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Case report

Solitary fibrous tumor of the urinary bladder: An unusual case report with literature review [☆]

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ABSTRACT

Solitary fibrous tumor is neoplasm of mesenchymal origin commonly involving visceral pleura however we are presenting an unusual case with involvement of urinary bladder. It is generally indolent in nature therefore proper diagnosis is required for complete characterization to avoid unnecessary extensive surgical resection. Our patient was a 64-year-old female who presented with lower abdominal fullness with change in her bowel movement pattern. On imaging partially necrotic mass with heterogenous enhancement was found which was later biopsied and resected with clean surgical margin. Solitary fibrous tumor is overall a benign tumor with satisfactory outcome. Physicians should keep it in a differential of pelvic masses and with the risk recurrence, 6 monthly follow up imaging are usually required after resection.

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Introduction

Solitary fibrous tumor (SFT) is an uncommon neoplasm of mesenchymal origin with slightly more prediction of occurrence in male than female. It generally involves visceral pleura but can be seen in other unusual locations such as orbit, meninges, paranasal sinuses thyroid, sali-

vary glands, lungs, mediastinum, pericardium, upper respiratory tract, gastrointestinal tract, urinary bladder, liver, kidney, peritoneum, adrenal gland, spinal cord, ovary, uterine cervix/vagina, prostate, scrotum, tunica vaginalis of testis, skin, soft tissue, and periosteum. It is generally a benign neoplasm and thus organ sparing surgical excision can be considered when possible. Therefore, proper identification and characterization of SFT through morphological and immunohisto-

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Fig. 1 – Axial (A), coronal (B) and sagittal (C) selected images of a non-contrast CT scan of the abdomen and pelvis. The images show a large central pelvic, heterogeneous mass. There is no fat or calcific density within the mass. Images a and c demonstrate close relationship of the mass to the bladder (thin arrows in A and C) and to the uterus, posterior to the mass (thick arrows in A and C). The right adnexa (Asterisk in A) is identified separate from the lesion but not the left adnexa.

chemical criteria on biopsy specimens are mandatory in the differential diagnosis from other more aggressive spindle-cell tumors, thus avoiding unnecessary radical surgery.

Clinical history

We present the case of a 64-year-old female with a relevant past medical history of hyperlipidemia and osteoporosis, who presents for consultation regarding a mass in the lower right abdomen that she is recently feeling. The patient first noticed a change in her bowel movement habits, with new constipation, thin caliber stools and new bloating. She denied melena or hematochezia and her last colonoscopy was 12 years ago and was normal. The patient also denied any vaginal bleeding or discharge. The patient denied any smoking history or alcohol abuse. Her past surgical history was relevant for tubal ligation and appendectomy in 1994. Her BMI was 20.49 Kg/m². Family history was negative for breast, ovarian or colon cancer.

Physical exam demonstrated a large midline suprapubic palpable mass, mildly tender that extended above the level of the umbilicus. There was no appreciable ascites on the physical exam. Laboratory results obtained at the initial visit were normal, and there was no anemia. The clinical suspicion was for a uterine, ovarian or bladder mass although the latter was considered less likely giving the lack of urinary symptoms. Computed tomography (CT) scan of the abdomen and pelvis

demonstrated a large pelvic mass that corresponded to the palpable abnormality (Fig 1). The origin of the mass was undetermined. Subsequently, Magnetic Resonance Imaging (MRI) was obtained that revealed a partially necrotic mass with heterogeneous enhancement (Figs. 2,3 and 4). Due to high concerns of malignancy and unclear origin of the mass, the patient was referred to gynecologic oncology team for further evaluation. The patient was sent for an image guided biopsy and histopathological correlation. The result showed interspersed hypocellular and hypercellular fibrous stroma with STAT6 and CD34 positive immunohistochemical markers suggests SFT (Fig. 5). Later it was surgically resected with clean surgical margins.

Discussion

SFT are infrequent metastasizing mesenchymal neoplasms that can be found in any anatomical part of the body with both pleural and extra pleural distribution, but most originate from the visceral pleura [1]. It accounts for less than 2 % of all soft tissue tumors and is extremely rare in the urogenital tract. Age at presentation varies between fourth and sixth decades of life with slightly high predominance in men. Approximately 18 cases have been reported occurring in the bladder with only 2 cases among females [2,3]. The first 5 cases of SFTs in mediastinum and pleura were reported in 1931 by Klemperer. In 1997, the first case in the urinary tract was reported [4]. It

