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CASE REPORT

Solitary Fibrous Tumor in Presacral Space- An Unusual Site of Presentation

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ABSTRACT

Presacral space is potentially important because of the presence of neural bundles, soft tissues and the axial skeleton. Any pathology in this space may hamper benign or malignant conditions. Demographics of the patient and correlation with imaging and subsequent confirmation on histopathology may provide a road map to diagnosis. Solitary fibrous tumor is tumor of mesenchymal origin. These present as solitary, well defined masses and mostly lobulated in contour. The typical location is in pleura however extrapleural sites have also been recognized. We present such case where solitary fibrous tumor is seen in presacral space. Diagnosis of which is confirmed on histopathology which showed proliferative spindle cells which are typical for these tumors.
Keywords: Presacral space mass, solitary fibrous tumor.

INTRODUCTION

Solitary fibrous tumor (SFT) as first described by Klemper and Rabin as pleural based tumors in 1931, are spindle cell neoplasms of mesenchymal origin.[1] This tumor is most commonly found in pleura but can occur anywhere in the body. Extra pleural sites although rare (mediastinum, pericardium, lung, retroperitoneum, pelvis and soft tissues of extremities), however are also reported.[2] Solitary fibrous tumors are slow-growing, painless tumors and identified mostly by the mass effect exerted on the local anatomic structures.[3] These tumors are mostly benign (78-88%) but some of these maybe malignant too (12-22%).[4] The histogenesis of SFTs has been debated for years, with early studies suggesting a mesothelial or mesenchymal origin. Recent immunohistochemical and ultrastructural studies strongly support the mesenchymal origin [5,6]

However, these tumors are silent in nature, smaller in size, variably located and easily mistaken with other commonly occurring neoplasm such as angio-sarcoma, lipo-sarcoma, adenolymphoma, schwannoma, hemangioma etc. With these varieties of differentials, patients with SFT's are initially subjected to imaging modalities before clinician advices for histological studies. Keeping this fact in mind, radiological scholars for the last decade have put in an enormous effort at diagnosing these SFT's by means of imaging techniques. Radiologists have been able to diagnose this tumor by means of CT that can accurately determine its size, morphology, location and topography while Magnetic Resonance Imaging, Ultrasonography, Positron Emission Tomography have been helpful too but less used.

CASE REPORT

A 47 years old male, presented with history of lower abdominal swelling, constipation and acute urinary retention for 6 months. On EUA and bimanual palpation, a large pelvic mass was identified. His routine blood chemistry revealed Hemoglobin 9.6 g/dl, Platelet count 409 x 10^9 per liter and Total Leucocytes Count was 6.7x 10^9 cells per liter.

Color Doppler imaging showed a large lobulated heterogenous mass with vascularity in pelvis (Figure 1). Further imaging with Computed

Tomography (CT) scan and MRI showed a large lobulated mass in presacral space with fatty components. This mass showed avid contrast enhancement with few feeding collaterals suggesting neovascularity. Compression and displacement of the urinary bladder and rectum was noted suggesting mass effect. Bilateral iliac vessels and pelvic side wall muscles were also abutted by this mass. No significant bony erosion was noted (Figure 2).

Microscopically, spindle cells comprising of patternless distribution were seen in the background of abundant fibromyxoid stroma. These cells exhibited mild to moderate oval nuclei and occasional prominent nucleoli. Intervening vascular channels of variable size was appreciated. No necrosis or mitosis seen. Immunohistology markers suggested CD 34, BCL 2 positive and S 100 negative (Figure 3).

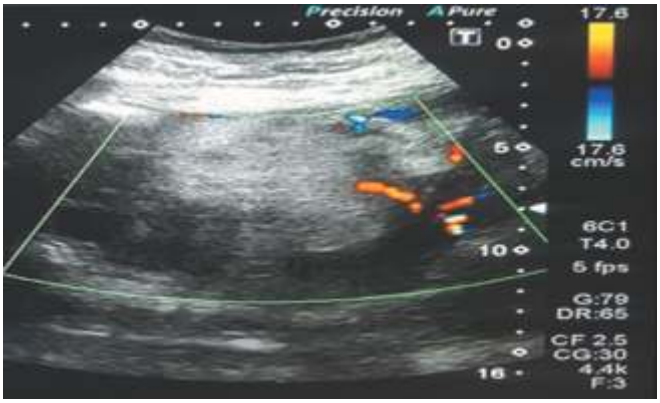

Figure 1: Color Doppler image showing heterogeneous mass in pelvis with vascularity

