Case Report

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Giant Leiomyosarcoma of the Urinary Bladder

Surgery Section

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ABSTRACT

The bladder leiomyosarcoma is a rare and agressive mesenchymal tumour, and adult women of reproductive age have a higher incidence of developing the bladder leiomyosarcoma. The pathophysiology of the disease is not certain, and its main symptoms are hematuria, dysuria and abdominal pain. There are not considerable amount of cases described in the literature. We report a case of a giant leiomyosarcoma of the urinary bladder in a 31-year-old woman.

Keywords: Cystectomy, Malignant mesenchymal tumour, Urinary Diversion

CASE REPORT

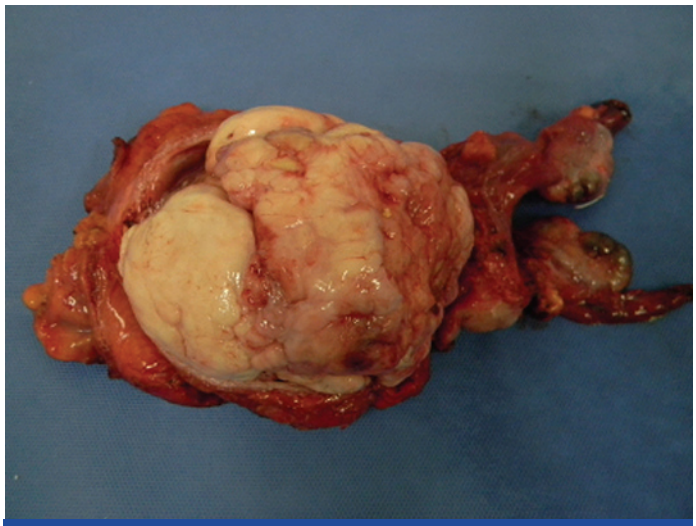
A 31-year-old woman presented progressive symptoms of vesical irritability associated with macroscopic hematuria, with a history of frequent urinary tract infections. Ultrasonography and CT-scan were performed and they both showed a mass of considerable size occupying a large portion of the urinary bladder [Table/Fig-1]. The patient did not show similar symptoms at any other site. Cystoscopy and biopsy were performed. Both tests revealed a low grade leiomyosarcoma. Pelvic exenteration with urinary diversion through the Bricker ileal conduit was performed. Macroscopically, it was found that the urinary bladder cavity was occupied by a voluminous pedunculated multinodular tumour of fasciculate mass measuring 10x9x7 cm [Table/Fig-2]. Complete resection along with free margins was done and the cut section was sent for histopathological examination, which confirmed the leiomyosarcoma, deeply infiltrated the smooth musculature of the urinary bladder. The uterus and nearby structures were free of neoplastic invasion. The surgical margins were negative. Microscopic examination showed a stage II leiomyosarcoma, compromising muscle bundles, the lamina propria and the mucosae. Vascular lesion was absent. Postoperative period was uneventful and no complementary treatment was given to the patient. During 12 months of follow-up the patient showed no signs of tumour recurrence.

