ISSN: 0976-3031

Available Online at http://www.recentscientific.com

CODEN: IJRSFP (USA)

International Journal of Recent Scientific Research
Vol. 15, Issue, 06, pp.4801-4805, June, 2024

International Journal of Recent Scientific Research

DOI: 10.24327/IJRSR

CASE REPORT

A RARE CASE OF RECURRENT SOLITARY FIBROUS TUMOUR OF THE MESENTERY

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DOI: http://dx.doi.org/10.24327/ijrsr.20241506.0901

ARTICLE INFO

Article History:

Received 14th May, 2024
Received in revised form 28th May, 2024
Accepted 12th June, 2024
Published online 28th June, 2024

Keywords:

Solitary fibrous tumour(SFT), mesentery, abdominal mass, computed tomography(CT), ultrasound(USG).

ABSTRACT

Solitary fibrous tumours are rare mesenchymal growths typically found in the pleura, although they can also originate from the mesentery, albeit rarely. Here, we present a case of a 38-year-old individual who presented with a mass in the left hypochondrium since 12 months. The patient occasionally experienced abdominal discomfort and pain. Upon physical examination, an well-defined hard mass was identified, leading to USG and computed tomograph investigations, which revealed a fibrous tumour. In September 2022 patient underwent surgery. Subsequent biopsy results identified the mass as a solitary fibrous tumour originating from the mesentery of the small intestine. Following surgery, the patient underwent regular monitoring without experiencing any symptoms. However, a recurrence of the solitary fibrous tumour was detected in June 2023 based on CT imaging.

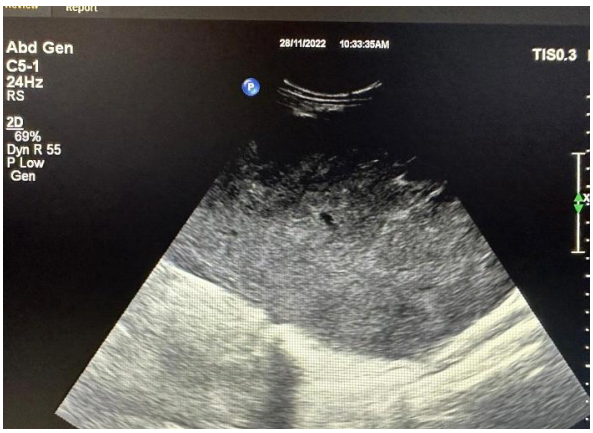
INTRODUCTION

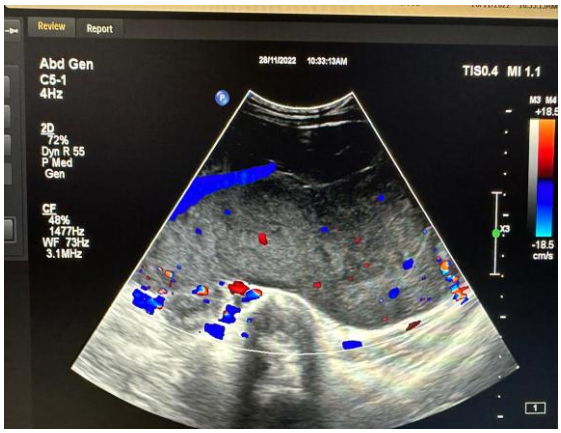
Solitary fibrous tumours (SFTs) represent a rare subset of spindle cell mesenchymal tumours primarily found in the thoracic pleura. These tumours, stemming from the mesenchyme, are distinguished by their notable fibrous composition and heightened vascularity. While instances of abdominal SFTs exist, involvement of the mesentery is infrequent. Typically benign, SFTs often go unnoticed unless they exert pressure on neighbouring tissues, complicating detection. Both computed tomography (CT) and magnetic resonance imaging (MRI) can aid in diagnosis, yet definitive confirmation relies on immunohistopathology, utilizing STAT6 and CD34 as specific markers.

Macroscopically, SFTs are commonly well-defined, encapsulated masses, presenting with a tan-white, firm, multinodular appearance, sometimes displaying haemorrhagic and myxoid alterations. Malignant transformations and tissue invasion can occur, underscoring the importance of complete surgical excision with clear margins. Given the potential for recurrence, even decades post-treatment, extended surveillance is advisable.

