CASE REPORT

Inflammatory myofibroblastic tumor of the ureter in a paediatric patient

Zehra Kazmi*[1], Fatima Ambreen[2], Ummiya Tahir[3], S. Zafar Zaidi[1]

[1] *Paediatric Urology, The Indus Hospital, Karachi, Pakistan.*
[2] *Paediatric Oncology, The Indus Hospital, Karachi, Pakistan.*
[3] *Department of Histopathology, The Indus Hospital, Karachi, Pakistan.*

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare soft-tissue neoplasm which has been described in a variety of locations. In the urogenital system, it occurs mainly in the bladder and the kidney. IMT arising from the ureter is exceedingly rare. We report an exceptional case of IMT arising from the ureteric submucosa in a three-year-old female child. The patient presented with pan-hematuria and no other symptoms. Urinalysis revealed numerous red blood cells, culture was negative for infection. A CT Urogram showed the lesion was involving most of the ureter. A left radical nephroureterectomy was performed, and she has remained well to date. Ureteral IMT is extremely rare and often asymptomatic, which can result in a delayed diagnosis. Radical excision is the best therapeutic approach with excellent survival outcomes.

Key Words: Inflammatory myofibroblastic tumor, Ureteric tumor, Spindle cell neoplasm, Paediatric urology

1. INTRODUCTION

Inflammatory myofibroblastic tumor (IMT)is also known as an inflammatory pseudotumor.[1] It is an extremely rare benign lesion with certain controversial clinical and imaging characteristics, which make it seem like a malignant neoplasm.[2]

It may arise from any primary site including the brain, lung, genitourinary tract, abdominal cavity, mesentery or the bladder. The lung is the commonest reported site.[3,4] This is such a rare tumor, that no clear incidence rate exists in the literature. The symptoms are often nonspecific, which often make it difficult to diagnose.[5]

2. CASE PRESENTATION

This is the case of a three-year-old female child born full term via normal vaginal delivery, with an age appropriate developmental history. She presented to us in the Paediatric Urology clinic with the complain of gross, painless hematuria for one week. The bloody urine came with clots and on and off mild left flank pain. She had no other lower urinary tract or constitutional symptoms.

General physical, abdominal and genital examinations were unremarkable, with a normal haematologic and biochemistry profile. An ultrasound showed moderate left sided hydroureteronephrosis. This was followed by a CT Urogram showing gross left hydronephrosis with a massive distortion of the left ureter in all its entirety, sparing only distal 1-2 cm, Figure

*Correspondence: Zehra Kazmi; Email: zehra-kazmi@hotmail.com; Address: The Indus Hospital, Plot C-76, Sector 31/5, Opposite Darussalam Society, Korangi Crossing, Karachi, Pakistan.

1. Some pelvic lymphadenopathy was also noted, while the right kidney and ureter were normal. The coronal and sagittal sections reveal an extensive infiltrative mass involving lower 2/3rd of left ureter with complete obstruction.

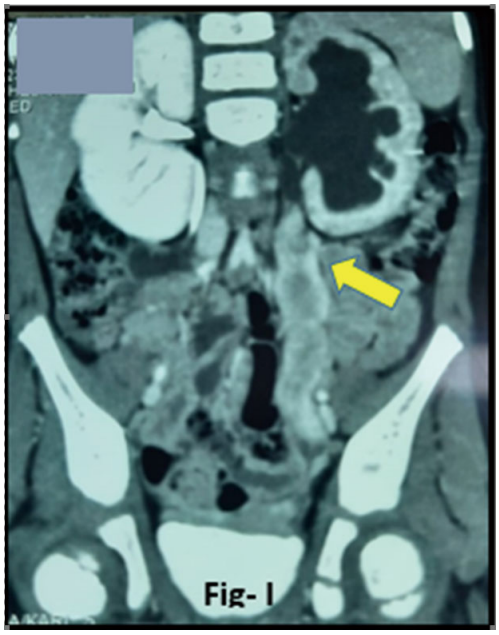

Figure 1. Computed Tomography Urogram (coronal & sagittal views) demonstrating a normal right kidney gross hydroureteronephrosis on the left side

