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SARCOMA OF THE PROSTATE: A REPORT OF TWO CASES AND A REVIEW OF THE LITERATURE

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ABSTRACT

The sarcomas of the prostate are very rare pathologies and to the poor prognosis. Their association is possible with a prostatic adenocarcinoma, and the diagnosis is then revealed only during the anatomopathological examination of the operative specimen with the immunohistochemical analysis which makes it possible to characterize them. Because of their rapidly invasive potential, it is important to be able to detect them early, especially in young people. We report in this article two cases of prostatic sarcomas treated and we review the literature concerning this pathology.

KEYWORDS: Sarcoma, Prostate, Immunohistochemistry, surgery, chemotherapy, prognosis.

INTRODUCTION

Prostate cancer is common in men after the age of 50, the vast majority of whom are prostatic adenocarcinoma. Its frequency reaches 67% of the male population during the eighth decade, with an incidence of 40/100000 in France. Prostate sarcomas are rare tumors (0.1 to 0.2% of malignant tumors of this gland), which can affect many cellular contingents and all ages of the population, including young subjects.[1] Among these sarcomas are tumors derived from muscle cells, namely rhabdomyosarcomas more often found in children and leiomyosarcomas affecting rather the adult, Associations of different neoplastic contingents are possible.

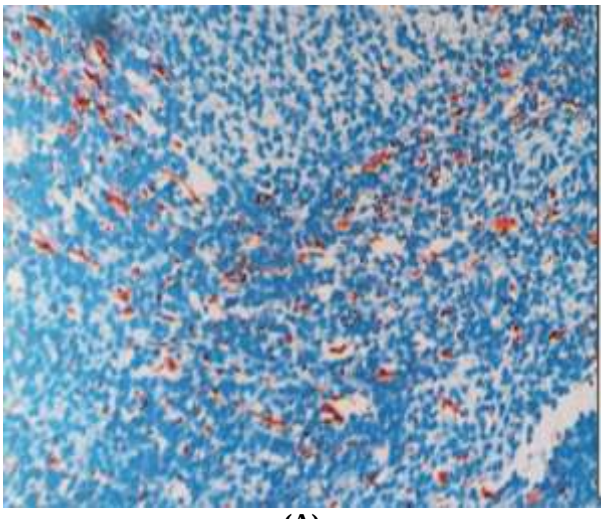
The origin of these cancers remains uncertain, pelvic irradiation seeming to play a role in the genesis of some,[2] genetic factors have also been implicated, and a link with myelodysplastic syndromes is evoked for some granulocystic sarcomas.

2. THE CASE REPORTS

2.1 Case 1

A 38 years old, with no significant pathological antecedents; who had consulted for urinary disorders like dysuria and pollakiuria for 06 months. Digital rectal examination had objectified a hypertrophied, irregular and indurated prostate, the rest of the physical examination was unremarkable. Blood analysis showed PSA value: 4.54 ng/m, all other blood routine indexes were within the normal limits, A trans-urethral resection of the prostate was performed, histopathological examination revealed a primary embryonic rhabdomyosarcoma of the prostate (figures 1), Abdomino-pelvic Magnetic resonance imaging (MRI) revealed a heterogeneous mass of the prostate with invasion of the seminal vesicles (Figures 2).

The patient had benefited from radical cystoprostatectomy; the fact that the resection was incomplete because of the pelvic shielding, he started three weeks after the surgery chemotherapy by the administration of three courses of vincristine, doxorubicin and cyclophosphamide, The evolution was marked by the death of the patient three months after the initial diagnosis.



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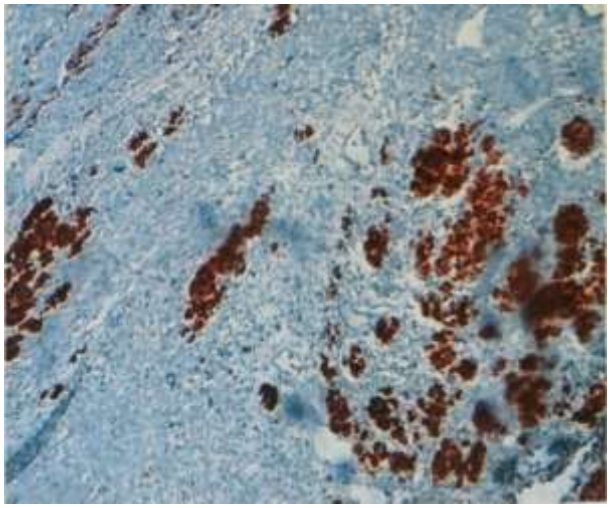

