**Inflammatory myofibroblastic tumor of the urinary bladder: A diagnostic challenge and therapeutic dilemma**

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**Abstract**

Inflammatory myofibroblastic tumor (IMT) is a rare, aggressive tumor of indeterminate malignant potential with myofibroblastic differentiation. Though bladder location is very uncommon, it arises from the bladder submucosal stroma as a polypoidal growth and is easily mistaken for a malignant neoplasm—clinically, radiologically and histologically. Essential criteria for the diagnosis of IMT are: spindle myoepithelial cell proliferation and lymphoplasmacytic infiltrate. Here we report the case of a 30 years old man who presented with painless gross haematuria for 2 weeks. The patient underwent open partial cystectomy and the final pathological diagnosis was IMT of bladder.

**Key words:** Inflammatory myofibroblastic tumor, lymphoplasmacytic infiltrate, pseudosarcomatous proliferation, urinary bladder neoplasm

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**Introduction**

Nonepithelial tumors account for 25% of all primary urinary bladder neoplasms. Inflammatory myofibroblastic tumor (IMT) is a rare tumor with a generally indolent, but sometimes aggressive behavior. IMT has been described in major organs, including lungs, liver and skin, along with mesentery and retroperitoneum. Lung is the most common site of occurrence for this tumor. In the genitourinary system, IMT has been found most commonly in the urinary bladder but has also been reported in the kidneys, prostate, ureter, and epididymis. The first case of IMT of the bladder was reported by Roth in 1980.[1] The largest clinicopathologic study comprising of 42 cases of IMT of the bladder have been reported by Harik et al. in 2006.[2] Here, we report a case of IMT of the urinary bladder and discuss its clinical presentation, diagnosis, and management.

**Case Report**

A 30-years-old man presented in the surgery outpatient department with painless gross hematuria for 2 weeks. There was no history of fever, trauma, bladder instrumentation, recurrent urinary tract infections, sexually transmitted disease's or weight loss. Laboratory studies were normal, except for severe microscopic hematuria. Cytological analysis of urine did not detect any malignant cell. Initial abdominal ultrasound showed a 6 cm × 4 cm × 4 cm sized, broad-based polypoidal growth arising from the posterior wall of urinary bladder which was confirmed on computed tomography abdomen as having deep muscle invasion and nonuniform dye uptake. No suspicious lymph nodes were observed. Cystoscopy was done, and multiple biopsies were taken from the tumor. Microscopically, the submitted material showed urothelium with underlying loose spindle cell proliferation with tissue culture appearance. The tumor was composed of plump spindle cells with abundant eosinophilic cytoplasm and elongated nuclei (without nuclear atypia) in a myxoid and inflammatory background of plasma cells and lymphocytes [Figure 1]. Abundant extravasated red blood cells were noted. Mitotic activity was inconspicuous. In addition, tumor cells surrounded by smooth muscle was also seen [Figure 2]. On immunohistochemistry, these spindle cells were strongly positive for AE1/AE3 and focally positive for α-smooth muscle actin (α-SMA). Anaplastic lymphoma kinase (ALK) showed weak reactivity in some cells. The tumor was diagnosed as IMT and open partial cystectomy was done.

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Discussion

Inflammatory myofibroblastic tumor of bladder is an uncommon tumor of controversial nosology; at the edge between benign and malignant tumors and has also been variously named as inflammatory pseudotumor, atypical myofibroblastic tumor, atypical fibromyxoid tumor and Plasma cell granuloma. The term Inflammatory fibrosarcoma has been proposed for the more aggressive tumors of this group. Though any age can be affected, it is more common in the pediatric age group. It is characterized by proliferation of plump, bland spindle cells arranged in a vaguely fascicular fashion in a fibromyxoid and inflammatory background of plasma cells, lymphocytes, and other inflammatory elements. There is a lack of unequivocal malignant features. Anaplastic or pleomorphic features, as well as atypical or bizarre mitotic figures, are absent.

Postoperative spindle cell nodule is a histologically similar, reactive lesion that occurs weeks to months after transurethral resection (TUR) of prostate or bladder lesions. Pseudosarcomatous proliferation is another similar lesion, which shows higher cellularity, more prominent hyperchromasia, prominent nucleoli and nuclear pleomorphism; is more infiltrative and shows strong, diffuse ALK positivity.

There are no known predisposing conditions for its occurrence in the urinary bladder. It is accompanied by fever, anemia and weight loss, all of which remit after tumor excision. IMT shows immunohistochemical positivity for vimentin (strong, diffuse), αSMA, muscle specific Actin, calponin and ALK. Rearrangement of ALK gene on chromosome 2p23 has been noted in these tumors. The pathogenesis of IMT is still in doubt—some regard this entity as a reactive or inflammatory condition, while others believe that it represents a low-grade mesenchymal malignancy. Recent evidence suggests that it is a neoplastic process of low-grade nature because of its aggressive behavior, deep infiltration, occasional coexistence with urothelial carcinoma and the demonstration of a nonrandom chromosomal translocation involving chromosome 2p23 and cytogenetic monoclonality. It has the potential for recurrence and persistent local growth. The therapy of IMT usually includes TUR, partial cystectomy and radiotherapy. Close follow-up is required and complete surgical resection is important to avoid local recurrence.

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