

Solitary Fibrous Tumor of Rectus Sheath: A Rare Case Report.

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Received: March 2017

Accepted: March 2017

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ABSTRACT

Solitary fibrous tumor represents a spectrum of mesenchymal tumors encompassing tumors previously termed haemangiopericytoma, which are classified as having intermediate biological potential in the 2002 WHO classification scheme. The most common site of occurrence is pleura. Abdominal localization is quite rare. We report a case of extrapleural site of occurrence of solitary fibrous tumor in the posterior rectus sheath of a 45 year old male patient. The patient presented with a hypogastric midline swelling since one month. Intraoperative finding revealed a mass attached to the posterior rectus sheath. Histopathology revealed oval to spindle tumor cells having round to oval vesicular nuclei, nuclear pleomorphism with moderate mitotic activity; morphologically mimicking Extragastrointestinal stromal tumor. Immunohistochemical staining and FISH studies were performed. The results of IHC staining are given in Table 1 and in figure 3. The term Solitary fibrous tumor is favored by soft tissue pathologist to describe a rare heterogeneous group of benign & malignant neoplasm along a morphologic continuum. We have described a rare case of malignant solitary fibrous tumor of rectus sheath.

Keywords: Extrapleural, Fibrous tumor, IHC, Rectus sheath.

INTRODUCTION

Solitary Fibrous tumor represents a single spectrum of mesenchymal tumors of which haemangiopericytoma is now considered a cellular phenotypic variant. This tumor presumed to be a fibroblastic differentiation; most commonly affects adults and can occur at any site. Solitary fibrous tumors are categorized as intermediate biological potential with a low risk of metastasis & relatively indolent course under the 2002 WHO classification.^[1] Solitary fibrous tumor was first reported by Klemperer and Rabin in 1931 as a primary neoplasm of the pleura.^[2] By 2008, approximately 820 cases of SFT had been reported worldwide, which occurs commonly in pleura, but also rarely in extrapleural site.^[3] Extrapleural SFT account 0.6% of all soft tissue tumors. SFT of abdominal wall are extremely rare with only 16 cases till date.^[4]

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As per latest WHO classification 2013 the term “haemangiopericytoma” is abandoned. It is now

used only to describe a morphological pattern that is shared by different entities. Currently, solitary fibrous tumor, haemangiopericytoma, lipomatous haemangiopericytoma, and giant cell angiofibroma are all lumped under the category of “extrapleural SFT”.^[3-6]

CASE REPORT

A 45 year old male presented to the surgery outpatient department with lump in hypogastrium since one month & pain in hypogastrium since 15 days. There was no history of fever, vomiting, decreased appetite & weight loss. Bowel & bladder habits were normal. On physical examination a spherical mass measuring 9cm X 7 cm was present in the midline of hypogastric region, mobile more freely in horizontal axis than vertical axis. The mass was firm & non-tender. Routine haemogram & urine examination appeared normal.

Intraoperative finding – The lump was found to be attached to posterior rectus sheath. There were dilated tortuous veins over the parietal surface of the mass. After removal of the tumor, it was found to have a well formed capsule, globular, measuring 10cmx9cmx6cm. Cut section was grayish white, firm with lobulated appearance, mostly solid with few cystic spaces along with haemorrhage & necrosis. Microsections showed hypercellular areas of biphasic pattern – mostly spindled shaped cells

intermingled with few epithelioid nodules having round polygonal cells. The cells having spindle shaped hyperchromatic nuclei with scant cytoplasm (fibroblastic appearance) were separated by parallel arrays of dense collagen. There were gaping staghorn vessels suggesting pericytic vascular pattern with perivascular hyalinization. Some areas showed patternless pattern consisting of small fusiform cells randomly arrayed between collagen bundles & giant cells. There was moderate to marked nuclear atypia & infiltrative borders. Mitotic figure were 5/10HPF with focal areas of haemorrhage & necrosis. Hence a diagnosis of malignant solitary fibrous tumor was made.



Figure 1: Gross Globular tissue mass & cut surface - solid areas, hemorrhage & necrosis.

