Inflammatory myofibroblastic tumour of the urinary bladder in a paediatric patient: a rare case

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Introduction

Inflammatory myofibroblastic tumor (IMT), also referred to as inflammatory pseudotumor or inflammatory fibrosarcoma, is an exceptionally rare condition which typically occurs in sites like the lung, orbit, and peritoneum, its occurrence retroperitoneally is infrequent. This tumor typically manifests in children and young adults, and its presentation and imaging characteristics can vary based on size and location, often resembling malignant neoplastic processes due to its aggressive nature [1]. This case report presents a unique occurrence of IMT in a 6-year-old female, emphasizing the importance of considering unusual diagnoses in paediatric patients with hematuria.

Case presentation

A 6-year-old female with no underlying medical illnesses was referred from her general practitioner for a 3 day history of gross haematuria associated with suprapubic pain and dysuria. This includes 2 days of initial drops of blood, followed by 1-day-history of passing blood clots. The child has no history of trauma or infectious illness. Family history and past medical history are unremarkable.

Physical examination shows the patient appears generally alert, with fair hydration. Vital signs such as blood pressure and heart rate are within normal range, and the patient is afebrile. On abdominal examination, the abdomen appears soft and non-tender. There is a vague mass palpable at the suprapubic region - it is non-mobile, non-tender, with a smooth surface, ~2.0 x 2.5 cm in size (note non-palpable lower margin of mass). Upon genital examination, the urethra and vaginal openings appear normal.

Bladder mass was suspected, and she was referred to paediatric surgery.

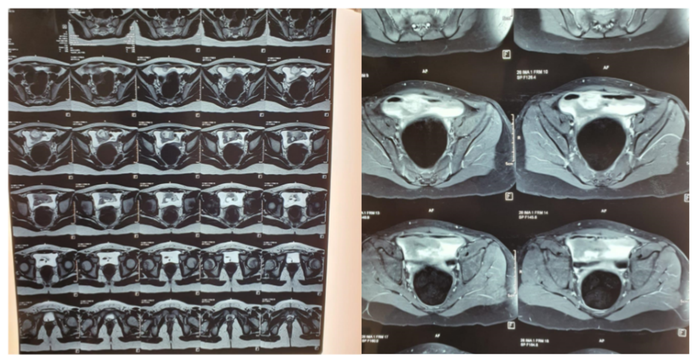
Investigations

Initial investigations done include a urine dipstick, showing gross hematuria. Full blood count, coagulation screen, and renal function tests are normal at admission. Upon a repeated set of bloods after the patient has passed large quantities of haematuria with blood clots, haemoglobin has dropped from 13.4 to 10.7 g/dl.

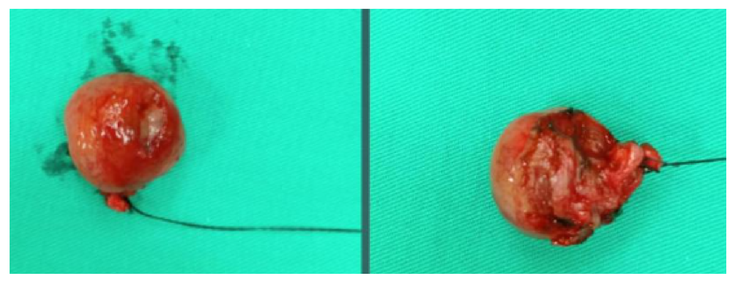
Initial management to stabilise the patient involved empirical treatment with IV fluids, NSAIDs, transfusion of fresh frozen plasma and packed cells, and urinary catheterisation for CBD. Further investigations were performed including an ultrasound of the bladder and MRI. Both reveal a fungating mass within the urinary bladder arising from the anterior abdominal wall, measuring 2.0 x 2.5 x 2.9 cm with adjacent blood clots. No other suspicious nodes or lesions were found.

