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CASE REPORT: A RARE CASE OF RETROPERITONEAL SOLITARY FIBROUS TUMOR

Radio-Diagnosis

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ABSTRACT

Making the diagnosis of solitary fibrous tumor can be challenging, considering its rare nature, however not it is not impossible. Features such as large, fairly well-defined solid masses which show progressive and prolonged enhancement through the phases, capsule however the features of cystic degeneration, myxoid degeneration & necrosis make the diagnosis challenging on cross sectional imaging. Hence, histopathology is the gold standard of diagnosis in such cases and plays an essential role in guiding further management. Solitary fibrous tumors, when examined histologically, consist of spindle-shaped cells embedded in a collagen-rich stroma.

KEYWORDS

Retroperitoneum, Retroperitoneal solid mass, Solitary fibrous tumor, Computed tomography, Magnetic resonance imaging, Abdominal Radiology

INTRODUCTION:

Solitary fibrous tumors (previously known as hemangiopericytomas) are uncommon mesenchymal neoplasms, accounting for less than 2% of all soft tissue tumors. Their behaviour varies from slow-growing to more aggressive forms.

Solitary fibrous tumors (SFTs) can grow to a considerable size and may develop in almost any location, though certain sites are more commonly affected than others- pleura and meninges being the most common sites. However, they can also be encountered in other locations such as retroperitoneum, trunk and extremities.

Cross sectional imaging such as Computed tomography and magnetic resonance imaging play an essential role in pre-operative evaluation of the lesions in their extent, tissue characterization and any potential complications.

Histologically, SFTs are sharply marginated multilobulated firm masses. The cut surface is dense and pale in colour however higher-grade lesions may show varied appearances due to changes of fat, haemorrhage, necrosis and myxomatous changes. These more aggressive variants can also exhibit ill-defined margins, loss of fat planes with infiltration into adjacent tissues.

CASE REPORT:

A 44-year-old hypertensive female presented to the surgery OPD with complaints of left lower limb pain and difficulty in walking since the past two years. The pain was insidious in onset and gradually progressive, radiating from the hip to knee & was aggravated on standing for long durations, relieved on rest. The patient also presented with intermittent left sided lower quadrant abdominal pain.

General physical examination and vitals were within normal limits. Lower Limb examination was normal and peripheral pulses were palpable.

On palpation of abdomen, a non-tender, firm and non-mobile mass was felt in the left lumbar region

Blood investigations such as CBC, LFT & RFT were normal.

A contrast enhanced CT was requested for the patient which showed:

A large heterogeneously enhancing solid-cystic retroperitoneal mass lesion with internal thick enhancing septations with lobulated margins in the left posterior pararenal space and left lumbar paravertebral region. The lesion shows progressive enhancement of its solid and cystic component with no evidence of washout. No evidence of hemorrhage/calcification was seen within it. No obvious neural foraminal widening is present.

The lesion is seen causing anterior displacement of the left kidney with loss of fat planes with its posterior aspect at places and anterior displacement of the descending colon.

There is dilatation of the left pelvicalyceal system and the left proximal & mid ureter- likely due to compression between the mass and the uterus in its distal portion.

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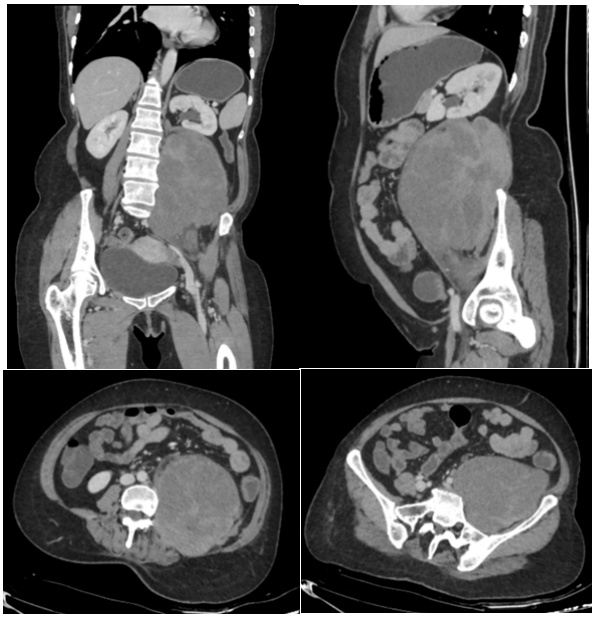
The lesion is seen to cause medial displacement of the left common iliac vessels.