CASE REPORT

A 38-year-old male patient came with the complaints of progressive swelling in the left side of abdomen and intermittent pain since 12 months. He underwent ultrasound





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scan suggested by the clinician which revealed large heterogenous hypoechoic mass in left lumbar region measuring 11x10x9cms taking mild flow on application of colour doppler. However there is no evidence of calcifications. Possibilities of GIST and Desmoid were considered and patient was advised for computed tomography.

On computed tomography

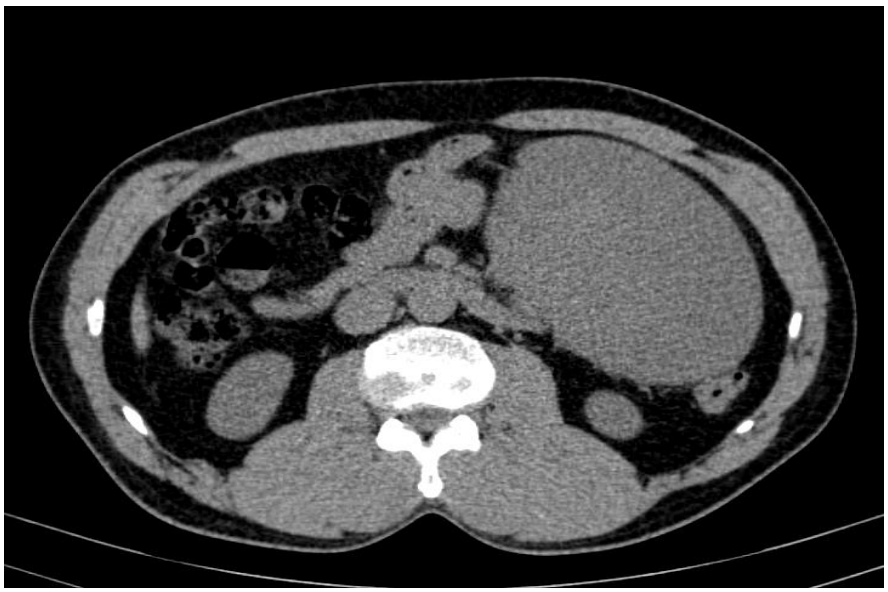
A large homogenous soft tissue ovoid mass with smooth margins in left lumbar region extending partially into left iliac fossa and left hypochondrium measuring approximately 11.5x10.6x9.7cms (APxMLxCC).

