Case Report

Inflamatory Pseudotumor of the Bladder: A Histopathological Diagnostic Challenge from Its Malignant Variant

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Abstract

Inflammatory Pseudotumor (IP) of bladder is one of a variety of lesions which may arise from spindle cell lesions in the bladder. It is benign in nature compared to the other end of the spectrum of spindle cell lesions, such as sarcoma, sarcomatoid carcinoma and leiomyosarcoma. The diagnosis of IP and its differentiation from a malignant pathology is a diagnostic dilemma for Urologist and Pathologist as both entities share certain similar morphological and histo-pathological appearance as well as immuno-histo chemistry staining. It is however crucial to avoid misdiagnosis as the treatment option varies significantly between a radical or partial cystectomy with future surveillance cystoscopes. Hereby, we highlight a case of a benign variant of spindle cell tumour of bladder which needed second pathology review for confirmation and thus avoiding a radical cystectomy. We also review the literature on its presentation and emphasis characteristics differentiating a benign from a malignant pathology.

Keywords: Bladder tumour, pseudotumor, myofirbroblastic, spindle cell

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Introduction

Spindle cell tumors of the bladder is not uncommon and it consists of a spectrum of benign to malignant variety. Inflamatory pseudotumor (IP) is a benign type of spindle cell tumor and is rare (1). It was originally described by Roth et al in 1980 (2). Patients usually present with painless hematuria, recurrent cystitis (3) and rarely urinary obstruction resulting from the large bladder mass. Uncommon presentations include bladder calculi and large palpable abdominal mass (4). It has no sex predilection and has been reported in age ranges from 15 to 74 years (3). However it is commonly seen in younger women (1). Cystoscopy findings may reveal a polypoidal , intraluminal or a submucosal mass. The dilemma of diagnosis in IP is in its certain similar histopathology characteristics with a malignant sarcoma. This dilemma is shared by the urologist as well since subsequent management differs greatly between a radical cystectomy or a local resection and regular surveillance. Here we report a middle age lady with the above mentioned problem and review the literature on pathological characteristics which can aid to differentiate a benign IP from a malignant sarcoma.

Case Report

A 46- year- old lady presented with painless hematuria of one month duration to a private medical center. There were no symptoms to suggest urinary tract infection (UTI) or previous episodes of UTI. There

