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INFLAMMATORY MYOFIBROBLASTIC TUMOR OF THE BLADDER: A CASE REPORT

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Abstract: Inflammatory myofibroblastic tumor (IMT) is a rare benign neoplasm characterized by proliferation of spindle cells with a characteristic fibroinflammatory and pseudosarcomatous appearance and has a low potential for malignancy or metastasis, despite being locally aggressive, since current evidence suggests recurrence or multicentricity. Of still unknown etiology, it may be related to infections, trauma or previous surgeries. It occurs between the second and fifth decades of life, more frequently in males. The clinical, radiological and macroscopic appearance simulates a malignant disease, and it is important to differentiate them before any definitive surgical procedure. Due to the rarity of bladder IMT, the purpose of this study is to report the case of a patient in our service, with the purpose of providing information related to the diagnostic and treatment challenges in IMT, thus providing knowledge to make better clinical decisions when faced with similar cases that may arise in daily clinical practice.
Keywords: inflammatory myofibroblastic tumor, spindle myoepithelial cell proliferation

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare benign neoplasm characterized by proliferation of spindle cells with a characteristic fibroinflammatory and pseudosarcomatous appearance1,2 and presents with low malignant potential. Mortality usually occurs due to tumor extension to adjacent organs3.

The first description of IMT of the bladder was in 1980 by Roth4, as a pseudosarcomatous inflammatory response that mimicked a malignant tumor in a patient with chronic cystitis.

Of unknown etiology, it may be related to infections, trauma or previous surgeries5.

It is also known as pseudosarcoma, atypical myofibroblastic tumor, atypical fibromyxoid tumor, plasma cell granuloma, etc.5

The clinical presentation includes nonspecific symptoms of the urinary tract. It occurs between the second and fifth decades of life, and is more frequent in males (5:1) 2,3,5. The clinical, radiological and macroscopic appearance simulates a malignant disease, and it is important to differentiate them before any definitive surgical procedure.

A gene associated with the formation of IMT was identified in immunohistochemistry assays, as well as its decoded protein, ALK1 (Anaplastic Lymphoma Kinase) 4,5,6,7,8.

This report aims to describe a case of IMT of the bladder diagnosed and treated in our service (Uro-Oncology of the Alfredo Abrão Cancer Hospital) located in the city of Campo Grande, MS. We describe information related to the diagnostic and treatment challenges in IMT, thus providing knowledge to make better clinical decisions when faced with similar cases that may arise in daily clinical practice.

CLINICAL CASE

A 45-year-old white, unemployed female patient was taking antihypertensive medication (Losartan) for systemic arterial hypertension. Her history included breast cancer with resection of the left breast and local radiotherapy and total hysterectomy for uterine fibroids. Her family history included a mother with a history of breast cancer and a father with skin cancer (squamous cell carcinoma). Her sole and main complaint was dysuria. She was diagnosed with a lower urinary tract infection and prescribed antibiotic therapy for 10 days. With no improvement in her condition and the emergence of a new symptom, urinary incontinence, she decided to seek medical help again.

Laboratory tests showed 3+/4+ hematuria, with no other alterations.

A computed tomography scan of the abdomen and pelvis revealed the presence of a large intravesical endophytic formation. She was referred for follow-up with the Uro-oncology team at our service.

After clinical, laboratory and radiological evaluation, with high suspicion of primary or secondary malignant etiology, a total abdominal MRI was indicated to plan the biopsy approach.

The total abdominal MRI showed a polypoid lesion in the urinary bladder on its right posterior wall measuring 6.5 x 5.6 x 4.7 cm, suspicious for neoplastic involvement, with involvement of the right ureterovesical junction (UVJ), causing marked hydronephrosis upstream on this side (Figures 1 and 2).

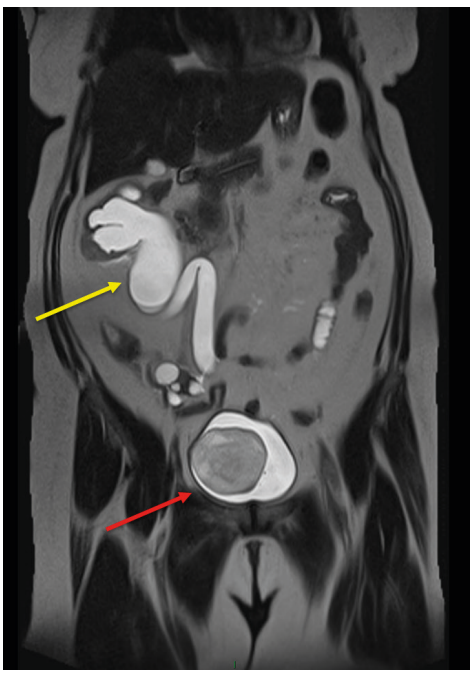

Figure 1: Coronal section with T2 weighting of the total abdominal MRI, demonstrating the intravesical mass (yellow arrow), causing dilatation of the collecting system on the right (red arrow)