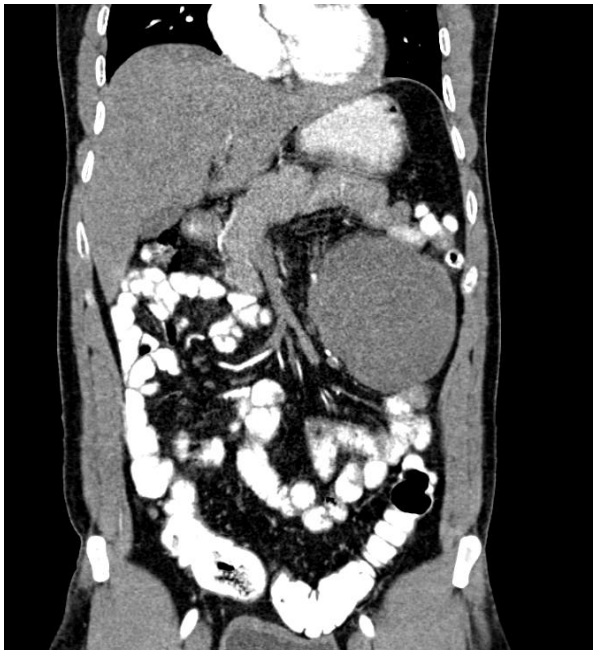
On post contrast

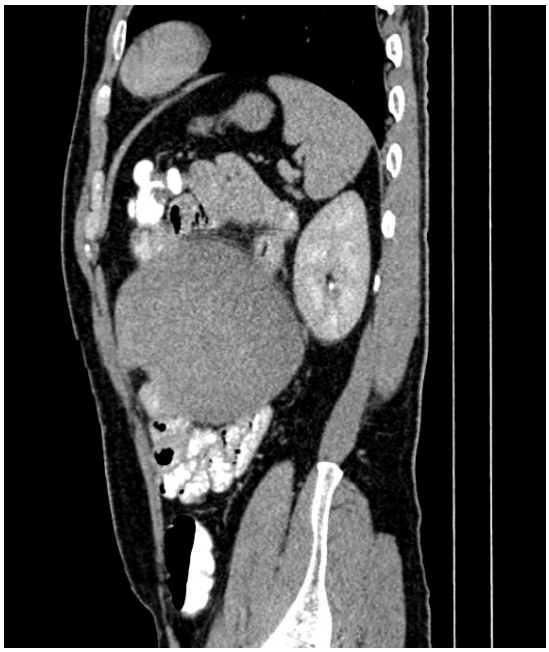
The mass shows multiple arterial feeders within the mass but the mass shows very poor enhancement in arterial phase, minimal heterogenous enhancement on venous phase, the mass shows delayed excretion of retained contrast with hyperdense appearance on 5 minutes delayed scan with CT attenuation value of 80-85 HU.

Arterial feeders from SMA and venous drainage into SMV noted.
No invasion into adjacent structures

Above imaging features were suggestive of **Solitary fibrous tumor.**







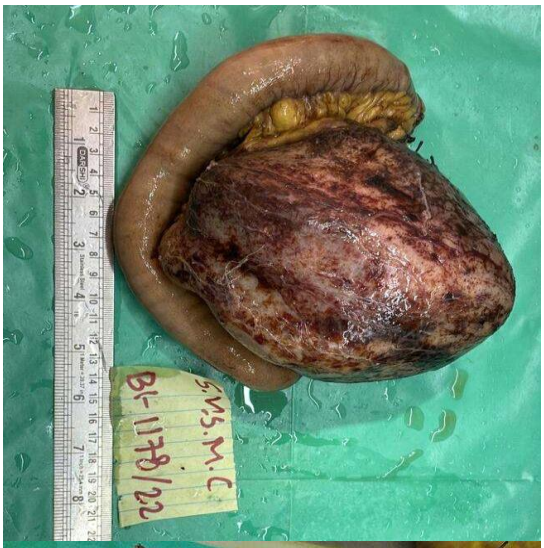
The mass displaces descending colon postero-laterally, small bowel loops medially and inferiorly.

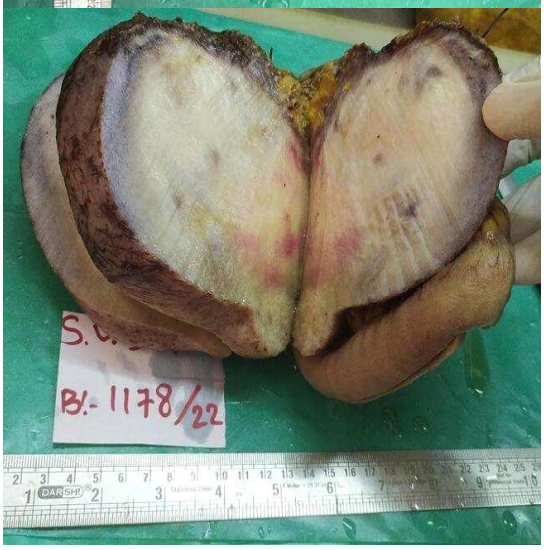
The mass abuts tail of pancreas medially with maintained fat planes.

Anteriorly the mass abuts anterior abdominal wall with maintained fat planes.