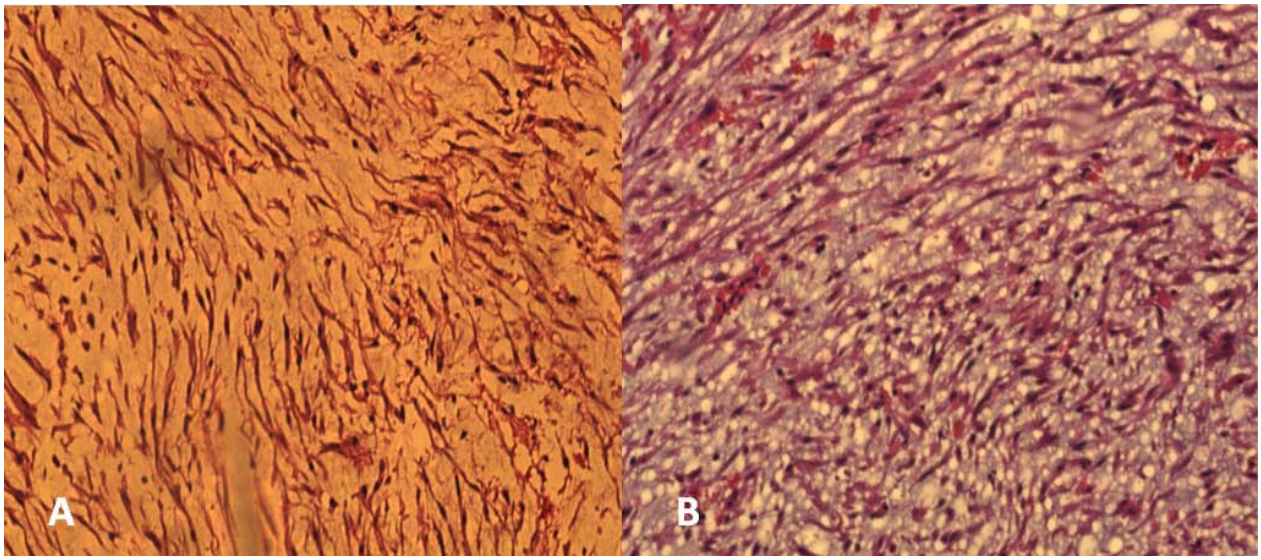

Figure 1: A) Proliferation of slender spindle cells (high power x 10), B) Myxoid and oedematous stroma (high power x 10)

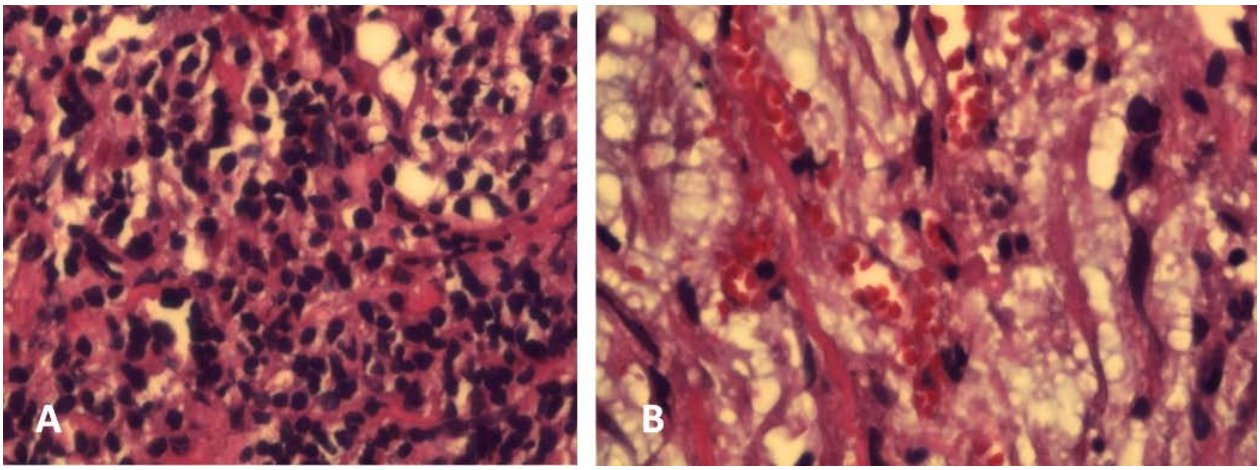

Figure 2: A) Foci of inflammatory mononuclear component composed of lymphocytes and plasma cells (high power x 40), B) areas of vessels proliferation (high power x 40)

was no previous bladder or urinary tract instrumentation or trauma. There were no constitutional symptoms. Cystoscopy revealed a 3 x 2 cm polypoidal mass arising from the left lateral wall of the bladder with superficial ulceration. In view of a possible malignant pathology a transurethral resection of bladder tumour (TURBT) was performed. Ultrasound of the upper urinary tracts were normal.

An initial histopathology diagnosis of a malignant spindle cell tumour of the bladder was made. The patient was offered cystectomy with an ileal conduit reconstruction. The patient sought a second opinion in our medical institution and the histopathology slides were reviewed. It showed that the mass consisted of proliferation of slender spindle cells (Fig. 1A) in a myxoid and oedematous stroma (Fig. 1B). There were foci of inflammatory mono-nuclear cells composed of lymphocytes and plasma cells (Fig. 2A) and micro vessels proliferation (Fig. 2B). Mitotic count is one per 10 high power field (hpf). Immuno-histo-chemistry staining showed positivity to vimentin, S100, CK and CD31. It was negative for MyoD1, Desmin, HMWCK and ALK. With this, a diagnosis of a benign inflammatory pseudotumour of the bladder was diagnosed and the patient was spared a radical cystectomy. A repeat cystoscopy six weeks after TURBT showed no recurrence of tumour. The patient is currently on three monthly surveillance cystoscopy with no recurrences detected over the past one year.

