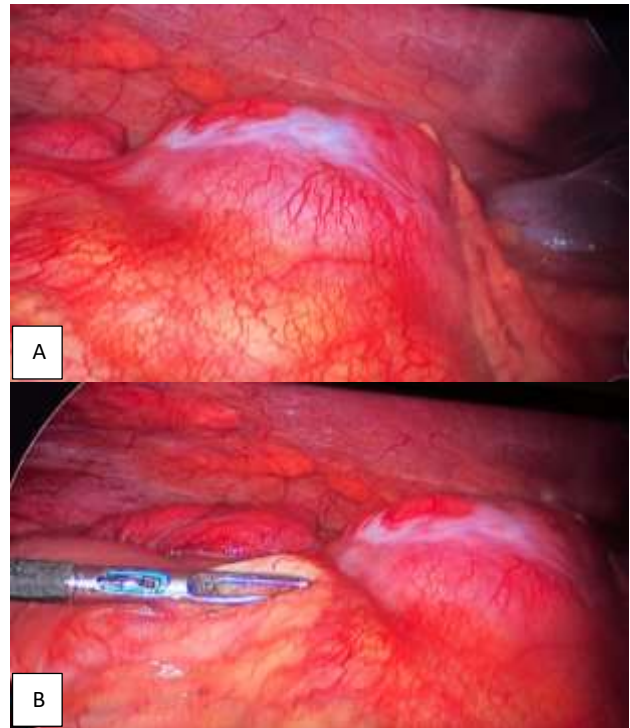


Figure 2 (A and B): Intraoperative laparoscopic view of tumor with visible serosal surface of the tumor in right lumbar region.

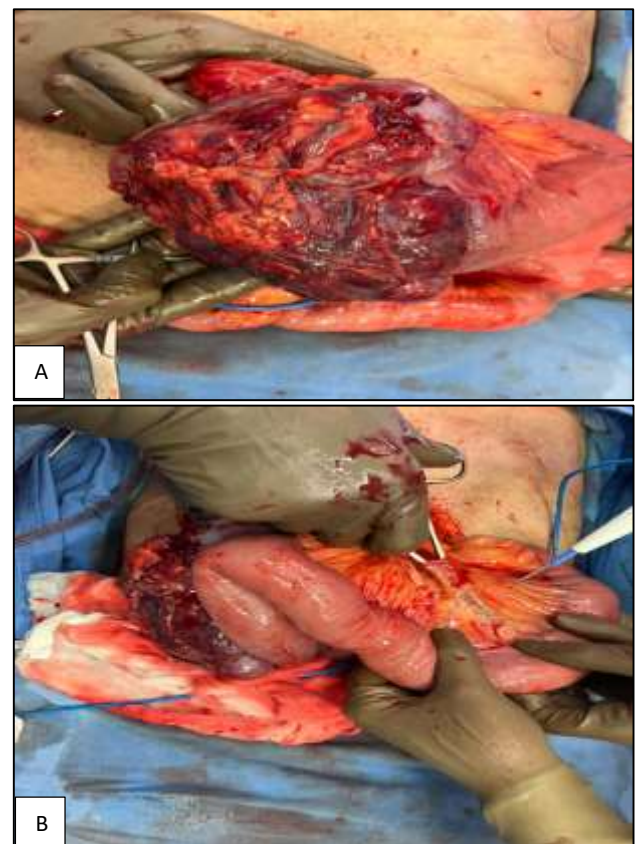


Figure 3 (A and B): Intraoperative image of mesenteric tumor densely adherent to jejunal loop with division of mesentery.

