Clinical Case Report

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Inflammatory myofibroblastic tumor of urinary bladder with severe hematuria

A Case report and literature review

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Abstract

Rationale: Inflammatory myofibroblastic tumor (IMT) is a rare intermediate soft tissue tumor. Rarely occurring in the urinary bladder, MTs is composed of myofibroblast differentiated spindle cells and accompanied by numerous inflammatory cells, plasma cells and/or Lymphocytes.

Patient concerns: A 28-year-old female, with history of 2 cesarean sections 8 years ago and 2 years ago, was admitted to the emergency department for a 7-day ongoing gross hematuria with clots.

Diagnoses: A cystoscopy showed a tumor near the bladder neck, computed tomography (CT) showed a mass and massive blood clots in the bladder. Finally, postoperative pathological examination confirmed a diagnosis of inflammatory myofibroblastic tumor of bladder.

Interventions: The patient was given rehydration and blood transfusion to improve the general condition. Then, emergency transurethral resection of the bladder tumor was performed.

Outcomes: No metastases to lymph nodes and other organs were found, and no tumor progression was revealed during 3-months of follow-up.

Lessons: IMTs rarely occur in the urinary bladder. Due to the lack of specificity in clinical symptoms, it's difficult to arrive at a definite diagnosis before operation. Therefore, usually, the final diagnosis depends on histomorphological features and the immunohistochemical profile. Further case studies are required to study the biological behavior of this condition.

Abbreviations: ALK = anaplastic lymphoma kinase, CK = cytokeratin, CT =computed tomography, IMT = inflammatory myofibroblastic tumor, RBC = red blood cell, SMA = smooth muscle actin, TURBT = transurethral resection of the bladder tumor.

Keywords: bladder, Inflammatory myofibroblastic tumor, severe hematuria

1. Introduction

Inflammatory myofibroblastic tumor (IMT), a rare intermediate soft tissue tumor.

Consequently, many different names have been assigned to this condition, including inflammatory pseudotumor, nodular fasciitis, and pseudomalignant spindle cell proliferation.[1,2] It arises from various organs, such as the lung, retroperitoneum, and pelvisand. Inflammatory myofibroblastic tumor of urinary bladder is extremely rare.[3] The first report of an Inflammatory myofibroblastic tumor of the urinary bladder was made in 1980.[4] Most patients are young, a mean age of 38.9 years and the incidence of female was higher (51.9%).[7] It manifests with hematuria, frequency–dysuria syndrome, and bladder outlet obstruction.[3] Surgical resection is the treatment of choice and the prognosis of IMTs of bladder is relatively good. The patient we reported was hospitalized for severe anemia caused by hematuria, who eventually recover smoothly after emergency surgery and other related treatments. We have further clarified the pathogenesis of IMT through this case report and related literature review. And we think we should not hesitate to patients with severe hematuria—timely surgery is extremely helpful to patients.

2. Case presentation

A 28-year-old female, with history of 2 cesarean sections 8 years ago and 2 years ago, was admitted into the emergency

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Daoqing Song and Wei jiao contribute equally to this paper.

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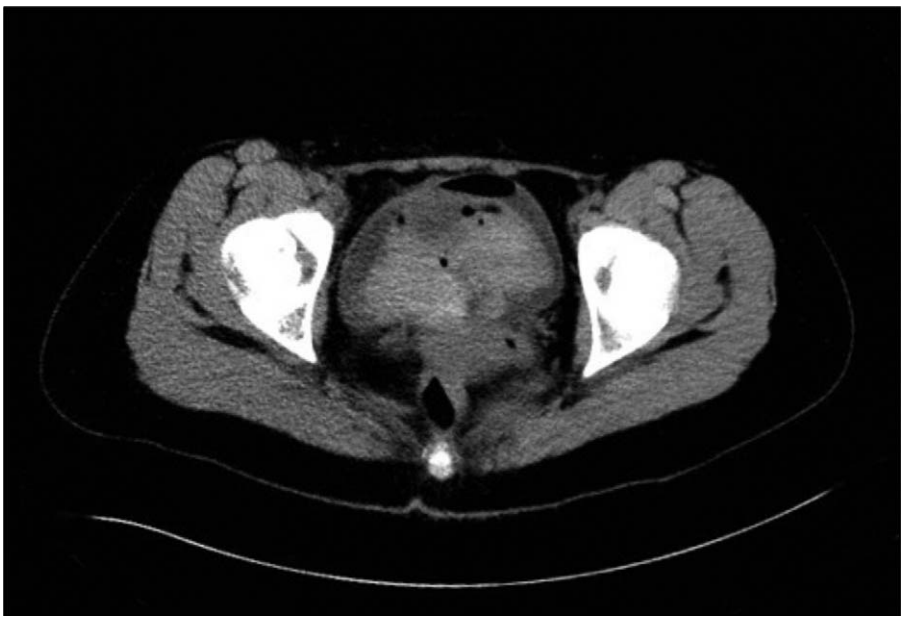

