

A Rare Case of Solitary Fibrous Tumor of the Pre-Sacral Space

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Abstract: A 21 years old young healthy male patient presented to surgical OPD of Sir Ganga Ram Hospital, Lahore with complaint of peri anal discharge. He was subjected to MRI pelvis for the evaluation of perianal sinus/fistula. His MRI revealed no evidence of a sinus or fistulous track, rather a well demarcated heterogeneous pre sacral mass was detected. Correlative abdominal and trans-rectal ultrasound were performed which demonstrated a highly vascular heterogeneous mass, later subjected to trans-rectal biopsy. Tissue biopsy came out to be positive for Solitary fibrous tumor. Patient was then referred to surgical team for excision of pre sacral mass which was completely excised and sent for histopathological analysis that confirmed it to be a Solitary fibrous tumor as well.

Key words: Solitary fibrous tumor • Pre-sacral mass • Rare location • Extra-thoracic fibrous tumor

INTRODUCTION

Solitary fibrous tumor (SFT), constituting less than 2% of all soft tissue tumors is a rare, ubiquitous lesion of a probable fibroblastic or myofibroblastic origin with a hemangiopericytoma-like vascular pattern, previously thought to arise from either mesothelial cells or submesothelial fibroblasts and hence restricted to mesothelium-lined surfaces. The majority are benign, although up to 20% may be malignant. They are usually asymptomatic or may present with secondary symptoms of compression due to their mass effect. This tumor has been successively documented in almost every anatomic site, so presently pleural and extrapleural SFTs are now considered a single entity since they share similar pathologic morphology and immunophenotype [1-3].

Intra thoracic, the commoner variety of tumor, usually presents in old age, however extra thoracic variety can present in any age group. Approximately 40% of solitary fibrous tumors are found in subcutaneous tissue, while others are observed in deep soft tissues of the extremities or in the head and neck region, thoracic wall, mediastinum, retroperitoneum and abdominal cavity. Other locations described include meninges, spinal cord, salivary gland, lung, thyroid, and liver [4] the gastrointestinal tract, adrenal glands, kidney, urinary bladder, prostate,

spermatic cord, testis, nasal turbinate [5] and skin [6]. SFT is most commonly found in adults and affects both sexes equally [7].

Usually, the tumor follows an indolent clinical course with no recurrence or metastasis, yet its elusive clinical behavior makes it impossible to provide an exact prognostic prediction and between benign and malignant SFTs.

Case Report: A completely healthy 21 year young man had complaints of peri anal discharge on and off from last one year. On inquiring, he had vague history of episodic abdominal pain. He was evaluated by surgical team for peri anal discharge. They advised MRI fistulogram to the patient for the probability of peri anal sinus/fistula. His MR fistulogram showed no evidence of peri anal sinus or fistula however a rounded well marginated soft tissue intensity mass occupying pre sacral region identified, measuring 8x7x6.5cm in transvers x CC X AP dimensions respectively which was not in communication with any viscera, gut or anterior abdominal wall.

Correlative abdominal and trans rectal ultrasounds were performed on the patient which showed a highly vascular solid heterogeneous mass located in pre sacral region, not in alliance with any superficial or deep structure.

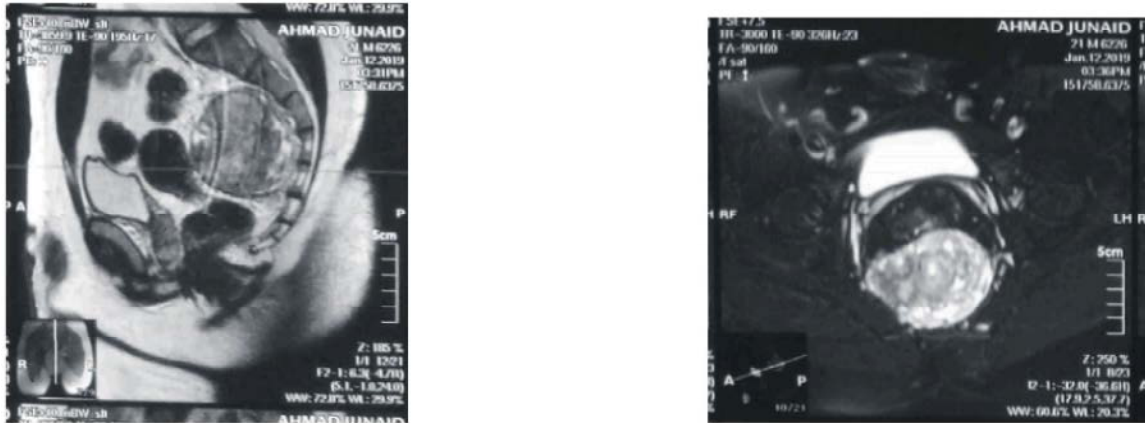


Fig. 1: Sagittal T2WI (a) and Axial T2 Fat-Sat (b) images showing a well-defined heterogeneous pre sacral mass being heterogeneous on T2WI slightly displacing rectum anteriorly with clear plain of separation. It is demarcated from sacrum

