Solitary Fibrous Tumor Originating in the Pelvis: A Case Report

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ABSTRACT

A 52 year old male presented with changes in bowel movements, and a mass was detected on digital rectal exam. Both CT and MRI revealed a large pelvic and gluteal mass filling the pelvic cavity displacing the adjacent pelvic structures. After surgical removal, pathology revealed solitary fibrous tumor; a rare neoplasm uncommonly discovered in the thorax, and even less commonly in extrapleural locations. We discuss in this article imaging findings and histological features of extrapleural solitary fibrous tumor.

CASE REPORT

CASE REPORT

A 52 year old Native American male was found to have a pelvic mass by digital rectal exam after presenting with changes in bowel movements. A CT scan of the abdomen and pelvis was obtained at a local hospital (Fig. 1) characterizing the mass as being vascular and inhomogenous without obvious fat or calcium. A core needle biopsy suggested a solitary fibrous tumor of the pelvis. He was subsequently referred to our hospital for further management, where an MRI was performed for staging and assessment of vascularity of the lesion.

On MRI, a large mass filling the pelvic cavity displacing adjacent pelvic structures was found. It was 12 cm x 9 cm x 20 cm in size (antererior-posterior, width, craniocaudal), 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. The mass showed heterogeneous signal on T1 and T2 weighted sequences with prominent areas of low T2 signal. A thin T2 dark outline was noted around some of the mass suggestive of a capsule (Fig. 2 a,b,c). Most of the tumor demonstrated high vascularity with patchy areas of necrosis on post Gadolinium contrast T1 sequences (Fig. 3 a,b,c,d). There was no fat within the lesion. Because of the rich vascular supply, the mass was initially treated with embolization (Fig. 4 a,b,c) followed by complete surgical resection.

Surgical excision reveals a 779 gm well defined mass filling >85% of the pelvis and eroding into the right ischiorectal fossa and perineal skin. None of the pelvic organs were invaded but the tumor could only be removed in multiple pieces. Pathology of the mass reveals spindle cell neoplasm consistent with solitary fibrous tumor. Microscopy of the tumor demonstrates heavily collagenized soft tissue with areas of necrosis. In some areas of preserved architexture, a cordlike arrangement of moderately atypical cells varying from spindle to epithelioid is demonstrated. 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. 5,6). The tumor is noted to be surrounded by an incomplete fibrous capsule. In some areas the capsule is absent and tumor cells extended into the resection margins. Immunohistochemical stains reveal positive expression for CD34 (Fig. 7), Bcl2 (Fig. 8), CD99 (Fig. 9), and Vimentin. Stains are negative for CD117 (C-Kit), keratin, S-100, Calretinin, Desmin, and Actin.

DISCUSSION

Solitary fibrous tumors are uncommon tumors arising from mesenchymal cells of the pleura with rare reports of occurrences outside the thorax [1,2,3,4,5,6]. Less than 1000 [7] cases of solitary fibrous tumor of the pleura and less than 100 [2] 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 individuals [7,9]. 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 structures [3,4]. Treatment for most cases has been surgical excision. Recurrence is a possibility, and the role for adjuvant therapy has been suggested [7].

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 changes [2,9] with signal characteristics similar to that of a malignant tumor [6]. They are usually visualized as heterogeneous signal intensity on T1 and T2 weighted images with post contrast enhancement following gadolinium administration. Areas of low signal on T2 weighted sequences might represent mature fibrous tissue in the tumor. Areas of intense enhancement are related to rich vascularity [1,2,9]. Benign and malignant tumors have both been reported to exhibit heterogenous, isointense, and hyperintense signal on T2 weighted sequences along with heterogeneous enhancement following gadolinium injection [1,2,6,9].

On CT, solitary fibrous tumors have been described to demonstrate heterogeneous attenuation [7,6]. Approximately 20-30% of tumors demonstrate calcification on CT [7]. Most lesions demonstrate rich vascularity and enhancement on CT and MRI [6,7,9]. 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 seen [10].