Figure 1. CT-scan showed a low-density mass near the bladder neck and a large number of blood clots in the bladder.

department for a 7-day ongoing gross hematuria with clots. Full blood count analysis on admission revealed a hemoglobin of 62.0 g/L. Then two units of red blood cells 200 ml plasma were infused to observe the condition continually while defining diagnoses and determining the treatment plan. A brief cystoscopy reported from the previous hospital showed a tumor approximately 2*3*4 cm in size near the bladder neck at 11 o'clock (lithotomy position). A computed tomography (CT) scan of the abdomen and pelvis showed a mass near the bladder neck and massive blood clots in the bladder (Fig. 1). Aiming to wait for hematuria to be controlled and then re-cystoscopy before we can perform the treatment, we consulted the interventional department for bladder tumor vascular embolization (Surgery related images shown in Fig. 2) in the hope of controlling bleeding. Nevertheless, her hemoglobin reduced to 39 g/L due to continuous hematuria prompting transfusion of 3 units of packed red blood cells and 400 ml plasma urgently the next morning. Consequently, the patient underwent an emergency transurethral resection of the bladder tumor (TURBT), demonstrating a lot of clots and a solid tumor of about 4 cm near the bladder neck. Once again, 2 units of red blood cells were injected and we gave other symptomatic supportive treatment after surgery to improve the general condition of the patient. To elaborate the change of hemoglobin in patient more intuitively, we specially created a line chart (Fig. 3). Finally, the patient's condition improved and she was discharged smoothly.