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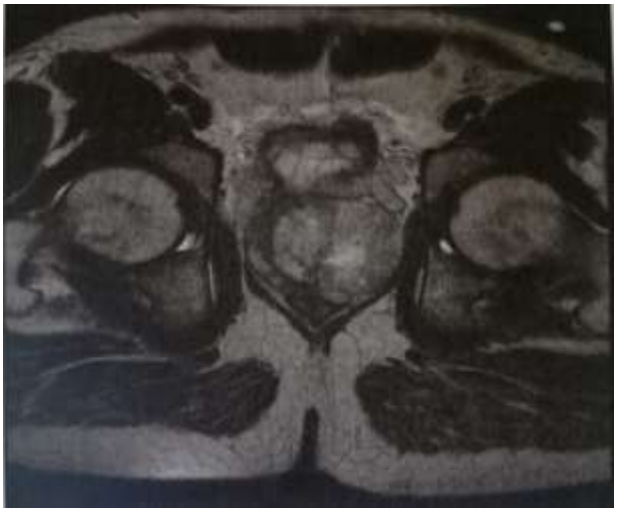
Figures 1: Embryonic rhabdomyosarcoma prostate; Immunohistochemistry showed: positive desmin (A) and positive myogenin (B).



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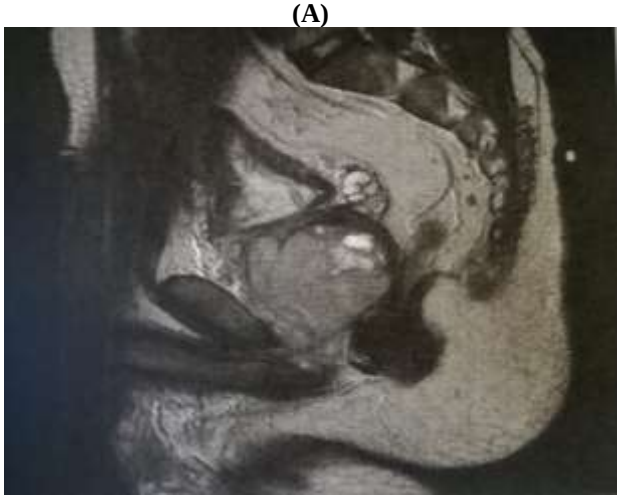


(B)

Figures 2: Abdominopelvic MRI; Cross section (A) showing: the presence of a heterogeneous mass of the prostate with invasion of the seminal vesicles. Sagittal section (B): showing expansive prostatic growth with invasion of the seminal vesicles.

2.2 Case 2

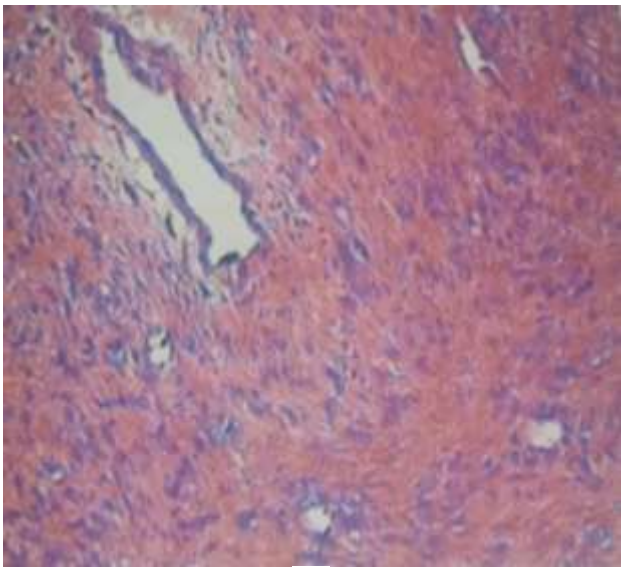
A 49 ans years old**,** with no significant pathological antecedents, who had consulted for urinary disorders like a dysuria for 6 months, with a weight loss not quantified, The physical examination had found a patient with an altered general condition; Digital rectal examination had objectified: firm prostate; nodular and forming part of the bladder base, the rest of The physical examination was unremarkable. Blood analysis showed obstructive renal failure that improved after bilateral nephrostomy, the value of PSA was normal.

A transperineal prostatic biopsy was performed for our patient, whose anatomopathological examination (Figures 3) concluded that prostatic leiomyosarcoma.