The mass described in this case report exhibits mixed signal intensity with areas of extremely low T2 signal on MRI and inhomogenous 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. An incomplete fibrous capsule is present on the pathological examination and may account for an incomplete rim of low signal intensity on MRI. Avid post contrast enhancement of the mass is demonstrated on CT and MRI consistent with abundant vascularity.

Embolization followed by surgical resection resulted in uneventful complete removal of the tumor. Our patient demonstrates no evidence for residual tumor or recurrence on a four month follow-up MRI.

TEACHING POINT

Extraplural solitary fibrous tumors are rare tumors of mesenchymal origin. They are usually visualized as heterogeneous signal intensity on T1 and T2 weighted images with avid contrast enhancement following gadolinium administration. Areas of low T2 signal and lack of invasion of adjacent structures are some helpful features in suggesting this 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.

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FIGURES

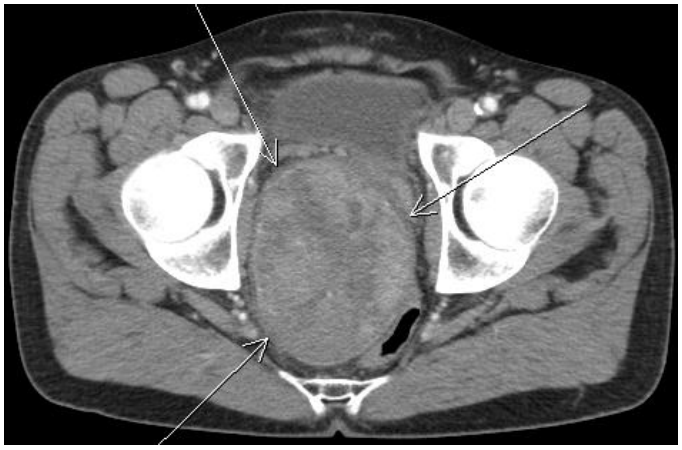

Figure 1: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. A 4 slice CT of the abdomen and pelvis from an outside facility was performed. Axial images were initially obtained of the abdomen following ingestion of 900 cc of Readicat with subsequent images obtained of the abdomen and pelvis following administration of 120 cc of Optiray 320 [4 mm slices, kVp 120, mAs 250].

