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Retroperitoneal Desmoid-type Fibromatosis of the Distal Ureter: A Discussion of the Imaging Differential Diagnoses

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Abstract

A 42-year-old man in remission for chronic myeloid leukaemia (CML) was found to have worsening renal function. Computed Tomography (CT) of the urinary system showed a soft tissue lesion of the right distal ureter which had an atypical appearance for primary malignancy. Due to concerns of recurrence, further evaluation with contrast-enhanced Magnetic Resonance Imaging (MRI) was performed revealing the fibrous nature of the lesion. Considering the resultant obstructive uropathy and the need to exclude recurrence of prior haematological malignancy, resection and histological examination was performed. Resection of the affected segment of the right distal ureter was performed with reimplantion (Psoas Hitch) and Heineke-Mikulicz Bladder reconstruction. Histological examination revealed desmoid-type fibromatosis involving the ureter which is extremely rare. We present the diagnostic process and describe the imaging characteristics through multi-planar CT and MRI images of the tumour obtained prior to resection. A discussion of the imaging differential diagnoses is included with companion images. This entity is important to recognise due to the local aggressiveness of this tumour with propensity for recurrence.

Keywords: Desmoid-type fibromatosis • Atypical ureter lesion • Retroperitoneal tumor • MRI kidneys

Introduction

A 42-year-old man was referred to the Urology department following incidental note of hydronephrosis on ultrasound scan which was done as part of the investigation for worsening renal function. He had a history of chronic myeloid leukaemia (CML), on remission since 2014. He was also known to have a history of left sided renal calculi complicated with urinary tract infection in the past.

The patient was asymptomatic and the physical examination showed no localizing signs. The blood test results showed a decline in the estimated glomerular filtration rate (eGFR) of 70 to 51 mls/min over a period of 6 months. Urine analysis and urine cultures were normal.

Case Presentation

In view of the hydronephrosis seen on ultrasound, CT Urography was performed to evaluate for the cause of obstruction. The CT urography (Figure 1A and 1B) showed a short segment of enhancing mural thickening of the distal right ureter with an aneurysmal dilation of the intralesional segment of ureter. Contrast flow through the affected segment is demonstrated despite upstream hydronephrosis (not shown, also seen on prior ultrasound). The lesion was in contact with the right seminal vesicle. No significant lymphadenopathy was detected. The lesion was deemed to be atypical in appearance for a primary ureteric malignancy due to the intralesional ureteric dilation despite upstream hydronephrosis. As Magnetic Resonance Imaging (MRI) provides superior soft tissue resolution, an MRI Urography was subsequently performed in 2 months after the initial CT to further characterise the ureteric lesion due to the concern of recurrence of haematological malignancy.

On MRI, the lesion showed apparent stability in extent with marked T2w hypointensity (Figure 1C and 1D, white arrow). The affected segment of the right distal ureter showed almost a kinked appearance. On other images not shown, there was uniform contrast enhancement was detected with no restricted diffusion. MRI also showed the lesion being inseparable from the right seminal vesicle, concerning for involvement.

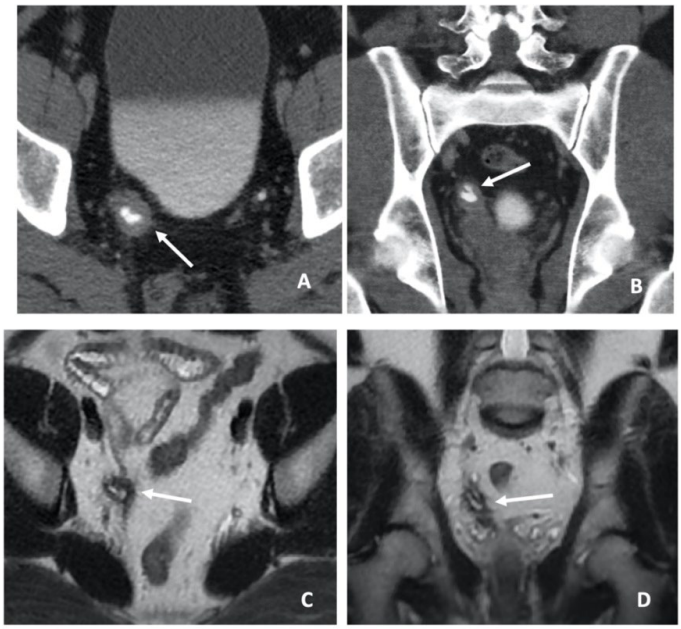

Figure 1. (A & B): Axial and coronal images of CT urography, excretory phase. (C & D): Axial and Coronal MRI of the abdomen & pelvis, T2-weighted images. White arrows indicate the soft tissue lesion encasing the right distal ureter with apparent intralesional ureteric dilation. Note is made of the intrinsic low T2w signal of the lesion.