Abdominopelvic CT scan was performed: the prostate had irregular contours, and raised heterogeneously, it came into contact with the rectum and the bladder forward without greasy separation between them, with infiltration of the neighborhood grease, Uretero-hydronephrosis was moderate with nephrostomy tubes in place, Pelvic MRI: (Figure 4) showed an enlarged prostate, with a heterogeneous hyposignal at T1 and T2, with moderate enhancement. It extended to the pelvic floor. It appeared separated from the bladder by a greasy border. On the other hand, there was no edging separating it from the rectum.

Given the alteration of the general condition of our patient and the importance of local and distant evolution of the prostatic tumor, a trans-ureteral resection was performed, associated with adjuvant chemotherapy.

The postoperative follow-up was marked by the death of the patient after his second course of chemotherapy (8 months after the beginning of the symptomatology).



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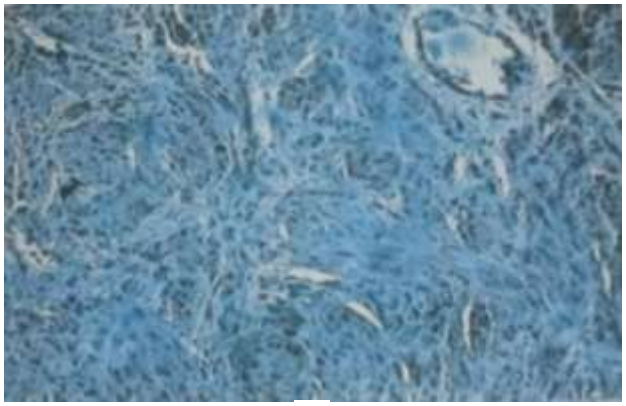
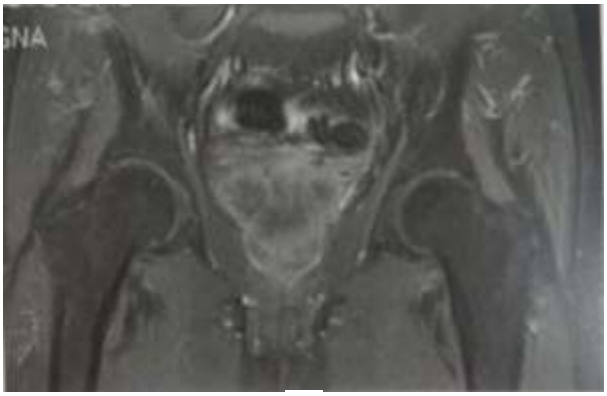


(B)

Figures 3: Prostatic leiomyosarcoma; histological section (× 20). (A): histological section with hematin-eosin staining showing normal prostatic glands surrounded by a heterogeneous tumor-like cell population.

(B): Immunohistochemistry shows a tumor proliferation expressing at the cytoplasmic level the anti-smooth muscle antibody and partially the anti-desmin antibody.



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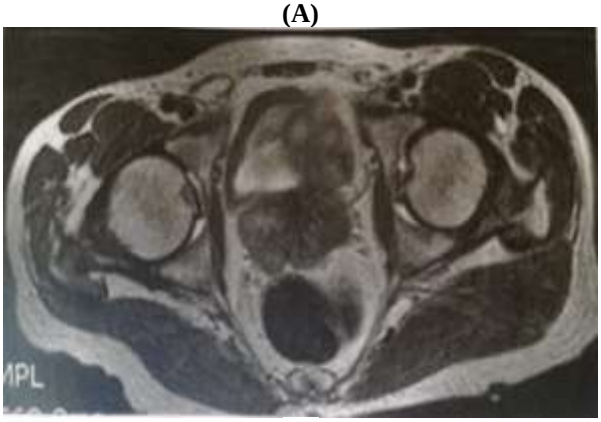

(B)

Figures 4: Pelvic MRI: (A): in T1-weighted axial section after Gadolinium injection: the tumor takes contrast heterogeneously and extends to the pelvic floor, without fatty edging separating it from the rectum, causing suspicion of an extension towards this organ. (B) T2-weighted cross section: Heterogeneous prostate, bumpy contours, contracting intimate relations with bladder and rectum.

3. DISCUSSION

These two observations lead us to consider two pathologies occurring in very different contexts, grouped under the term of prostate sarcomas and which represent 0.1 to 0.2% of all prostatic neoplasias. The field, the symptomatology, the treatment and the prognosis are in each specific case of the cellular contingent concerned.

Primary embryonic rhabdomyosarcoma of the prostate (ERMP)**:** is a rare pathological and clinical entity in adults, These are aggressive tumors characterized by locoregional extension and a tendency to metastasize by regional blood and lymphatic circulation. In adult patients, available data on the management of **(ERMP)** are limited and most are from pediatric clinical trials of rhabdomyosarcoma. It is a common tumor in infants and children, with a median age of onset of 5 years, but is rare in adults.[3,4] They have a tendency to reproduce in tissues such as the middle ear, bile ducts, bladder and prostate, which lack skeletal muscle.