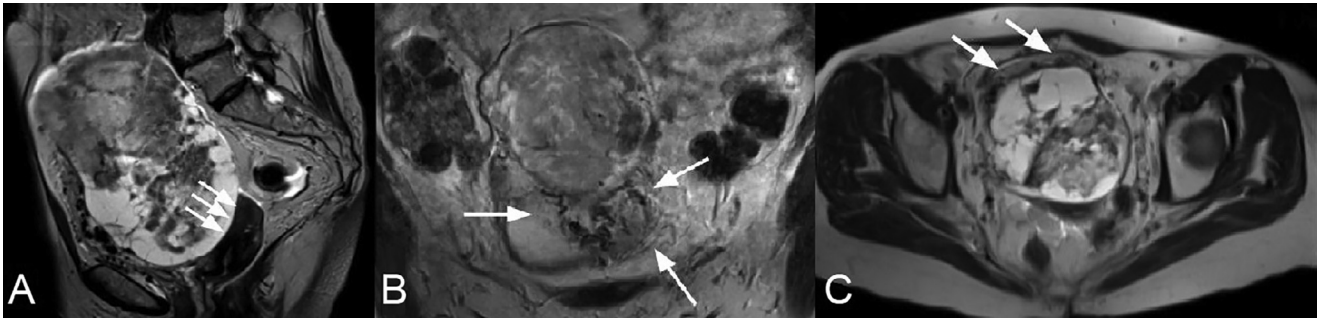


Fig. 2 – Sagittal T2 weighted image (A) demonstrates a large central pelvic mass, with heterogeneous signal that is abutting the anterior uterine wall without definitive invasion (arrows). Coronal T2 weighted image (B) demonstrates the mass in close relation with the bladder wall with enlarged vessels crossing the space between the mass and the bladder wall, suggestive of bladder origin or bladder invasion by the mass (arrows). Axial T2 HASTE image (C) demonstrates again the close relationship between the anterior mass wall and the bladder (arrows). Note that the mass was separate from the adnexa.

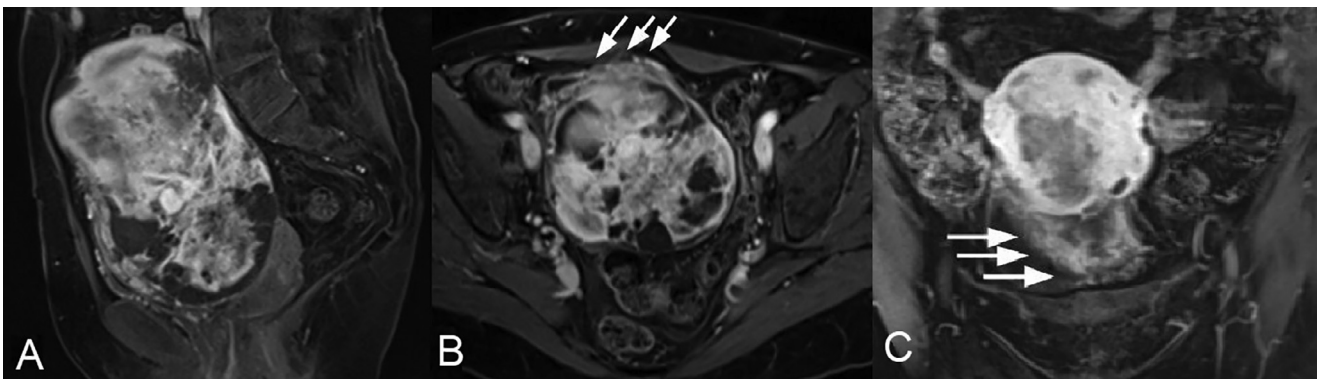


Fig. 3 – Sagittal (A), axial (B) and coronal (C) T1 VIBE post contrast sequences demonstrate heterogeneous enhancement of the mass with areas of solid enhancement and necrosis. Axial and coronal post contrast images (B and C) demonstrate the close relationship between the mass and the left lateral aspect of the bladder dome (arrows).

has an indolent biologic behavior that is independent of its size. Symptoms are mostly related to tumor volume and the extent of local invasion including dysuria, voiding difficulty, urinary urgency, hematuria and lower abdominal discomfort. Most tumors have a benign clinical course but 10%-20% can present rather aggressively. The factors associated with increased likelihood of malignancy include infiltrative borders with regions of necrosis, high mitotic count, tumor size >10 cm, and poor histology [5]. Additionally, it can cause paraneoplastic syndromes, like hypoglycemia secondary to insulin like growth factor [6]. Risk stratification into low, intermediate or high risk of metastasis is done based on scoring systems that account for age at diagnosis, tumor size, mitotic count and tumor necrosis.

Metastasis and local recurrence are uncommon after surgical resection with negative margins. However, when metastasis occurs, it most commonly involves lungs and pleura [7].

