Inflammatory myofibroblastic tumour of the urinary bladder: a rare entity

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Abstract

Inflammatory myofibroblastic tumour (IMT) is a type of neoplasm composed of myofibroblast and fibroblastic spindle cells, with presence of inflammatory aggregates of plasma cells, lymphocytes, and eosinophils. However, IMTs rarely occur in the urinary bladder. We report a 20-year-old man who presented with haematuria for two days. Cystoscopy revealed a solitary tumour arising from the dome of the urinary bladder. The patient underwent trans urethral resection of bladder tumour (TURBT). The tissue histopathology examination (HPE) of the bladder tumour was suggestive of IMT of the urinary bladder. IMTs in the lower urogenital tract are special type of IMT. They are usually associated with surgical trauma and have been proposed to be an exuberant reparative reaction. Some IMTs are very aggressive, spreading locally, recurring, and requiring pharmacotherapy. A typical IMT can be locally aggressive and may require radical surgical resection (radical cystectomy) with close follow-up.

Introduction

Inflammatory myofibroblastic tumour (IMT) is a tumour of fibroblastic and myofibroblastic spindle cells associated with inflammatory infiltration of plasma cells, lymphocytes, and eosinophils.[1,2] Urinary bladder IMT is a rare entity.[1,2] It is important to differentiate this tumour from malignant spindle cell tumours.[1] The histopathological examination for IMT routinely shows proliferation of spindle-shaped cells with infiltration of plasma cells and lymphocytes. Immunohistochemical staining of the tumour will be positive for anaplastic lymphoma kinase (ALK), smooth muscle actin, and vascular endothelial growth factor (VEGF). If the immunohistochemical staining is positive, it is indicative of an IMT and it is suggested to use the inhibitors of ALK and VEGF as pharmacotherapy.[1]

The first case of IMT of urinary bladder was reported in 1980.[3] It was characterized by atypical spindle cell proliferation and inflammatory cell infiltrates primarily involving lymphocytes and plasma cells. Although it has been associated with trauma, surgery, and infection, the majority of IMT cases occur spontaneously. IMT is classified as intermediate (rarely metastasizing) tumour according to the WHO classification of soft tissue tumours.[1] Common involvement of IMT tumours are omentum, retroperitoneum, pelvis, and abdominal soft tissues in 73% of cases.[3] It is highly unusual to occur in the bladder.[2]

Case report

A 20-year-old man presented with gross haematuria with blood clots for two days. He had no abdominal pain or other bleeding tendencies. He had no history of taking any traditional medications, anticoagulant, or antiplatelet agents. He was a non-smoker. The patient had no past medical history. He denied any previous history of hospitalization. He also denied any urinary tract symptoms. Clinically the patient was pink with normal vital signs. No abdominal mass was palpable. Ultrasound abdomen done after haematuria resolved noted the presence of a urinary bladder mass (Figure 1). Computed Tomography (CT) scan staging revealed a 3.6 cm x 3.2 cm well-defined, macrolobulated lesion arising from the right anterolateral aspect of the urinary bladder with no evidence of metastasis (Figure 2). Cystoscopy demonstrated a solitary tumour arising from the dome of the bladder with query involvement of detrusor muscle (Figure 3). The patient underwent trans urethral resection of bladder (TURBT) on the same setting. The tissue histopathological examination (HPE) of the bladder tumour was suggestive IMT with no muscle involvement (Figure 4). Thus, he was planned for surveillance cystoscopy every 6 months for the first 3 years. No additional therapy was administered for this patient.

Discussion

IMTs range from benign to locally invasive and they have been proposed to be caused by chronic infection, an immune or autoimmune condition, trauma, surgery, or other malignancies. Commonly these tumours are found as isolated nodular lesions in the lungs, mesentery, retroperitoneum, or omentum.[5] They rarely occur in the genitourinary tract. IMTs in the lower urogenital tract

may be a special type of IMT associated with surgical trauma and have been proposed to be an exuberant reparative reaction.[6] We were unable to determine the likely cause of the IMT in our patient. Von Recklinghausen disease has also been reported to be associated with bladder IMT, but other typical characteristics of this disease such as orbital pseudotumour and thyroid lesions were absent in our patient.[2]

