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Inflammatory myofibroblastic tumor: an enigma

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Abstract

Background Inflammatory myofibroblastic tumor (IMT) of urinary bladder is a rare entity of genitourinary tract which has baffled urologists worldwide. Sign and symptoms are site specific. Usually diagnosed on the basis of immunohistochemistry findings.

Case presentation 35-year-old female patient presented with gross hematuria since 10 days. Ultrasound revealed a hyperechoic mass from right lateral wall of urinary bladder. Patient was managed with complete transurethral resection of bladder tumor. Histopathology and immunohistochemistry was diagnostic for inflammatory myofibroblastic tumor. 2 months post-surgery patient developed recurrence, following which she was managed with partial cystectomy.

Conclusion Inflammatory myofibroblastic tumor diagnosis is very challenging because of rarity of the tumor with non-specific presentation, and confirmation is done only by immunohistochemistry. Patients are usually managed with complete resection of tumor with regular follow-up.

Keywords Inflammatory myofibroblastic tumor, IMT, Urinary bladder, Partial cystectomy, Case report

1 Background

Inflammatory myofibroblastic tumor (IMT) is one of the rare soft tissue tumors which is composed of inflammatory infiltrate and spindle-shaped myofibroblasts. IMT has been puzzling the medical world since its inception when Brunn described two cases of myoma of lung in 1939, and many different terminologies have been assigned to it including "inflammatory pseudotumor", "pseudosarcomatous myofibroblastic proliferation", "inflammatory myofibrohistiocytic proliferation", "atypical fibromyxoid tumor", "plasma cell granuloma". Because of low risk of distant metastasis, World Health Organization (WHO) has grouped IMTs under the tumors of intermediate biological potential.

Lung is the most common organ involved and in the genitourinary tract, bladder is the most common site. The incidence of IMT is reported to be 6 out of 2050 bladder tumors [1]. Although etiology is unclear, but association with bladder instrumentation, inguinal hernia mesh repair, or urachal cysts, urinary tract infection (UTI), diabetes mellitus (NIDDM), steroid use, and immune disorders have been proposed [2]. Majority of the patients with IMT of bladder presents with gross haematuria, irritative voiding symptoms, and dysuria. Less reported symptoms include lower abdominal

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pain, clot retention, and loin pain. Very rarely, a large IMT can lead to bladder outlet obstruction and hydronephrosis. IMT expresses vimentin, smooth muscle actin and CK18 in up to 100% of cases [3]. In addition, it commonly displays muscle-specific actin, desmin, CAM5.2, and cytokeratin [3]. Approximately, 50–60% of IMTs exhibit expression of ALK-1, and correlated with ALK rearrangement in about 30–67% of these tumors on fluorescence in situ hybridization (FISH) testing. IMT does not express S-100 or MyoD1, which are characteristically found in rhabdomyosarcomas [3]. Histologically, IMT has less nuclear and cellular atypia and fewer mitotic figures when compared to these other malignant lesions [3].

IMTs usually have a benign morphological appearances, however, they can be aggressive, leading to metastatic and recurrent cases [4]. Complete resection is recommended because of risk of local recurrence [5]. Majority of the cases are managed with TURBT with regular follow-up to monitor for recurrence. Partial cystectomy has been employed for recurrent cases. Radical cystectomy has also been performed for muscle invasive IMTs. On the other hand, there have been some reports of tumors treated by steroids or anti-inflammatory drugs without radical treatment.

Follow-up with regular cystoscopies and radiological imaging in the form of regular computed tomography scan has been suggested for assessment of local tumor recurrence and distant metastases [4].

2 Case presentation

Our patient is a 35-year-old female, homemaker, non-smoker, non-alcoholic, known case of bronchial asthma, on treatment. She presented to us with chief complaints of gross haematuria with passage of amorphous clots since 10 days. Patient has history of lower segment cesarean section (LSCS) 3 years back. There was no history of dysuria, lower urinary tract symptoms (LUTS), abdominal pain, UTI, and clot retention.

On examination, patient was conscious oriented with general examination revealing pallor. Per abdominal examination revealed previous surgery scar, bladder not palpable. Rest of the systemic examination was normal.

