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Inflammatory myofibroblastic tumor of the urinary bladder: A prognostically favorable spindle cell neoplasm

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare, benign spindle cell neoplasm of the urinary bladder with a presentation concerning a malignant disease. Oftentimes, these tumors pose a diagnostic dilemma because of a significant overlap with malignant spindle cell tumors in terms of clinical presentation, gross findings, and immunohistopathologic profile. A 28-year-old female presented to us with gross hematuria. Upon work up, the presence of a bladder mass was noted. Cystoscopy and transurethral resection of bladder tumor were done and histopathologic results revealed an IMT. Partial cystectomy was then performed for complete surgical resection.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare benign spindle cell neoplasm that has been identified in various organ systems. In the genitourinary (GU) tract, it most commonly involves the urinary bladder.[1] IMT is often misdiagnosed with other malignant spindle cell tumors based on its their similar cell morphology.[2,3] Hence, it is of paramount importance that these lesions be differentiated from sarcomas of the urinary bladder to avoid unnecessary chemotherapy or radical surgery and their deleterious consequences. Reporting cases of IMT will bring awareness to the features of these rare tumors and will help in overcoming limitations in diagnosis and treatment protocols. We report a case of a 28 -years-old female diagnosed with inflammatory myofibroblastic tumor arising from the urinary bladder.

Case presentation

A 28 year-old female presented at our urology clinic with a 3 months history of episodes of gross hematuria. Patient was initially investigated through Computed Tomography (CT) scan which showed a mass arising from the right superior anterior wall of the urinary bladder measuring 4.3 × 4.7 × 4.6 cms (Fig. 1A). No extravesical component was seen in the CT Scan. Patient is a non-smoker and had a caesarean section 3 months prior to the onset of hematuria. Cystoscopy revealed a smooth walled mass involving the right superior bladder wall (Fig. 1B). Transurethral resection of bladder tumor (TURBT) was performed. Histopathologic examination of the specimens revealed edematous spindle cells with thickened transitional epithelial lining (Fig. 2A). Immunohistochemistry revealed expression of Anaplastic lymphoma kinase (ALK) (Fig. 2B), Smooth muscle actin (SMA) (Fig. 2C), and desmin (Fig. 2D) by tumor cells. The tumor cells were noted to be negative for myogenin. Results were consistent with the diagnosis of inflammatory myofibroblastic tumor.

Patient underwent partial cystectomy and the gross specimen revealed a 4 × 4 cm ovoid mass encased by a pink fibromuscular membrane. Microscopic findings revealed spindle cells with mixed inflammatory infiltrates involving the mucosa, submucosa, and some within the muscle layer without involvement of the adjacent bladder wall and the urachus. Up to date, the patient showed no signs of recurrence after 18 months of follow-up.

Discussion

Inflammatory myofibroblastic tumor (IMT) is a rare benign spindle cell lesion of unclear etiology and malignant potential. Cases have been reported in multiple anatomic locations, with the most frequent site being the lungs. To this point, reports of IMT of the GU tract have been very limited and the most common site reported was in the urinary bladder.[2] These tumors are most commonly observed in adults in their 20s–40s, with a slight female predominance. The most common presenting symptom is hematuria followed by irritative voiding symptoms, and lower abdominal pain. Currently, there are no known predisposing factors for the development of these lesions, however, previous literatures suggest associations between infection, trauma, and previous

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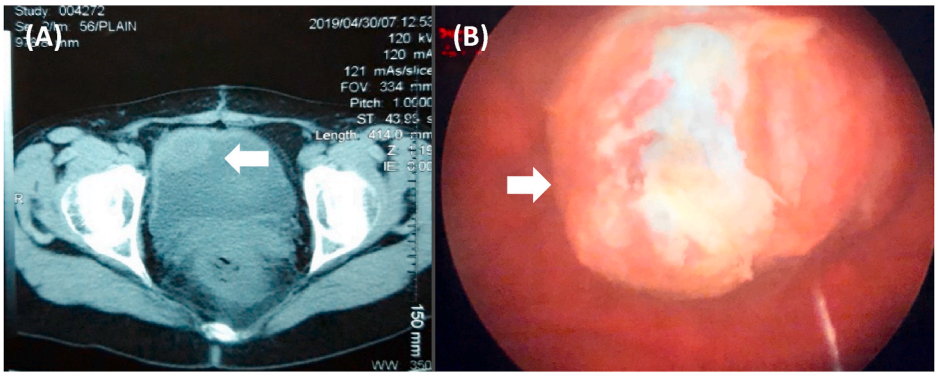

Fig. 1. Preoperative and intraoperative images of the bladder mass (indicated by arrow). A.) Computed tomography (CT) scan, B.) Cystoscopic appearance.