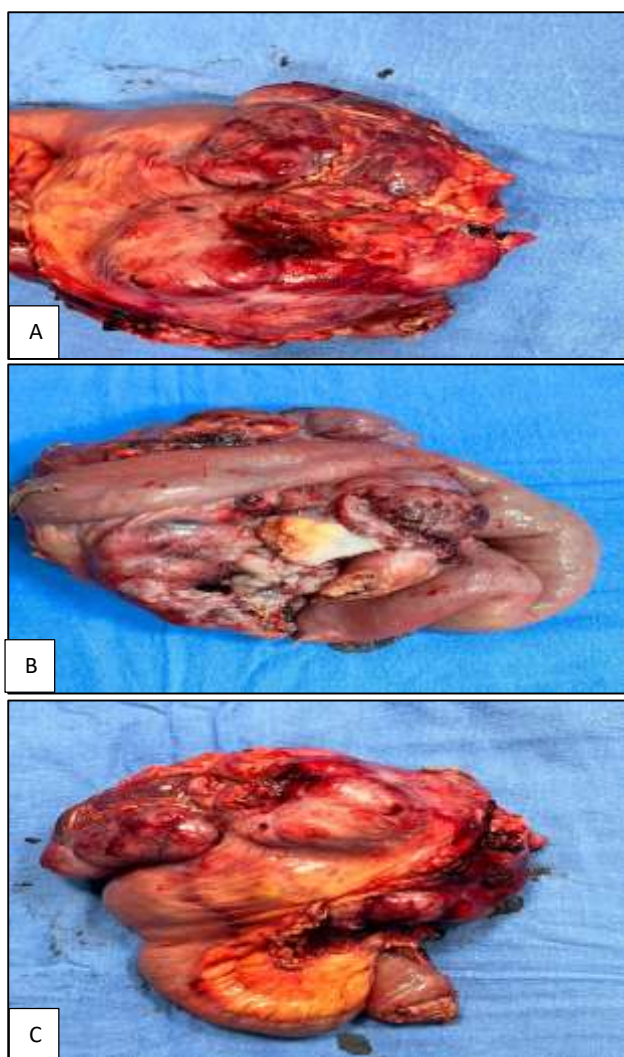


Figure 4 (A-C): Post operative resected specimen of mesenteric tumor along with involved adherent and jejunal loop.

DISCUSSION

First described by Klemperer and Rabin in 1931, a SFT is a rare mesenchymal spindle cell neoplasm believed to arise from pleural mesothelial tissue.¹ Though extrapleural locations such as pericardium, mesentery, peritoneum, extraperitoneal space, brain, spinal cord and paranasal sinus have been reported, these extra pleural occurrences often go unrecognized/misdiagnosed because of their rarity.² Mesenteric origin of such tumor is *recherché* and seems to occur in ileum, more commonly affecting males of age group 40-50 years according to existing literature; however, in our patient, it emanated from jejunum. Liu et al have reported a similar jejunal origin of this tumour.³ SFTs are usually steadily growing and present with little to no clinical symptoms unless it compresses adjacent tissues/structures nonetheless disastrous complications like torsion, hemorrhage/infarction may pose a threat.⁴ SFTs, generally, are round/lobulated soft tissue with clear boundaries, but in our case, tumour boundaries were not well-defined. Chiu

et al had a similar finding in their case.⁵ Most cases are sporadic, but they can be associated with Gardner syndrome, familial polyposis coli, bilateral ovarian fibromatosis, previous intra-abdominal surgery and estrogenic stimulation.

CT and MRI are effective modalities for diagnosing SFTs, but they might not suffice. On CT, these tumours usually appear round or lobulated with clear margins, heterogeneously enhancing, multiloculated mass with/without calcification or necrosis. On both T1 and T2-weighted MRI, they seem heterogeneous, with solid areas appearing iso to hypointense relative to skeletal muscle and cystic areas appearing hyperintense on T2 sequence. Histopathologically, spindle cells are abundant in SFTs with “pattern-less” growth, benign in nature, although 20% can be malignant. IHC reveals CD34, Bcl2, vimentin and STAT6 positivity as specific makers for SFTs. Differentials for SFTs might be desmoid tumors, (GIST) leiomyomas, leiomyosarcomas, soft tissue sarcomas, inflammatory pseudotumor, sclerosing mesenteritis, mesenteric panniculitis and metastases. SFTs are classified as benign/malignant according to high mitotic activity >4 mitoses per 10 high-power fields, high cellularity, necrosis, and pleomorphism. Tumours are considered malignant if >1 criterion was present and these factors are positively correlated with unfavourable clinical outcomes.⁶

The definitive treatment is surgical resection of the tumour; however, a few literature reports have exhibited radiotherapy as an effective therapeutic modality for reducing tumour size.^{6,7} Cytotoxic chemotherapy, anti-estrogenic drugs and novel targeted therapy might exert some effect on SFTs.⁸ Owing to its tendency towards involving adjacent visceral abdominal structures, wide local excision of the tumour and involved structures is advocated, as was followed in our case, too. Burke et al reported a strong association between patients with Gardner syndrome and recurrence and a 23% recurrence rate in all mesenteric SFTs.¹⁰ Unpredictable behaviour of SFTs requires a careful, close, long-term follow-up.

While reviewing literature, we compiled cases reported worldwide and found that only 24 cases, including ours, have been reported in the past 35 years. Mean age of patients was 54 years (range 26-83 years), predominantly males (male:female 4:1). Most of the patients were diagnosed either incidentally or had a painless mass in abdomen. Tumour size varied from 3-25 cm with most patients having a mass greater than 10 cm. Nearly 60% of tumours were located in small bowel, predominantly in ileum. All patients were treated with surgical removal of tumour and 2/3rd of patients required intestinal resection. Only 1 case underwent laparoscopic removal of tumour, while all others had a midline laparotomy. Recurrence was seen in 1 case; 3 cases were malignant and metastasis was observed in none. Radio-chemotherapy was instituted in 2 cases, and cytoreductive surgery was done in 1 case, which was a recurrent tumour.

