

# An inflammatory myofibroblastic tumor of the urinary bladder in a young girl presenting as acute kidney injury due to obstructive uropathy

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Submitted: 06-05-2015

Revised: 22-05-2015

Published: 01-07-2015

## ABSTRACT

Inflammatory myofibroblastic tumor (IMT) of the bladder is an uncommon benign tumor of the urinary bladder, which is of unknown neoplastic potential, characterized by spindle cell proliferation with characteristic fibro inflammatory and pseudosarcomatous appearance of mesenchymal origin. Myofibroblastic tumor, is also known as inflammatory pseudotumor or pseudosarcoma. Bladder location is very uncommon. We report a case of a young girl who presented with acute kidney injury due to obstructive uropathy. On evaluation found to have Inflammatory myofibroblastic tumor at the base of the urinary bladder, which gradually improved with hemodialysis and successive urological interventions.

**Key Words:** Inflammatory myofibroblastic tumor, Urinary bladder, Pseudotumor

### Access this article online

**Website:**

<http://nepjol.info/index.php/AJMS>

**DOI: 10.3126/ajms.v6i6.12535**

## INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) of the urinary bladder is a rare benign tumor of nonepithelial lesion. This type of tumor has been described in several organs and anatomical sites including lung, head, neck, and brain. It was Roth in 1980 who first described such a lesion in the genitourinary tract, followed by Proppe et al. who reported similar findings in eight patients who had undergone a recent surgical procedure (postoperative spindle cell nodule).<sup>1,2</sup> In the genitourinary system, the tumor usually originates in the bladder, but it has also been reported in the kidney, urethra, prostate, ureter, and rete testis.<sup>3,4</sup>

This case report highlights the rare tumour of the urinary bladder which can present as acute kidney injury due to obstructive uropathy.

## CASE REPORT

A 7-year old young girl presented with history of fever with chills and rigors of four days duration followed by pain abdomen, puffiness of face and swelling of both

lower limbs of two weeks duration and also had history of decreased urine output of two days duration. No history suggestive of lower urinary tract symptoms or trauma or hematuria in the past.

On investigating, the following findings were revealed, urine showed traces of protein, 10 to 12 pus cells, isomorphic RBC of 8 to 10, Blood urea of 209 mg/dl and serum creatinine of 10.5 mg/dl and serum potassium of 7.0 mg/dl. Serum albumin was 3.8 gm/dl, Ultrasound abdomen and CT scan [Figure 1] showed bilateral hydroureteronephrosis with an hypochoic growth of 3.9 cm × 1.5 cm at the base of the bladder.

Patient was initiated on hemodialysis. Renal function stabilized to normalcy with successive hemodialysis and urologist opinion was taken. Cystoscopy was done which revealed growth in trigonal area and right posterolateral area of bladder. Patient was subjected for percutaneous nephrostomy (PCN) of left and right side successively to relieve the obstruction. Transurethral (TUR) biopsy was taken. Histopathology report showed fragments of fibromuscular tissue lined by urothelium and subepithelial

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stroma showed dense mixed inflammatory infiltrate including plasma cells, eosinophils, foam cells and occasional multinucleate cells, reactive lymphoid aggregates and fibroblastic proliferation were seen [Figure 2].

On immunohistochemistry, tumor expressed Vimentin, SMA (smooth muscle actin), ALK-1 (Anaplastic lymphoma kinase) and Calponin. It was negative for CK (cytokeratin), Myf 4 (myofibroblast). Finally patient underwent bladder mass excision with bilateral PCN (percutaneous nephrostomy) with DJ (Double J) stenting, urine output gradually improved and became normal and patient did not require any more hemodialysis.

## DISCUSSION

An IMT of bladder is an uncommon benign tumor of bladder of unknown neoplastic potential characterized by spindle cell proliferation with characteristic fibroinflammatory and pseudosarcomatous appearance. It is also known as pseudosarcoma, atypical myofibroblastic tumor, plasma

cell granuloma, etc. The most common site for this tumor is lung.<sup>4</sup> It can affect any age group, but is more common in children and young adults with female preponderance (F:M ratio 4:3). It is rare in the genitourinary tract with the most common site being urinary bladder.

Inflammatory myofibroblastic tumor is composed of spindle cells in a myxoid stroma, with scattered chronic inflammatory cells with typical mitotic figures and the lesion infiltrates the muscle. The most frequent presenting symptom is hematuria, other symptoms include irritative and or obstructive voiding symptoms, abdominal pain, or the discovery of a mass lesion. Rarely, constitutional symptoms including fever and weight loss have been reported, this is possibly due to the release of cytokines. Grossly, the lesion is either a polypoid mass or a submucosal nodule. Microscopically, the tumor is classically described as a spindle cell proliferation with elongated, eosinophilic cytoplasmic processes in a loose and edematous, or myxoid.