From the literature review, IMT is characterized histologically by an inflammatory infiltrate, and various microbes have been isolated from lesions (such as mycobacteria, corynebacteria, the Epstein-Barr virus, and human herpes virus). Infection has long been suspected to play an important role in IMT pathogenesis.[7] Although some IMTs are very aggressive, spreading locally, and recurring after successful excision, the IMT lesion in our patient

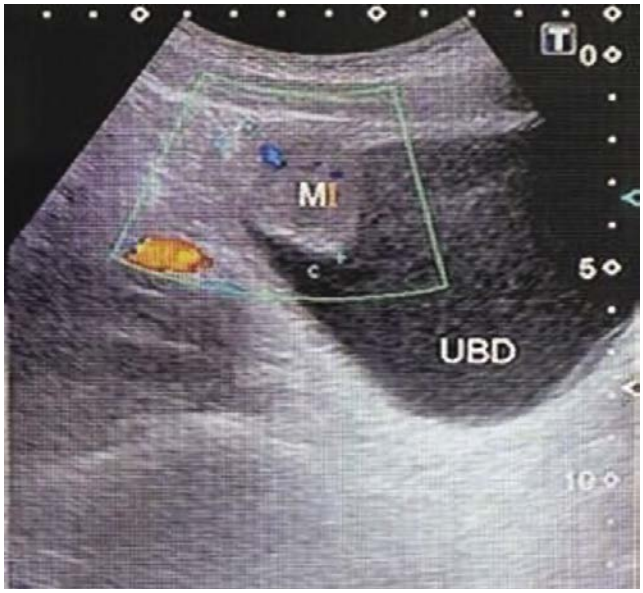

Figure 1. Ultrasound showed presence of urinary bladder mass

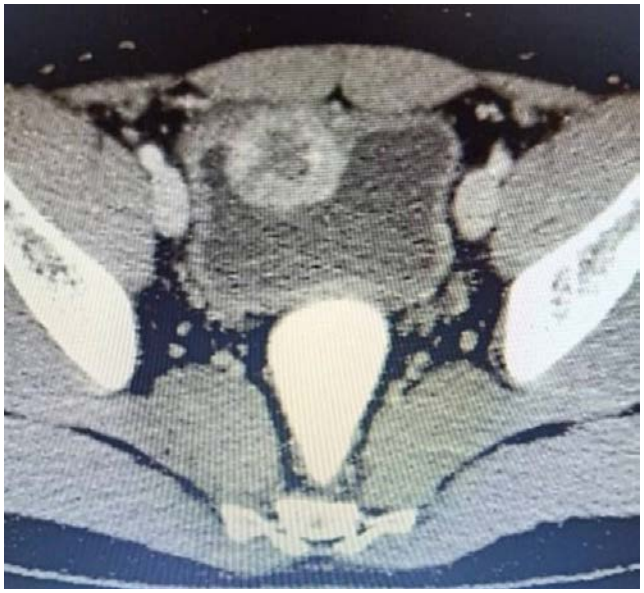

Figure 2. CT scan staging revealed a 3.6 cm x 3.2 cm well-defined, macrolobulated lesion arising from the right enterolateral aspect of the urinary bladder

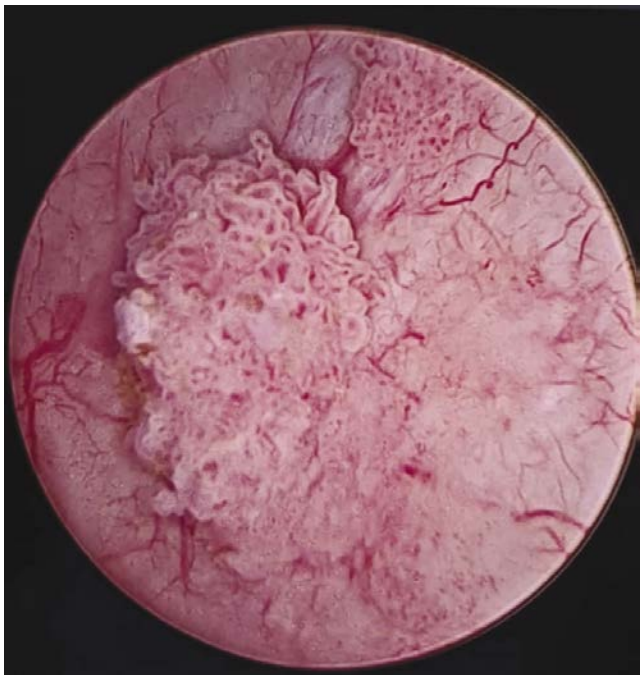

Figure 3. Cystoscopy showed solitary tumour arising from the dome of the urinary bladder

