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Research Paper

Extra Pleural Solitary Fibrous Tumor – A Rare Case Report

Medical Science

KEYWORDS : Mesenchymal, Mesothelial, Immunohistochemical,Extrapleural.

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ABSTRACT

A 52 year old lady presented with changes in bowel habits and heaviness in the Lower Abdomen to our Gynaecology OPD. On Vaginal examination she was found to have a pelvic mass. Ultra sound and CT scan of the abdomen and pelvis was obtained in our hospital showed the features characterizing the mass as vascular and homogenous. A core needle biopsy suggested a solitary fibrous tumor of the pelvis. Solitary fibrous tumors are relatively rare mesenchymal neoplasms that were originally described as pleural- or peritoneal-based lesions. They were considered as a form of Mesothelioma.

Introduction

Solitary fibrous tumor (SFT) is a rare Mesenchymal Tumor originating in the Pleura or at virtually any site in the soft tissue including seminal vesicle. Approximately 78% to 88% of SFT's are benign and 12% to 22% are malignant. Extra pleural solitary fibrous tumors are rare tumors of mesenchymal origin. Over the years this entity acquired a number of synonyms, including localized fibrous tumor, benign mesothelioma, localized fibrous mesothelioma, submesothelial fibroma, and pleural fibroma. The use of names that include 'mesothelioma' for this tumor is discouraged because of potential confusion with diffuse malignant mesothelioma.

Case Report

A 52 year old lady presented with changes in bowel habits to our Gynecology OPD. On Vaginal Examination was found to have a pelvic mass. Ultra sound and CT scan of the abdomen and pelvis was obtained in our hospital characterizing the mass as being vascular and homogenous. On MRI, a large mass filling the pelvic cavity displacing adjacent pelvic structures was found. It was 12 cm × 9 cm × 20 cm in size (anterior-posterior, width, cranio-caudal), with well defined borders, and extended cranio-caudally from the level of L5/S1 to the right perineum. The mass filled the pelvic cavity displacing the rectum, bladder, and seminal vesicles to the left side of the pelvis but demonstrates no obvious invasion of the pelvic wall or adjacent structures. . Most of the tumor demonstrated high vascularity with patchy areas of necrosis. There was no fat within the lesion. None of the pelvic organs were invaded .

A thin T2 dark outline was noted around some of the mass suggestive of a capsule With the help of General Surgery and Gynec Department, biopsy was done through the pouch of Douglas approach and sent to Histopathology. Because of the rich vascular supply, biopsy was done with much difficulty.

Histo Pathology of the mass reveals spindle cell neoplasm consistent with solitary fibrous tumor. Microscopy of the tumor demonstrates heavily collagenised soft tissue with areas of necrosis.[Fig-1] In some areas of preserved architecture, a cordlike arrangement of moderately atypical cells varying from spindle to epithelioid is demonstrated.

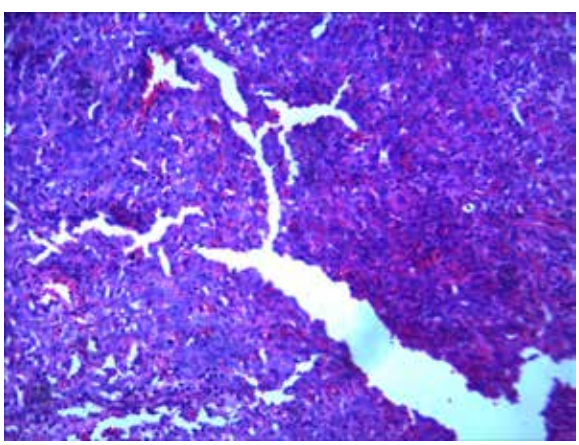

Fig -1 Low power view of SFT

The bands of colloid between the tumor cells demonstrate a keloid-like appearance. Some nuclear atypia is demonstrated, but there are no areas with an increased mitotic rate .[Fig -2].

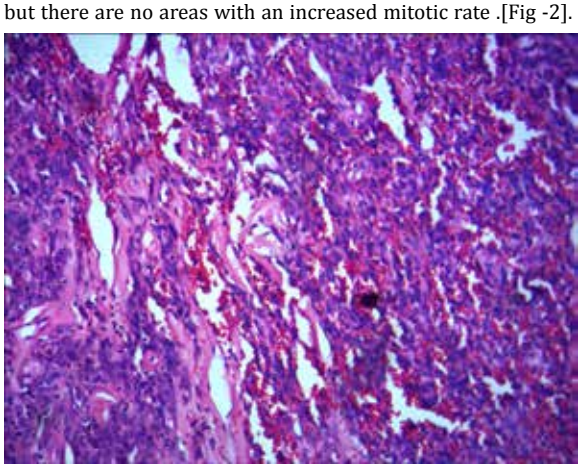

Fig – 2 High power view showing high vascularity

Immunohistochemical stains reveal positive expression for CD34,[Fig -3]