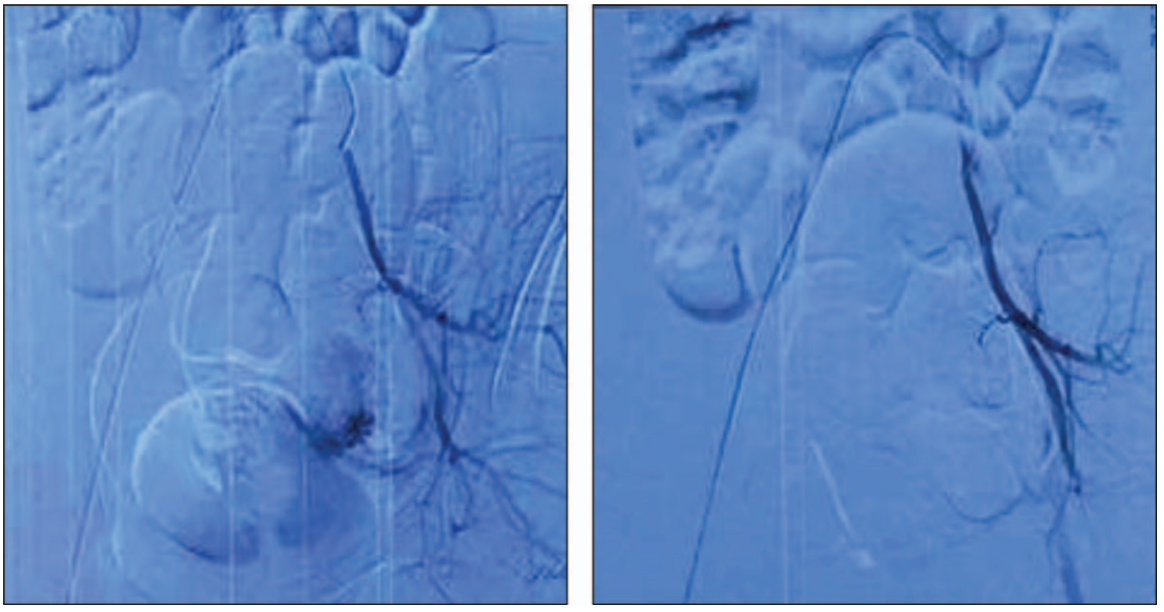

Figure 2. Angiographic results before (a) and after vascular embolization (b).

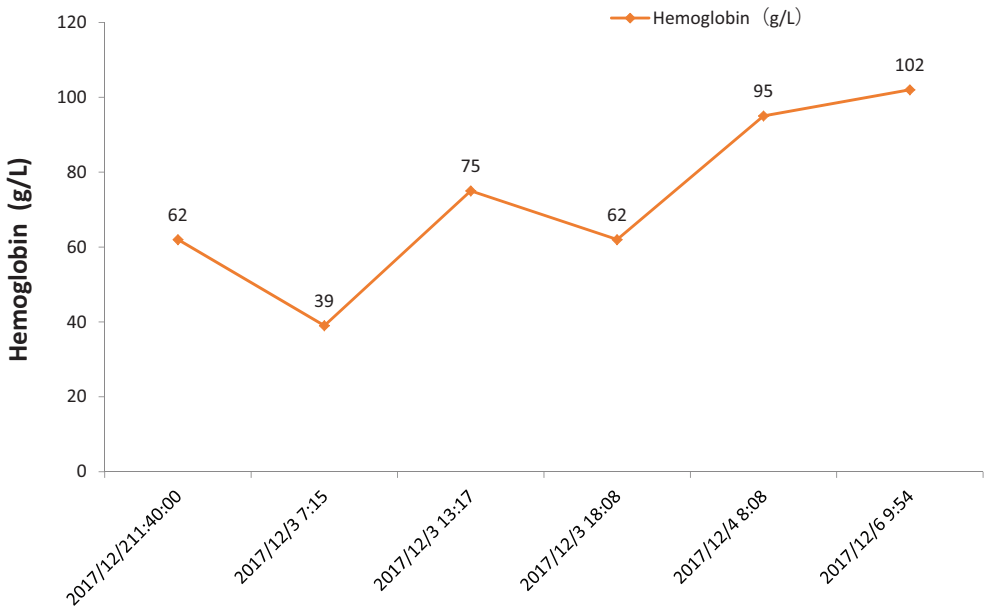

Figure 3. Changes in hemoglobin since admission to hospital.

Postoperative pathological results show that it is bladder myofibroblastic tumor with muscle layer infiltration, finding tumors in the basement (Fig. 4). The results of immunohistochemistry examination showed: SMA (+), ALK (+), Demsin (-); CK (-), with about 20% the tumor cells as Ki-67 positive (Fig.5). Three months after surgery, computed tomography and cystoscopy were reviewed without signs of recurrence and no symptoms such as dysuria appeared in patient.