She underwent her first transurethral resection (TUR) (July 2023) of the bladder in order to obtain a sample of the lesion for biopsy. During surgery, a large solid lesion with a smooth surface and polypoid appearance was seen, measuring approximately 7 cm in diameter. Partial resection of the lesion was performed (Figure 3).

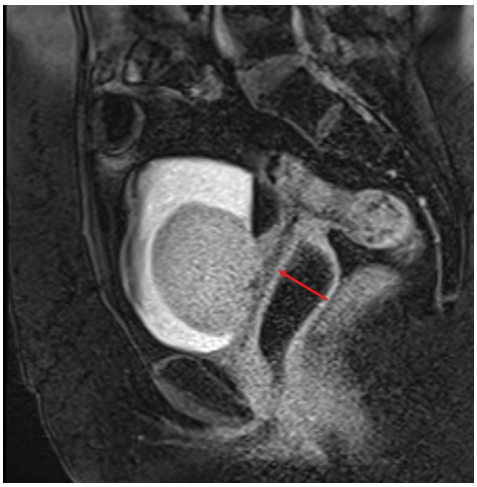

Figure 2: Sagittal section with T1 weighting after contrast of the total abdominal magnetic resonance imaging, demonstrating an intravesical mass in close contact with the inner layer of the urinary bladder (red arrow)

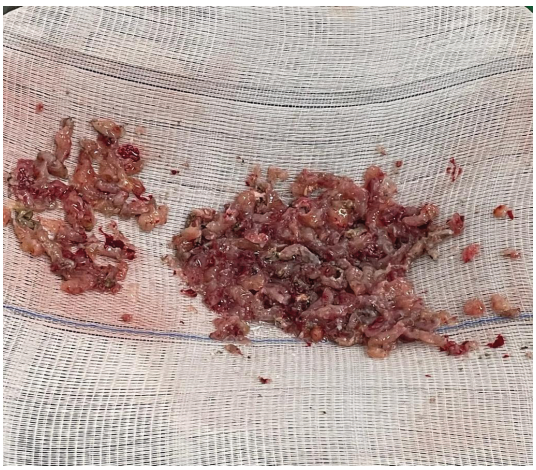

Figure 3: Macroscopy of the lesion, product of TURP of the urinary bladder, characterized by irregular fragments of brownish and friable tissue.

The anatomopathological result demonstrated the existence of spindle cell neoplasia amidst myxoid stroma, weighing approximately 118 g, with a suggestion for immunohistochemical analysis.

In the immunohistochemistry, the tumor revealed focal positive AML marker and diffuse positive ALK. Other specific markers were negative. The panel associated with the histological aspects was suggestive of ALK-positive inflammatory myofibroblastic tumor.

In the control cystoscopy, a new large mass was visualized, with a new approach scheduled and performed for complete resection by TUR in August 2023, which was performed with technical success, with 78 grams of neoplastic material being removed.

In the control cystoscopies (April/2024 and August/2024), no masses or nodules were visualized, and urine cytology was negative for malignant cells.

In the following months, the patient progressed without complaints, with improvement of symptoms in the postoperative period, and continues to be monitored by the Uro-oncology team at the cancer hospital for periodic cystoscopies and imaging exams, the last one performed in August 2024, without detecting local recurrences (Figure 4), maintaining slight dilation of the collecting system on the right (Figure 5) to date.