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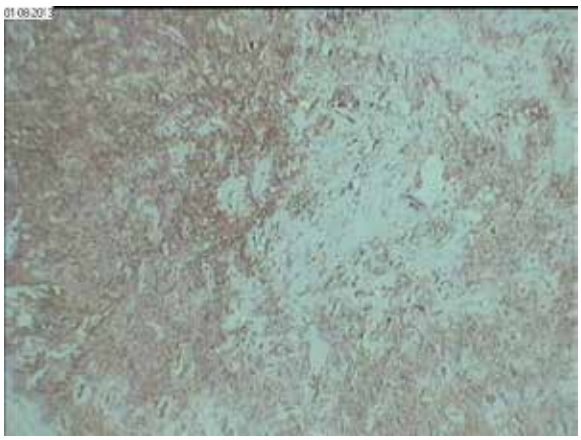

Fig -3 showing CD 34 Positivity

Other sites of this tumor include Orbit, dura/meninges, paranasal sinuses and upper respiratory tract, thyroid, sublingual gland, lung, mediastinum, pericardium, gastrointestinal tract, liver, kidney, peritoneum, retroperitoneum/pelvis, adrenal gland, cauda equina/spinal cord, ovary, uterine cervix/vagina, bladder, prostate, scrotum, testicular tunica vaginalis, trunk/skin, soft tissue, periosteum, and extremities.

Discussion

Solitary fibrous tumors are uncommon tumors arising from mesenchymal cells of the pleura with rare reports of occurrences outside the thorax1,2,3,4,5,6. Less than 10007 cases of solitary fibrous tumor of the pleura and less than 1002 cases of extrapleural solitary fibrous tumor have been reported. Men and women appear to be affected equally and the tumor appears to develop primarily in middle age to older individuals7,9 .The exact etiology has not yet been clearly identified with no known causative/contributing factor or genetic predisposition. Patients may be asymptomatic at presentation or may present with abdominal fullness, abdominal pain, or symptoms related to compression of adjacent structures3,4 Treatment for most cases has been

surgical excision. Recurrence is a possibility, and the role for adjuvant therapy has been suggested7 .Most of the solitary fibrous tumors are benign, however up to 20% of tumors have been reported to be malignant 3.MRI features of this tumor can be nonspecific as these tumors can exhibit necrosis, hemorrhage, and myxoid and cystic changes2,9. with signal characteristics similar to that of a malignant tumor6. . On CT, solitary fibrous tumors have been described to demonstrate heterogeneous attenuation6,9. Approximately 20–30% of tumors demonstrate calcification on CT. Most lesions demonstrate rich vascularity and enhancement on CT and MRI6,7,9. Areas of low attenuation are felt to represent cystic or necrotic changes 6,7,8,9.There is limited description of the ultrasound findings of extrapleural solitary fibrous tumors. Most of those within the thorax have been found to have heterogeneous echotexture but without cysts or calcifications 6.Heterogeneous uptake on FDG PET imaging may be seen10 .

The mass described in our case exhibits mixed signal intensity with areas of extremely low T2 signal on MRI and inhomogeneous attenuation on CT consistent with the pathological finding of necrosis and fibrosis. These findings on MRI are nonspecific and can be seen with other abdominal/pelvic solid masses including but not limited to gastrointestinal stromal cell tumor, myxoid liposarcoma, inflammatory pseudotumor, and neurogenic tumor.Embolization followed by surgical resection results in uneventful complete removal of the tumor. Complete surgical resection is advocated considering the malignant potential in about 20% of the cases and preoperative embolization may be helpful in highly vascular tumors.

Conclussion

The histological variability of solitary fibrous tumors may contribute to the difficulty in diagnosing these neoplasms, especially when they arise in extrathoracic sites. Like intrathoracic lesions, the behavior of extrathoracic solitary fibrous tumors is currently unpredictable because these types of tumor have only recently been recognized. Long-term clinical follow-up is recommended for our patient with solitary fibrous tumor because of the potential adverse biological behavior of this tumor, which may lead to repeated recurrences and/or malignant transformation.

Differential diagnosis table for solitary fibrous tumor

1. CD 34- Positive Myxoid Dermal Dendrocytoma.
2. Cellular digital fibromas: distinctive CD34-positive lesions that may mimic dermatofibrosarcoma protuberans.
3. CD34-positive Cutaneous eruptive fibromas.

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