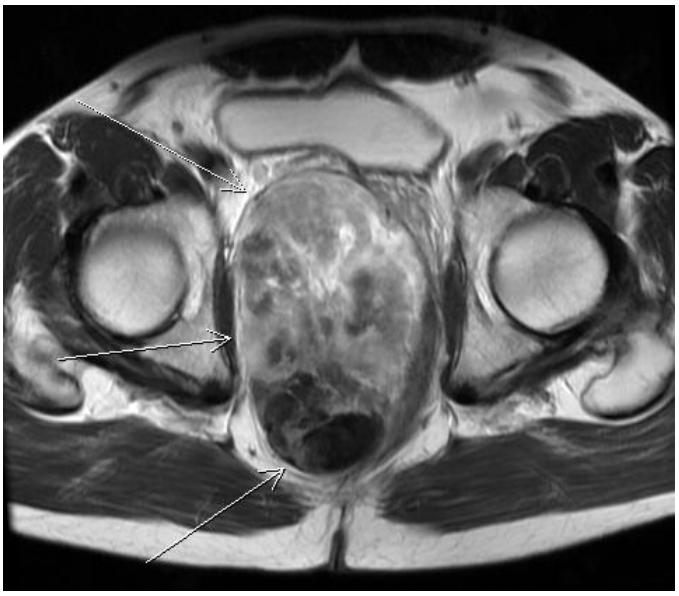

Figure 2a

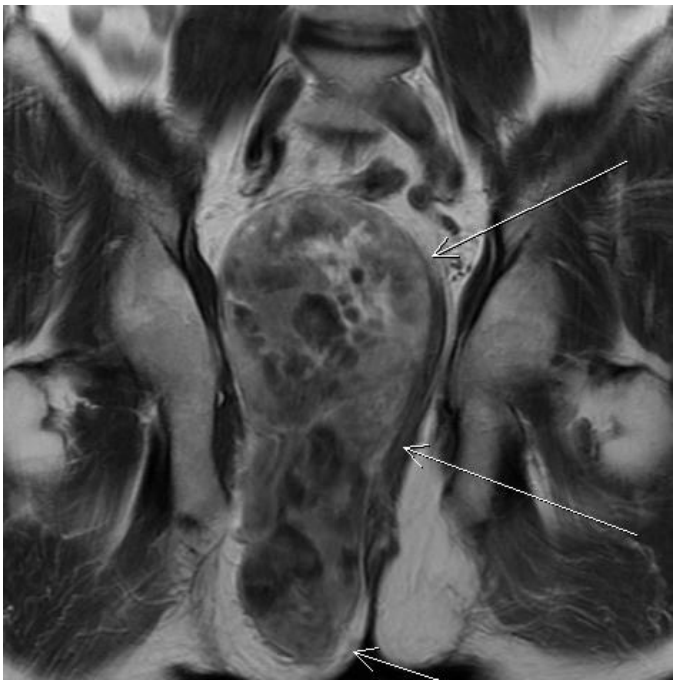

Figure 2b

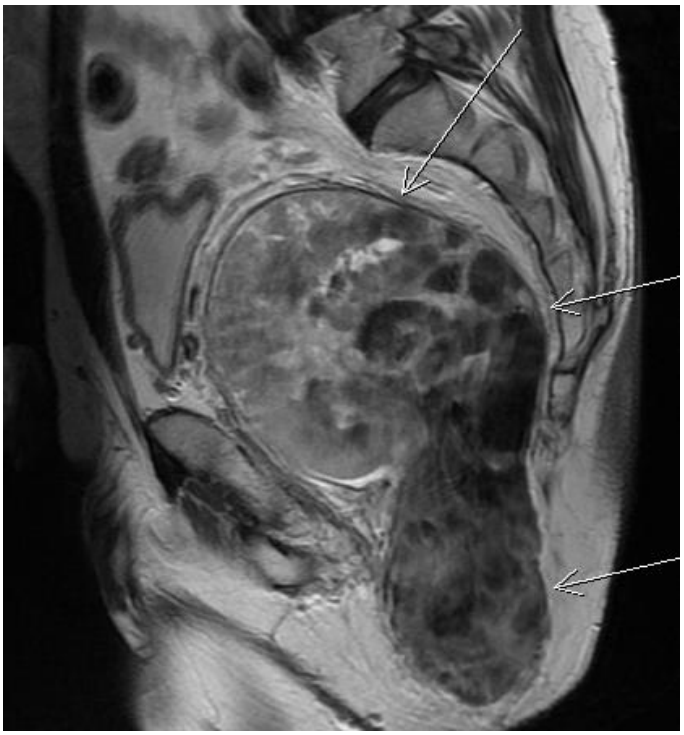

Figure 2c

Figure 2: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. Axial (a), coronal (b), and sagittal (c) T2 MRI images obtained using a 1.5 Tesla magnet [TR 5,000, TE 71 ms, 7 mm slices] reveal a large mass occupying most of the pelvis displacing adjacent structures and extending into the right gluteal region. The mass exhibits heterogeneous signal compatible with necrosis/cystic change with areas of low signal consistent with fibrosis.

