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A case of large, benign, solitary fibrous tumour of the prostate causing obstructive uropathy

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Introduction

Solitary fibrous tumour (SFT) is a rare fibroblastic mesenchymal tumour with a characteristic histology of a 'patternless', storiform or herringbone distribution of spindle cells within a collagenous stroma. It was initially described in 1931 as occurring commonly in the pleura.[1] During the last few decades, it has been recognised in various other extrathoracic sites, including the urinary tract.[2,3] Conventional SFT is considered as an intermediate, rarely metastasising neoplasm with variable clinical course.[4] A more aggressive form is the malignant SFT, which recurs and metastasises more frequently. Recently another variant has been described as dedifferentiated SFT, in which a sudden transition from a conventional SFT to a high-grade sarcoma may occur.[5] SFTs originating from the prostate gland are rare with fewer than 40 cases reported in the literature.[6–8] SFT of the prostate shows a wide spectrum of morphologic variation.[9,10] Its exact biological behaviour and predictability of long-term outcome are not well understood because of its rarity and small numbers reported.

Case report

A 54-year-old man presented with poor urinary stream, urinary frequency, urgency and lower abdominal discomfort of two years' duration. Examination revealed a large, firm and smooth intraabdominal lump arising from the pelvis. His prostate gland was enlarged and clinically benign. Serum prostate-specific antigen (PSA) level was 1.19 ng/ml. Abdominal ultrasonography and contrast-enhanced computed tomography (CT) of the abdomen and pelvis confirmed the large mass to be in continuity with the prostate gland (Figure 1). Urethra was elongated and bladder was compressed with bilateral hydronephrosis. Serum creatinine level was 1.7 mg/dl. Flexible cystoscopy revealed a compressed, elongated and distorted urethra, massively enlarged lateral lobes of the prostate, normal bladder mucosa and displaced ureteric orifices.

Transrectal ultrasound (TRUS)-guided core biopsy of the prostate showed features of a benign spindle cell tumour composed of uniform spindle cells arranged in a vague storiform pattern with haemangiopericytomatous vessels. The neoplastic cells showed monomorphic oval nuclei and moderate eosinophilic cytoplasm. Mitotic figures were not identified and there was no nuclear atypia or tumour necrosis. Neoplastic cells were positive immunohistochemically for CD34, CD99 and Bcl-2. In addition, CD34 accentuated the haemangiopericytomatous vascular pattern. The tumour was negative for epithelial membrane antigen, desmin, S-100 and cytokeratin. Ki-67 cell proliferative index was 3%. A possible diagnosis of SFT of the prostate with benign features was made.

Enucleation of the tumour mass similar to the Millin open retropubic prostatectomy was performed to remove a mass measuring 14.5 cm × 9 cm × 7 cm in diameter. Through a Pfannenstiel incision, the tumour was accessed. A transverse incision was made over the stretched prostatic capsule after ligating the longitudinally crossing blood vessels. Then a clear plane was found between the tumour capsule and the prostatic tissues and this enabled complete