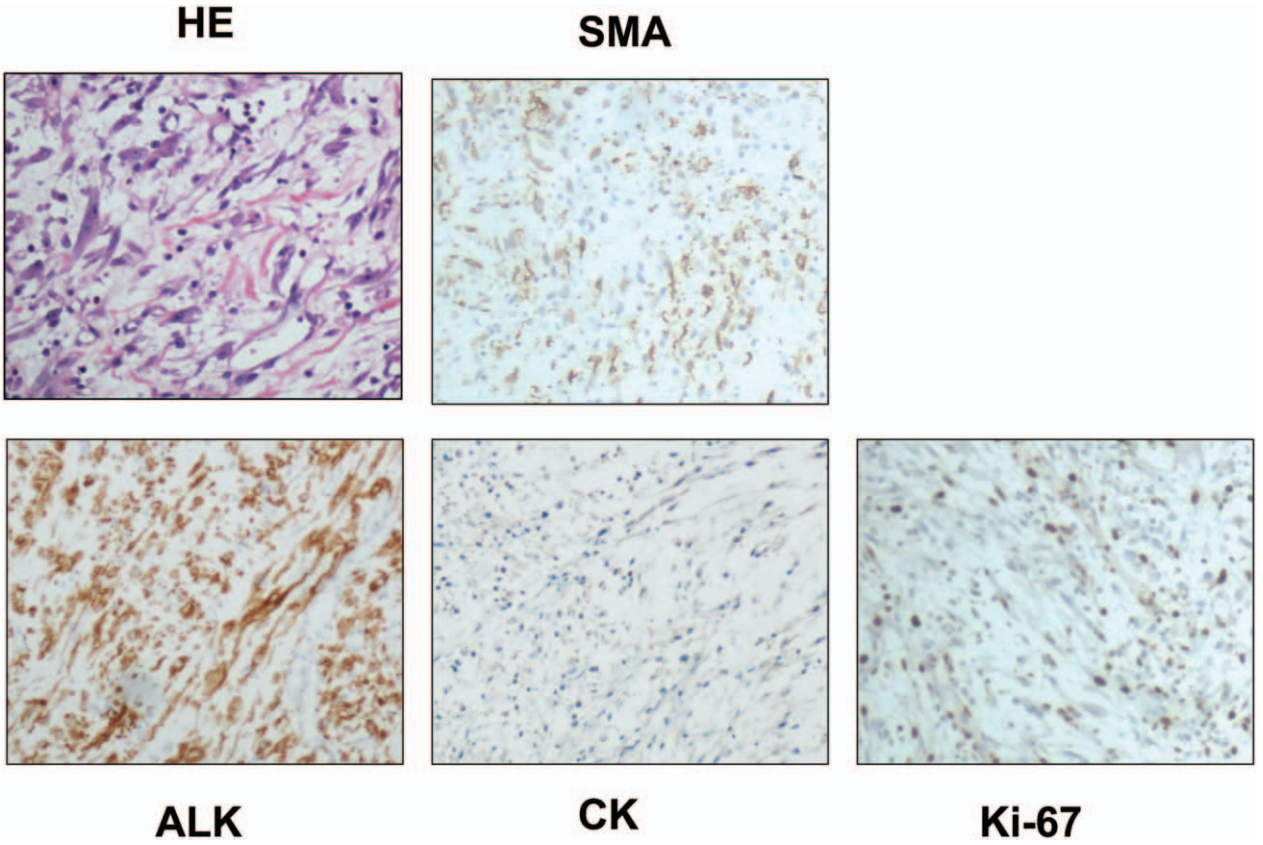

Figure 4. Histological examination (hematoxylin-eosin staining; magnification 10 × 40) shows spindle cell proliferation with a fascicular pattern admixed with inflammatory cells on a myxoid background. And the immunohistochemistry examination results: positive SMA staining (SMA stain, × 400); positive ALK staining (ALK stain, × 400); negative CK staining (CK stain, × 400); Ki-67 staining, with Ki-67 index about 20%. CK = cytokeratin, SMA = smooth muscle actin.

Table 1

Summary of the clinical features and treatment of bladder of IMTs in the literature.

Cases	Age/sex	Clinical manifestations	Hemoglobin (g/L)	Tumor size (cm)	Management
1[12]	38/F	Dysuria and pelvic pain			Urinary bladder transurethral resection
2[13]	26/F	Hematuria; severe anemia		3.3	TUR-BT Radical cystectomy
3[14]	40/M	Hematuria, dysuria, abdominal pain		5	TURBT
4[15]	38/M	Burning micturition; Terminal macroscopic hematuria		3.2	Partial cystectomy
5[15]	56/M	Hematuria	86	6.0	Urgent transurethral bladder resection
6[16]	17/F	Gross hematuria; lower abdominal pain		10*5*4.5	TURBT Partial cystectomy
7[17]	29/M	Painless gross hematuria		4	Robot-assisted partial cystectomy
8[18]	62/F	Visible hematuria	58	4	Urgent TURBT Partial cystectomy
9[19]	71/F	Massive visible hematuria; Suprapubic pain; dysuria	86	3	Transurethral resection
10[20]	52/M	Gross hematuria		3	TURBT Partial cystectomy
11[21]	36/M	Gross hematuria		4*2.5	TURBT
12[22]	31/F	Abdominal pain; dysuria; nocturia; frequency; hematuria		2.3*2*2	Partial cystectomy
13[23]	19/F	Hypogastrium pain		3*4	Partial cystectomy
14[24]	23/F	Gross hematuria; hypovolemic shock		3	TUR Partial cystectomy
15[25]	31/F	Painful urination		4.5	TURBT
16[26]	39/F	Severe hematuria;	48	3.2 * 2. * 2.7	TURBT