Laboratory investigations showed Hb of 8.6 g/dl, total leucocyte count of 7700/mm^3 with platelet count of 1.07 lacs/mm^3. Serum creatinine was 0.7 mg/dl with normal coagulogram. Ultrasound of abdomen–pelvis revealed a 4 cm × 3.2 cm irregular hyperechoic mass arising from right lateral wall of urinary bladder with internal vascularity on color Doppler. There was no associated hydronephrosis (Fig. 1).

After getting anesthetic fitness, patient was subjected to transurethral resection of bladder tumor (TURBT) with complete resection of the bladder mass. Patient was discharged on post-operative day 2.

Histopathological examination (HPE) of TURBT specimen showed normal transitional epithelium and focal areas of surface ulceration and granulation tissue. The subepithelium showed proliferation

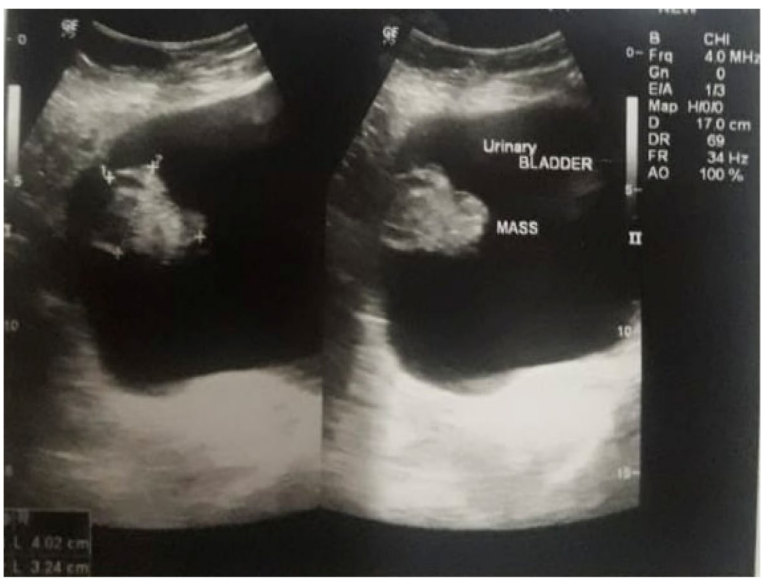

Fig. 1 Ultrasound image showing filled urinary bladder with hyperechoic mass lesion from right lateral wall

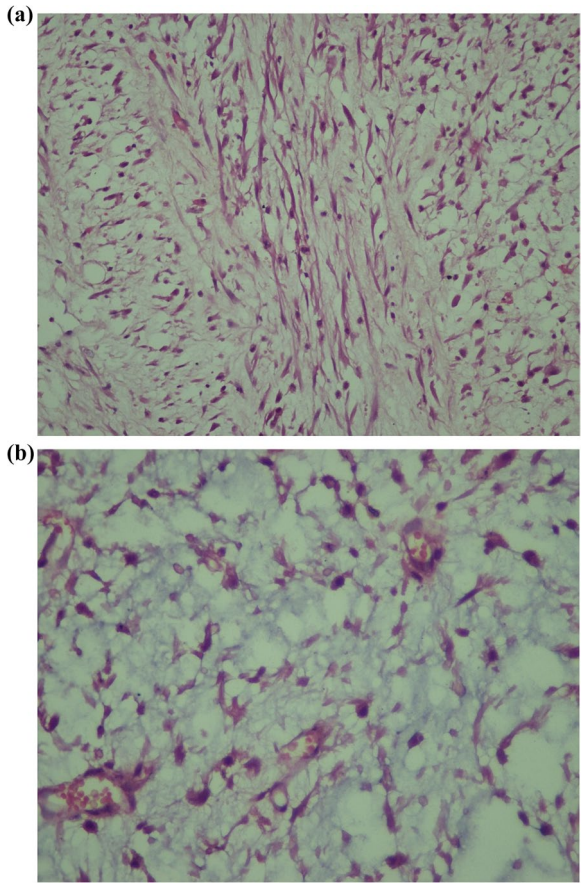

Fig. 2 **a** Spindle cells arranged in fascicles along with inflammatory cells on a myxoid background (H&E ×200). **b** Plump myofibroblastic cells along with plasma cells on a myxoid background (H&E ×400)

of spindle cells arranged in fascicles and haphazardly arranged at places. Individual cells showed mild pleomorphism having vesicular chromatin, prominent nucleoli with abundant eosinophilic cytoplasm. Occasional mitotic figures were noted. Background showed areas of myxoid changes and intermixed dense inflammatory infiltrate comprising of lymphocytes, plasma cells and few eosinophils. One tiny bit showed areas of necrosis (Fig. 2).