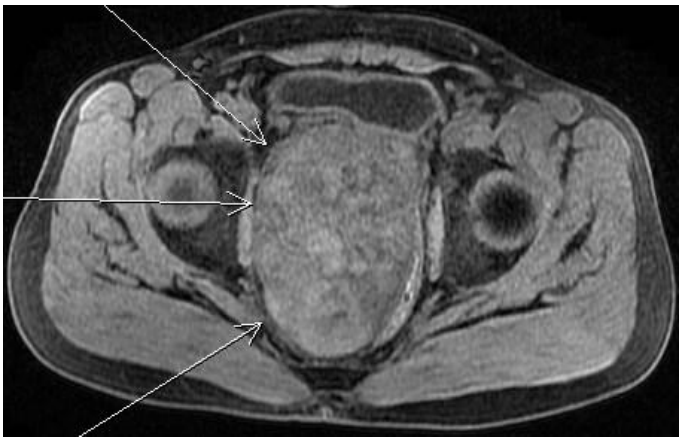

Figure 3a

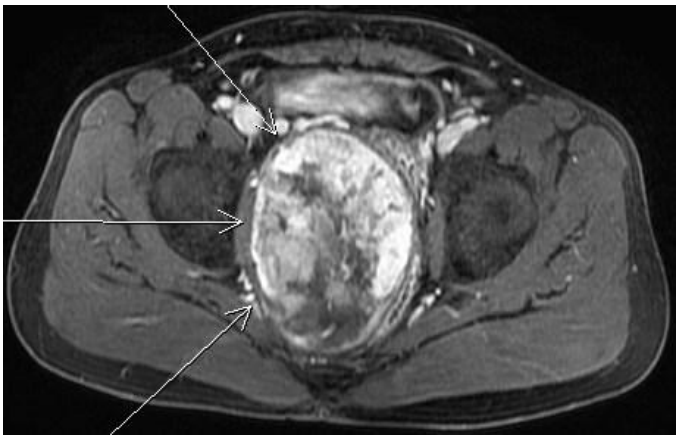

Figure 3b

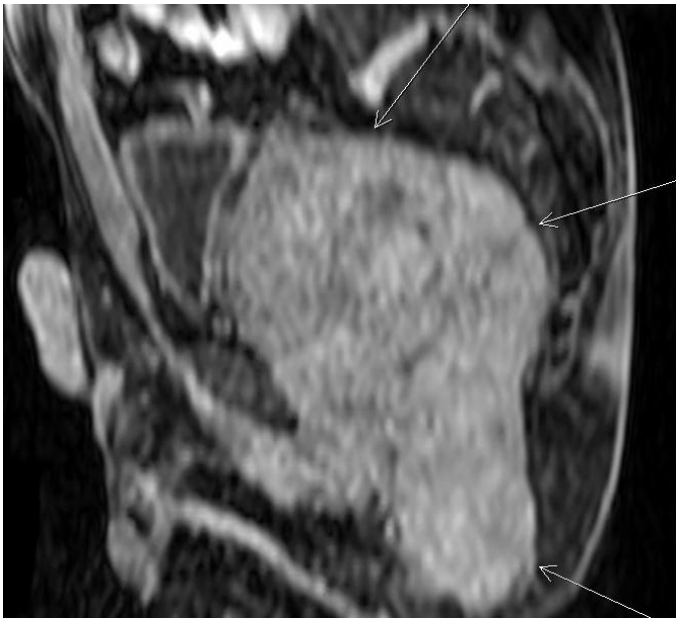

Figure 3c

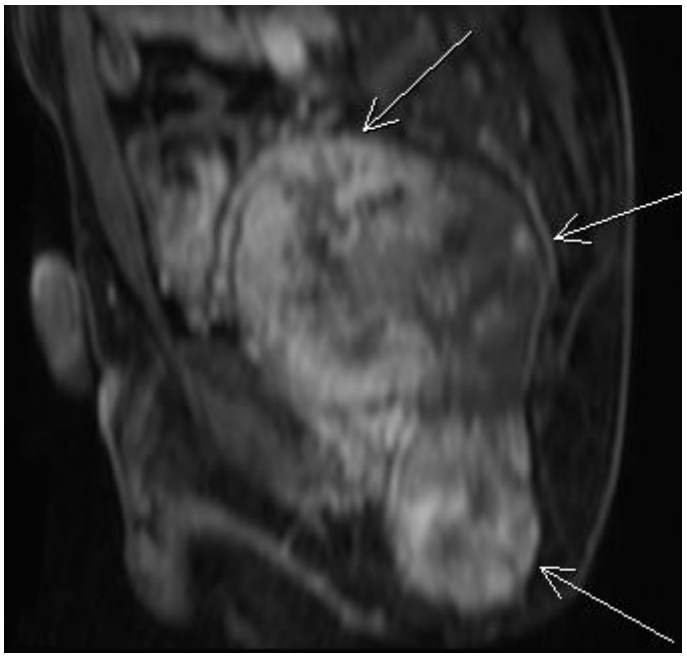

Figure 3d

Figure 3: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. Axial precontrast (a), axial postcontrast (b), reformatted sagittal precontrast (c), and reformatted sagittal postcontrast (d) T1 weighted MRI images obtained using a 1.5 Tesla magnet pre and post infusion (arterial phase) of 20 cc of Magnevist [TR 4.8 ms, TE 2.1 ms, 2.5 m slices] and with fat saturation reveal a mass of heterogenous signal with heterogeneous enhancement occupying most of the pelvis and displacing adjacent pelvic structures. Note the areas of avid enhancement that has been described with these neoplasms. Nonenhancing areas likely represent necrotic/cystic regions of the mass.

