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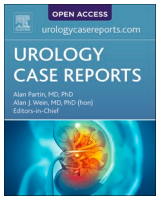

Pediatrics

Inflammatory myofibroblastic tumor of the bladder in an adolescent: Case report

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ABSTRACT

Inflammatory myofibroblastic tumors of the bladder (IMTB) are rare neoplasms that can occur in children. These tumors have uncertain malignant potential and can present similarly to bladder sarcomas. It is important to differentiate between IMTB and bladder sarcomas using a careful immunohistochemical approach. We report a case of IMTB in a 12-year-old girl who presented with presyncope and gross hematuria. IMTB was diagnosed through immunohistochemical analysis, and clinical improvement was observed after resection of the tumor.

1. Introduction

Inflammatory myofibroblastic tumors of the bladder (IMTB) are neoplasms of uncertain malignant potential composed of proliferations of myofibroblasts with inflammatory infiltrates. About 25% of IMTB cases have been found to occur in children and most commonly present with hematuria, dysuria, and abdominal pain.[1] Inflammatory myofibroblastic tumors of the bladder are exceedingly rare. We present a case of IMTB in an adolescent presenting with massive hematuria and review the literature.

2. Case presentation

A 12-year-old girl presented to the emergency room with episodes of presyncope and lightheadedness. The patient started her menstrual period 5 days prior to admission, but her mother mentioned that the patient was also experiencing episodes of gross hematuria. Physical examination revealed orthostatic hypotension. Laboratory evaluation revealed normocytic anemia with a hemoglobin of 8.2 g/dL. The patient was admitted for resuscitation and further workup.

Ultrasound revealed a heterogeneous space-occupying lesion measuring 5.7 × 4.5 × 3.9 cm (Fig. 1). The lesion was localized to the inferior posterior bladder.

CT Urogram did not reveal renal or ureteral pathology. The patient was taken to the operating room for cystoscopy with clot evacuation and tumor resection. At the time of cystoscopy, a bladder mass was identified arising from the right lateral wall that was actively bleeding (Fig. 2). Complete resection of the mass was performed, and the patient's symptoms resolved. Tissue specimens were analyzed by pathology after resection of the bladder tumor. Histology revealed a low grade, bland spindle cell tumor within a myxoid matrix and scattered inflammatory cells (Fig. 3a). On further immunohistochemical analysis, the tumor cells were positive for ALK-1 (Fig. 3b) and vimentin, with focal pan-cytokeratin positivity. The tumor cells were negative for s100, actin, desmin, calponin, myogenin, myo-D1, DOG-1, CD117, CD34, and CD31.

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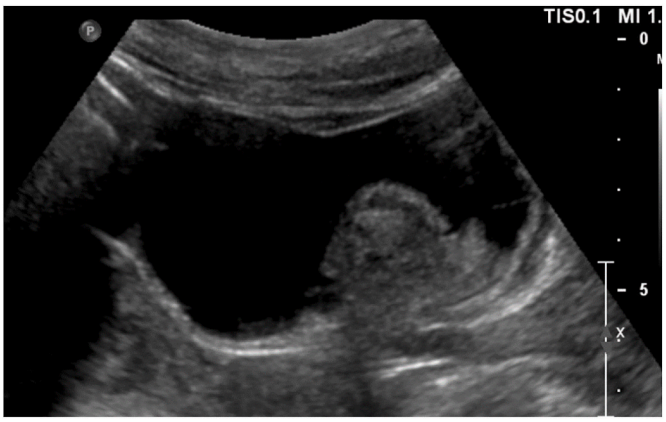

Fig. 1. Bladder tumor on ultrasound.

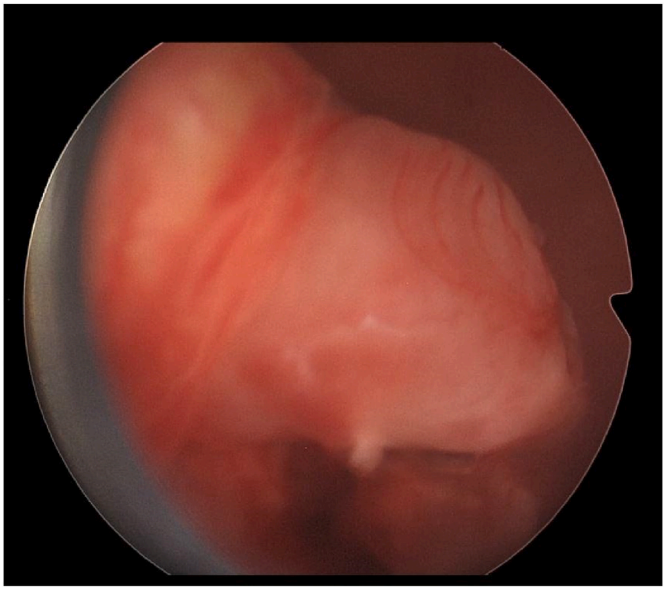

Fig. 2. Bladder tumor on cystoscopy.

