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Inflammation and Infection

Inflammatory Myofibroblastic Tumor: A Case Study☆

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare benign lesion found in many locations throughout the body and genitourinary tract. Endoscopically and radiographically, these solid lesions cannot be distinguished from malignant bladder tumors. We present the case of a 21-year-old woman who presented with painful obstructive and irritative voiding symptoms of short duration. After extensive preoperative evaluation failed to reveal a definitive diagnosis, the patient underwent partial cystectomy. Final pathology revealed IMT. A high index of suspicion is required for diagnosis of IMT as it is often difficult to distinguish from its malignant counterparts.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare benign lesion found in many locations throughout the body and genitourinary tract. Endoscopically and radiographically, these solid lesions cannot be distinguished from malignant bladder tumors. Diagnosis is based on full resection with histologic evaluation of atypical spindle cell proliferations. We present the case of a 21-year-old woman who presented with painful obstructive and irritative voiding symptoms of short duration. The case and literature review, including presentation, radiographic and histologic findings, and management, are presented.

Case presentation

A 21-year-old G0P0 woman presented to our clinic with severe dysuria, pressured voiding, urgency, and hourly urinary frequency of 3-week duration. She denied fevers, chills, sweats, nausea, and vomiting. She described severe dysuria and low abdominal and perineal pain after micturition. She had no significant urologic history. She was referred with a positive pyridium tampon test (this would indicate a fistula) and difficulty with passage of a Foley catheter for urine culture when she was unable to void. Physical examination revealed a mildly overweight woman appearing in good health. She was afebrile and hemodynamically stable. Pelvic examination was significant for left forniceal tenderness and urine appearing fluid in the introitus. Her laboratory workup was unremarkable. In-office flexible cystoscopy revealed fullness of the left bladder wall including benign-appearing cystic edematous changes. Vaginogram and voiding cystourethrogram did not reveal a fistula, but were remarkable for a left, lateral bladder base filling defect. Computed tomography (CT) urogram revealed eccentric mural thickening of the left bladder base with varicoid enhancement and extravesical stranding surrounding the left fallopian tube (Fig. 1). A delayed left nephrogram was present on a scout film (Fig. 2). A CT-guided percutaneous needle biopsy was performed, which revealed benign smooth muscle.

The patient was counseled on the differential including benign and malignant pathologies. She was subsequently taken for the operating room for exploratory laparotomy with resection of the mass. Examination of the bladder revealed extensive grape-like lesions involving the mucosa of the left bladder wall, base, and trigone. The left ureteral orifice was unable to be visualized. Through a midline incision, multiple open bladder biopsies were sent from the involved region. Initial pathologic diagnoses included both normal urothelium and inverted urothelial papilloma. A 2-cm, full-thickness, solid mass was palpated at the left lateral bladder base in close proximity to the left trigone. Left ureteral access was obtained and a stent was placed. A partial cystectomy was performed and the lesion was resected in its entirety. Gross specimen consisted of a tan-pink rubbery tissue measuring 2.5 × 2.1 × 2.0 cm. Acute and chronic inflammation with benign-appearing spindle cells (Fig. 3) was found,

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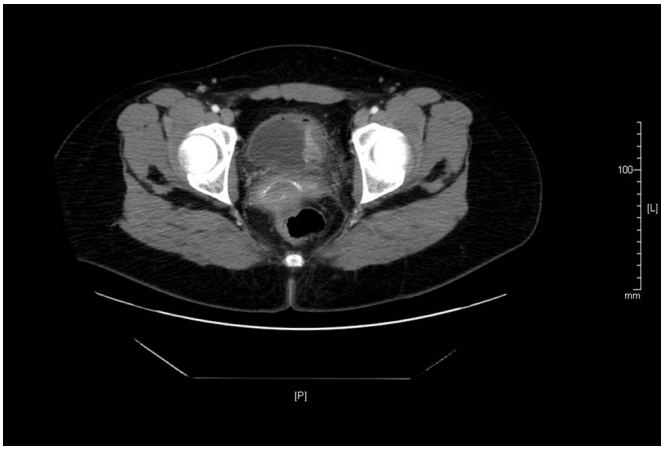

Figure 1. Contrast-enhanced computed tomography during venous phase shows nodular enhancing mass in the left lower bladder with perivesicular soft tissue fat stranding on the left.

