

Spindle cell tumours of the ureter: Operation or observation?

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ABSTRACT

We describe a rare case of inflammatory spindle cell tumour of the ureter in a patient who presented with renal colic and macroscopic haematuria. Pyeloscopy revealed a partially obstructing mass at the proximal right ureter which confirmed a myofibroblastic tumour on biopsy. Radical nephrectomy was performed which confirmed a spindle cell tumour of the ureter confined to the resection margins. Follow-up imaging in 12 months did not illustrate recurrence or metastasis. The decision to perform a nephrectomy was due to the limited experience with this tumour. Reports illustrate that this tumour is unlikely to metastasize, and thus be managed conservatively.

1. Introduction

Spindle cell tumours are an extremely rare subset of inflammatory tumours.¹ Also known as Inflammatory myofibroblastic tumours/pseudotumours (IMT) they are known to occur in the lung, spleen, heart, and mesentery.² Within the urinary tract they have been described mainly in the bladder and kidney and are exceedingly rare to be within the ureter.² To date only 11 other cases have been published of these tumours occurring within the ureter, of which 6 are in children.³

2. Case presentation

A 38-year-old male presented to our emergency department with sudden onset right flank pain and macroscopic haematuria for 48 hours duration. He denied any infective or systemic symptoms. There was no previous history of stone disease, recurrent UTIs, lower urinary tract symptoms (LUTs) or childhood developmental issues. He had a significant past medical history of smoking and recreational drug use. Physical examination revealed normal observations with a soft abdomen and right loin tenderness without guarding. A full ward test (FWT) illustrated microhaematuria and proteinuria and culture were negative for infection. His haemoglobin (Hb) was 135 g/L and an eGFR (estimated glomerular filtration rate) was 70 ml/min/1.73 m² on presentation (normal >90 ml/min/1.73 m²). Urine cytology was negative for urothelial malignancy. A contrast enhanced CT (computed tomography) abdomen and pelvis in the delayed phase was performed illustrating

moderate right hydronephrosis, delayed nephrogram with a density within the proximal ureter (Fig. 1).

The patient subsequently underwent a rigid cystoscopy, right retrograde pyelogram and a ureteropyeloscopy and biopsy with revealed a filling defect in the proximal ureter consistent with the CT images. Direct visualisation of the lesion revealed a partially obstructing mass arising from the urothelium, measuring 2cm in maximum dimension, while also not displaying the typical features of a urothelial carcinoma (Fig. 2). Multiple biopsies of the lesion were taken which revealed an inflammatory myofibroblastic tumour. Subsequently the patient underwent a laparoscopic right nephrectomy with the ureter dissected down to the iliac vessels. The patient was discharged day 2 post the operation without complication.

Histopathology illustrated a myofibroblastic spindle cell tumour of the proximal ureter, with no evidence of invasion, clear of all margins, no features to suggest IgG4-related disease, with a TNM staging pT1aNXMO. After 12 months the patient underwent a surveillance CT of the abdomen which did not illustrate any recurrence or distant metastasis (Fig. 3).

3. Discussion

The aetiology and pathology of spindle cell tumours remains unclear, with some studies suggesting a chronic inflammatory response to trauma and infection,¹ recent studies have also suggested a genetic predisposition to these tumours.² However, in our case there was no

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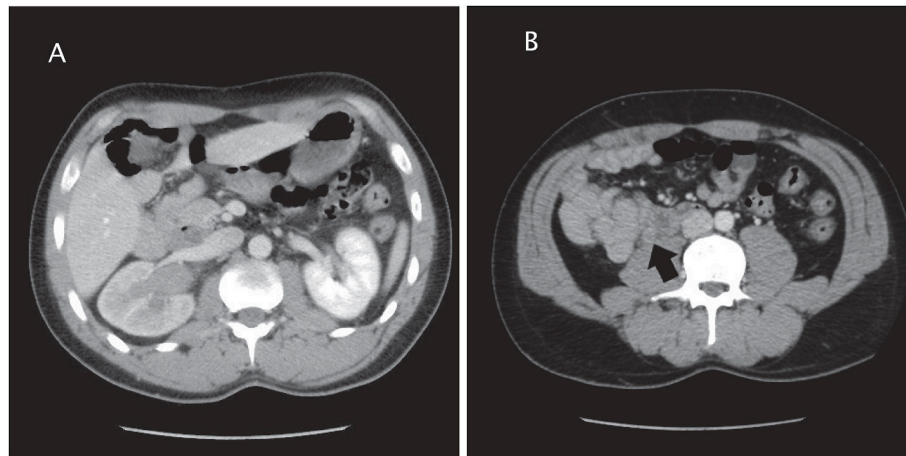


Fig. 1a. Cross-sectional CT Images of the abdomen & Pelvis illustrating the moderate right hydronephrosis

Fig. 1b: delayed nephrogram illustrating the density within the proximal ureter as highlighted by the black arrow.

