

Pseudosarcomatous Myofibroblastic Proliferation of the Urinary Bladder

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Pseudosarcomatous myofibroblastic proliferation (PMP) of the bladder is a rare, benign, and proliferative lesion of the submucosal stroma. We report a 38-year-old female patient who was initially diagnosed with urothelial carcinoma of the urinary bladder under intravenous pyelography. Bladder tumor was resected by the transurethral method, and pathology disclosed a picture compatible with pseudosarcomatous myofibroblastic proliferation. However, local recurrence was found 2 months later, and tumor resection was performed again. The patient has been followed-up at our outpatient department for a year without any evidence of recurrence. [*J Chin Med Assoc* 2008;71(8):431–434]

Key Words: pseudosarcomatous myofibroblastic proliferation, urinary bladder

Introduction

Pseudosarcomatous myofibroblastic proliferation (PMP) of the bladder has been described by many. Roth¹ first described the lesion as a reactive pseudosarcomatous response in 1980. In the past, these lesions have often been initially misdiagnosed as malignancies such as sarcomatoid urothelial carcinoma, leiomyosarcoma, and rhabdomyosarcoma. The patient with PMP presents with symptoms that include urgency, urinary frequency, dysuria and hematuria, urinary obstruction, and pelvic pain.² These tumors can occur at any age, but are usually seen in young adults.^{3,4} PMP usually appears as a polypoid or nodular, sometimes ulcerated, exophytic mass with broad attachment to the bladder wall.^{2,4} Treatment usually consists of transurethral resection or partial cystectomy.⁴ PMP can grow extensively through the muscularis propria to invade the perivesicular adipose tissues, peritoneum, and omentum.⁴ Recurrence has recently been reported in 3 cases,⁴ but it has not been reported to metastasize.^{4,5} We present a female with recurrent PMP of the urinary bladder and review the literature.

Case Report

A 38-year-old woman had sudden onset of gross hematuria for 1 day, and intravenous pyelography showed a large filling defect on the lateral wall of the urinary bladder (Figure 1). Cystoscopy showed a large ovoid tumor on the left lateral wall of the urinary bladder near the left ureteral orifice. Then, abdominal computed tomography was arranged and disclosed a 5.6 × 3.5-cm mass lesion on the left posterior wall of the urinary bladder with muscle layer invasion, but no lymph node lesion was noted (Figure 2). Initially, urothelial carcinoma or tumor of other pathology was suspected. Transurethral resection of bladder tumor (TURBT) was performed, and pathology disclosed a picture of spindle cell proliferation in the submucosal and mucosal layers of the urinary bladder. The cellular proliferation was associated with mild nuclear atypia and eosinophilic cytoplasm within a fibrillary or myxoid background (Figure 3). It was compatible with PMP.

Cystoscopy was performed again 2 months later due to gross hematuria. Local recurrence was found (Figure 4), and TURBT was performed again. The



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Figure 1. Intravenous pyelography revealed an ovoid large filling defect on the left lateral wall of the urinary bladder.



Figure 2. Computed tomography showed a 5.6×3.5-cm mass lesion on the lateral posterior wall of the urinary bladder.