They derive from undifferentiated mesenchymal cells, which are found in the distal urogenital tract surround the Wolff duct; then they are incorporated into the bladder and prostate during embryogenesis.[5] These cells, which persist in later life, have the capacity for rhabdomyoblastic differentiation, and may explain the occurrence of these tumors, although extremely rare, in adults.It is characterized by a high degree of malignancy due to rapid local growth with the formation of large pelvic masses that often leads to renal failure due to urethral obstruction, and systemic spread, usually in the lungs, liver and regional lymph node bones.[6]

The initial symptoms of ERMP published in the literature are often related to obstruction of the urethra. biological analyzes the value of PSA is normal.

For our patient, the reason for consultation was urinary disorders like dysuria and pollakiuria and rectal examination found an enlarged, irregular and indurated prostate with a normal PSA level.

Radiological evaluation of the genitourinary organs and potential intra-abdominal metastatic sites can be performed with computed tomography (CT) of the abdomen and pelvis.[7,8] Recently, the best contrast resolution of magnetic resonance imaging (MRI) has offered new possibilities for staging primary tumors.[7] However, differential radiological diagnosis with adenocarcinoma of the prostate can be very difficult.

Our patient: an Abdomino-pelvic MRI (Figures 2) revealed in our patient: a heterogeneous mass of the prostate with invasion of the seminal vesicles.

The diagnosis is made by transrectal biopsy or transurethral resection.[9] Histologically, RMS characterized by a rather monomorphic tumor proliferation of undifferentiated cells. The appearance

varies from the totally undifferentiated primitive or rhabdomyoblast cell to the well-differentiated cell with cytoplasmic striations, Fusiform or myxoidal aspects can be observed.

Immunohistochemical (Figures: 1) studies show that cells are not labeled by cytokeratin (KL1, AE1 / AE3), smooth muscle actin, global muscle actin (HHF35), EMA epithelial membrane antigen, or lymphoid markers. (ALC, CD 220 and CD 3), or neuroendocrine (Chromogranin A, Synaptophysin, Leu 7, NSE). They express vimentin, and especially desmin and myoglobin confirming the rhabdomyoblastic nature of tumor proliferation.

The embryonic rhabdomyosarcomas have a high cytological variability, which represents several stages of the morphogenesis of the muscular skeleton. Desmin and specific actin muscle (muscle specific actin) are the typical spots used to identify rhabdomyosarcoma.

For our patient the results of trans-urethral resection had also shown fusiform cells containing round rhabdomyoblasts with eosinophilic cytoplasm and multiple hyper chromatic nuclei; and on Immunohistochemical examination had shown: positivity desmin (A) and myogenin (B) (Figures 1).

A multimodal therapy is essential for curing patients with RMS, but different uses of surgery, radiation and chemotherapy, and their intensity, need to be selected and modulated to different risk groups of patients.

The prognosis of rhabdomyosarcoma (RMS) in adults remains poor. Recently, Ferrari et al.[10] reported in a large retrospective analysis of 141 adults with rhabdomyosarcoma, 36 RMS subjected to multimodal treatment according to RMS childhood guidelines (RMS guidelines). Other previous series of adult prostate rhabdomyosarcoma reported poor results.[11,12] In this report: 5 years of event-free survival and 5 years of overall survival were 32.9 months and 45.7 months respectively, suggesting the worst outcome of adults with RMS as children; however, subgroup analysis showed that genitourinary RMS; were not associated with reported results worse. Our patient died three months after the initial diagnosis.

The therapeutic options used are generally derived from pediatric clinical trials on RMS conducted by international cooperation groups, but no definitive conclusion has been drawn so far regarding the best treatment option available for ERMP in adults.[13]

Treatment depends largely on the stage of the disease at the time of onset: Localized RMS of the prostate require prostatectomy with an attempt to salvage the bladder with or without urethral reconstruction.[14] Unfortunately, the functional morbidity of initial genitourinary tract tumor surgery has prompted patients to avoid advance surgery in favor of multimodal approaches,[13] RMS generally respond well enough to induction chemotherapy to make them locally resectable.[15,16] Our patient had benefited from a radical cystoprostatectomy; the fact that resection was incomplete because of pelvic shielding, the patient started three weeks after surgery adjuvant chemotherapy.

More data and further studies are needed in the future to improve the understanding of the biological behavior of ERMP in adults and to define the most appropriate therapeutic approach, as well as the local treatment protocol in a locally advanced disease.

