CASE REPORT

WHEN A BLADDER TUMOR IS NOT WHAT IT SEEMS TO BE!

Pseudosarcomatous Fibromyxoid Tumor of the Urinary Bladder in a 43 year-old Male

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Pseudosarcomatous fibromyxoid tumors are benign tumors of the urinary bladder with clinical presentation similar to that of malignant lesions. This is a case of a 43 year-old male presenting with painless gross hematuria. Personal history and CT urogram are suggestive of a malignant bladder tumor. On cystoscopy, a 1cm x 2.5cm pedunculated polypoid mass was appreciated at the lower lip of the right ureteral orifice. Transurethral resection of the bladder tumor was subsequently done. Ureteroscopy of the right distal ureter was also done revealing no gross involvement of the tumor. Histopathologic examination of the mass revealed pseudosarcomatous fibromyxoid tumor. Review of current literature highlights the rarity of this disease entity, the relative technical difficulty of distinguishing it from malignant lesions, and its benign clinical course excluding the need for more aggressive treatment regimens.

Key words: Pseudosarcomatous fibromyxoid tumor, inflammatory pseudotumor

Introduction

Pseudosarcomatous fibromyxoid tumor (PFT) of the urinary bladder is a rare reactive proliferation of myofibroblast. This was first documented by Roth in 1980 in a 32 year-old woman with recurrent cystitis presenting with gross hematuria and was founded to have a 1.5 cm friable lesion on cystoscopy. Histopathology of the lesion revealed a spindle cell proliferation within a myxoid background with prominent vasculature, and a mild infiltrate of lymphocytes, plasma cells, eosinophils and mast cells.[1] These lesions are frequently composed of atypical spindle cells, making their potential misdiagnosis as a bladder sarcoma or sarcomatoid carcinoma. Despite this, PFT follows a benign course. Hematuria is the most common presenting symptom (60%), followed by pelvic pain (7%), mass lesion (7%), obstructive symptoms (4%) and urinary tract infection (4%).[2] Treatment entails transurethral resection of the tumor with no current reports of local recurrence, metastasis, or disease-specific deaths.

Presented is a case of an adult male with painless gross hematuria. The epidemiology diagnosis and treatment this disease entity in a patient who, initially, was suspected of having a malignant urinary bladder pathology based on clinical presentation.

The Case

Patient is a 43 year-old male, who sought consult due to 2-week history of painless gross hematuria and hesitancy. No fever nor dysuria was noted. He is a tobacco smoker but notes no previous history of urological procedures done nor instrumentation of the genitourinary tract. CT urogram was requested by a private physician revealing a non-specific polypoid nodule in the urinary bladder. The kidneys, ureters, and prostate were unremarkable; no urolithiases were seen.

He was subsequently scheduled to undergo transurethral resection of bladder tumor. Initial cystoscopy revealed a pedunculated polypoid tumor at the lower lip of the right ureteral orifice measuring about 1cm x 2.5cm (Figure 1.) The rest of the bladder mucosa was unremarkable. Resection of the tumor was done up to the muscular layer. Ureteroscopy of the right distal ureter was also done showing no gross involvement of the tumor (Figure 2.)

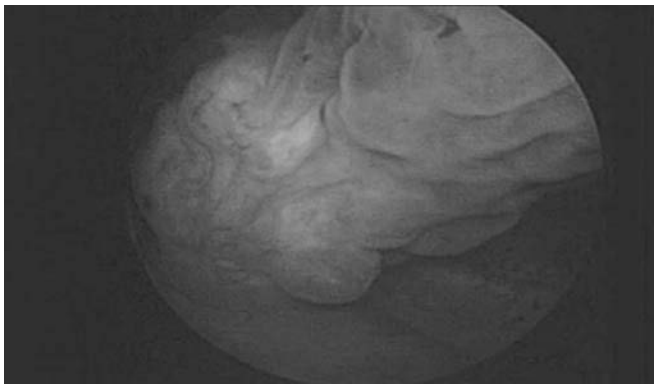

Figure 1. Cystoscopic identification of the bladder tumor at the lower lip of the right ureteral orifice.