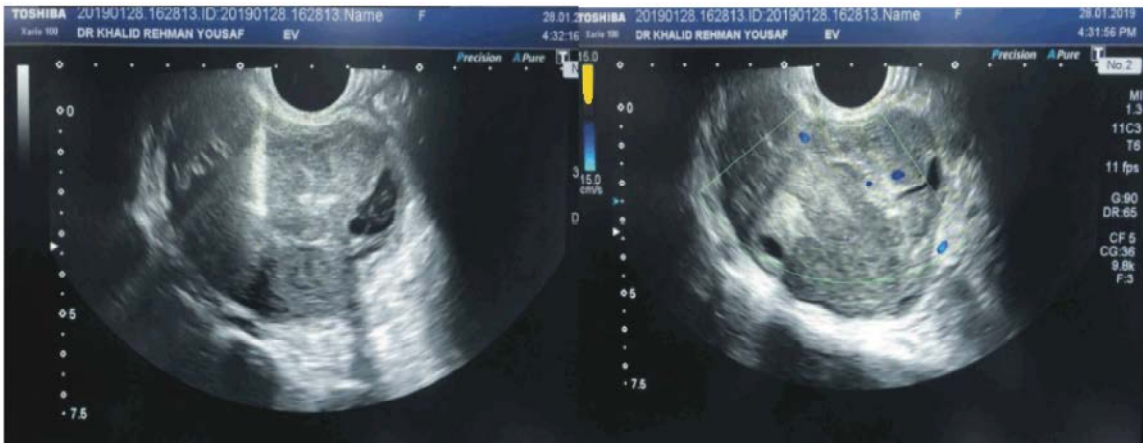


Fig. 3: (a) Transrectal ultrasound showing a large solid mass in pelvis, pre sacral region having heterogeneous internal contents with biopsy needle seen in situ (b) On Color Doppler analysis, lesion showing excessive blood flow.

Trans rectal biopsy was performed, histopathology revealed it to be a solitary fibrous tumor.

Patient was then referred back to the surgical team for excision of this mass. Complete excision was achieved and histopathological analysis further confirmed fibrous nature of the mass.

DISCUSSION

SFTs, first reported by Klemperer and Rabin in 1931 [8] as rare mesenchymal neoplasms that account for <2% of all soft tissue tumors. Gold *et al.* [2] Although SFTs were previously thought to exclusively involve the pleura, it is now established that SFTs can originate in almost any part of the body, with ESFTs (Extra Thoracic Solitary Fibrous Tumor) being more common than pleural SFTs [9-12].

SFTs in the abdomen and pelvis are primarily tumors of adult life which affect both genders equally. Clinically, SFTs manifest as slow-growing, often asymptomatic masses. Common symptoms include abdominal pain, a palpable mass with neurological or vascular symptoms. Symptoms due to mass effect, including urinary retention, bowel obstruction or constipation, and abdominal distention, may be observed with tumors in the abdomen or pelvis [13].

On ultrasound examination, pelvic SFT can often be seen as a hypoechoic mass, but occasionally it is heterogeneous. The latter finding corresponds to the heterogeneity identified using other imaging modalities and likely represents areas of myxoid degeneration. Since SFT is a highly vascular neoplasm, the lesion exhibits flow during Doppler imaging, as was seen in our patient [14].

On CT, pelvic SFTs appear as well-circumscribed masses that often compress adjacent tissues and organs. Large pelvic SFTs have been reported to result in large bowel obstruction and various urinary symptoms including urinary retention and bilateral hydronephrosis [13-15].

MRI is a useful complementary test for characterizing the primary lesion and assessing the disease burden. On T1 weighted images, SFT usually appears as variable signal intensity lesion. Areas of subacute haemorrhage can be identified by T1 weighted signal enhancement. On T2 weighted images, flow voids can be seen as areas of heterogeneous low-signal intensity, as was the case in our patient. Wat *et al.* [16] Gadolinium-enhanced, fat-suppressed T1 weighted MR shows intense heterogeneous enhancement of the pelvic mass in the arterial phase, with progressive enhancement in the venous phase, which are findings consistent with the predominant fibrous content of the tumor [17].

CONCLUSION

In conclusion, the imaging features of SFTs in the abdomen and pelvis predominantly are well-defined, hypervascular masses with variable degrees of necrosis, cystic change, or hemorrhage. They are usually manifest heterogeneously bright on T2WI with low signal intensity areas representing flow voids, fibrosis, or collagen. Although we believe that the radiologist may diagnose SFT when a mass presents with the aforementioned imaging features, histopathologic examination however remains necessary for confirmation.

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