Inflammatory myofibroblastic tumour of the bladder: Case report and review of the literature

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Abstract

Inflammatory myofibroblastic tumour (IMT) is a rare tumour with malignant potential, and has been described in many major organs. However, bladder location is very uncommon. We report the case of a 23-year-old woman who presented with painless gross hematuria for 2 weeks. Contrast-enhanced computed tomography revealed a bladder tumour. The patient underwent an open partial cystectomy and the final pathologic diagnosis was IMT of bladder. Typical IMTs can be locally aggressive, therefore close follow-up is necessary.

Introduction

Inflammatory myofibroblastic tumour (IMT) is a rare tumor with malignant potential; it is also known as inflammatory pseudotumour. IMT has been described in many major organs, including lungs,1 liver2 and skin.3 In the genitourinary system, IMT likely starts in the bladder, but the lesion has also been reported in the kidneys, prostate, ureter4 and epididymis.5

Roth reported the first case of IMT of the bladder in 1980.6 In the past 10 years, more than 20 cases of bladder of IMT have been documented. The largest clinicopathologic study was reported by American pathologists in 2006. This study included 42 cases of IMT of the bladder.7 We report a new case of inflammatory myofibroblastic tumour of the bladder, in which the patient presented with gross hematuria. We review the tumour’s clinical presentation, diagnosis and pathological features.

Case report

A previously healthy 23-year-old woman presented with painless gross hematuria in 2 weeks. She had no history of urinary tract infection, calculi, trauma and other urological abnormality. Laboratory studies were normal, except for severe microscopic hematuria. Initial abdominal ultrasound showed a mass on the bladder dome, but the results of intravenous pyelography were normal. Cytological analysis of urine did not show malignant cells. Contrast-enhanced computed tomography (CT) showed a round mass arising from the wall of the bladder dome, and with deep muscle invasion, the mass could be enhanced non-uniformly (Fig. 1). A suspicious pelvic lymph node was observed. Cystoscopy showed a broad-based tumour in the superior wall of the bladder. The tumour was 3 cm in diameter; it was bleeding and had necrosis on its surface. We decided to perform transurethral resection (TUR) for hemostasis and because we wanted to keep the tumour tissue for pathological examination. Unfortunately, the effect of hemostasis by TUR was not satisfied and severe hematuria induced hypovolemic shock. Open partial cystectomy was performed to remove the tumour in this emergency.

As this is a rare case, our initial diagnosis was carcinoma of urachus due to the tumour’s site, so we removed the partial urachus nearby the bladder simultaneously. Although abnormal lymph node was not detected during the operation, bilateral pelvic lymph tissue was biopsied considering the CT results. The final pathologic diagnosis was inflammatory myofibroblastic tumour of bladder (Fig. 2). The maximum diameter of the tumour was 3 cm and invasion of muscularis propria was observed (Fig. 3), however, serous membrane of bladder was intact. The necrosis and bleeding were seen in the tumour tissue. There were many ganglion-like cells, spindle cells and epithelioid cells in the lesion. Mitoses were only occasionally found (2-3/10 hpf) (Fig. 4). The results of immunohistochemistry (IHC) examination showed: CD117 (-); DOG-1(-); CD34(-); ALK(+); Galdesmon(-); SMA(+); about 25% the tumour cells were Ki-67 positive. There was no histologic evidence of extension to the lymph tissue and urachus.
IMT is a rare pathologic entity composed of myofibroblasts and an accompanying inflammatory infiltrates. IMT was first recognized in the lung. Although some cases of IMT are considered to represent an inflammatory response to infection, trauma or surgery, the etiological factors are not clear. We reviewed 17 cases of IMT of the bladder (Table 1); patients’ ages ranged from 3 to 72 years (mean 37) and males were represented slightly more than females (ratio 9:8). The most common symptom of IMT is hematuria. The data are consistent with Iczkowski and colleagues. They researched clinic data of IMT in 36 patients, whose chief complaint was hematuria (n = 27). The diagnosis of IMT may remain a dilemma for urologists, radiologists and pathologists. Because the IMT of the bladder has similar clinical features to uroepithelial cancer and it is sometimes aggressive on imaging, this lesion is often mistaken as a malignant process in the diagnostic procedure and during surgery. In our case, according to cystoscopic evaluation and CT results, invasive bladder cancer was established and a radical cystectomy was the next therapeutic option.

The diagnosis of IMT is identified by pathological examination. Immunohistochemical stain can help pathologists confirm the diagnosis. Histologically, the tumour cells were spindle to stellate in shape, widely separated or showed a compact fascicular pattern. They were often mixed inflammatory infiltrates and an irregular meshwork of small dilated vessel. Mitoses were typically scant. Montgomery and colleagues reviewed a series of IMTs arising in the ureter, bladder and prostate derived primarily from a large consultation practice. They found necrosis in 14 (30%) cases.

**Fig. 1.** Contrast-enhanced computed tomography showed a round mass arising from the wall of the bladder dome and with deep muscle invasion, the mass could be enhanced non-uniformly.

**Fig. 2.** Inflammatory myofibroblastic tumors cells were spindle to stellate in shape, widely separated or showed a compact fascicular pattern (hematoxylin and eosin stain, ×100).

**Fig. 3.** Image shows the invasion of muscularis propria (hematoxylin and eosin stain, ×100).

**Fig. 4.** Image shows typical mitotic figures (white arrow, hematoxylin and eosin stain, ×200).
Inflammation of the muscularis propria was found in 19 (41%). By IHC, lesions at least focally expressed anaplastic lymphoma kinase (ALK) (20/35, 57%), CAM5.2 (10/15, 67%), CK18 (6/6, 100%), actin (23/25, 92%), desmin (15/19, 79%), caldesmon (4/7, 57%, rare cells), p53 (10/13, 77%); most lacked S100 (0/14), CD34 (0/13) and CD117 (2/13, 15%). The histology and immunohistochemistry of our case were consistent with previous studies. However, there was not enough evidence to diagnose sarcoma, as in our case only 2-3/10 hpf typical mitotic figures were found even in most active mitotic area.

The therapy of IMT usually includes TUR, partial cystectomy and radiotherapy. Complete surgical resection is important to avoid local recurrence.16 Compared with TUR, partial cystectomy is used for most patients (Table 1). One patient had a radical cystectomy due to the uncertain pathogenesis of the inflammatory myofibroblastic tumour and the rarity of Von Recklinghausen’s disease. In our case, the patient finally underwent partial cystectomy. Considering the muscle invasive feature of IMT, we think the partial cystectomy may be more reliable to avoid tumour residue. If the tumour recurred after endoscopic resection, a partial cystectomy was suggested. In our review of the 17 cases, nearly half of the patients received regular follow-up for at least 6 months, without evidence of local recurrence (data not shown). Iczkowski and colleagues reported no recurrence of inflammatory pseudotumours after TUR or partial cystectomy in 36 patients.21 Additionally, Berger and colleagues reported the first case of a bladder inflammatory myofibroblastic tumour that responded to an anti-inflammatory regimen (prednisone and Cox-2 inhibitor) before surgical extirpation.22

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**Conclusion**

IMT is a rare neoplasm with unknown malignant potential. Typical IMTs can be locally aggressive, and may require radical surgical resection, therefore close follow-up is warranted.

**Competing interests:** None declared.

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**References**


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