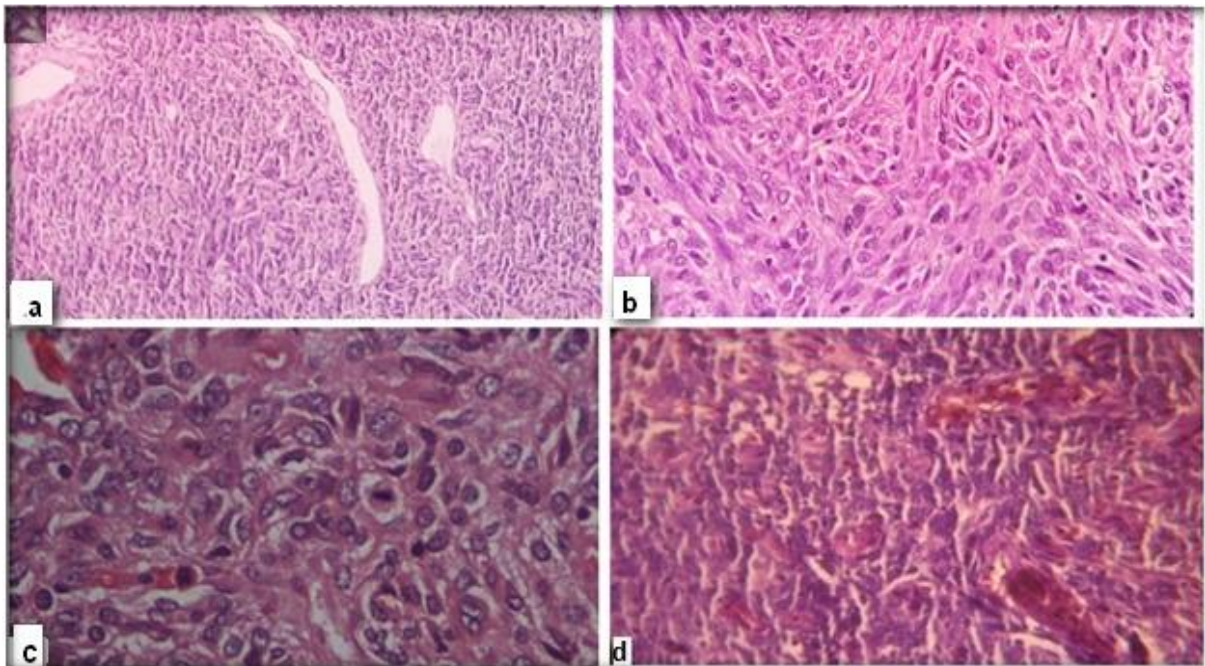
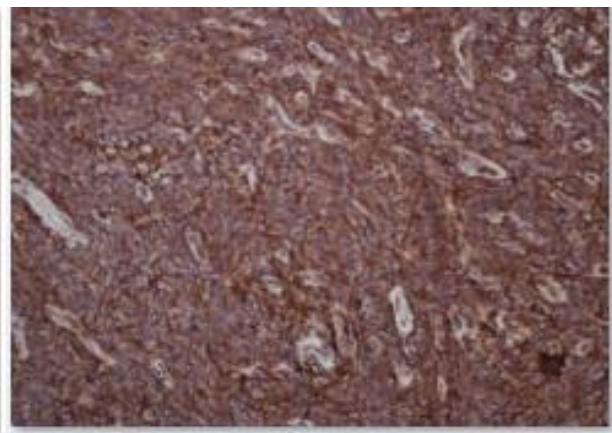


Figure 2: (a) patternless arrangement of fusiform spindle cells with admixture of hyaline collagen and hemangiopericytoma-like area (Hematoxylin and eosin, x200). (b) Epithelioid cell nodule (H and E, X400) (c) Pleomorphic spindle cells with increased mitosis (H & E, X400) (d) cells show hyperchromasia and marked atypia (hematoxylin and eosin, X400).