Superiorly, the lesion shows anterior displacement of the superior portion of the left psoas muscles however rest of the muscle can't be delineated separately seen from the lesion- likely suggestive of its infiltration.

The lesion shows infiltration into the iliacus, quadratus lumborum and the posterior paraspinal muscles on the left side.

There is seen associated erosion of the left transverse process of the L5 vertebra.

No evidence of visualized pulmonary or skeletal metastasis was present.



On subsequent MRI for the patient, there was a large solid-cystic retroperitoneal mass lesion with lobulated margins, in the left lumbar paravertebral region. The lesion shows internal T2/STIR hypointense thick septations and internal T2/STIR hyperintense & T1 iso-hypointense cystic areas.

There was no evidence of signal loss on chemical shift imaging.

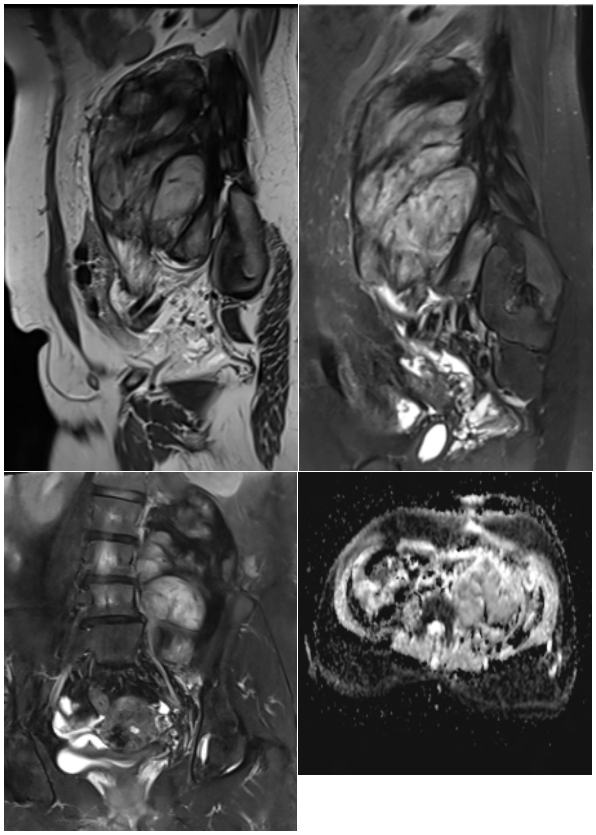
There was evidence of a focal area of signal loss on all sequences which shows blooming on GRE sequence-likely representing calcification.

There was evidence of restricted diffusion on DWI sequence.

The lesion was seen causing anterior displacement of the left kidney and descending colon.

The lesion showed mass effect and medial displacement of the common iliac vessels on the left side.

The lesion showed infiltration into the psoas, quadratus lumborum & iliacus muscles on the left side.

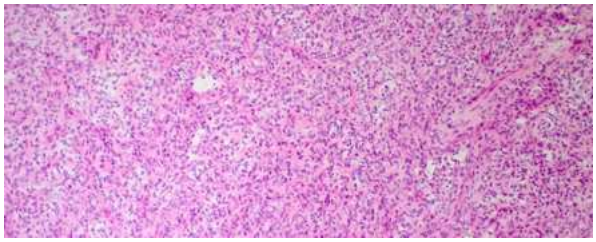


The figures depict a large solid-cystic retroperitoneal mass lesion with lobulated margins in the left lumbar paravertebral region. The lesion shows internal T2/STIR hypointense thick septations and internal T2/STIR hyperintense & T1 iso-hypointense cystic areas. The lesion shows areas of restricted diffusion with it.

The following differential diagnosis were considered:

1. Solitary fibrous tumor: The points in favour were well, defined, lobulated mass lesion with internal fibrous tissue showing delayed enhancement, non-enhancement of the internal cystic/myxoid change, prolonged enhancement of the cellular component.
2. Undifferentiated pleomorphic sarcoma/myxoid fibrous histiocytoma: The points in favour were Well defined mass with cystic or necrotic change spreading across the fascial plane and between muscle fibres with heterogeneous signal intensity and absence of intratumoral fat. However, UFH are more commonly seen in the 6-8th decade of life and have high tendency towards metastasis which were not present in the patient.
3. Neurogenic tumor- schwannoma: The points in favour were well defined encapsulated mass with cystic change, calcification in a paravertebral location. However, neural foraminal widening or extension of the lesion into it was not present. No nodular extension along the neural course was present.