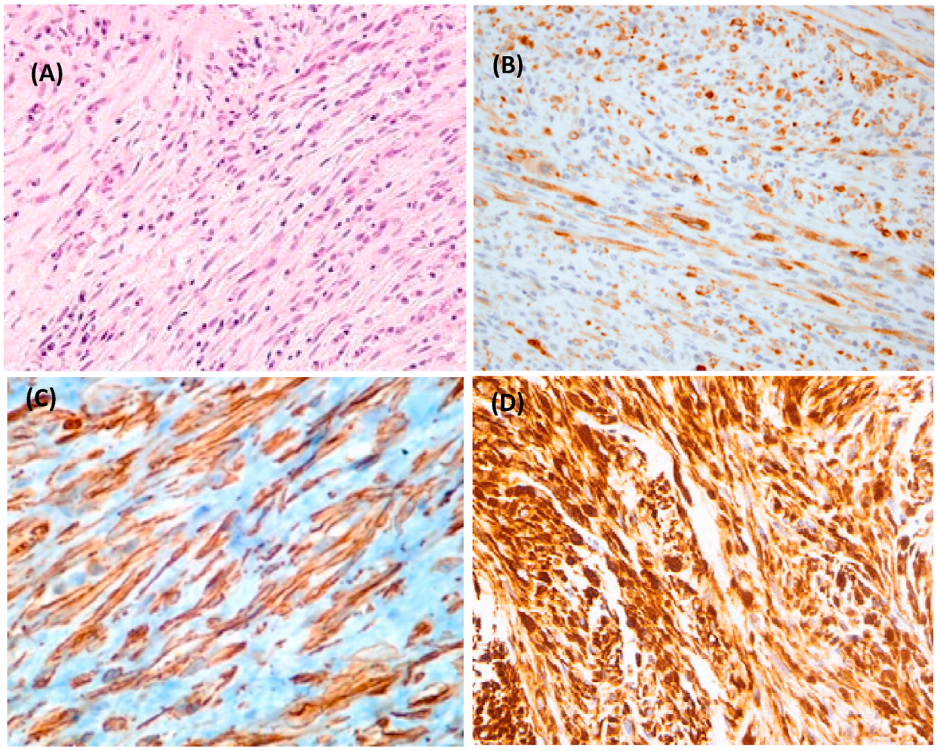

Fig. 2. Immunohistopathologic findings of bladder tumor. A.) Hematoxylin and eosin (H&E) stain, 400x magnification, shows spindle cells in a background of transitional epithelial lining, B.) ALK expression. 400x magnification; C) SMA expression, 400x magnification, D.) Desmin expression, 400x magnification.

surgery prior to development.[2,3] In our case, the patient had a history of previous caesarean section 3 months prior to the onset of hematuria.

IMTs of the urinary bladder are characterized by atypical spindle cell proliferation, accompanied by inflammatory cell infiltrates comprised primarily of lymphocytes and plasma cells. These tumors exhibit morphologic and immunohistopathologic overlap with malignant spindle cell tumors such as leiomyosarcoma, rhabdomyosarcoma, and sarcomatoid carcinoma.[1,3] Hence, the diagnostic distinction can be problematic at times because epithelial and myogenic markers can be both expressed by inflammatory myofibroblastic tumors. Final and definitive diagnosis can only be made by immunohistopathologic studies.[1,3] Immunohistochemical staining for IMT may demonstrate positive results for Anaplastic lymphoma kinase (ALK), smooth muscle actin (SMA), desmin, cytokeratin, p53, and vimentin.[3] The most defining feature of IMT is the positive immunostaining for ALK which is absent in other spindle cell tumors of various anatomic locations.[4] However, only 67% of documented cases of IMT showed positivity for ALK by immunohistochemistry.[5] Based on a systematic review of 186 cases of IMT of the bladder by Theo et al. (2016), Immunohistochemical staining results revealed positive results for ALK (65%), vimentin (96.3%), p53 (77.8%), CK AE1/AE3 (75.3%), SMA (71.9%), desmin (43.8%). In addition, all cases were negative for myogenin, CD 21, CD 34, and CD 35.[5] Consistently, in our case presented, immunohistochemical staining results revealed positive for ALK, SMA, and desmin, while negative for myogenin. Hence, ALK immunohistochemical staining can be a useful adjunct in the differentiation of IMT from other spindle cell lesions of the urinary bladder.

IMT of the urinary tract has been associated with a more indolent course and a relatively good prognosis due to its low risk of distant metastasis. However, due to the limited cases documented in the literature, no current guideline regarding the treatment strategies for managing IMT. Complete local resection with negative surgical margin is the more preferred treatment. This can be achieved by transurethral resection or by partial cystectomy.[3,5] Of the 182 patients in Teoh et al.'s systematic review, 60.8% were treated with TURBT, 29.2% with partial cystectomy, and 9.2% with radical cystectomy. Despite a typically

benign course, 5 of these 182 patients developed local recurrences, representing an overall 2.7% rate of local recurrence. Two of these five recurrences occurred within a 6-month window after the primary excision and required treatment with a second TURBT. Only one single case of metastases related to IMT was reported.[5] As in our case, transurethral resection was done to establish the diagnosis and subsequently a partial cystectomy was performed for complete surgical resection.

Conclusion

Inflammatory myofibroblastic tumor is a benign neoplasm that presents similarly to malignant spindle cell tumors of the urinary bladder. Differentiation from malignant spindle cell tumors through immunohistopathologic studies is of paramount importance to avoid overdiagnosis and the consequence of treatment with radical surgery and chemotherapy. ALK staining can be a useful adjunct to differentiate IMT from other spindle cell lesions of the bladder. Management is aimed at complete local resection through transurethral resection or partial cystectomy is the more advisable treatment. Due to their unknown malignant potential, these tumors warrant close follow-up with CT scans and surveillance cystoscopy.

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