Immunohistochemistry (IHC) demonstrated ALK-1 immunoreactivity in majority of tumor (diffuse cytoplasmic pattern, 4+). Smooth muscle actin (SMA) and cytokeratin (CK) immunoreactivity was 3+. Ki-67 immunoreactivity was seen in 6–8% of tumor cells. Tumor cells were negative for CD-34 and S-100. On the basis of histopathology and IHC, diagnosis of IMT was made (Fig. 3).

Patient was advised for follow-up visit after 3 months. Subsequently, patient developed gross hematuria again approximately 2 months from surgery. Cystoscopy revealed a small 4 mm × 4 mm nodular growth at previous resection site. Patient was managed with partial cystectomy with adequate margins from previous resection site. Patient was discharged in stable condition with indwelling catheter. Per urethral catheter was removed on post-operative day 10.

HPE showed normal transitional lining epithelium and ulcerated mucosa with underlying submucosa showing foreign body giant cell reaction and inflammatory myofibroblastic areas infiltrating upto muscle. However, no muscle invasion seen. Resection margins were histologically unremarkable.

3 Conclusion

IMT of urinary bladder represent a diagnostic and therapeutic dilemma for urologists and pathologists across the globe. The rarity of this tumor with nonspecific clinical presentation and the radiological imaging studies makes it an enigmatic entity. IMTs are usually diagnosed on histopathological and IHC grounds. Although it has been considered as benign entity, recurrent and metastatic cases have been reported in the literature. Complete resection is usually considered as the procedure of choice with close follow-up of the patient.

This case report revives the medical fraternity about this rare entity and understanding the diagnostic and therapeutic challenges in such scenarios.

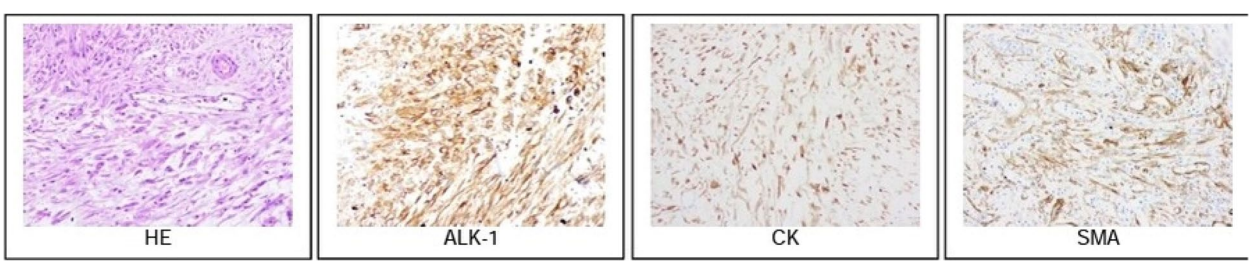

Fig. 3 IHC demonstrating ALK-1 immunoreactivity in majority of tumor (4+), along with SMA and CK immunoreactivity (3+)

Abbreviations

IMT	Inflammatory myofibroblastic tumor
UTI	Urinary tract infection
NIDDM	Non-insulin dependent diabetes mellitus
LUTS	Lower urinary tract symptoms
LSCS	Lower segment cesarean section
TURBT	Transurethral resection of bladder tumor
HPE	Histopathological examination
IHC	Immunohistochemistry
ALK	Anaplastic lymphoma kinase
SMA	Smooth muscle actin
CK	Cytokeratin
WHO	World Health Organization
MRI	Magnetic resonance imaging
ALCL	Anaplastic large cell lymphoma
FISH	Fluorescence in situ hybridization
TPM3-ALK	Tropomyosin3-ALK

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Author contributions
FMA—data acquisition and analysis. SAP—conceptualization of idea. MSW—design of the work. AHB—drafted the work and substantially revised it. ARK—data curation and methodology. ZZ—data acquisition and analysis. All the authors of this case report have read and approved the final version submitted.

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Declarations

Ethics approval and consent to participate
This work is approved by the institutional ethics committee of Sher-I-Kashmir Institute of Medical Sciences, with protocol number IEC/SKIMS/Protocol #EC36/2023.

Consent for publication
Written informed consent was taken from the patient for publishing of the data related to her.

Competing interests
The authors declare that they have no competing interests.

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