Benign Solitary Fibrous Tumor Growth in the Wall of Urinary Bladder, Reporting Two New Cases from Yemen

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Abstract

A solitary fibrous tumor (SFT) of the urinary bladder is a rare tumor with only 25 reported cases in the English literature. Because of its rarity and variable clinical presentation, diagnosis and treatment of these tumors represent a real challenge. Although in most of the cases, the tumor has benign behavior, malignant presentation and co-association with other malignant tumors have been reported also. The diagnosis relies mainly on immunohistochemical staining of the specimen. Proper histological diagnosis is very important as the treatment approach is quite different from malignant bladder tumors. Herein, we report about 2 new cases with SFT presented to us in different periods with different clinical scenarios. In both cases, we faced challenges in diagnosis and treatment for which we are reporting.

Keywords: Solitary fibrous tumor; urinary bladder tumors; rare tumors; spindle cell tumors;

Introduction

A solitary fibrous tumor (SFT) is a rare benign mesenchymal tumor which had been described first as pleural tumor [1]. Later on, it showed to have extra-pleural presentation including urinary tract system. The 1st report about urinary bladder involvement was in 1997 [2]. And since then only 25 cases were reported in the English literature [3, 4]. Because of its rarity, large tumor size and the unusual presentation, the tumor may be confused with other malignant soft tissue tumors mainly sarcomas thus representing diagnostic and therapeutic challenge [3, 4]. The pathological diagnosis usually shows spindle cell tumor and the final diagnosis is confirmed only by immunohistochemical staining which confirms the presence of specific cellular markers for fibrous tissue [5, 6]. Here we present 2 new cases of SFT of the urinary bladder presented with different scenarios and represented for us diagnostic and therapeutic challenge.

Case presentation 1

A 45-year-old female who presented complaining of recurrent painless hematuria associated with dull left loin pain for several months. Radiological evaluation revealed large mass 7x5cm in the left lateral bladder wall causing severe left hydroureteronephrosis with loss of parenchymal thickness (Figure 1). Cystoscopy showed a solid mass in the left lateral wall with intact smooth mucosal surface and non-visualized left ureteric orifice. Transurethral resection of the bladder tumor (TURBT) was done for biopsy purpose. This, unfortunately, was complicated by bladder perforation which was not recognized during the procedure and picture of acute abdomen developed on the 3rd day. We were in a critical situation as we don’t have a pathological diagnosis yet and have a serious complication. We contacted the pathologist for quick diagnosis and it showed benign spindle cell tumor with the recommendation for immunohistochemical staining for more confirmation. We proceeded to immediate laparotomy. Peritoneal lavage was performed first and then the bladder was opened and the tumor was easily enucleated and the bladder perforation was repaired. The definitive histopathology confirmed the diagnosis of a solitary fibrous tumor of the urinary bladder by immunohistochemical staining. The patient passed a smooth postoperative period with favorable follows up.

Case presentation 2

A 30-year-old non-married male presented with obstructive and irritative lower urinary tract symptoms (LUTS), deep pelvic and lower limbs pain, and general fatigue, loss of appetite and difficulty of defecation. Primary ultrasound study showed large pelvic mass with bilateral hydronephrosis. Computed tomography (CT) showed large enhancing pelvic mass 12x8cm protruding into the bladder cavity and compressing the rectum posteriorly (Figure 2a) and causing bilateral hydroureteronephrosis more on the right side. Digital rectal examination (DRE) under anesthesia
**Figure 1:** CT scan abdomen showed a solid mass in the left lateral wall with intact smooth mucosal surface and non-visualized left ureteric orifice.

**Figure 2a:** CT scan abdomen showed large pelvic mass with bilateral hydronephrosis.
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Figure 2b: Showed the tumor was filling the pelvic cavity. Its weight was 37.6g and length was 11x7 cm

revealed large pelvic mass filling the rectum with limited mobility. Cystoscopy showed large mass deviating the prostatic urethra and bulging inside the bladder cavity with smooth intact surface preventing adequate movement and examination of the bladder cavity. TUR biopsy was taken and the pathology after immunohistochemistry showed solitary fibrous tumor of the urinary bladder with positive tissue markers CD 99, CD 34, BCL2, very low proliferation index ki-67 and negative for S-100 and Action. After consultation with the colorectal surgeon, the decision was for surgical excision. The tumor was filling the pelvic cavity but fortunately, it was easily enucleated through midline transperitoneal and trans-vesicle approach. Its weight was 376g and length was 11x7 cm (Figure 2b). After excision, both orifices were identified and bilateral DJ stents were inserted. The patient passed a smooth postoperative course and the final histopathology also confirmed the diagnosis of SFT.

