

Inflammatory myofibroblastic tumour of the bladder: a case report and review of the literature

Bernard Marais^{ID}, Paula Eyal, Ken Kesner and Jeff John*^{ID}

Abstract: Inflammatory myofibroblastic tumours (IMTs) are rare neoplasms of uncertain malignant potential that closely resemble other more aggressive spindle cell tumours. The distinction of IMT from the latter is of importance. We report a case of IMT in a 27-year-old man who presented with intermittent painless, macroscopic haematuria and was found to have a large bladder mass arising from the dome of the bladder. The tumour was resected transurethrally, and histology and immunohistochemistry were consistent with an IMT of the bladder. Our patient remained asymptomatic at follow-up 3 months later, when cystoscopy noted no regrowth of the residual tumour. Transurethral resection of bladder tumour, partial cystectomy and radical cystectomy form the mainstay of treatment of IMT. However, the optimal management of this condition remains uncertain due to the sparsity of reported cases.

Keywords: Inflammatory myofibroblastic tumour, bladder mass, haematuria, transurethral resection of bladder tumour, anaplastic lymphoma kinase

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Introduction

Inflammatory myofibroblastic tumours (IMTs) are rare neoplasms that may be infrequently encountered in the bladder. IMT is a rarity and poses a unique diagnostic challenge because it shares characteristics with other malignant neoplasms.¹ Histologically, IMTs are characterised by spindle cell proliferation with fibroinflammatory and pseudosarcomatous appearance and the many different names assigned to this condition, including inflammatory pseudotumour, plasma cell granuloma, pseudosarcoma, lymphoid hamartoma and benign myofibroblastoma² attest to the uncertainty regarding its true biologic nature. Although generally regarded as benign, they tend to recur locally, and the true malignant potential of IMT is not well established. We report a case of a young male diagnosed with an IMT of the bladder and review the literature on this rare lesion.

Case presentation

A 27-year-old male smoker presented with a 3-month history of suprapubic discomfort and

intermittent, painless macroscopic haematuria. He gave no history of trauma, urinary tract infection, surgical procedures or recent urological instrumentation, and the history and clinical examination were further unremarkable. His urine was frankly blood-stained with small clots, while urinalysis confirmed blood (4+) and the absence of leucocytes or nitrites. A full blood count revealed normocytic, normochromic anaemia with haemoglobin 10 g/dl and a normal white cell count and platelet count. His serum urea, creatinine and electrolytes were normal. Urine culture proved negative. Bed-side ultrasound noted a large bladder mass at the dome of the bladder and no upper urinary tract dilatation. A staging computed tomography (CT) of the abdomen and chest (Figure 1) confirmed a large, lobulated mass arising from the superior and anterior portion of the bladder measuring 4.9 cm × 4.5 cm in size, which contained some low-density central changes in keeping with necrosis and areas of peripheral calcification. No local or distant metastases were demonstrated. Cystoscopy revealed a large, sessile nodular mass

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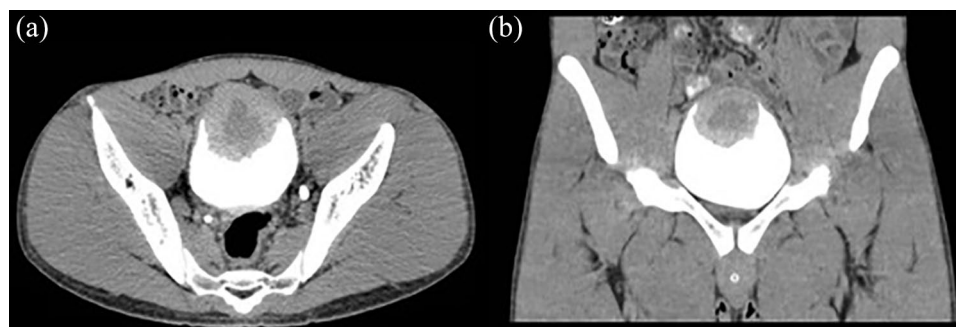


Figure 1. Computed tomography (CT) images – axial (a) and coronal (b) views – showing a large mass involving the dome and part of the anterior wall of the bladder.

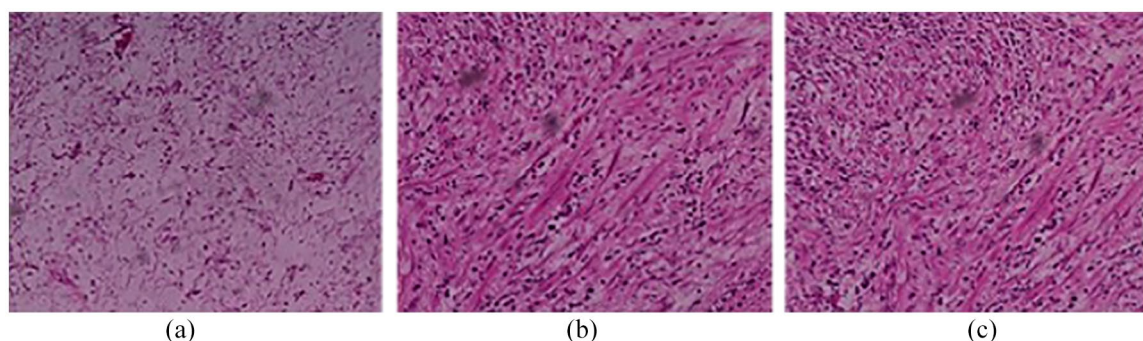


Figure 2. Microscopic images showing spindle and plump polygonal myofibroblasts dispersed in an oedematous and myxoid background with intermingled inflammation (a); compact spindle cell proliferation with a variable myxoid and collagenised stroma with distinctive inflammatory infiltrate (b) and interlacing bundles of spindle cells intermingled with plasma cells and lymphocytes (c).