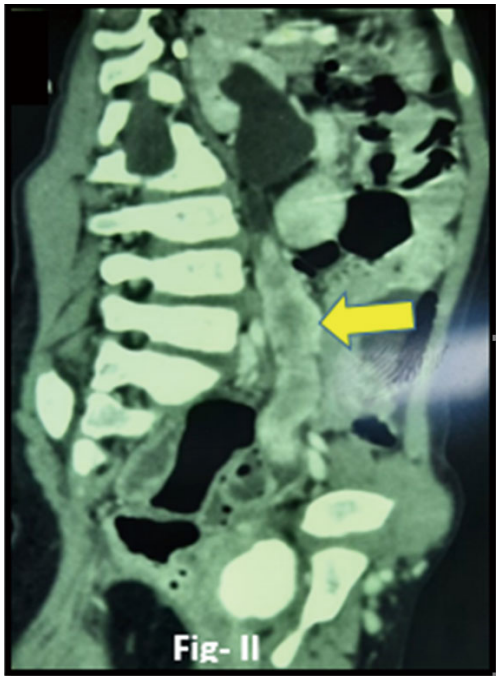

Figure 2. There is an extensively infiltrating mass lesion involving the lower two-thirds of the left ureter resulting in complete obstruction and no contrast uptake.

A DMSA scan was done to evaluate the functional status of the left renal unit. It revealed less than 10% function on the affected side. This case was discussed in our Paediatric Tumor Board meeting where a unanimous decision was made to proceed with surgical intervention. We performed a left radical nephroureterectomy via a left flank incision, and the gross specimen is shown in Figure 3.

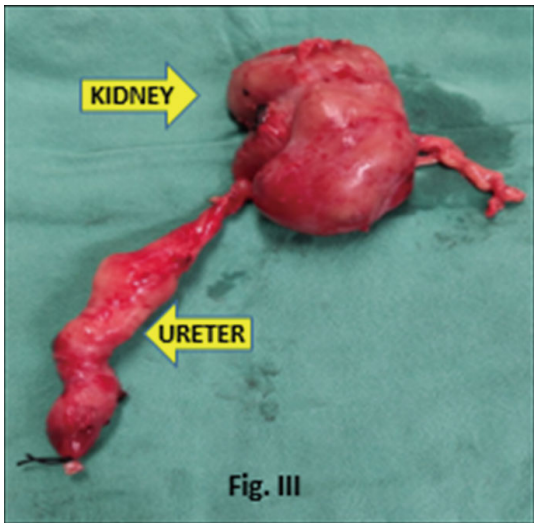

Figure 3. An image of the gross specimen, showing left kidney and a grossly dilated and distorted left ureter

There were no post-operative complications and she was discharged uneventfully on day 2. Histo-pathology revealed negative left ureteric margins, and a final diagnosis of benign Inflammatory Myofibroblastic Tumor. The histo-pathologic details are shown in Figures 4-7.

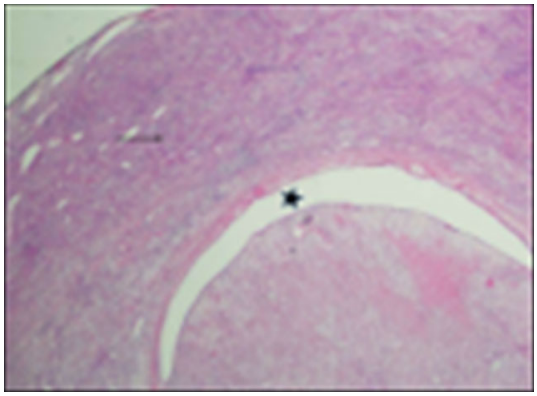

Figure 4. The arrow indicates tumor infiltration of the left ureteric wall, the asterisk indicates ureteric luminal compression

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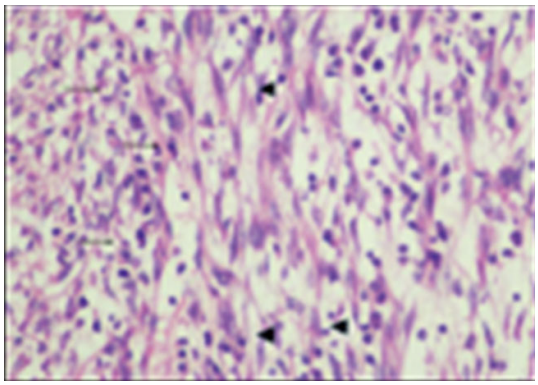