Immunohistochemistry



bcl2 20X



CD 34 20X

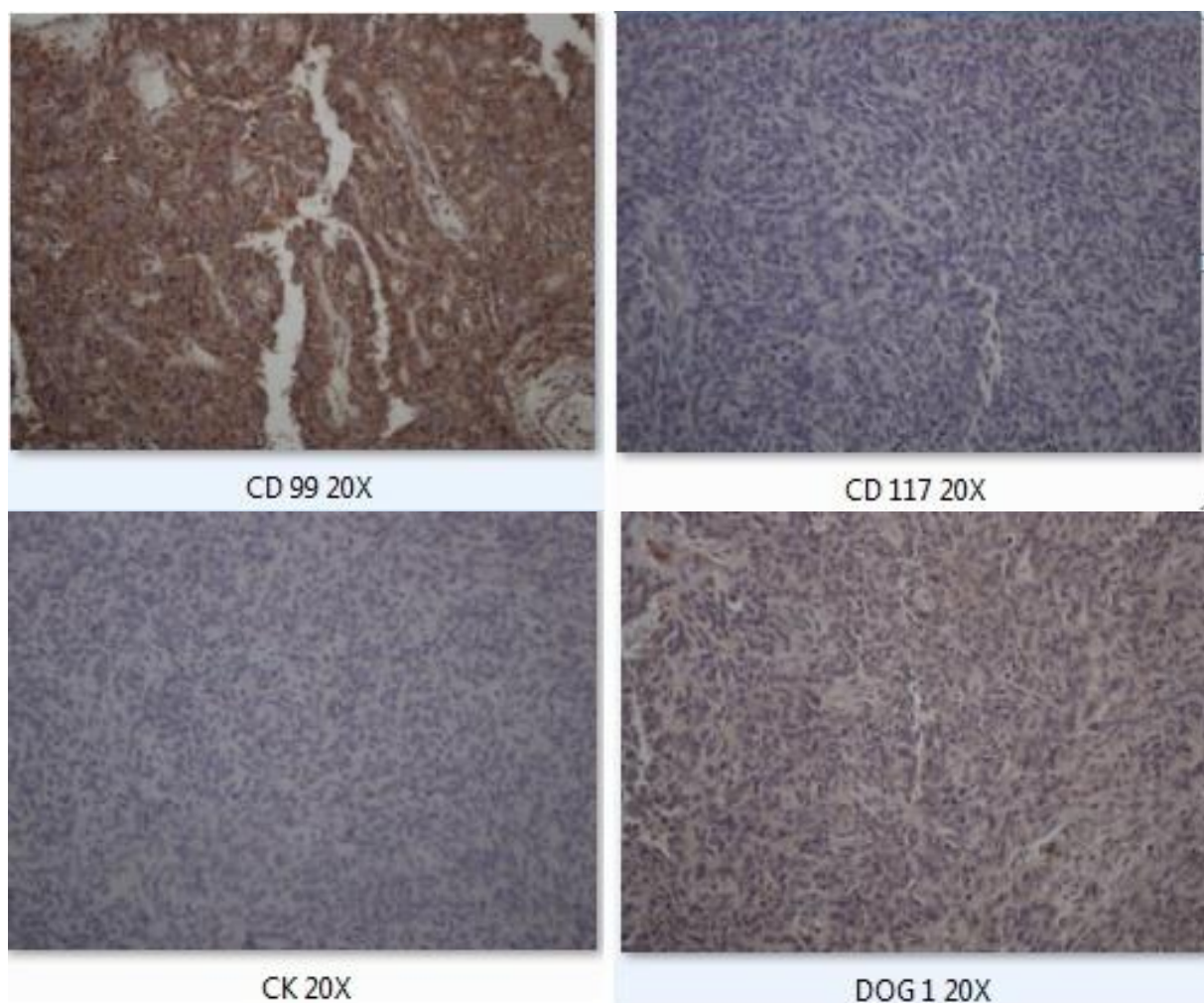


Table 1: Immunohistochemical staining pattern of SFT of the rectus sheath.

Antibody	Source & Clone	Staining Pattern
CD34	DaKoQBEnd 10	Diffuse Strong Positive
Ki67	DaKo MIB-1	Index-8%
CD99	DaKo 12E7	Diffuse Moderately Positive
bcl-2	Biogenex 100	Diffuse Moderately Positive
CD117	Biogenex R145	Negative
DOG1	Biogenex 1.1	Negative
Pan-CK	DaKo AE1/AE3	Negative
EMA	DaKo E29	Negative

Fluorescence in situ hybridization (FISH) for F1P1-Chic 2 – PDGFRA was done. There is no evidence of deletion/translocation involving the 4 12 region. FISH for PDGFRB showed no evidence of translocation involving the 5q 33 (PDGFRB) region.