Discussion

The rare incidence and the variable nonspecific presentation of solitary fibrous tumor of the urinary bladder are behind the confusion and challenge in patients who present with this tumor. In many cases, the tumor origin couldn’t be defined precisely and usually the 1st primary diagnosis comes to mind is pelvic sarcoma [3, 4]. Because of this confusion, proper pre-operative pathological diagnosis is of utmost importance for proper planning of the treatment which in most of the cases is organ sparing surgical excision avoiding any unnecessary radical interventions.

In 2016, Tanaka et al reported 2 new cases and reviewed all previously reported 22 cases. They found different clinical presentation according to the size and location of the tumor. These include voiding LUTS in 36%; hematuria in 32%, incidental finding on imaging in 18%; and lower abdominal discomfort in 14% [3].

After this report, only one case was reported in 2017 and with our report, the total number of reported patients with SFT of the urinary bladder will be 27 cases [4]. The para-neoplastic syndrome has been described for the tumor mainly with hypoglycemia due to insulin-like growth factor production by the tumor. It resolves after surgical excision of the tumor [5, 7]. Our patients presented with mixed irritative and obstructive LUTS because of the location and large size of the tumors that occlude the bladder outlet. In addition hematuria developed in the first patient.

The main important diagnostic tool is immunohistochemical staining of the tumor for the detection of specific cellular markers. These include BCL-2, CD34 (90–95%), CD99 (70%), and vimentin [6, 3]. In the 1st case of our patients, the diagnosis of the biopsy specimen was benign spindle cell tumor and no immunohistochemistry was done because the patient developed an acute abdomen and we had to operate on her. However final histopathology confirmed the diagnosis with immunohistochemistry with positive cellular markers for SFT. The 2nd case was managed better and the diagnosis was confirmed in the biopsy specimen which was positive for CD 99, CD 34 and BCL2 and negative for other tumors.

The recommended treatment approach for SFT of the bladder is different. Because of the large size, deep pelvic location with the uncertain origin of the tumor, unclear preoperative diagnosis or there is suspicion to be malignant, open surgical excision of the tumor with bladder preservation is the most commonly reported approach [3, 4, 8]. Nevertheless, if the tumor is of average size, in a suitable location inside the bladder and the diagnosis is confirmed histologically, TURBT is the preferred less invasive treatment approach. However, repeated TURBT may be needed for residual or recurrent tumor [9]. The 1st case of our patients might be suitable for TURBT if the diagnosis was confirmed and no
complication developed. The 2nd case was typical for open surgical intervention because of the large size and deep pelvic location of the tumor.

Despite its benign course in most of the cases, SFT may have malignant behavior in 10-20% of cases [3]. Cheng et al, 2012 reported about malignant SFT of the bladder which required excision and partial cystectomy with favorable outcome [8]. Thus, the prognosis of the disease is favorable in most of the reports with long term follow up approaching 10 years in one report [3]. However, malignant variants and co-association of other malignancies should be considered [8, 3, and 10]. This may require adjuvant chemotherapy and close follow up.

Conclusion

Despite its extremely rare incidence, the solitary fibrous tumor of the urinary bladder should be put in the differential diagnosis of any bladder or pelvic mass which looks well circumscribed on radiological imaging. Clinical suspicion and good communications with radiologists and pathologists are the main clue for proper diagnosis and good planning for treatment.

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Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure statements

The authors have no conflicts of interest to declare.

Authors’ contribution

Khaled M. Al-Kohlany: Idea of the manuscript, writing and literature review.

Waheeb R. Al-Kubati: Revision and supervision of the manuscript

Husam A. Alhamss: Data collection, literature review.

Eman Al-Salamy: Pathological interpretation, supervision of the manuscript.

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