DISCUSSION

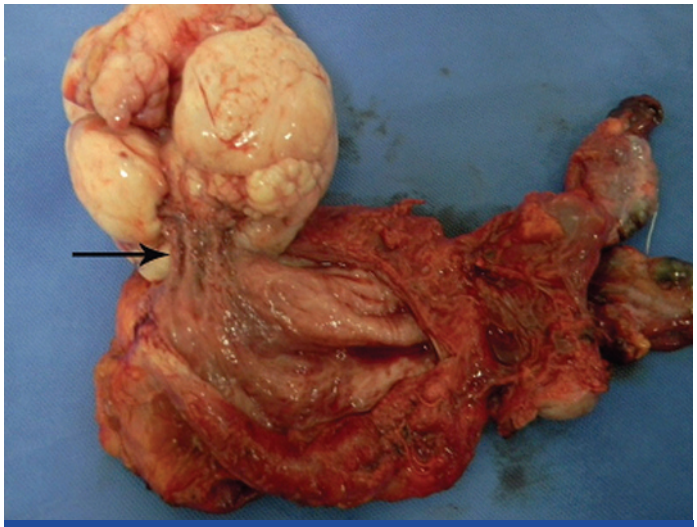
Mesenchymal tumours (benign or malignant) of the bladder accounts for 0.1% in adults, and the bladder leiomyosarcoma is the most prevalent. Less than 200 cases of this tumour were reported in the literature [1,2]. The distinctive macroscopic characteristics of the disease were described primarily by Bergman et al., [3]. The first case of leiomyosarcoma was described by Gussembauer in 1875 [4,5]. Most of malignant bladder tumours are from epithelial origin. Sarcomas are rare, and they represent less than 1% of malignant bladder tumours [6-8]. They can often be caused by radiotherapy, chemotherapy or hereditary causes [1,2]. Since, our patient presented history of frequent urinary infections, we believe that the chronic irritation and stimuli could have triggered cellular mutation, thus causing the tumour.

Since it is a relatively rare tumour, there is a lack of consensus regarding its pathophysiology, treatment and prognosis, although the leiomyosarcoma is generally resected through an aggressive surgery (by the oncologic point of view). The radical cystectomy with ample surgical margins seems to be the best treatment course and should be performed whenever possible, as there have been low rates of tumour recurrence [6].

The bladder leiomyosarcoma is an extremely rare tumour that can develop in any site of the bladder, and, as opposed to urothelial tumours, it hardly affects the ureters or the renal pelvis. In 1989,



[Table/Fig-1]: Superior view of the tumour, note that it occupies a large portion of the bladder's lumen.



[Table/Fig-2]: Lateral view of the tumour, it is fixated to the bladder wall through a shaft (black arrow).

15 cases of bladder leiomyosarcoma were reported, it was stated, then, that the macroscopic hematuria is the most common clinical manifestation, followed by symptoms produced by an abdominal mass, loss of urinary flow and suprapubic discomfort [9,10].

More than 60% of tumours have aggressive characteristics, and they usually lead to metastasis - even low stage tumours. The bladder leiomyosarcoma usually appear during the fifth decade of life, with a higher incidence in women of reproductive age, which can suggest the hypothesis that the hormones perform a role during the manifestation of the tumour [10,11].

When it is detected, most of the tumours are in an advanced stage. Furthermore, less than 15% of tumours are identified early (T1 stage). When discovered, the metastatic signs of the illness announce a morbid prognosis. In addition, the size of the tumour, the degree of tissue involvement and the free margins seems to be good parameters to evaluate the prognosis [8-11].

In an analysis of 35 patients with bladder leiomyosarcoma, it was shown that there is a specific survival rate of 62% in 5 years, and a recurrence rate of 34% in 38 months [12].

The treatment can be either a radical cystectomy with ample surgical margins, as it is the best choice in cases where surgical resection is possible, although the radical excision of the tumour associated with the removal of organs such as the prostate, seminal vesicles, uterus, cervix and vaginal cuff results in lower rates of positive surgical margins and local recurrence of the tumour, even though there could be a loss of bladder function and subsequent loss of quality of life [13].

In this particular case, we opted to perform a pelvic exenteration, since, the patient had a very large mass, with a diameter of 10cm appx. that completely occupied the urinary bladder [Table/Fig-1]. The tumour presented itself in a pedunculated form, attached to a unique insertion point at the wall of the bladder [Table/Fig-2]. Positive surgical margins and compromised lymph nodes were not found. We also decided to perform a radical cystectomy, because we only discovered that the tumour was pedunculated during the surgical act.

The patient was in her 12 month of postoperative period and didn't manifest any secondary complication.

CONCLUSION

Early detection and surgical intervention of the urinary bladder leiomyosarcoma can significantly improve the treatment efficacy. Partial cystectomy can be a reliable option to treat smaller leiomyosarcomas (below 4cm), since it can provide a similar therapeutic efficacy and a better quality of life to the patient, although the radical cystectomy still seems to be the best choice of treatment.

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