DISCUSSION

Extrapleural SFTs are most commonly diagnosed between the fifth and seventh decades of life. They are typically slow growing and asymptomatic.^[7] Specifically, extrapleural SFT has been described within the head and neck region, central nervous system, solid organs, the retroperitoneum, pelvis and the genitourinary tract. SFT's are usually benign, although cases of malignant extrapleural SFTs have

been described, they are extremely rare. The majority of HPC-SFTs are histologically benign. However, a small percentage of HPC-SFTs possess atypical features. Approximately 78% to 88% of SFT's are benign and 12% to 22% are malignant.^[8]

The criteria for malignancy in HPC vary from study to study. The criteria proposed by Enzinger & Smith for malignancy in classic hemangiopericytoma identify overtly malignant or high grade lesions but fail to address low grade lesions. In their study, large size (>5cm), increased mitotic rate (>=4MF/10HPF), high cellularity, presence of immature & pleomorphic tumor cells & foci of hemorrhage and necrosis predicted a highly malignant course.^[9] It is important to note that malignant histologic features are not always an indicator of aggressive tumor

behavior, as benign tumor can act locally aggressive and recur, while alternatively malignant tumors can proceed with an indolent course. Nonetheless, with respect to benign SFTs, malignant SFTs do account

for higher rates of local recurrence (63 versus 8%) and metastasize to distant soft tissue sites and the lungs.^[10]