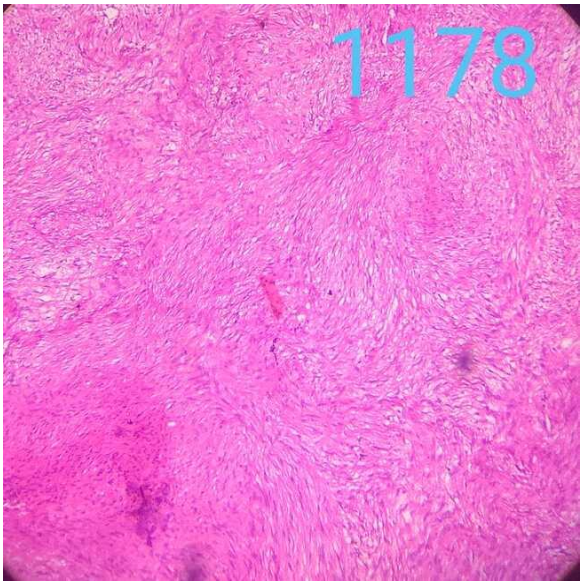
Histopathological Findings

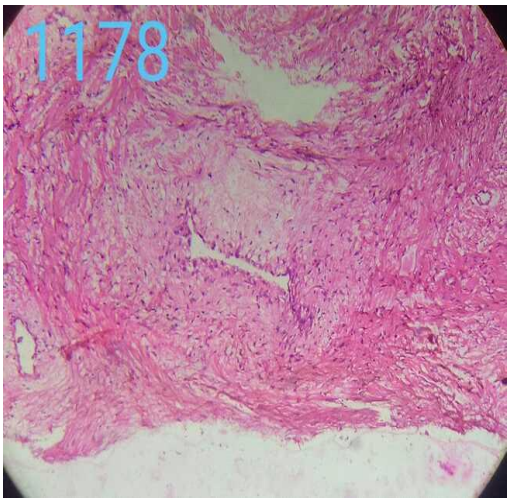
Gross Examination: Received intestinal specimen measuring 16cms, along with grey brown mass in the mesentery measuring 13x10x8cms and grossly showing congestion. Cut section of intestinal segment shows no loss of rugae, no strictures, no ulcers, overall normal. Mass cut section shows-homogenous grey white to focal grey brown areas.





MICROSCOPIC: Sections studied show spindle cell lesion. Cellularity is variable with hypo and hypercellular areas. Focal myxoid areas noted. Cellular areas show cells arranged in random and pattern less fashion. Vague areas of storiform pattern noted. Fibrous areas show thick collagen bundles at places organized as amianthoid fibers. Prominent blood vessels noted surrounded by spindle cells. Mast cells noted in stroma. Hemangiopericytomas pattern noted. No evidence of malignancy**.**

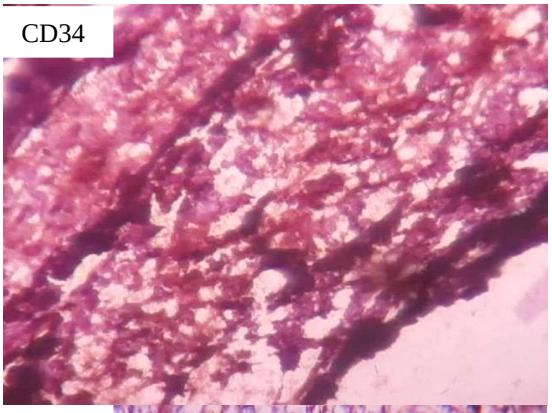




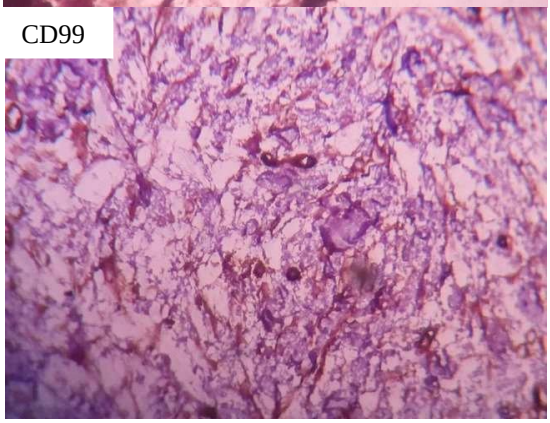
IMMUNOHISTOCHEMISTRY

CD34 and CD99 moderate positivity are in favor of solitary fibrous tumor.

