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Leiomyosarcoma of Bladder

Ousmane Nago Dembele, Kallat Adil, Ali Beddouch, Hachem Elsayegh, Ali Iken, Lounis Benslimane, and Yassine Nouini.

Department of Urology, Chu Ibn Sina, Rabat Morocco.

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ABSTRACT

We report a case of bladder leiomyosarcoma. This is a rare infiltrating bladder tumor, highly malignant, very poor prognosis due to a very early loco regional invasion. Only the pathological examination can bring positive diagnosis. The therapeutic modalities for this histological type are not codified. However treatment is mainly based on surgical excision Solid sometimes preceded by chemotherapy when the patient's condition .

Introduction

Mrs. BH, aged 65, housewife, non- smoking in his background having an appendectomy at age 49 years consulted for a total hematuria, with intermittent presence of blood clots lasting for seven months in a context of apyrexia and conservation condition. Clinical examination was normal.

Before this hematuria, realized a biological assessment showed anemia to 10.4 g/ dl in blood counts and sterile urine to cytobacteriological urinalysis. Bladder ultrasound objectifying a process of the anterior wall of bladder 58 * 36 mm

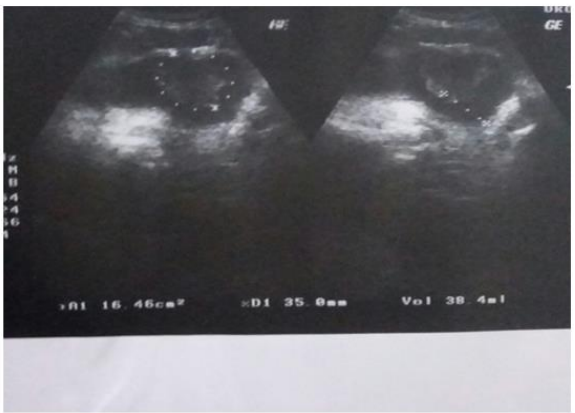

Figure 1. Ultrasound objectifying the intravesical process.

Endoscopic exploration of the bladder, on sterile urine highlighted a huge sessile tumor taking almost all of the front face of polypoid; the ureteral meatus were free. Transurethral resection of the tumor was incompletely performed, and pathologic study of resection chips was in favor of a high grade infiltrating and ulcerating mucosal leiomyosarcoma.

Immunohistochemical study was performed to confirm the diagnosis. (Fig. 2) thoraco- abdominal CT scan in the staging was performed and bladder objectified process; no secondary location.

An earlier pelvectomy was performed in the patient with cutaneous urethrostomy. The postoperative course was uneventful.

Microscopic examination of the specimen had found a high grade leiomyosarcoma infiltrating the bladder wall without injury to the tissue peri- bladder with negative margins, absence of lymph node.

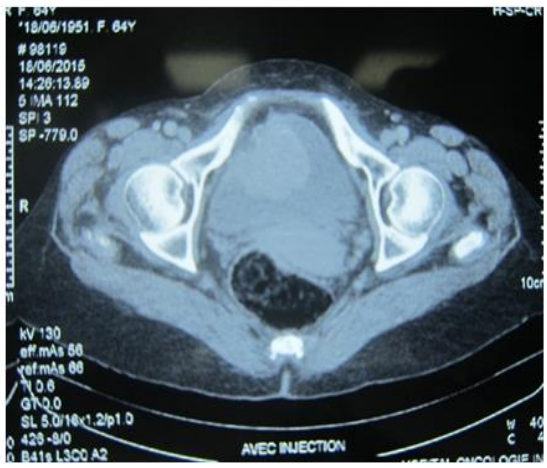

Figure 2. CT scan showing pelvic tumor.

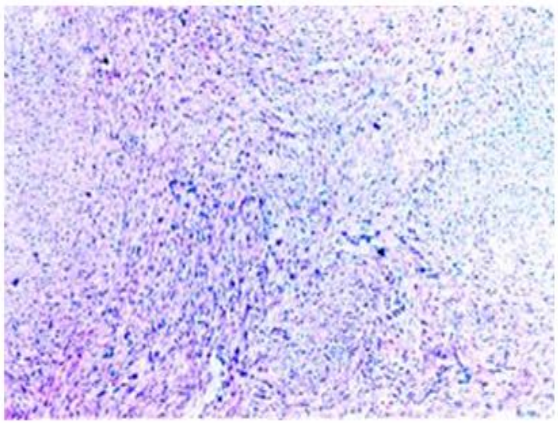

Figure 3. Proliferation to high cell density , made of spindle-shaped cells that form perpendicular beams (HES X 40).

Tele:
E-mail address: nagodembele@gmail.com

The clinical outcome was favorable.