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Overall, the MRI suggested that the lesion showed signal intensities in keeping with a fibrotic process. Considering that the patient had completed treatment for prior CML, prior sites of extra-nodal disease could have appeared fibrotic after treatment. Due to the recent onset of mass effect and hydronephrosis, an important differential was an extra-nodal recurrence on background of prior fibrotic change. It was important to exclude a potential recurrence of prior haemotological malignancy to avoid further chemotherapy. In view of the lesion causing obstructive uropathy as well as the indeterminate risk of recurrence based on imaging features, we decided to perform an open excision of the right distal ureteric mass with reimplantion of the right ureter (Psoas Hitch) and Heineke-Mikulicz ureteroplasty.

Intraoperatively, the mass was hard and diffuse with no distinct borders. It densely adhered to the posterior denonvillers fascia, right seminal vesicle and right vas deferens as well as the posterior peritoneal reflection. The affected segment of the right ureter, vas deferens and seminal vesicle were resected. The right ureter was subsequently reimplanted (Psoas Hitch) and Heineke-Mikulicz ureteroplasty was performed. All resected specimens were sent for histological analysis.

On histology, the ureteric mass was composed of spindle cells arranged in long fascicles within a myxoid to collagenous stroma (Figure 2A). No nuclear pleomorphism or atypical mitosis was detected. There was diffuse nuclear expression of Beta-catenin (E) (Figure 2B). The specimen showed features consistent with desmoid-type fibromatosis circumferentially involving the ureter and the adjacent seminal vesicle (Figure 2). There was no involvement of the right vas deferens. Oncologic resection was achieved with all margins showing no evidence of tumour involvement. The patient made an unremarkable expected recovery after surgery.

Discussion

In the evaluation of ureteric lesions, desmoid-type fibromatosis is not often considered due to its extremely rare occurrence. Desmoid-type fibromatosis also known as Desmoid tumours are locally aggressive fibrous neoplasms of mesenchymal origin [1]. The reported incidence is 2-4 per million per year, often occurring in the third and fourth decades of life [2]. Established genetic predisposition is seen in Familial Adenomatous Polyposis (FAP) and Gardner Syndrome [2]. These tumours may occur in any part of the body, though those associated with hereditary syndromes tend to occur in the intra-abdominal compartment [3]. Otherwise, the tumours mostly occur in muscle and fascia [4].

The infiltrative and locally aggressive nature of these tumours make pre-treatment imaging important in determining resection potential. While asymptomatic incidental desmoid-type fibromatosis may be observed, often treatment is required due to local mass effect on critical structures, as in the ureter in this case [5]. To date there has been only a handful of reports of ureteric involvement, Lahoud J, et al. [6] reported a patient with urinary obstructive symptoms with a lesion resulting in hydroureteronephrosis, another report showed a patient with spontaneous ureteric rupture and urinoma formation and an incidentally discovered case involving the ureter was found during gynaecological surgery [7,8].

In our patient the imaging findings showed a lesion of the ureteric wall which was unlikely of urothelial origin considering the lack of intra-luminal polypoid components, lack of complete obstruction to the collecting system and with apparent luminal dilation.

Due to the significant history of CML, focal leukemic deposits and infiltration of the ureter was of concern. Pulik M, et al. previously reported two patients who developed desmoid tumours with prior chronic lymphocytic leukemia (CLL), otherwise, there is no known reports or associations of CML and desmoid tumours to date [9]. In this patient, the biochemical and genetic analysis of the patient's recent blood samples suggested good response which reduced its likelihood. Likewise, lymphoma of the ureter was considered due the infiltrative appearance and apparent circumferential thickening. However, it would be unusual for lymphoma involving the ureter to exhibit slow growth over 18 months, considering the prior CT scans performed for an unrelated issue (Figure 3). Furthermore, MRI of lymphomatous involvement would show mild T2w hypointensity compared to adjacent renal cortex, but the lesion was markedly hypointense, corresponding to the usually low T2w signal in fibrous tissue, consistent with desmoid-type fibromatosis. Moreover, lymphoma would show restricted diffusion which was absent. An example of a patient with lymphomatous involvement of the ureter is as depicted in Figure 4.