Figure 5. Spindle shaped cells (arrow heads)

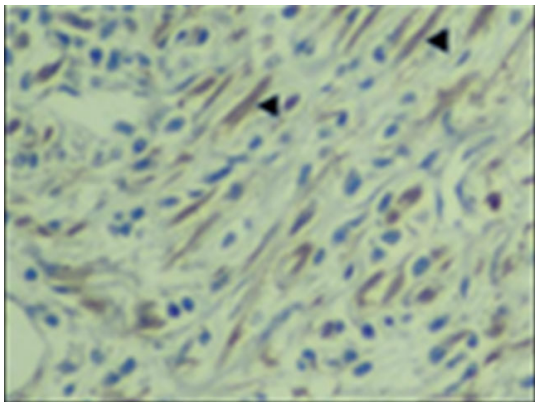

Figure 6. ALK protein

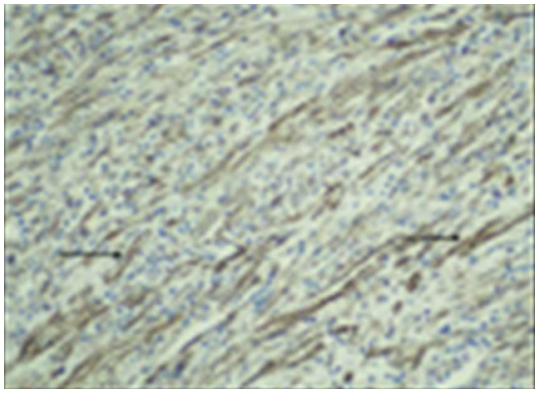

Figure 7. ASMA positive tumor cells

She has been well on subsequent follow up visits, and thus far no recurrence has been detected. Sparse data is available, and there is currently no uniform regime for follow up visits in these cases. We intend to keep her on a six monthly follow up with CT Urogram for two years, followed by an annual ultrasound.

3. Discussion

Inflammatory myofibroblastic tumors are histologically composed of myofibroblasts and fibroblasts with a dense inflammatory infiltrates of plasma cells, lymphocytes, and eosinophils,[6] as was also seen in our case.

Recent research has shown that this tumor is classified as a neoplasm with a high potential forlocal aggressiveness, recurrence, metastasis, and malignant transformation.[7] The common methods for diagnosis of IMT are ultrasound, abdominal computed tomography (CT) or magnetic resonance imaging.[8,9] The primary and curative treatment for IMT is complete surgical excision.[10]

Literature review revealed that this ureteric IMT has been identified in all age groups, across the world. We extensively reviewed the cases, and compared patient's treatment course with the most recently reported age matched cases. It was interesting to note, that most of the reported IMTs have occurred in male children,[11] whereas our patient is a three-year-old female which is quite a unique presentation.

Approximately 90% of the reported cases in literature performed a cysto-ureteroscopy with or without ureteric wall biopsy, to confirm the diagnosis.[12] The CT Urogram of our patient as shown in Fig. 1 and 2 revealed considerably thinned out cortex on the left side. Based on this finding we proceeded with a DMSA radionuclide scan, which confirmed the suspicion that this was indeed a poorly functioning unit. Hence the decision to perform an open left radical nephroureterectomy was made, and this management strategy is also supported by the available literature. Currently, there are no guidelines suggesting which approach is superior to the other.[13]

Certain reports highlight the importance of stringent long-term follow-up to detect local recurrence, which is quite common especially for lesions involving the lung.[14] After a detailed discussion in our multi-disciplinary tumor board meeting, it was decided that we will initially keep the patient on a close 3 monthly follow up with an ultrasound of the kidneys & urinary bladder for 2 years and then six monthly for three years. As indicated in the literature, this is the period when most IMTs are expected to recur, albeit in a new location.[15] After a period of two years, she will adhere to a 6 monthly follow up with only an ultrasound of the kidneys and bladder.

Children presenting with urinary tract IMTs fare well after radical surgical excision, and have an excellent prognosis. These patients need to remain on long-term follow up. We conclude that the commonest presentation of an inflammatory myofibroblastic tumor involving the kidney or the ureter is with hematuria, gross hydro-ureteronephrosis and a solid lesion involving the kidney or the ureter.

Conflicts of Interest Disclosure

There is no conflict of interest.

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