

Original Article

Inflammatory myofibroblastic tumor of the urinary bladder: report of a case and review of literature

Tao Wu^{1*}, Shulin Cheng^{1*}, Xi Duan², Bo Liao¹, Tielong Tang¹

¹Department of Urology, Affiliated Hospital of North Sichuan Medical College, Nanchong, China; ²Department of Dermatovenereology, Affiliated Hospital of North Sichuan Medical College, Nanchong, China. *Equal contributors.

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Abstract: We present a case of inflammatory myofibroblastic tumor of the urinary bladder. A 15-year-old boy with no previous history of malignancy presented to our hospital with hematuria, frequent micturition and urgent urination for two weeks; on contrast enhanced CT scan of the pelvis, we found a bladder tumor. Immunohistochemistry and fluorescent in situ hybridization confirmed the diagnosis. The patient underwent partial cystectomy. During the follow up, the patient was free of recurrence within 25 months. Inflammatory myofibroblastic of the urinary bladder may have a good prognosis after surgical resection.

Keywords: Inflammatory myofibroblastic tumor, urinary bladder, case report

Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal tumor. IMT was first recognized in the lung [1], and then was reported in many other sites including genitourinary tract [2]. IMT rarely occurs in the urinary bladder. It was Roth in 1980 [4] first described such IMT in the genitourinary tract. IMT of the urinary bladder is difficult to distinguish from other malignant spindle cell proliferation. We report one case of an inflammatory myofibroblastic tumor of urinary bladder. And we also performed a literature review of previous cases.

Case report

A 15-year-old boy with no previous history of malignancy presented to our hospital with hematuria, frequent micturition and urgent urination for two weeks. There was no history of preceding bladder instrumentation, urinary tract infection, or local trauma. There were no palpable lymph nodes. Contrast-enhanced computed tomography (CT) showed a 3*3 cm, solitary, heterogeneous density mass with no enlargement inguinal lymph nodes (**Figure 1**). Other radiologic and ultrasound examinations showed no obvious lymphadenopathy or distant metastases. The patient received an open

partial cystectomy. Histopathology of the operation specimens showed the tumor has a spindled myofibroblastic appearance with an inflammatory component (**Figure 2A**). Immunohistochemical study revealed diffuse staining with anaplastic lymphoma kinase (ALK) 1, smooth muscle actin (SMA), desmin, Ki67 (5-10%) (**Figure 2B**). Cells were negative for all other antibodies tested, including S-100, myogenin, EMA, CD117, myoglobin. Fluorescent in situ hybridization (FISH) showed ALK1 gene translocation on chromosome 2p23. All of the results confirmed the diagnosis of IMT. After treatment, the patient was free of recurrence within 25 months.

Discussion

There are many different names including Inflammatory myofibroblastic tumor, pseudosarcomatous myofibroblastic proliferation, inflammatory myofibrohistiocytic proliferation, inflammatory pseudosarcomatous fibromyxoid tumor and atypical fibromyxoid tumor have been described this neoplastic lesion [3, 4]. Regardless of many terminologies, the most often used name is IMT. It is an enigmatic, uncommon spindle cell lesion that can occur in the urinary bladder and multiple other sites. IMT of the urinary bladder was first reported in 1980

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Figure 1. Contrast-enhanced CT image: a solitary, heterogeneous enhanced density mass with no enlargement inguinal lymph nodes.

[5], which was a rare benign or potentially malignant condition of unknown etiology. IMT is often difficult to differentiate from malignant tumors including teratoma, liposarcoma and lymphoma prior to surgery, due to the nonspecific radiological features exhibited by the tumor. IMT occurs more often in the soft tissues and organs of children and young adults [6].