Radiological imaging is the crucial for diagnosis of SFTs. Imaging findings can be nonspecific, but they raise concern for a potentially malignant mass that warrants further work up. CT and MRI scans help to identify hypodense areas caused by

necrotic, myxoid or cystic degeneration. On X ray sometimes it appears as a moderately radio-opaque mass [8]. On ultrasound SFT generally presents a variable pattern of echogenicity described as hypochoic mass with relatively well-defined margins. On CT scan it shows as heterogenous enhancing mass with scatter areas of associated cysts, regions of hemorrhage, or necrosis.

On MRI, SFT shows predominant low or intermediate signal intensity on both T1- and T2- weighted images, which is believed to be related to high content of fibrous collagenous tissue, hypocellularity, and relatively small numbers of mobile protons. The low signal intensity on T2 weighted images may be characteristic. Recent onset and malignant fibrous tissue tend to show high signal intensity on T2 weighted images. The preoperative diagnosis of benign lesions using either particular imaging findings or imaging guided biopsy is important to avoid unnecessary radical resection [9].

There are few descriptions in the literature regarding the PET manifestations of SFT. We have found that benign SFT exhibit low-grade activity at PET and that malignant SFT tend

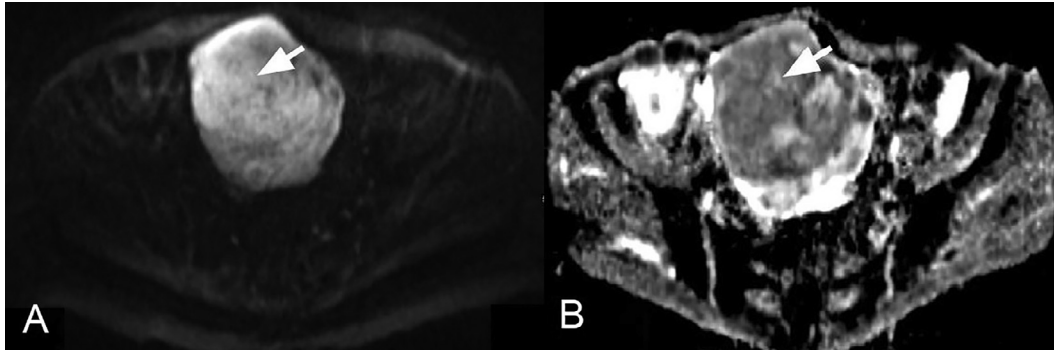


Fig. 4 – Axial diffusion B800 diffusion image (A) and corresponding ADC map (B) demonstrate significant increase in the diffusion signal of the lesion (arrow) with corresponding areas of low ADC (arrow) suggesting hypercellularity of the mass.