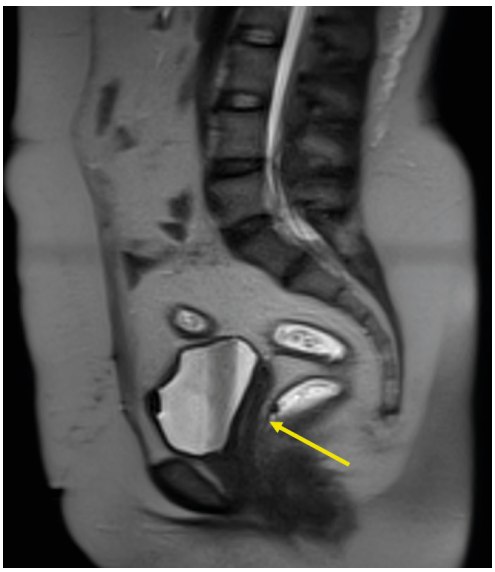

Figure 4: Sagittal section in T2 weighting demonstrating absence of intravesical tumor lesion (yellow arrow).

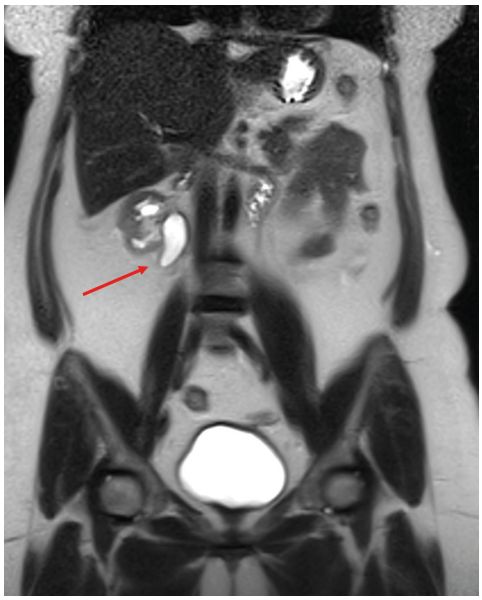

Figure 5: Coronal section with T2 weighting of the total abdominal MRI demonstrating pyelocaliceal dilatation in the right kidney, with signs suggestive of narrowing at the ureteropelvic junction, without characterization of an obstructive process.

DISCUSSION

IMT is a rare tumor that affects children and young adults between 3 and 89 years of age 5,7,8,9. It is a neoplasm described in several organs, such as the liver, bile duct, lungs, and gastrointestinal tract. In the urinary system, the bladder is the predominant location 10,11.

There are several hypotheses about the pathogenesis of IMT, such as immunological, allergic, multifactorial mechanisms, and inflammatory response and aberrations. It may be associated with recurrent infections, repeated cystoscopy, and use of medications such as cyclophosphamide or foreign bodies in the bladder 7,8,9,11,12.

Preoperative diagnosis is challenging due to the nonspecificity presented in the images and similarity to the symptoms of urothelial carcinoma. On ultrasound, IMT in the urinary bladder and ureter presents as a hypo- or isoechoic mass 12,13,14. CT or MRI are always necessary when a mass is detected by ultra-

sound. CT scans usually demonstrate masses with hybrid density originating from the bladder wall, which may or may not determine mass effect in the ureteral meatus 12,15.

On MRI, the intensity of the IMT signals may be hypo- to hyperintense. Given the large overlap of radiological findings between IMT, rhabdomyosarcoma and leiomyosarcoma 15,16,17, anatomopathological differentiation is urgently needed in view of the findings described, with the aim of reducing the number of unnecessary radical surgeries. Within this context, the immunohistochemistry result showed a positive expression for the ALK-1 gene, considered a good marker in the differential diagnosis between IMT and malignant tumors of the urinary bladder, avoiding cystectomy in the patient in question.

To date, there is no standard protocol for the treatment of patients with IMT. The best treatment for this lesion is TURP and follow--up cystoscopy. 17 Previously, inflammatory myofibroblastic tumor was thought to be malignant; therefore, unnecessary radical surgeries, such as cystectomy, were performed. Currently, nonsurgical management is an option in selected patients and can serve as neoadjuvant treatment with the aim of preserving urinary bladder function, as occurred in our case. An ALK-1 inhibitor, crizotinib, has demonstrated favorable clinical responses in patients with metastatic or unresectable IMT. 17 The drug could become an alternative treatment for the disease.

CONCLUSION

This case report is an example of IMT of the urinary bladder. Classical clinical presentation of hematuria. Magnetic resonance imaging showed a polypoid lesion on its right posterior wall with involvement of the ureterovesical junction and consequent marked hydronephrosis upstream. Based on the immunohistochemistry results, there was also a positivity for ALK-1. A transurethral resection of the tumor was performed to completely remove the IMT.

As a future perspective, it is suggested that observational studies or case series be carried out in order to contribute to medical learning about this rare tumor formation.

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