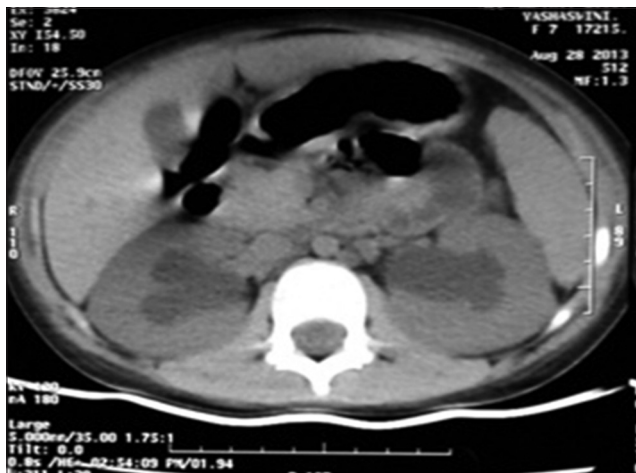
Information on follow-up of cases of IMT has revealed no incidences of metastasis.

Immunohistochemical reactivity in differentiating IMT from other spindle cell lesions of the bladder is indeterminate.<sup>5</sup> Some reports have shown immunoreactivity of IMT for actins, vimentin and for desmin. IMT shows variable staining for epithelial membrane antigen. The main differential diagnosis of IMT includes rhabdomyosarcoma (RMS), leiomyosarcoma and postoperative spindle cell nodule.<sup>6</sup>

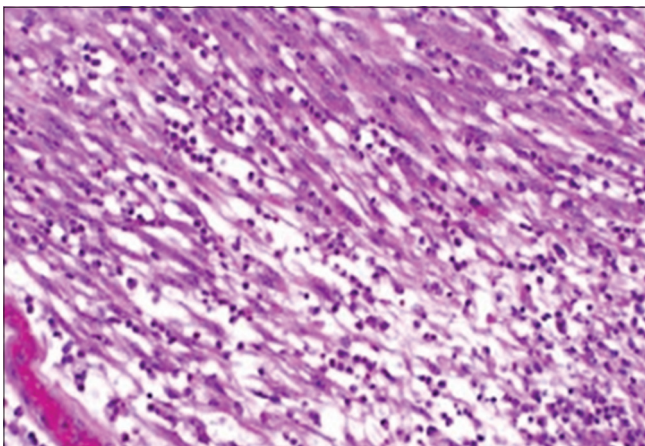
Rhabdomyosarcoma rarely expresses smooth muscle actin, but leiomyosarcoma often does. Myogenin, a potent marker for RMS, helps in exclusion of this tumor.

McKenney states that strong co-expression of smooth muscle actin and cytokeratin is characteristic of vesical myofibroblasts which is typically seen in IMT. IMT also expresses actins, vimentin and desmin.<sup>7</sup> Iczkowski et al reported the death of one patient who died of urosepsis secondary to obstruction by a tumor that had grown to 37.5 cm over 5 months. The treatment of choice is surgical resection.<sup>8</sup>

IMT can be confused with myxoid leiomyosarcoma because both may have a myxoid stroma. Morphologically, leiomyosarcoma is more uniform in its cellularity with more cytologic atypia, whereas IMT has a more prominent network of small blood vessels and a more abundant inflammatory infiltrate.<sup>9,10</sup> The reported extent and location of necrosis in IMT have been variable. In one series, although infiltration of detrusor muscle was present, only superficial necrosis underlying surface ulceration



**Figure 1:** Picture showing bilateral hydronephrosis



**Figure 2:** Picture showing multiple spindle cells with fibroblastic proliferation with scattered inflammatory cells

was observed.<sup>11</sup> Although the overall histologic and immunohistochemical findings must be taken into account, evidence of Anaplastic lymphoma kinase (ALK) -1 expression supports the diagnosis of IMT in the urinary bladder, which was also noted in our case.

IMT is often associated with systemic symptoms, such as fever and weight loss, as well as laboratory abnormalities, such as anemia, thrombocytosis, polyclonal hypergammaglobulinemia, and elevated erythrocyte sedimentation rate. Complete surgical resection is the main mode of treatment and it is also performed to avoid local recurrence.

## CONCLUSION

An IMT is a rare tumor of the urinary bladder. It can present with constitutional symptoms and also as an acute renal failure due to urinary obstruction. Hence high index of suspicion is required for diagnosis. Biopsy is the gold standard for diagnosis and immunohistochemistry is very important to confirm the diagnosis. Surgical resection is the treatment of choice.

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### Authors Contribution:

**UL:** Concept and design of the case report, reviewed the literature, manuscript preparation and critical revision of the manuscript. **SSM:** Concept and review of literature and helped in preparing first draft of manuscript. **NMR:** Conceptualized the rare case report, literature search and interpreted, prepared first draft of manuscript and critical revision of the manuscript. **SKMM:** Concept, literature search, prepared first draft of manuscript and critical revision of the manuscript **LV:** Concept and review of case report. **SCG:** Concept and review of case report.

**Source of Support:** Nil, **Conflict of Interest:** None declared.