Discussion

The bladder leiomyosarcoma (LMS) is a malignant tumor of connective guy with smooth muscle differentiation. It is rare and represents 20% of non-epithelial bladder tumors [9] is 0.38% to 0.67 % of all bladder tumors [3], 192 cases leiomyosarcoma have been described in the literature [8]. It occurs both in children than in adults, with a peak incidence beyond 60 years [3, 5, 7, 8]. It seems preferentially reach the male with a sex ratio of 3/1 [8, 9].

It can be associated with other lesions such as bladder transitional cell carcinoma or papillomatosis [9], or to certain general conditions like Van Recklinghausen disease [15]. The etiology is unknown, though several cases of leiomyosarcoma occurred after long-term courses of chemotherapy in this case by cyclophosphamide; this remains a scarce factor and the relationship cause and effect is not yet well elucidated [7, 8, 9, 11]. Recent studies [4, 17] described the high risk of developing leiomyosarcoma in patients with retinoblastoma; This is due to a genetic predisposition one hand, on the other hand to the potentiating effect of radiotherapy on oncogenesis by mutation of a second allele retinoblastoma gene.

Clinically, the LMS bladder often manifested by massive hematuria in 57% of cases that can be isolated or associated with signs of bladder irritation or a hypogastric mass [3, 14].

A cystoscopy [9], the tumor appears as a well-circumscribed mass, smooth or nodular multilobed widely implanted in the bladder wall, often sitting at the trine in 67 % of cases.

There are mainly two forms of scalable LMS bladder

-a form early invasive with extensive infiltration of tissues and organs peri bladder.

-a vegetating and exophytic form, initially somewhat invasive, often larger. This was the case of our patient.

The diagnosis of bladder LMS is based on the histological finding of a proliferation of spindle cells with nuclear atypia and abnormal mitosis and areas of necrosis. There are three types of spindle cells [9]:

-the cells in time, accumulated variously shaped beams available plexi form or palisades. The nucleus of these cells is oval or rod-shaped and plunged into a cytoplasm of myofibrillée structure.

-the spindle cells shorter than the previous, with oval nuclei more or less globular .

-The atypical giant cells with nuclear monstrosities high mitotic index

In undifferentiated forms of diagnosis difficult [12, 14] will be sought by immunohistochemistry techniques a cell reactivity for vimentin, desmin and smooth muscle actin and negativity of epithelial markers; otherwise we can highlight electron microscopy of micro filaments, dense plates and pinocytosis vesicles reflecting the smooth muscle differentiation.

The differential diagnosis [3, 12,14] will land with a number of injuries , including the pseudo inflammatory sarcoma that may be infiltrating and rich in mitosis , but without necrosis area, the fibro- myxoid sarcoma tumors nickname because of their cellular pleomorphism and character infiltrating the slightest leiomyoma cell density without cellular atypia or necrosis area; as the bladder leiomyosarcoma can lend confusion with myxoid leiomyosarcoma, a very rare variety of smooth muscle sarcoma , only one case was described by Young [18] it consists of relatively well-differentiated muscle cells, little mitotic immersed in a stroma myxoid.

Given the scarcity of LMS bladder, the therapeutic conduct is not codified.

However different therapeutic modalities are used such as external radiotherapy; cryotherapy; phototherapy; [7, 8]. Van Thillo and Narayana, are state of healing after a transurethral resection of the tumor with a survival rate of 2 to 4 years [13, 16].

Indeed, treatment is surgical, it relies on a partial cystectomy, when the tumor is less than 3 cm, and not invasive; beyond cystectomy is required [8, 16]. Albaster [2] recommends additional urethrectomy to avoid urethral recurrence.

Some authors [3] propose adjuvant chemotherapy based on doxorubicin and cis platinum mainly in the invasive tumors and in cases of lymph node involvement, others [6, 14] the reserve for metastatic forms. Radiation therapy seems less effective [3, 9]. For the poor prognosis of tumors (invasive tumor size greater than or equal to 5 cm, affected lymph node, of high grade according to the criteria of (Memorial Sloan Kettering Cancer Center MSKCC) Charles indicates a pre chemotherapy or postoperative. once the tumor is considered extirpable; . radical cystectomy can be performed [6] After surgical excision, the recurrence rate is 42 % for low grade tumors [10] and 34% for high- tumors grade [6].

In our patient, the LMS bladder is diagnosed at an early stage, taking into account the nature of the malignant tumor and its invasive character; an earlier pelvectomy with urethrectomy is performed immediately, pathologic examination of the surgical specimen , iliac nodes and urethral slice showed no invasion. Urethrostomy skin was performed as derivation type.

Conclusion

Leiomyosarcoma is a rare tumor of the bladder, prognosis. The diagnosis is based on pathological study.

Only the passage of time and the many series would see more clearly in order to establish an adequate therapeutic strategy to improve the prognosis. Currently, treatment is recommended that seems to neoadjuvant chemotherapy and radical cystectomy association.

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