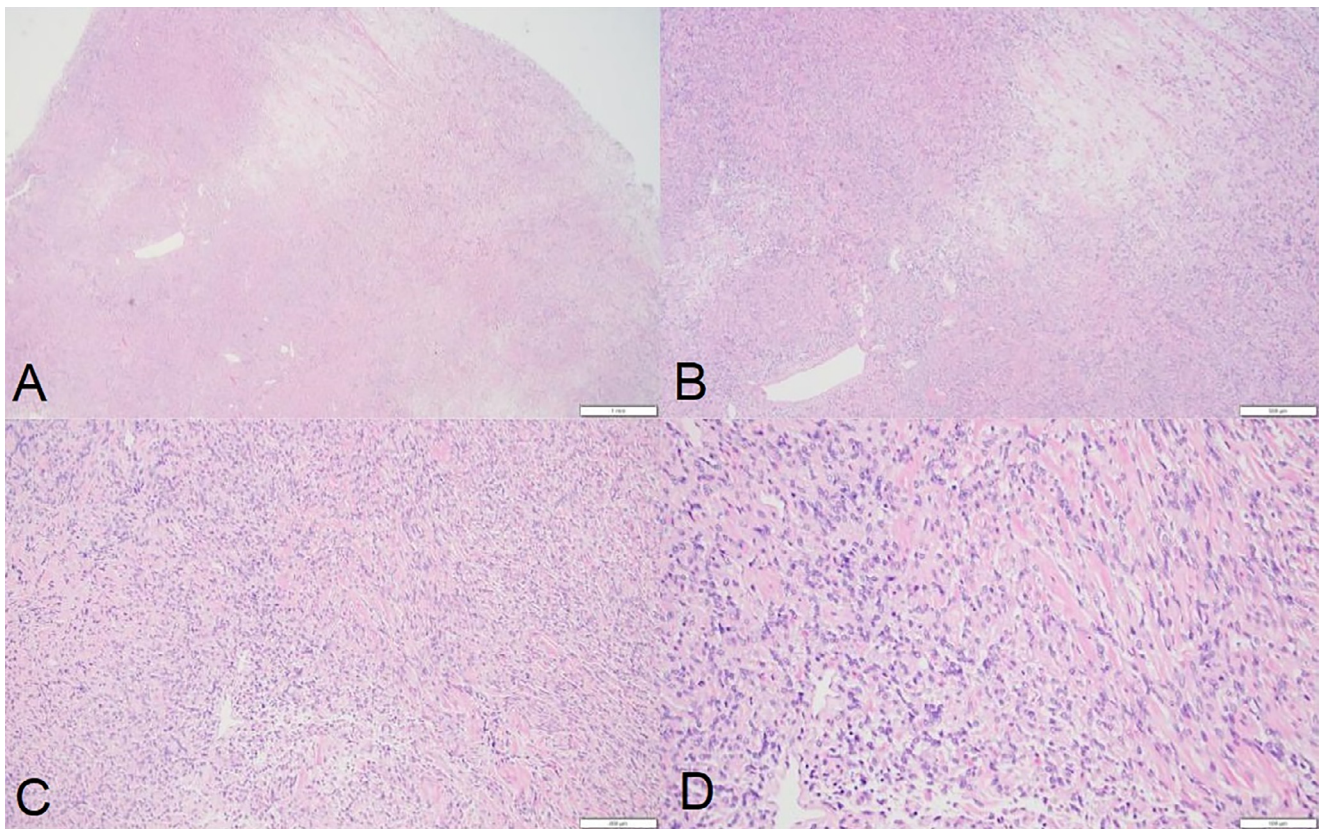


Fig. 5 – There are alternating zones of hypocellular stroma and hypercellular areas of fibrous stroma in 1 mm (A), 500 μm (B), 200 μm (C) and 100 μm images. The tumor cells are small and oval to spindled with limited cytoplasm, juxtaped with scattered thick fibers of collagen and branching vessels. The immunohistochemical profile (STAT6 positive and CD34 positive, see below for details) and the morphology are in keeping with classifying this tumor as a solitary fibrous tumor.

to be strongly hypermetabolic and homogeneous. Furthermore, the presence of multiple SFT and high-grade 18F-FDG metabolism at PET should raise the suspicion of a malignant variety of SFT [10].

The differential diagnosis is broad and includes many solid tumors like hemangiopericytoma, sarcoma, leiomyoma, nodular fasciitis, inflammatory myofibroblastic tumor, fibromatosis and benign peripheral nerve sheath tumor [11].

SFTs involving the urinary bladder and thus should be differentiated from the primary bladder neoplasm based on particular imaging findings. Transitional cell carcinoma on MRI shows intermediate in signal intensity on T2-weighted images, in contrast to benign SFT which shows low signal on T1 and T2 weighted sequences. The transitional cell carcinoma also has predilection to involve ureters and also involve renal pelvicalyceal system as secondary site [12].

Macroscopically, SFT of the bladder is well circumscribed, tan colored mass that may be encapsulated by a thin and vascular pseudocapsule. It is slow growing polypoid intraluminal or submucosal mass with or without extension into the perivisceral fat. Histologically under light microscope, mesenchymal neoplasms are composed of predominantly fibrous lesions containing proliferation of fusiform tumor cells (spindle shaped tumor cells) with large collagenized areas alternately distributed between the dense region (less collagen), and the loose region (abundant collagen), and hyalinized vessels [6]. One of the most common patterns consists of alternating hyper and hypocellular areas with a hemangiopericytoma like appearance [9].

Immunohistochemistry helps to differentiate SFTs from other microscopically similar neoplasms. The most consistently positive tumor markers in SFT are CD34, CD99, Bcl-2 and STAT-6 [13]. STAT6 is a highly sensitive and specific protein that has demonstrated 90–100% positivity in SFTs.

There is a reported role of immunohistochemistry in determining the malignant potential of the SFTs through biomarkers such as p53 and Ki-67 but this remains controversial [14,15].

Surgical resection of the tumor mass with curative intent is the mainstay of treatment achieved using different methods including transurethral resection and partial cystectomy. Chemoradiation therapy is utilized in cases of nonresectable tumors. Long term follow-up is recommended as bladder SFTs with sarcomatoid changes have been reported [1]. Prognosis is considered favorable after complete surgical resection with evidence of 5-year survival.

Conclusion

SFT of the bladder is a rare entity. It is usually benign with a favorable prognosis but can be malignant in a subset of cases. It is imperative for diagnostic radiologists to familiarize themselves with the imaging findings of this tumor. This will help in making an early diagnosis and therefore, guide management. Due to the rarity of the tumor, there are no formal guidelines available for follow up. However, due to risk of recurrence of the tumor, follow up with physical exam and imaging such as abdominal CT should be performed at 6 monthly intervals.

Ethical statement

The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Patient consent

Verbal consent was obtained from patient during the hospital stay during surgery.

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