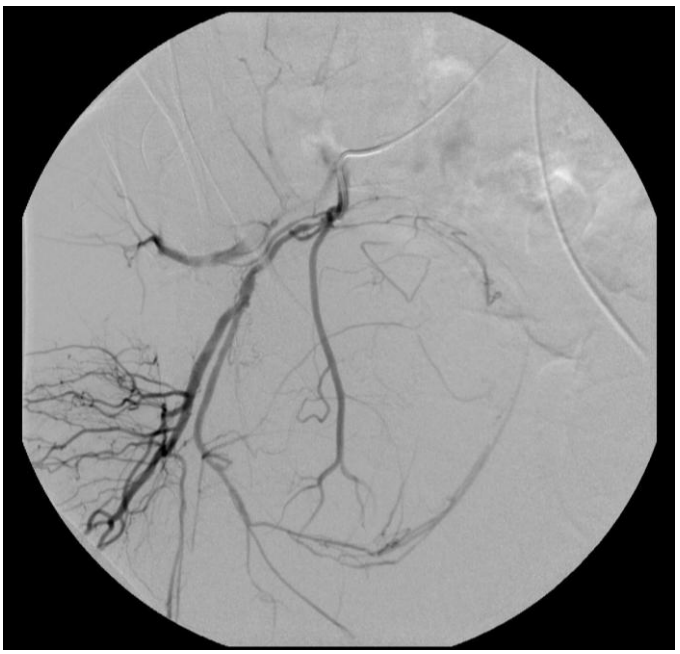

Figure 4a

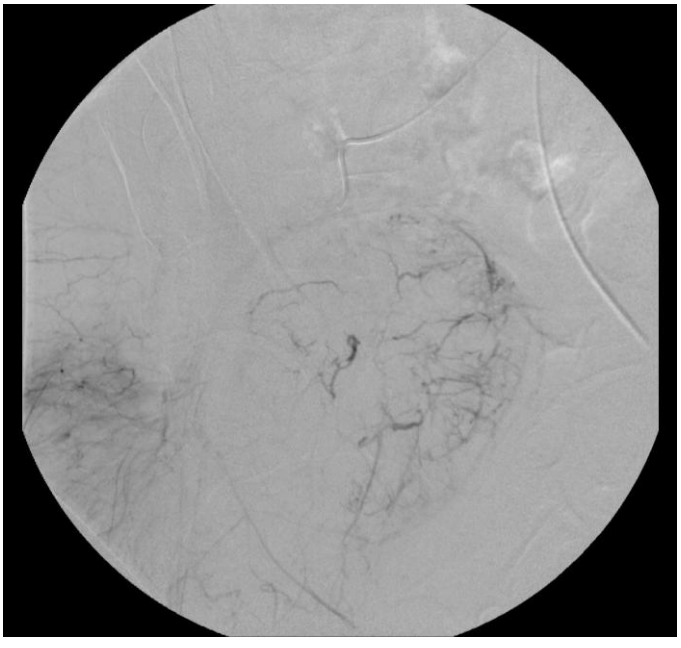

Figure 4b

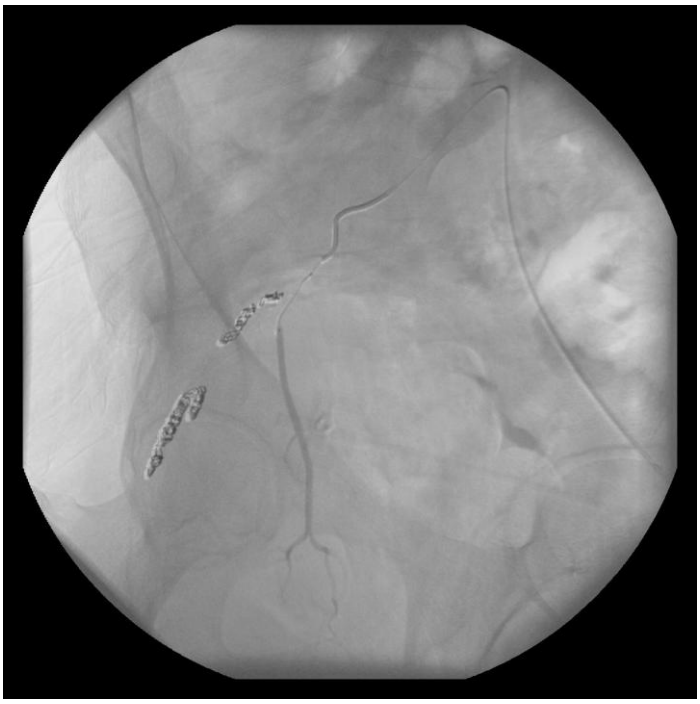

Figure 4c

Figure 4: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. Oblique coronal digital subtraction images are obtained during partial embolization at an outside facility. Arterial (a) and capillary (b) phase images prior to embolization