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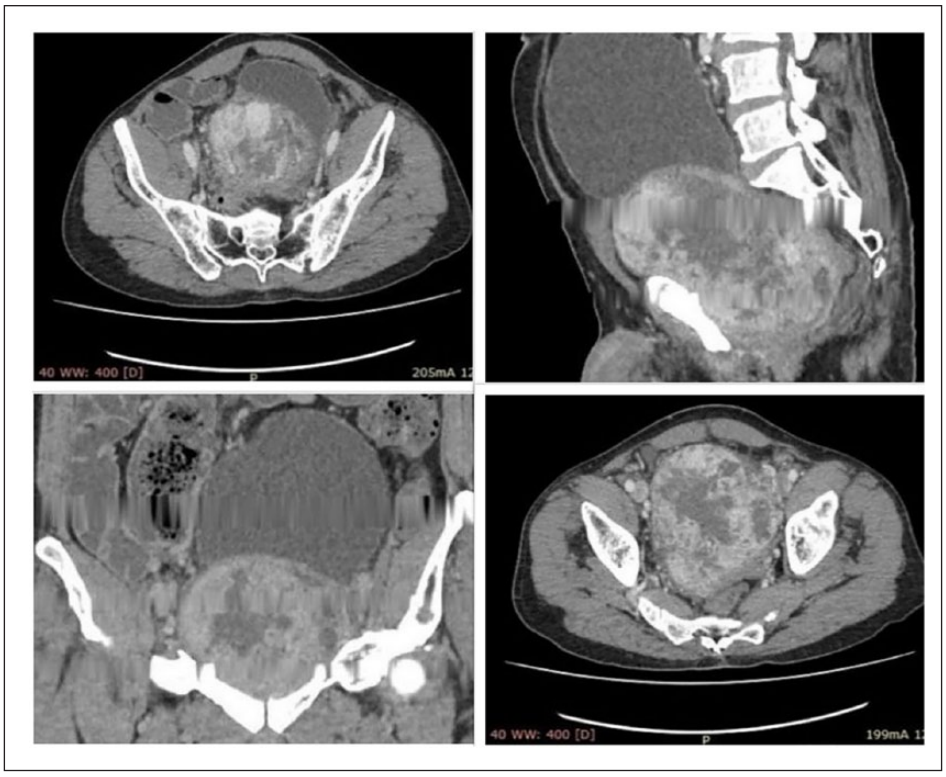

Figure 1. Computed tomography images showing a large mass in the area of the prostate with distended bladder and compressed urethra.

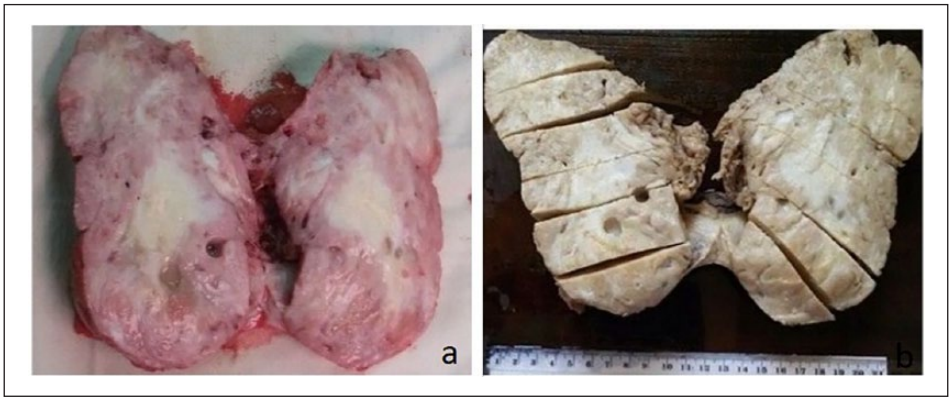

Figure 2. Macroscopic images of the (a) fresh specimen, and (b) fixed specimen showing tan-coloured surface without areas of haemorrhage or necrosis.

enucleation of the mass. Many large blood vessels supplying the tumour were ligated during the process. Blood loss was estimated to be 2 l, and two units of packed cells were transfused. The duration of the surgery was two hours. Postoperative period was uneventful and the urethral catheter was removed after two weeks.

On cut section, the tumour showed tan-coloured zones and cystic spaces without haemorrhagic or necrotic areas (Figure 2). Most of the tumour was surrounded by a fibrous capsule. The histology and immunohistochemistry confirmed SFT of the prostate without nuclear pleomorphism, detectable mitotic activity or necrosis (Figure 3). One year after the operation, the patient is symptom free and continent without any recurrence of the tumour. His serum creatinine is 0.9 mg/dl.