arising from the dome of the bladder, which was transurethraly resected in its entirety. Histopathology (Figure 2) noted an admixture of spindle cells, fibroblastic/myofibroblastic cells and inflammatory cells. In some areas, loosely arranged, plump to spindle myofibroblasts were present in an oedematous and myxoid background associated with an infiltrate of plasma cells and lymphocytes. Special stains confirmed the presence of patchy stromal mucin. Immunohistochemical staining showed stromal positivity for smooth muscle actin (SMA), and the anaplastic lymphoma kinase (ALK) was positive. The histological and immunohistochemical features were in keeping with an IMT. No obvious detrusor muscle invasion was noted.

Our patient's postoperative course proved uneventful, and he was discharged 2 days after surgery. At follow-up, 6 months later, he remained asymptomatic. Repeat cystoscopy revealed no regrowth or residual tumour.

Discussion

IMT is a rare tumour generally regarded as benign but of uncertain malignant potential. Most cases appear idiopathic, and no specific cause or risk factors have been identified. Although rare, IMT most commonly occurs in the lungs, liver, gastrointestinal tract and involvement of the urinary system rarely occurs. When it does, however, the urinary bladder is most commonly involved.³

IMT was first reported in 1939 in the lung, and the first case involving the urinary system was described in 1980.^{4,5} Histologically, the condition is characterised by atypical spindle cell proliferation, and inflammatory cell infiltrates with a characteristic fibroinflammatory and pseudosarcomatous appearance. This is often accompanied by the presence of plasma cells and lymphocytes. IMTs may display one or more of three basic tissue patterns: a loosely arranged pattern such as in Figure 2(a), a more compact proliferation such as in Figure 2(b) and (c), and a third pattern with

dense collagen and low cellularity that we didn't see in our reported case.

Immunohistochemical staining may be positive for ALK, which is considered pathognomonic for IMT. ALK, a protein overexpressed in anaplastic large cell lymphomas, has recently been reported as overexpressed in IMT in contrast to other spindle cell tumours, and this may be useful in differentiating it from the latter.¹ It is important that we have means of distinguishing IMT from other mesodermal tumours because other sarcomatoid carcinomas, leiomyosarcomas and rhabdomyosarcomas can resemble IMT clinically, radiologically and histopathologically and have vastly different treatment options and prognoses.

Cases of IMT have been reported at any age, but a recent review found the mean age of presentation to be 38.9 years with a slight female predominance. Patients most commonly presented with macroscopic haematuria and dysuria, and 3.1% of patients were hemodynamically unstable on presentation. No abnormal urine cytology was noted.²

While generally regarded as a benign disease with unknown malignant potential, IMT can be locally aggressive, and invasion into the muscularis propria of the bladder frequently occurs. There have been reports of invasion of the prostate and other pelvic structures.⁶

Because IMT can be locally aggressive, complete excision of the tumour is the cornerstone of treatment. However, there is no consensus about the ideal procedure due to the rarity of the condition, and no guidelines are available. The systematic review by Teoh *et al.* reflects the heterogeneity of management of these patients. A transurethral resection of a bladder tumour (TURBT) was deemed adequate therapy in 60.8% of patients, while 29.2% and 9.2% of patients were treated with partial and radical cystectomy, respectively.² Although partial or even radical cystectomy can ensure complete tumour excision, the course of the disease is generally benign, and a less invasive endoscopic resection (TURBT) may be preferable, particularly in patients hesitant to undergo radical surgery or for patients who are poor candidates for major surgery.^{2,7} Recurrence is seen in as many as 10–25% of cases, but such recurrences have been observed to have a relatively benign course.⁷ Tumours that have been incompletely excised have been seen to regress or remain

stable.⁸ There is a lack of evidence to support chemotherapy or radiotherapy in the treatment of these tumours.⁹

The prognosis is generally good irrespective of the choice of surgery, even with local recurrence. Distant metastases are exceptionally rare. To our knowledge, there have only been two cases of distant metastases related to IMT recorded to date.^{6,8}

Despite the rarity of metastases and the relatively benign, indolent course of the disease, close clinical and cystoscopic follow-up is warranted after surgical resection because much remains unknown about IMT and its actual biological behaviour. All cases of IMT of the bladder and urinary tract must be reported in the literature to improve understanding of this condition so that treatment options can be standardised and optimised. Our current knowledge of IMT is sourced only from sporadic case reports and a single review of 182 cases.

Conclusion

IMTs are rare neoplasms with a generally benign course but as yet uncertain malignant potential, closely resembling other more malignant spindle cell tumours. Differentiation of IMT from these more serious cancers by immunohistochemical staining is key to avoiding unnecessary and potentially harmful overtreatment. The preferred treatment of IMT is complete or near-complete surgical resection (TURBT) with close clinical and cystoscopic follow-up.

Ethical approval

Our institution does not require ethics approval for reporting individual case reports.

Informed consent

Written informed consent was obtained from the patient for the anonymised information and the accompanying images to be published in this article.

Author contribution(s)

Bernard Marais: Conceptualisation; Writing – original draft.


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