Prostatic leiomyosarcoma is a very rare tumor in adults accounting for 38% to 52% of prostate sarcomas,[17] which constitute 0.1% of the malignant tumors of this gland.[17,18]

The first review of prostatic leiomyosarcoma was published by Fitzpatrick and Stump in 1960.[19] it occurs in adults, with an average age of 58 years,[17,20] his diagnosis is usually made at a late stage, because of the clinical and radiological nonspecificity, his prognosis remains dark. Imaging, represented by CT and especially MRI plays a vital role in extension evaluation and follow-up, Our patient was a 74 year old man.

physical examination: It presents itself as a prostatic mass, with a rectal touching a regular prostate, lobulated, of firm and elastic consistency.[18] An obstructive syndrome of the lower urinary tract is the most common reason for consultation.[20,21] Prostate marker (PSA, PAD) assay is typically normal.[17]

The symptomatology of our patient was a dysuria; with rectal examination: the prostate was enlarged, firm; nodular and forming part of the bladder base, and the total PSA value was normal.

The diagnosis is confirmed by a histological study performed following an echo-guided transrectal biopsy in most cases and less often by a transperineal, CT-guided biopsy, or a suprapubic prostatectomy.[22]

The anatomopathological diagnosis, completed by an immunohistochemical study,[21,23] is essential for the classification of these tumors.[24] The majority of leiomyosarcomas are hypercellular, high grade, composed of fusiform eosinophilic cells with increased mitotic activity and severe to moderate nuclear atypia. Low grade Leiomyosarcoma, with moderate atypia, and scattered mitosis is very rare. Neoplastic cells generally express vimentin, smooth muscle actin and desmin, whereas cytokeratin expression is observed in only 25% of cases.[20]

The diagnosis of certainty for our patient was histological: a transperineal prostatic biopsy whose pathological examination (Figures: 3) had concluded to a

prostatic leiomyosarcoma; in immunohistochemistry: positivity of anti-smooth muscle antibody and partially anti-desmin antibody.

CT and MRI (Figures: 4) are used to evaluate the local and general extension of the tumor. The radiological appearance of leiomyosarcoma is not specific.[18] It is usually a large, heterogeneous tumor that can elevate or invade the bladder floor. Endo-rectal ultrasound is superior in the evaluation of local extension, which allow precise delineation of the tumor in relation to healthy tissue, an important element for surgically resectable tumors.[17]

The frequency of metastases at the time of diagnosis varies from 20 to 40%, while lymphadenopathies are found in only 9.8% of cases. MRI and CT The disappearance of the fatty border with the bladder and the rectum is not a formal sign of extension.[18] Our patient had abdominopelvic CT and pelvic MRI (Figures: 4); who had objectified an enlarged prostate of irregular contours, and raised heterogeneously with infiltration of the pelvic floor; with nodular liver lesions.

The treatment of this tumor is not yet codified, it includes surgery, pre- or post-operative radiotherapy and neo-adjuvant or adjuvant chemotherapy. It depends on age, general condition, tumor volume, grade of malignancy and extension status.[18] Curative surgery includes radical retro-pubic prostatectomy, radical cysto-prostatectomy, suprapubic prostatectomy, and pelvic exenteration.[22] The chemotherapy protocols used are diverse, but most patients receive anthracyclines, alkylating agents and/or alkaloids.[25]

In front of the deterioration of the general state and the importance of the local and distant evolution of the prostatic tumor, our patient had benefited from a trans-ureteral resection, associated with an adjuvant chemotherapy.

The prognosis of prostatic leiomyosarcomas is often unfavorable.[26,27] Survival is highly variable between series, averaging less than 10% at 5 years.[17] The post-operative consequences of our patient; were marked by death after his second course of chemotherapy (8 months after the onset of symptoms).

The therapeutic management of leiomyosarcoma is not codified at present and their prognosis remains very dark and can only be improved with a multi-disciplinary approach and early diagnosis, allowing complete radical surgery to be performed alone. effective therapeutic. Advances in immunohistochemistry have made it possible to confirm the diagnosis of these different tumors and consequently to adapt the therapeutic strategy and to establish adequate monitoring methods.

4. CONCLUSION

Sarcomatous tumors of the prostate are very rare. It is necessary to mistreat everything you need to know before contracting a rectal symptomatology in an elderly subject or with rapid obstructive urinary signs in a young subject. The anatomopathological diagnosis completed by an immunohistochemical study is then indispensable for the classification of these tumors. Their therapeutic management is not codified at the moment and their prognosis remains very dark.

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