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Oncology

Prolapsing inflammatory myofibroblastic tumor of the urinary bladder: A case report and a review of literature

A. Higazy *, Mohamed Kandel

Urology Department, Ain Shams University, Cairo, 11361, Egypt

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) of the urinary bladder is a rare soft tissue benign tumor usually presents with hematuria, dysuria or obstructive urinary symptoms. No distant metastasis has been reported except in a single case. There is no clear consensus on the management plan yet. However, complete surgical resection is the mainstay management. We reported a case of a female patient presented with a painless prolapsing urethral mass that bleeds of touch with no signs of local invasion or distant metastasis, complete resection and pathological evaluation of the specimen revealed an IMT. The postoperative period was uneventful for 1 year.

Introduction

Inflammatory myofibroblastic tumor (IMT) of the urinary bladder is a rare genitourinary tumor, it is considered a soft tissue benign tumor with a poorly understood etiology. Here is no clear consensus till now regarding the proper management and follow up of such urinary bladder tumors. However, complete surgical resection through a transurethral approach is the mainstay practice in most of the report cases and in a few cases partial or radical cystectomy was performed.[1]

We report a case of (IMT) in a 40 years old female patient presented with a painless prolapsing mass through the urethra that was completely resected and was shown to be an IMT. It is the first case of a prolapsing tumor to be reported.

Case presentation

A 40 years old female patient presented to our clinic with a protruding soft tissue lesion through the urethra causing difficult micturition, dyspareunia. On examination, the protruding painless mass about 4 cm that bleed on touch, freely mobile not attached to the external urethral meatus as shown in (Fig. 1), preoperative ultrasonography and contrast CT were done showing a soft tissue lesion in the bladder protruding through the urethra with no back-pressure changes or any signs of extravesical involvement. Diagnostic cystoscopy was done showing a mass on the later wall of the bladder encroaching the left ureteric orifice and prolapse through the urethra, transurethral complete resection of the lesion was done and left ureteric stenting was done as showing in (Fig. 2). The postoperative period went uneventful and pathological assessment of the specimen showed myofibroblastic proliferation with spindle cell lesion and prominent myxoid and vascular stroma with no signs of malignancy. Immune histochemistry staining was positive with anaplastic lymphoma kinase (Fig. 3). The patient was followed up with cystoscopy after 3 months and sonography every 3 months for 1 year and no signs of recurrence were noted.

Discussion

IMT of the urinary bladder is a rare soft tissue benign tumor of unknown etiology characterized by spindle cell proliferation giving an appearance like sarcomas, they have been also known as pseudosarcomatous tumors, atypical fibromyxoid tumors, plasma cell granuloma or spindle cell tumor.[1]

Although it is histologically benign, it may carry a risk of sarcomatous change especially pulmonary IMT with metastatic potentials. Regarding urinary bladder IMT, only one case has been reported to metastasis to the colon following radical cystectomy. Local recurrence in the urinary bladder usually due to incomplete resection rather than a true recurrence.[1]

IMT can affect any age group, but it is more common in children and young adults with slight female preponderance. It is postulated that IMT differs in behavior according to the age group. Harik et al. suggested that the pediatric IMT had a more aggressive course with greater recurrence

* Corresponding author.
E-mail addresses: Ahmedmaherhigazy@gmail.com, ahmedmaher@med.asu.edu.eg (A. Higazy), Kandil1981@gmail.com (M. Kandel).

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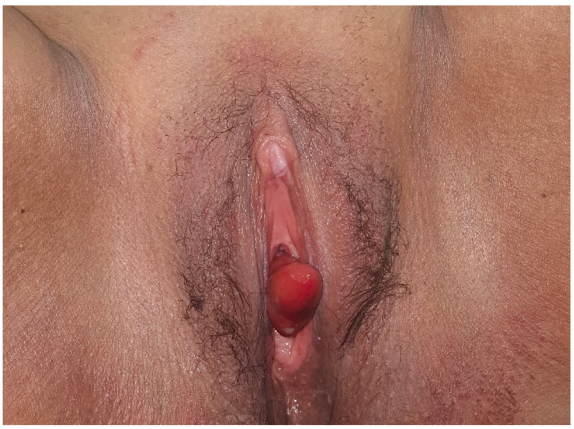

Fig. 1. (Prolapsing tumor).

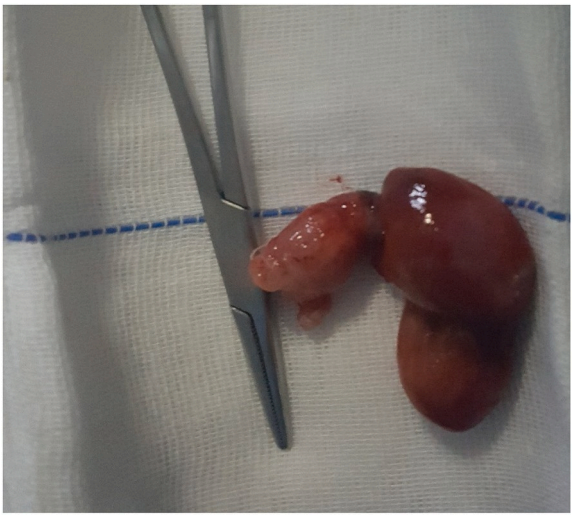

Fig. 2. (Resected tumor).

than the adult population.[2]

Genetic changes have been seen in IMT through rearrangements of the anaplastic lymphoma kinase (*ALK*) locus on chromosome 2p23, causing aberrant ALK expression especially in pediatrics and young adults while the expression of cytokeratin is comparatively more frequent in adult IMT.[3]

IMT usually presents with gross hematuria which may be absent if the lesion is confined to the submucosa. In our case, the presentation was a painless lump protruding from the urethra that bleeds on touch and causing dyspareunia and obstructive urinary symptoms.[2]

The commonest site of IMT in pediatrics is the dome of the bladder, compared to the lateral bladder wall in adults.[1] In our case, she was an adult female with a prolapsing pedunculated mass arising from the left lateral wall of the bladder.