CD34

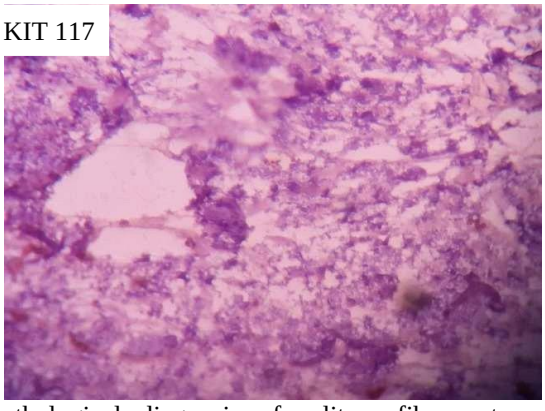


CD99



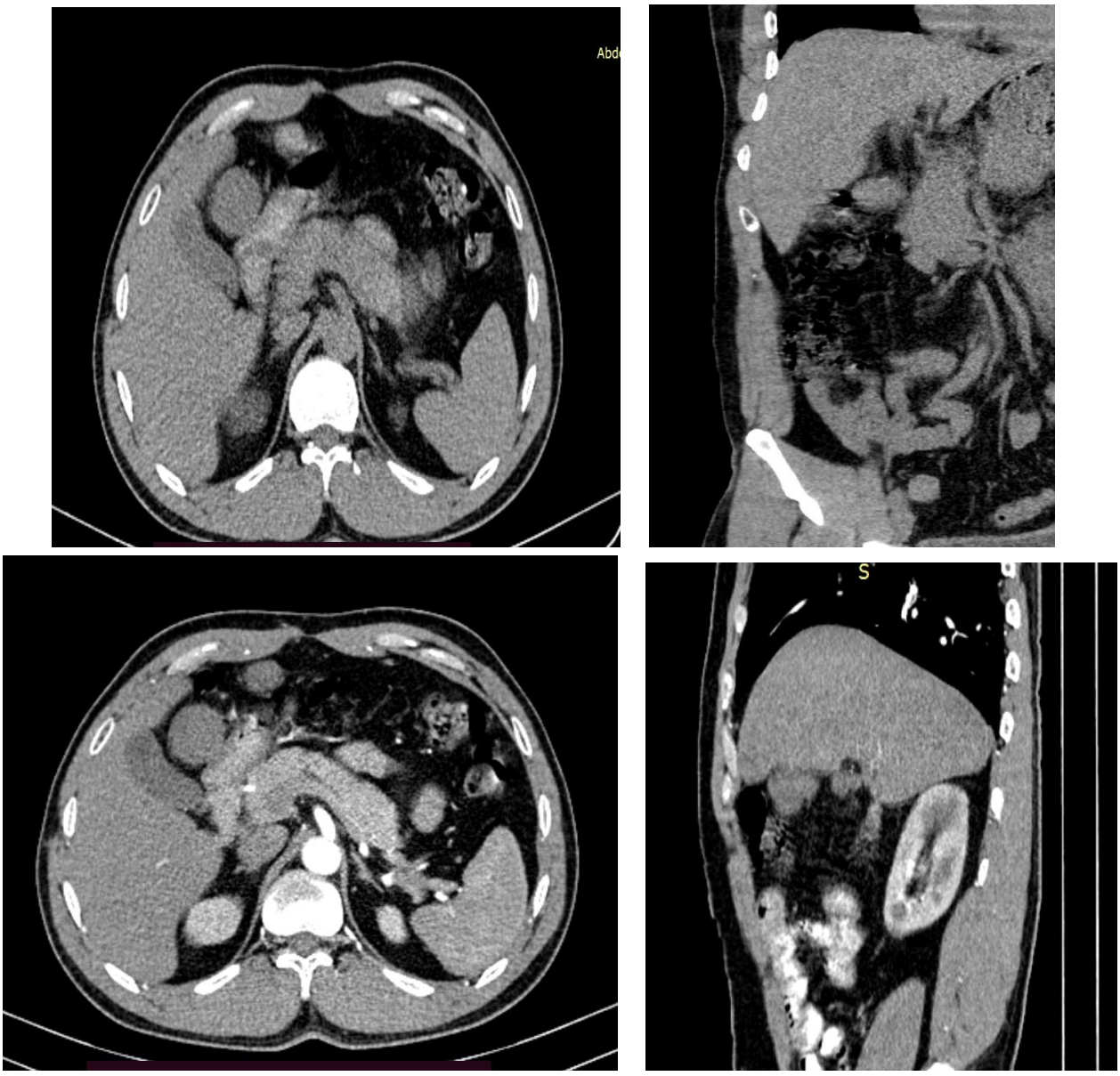
Kit 117 NEGATIVE was done to rule out GIST.

KIT 117



Histopathological diagnosis of solitary fibrous tumor was made.

FOLLOW UP SCAN AFTER 9 MONTHS



Two small soft tissue mass lesions noted in the sub hepatic region, largest measuring approximately 2.5x2.1x1.8 cm, which are radiologically similar to the previously discussed lesion.

DISCUSSION

Solitary fibrous tumors (SFTs) are rare tumors arising from mesenchymal cells, predominantly found in the pleura. However, they can also manifest in other areas such as the paranasal sinuses, pericardium, lung, mediastinum, peritoneum, extraperitoneal spaces and mesentery. When they develop in the peritoneum or mesentery, they are termed extra pleural SFTs or peritoneal SFTs. Peritoneal SFTs tend to affect males slightly more often, typically appearing around the age of 54. The clinical presentation varies depending on the tumor's size and its specific location.

