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Solitary fibrous tumour of the urinary bladder – A rare and potentially malignant neoplasm

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ABSTRACT

Solitary fibrous tumours (SFTs) of the urinary bladder are a rare mesenchymal neoplasm that occasionally has malignant potential. The tumour is characterised by haphazardly arranged spindle-shaped to ovoid cells, with a prominent, branching, thin-walled, dilated vasculature and NAB2-STAT6 gene rearrangement. While most SFTs are indolent in nature, difficulty arises predicting which SFTs are potentially malignant. There are now validated risk stratification tools to help identify which SFTs are likely to metastasize and help clinicians determine management. The mainstay of treatment for SFTs remains surgery, with emerging evidence in the combined use of surgery and radiotherapy.

1. Introduction

Solitary fibrous tumours (SFTs) are rare neoplasms of mesenchymal origin.[1] The morphological features of SFTs were first described in 1931, with the term 'solitary fibrous tumour' first used in 1951.[2] Haemangiopericytoma was a tumour of similar morphological features described in 1942,[2] and in 2013 haemangiopericytoma and SFTs were combined into a single tumour classification.[1,2] While initially described to be of pleural origin, awareness of extra-pleural solitary fibrous tumours is increasing and have been found in areas such as the prostate, seminal vesicle, orbit, nasal septum, and abdominal organs.[2,3] Majority of SFTs are usually benign, however, a small proportion of these tumours are malignant and can metastasize.[2]

There have been less than 40 cases reported in the literature on SFTs in the urinary bladder, and standard treatment of SFTs has been with surgery. The combined use of radiotherapy and surgery has been used to treat SFTs,[4] however prospective evidence is lacking. We discuss a case of a patient with a SFT of the urinary bladder, his management, and a review of the literature.

2. Case presentation

A 68-year-old male was initially investigated for lower urinary tract symptoms (LUTS). His past medical history included hypertension, a cerebrovascular accident, and asbestosis with a right-sided pleural effusion. The patient had a very enlarged prostate with a prostate specific antigen (PSA) blood test of 0.21μg/L and underwent transrectal ultrasound (TRUS) guided prostate biopsies. The histopathology revealed benign prostatic hyperplasia. He was started on a combination of dutasteride and tamsulosin medication and was discharged to his general practitioner (GP) after his LUTS improved.

He was referred again for worsening LUTS five years later. A CT and FDG-PET scan performed for his pleural effusion showed a large mass within the bladder, unable to differentiate between a bladder tumour or prostate median lobe. Flexible cystoscopy confirmed this was an enlarged median lobe and the patient went on to have a transurethral resection of prostate (TURP).

The histopathology from his TURP returned showing atypical solitary fibrous tumour of the prostate, which was characterised by a cellular spindle shaped lesion comprised of haphazardly arranged spindle cells (Figs. 1 and 2). There was a high mitotic count (7–10 per high powered field [HPF]), increased cellularity and mild pleomorphism, with diffuse staining for STAT6.

As the tumour showed high risk features such as large tumour size on imaging, high mitotic count, nuclear pleomorphism and increased cellularity, the recommendation from the Urology-Oncology

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Multidisciplinary Meeting was for the patient to undergo a cystoprostatectomy, however he declined this initially.

Over the next two years, the tumour continued to enlarge and subsequently obstructed his left kidney. After further discussions, the patient agreed to undergo cystoprostatectomy, pelvic lymphadenectomy and ileal conduit formation. Final histopathology from his cystoprostatectomy revealed an atypical solitary fibrous tumour of the bladder measuring 9.5cm, clear of margins. Specifically, there was a large cellular mesenchymal spindle cell lesion expanding the bladder wall, subadjacent to the lamina propria. The highly cellular population of spindled cells showed 3 to 4 mitoses per 10HPF, with focal areas of necrosis. The tumour showed positive expression for STAT6. His prostate did not show any evidence of solitary fibrous tumour but did contain Gleason 3 + 3 = 6 prostate cancer, clear of margins. All lymph nodes were negative for metastatic disease.

His follow-up CT scans up to two years post-operatively has shown no evidence of local recurrence or metastatic disease.

3. Discussion

SFTs are defined by the World Health Organisation (WHO) as a fibroblastic tumour, characterised by prominent branching, thin-walled, dilated vasculature and NAB2-STAT6 gene rearrangement, with intermediate biologic potential (relatively indolent with low risk of metastasis).[1] They have been found at almost any anatomical site and are more common at extra-pleural sites with only about 30% of SFTs occurring in the pleura.[1]

The gender distribution of SFTs is equal in both men and women, with a peak incidence between 40 and 70 years of age.[1] The tumours are generally asymptomatic, painless, and slow growing, and often an incidental finding on imaging for other conditions.[1] As the tumour enlarges, patients may develop symptoms related to mass effect on other organs.[1] Currently there are no known risk factors that influence the development of SFTs.[1]