demonstrate the rich vasularity of the mass. Following placement of vascular coils (c), decreased vacularity is demonstrated.

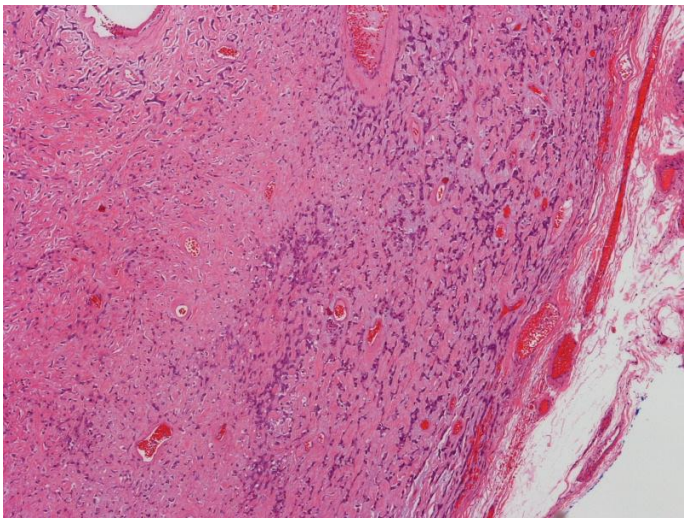

Figure 5: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. Hematoxylin and eosin stain reveals a well circumscribed tumor with cord-like arrangement of mildly atypical short spindle cells in a fibrotic stroma (40x magnification).