The most common clinical presentation of IMT is painless gross hematuria [7]. Other symptoms may include frequency of urination and dysuria. Clinical symptoms are not distinct from other bladder carcinoma. Contrast-enhanced CT and MRI usually demonstrate early peripheral enhancement. This finding can be explained by the histological arrangement of the lesion, with spindle cells, blood vessels and inflammatory cells at the peripheral area [8]. Delayed imaging often shows increasing enhancement due to the presence of fibrosis [9]. Distinctly to urothelial carcinoma, IMT occurs in a younger age group, usually appears as a single urinary bladder mass. Enhancement characteristics may help to distinguish these two entities as urothelial carcinoma enhances avidly and IMT has ring like enhancement with a hypo-enhancing central region [10].

However, histological confirmation is essential to distinguish IMT from other urinary bladder carcinoma. The positive finding of ALK by immunohistochemistry in up to 87.5% of the IMTs can be useful for the differentiation of IMTs from other malignant spindle cell lesions [11]. And, 30%-67% of IMTs were also noted to have ALK gene rearrangements on FISH testing [12]. The results of ALK expression in IMT have made

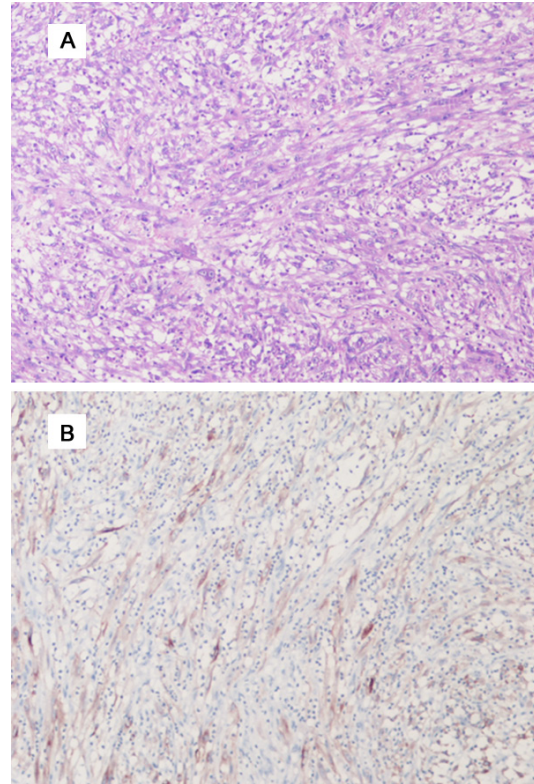


Figure 2. A: HE (hematoxylin-eosin) staining: spindled myofibroblastic appearance with an inflammatory component (Original magnification $\times 100$). B: Immunoperoxidase staining: ALK1 reactivity (ALK antibody, original magnification $\times 100$).

ALK a hallmark marker for diagnosing IMT [7]. Chun et al identified 41 studies containing 182 patients. Of the IMTs, 65% were ALK-positive. Regardless of its ALK status, IMT of the urinary bladder has a good prognosis after surgical resection [7]. Immunohistochemistry and FISH all confirmed ALK1 positive in our case.

According to many studies, IMT of the urinary bladder seems generally run a very benign disease course. The largest systematic review by now reported local IMT recurrence rate was 4% and no cases of distant metastases [7]. However, Wang reported a malignant IMT of 14 year old boy. The tumor had infiltrated the greater omentum, appendix, and the rectus muscle. The patient treated with radical resection and perioperative chemotherapy. He had no evidence of recurrence or metastasis 15 months after initial diagnosis [6]. In most reported cases of IMTs of the urinary bladder, surgical resections, including transurethral resection, partial and radical cystectomy were

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performed [13]. We performed partial cystectomy and the patient was free of recurrence within 25 months. However, long-term surveillance and implication of this lesion is unclear. We need more multi-center cases and long-term followed data to guide a better clinical decision about diagnosis and treatment for IMT of the urinary bladder.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Tielong Tang, Department of Urology, Affiliated Hospital of North Sichuan Medical College, Nanchong 637000, China. Tel: 86-8172262409; Fax: 86-8172262409; E-mail: scimail@126.com

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