Fibro-inflammatory lesions that exhibit low T2-weighted signal on MRI were considered. These include IgG4-related disease of the ureter, though this usually presents extensively and bilaterally as seen in previous cases in our institution for example in Figure 5. Erdheim-Chester disease was also considered, however the patient's disease was predominantly unilateral with also no other organ systems involved. In addition, focal ureteric amyloidosis also shows T2w hypointensity. This is a rare entity with prior case reports describing ureteric involvement [10]. However, it was noted that the deposits of amyloidosis would show typical calcification which was absent in our case.

MRI characteristics of desmoid-type fibromatosis include internal T2w low signal intensity bands due to its fibrous nature and inherent collagen bands. The T2w signal may be heterogenous due to inherent variable composition of spindle cells, collagen and myxoid components [11]. On T1w imaging these lesions show intermediate signal intensity, appearing isointense to skeletal muscle. Gadolinium contrast enhancement may be heterogenous [11]. Margins may be ill-defined in up to 50% of tumours [11,12]. These imaging characteristics often differ from the urothelial carcinomas, thus alluding to atypical ureteric lesions, differentials listed above. Often, CT and US findings are non-specific though sensitive in picking up abnormalities, subsequently further evaluation with MRI can be considered. In evaluation of atypical ureteric

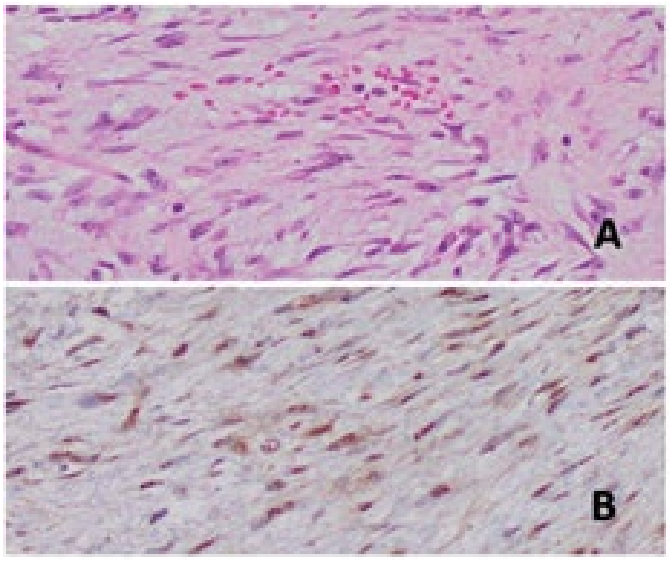

Figure 2. Histology of resected lesion at 100X magnification, H&E staining (A) and beta-catenin staining (B).

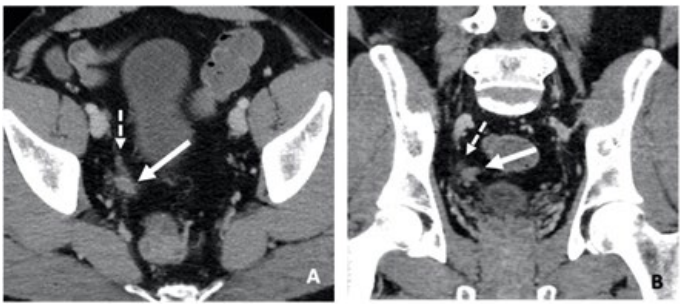

Figure 3. A & B: Axial and coronal images of a CT Enterography performed 18 months earlier showing the lesion (thick white arrow) as an ill-defined soft tissue density within the retroperitoneal fat adjacent and distinct to the right distal ureter (thin dashed arrow). This was distinct from the right seminal vesicle.

