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Case Report

Inflammatory Myofibroblastic Tumor of the Bladder: Dramatic Presentation of an Unusual Tumor

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ABSTRACT

We report a case of an inflammatory myofibroblastic tumor of the bladder (IMT) in an adult male who presented with recurrent hematuria. He required partial cystectomy which revealed perivesical fat infiltration. In spite of this, the tumor was categorized as benign and the patient remained symptom- and tumor-free 18 months post-operatively.

Key Words: Myofibroblastic tumor, bladder, IMT

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INTRODUCTION

Inflammatory myofibroblastic tumor, also known as pseudosarcomatous fibromyxoid tumor, plasma cell granuloma, or inflammatory pseudotumour can affect the urinary bladder. Since first reported in 1980, about 100 cases of inflammatory myofibroblastic tumors of the bladder have been described[1]. We describe a case with unusual presentation and review the literature to provide a clear picture of a rare pathological entity.

CASE REPORT

A 41-year-old male patient presented with a 3 weeks history of severe frank painless hematuria, which did not respond to initial treatment with antibiotics. He is a fit non-smoker and has had no previous urological problems.

He was severely anemic with hemoglobin of 5.1 g/dl and normal renal function. He was catheterized for bladder washout and received a blood transfusion.

Urine culture showed no pyuria, bacterial culture was negative and urine cytology was inconclusive. An intravenous urogram revealed normal upper tracts. A CT scan of the abdomen showed a 2 x 1.8 cm solitary lesion located at the dome of the bladder. The rest of the abdominal organs appeared normal with no regional lymphadenopathy. Cystoscopy under general anesthesia showed active bleeding from a solid tumor at the bladder dome. The lesion did not have the gross appearance of transitional cell tumors and it was only possible to perform a limited resection due to poor visibility.

Intermittent hematuria continued and MRI of the pelvis showed a bladder tumor with extension into the adjacent peri-vesical fat (Fig 1,2). A further tumor

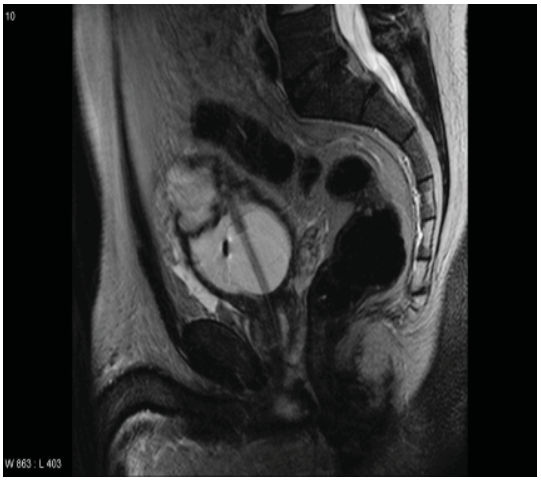

Fig. 1: Sagittal MRI scan demonstrating the presence of the myofibroblastic tumor on the dome of the bladder with extension into the perivesical fat (catheter balloon is also shown).

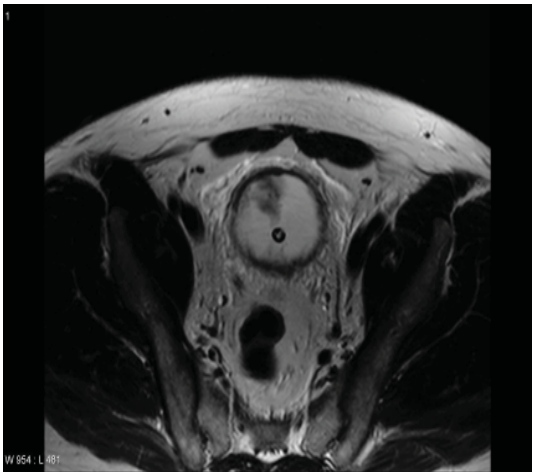

Fig. 2: Transverse MRI scan demonstrating the same tumor.