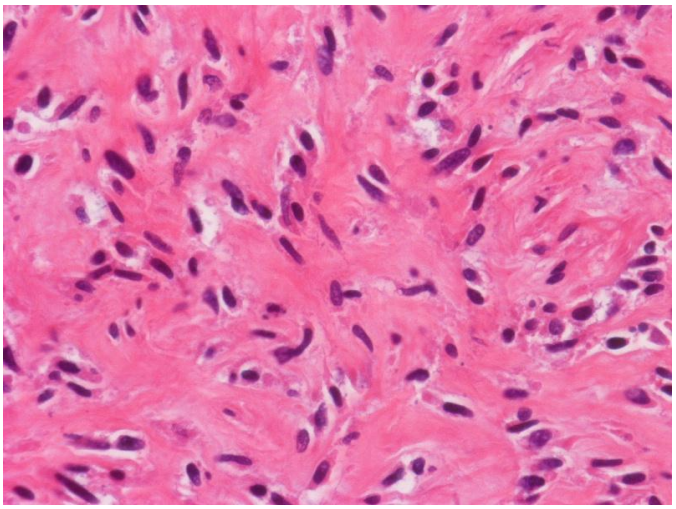

Figure 6: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. Hematoxylin and eosin stain reveals small, short spindle, mildly atypical tumor cells in a random or "patternless" arrangement in a fibrotic stroma (200x magnification).

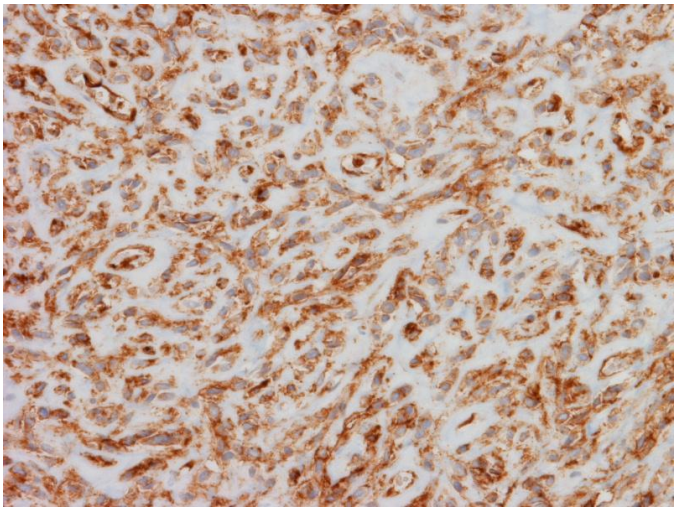

Figure 7: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. Tumor cells stain positive for CD34 (200x magnification).

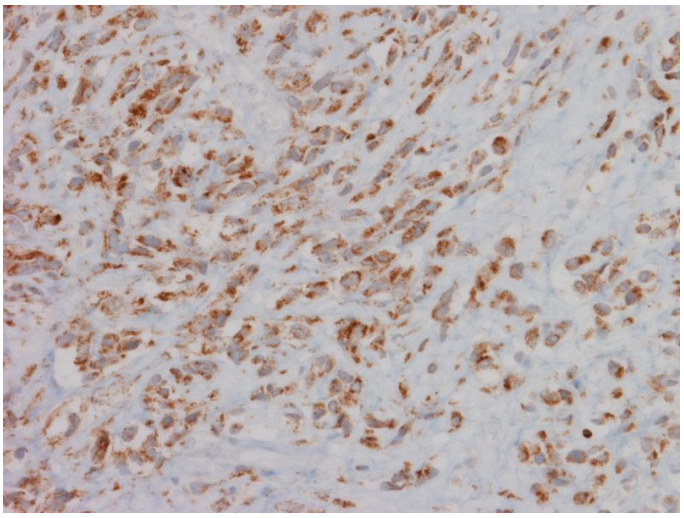

Figure 8: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. Tumor cells stain positive for Bcl-2 (200x magnification).