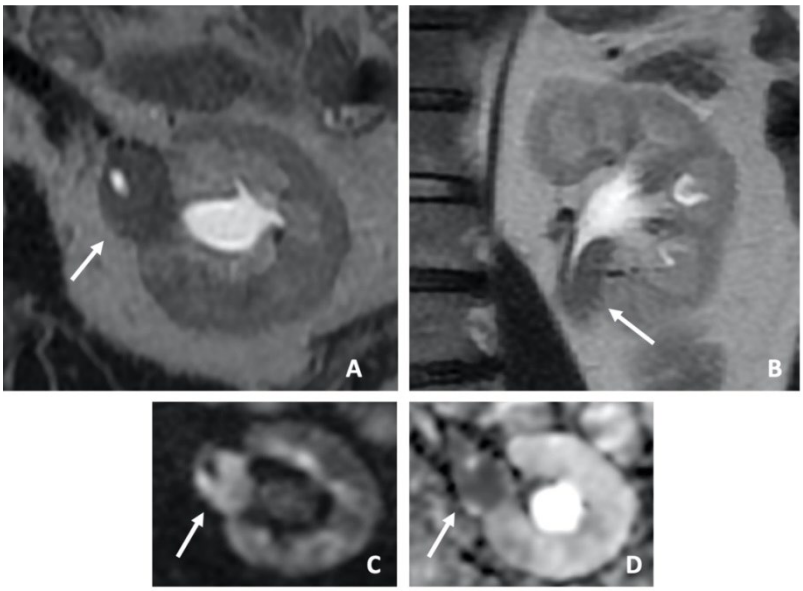

Figure 4. MRI images of a histologically proven left ureteric lymphoma. A & B: T2w Axial and Coronal MRI images showing a lesion (arrow) of the left pelvi-ureteric junction appearing mildly T2w hypointense, surrounding the left ureter. C & D: DWI and ADC images shows restricted diffusion of the ureteric lesion (arrow), suggestive of lymphoma.

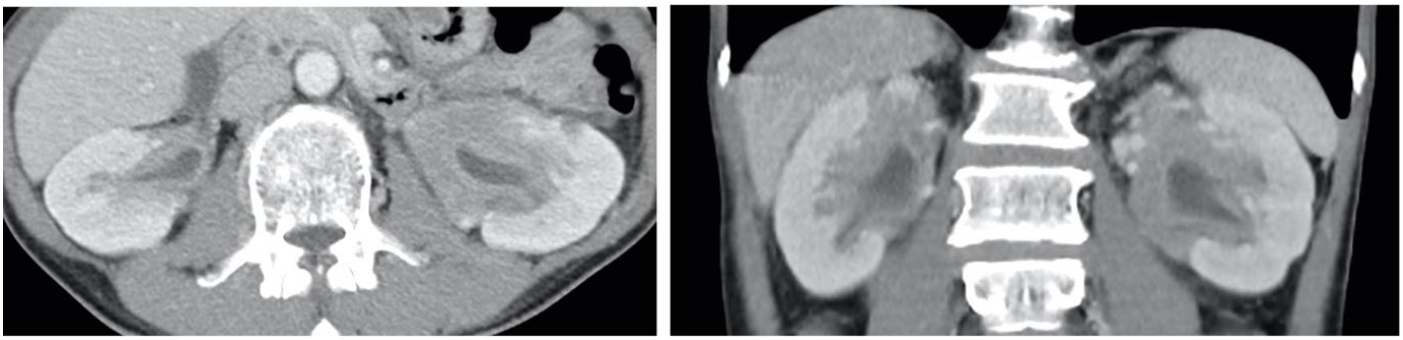

Figure 5. Contrast enhanced CT scans of a histologically proven IgG4-related disease of the collecting system. Axial and coronal images showing relatively symmetrical involvement of both kidneys with diffuse thickening of the pelvicalyceal systems with no significant hydronephrosis. Soft tissue thickening shows an infiltrative appearance into the adjacent renal parenchyma.

lesions, MRI offers superior soft tissue contrast and allows for delineation of margins for surgical planning to avoid critical adjacent structures.

With regards to treatment, multidisciplinary consensus should be obtained concerning feasibility for oncological resection. Surgical resection may be precluded due to involvement of vital structures. Consequently, consideration of radiation and systemic therapy can be explored. There have been reports using molecular targeted agents such as imatinib and interferon in the treatment of unresectable tumours [2,3,13]. Radiation therapy has shown promising efficacy and potential for adjuvant treatment to reduce recurrence rates [14].

Radiology plays an important role in the follow up of these tumours regardless of treatment strategies due to its propensity for recurrence, found to be as high as 77% [15,16]. There are currently no published guidelines on suggested follow up regimens, but these would most likely be based on consensus allowing for institutional practice, extent of the original tumour and the adopted treatment method.

Author Contributions

Case report concept and design

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Data acquisition

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Drafting of manuscript

Lim Shi Wei Desmond.

Critical revision of the manuscript

Lim Mei Chin.

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Conflicts of Interest

The authors have no conflicts of interest to declare.

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