Case series

Case series: Inflammatory myofibroblastic bladder tumor in regional Australia

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Abstract

Introduction and importance: Inflammatory myofibroblastic tumor (IMT) is a rare tumor subtype that affects multiple organ systems. This case series adds a regional perspective to the literature, demonstrating rare urological cancers can be managed excellently in regional settings.

Case presentation: We report a case series of two patients in regional Australia who were diagnosed with IMT of the urinary bladder. Both patients received computed tomography (CT) and ultrasound (US) imaging to investigate lower urinary tract symptoms (LUTS). Following initial diagnosis of a bladder tumor, both patients underwent surgery, one having a transurethral resection of bladder tumor (TURBT) with the other receiving a partial cystectomy. Histology from both surgeries revealed IMT of the urinary bladder, with the first case going on to receive a partial cystectomy to ensure clear surgical margins. Both patients recovered well postoperatively, with CT cystograms revealing water-tight bladders. Initial follow up reveals no recurrence of disease.

Clinical discussion: IMT of the urinary bladder is an exceedingly rare clinical entity which is fortunately benign and can be well managed with appropriate surgical intervention.

Conclusion: IMT can be well managed in regional hospitals equipped with appropriate surgical, pathological and oncological services.

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1. Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare, benign tumor which commonly involves the abdomino-pelvic region, lung and retroperitoneum. IMT sits within the broader category of inflammatory pseudotumors, however in recent years IMT has begun to separate from pseudotumors after the discovery of distinct histological and molecular features, specifically characteristic cellular spindle cell proliferation alongside mutations in the ALK gene loci [1]. We present the cases of two female patients diagnosed with IMT of the bladder who were managed in a regional Australian public hospital. This case series has been reported in line with the SCARE 2020 criteria [2].

2. Case presentation

The first case involves a 20 year old female G1P1, three months postpartum who presented to her general practitioner with lethargy and on-going vaginal bleeding post an uncomplicated vaginal delivery. She had no past medical conditions and no relevant family history. Following two courses of oral trimethoprim for a presumed urinary tract infection (UTI) without improvement, the patient began to experience lower urinary tract symptoms (LUTS), mainly incomplete voiding and the urge to double void. After visiting a different general practitioner, an ultrasound scan (USS) was ordered to investigate further, which revealed a 34 × 27 mm lesion arising from the left anterior surface of the bladder. Following this she was referred to the regional urology department where she underwent a computed tomography intravenous pyelogram (CT-IVP) to assess for any upper tract lesions and further typify the lesion (Fig. 1).

After radiological diagnosis of a bladder mass, transurethral resection of bladder tumor (TURBT) was performed. Histology demonstrated marked spindle cell proliferation with morphological variation, some areas demonstrating the spindle cells in well formed fascicles, whilst others were more abundant in myxoid ground substance. There were some areas of tumor necrosis, with the tumor invading into muscularis propria. The tumor stained positive for ALK-1, vimentin, CD10, B-Catenin and SMA, which alongside the microscopic features is suggestive of IMT. Further staining of the specimen revealed ALK-1 positive residual tumor at the site of TURBT with a similar histological pattern. Following this tissue diagnosis, the patient underwent a partial cystectomy by the consultant Urologist which confirmed residual IMT. The repeat resection was clear of all surgical margins. After 5

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days with an indwelling catheter, a CT cystogram was performed which demonstrated a water-tight bladder. Following this, the patient was discharged home (Fig. 2).

The second case involves a 50 year old female who presented to her general practitioner with suprapubic tenderness just prior to, and during urination. Her past history is significant only for hypertension, hypercholesterolaemia and depression. After two courses of oral antibiotics for what a presumed UTI without any clinical improvement, an USS was ordered to assess for any bladder pathology. This USS revealed a presumed urachal cyst and the patient was referred to a regional urologist. A CT-IVP was ordered to assess for any extension of the mass into surrounding tissues, of which there was none (Fig. 3).

A partial cystectomy was performed by the consultant Urologist, with histology demonstrating fascicles of spindle cells in a fibromyxoid stroma with an associated inflammatory infiltrate. There was no evidence of necrosis within the specimen and characteristic features of a urachal cyst were not identified. Immunohistochemistry staining was positive for ALK, SMA and Desmin with resection being clear at all surgical margins. Immunohistochemical staining alongside characteristic morphological appearance was in keeping with an IMT. Post operatively the patient recovered well and following a period with an IDC the patient remains asymptomatic and repeat CT-IVP does not demonstrate any recurrence of disease.

3. Discussion

Inflammatory pseudotumors were first described in 1939 in the lung, where it was initially seen as a reactive post-inflammatory condition as opposed to a neoplasm [1]. Histopathologically similar lesions were subsequently discovered at various extrapulmonary sites, with a predilection for children and adolescents. In 1995 Coffin et al. described three different histological patterns of IMT which may be present simultaneously within a single specimen, a myxoid/vascular pattern, a compact spindle cell pattern and a hypocellular fibrous pattern [3]. IMT of the bladder is an exceedingly rare clinical entity with only 182 cases recorded up to 2014. The average age of these affected is 39 years ± 16.6 years with 51.7% of patients being female. Clinically IMT of the bladder presents with haematuria most frequently (72% of cases) followed by dysuria (20%) and urinary frequency (19%) [4]. IMT tumors have been associated with trauma, surgery and infection, however the majority occur spontaneously. In one of our cases it is possible the patient’s recent vaginal delivery was a contributing factor with regards to bladder trauma (Fig. 4).

Detection of transformations at the ALK gene loci has formed an important part of molecular diagnosis with 33–89% of tumors demonstrating ALK expression, with a particular preference for younger patients.
Alongside ALK expression, vimentin, p53, cytokeratin AE1/AE3 and smooth muscle actin are useful markers for molecular diagnosis, being present in 98.3%, 77.8%, 75.3 and 71.9% of cases respectively [4,5].

With regards to management options, surgical resection is the gold standard. Following diagnosis, the majority of reported cases are managed with TURBT (60.8%) with systematic review suggesting as little as 4% of cases have local recurrence following TURBT, most of which were managed successfully with a repeat TURBT. There are no reported cases of distant metastasis of IMT of the bladder. Cystectomy (Partial or radical) ensures complete resection of disease, however given the benign course of Bladder IMT, TURBT remains an option for those reluctant or unable to undergo major surgery.

Both of the cases were managed in a regional hospital equipped with oncological services that utilise a multidisciplinary team approach to patient care. Expert pathological advice was sought following this rare diagnosis and gold standard management was able to be conducted safely in a regional hospital. This affords the patients the support of family and friends alongside negating the financial burden of travel and accommodation that comes with accessing healthcare when living in a regional or rural area.

### 4. Conclusion

IMT of the urinary bladder is a rare clinical entity. Characteristic morphological appearance alongside immunohistochemistry, particularly ALK staining can aid pathological diagnosis. Surgical excision is the gold standard with TURBT or cystectomy (partial or radical) both appropriate options given the conditions benign course. Patients with bladder IMT can be safely and effectively managed in regional hospitals with appropriate surgical, pathological and oncological services.

**Ethics**

Ethics approval was not required for this case series.

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**Author contribution**

Dr. Tatenda Nzenza: Writing the paper, literature review.

Dr. Sagarika Tripathy: Paper review, Expert opinion.

Mr. Rohan Hall: Paper review, Expert opinion.

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Consent

Written informed consent was obtained from both patients for publication of this case series and accompanying images. A copy of the written consent is available for review by the editor in chief on request.

Declaration of competing interest

The authors of this paper declare no conflicting interests.

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