Discussion

A variety of names have been given for the benign variant of bladder spindle cell tumour such as IP, inflammatory myofibroblastic tumour (IMT), pseudo-sarcomatous myofibroblastic proliferation (PMP) and post operative spindle cell nodule (PSCN) (5). Malignant variant of them include leiomyosarcoma, rhabdomyoarcoma and spindle cell carcinoma. All the nomenclature given above reflect the fundamental pathological abnormality detected in these lesions that is the presence of an inflammatory process, proliferation of fibro-collagenous tissues, abundant spindle cells and high vascularity (1).

Spindle cell tumour of bladder is reportedly associated with neurogenic bladder and those with a history of bladder instrumentation (1). The term PSCN is applied for lesions similar to IP that arises within a 3 month time frame from bladder instrumentation. This produces an inflammatory pseudotumour which arises from the submucosa of the bladder under a normal transitional cell epithelium (6).

IP is benign, does not metastasis and is not a pre malignant condition. It is associated mortality previously reported was from a large bladder mass causing urinary obstruction with urosepesis. However, synchronous urothelial carcinoma has been reported with IP (7). Hence surveillance cystoscopy is necessary even if IP is diagnosed.

IP was initially considered as a non-neoplastic lesion and represented an aberrant inflammatory response. The dilemma occurs as several aspects of IP are neoplastic in nature, that in its potential for recurrences, possible multifocal involvement, infiltrative local growth and vascular invasion. Additionally it may show muscularis propria involvement (3).

Pathological characteristics that support a benign IP include the absence of nuclear hyperchromasia, absence of atypical mitosis and absence of necrosis at tumour - detrusor muscle interface. Benign IP however may demonstrate mitosis of 0-1 per 10 high power field (hpf) as compared to sarcoma (1-16 per 10hpf) (1). Presence of necrosis, myxoid degeneration and nuclear atypia were the best criteria to differentiate a benign from malignant pathology. The malignant sarcoma is reportedly to have less prominent microvasculature and without a history of prior instrumentation.

Immunohistochemistry staining aids slightly in differentiating the two. IP demonstrates positivity towards vimentin and actin. Regarding sarcoma p53 is the common denominator and is absent in IP (5). Other markers like desmin, is usually not positive in IP. Fibronectin has been reported to be specific to IMT.

Hematuria is a common presentation in IP lesions as in our patient. It is due to its high tumour vascularity and associated superficial ulceration (1). Our patient did not have any history of bladder instrumentation making PSCN an unlikely diagnosis.

Tamoxifen was found to be effective for IP. It acts by modulating production of growth factors such as epidermal growth factor, insulin like growth factor and transforming growth factor. This results in reduction of fibroblast and spindle cell proliferation (1).

Surgery, however, is the main stay of treatment. Surgical treatment of IP is usually sufficient with a transurethral resection. Recurrence is uncommon though it is thought to be more of a case of incomplete resection rather than a true recurrence itself. Surveillance cystoscope is recommended to look out for local recurrence and pick up any missed concurrent malignant lesion. The surveillance protocol would avoid any mismanagement if a wrong initial pathtological diagnosis was made.

Our patient avoided a radical cystectomy and its associated morbidities after careful re-inspection of the pathology specimens. Currently the surveillance protocol has yield no recurrence over the past one year.

Conclusion

IP of the bladder is a benign condition that has a good prognosis and should be carefully diagnosed and differentiated from its malignant counter-part, spindle cell sarcoma. However, the complexity and large variant of spindle cell lesions poses a diagnostic challenge and potentiates a misdiagnosis. Here, we stress of the importance and awareness in differentiating benign and malignant bladder spindle cell lesion by careful inspection of the presenting history, clinical presentation, it's pathological and immunohistochemistry characteristics to avoid error in management.

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