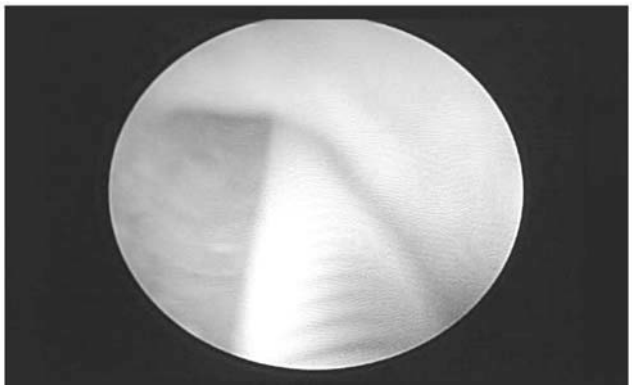

Figure 2. Ureteroscopy of the right distal ureter.

Histopathologic examination of the tumor showed bladder mucosal tissues with proliferation of spindle cells arranged in haphazard fashion in a myxoid matrix. Inflammatory cells, notably eosinophils, were present. Few mitoses are seen. The urothelial lining shows no dysplastic changes. This was signed-out as Pseudosarcomatous Fibromyxoid Tumor (Inflammatory Pseudotumor.) (Figures 3 & 4)

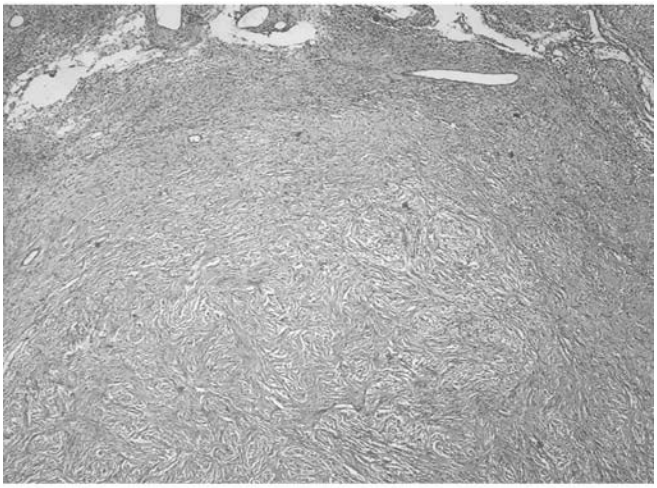

Figure 3. LPO view of the bladder tumor.

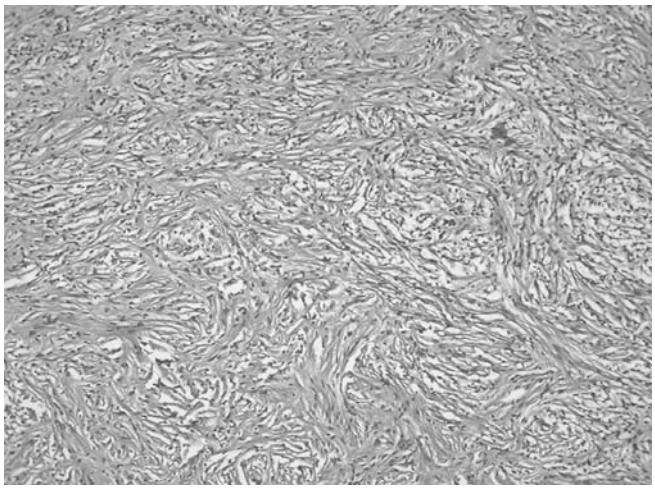

Figure 4. HPO view of the bladder tumor.

Post-operatively, the patient did not note any recurrence of gross hematuria. He followed-up 1, 3, and 10 months post-op wherein routine KUBP ultrasound and urinalysis were done revealing no tumor along the urinary tract and no microscopic hematuria, respectively.

