Abstract:
Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm mainly originating in the pleural cavity. Here, an unusual case of a large SFT in the retroperitoneum is reported.

Keyword: Solitary fibrous tumor, retroperitoneum, stag horn, spindle cells.

INTRODUCTION:
Solitary fibrous tumor (SFT) is a mesenchyme-derived tumor originating mainly in the pleura and rarely in extrapleural sites of origin[1]. Cases of SFT in various extrapleural sites, such as the mediastinum[2], pericardium[3], nasal cavity[4], liver[5], renal capsule[6], thyroid gland[7], salivary gland[8], orbit[9], peritoneum[10], and retroperitoneum[11] have also been reported. Most of SFT are benign, but SFT has also the potential for malignant behavior. Immunohistochemical analysis is of adjunctive importance in making the differential diagnosis and predicting the behavior of this disease. Here, an unusual case of large retroperitoneal SFT presenting in the retroperitonium is described.

CASE REPORT:
A 55 year old male was admitted with complaints of chronic lower abdominal pain, constipation, loss of appetite and weight loss since 8 months. CT abdomen showed a retroperitoneal, lobulated hyper dense lesion
of size 22 x 18 x 8 cms displacing the intestinal loops on the left side. A USG guided biopsy was done and sent for HPE. Grossly, the biopsy tissue was a linear grey white soft tissue fragment of about 1 cm in length, all embedded. Histopathology showed a fragment of neoplasm composed of spindle shaped cells with moderate cytoplasm, elongated nuclei arranged in diffuse sheets and fascicles interspersed with numerous congested blood vessels. No mitotic figures, necrosis were made out. An impression of spindle cell neoplasm was given. The surgeons proceeded with total excision. The mass was found adherent to bowel loops and removed in two pieces.

GROSS FINDINGS:

Two separate soft tissue masses one measuring 15 x 10 x 4 cms and another measuring 8 x 8 x 4 cms were received. The external surface was nodular and grey white (Fig 1). The cut surface was grey white and showed hemorrhagic foci. Representative samples were taken from different areas and processed for H & E and IHC (Fig 2).

Figure 1 : Gross – Soft tissue mass. External surface – nodular and grey white.
Figure 2 : Cut surface – grey white with hemorrhagic foci.

HISTOPATHOLOGICAL FINDINGS:
Sections showed a poorly circumscribed neoplasm composed of spindle shaped cells with moderate cytoplasm and elongated spindle shaped nuclei arranged in diffuse sheets and short fascicles. Blood vessels exhibiting staghorn pattern were found interspersed among the tumor cells. Mitotic figures were 2-3/10 hpf. Occasional cells exhibited nuclear pleomorphism. Focal areas of hyalinization, hemorrhage were made out (Fig 3 & Fig 4).
IMMUNOHISTOCHEMISTRY:
Immunohistochemical study of the tumor showed positivity for CD34, vimentin and bcl-2 (Fig 5, 6 & 7). The tumor cells were negative for S-100, CD117 and CD31.

DISCUSSION:
SFT is an uncommon soft tissue tumor that usually develops in the pleura. Approximately 30% of SFT arise in extra-pleural locations [1]. SFT was initially thought to be of mesothelial origin and therefore termed solitary/ localized mesothelioma, but its histogenesis has been controversial [12]. Recent histological and immunohistochemical studies have described fibrous tumors in this category as solitary/localized fibrous tumors derived from ubiquitous dendritic interstitial cells such as submesothelial stromal cell or fibroblast like mesenchymal cell [13]. The histological characteristics of SFT were the so-called “patternless pattern” characterized by a haphazard, storiform arrangement of spindle cells, and “hemangiopericytoma-like appearance” with prominent vascularity [14]. Our case showed the same histological features of a solitary fibrous tumor. But it was seen to arise from the retroperitonium. A definitive diagnosis of SFT needs confirmative IHC to exclude other spindle cell lesions. Therefore, immunohistochemical staining was used for establishing the diagnosis, in particular CD34, which is considered as a positive marker for SFT [15]. CD34 showed diffuse positivity.
Positivity for vimentin, a general marker of cells originating in the mesenchyme, supported the diagnosis of SFT. Other similar spindle cell neoplasms mixed with adipose tissue, such as dendritic fibromyxolipoma, gastrointestinal tumor, lipomatous hemangiopericytoma, and spindle cell lipoma, should be distinguished from SFT. The histological and immunohistochemical features are helpful for excluding other CD34-positive tumors, such as neural tumors and smooth muscle tumors. Immunoreactivity of the present tumor for CD34 and vimentin, coupled with the absence of immunoreactivity for CD31, CD117, cytokeratin, desmin, and S-100, strongly favored a definite diagnosis of SFT.

Hasegawa et al. reported that 75% of extrapleural SFT showed positive reactivity for Bcl-2, supporting the view that the expression of Bcl-2 can be used together with CD34 in the diagnosis of SFT[16]. Our case also showed bcl-2 positivity which further supported the diagnosis.

In summary, an unusual case of large retroperitoneal SFT is described. The possibility of an extrapleural location of SFT must be kept in mind to set up the correct diagnosis, because symptoms and radiologic findings do not lead support to a diagnosis of retroperitoneal SFT. Most extrapleural SFTs usually have a favorable prognosis after complete local excision, but may have a potential to recur or metastasize. Hence, careful clinical long-term follow-up is necessary for all patients with SFT.

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