

A Rare Case of Benign Solitary Fibrous Tumour Arising From Retroperitoneum

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Abstract: Solitary fibrous tumors (SFTs), are a group of rare, spindle-cell derived fibrous neoplasms most commonly found in the thoracic (pleural) cavity (80%). However, it has been described that SFTs can affect a wide range of anatomic sites outside the pleura, including retroperitoneum, meningeal, nasal cavity and others (20%). Solitary fibrous tumors are usually described in patients between 40 and 70 years of age, and are primarily benign. We present the case of a 46-year-old male presented with a vague abdominal mass, its radiological features, its surgical resection and histopathological findings and immunohistochemistry study.

Keyword: sextra-thoracic, KI-67, retroperitoneum, Solitary fibrous tumor

I. Introduction

Solitary fibrous tumor (SFT) is an extremely infrequent tumor first reported as a pleural tumor in 1898 [1]. This heterogeneous group of tumors includes both benign (80%) and malignant neoplasm, with an ongoing debate concerning their cell of origin. SFTs are commonly described in mid aged and elderly people, and no risk factor has been identified [2]. Many SFTs are found as asymptomatic incidental masses, while others are aggressive symptomatic tumors; depending primarily on their size and location. Because of their histological variety and rarity, it is almost impossible to predict the individual clinical behavior, therefore immunohistochemical analysis, is the most important pillar for diagnosis and prognosis of this disease [3]. Hematoxylin -eosin staining alone may not be sufficient and rather makes it difficult to separate this tumor from other spindle-cell tumors, for which CD34 stain has been described as the most important immunohistochemical marker. Enbloc resection of the tumor remains to be the cornerstone of treatment, radiotherapy may be useful as an adjuvant therapy for malignant forms, while no evidence supports the use of chemotherapy [2].

1.1Case report

A 46-year-old male with no significant previous illness or acute symptoms, was referred to us with the incidental finding of a 20x15 cms vague abdominal mass.No h/o pain,vomiting,constipation and weight loss.Bladder and bowel habits normal.On examination 20x15 cms mass occupying predominantly the umbilical and left lumbar region extending to left hypochondrium and epigastricregion.swelling firm in consistency and does not moves with respiration.

Ultrasound abdomen shows a heteroechoic mass lesion measuring of size 19 x 13 x11 occupying the region of body, tail of pancreas and upper pole of left kidney possibility of GIST,PANCREATICTUMOUR,and RENAL TUMOUR. CECT abdomen shows Evidence of 18 x 15 x10 cm heterogeneous mass with irregular enhancing focus lesion in the region of tail of pancreas, left kidney region.lower pole of left kidney normal.



Fig 1

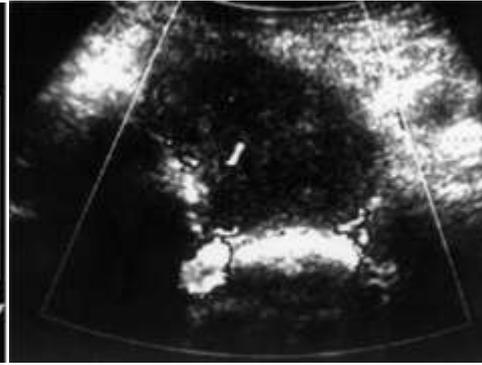


Fig 2

After obtaining surgical gastroenterologist and urologist opinion patient posted for laparotomy and proceed under ETGA with epidural catheter with upper midline incision laparotomy done. Mass found to be occupying the epigastrium, left lumbar and umbilical region.



Fig 3



Fig 4



Fig 5

The mass adherent to transverse colon, descending colon and ileum. Stomach was free. Tumor found to be originating from the retroperitoneum. With meticulous dissection, the bowel loops separated from the tumor. The soft tissue attachment all around the tumor were dissected, ligated and divided. Since the tumor was adherent to Gerota's fascia, the tumor was excised along with Gerota's fascia. Left kidney found to be free. Left Renal Hilum, pelvis were identified and protected. Great vessels found to be free from the tumor. Surface of the tumor was homogenous and the tumor was bilobed. Tumor excised in toto. Postoperative period uneventful. Wound healed well.



Fig 6



Fig 7

HPE Report: Gross huge encapsulated nodular mass measuring 30 x 15 x 10 cm. External surface nodular. Cut surface grey white, Firm bulging appearance. Whorling seen. Glistening in focal areas.