Ultrasound evaluation and computed tomography (CT) scanning are the usual investigations to evaluate any bladder mass. However, transurethral resection and tissue diagnosis is mandatory to confirm the diagnosis.

Histological evaluation of IMT shows spindle myoepithelial cell proliferation and lymphocytic infiltrate. It can be confused with malignancy especially for rhabdomyosarcoma in pediatric patients and sarcomatoid carcinoma, leiomyosarcoma and malignant rhabdomyosarcoma in adult patients, immunohistochemical staining is positive for anaplastic lymphoma kinase (ALK) also Vimentin, smooth muscle actin and cytokeratin are expressed in IMT, while in rhabdomyosarcomas, Myogenin is a potent marker for rhabdomyosarcoma, and only 20% is positive for ALK. Thus, making ALK an important diagnostic marker for IMT in the absence of Myogenin. Leiomyosarcomas and sarcomatoid carcinomas do not express ALK especially with the presence of necrosis at the interface between the tumor and detrusor muscle should raise the concern of sarcoma.[4]

Most of the studies have agreed that the treatment of choice for IMT in both adult and pediatric patients is the complete transurethral resection of the lesion, local recurrence has been reported in adults following incomplete surgical resection otherwise, no proven recurrence or malignant episodes have been reported following surgical treatment.[1]

Complete surgical resection is important to avoid local recurrence, for a nonresectable IMT, pharmacotherapy has also been reported. COX2 and VEGF expression have been detected in IMT and are thought to be therapeutic targets. Pretreatment with non-steroidal anti-inflammatory (COX-2 inhibitors) to shrink the tumor size prior to resection has also been described in both adults and pediatric patients,[1] also An ALK inhibitor (Crizotinib) has also been used in a couple of cases in the treatment of an aggressive form of non-genitourinary IMT with a promising outcome.[5]

Post resection follow up is recommended by regular imaging and cystoscopy for local recurrence, although no specific protocol is agreed for the follow-up. Our institute adopted a 3 month follow up cystoscopy combined with ultrasonography every 3 months for up to 1 year.

Conclusion

IMT of the urinary bladder is a rare soft tissue benign tumor, usually presents with hematuria, dysuria or obstructive urinary symptoms. Histological evaluation and immunohistochemistry with the characteristic marker ALK are mandatory for accurate diagnosis and to differentiate it from more aggressive malignant tumors like rhabdomyosarcoma.

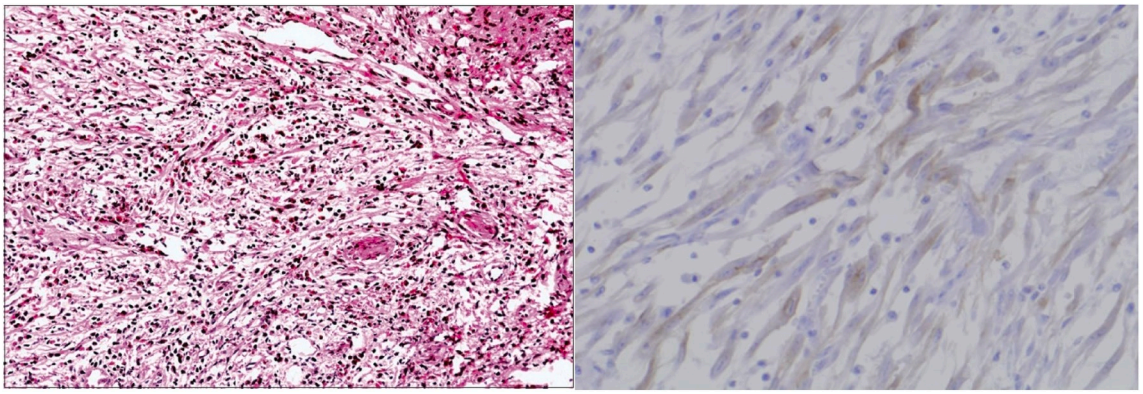

Fig. 3. (H&E staining on the right showing spindle myoepithelial cell proliferation and lymphocytic infiltrate - ALK positive staining of spindle cells on the left).

Transurethral complete resection is the preferred way of management of IMT. We recommend early follow up cystoscopy and radiological follow up for 1 year for detection of local recurrence.

Consent for publication

Written consent was obtained from the patient prior to surgery including approval for publication without breeching patient's confidentiality.

Funding

We received no fund to declare.

Declaration of competing interest

No competing interest to declare.

List of abbreviation

IMT	Inflammatory myofibroblastic tumor
ALK	anaplastic lymphoma kinase

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