Discussion

SFT is a rare neoplasm of mesenchymal origin that should be considered in cases of prostatic tumours with a spindled histology.[4,10] However, the exact diagnosis is challenging as most mesenchymal tumours of the prostate contain spindle

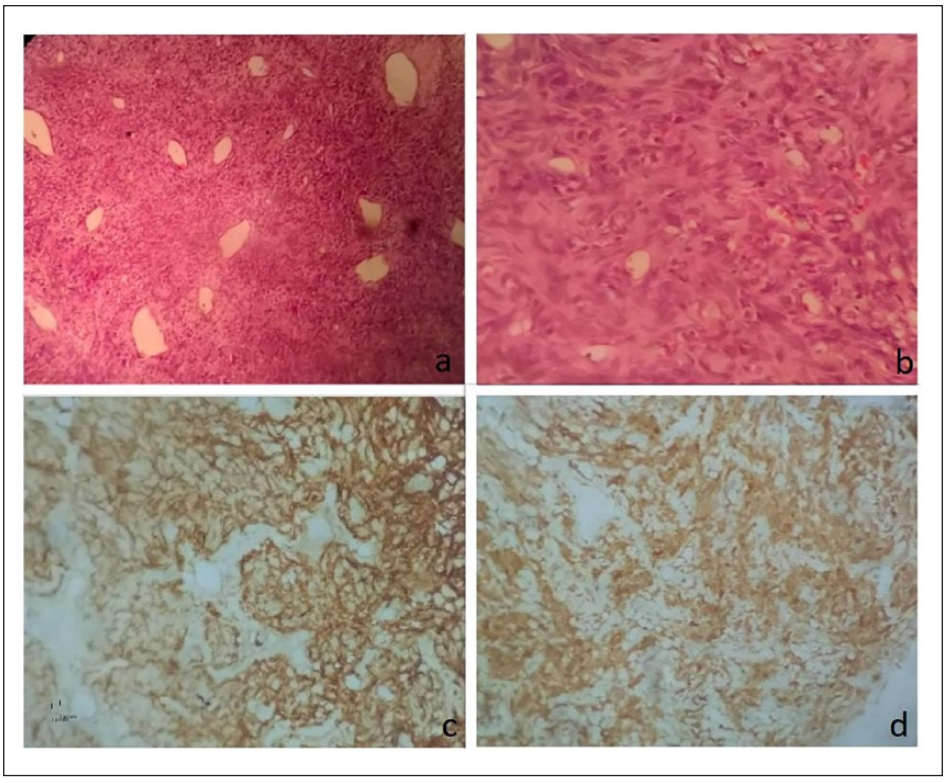

Figure 3. Microscopic images of the spindle cell tumour (a) haematoxylin and eosin (H&E) stain (×100), (b) H&E stain (×400), (c) CD99 positivity (×400), and (d) CD34 positivity (×400).

cell morphology with a lot of overlapping histological features. They are consistently associated with *NAB2-STAT6* gene fusions and are thought to arise from CD34-positive dendritic interstitial cells.[6,9,11] Prostatic SFTs have a varying biological behaviour ranging from benign to malignant. About 20% to 30% of SFTs originating from organs all over the body develop recurrences or distant metastases.[6,7] Despite nearly half of the reported prostatic SFTs showing malignant features in histology, recurrences and distant metastases have been reported rarely.[6] Surprisingly, the extent of surgical resection does not show a clear correlation with recurrences. Whether the histological features correlate well with actual outcome and behaviour of prostatic SFTs is also not well understood because of the small number of cases and short follow-up periods. Benign SFTs show a clear boundary and expansive growth compared to undefined boundary and invasive growth of malignant SFTs. Immunohistochemical analysis of basic fibroblast growth factor and Ki-67 expression may help in differentiating benign SFTs from malignant. However, because of the rarity of cases, molecular markers helpful in diagnosis and prognostication are not yet defined. Hence targeted therapy for malignant SFTs is still speculative.

The reported cases of SFT of the prostate have an age range of 33–78 years with a mean of about 55 years.[6–8] The common clinical features of prostatic SFTs include urinary retention, urinary frequency, dysuria and haematuria. Hypoglycaemic episodes have been reported rarely in the SFT of serosal origin due to the production of insulin-like growth factors but not with prostatic SFTs.[12] The predominant symptoms of our patient were poor stream and urinary frequency. Obstructive uropathy with upper urinary tract dilatation and renal impairment due to a prostatic SFT, as seen in our patient, is unusual and has not been reported before.