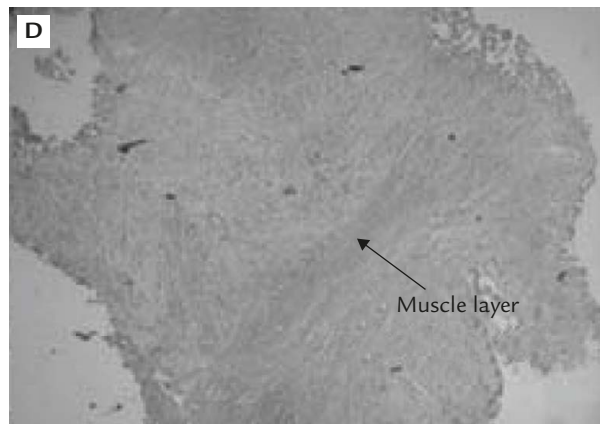
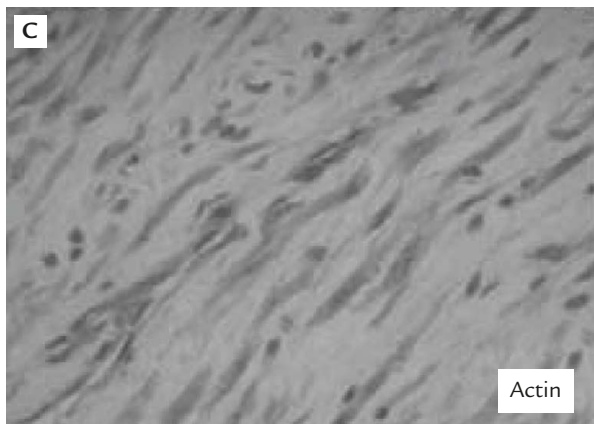
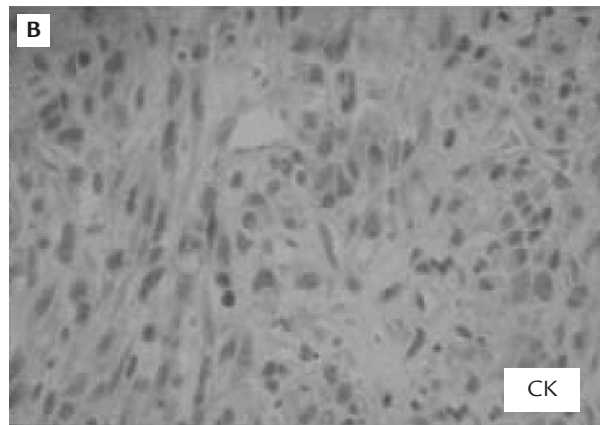
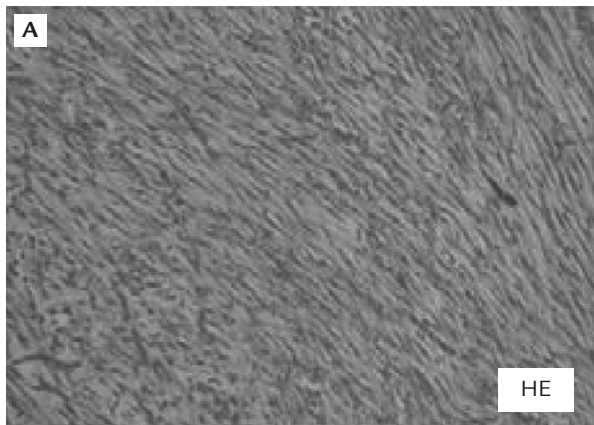


Figure 3. (A) Spindle cell proliferation in the submucosa and mucosa with mild nuclear atypia and eosinophilic cytoplasm within fibrillary or myxoid background was observed (hematoxylin & eosin, 100×). The cells were weakly positive for: (B) cytokeratin and (C) smooth muscle actin. (D) The tumor did not involve the muscle layer (arrow).

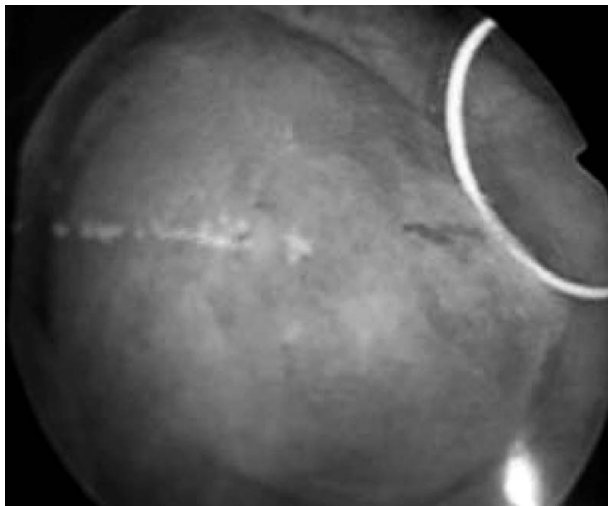


Figure 4. A large ovoid tumor (2.5 × 2.5 cm) on the left lateral wall of the urinary bladder was seen under cystoscopy.

patient has been followed-up at our outpatient department, with no evidence of recurrence for 1 year.