Table 1: Reports of SFTs of mesentery in literature.

References	Age/ gender (in years)	Symptoms	Location	Tumour size	Treatment	Follow up (months)	Outcome
Our case	64/M	Pain in abdomen	Jejunum	25×22 cm	Tumor resection and bowel resection anastomosis	6	Alive and well
Chiu et al⁵	69/M	Incidental finding	Ileum, colon and urinary bladder	Multiple nodules	Cytoreductive surgery	2 years	Alive and recurrence of tumor
Liu et al³	66/M	Abdominal pain	Jejunum	10×7×8 cm	Tumor resection and partial omentectomy	4	Alive and well
Kesar et al⁴	35/M	Painless, palpable mass	Ascending mesocolon	9.6×8.5×5 cm	Tumor resection and partial omentectomy	2	Alive and well
Zhang et al¹¹	41/M	Painless, palpable mass	Distal ileum	9×9×7 cm	Tumor resection with resection anastomosis of distal ileum	Not reported	Not reported
Nishida et al¹²	65/M	Abdominal pain	Distal ileum	25×13×10 cm	Tumor resection and distal ileum resection anastomosis	Not reported	Not reported
Val- Bernal et al¹³	61/M	Incidental finding	Small bowel	3 cm	Laparoscopic tumor resection	Not reported	Not reported
Val- Bernal et al¹³	32/M	Incidental finding	Mesosigmoid colon	13×11×9 cm	Radical tumour excision with sigmoidectomy	Not reported	Not reported
Chiroque et al¹⁴	82/M	Abdominal pain	Distal ileum	13×5×5 cm (malignant)	Tumour resection and distal ileum resection anastomosis	1	Alive and well
Nishikawa et al¹⁵	36/M	Constipation and increased urinary frequency	Rectum	15×8×7 cm	Radiotherapy followed by laparoscopic tumour resection	26	Alive and well
Cantarella et al¹⁶	26/M	Increased urinary frequency	Proximal ileum	12 cm	Tumour resection with resection anastomosis of ileum	18	Alive and well
Muroni et al¹⁷	73/M	Abdominal pain	Small bowel	24×23 cm	Tumour resection	Not reported	Not reported
Kudva et al¹⁸	41/M	Mass in abdomen	Small bowel	23×20×9 cm	Tumour resection	7	Alive and well
Bouhabel et al¹⁹	71/M	Painless palpable mass	Small bowel	15×14×9 cm	Tumour resection and bowel resection anastomosis	12	Alive and well
Medina-Franco et al²⁰	60/F	Pain in abdomen	Transverse colon	16 cm (malignant)	Tumour resection and bowel resection anastomosis	84	Alive and well
Lau et al²¹	53/M	Painless palpable mass	Ileum	22 cm	Tumour resection and right hemicolectomy	1	Alive and well
Soda et al²²	27/F	Pelvic pain	Rectum	16 cm	Tumour resection with aortic balloon occlusion	12	Alive and well
Balaji et al²³	68/M	Painless mass	Sigmoid	18×15 cm (malignant)	Tumour resection	Not reported	Not reported
Ben-fadhal et al²⁴	30/F	Pain in abdomen	Small bowel	12 cm	Tumour resection	5	Alive and well
West et al²⁵	61/M	Painless mass	Sigmoid	10 cm	Tumour and sigmoid resection	Not reported	Not reported
Prathima et al²⁶	38/F	Painless mass	Ileum and caecum	20 cm	Tumour with ileum and caecum resection	Not reported	Not reported
Nakagawa et al²⁷	83/M	Painless mass	Sigmoid	16 cm	Tumour and sigmoid resection	11	Alive and well
Hardisson et al²⁸	33/M	Pain in abdomen	Small bowel	8 cm	Tumour resection	6	Alive and well
Perez et al²⁹	45/M	Pelvic pain	Rectum	9 cm	Tumour and rectal resection	Not reported	Not reported

CONCLUSION

SFTs rarely arise from mesentery and are usually benign and asymptomatic which makes its diagnosis difficult. Although CT and MRI may help in identifying the tumour, histopathology and immunohistochemistry provide an exact diagnosis. Surgical resection of the tumour and adjacent involved viscera is the definitive treatment for the condition. Chemo-radiotherapy might help in the shrinkage of tumours to some extent. These tumours can recur; therefore, long-term follow-up is essential.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Sharma A, Singh I, Wig JD, Verma SP, Basude AK. Solitary fibrous tumour of mesentery: a rare tumour-case report and a comprehensive review of literature. *Int Surg J* 2024;11:1392-7.