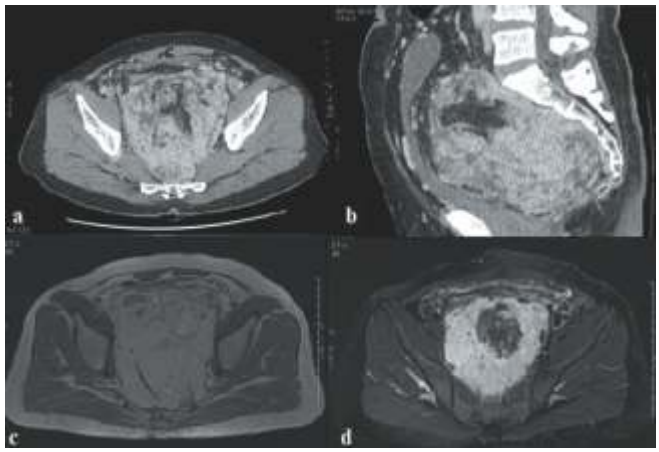

Figure 2: CECT axial and sagittal images: Heterogeneously enhancing mass in pelvis with fatty areas (a & b). Pre and post contrast MRI Fat Sat axial images showing heterogeously enhancing mass in pelvis with fat suppression areas c & d)

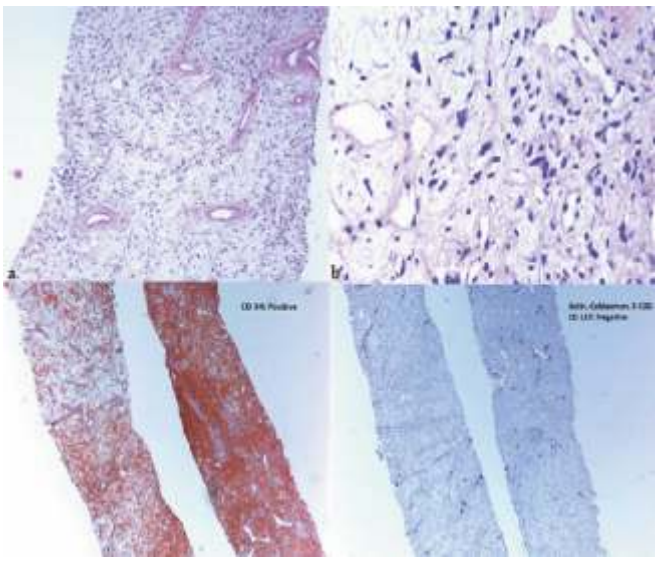

Figure 3: Medium power viewer showing numerous vessels and patternless arrangement of spindle cells with mild to moderate nuclear atypia (a & b). Immunohistology stained CD 34 positive and CD 117 negative (a & d)

DISCUSSION

Solitary Fibrous Tumors are rare mesenchymal spindle cell tumors based in pleura. Imaging plays a vital role in tumor characterization, presurgical assessment and evidence of metastasis. This mass appears as a well demarcated and lobulated lesion causing compression and displacement of adjacent viscera without any significant osseous infiltration and bony erosion. This clarifies its mesenchymal nature. Ultrasound appearances include hypoechogenecity, however heterogeneity is probably secondary to myxoid degeneration.[7] CT demonstrates well circumscribed, smoothly marginated mass with lobulated contours showing intense post contrast enhancement. Some areas of necrosis and fat are also appreciated. MRI being more sensitive shows low to intermediate signals on T1WI and T2WI which are lower on T2WI. Contrast enhancement is noted owing to the fibrous and collagenous nature of the tumor.[8] Radiological imaging can also demonstrate the local extensions and distant metastasis. These are highly vascular tumors. Assertive diagnosis is made by characteristic positive immunohistochemical staining for CD34, BCL 2 and

negative staining for S-1006. Histologically, spindle cells are seen in a background of whorled pattern or appear patternless.[9]

Solitary fibrous tumor although benign in nature, however surgical excision proves to be the gold standard with better survival prognosis.[10]

In our case, CT and MRI features suggested solitary fibrous tumor in presacral space showing avid enhancement on post contrast images with feeding collaterals signifying its fibrous nature and vascularity. Areas of fatty component showed suppression on fat sat images. These were further confirmed by Immunohistology which showed CD 34 positive and S 100 negative. Treatment of choice is surgical resection which is still to be taken after complete workup of the patient.

CONCLUSION

We described imaging features of solitary fibrous tumor existing outside pleura and in presacral space. Keeping in mind the demographics of patient and the particular imaging features, differential diagnosis can be made which can be confirmed by Immunohistology. This will help the radiologist in guiding the clinicians about the tumors located in presacral space.

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