Reported tumour sizes range from 5 cm to 25 cm with normal serum PSA levels in almost all cases.[6,13] Imaging modalities including ultrasonography and CT/magnetic resonance imaging (MRI) of the abdomen and pelvis are used to confirm the organ of tumour origin, staging of the tumour and detection of associated complications such as upper urinary tract dilatation. Urinary tract ultrasonography will show an echogenic mass in the prostate and is nonspecific. Characteristically CT imaging shows a mass of low density while MRI images show T1-weighted low-density mass and T2-weighted high-density mass. In the presence of these imaging findings, TRUS-guided core biopsy of the prostate should be performed as SFT is likely. Nevertheless, there are no characteristic features in imaging to differentiate SFT from other tumours of the prostate.[10] Differential diagnoses include prostatic adenoma and adenocarcinoma, leiomyoma, leiomyosarcoma, stromal tumours of uncertain malignant potential, inflammatory myofibroblastic tumour, gastrointestinal stromal tumour and other spindle cell

tumours arising from adjacent organs such as the bladder, seminal vesicles and rectum.[14]

Diagnosis can be made by histology and immunohistochemistry of the lesional biopsies. Macroscopically, SFTs show homogeneous white to tan-coloured masses with areas of cystic changes and may have a capsule similar to our case. Malignant variants can have additional zones of haemorrhage, necrosis and infiltrative borders. Microscopy is characterised by spindled fibroblastic cells showing a patternless distribution within collagenous stroma containing large, branching haemangiopericytic vessels. Malignant histology may show additional features of hypercellularity, focal pleomorphism, mitotic index greater than 4 per 10 high-power field and necrosis. Immunohistochemistry shows strong CD34 positivity and diffuse expression of CD99 and Bcl-2, while it is negative for CD31, desmin, h-caldesmon, S-100 protein, and cytokeratins.[4,11] *NAB2-STAT6* fusion is a more accurate immunohistochemical marker with a greater sensitivity and specificity.[6,9] Hence nuclear expression of *STAT6* is the most reliable marker for differentiating SFTs from histological mimics. Fluorescence *in situ* hybridisation or polymerase chain reaction confirmation of the *NAB2-STAT6* fusion is available for uncertain cases.[9]

Current understanding of surgical strategy is based on the unpredictability of clinical behaviour of the tumour. Thus, complete tumour resection with negative surgical margins has been attempted in previously reported cases.[6,7,10] Since our patient had a very large tumour, it was technically difficult to perform a radical prostatectomy. We decided to perform enucleation of the tumour mass similar to a Millin open retropubic prostatectomy as we expected the postoperative morbidity and mortality to be unacceptably higher after radical prostatectomy, especially in relation to urinary incontinence and erectile dysfunction. Surprisingly, even after radical forms of surgery recurrences of the tumour can occur; hence long-term follow-up is mandatory in all cases of SFT.[6] This would help to identify recurrences early, enabling less-invasive surgery. After one year of follow-up, our patient maintains normal erectile, voiding and renal functions with no recurrences or metastases. Long-term follow-up and documentation of all SFTs of the prostate are needed to establish prognostic patterns to identify tumour behaviour and to develop appropriate management strategies.

Conflicting interests

The Authors declare that there is no conflict of interest.

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Ethical approval

Because this is a case report and the patient has given informed written consent for publication of the article, the ethics committee of Colombo South Teaching Hospital decided that its formal approval is not necessary for publication of the case report.

Informed consent

The patient gave informed, written consent for publication of this case report. The document is available from the editors for perusal.

Guarantor

AMA.

Contributorship

AMA and CPS conceived the idea of reporting this case, GPUDS, ALAMCA, MGSRK, BB and AMA were involved in the management of the patient, AMA and GPUDS wrote the manuscript, CPS and WMMAW modified the manuscript and all authors approved the final manuscript.

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