Management

Patient was prepared for surgical management. Cystoscopy in theatre showed suboptimal visualisation due to numerous blood clots present within the bladder. Surgery was proceeded with lower midline incision and vesicostomy. Findings included a fungating bladder tumour arising from the dome of the bladder 2.5 x 2.0 cm in size, with a smooth surface and wide base, not eroding to the outer serosa layer. The specimen was sent for histopathology. Microscopy revealed a polypoidal tumour with proliferation of atypical spindle cells with mixed inflammatory cells, areas of focal mucosal ulceration, and resection margin which was clear from the tumour. Immunohistochemistry showed atypical spindle cells positive to anaplastic lymphoma tyrosine kinase (ALK-1) and vimentin - confirming inflammatory myofibroblastic tumour of the urinary bladder



Figures 1 and 2: Axial scans of the urinary bladder on T1 MRI, showing the presence of a bladder mass



Figures 3 and 4: Tumour excised from the urinary bladder, sized 2.0 x 2.5 x 2.9 c

Discussion

I. Haematuria in children:

Painless haematuria in children is related to a wide spectrum of conditions mostly unrelated to malignancy. Differentials include trauma, congenital anomalies, infectious diseases and inflammatory processes, vigorous exercise, malignant tumours, metabolic disorders, systemic and autoimmune disorders, bleeding disorders, and vascular disorders.

Gross haematuria, specifically, is commonly caused by urethrorrhagia (15%), urinary tract infection (14%), trauma (14%), congenital urinary tract anomalies (13%), and stones (6%) [2].

II. Inflammatory myofibroblastic tumours (IMT):

IMT is a very rare tumour with an incidence of less than 1 in 1 million people. Most commonly found in the lungs, it may also be found in the uterus, stomach, liver, intestine, larynx, or bladder. It originates from myofibroblasts and histiocytes. IMT is usually benign but can be locally invasive, and is hence known as 'the great mimicker' for its malignancy-mimicking-behaviour [3]. IMTs are usually aggressive and highly cellular, similar to malignant tumours. Recurrence is often local, and it has rarely been reported to metastasize to distant sites.

Bladder IMT is more common in young adults than children. It is important to consider rhabdomyosarcoma as a differential due to similar presentation. Bladder IMT can present with gross haematuria, anaemia, dysuria, frequency, and bladder outflow obstruction symptoms. Other symptoms that may be present depending on the size and location of tumour include fever, night sweats, weight loss, lethargy, pain.

Diagnosis involves imaging modalities, preferably MRI (as it is able to distinguish between bladder wall muscle layers) and biopsy (GOLD standard) showing lymphocytic proliferation, positive ALK-1, and spindle myoepithelial cell proliferation. It is important to have a high index of suspicion for such tumours, as they can mimic malignant tumours or infections.

Treatment modalities include surgical excision, non-steroidal cyclooxygenase 2 (COX2) inhibitors, with or without chemotherapy. Complete surgical excision of the tumour is required to prevent recurrence. In several cases, it may be difficult to preserve urinary function depending on the location of the tumour, especially if there is extensive bladder neck or ureter involvement. Supplemental NSAID use can shrink the tumour prior to excision. Monitoring involves follow-ups with the use of ultrasound or MRI to observe for recurrence. If recurrence is suspected, chemotherapy is indicated. Literature has supported the use of non-steroidals and chemotherapy in the treatment of IMT [4].

In terms of prognosis, the systematic review published in the Journal of Urology finds that urinary bladder IMT has a good prognosis post-surgical excision, despite ALK-1 status [5].

Conclusion

Inflammatory myofibroblastic tumour of the bladder is a rare benign tumour occurring mainly in young adults and children, presenting with gross haematuria and blood clots. In children presenting with such symptoms, clinicians must keep a high index of suspicion for bladder tumours. Whenever it is suspected and possible, complete surgical excision of the tumour is performed for successful outcome and the condition can be confirmed only by histopathological examination of the specimen. NSAIDs play a key role in treatment, and chemotherapy is recommended for especially aggressive or recurring tumours.

References

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