IMT = inflammatory myofibroblastic tumor, TURBT/TUR = Urinary bladder transurethral resection.

3. Discussion

Inflammatory myofibroblastic tumor (IMT), a rare borderline tumor, is composed of myofibroblast-differentiated spindle cells and accompanied by numerous inflammatory cells, plasma cells, and/or Lymphocytes.[5] Although it occurs mainly in the lungs, it can also be found in head and neck soft tissue, abdominal cavity, omentum, retroperitoneum and other tissues and organs.[6] But inflammatory myofibroblastic tumor of the urinary bladder is even rarer. A systematic review by Teoh et al evaluated 182 IMT cases and found a mean age of patients of 38.9 years.[7] Hematuria and dysuria are common clinical manifestations, and even more, some patients will also experience severe anemia.[8] Bladder IMTs are more frequently found in young women than in men and are rare in children.[9] Tumor size descriptions range from 1.5 to 13 cm.[8] IMTs in the urinary system commonly occur in the superior wall or the front wall of the bladder.[10] The observation that polypoid nodules on the bladder walls show ring enhancement on contrast-enhanced CT may be valuable in the diagnostic imaging of IMTs of the urinary system.[10] Despite this, it is difficult to make an effective identification of such a rare disease by imaging examination in clinical practices. And the final diagnosis often depends on histopathological features and the immunohistochemical profile. It is important to distinguish this tumor from other malignant spindle cell tumors, such as the sarcomatoid variant of urothelial carcinoma and leiomyosarcoma. We believe that the diagnosis of inflammatory myofibroblastic tumor of the bladder in patients with massive blood loss due to hematuria, especially among young patients, should be taken into consideration. Transurethral resection of bladder tumor (TURBT) is often performed for pathological examination, and additional TURBT or partial cystectomy for radical resection is selected according to histologic outcomes.[7] IMT has a relatively good prognosis and is considered to be a tumor with intermediate biologic potential because of its low risk of distant metastases.[11] IMTs of the bladder have a local tumor recurrence rate after surgery of only 4%, and no patients with distant metastases have been reported.[7]

To study and understand IMT of the urinary bladder further, we reviewed the relevant case reports published since 2013, including a total of 15 articles reported 16 patients (Table 1). Patients' ages ranged from 17 to 71 years (mean 38), in which, females were represented more than males (ratio 5:3). Hematuria (n = 13) are common clinical manifestations in patients. The data are consistent with that of Li Wei, MD and Liang Jianbo, MD,[24] who collected clinic data of IMT through the research of 17 patients, whose chief complaint was hematuria (n = 14). Most of the first treatment options selected TURBT (n = 10), followed by partial bladder resection (n = 8), and then regularly review. What is worth mentioning is that there are 4 cases of emergency surgery due to severe hematuria, in which, the most serious patient is exposed to the hypovolemic shock. Based on previous case reports and the characteristics of this patient, we believe that we should take the possibility of IMT of the bladder among patients, especially young people, with persistent hematuria into consideration in clinical practices, and make timely responses based on the patient's condition to guarantee timely treatment.

4. Consent

Written informed consent was obtained from the participant for publication of this case report. A copy of the written consent is available for editorial review.

Author contributions

Conceptualization: Shimin Zhang, Yuqian Zong.
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Writing – review & editing: wei Jiao, zhiqing fang.

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