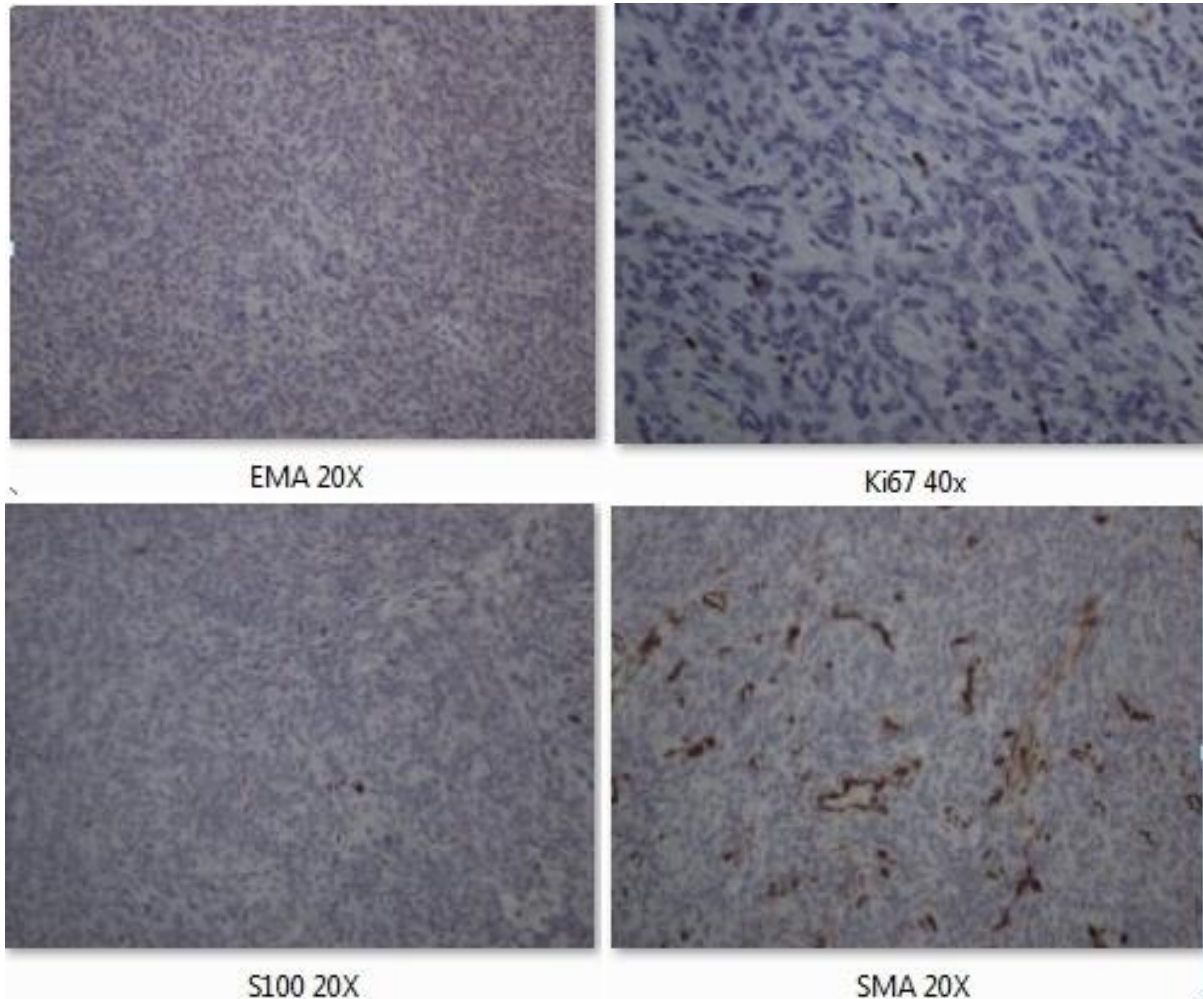


Figure 3: CD34 has taken diffuse strong positivity in tumor cells. MIB-1 index 8%, CD99 positive & bcl2 positive where as CD117, Dog-1, PanCK and EMA S-100, SMA are negative.

SFT of pleural and extrapleural origin typically express CD34 (80-90%), CD99 (70%), bcl2 (30%), EMA (30%) & actin (20%). Desmin, cytokeratin & S100 protein are usually absent. The high sensitivity of CD34 for solitary fibrous tumour has resulted in a more accurate & consistent diagnosis of the entity, undoubtedly accounting for the increasing number of solitary fibrous tumors now diagnosed at extrathoracic sites.^[9]

In our case, the biphasic pattern i.e presence of epithelial & spindle cell prompted us to bring Extra GastroIntestinal Stromal Tumour (EGIST) as differential diagnosis. There is perivascular hyalinization as seen in hemangiopericytoma, also seen in EGIST. The cells have short fusiform shape, collagen interrupted by delicate vasculature. The differential diagnosis of gastrointestinal stromal tumor (GIST) and solitary fibrous tumors may be of

diagnostic challenge because of overlapping clinicopathologic features. Approximately 5% of GIST are KIT-negative & tend to be located in the stomach /peritoneum and have an epithelioid appearance. Most harbor platelet derived growth factor A (PDGFA) mutation while a minority (20%) harbor KIT mutation, but not both. DOG1, a protein of unknown function is expressed in GIST irrespective of mutational status.^[11] In our study we have excluded EGIST by immunohistochemistry which showed CD117, DOG1, pan CK & EMA negativity as well as absence of translocation in PDGFRA & PDGFRB in FISH study. Further CD34 strong positivity, MIB-1 index 8%, CD99 & bcl 2 positivity are in strong favour of diagnosis of solitary fibrous tumor. CD99 is a marker for SFT, and its expression in GIST has not been evaluated.

Recently, genomic studies have revealed a gene fusion that may have diagnostic and therapeutic implications for patients with SFT. The NAB2 gene which indirectly represses transforming growth factor (TGF)- β , and the STAT6 gene which is a transcriptional factor that modulate signaling through interleukin-4 and interleukin-13, have both been described as oncogenic. A recent study of 53 patients undergoing whole exome tumour sequencing revealed that when fused, these two genes (located on chromosome 12) represent a distinct molecular feature in SFT. This characteristic fusion transcript occurred in 55% of tumors, creating the opportunity to develop drugs that specifically target the fusion gene product^[12]

We have diagnosed malignant extrapleural SFT on the basis of results of morphological and immunohistochemical studies.

CONCLUSION

Solitary fibrous tumor should be considered in the differential diagnosis of mesenchymal tumor in various serosal and extraserosal locations. A combination of light microscopy and immunostaining helps to differentiate solitary fibrous tumor from other tumors.

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How to cite this article: Sahu A, Nahak SR, Nayak SK, Agrawal KC. Solitary Fibrous Tumor of Rectus Sheath: A Rare Case Report. Ann. Int. Med. Den. Res. 2017; 3(3):PT25-PT29.

Source of Support: Nil, **Conflict of Interest:** None declared