The exact causes and mechanisms underlying solitary fibrous tumors (SFTs) remain uncertain, although there are theories proposing complex genomic alterations. Histologically, SFTs present as well-defined masses characterized by a collagen-rich matrix hosting arrays of spindle cells with varied histological appearances. A prevalent pattern observed in SFTs resembles hemangiopericytoma, while occurrences of mitoses and necrosis are infrequent.

Immunohistochemically, Solitary fibrous tumors display robust and widespread staining for CD34, bcl-2, and vimentin, with occasional expression of epithelial membrane antigen (EMA) and smooth muscle actin (SMA). SFTs rarely exhibit positive results for S100 proteins, desmin, actin, and cytokeratin. The pathological behavior of SFTs is unpredictable, given the unclear correlation between morphology and clinical outcomes.

Histopathologically most solitary fibrous tumors (SFTs) are benign, approximately 20% demonstrate malignant characteristics. Interestingly, the morphological appearance of an SFT does not always correlate with its clinical behavior; a tumor labeled as "malignant" may exhibit benign behavior, and vice versa. Malignant SFTs typically manifest as large, hypercellular, invasive masses, exceeding 50 mm in diameter, tissue necrosis , nuclear pleomorphism and a high mitotic index, typically exceeding 4 mitoses per 10 high-power fields (HPF).

All SFTs possess the potential to undergo malignant transformation, highlighting the importance of gross tumor examination and mitotic count in prognostic assessment.

Drawing parallels with the risk stratification scheme used for gastrointestinal stromal tumors (GISTs), which considers tumor size and mitotic count, a similar consensus-based approach could be adopted for SFTs. The underlying principle is that smaller tumors with lower mitotic indices generally carry a more favorable prognosis compared to larger tumors with higher mitotic indices.

Tumor size has emerged as a notable independent prognostic indicator, with a reported 5-year survival rate of merely 20% for patients harboring SFTs larger than 10 cm. While imaging techniques such as ultrasonography, CT, and MRI have been documented in only a handful of cases within the literature, CT scans typically depict SFTs as smooth, lobulated masses occasionally exhibiting calcifications. In our specific case, CT imaging unveiled a well-defined mass measuring 11.5 x 10.6 x 9.7 cm. Given the relative scarcity of SFT occurrences, arriving at a pre-operative imaging diagnosis poses challenges, necessitating consideration of more prevalent neoplasms in the differential diagnosis, such as neurogenic tumors, malignant fibrous histiocytoma, sarcomas, and desmoid tumors particularly, when SFTs localize within the abdominal and pelvic regions. Furthermore, GISTs may present morphological similarities to SFTs, with larger GISTs (>5 cm) often displaying exophytic growth patterns and occasionally featuring necrosis and calcifications. Nevertheless, discerning features on CT scans are limited. Positive immunohistochemical staining for CD34 serves as a pivotal diagnostic marker for SFTs, although this marker is also encountered in other tumor types. Approximately 75% of GISTs express CD34, with 95% demonstrating positivity for CD117. Contrarily, SFTs characteristically exhibit positivity for CD34 and CD99 and negative for CD117. Consequently, negative staining with the CD117 antibody emerges as the primary discriminator between SFTs and GISTs.

Table below shows Immunohistochemical patterns of SFT, leiomyosarcoma and GIST of the gastrointestinal tract

Tumor	CD34	KIT117
SFT	++	--
GIST	+	++
LEIOMYOSCARCOMA	-	-

CONCLUSION

We have reported an uncommon occurrence of an SFT originating from the mesentery of the small intestine. On CT imaging, this lesion resembled SFT w
hich was cofirmed on biopsy. Gross examination suggested that the SFT was likely benign. Therefore, it's imperative to include SFT in the list of potential diagnoses for any mesenchymal lesion arising from the gastrointestinal tract.

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How to cite this article:

M. Rithvik Reddy.,Y. Sai Rahul.,SubhashReddy Doni.,G. Ramakrishna Reddy and K. Venkat Ram Reddy. (2024). A Rare Case of Recurrent Solitary Fibrous Tumour of the Mesentery. *Int J Recent Sci Res.*15(06), pp.4801-4805.

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