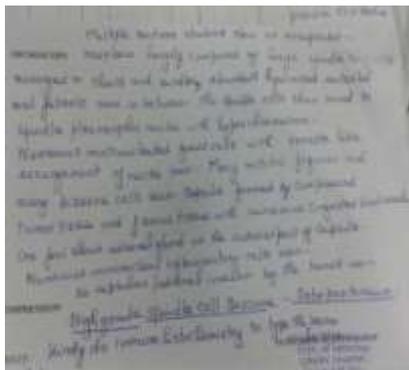


Fig 8



Fig 9

Microscopic picture shows large spindle cells with pleomorphic nuclei and hyperchromatism arranged in sheets and bundles. Numerous multinucleated giant cells seen. Many Mitotic figures and bizarre cells seen. Features suggestive of high grade Spindle Cell Neoplasm-Retroperitoneum. To do Immunohistochemistry to type the lesion.

Immunohistochemistry report shows positivity for cd34,ihc and ki-67 suggestive of benign solitary fibrous Tumour.

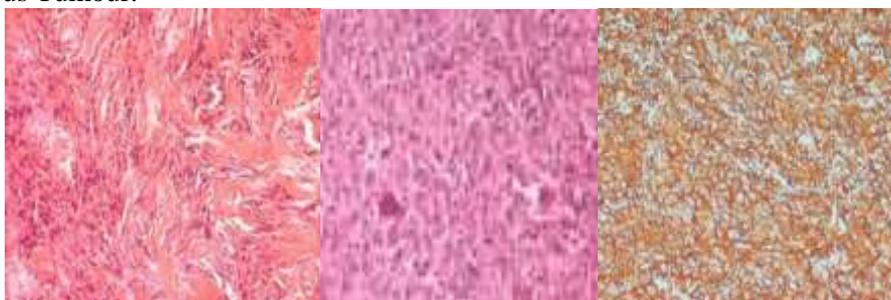


Fig 10

Fig 11

Fig 12

Patternless architecture with thick SFT with giant multinucleated Immunoreactivity for CD34 collagenous bundles stomal cells Oncologist opinion obtained and advised regular follow up.

II. Discussion

Solitary fibrous tumors, as well as related lesions such as lipomatous hemangiopericytoma or giant cell angiofibroma, are rare fibrous neoplasms originating in the pleura and occurring often in the thoracic cavity, although recent data suggests that as much as 20-30% of SFTs have extra-pleural origin [4,5,7]. Such as the case presented above, a solitary fibrous tumor with retroperitoneal origin, supporting a recent study of Wignall et al., which reviewed 34 cases with histopathologic confirmation of SFT, where only 3 cases (9%) were intrathoracic [11].

SFTs were initially considered to have mesothelial origin, therefore termed solitary/localized mesotheliomas [6] but its histogenesis has been controversial. Recent histological and immunohistochemical studies define that fibrous tumors of this category have a mesenchymal origin as the most probable one [6,7]. Because of the unspecific histological morphology of SFTs, (the so-called "patternless pattern") immunohistochemical analysis is vital for the final diagnosis, rather than hematoxylin-eosin staining alone [9].

Immunohistochemical staining has shown that most SFTs are negative for desmin or SL-100, and more than 90% are positive for CD34 or BCL2, making immunoreactivity for these antigens crucial for the final diagnosis of SFTs [10,11].

Solitary fibrous tumors principally affect middle aged adults, with no sex predilection, or other known risk factor [1,5,12].

Most SFTs are asymptomatic and present as a painless mass, rare and unspecific symptoms may present depending on location, most commonly as a local pressure effect [12].

Because of their variety in presentation, unspecific or null symptoms, SFTs are usually diagnosed as an incidental finding in radiographs or CT-scans [11].

On imaging studies, SFTs generally appear as a single, lobulated (62%), well defined mass, with well defined margins (100%) and occasional collateral feeding vessels (35%) [11].

Shin et al., reported that SFTs show heterogenous enhancements and areas of different uptake in CT-scans, probably due to the varying cellularity. On T2 MR images, benign SFTs tend to show low signal intensity, whereas malignant SFTs tend to reveal a higher signal intensity on T2-weighted images [13,14].

Encountering a large, solid, vascular tumor, with prominent feeding vessels should always alert the possibility of a solitary fibrous tumor. Percutaneous biopsies offer minimal risk and should be used for diagnosis. SFTs are usually benign (80%) in all sites, with overall good prognosis with surgical en-bloc resection, which remains to be curative in almost all cases; radiotherapy may be used as adjuvant therapy. Long term follow up is mandatory, with CT-scan or MR imaging [2,11].

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