Further biopsy was performed for the patient which showed spindle cells with no areas of cytologic atypia or mitosis. On immunohistochemistry, the tumor cells showed positivity for vimentin, STAT6, Calponin and variable positivity for SMA. The tumour cells were negative for CD34, S100, SOX10, Beta catenin and h-caldesmon.



The figure shows the histopathological sample which depicted spindle cells within the neoplastic lesion.

DISCUSSION:

Solitary fibrous tumors (previously known as hemangiopericytomas) are uncommon mesenchymal neoplasms, accounting for less than 2% of all soft tissue tumors. Their behaviour varies from slow-growing to more aggressive forms. (1)

Solitary fibrous tumors (SFTs) can grow to a considerable size and may develop in almost any location, though certain sites are more commonly affected than others- pleura and meninges being the most common sites. However, they can also be encountered in other locations such as retroperitoneum, trunk and extremities.(1)

Cross sectional imaging plays an essential role in pre-operative evaluation of the lesions in their extent, tissue characterization and any potential complications. Contrast enhanced CT (CECT) reveals large soft tissue mass lesions with well circumscribed, smooth or lobulated margins. These masses show avid enhancement on arterial phases, progressive enhancement in the delated phases. With the increased size of the tumor, there will be increased incidence of internal cystic/necrotic change. (2,3)

MR imaging is capable of detecting areas of fibrosis within the lesion as areas of homogeneous low to intermediate T1 & T2 signal intensity relative to the skeletal muscles which show delayed enhancement on dynamic MRI. However, literature has shown SFTs often have no pathological pattern, indicating there will be non-homogeneous distribution of areas of hyalinization, myxoid and cystic changes with areas of haemorrhage. MR imaging is considered better than CT in its ability to differentiate soft tissues and is helpful in detecting necrotic, haemorrhagic or cystic changes. (2,4)

Although SFT is rare, possibility of SFT should be put into consideration in the differential diagnosis of a well-defined predominantly solid retroperitoneal tumor with inhomogeneous T2 characteristics and a dynamic enhancement pattern.(5)

On gross examination, SFTs are sharply marginated multilobulated firm tumors of mesenchymal origin. The cut surface is dense and pale white-grayish in colour however higher-grade lesions may show varied appearances due to changes of fat, haemorrhage, necrosis and myxomatous changes. More aggressive variants of the tumor can also exhibit ill-defined margins, loss of fat planes with infiltration into adjacent tissues. (2,6)

SFTs show rrandomly arranged spindle or ovoid shaped cells within a collagenous stroma, intermixed with blood vessels in a pattern-less manner. The histological appearance of solitary fibrous tumors ranges from sparsely cellular areas with abundant stromal collagen to highly cellular tumors with minimal stromal content. Mitotic activity is generally low and plays a key role in assessing the risk of recurrence. Additional pathological features that may impact recurrence risk include nuclear pleomorphism, necrosis, and cellularity.(7)

Immunohistochemically, solitary fibrous tumors are distinguished by strong nuclear expression of signal transducer and activator of transcription 6 (STAT6) however the strong staining of STAT6 is also

observed in other more aggressive well retroperitoneal mesenchymal tumors such as differentiated and dedifferentiated liposarcoma. (7,8)

Other unspecific supportive immunostaining markers used in SFT diagnosis are CD34, bcl-2 and CD99. Majority of the SFTs show strong and diffuse CD34 expression, but in most aggressive SFTs, the expression can be lost. However, CD34 positivity is also seen in various other low grade fibroblastic and myofibroblastic tumors. (7,8)

MANAGEMENT

Complete surgical excision has a feasible and reasonable first line of therapy for retroperitoneal solitary fibrous tumors with minimal perioperative morbidity and mortality and overall good median survival above 4 years. (9)

The goal of surgical resection is to achieve a wide excision while preserving nearby organs unless they are encased, adhered to, or infiltrated by the tumor. Resection can be performed either en bloc or in a piecemeal manner, with some studies indicating no significant impact on prognosis between the two techniques.(10,11)

They exhibit diverse growth patterns, with alternating regions of high cellularity (dense with tumor cells) and low cellularity (abundant in collagen). The diagnosis is confirmed through distinct immunohistochemical staining can have varied results.

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