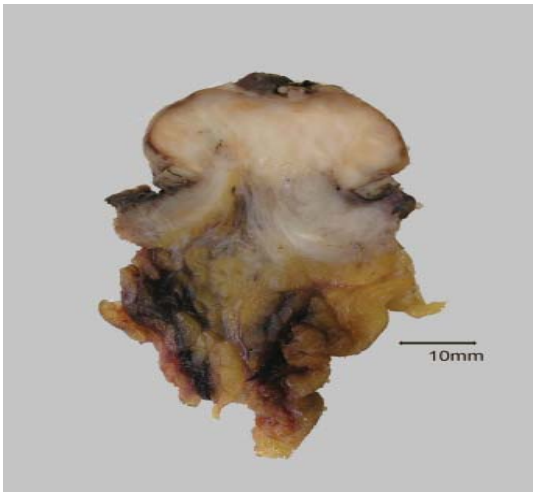

Fig. 3: Macroscopic appearance of the myofibroblastic tumor after partial cystectomy - note extension into the adjacent fat.

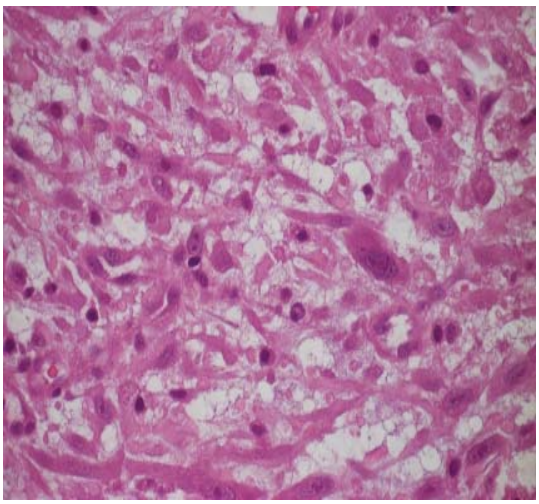

Fig. 4: Microscopic field showing spindle cell proliferation in addition to inflammatory cell infiltrate on a collagen background, typical of IMT.

resection was performed. The histology on both occasions showed a lesion with predominantly spindle cell morphology. Intermittent hematuria continued and it was decided to perform a partial cystectomy.

Abdominal exploration showed firm infiltration of the perivesical fat at the region of the tumor extending into the covering peritoneum. Part of the bladder dome and the covering perivesical fat was resected.

The procedure was assisted with flexible cystoscopy to mark the line of bladder excision and to take a healthy margin around the tumor.

Both the first biopsy and the subsequent partial cystectomy specimen exhibited the same histological features. Macroscopically there was an ulcerated polypoid mass projecting into the lumen of the bladder (Fig 3). The lesion appeared to be arising

from the lamina propria of the bladder wall and penetrated the muscularis propria to eventually blend into the perivesical fat, explaining the MRI scan appearance of possible extravesical fat involvement.

Histological examination showed a pleomorphic spindle cell tumor with a mixed inflammatory cell infiltrate (Fig 4). The tumor was focally positive for anaplastic lymphoma kinase (ALK-1) and the diagnosis of IMT was made.

DISCUSSION

Inflammatory myofibroblastic tumor (IMT) is defined in the 2002 WHO soft tissue classification as a "distinctive lesion composed of myofibroblastic cells accompanied by an inflammatory cell infiltrate of plasma cells lymphocytes and eosinophils"[2]. These were the findings in our case.

There is often expression of ALK-1 associated with genetic abnormalities in the tumor cells. In one series of bladder IMT, ALK-1 positivity was present in 10 of 14 cases[2] and the authors suggested that this supports the concept of IMT being a true neoplastic process rather than a reactive proliferation.

More than 100 cases have been reported in the literature since 1980[1]. One of the largest reported series of IMT involving the bladder included 62 cases[4]. The mean age was 47 years (range 7-77) with a male predominance (3.2:1).

The tumors are commonly associated with smoking and previous bladder instrumentation or surgery (trauma). The lesion can affect any part of the bladder wall in addition to other parts of the urinary system such as the ureters and prostate[5]. The presenting symptom is hematuria in most cases. In spite of its atypical histological features of muscle invasion the prognosis is usually good. Follow-up cystoscopy may be required as recurrence has been reported[4], although it is unusual following complete surgical resection[6].

In conclusion, IMT is an unusual benign tumor of the bladder. Its presentation can be drastic with recurrent severe hematuria. The standard treatment of this condition is not clearly defined. This tumor may grow deeply into the bladder wall as was the case in our patient and it may not be possible to perform complete endoscopic resection in all cases. Partial cystectomy is probably the treatment of choice in this context to achieve complete excision and reduce the risk of recurrence.

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