Morphologically, SFTs are comprised of haphazardly arranged spindled to ovoid cells.[1] They have indistinct, pale, eosinophilic cytoplasm within a variably collagenous stroma and branching staghorn-shaped blood vessels.[1] Most SFTs have low mitotic counts, without substantial nuclear pleomorphism or necrosis.[1] There are various subtypes of SFTs including lipomatous, giant-cell and anaplastic SFTs that each have unique histopathological features.[1] The hallmark immunohistochemical feature of SFTs is strong and diffuse expression of CD34 and STAT6 expression, with the fusion of the NAB2 and STAT6 gene.[1]

SFTs have a recurrence rate of 10–30% (either local or distal recurrences).[1] Traditionally tumours that had high mitotic counts, large tumour size, nuclear pleomorphism, or atypical, necrosis or infiltrative growth patterns were considered as having malignant potential, however the development of multivariate risk stratification models are now able to predict risk of metastases more accurately.[2] Demicco et al. developed a risk stratification tool that uses a combination of pathological features (mitotic count and necrosis) along with clinical data (patient age and tumour size) to risk stratify tumours into low, intermediate, or high risk and subsequently the patients' risk of metastatic disease at 5 and 10 years[5] (Table 1).

Surgery combined with radiotherapy (either pre- or post-operative) has been performed with good outcomes in local recurrence and distant metastases,[4] however evidence is only retrospective or anecdotal, and prospective studies are limited by the rarity of this disease.

4. Conclusion

SFTs are a rare and usually indolent neoplasm, but care must be taken as they can infrequently metastasize. Using a risk stratification tool can assist with determining which patients should have more aggressive treatment. Surgical resection is still the primary treatment option, however future studies may further consolidate the use of neoadjuvant or adjuvant radiotherapy.

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Declarations of conflicts of interest

None.

APPENDIX

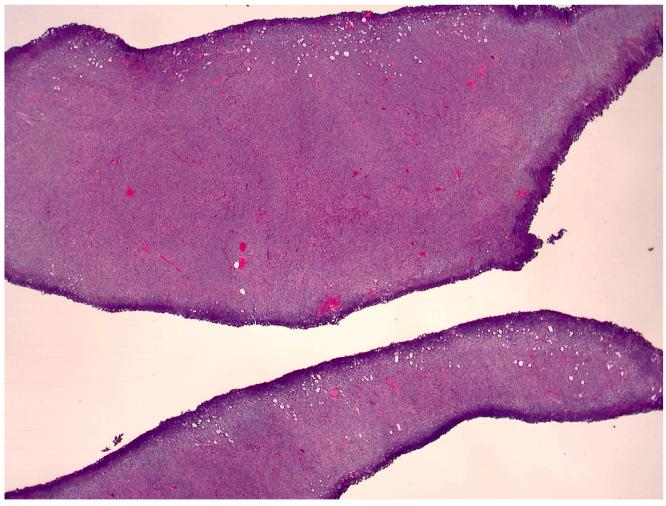

Fig. 1. Histological section demonstrating prostate SFT showing highly cellular tissue (Hematoxylin & Eosin stain, 4x magnification).

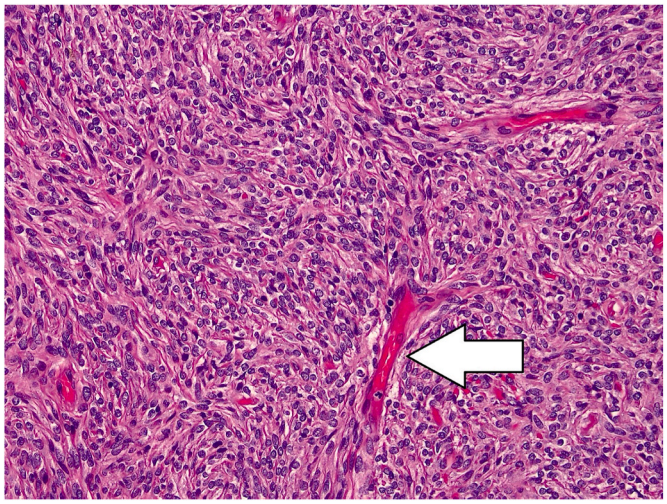

Fig. 2. Histological section demonstrating prostate SFT with closely packed spindled cells and prominent thing-walled branching blood vessels (red arrow) (Hematoxylin & Eosin stain, 20x magnification).

Table 1
Demicco et al. Risk stratification model for development of metastases in SFTs

Risk Factor		*Score*
Age		
	<55	0
	≥55	1
Tumour size (cm)		
<5		0
5 to < 10		1
10 to < 15		2
≥15		3
Mitotic count (/10 high-power fields)		
0		0
1–3		1
≥4		2
Tumour necrosis		
<10%		0
≥10%		1
Risk Class		***Total Score***
Low risk		0–3
Intermediate risk		4–5
High risk		6–7
Metastasis-Free	***5-years***	***10-years***
Low risk	100%	100%
Intermediate risk	90%	90%
High risk	27%	–

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