Discussion

In a study by Speiss, et al. involving 7 patients diagnosed with PFT, mean age of presentation was 52 years old (range 37-76) with a male to female ratio of 2:5. Of note in this study is that 71% of patients had previous pregnancies and 14% had previous radiotherapy, although the association of the aforementioned risk factors to PFT is not well established.[3] In contradistinction with Postoperative Spindle Cell Nodule, a disease entity similar to PFT, no history of previous urinary tract instrumentation was noted. In comparison, previous studies by Harick, et al. and Montgomery et. al. showed a male predominance (3:1) with a mean age of presentation of 47-54 years old (range 3-89).[4,5]

In a more recent systematic review of 41 studies done by Chun, et al. on inflammatory myofibroblastic tumors (encompassing PFT), a total of 182 patients were included with a mean age of 38.9 + 16.6 years. Of the total, 51.7% were female patients. The most common presentation was hematuria (71.9%), followed by dysuria (19.8%), urinary frequency (18.8%), lower abdominal pain (13.5%), and loin pain (2.1%).[6]

A study done by Cheng, et al. of inflammatory myofibroblastic tumors (IMT) revealed that about 50% to 60% of IMTs have clonal genetic aberrations in the short arm of chromosome 2 in region p21-p23, specifically a 2p23 rearrangement involving the anaplastic lymphoma kinase (ALK) gene, supporting the concept that IMT is a neoplasm.[7] In lieu of this, Fluoresence-in-situ hybridization (FISH) is used to detect ALK break apart rearrangements. Malignant differential diagnoses for PFT include leiomyosarcoma, sarcomatoid carcinoma, and rhabdomyosarcoma. Leiomyosarcoma can be differentiated histologically from PFT owing to its mild to moderate nuclear pleomorphism, necrosis, and atypical mitotic figures. Sarcomatoid carcinoma normally shows positive immunohistochemical staining for epithelial markers such as cytokeratin and less frequently express smooth muscle cell markers. In this regard, ALK immunostaining and FISH analysis could be valuable in the differential diagnosis. Rhabdomyosarcoma, particularly the embryonal subtype, is an important consideration in children but it can be differentiated from PFT by careful attention to morphological features and positive immunohistochemical staining for MyoD1 or myogenin.[7]

In the previous study by Speiss et. al., all PFT patients underwent transurethral resection of bladder tumor alone. After which, they had a median follow-up of 3 years (range 0.7-13.1.) During this period, no local recurrence, distant recurrence, and disease-specific deaths were noted.[3]

In congruency with this, in the systematic review of Chun, et al., 60.8% of cases of IMT were treated with TURBT, followed by partial cystectomy (29.2%), radical cystectomy (9.2%), and cystoscopic biopsy (0.8%). Among those patients who had TURBT performed, 24.7% of them had further treatment including partial cystectomy (17.8%), second TURBT (5.5%), and radical cystectomy (1.4%).[6] Mean follow-up was 30.0 months ± 28.2 months. Five of 120 patients (4%) were noted to have local tumor recurrence on follow-up. One of them was found to have local recurrence at 6 months, which was treated with TURBT and remained recurrence free afterward. One patient was found to have local recurrence at 1 month; TURBT was performed but unfortunately the tumor recurred again at 4 months and the patient finally underwent partial cystectomy as the definitive treatment and remained recurrence free afterward.[6] For that particular patient who underwent cystoscopic biopsy as the definitive treatment, no tumor recurrence was reported. There was no reported case of distant metastases from IMTs of the urinary bladder.[6]

Conclusion

In conclusion, pseudosarcomatous fibromyxiod tumors of the urinary bladder are rare myofibrobalstic tumors with clinical presentation similar to malignant lesions. Diagnosis of this disease entity can be established by meticulous histologic examination and utilization of immunohistochemical studies. At the present,

available literature confirms its benign course excluding the need for more aggressive treatment regimens.

References

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