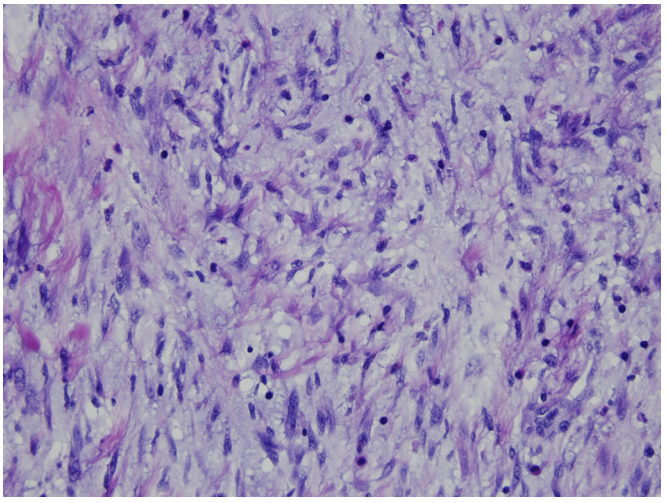

Figure 3. Spindle cells within fibromyxoid matrix and scattered inflammatory cells.

consistent with an IMT. Immunohistochemical staining is positive for calponin and smooth muscle actin and focally positive for desmin.

Discussion

IMT is a rare benign lesion found in many places throughout the body and genitourinary tract. IMT was originally described by Roth in 1980. Dr. Roth presented a case in which a 32-year-old woman was found to have an intravesical lesion composed of spindle cells in a myxoid stroma, with scattered chronic inflammatory cells. The lesion was resected in its entirety without recurrence.[1]

IMT has many designations including inflammatory pseudotumor, inflammatory pseudosarcomatous fibromyxoid tumor, nodular fasciitis, pseudosarcomatous myofibroblastic tumor, and fibromyxoid pseudotumor.[2] IMT most commonly occurs in the lungs but has been described in multiple organs including bladder, liver, colon, spleen, and heart. Although some studies have reported that this entity primarily occurs in young females, others have shown no sex or age predilection.[3] Presentation of bladder IMT most commonly involves painless hematuria, dysuria, frequency, and urgency.[2]

Imaging often provides no benefit in differentiating IMT from its malignant counterparts. Although most IMTs present as intramural lesions without necrosis or perivesical lymphadenopathy, Kim, et al described a mass that was broad based with an enhancing centrally necrotic core involving the bladder wall. Perivascular extension to other pelvic structures appeared to be present on CT.[4]

Histologic appearance is the mainstay of diagnosing IMT. It often reveals a proliferation of spindle cells, which show no atypia, mild nuclear pleomorphism, and rare mitotic activity with diffuse infiltration of acute and chronic inflammatory cells, specifically lymphocytes, eosinophils, and macrophages. Immunohistochemical staining often provides little assistance in diagnosis as similar malignant lesions such as leiomyosarcoma, rhabdomyosarcoma, and sarcomatoid transitional cell carcinoma have similar reactivities. Several recent studies have investigated the use of anaplastic lymphoma kinase (ALK) in the diagnosis of IMT. This is the result of chromosomal translocation of the ALK gene (chromosome 2p23) with a partner gene. These studies have reported positive ALK-1 staining in 30%-75% of IMTs.[5] Although this rate is widely variable, only lymphoma has previously been shown to express ALK-1.

Current standard treatment of IMT is complete surgical resection via either a transurethral approach if possible or an open procedure. Although care should be taken to appropriately counsel patients preoperatively regarding surgical therapies including partial and radical cystectomy with urinary diversion, it is important to recognize IMT and perform an organ-sparing procedure if possible. IMT has not been shown to respond to chemotherapy or radiotherapy. Alternative treatments are currently being investigated and include both anti-inflammatory agents and anti-tumor necrosis factor-α binding antibodies. Although early results are promising, larger prospective studies are needed.

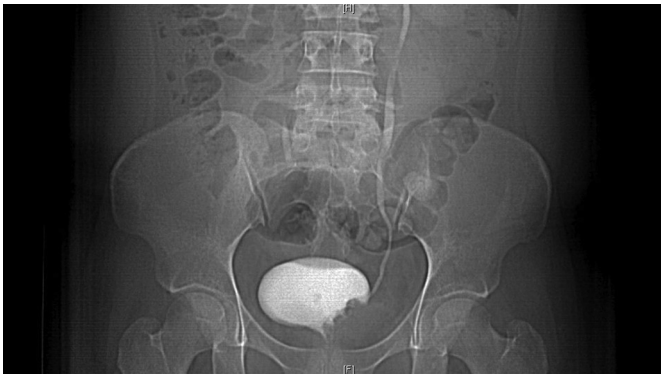

Figure 2. Delayed "scout" view from computed tomography shows nodular mass involving the left bladder base and mild left hydroureter.

Conclusion

In summary, IMT is a rare benign tumor that can present in the bladder. A high index of suspicion is required for diagnosis as it is often difficult to distinguish from its malignant counterparts. Surgical resection is the treatment of choice and care should be taken to appropriately counsel patients preoperatively regarding potential surgical therapies including the need for possible radical cystectomy and urinary diversion. New therapies are on the horizon; however, larger prospective studies are needed before these can be widely adopted.

Acknowledgment

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