3. Discussion

Inflammatory myofibroblastic tumors are not only found in the bladder but have been reported to occur in other organ systems such as the lungs and gastrointestinal tract.[1] The cause of IMTB is not fully understood, but it has been postulated that they can occur due to an abnormal immune response to an infection or injury. In some cases, genetic abnormalities have been reported in the development of these tumors. Trauma, vascular causes, and autoimmune disorders have also been linked to the pathogenesis of IMTB.[2]

Symptoms of IMTB in children include hematuria, dysuria, and abdominal pain. Our patient also complained of presyncope and lightheadedness, which in retrospect was likely due to anemia. It is extremely important to differentiate IMTB from other bladder sarcomas through careful histological and immunochemistry analysis. Aggressive therapy with radical cystectomy, radiation, or chemotherapy can be avoided with the appropriate diagnosis of IMTB.[1]

ALK-1 can serve as a useful marker in the diagnosis of IMTB and has been found to be expressed in 35–89% of cases of IMTB.[1,3] IMTB has a variably cellular spindle cell pattern on histology and also stains positive for vimentin, smooth muscle actin, desmin, calponin, and cytokeratin.[3] To exclude other mesenchymal tumors of the bladder, S100, CD34, CD117, myo-D1, and myogenin may be employed, as these stains are rarely positive in IMTB.[4] Malignant mesenchymal tumors can be excluded based on the aforementioned negative stains and the absence of significant pleomorphism, although malignant transformation following resection has rarely been reported.[5] Our patient's tumor, in comparison to the literature, consisted of a bland spindle cell neoplasm that stained positive for ALK-1, vimentin, and pan-cytokeratin. Her tumor was negative for desmin, calponin, and myogenin. Surgical resection of the tumor is typically the treatment of choice, and close follow-up is warranted as cases of inflammatory myofibroblastic tumors have been known to recur.[1]

4. Conclusion

Inflammatory myofibroblastic tumors are uncommon bladder neoplasms in children. They usually present with hematuria, dysuria and/or abdominal pain. Follow-up is recommended as these tumors can recur.

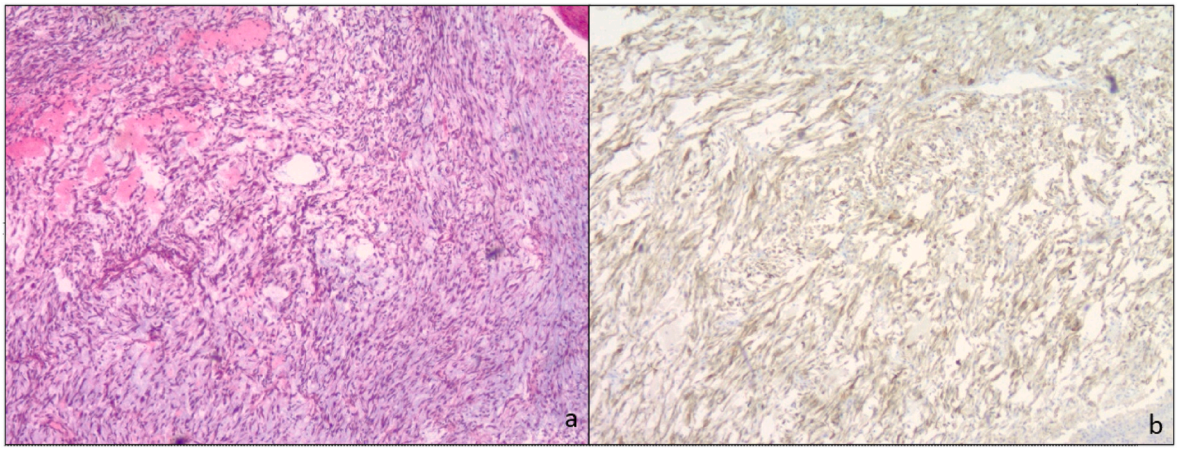

Fig. 3. a. Tumor with bland spindle cells in a myxoid stroma with mixed inflammatory cells b. Tumor cells are positive for ALK-1.

Consent

Yes.

Declaration of competing interest

None.

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