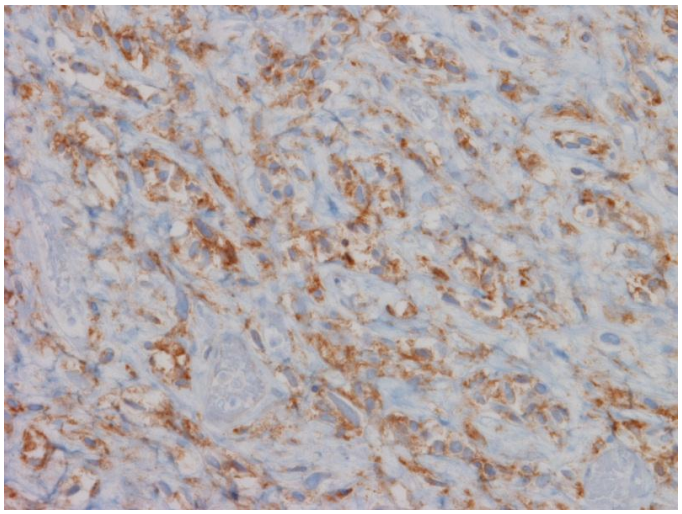

Figure 9: 52 year old male with a pelvic mass found to be an extrapleural solitary fibrous tumor. Tumor cells stain positive for CD99 (200x magnification).

Etiology	Mesenchymal cells of the pleura [7]
Incidence	Rare, less than 1000 reported cases in the thorax and less than 100 reported cases outside of the thorax [7]
Gender ratio	M = F [7,9]
Age Predilection	Middle age to older (40-80) [7,9]
Risk Factors	None known [7]
Treatment	Surgical excision and possible adjuvant therapy [7]
Prognosis	Good, most are benign [7]

Table 1: Summary table for solitary fibrous tumor

Mass	X-ray	US	CT	MRI T1	MRI T2	Pattern of Enhancement	FDG-PET
Solitary fibrous tumor	Soft tissue opacity with possible calcifications [6]	Heterogeneous echogenicity due to necrosis with possible areas of shadowing due to calcifications [6]	Heterogenous attenuation due to necrosis with possible calcification [6]	Heterogenous signal due to necrosis with areas of low signal corresponding to mature fibrous tissue [2,6,7]	Heterogenous signal due to necrosis with areas of low signal corresponding to mature fibrous tissue [2,6,7]	Heterogeneous [2,6,7]	Possible uptake in solid portions of mass [10]
Inflammatory pseudotumor	Soft tissue opacity with possible calcifications [11]	Not well characterized, but primarily hypoechoic [12,13]	Heterogeneous attenuation, may have calcifications [11]	Generally hypointense [11]	Generally hypointense [11]	Heterogeneous, homogeneous, or no enhancement [11]	Can have increased uptake [14]
Neurogenic tumor	Soft tissue opacity [15]	Heterogeneous echogenicity[16]	Heterogeneous attenuation [15]	Generally hypo intense [15]	Generally hyperi ntense [15]	Heterogeneous enhancement [15]	Malignant lesions may demonstrate increased uptake [17]
GIST	Soft tissue opacity [18]	Variable, most hypoechoic with heterogeneous or homogeneous echogenicity [19]	Heterogeneous attenuation, rarely may have calcifications [18]	Heterogeneous with solid components having low signal [18]	Heterogeneous with solid components having high signal [18]	Heterogeneous enhancement [18]	May demonstrate increased uptake [20]
Myxoid Liposarcoma	Soft tissue opacity [22]	Variable, can be hyperechoic and heterogeneous in echotexture [24,25]	Often heterogeneous in attenuation, can have cystic regions, rarely calcifications [22]	May have foci of increased signal from fat, cystic and solid regions usually have low signal [21,22]	Heterogeneous with increased signal in cystic regions [21,22]	Homogenous, heterogeneous, or little enhancement [21,22]	May not show increased uptake [23]

Table 2: Differential diagnosis table for solitary fibrous tumor

ABBREVIATIONS

CT = Computed Tomography
MRI = Magnetic Resonance Imaging
FDG = Fluorodeoxyglucose
PET = Positron Emission Tomography
US = Ultrasound
GIST = Gastrointestinal stromal tumor

KEYWORDS

solitary fibrous tumor; pelvis

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