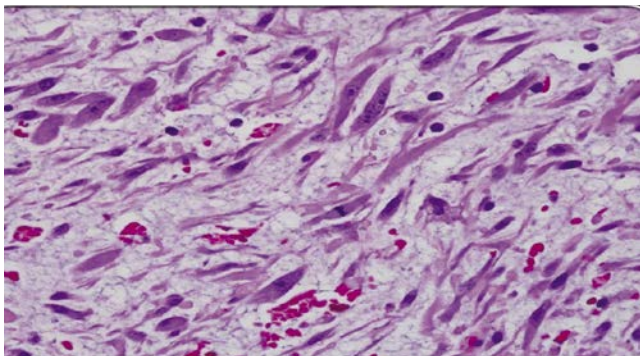

Figure 4. Microscopic features of markedly pleomorphism, plump spindly hyperchromatic to vesicular nuclei and coarse chromatin. Some with prominent macronucleoli. Elongated eosinophilic cytoplasm. Mitoses are present

appeared to be inflammatory rather than malignant. This is based on the tissue histopathological examination (HPE) of the bladder tumour which was suggestive IMT with no muscle involvement. The detrusor muscles of the bladder were preserved, which are usually involved when a bladder tumour is aggressively malignant like in sarcomatoid.[8] IMTs resemble malignant spindle cell tumours, such as sarcomatoid carcinoma, leiomyosarcoma, or rhabdomyosarcoma, making diagnosis difficult.[8]

Recent reports have indicated that ALK, which was originally identified as a protein overexpressed in anaplastic large-cell lymphomas, was overexpressed in a substantial proportion of IMTs of various anatomic location including urinary bladder.[9] A positive finding of ALK by immunohistochemistry in up to 87.5% of IMTs can be useful for the differentiation of IMTs from other spindle cell tumours in urinary bladder.[1] In this case, ALK immunohistochemistry was positive and useful for a definite final diagnosis.

Surgical resection is the initial therapy recommended for IMT of the bladder; complete surgical resection is important to avoid local recurrence.[9] In a systematic review undertaken in 2014 with total 120 patients, most patients underwent TURBT (60.8%), others had partial (29.2%) and radical cystectomy (9.2%). During follow up, 5 of the 73 patients who underwent previous TURBT experienced local recurrence.[6,10] Partial or radical cystectomy can ensure complete resection of the IMT, but in view of its benign disease course TURBT remained as an option for those patients who are reluctant to undergo a major surgery; the disease course of IMT should be explained thoroughly and the form of treatment should be highly individualized and tailored according case by case scenario. For locally aggressive or malignant IMTs, pharmacotherapy can be considered.[1] Cyclooxygenase-2 (COX-2) and VEGF expression have been detected in IMTs and may present therapeutic targets, thus enabling the use of COX-2 inhibitors such celecoxib.[1,11] An ALK inhibitor, crizotinib, has also been used in the treatment of IMTs.[1,11]

Crizotinib has been used successfully in the treatment of ALK-driven tumours in children, particularly IMT and anaplastic large cell lymphoma (ALCL). The clinical trials conducted by the United States Children Oncology Group (COG) demonstrated a complete response (CR) in 36% and partial response (PR) in 50% patients treated with crizotinib in ALK-positive IMT.[12,13]

Conclusion

In conclusion, IMT within the urinary bladder is a rare neoplasm of unknown malignant potential. A typical IMT of the urinary bladder can be locally aggressive and may require surgical resection (TURBT or radical cystectomy). Close follow-up with interval clinical and radiological monitoring for local recurrence and distant metastases is therefore warranted.

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