Discussion

PMP of the urinary bladder was first reported by Roth in 1980.¹ He described a 32-year-old woman with recurrent cystitis and hematuria associated with an ulcerated bladder lesion. Unique histologic features included tumor-like, atypical spindle cell proliferation. Histology and etiology were not defined in Roth's report, although chronic cystitis was characteristic in the history.

In subsequent years, many descriptive names have been used to describe histologically similar lesions, including inflammatory pseudotumor, nodular fasciitis, postoperative spindle cell nodule, pseudosarcomatous fibromyxoid tumor, PMP, and inflammatory myofibroblastic tumor. Unfortunately, no consensus on nomenclature seems imminent.⁴

Recent reports show a male predominance (3:1), with a mean patient age range at presentation of 47 to 54 years (range, 3–89 years).^{4,6} Hematuria is the most common presenting symptom (60%), and our case had the same presenting symptom. The following symptoms, pelvic pain (7%), mass lesion (7%), obstructive symptoms (4%) and urinary tract infection (4%) were often noted. Prior instrumentation was reported in 17–21% of cases, and a feature that led investigators to propose at least 2 categories of such lesions was separated chiefly on the basis of whether or not the onset of the lesion was preceded by previous instrumentation.⁴ However, such a separation may be invalid.

Most cases are limited to the bladder, although concurrent involvement of the prostate has been reported.^{4,6} Lesions range in size from 1 cm to 12 cm, and are polypoid or nodular, with variable degrees of mucosal hemorrhage and ulceration. The cut surface is gray-white to tan-pink and often gelatinous. Microscopically, the lesions vary from highly myxoid to highly cellular, and are composed of bland, spindle-shaped myofibroblastic cells, with an associated interlacing vascular network and a variable but polymorphous inflammatory infiltrate. Mitotic rates range from 0 to 20 per 10 high-power field. Invasion into the muscularis mucosa and muscularis propria is common, and infrequently the process may involve perivesical adipose tissue, peritoneum or even omentum. Necrosis, if present, is usually focal and associated with surface ulceration or deep muscularis propria invasion. Immunohistochemically, the spindle cells have expression for epithelial markers such as cytokeratin AE1/3 (94%), and smooth muscle markers such as smooth muscle actin (68%) and desmin (60%).^{4,6}

Transurethral resection or partial cystectomy has been reported as the treatment of choice.⁷

Whether these lesions are reactive or neoplastic is unresolved. It has been shown recently that a t(2;5) involving the *Alk-1* gene can be demonstrated by fluorescent *in situ* hybridization in a substantial number of cases with expression of *Alk-1* protein by immunohistochemistry, supporting the notion that a significant proportion of these histologically similar lesions are truly neoplastic, while the remainder may be either reactive or harbor a different genetic derangement.⁶ Most lesions can be managed by transurethral resection, although more extensive resection is sometimes required. Cases regarded as “typical” occasionally recur locally but do not metastasize.^{4,6}

Iczkowski et al⁵ reported the death of 1 patient who was not a candidate for definitive tumor ablation after transurethral resection of a 13-cm tumor. The tumor grew to 37.5 cm, resulting in urinary obstruction and urosepsis. Sandhu and Iacovou⁷ reported the case of a patient with full-thickness bladder invasion and tumor fixation to the rectus sheath who was treated with 4 months of oral antibiotics. They saw resolution of the lesion at 9 months of follow-up, and the patient was free of recurrence after 3 years.⁷

PMP of the urinary bladder has unique clinical and pathologic features that allow their distinction from primary bladder sarcoma and sarcomatoid carcinoma.⁸ The definition of PMP and inflammatory myofibroblastic tumor of the urinary bladder is still controversial because of the malignant potential.⁹ Transurethral resection or partial cystectomy is sufficient to eradicate

these lesions, with few patients requiring further additional resection. Further research is needed to establish a correct diagnosis among these lesions, which have varying biological potential with important therapeutic and prognostic implications.

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