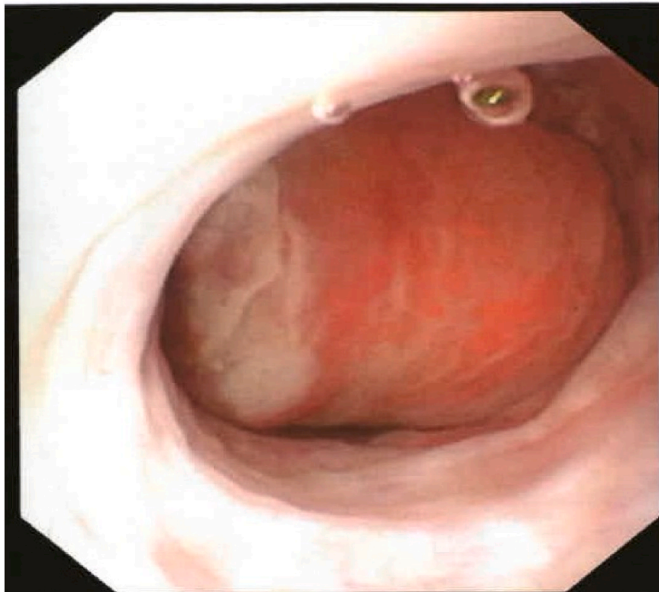


Fig. 2. Intra-operative image that shows the partially obstructing lesion arising from the urothelium.

history of recurrent infections, trauma or instrumentation, and no history of viral infections such as Herpes virus and Ebstein Bar Virus as has been described in published literature.

Clinical presentations of such primary tumours within the ureter are like those seen with urothelial carcinomas, including flank pain and frank haematuria.² Other benign conditions such as fibromatosis, fibrous histiocytoma and chronic cystitis cystica may be considered as differential diagnoses.⁴ Detailed pathologic examination of tumour biopsies is of utmost importance to identify this condition as sarcomas potentially may be mistaken on histological appearance.⁴

Like all urothelial tumours, CT IVP (intravenous pyelogram) remains the optimal imaging techniques to help identify and define the tumours, however endoscopic visualisation leading to biopsy and histological evaluation remains the only way to achieve a definitive diagnosis⁴

Spindle cell tumours generally are considered a neoplasm with low malignant potential and low recurrence rates.¹ The recurrence rates are related to large size, abdominopelvic location, and occurrence in the elderly, with the literature reporting a recurrence rate from 2 to 25%. Within the urinary tract recurrence appears potentially less common. Li

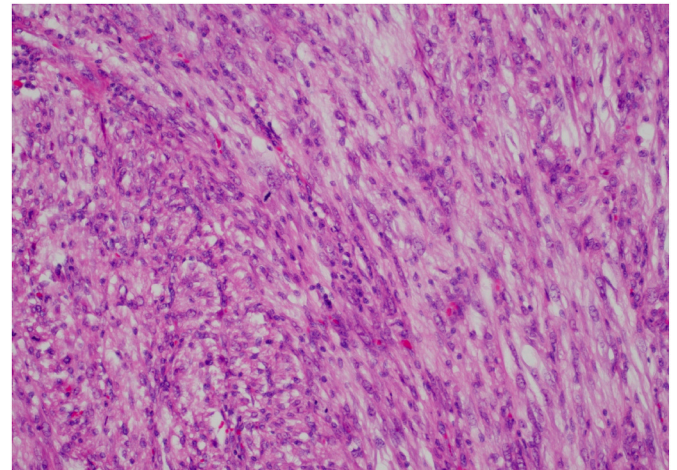


Fig. 3. High power view revealing elongate and spindled tumour cells arranged in vague fascicles, H&E stain, magnification $\times 200$.

et al. reported a case series of 11 cases of urinary bladder spindle cell tumours, with no evidence of recurrence or distant metastasis at routine surveillance.²

Despite the indolent nature of this disease, extirpative surgery remains standard of care. In all 11 cases within the literature, plus our case above, radical surgical management was offered, ranging from segmental uretectomy and ureteric reimplantation to radical nephrouretectomy.² Coffin et al. hypothesized that urothelial spindle cell tumours are indeed a low-grade inflammatory sarcoma, with other authors also describing similar histological features between the two tumour types.^{4,5}

4. Conclusion

Currently all evidence points towards spindle cell tumours of the urinary tract presenting little to no malignant potential, however some studies have described histologic similarities to low grade sarcomas of the ureter. Given most of the tumours occur in a young population, radical nephrectomy/nephrouretectomy will achieve a curative outcome, but with potential for future morbidity. Therefore, in the appropriate patients nephron conserving management may potentially be considered depending on the size of the tumour at diagnosis. Procedures like uretectomy and ureteric reimplantation, endoscopic enucleation of the tumour and indeed close clinical and radiological

observation have the potential to lessen the impact on patients of this diagnosis.

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Patient consent

- Obtained

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