PROSTATE SARCOMA: REPORT OF 2 CASES AND BIBLIOGRAPHIC REVIEW

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Summary.- OBJECTIVE: To report two cases of prostate sarcoma and perform a review of the published literature.

METHODS / RESULTS: The first case is a 21 year old patient who presented acute urine retention and lung metastases on diagnosis. He was diagnosed by TURP of rhabdomyosarcoma of the prostate dying 1 month after surgery. The second case was a 33 years old male who presented to the emergency room with anal pain, urinary symptoms, hematochezia and loss of 20 kg over the past 3 months. Abdominal CT scan showed an 11 x 10 x 9 cm mass in the lower pelvis that infiltrated the bladder and rectum, being unable to define its origin. CA 19.9, CEA and PSA were normal. The suspected diagnosis was a prostate sarcoma infiltrating rectum and bladder. A pelvic exenteration was performed with a wet colostomy. The pathologic diagnosis was a high grade sarcoma not clearly identified of the prostate. He was treated with Adriamycin as adjuvant chemotherapy, having local recurrence, nodal involvement and multiple pulmonary metastases after 3 months of follow up.

CONCLUSIONS: Prostate sarcomas are rare tumors. This makes difficult to know their natural history. Their rapid progression and systemic spread, despite multimodal treatment, gives a mean survival of 24 months. Main survival factors are grade, a complete resection of the tumor and a low local stage. There is a need to find new chemotherapy protocols to increase survival rates as it has been shown in extremities sarcomas.

Keywords: Prostate sarcoma. Spindle cell sarcoma. Rhabdomyosarcoma. High grade sarcoma.

Resumen.- OBJETIVO: Presentar dos casos de sarcoma de próstata y realizar una revisión de la literatura publicada.

METODO/RESULTADO:El primer caso es un paciente de 21 años de edad que acudió por retención aguda de orina y metástasis pulmonares al momento del diagnóstico. Fue diagnosticado por RTU de rhabdomyosarcoma de próstata falleciendo un mes después de la cirugía. El otro paciente era un varón de 33 años que acude a urgencias por proctalgia, síntomas miccionales, hematoquecía y pérdida de 20 kg durante los 3 últimos meses. El TAC abdominal muestra una gran masa de 11x10x9 cm en pelvis inferior que desplaza anteriormente la vejiga e infiltra el recto sin poder concretar su origen. Los marcadores CA 19.9, CEA y PSA/s no presentaban alteraciones. La RMN orientó hacia un sarcoma de próstata que infiltraba vejiga y recto. Se realizó una exenteración pélvica con colostomía húmeda. El diagnóstico definitivo fue de sarcoma fusocelular de próstata de alto grado con infiltración de recto y vejiga. Se administró tratamiento quimioterápico adyuvante con Adriamicina, presentando a los 3 meses recidiva local y múltiples metástasis pulmonares.

CONCLUSIONES: Los sarcomas de próstata son tumores poco frecuentes, lo cual dificulta el conocimiento de su historia natural. La progresión local y a distancia es muy rápida, con una supervivencia media de 2 años, a pesar del manejo terapéutico multimodal. El principal factor de supervivencia es una resección completa del tumor y un estadio local poco avanzado. Es necesario buscar nuevos
Global prognosis of sarcomas is bad, independently from the modality of treatment. The histological subtype of prostate sarcoma appears to have prognostic significance, being the most frequent types the rhabdomyosarcoma, leiomyosarcoma, carcinosarcoma and the undifferentiated sarcoma (1).

Here we present a case of a rhabdomyosarcoma and a spindle cell sarcoma of the prostate and a review of the literature published, marking the importance of a good diagnose and a correct management.

**INTRODUCTION**

Sarcomas are rare malignant tumors raised from mesenchymal tissue derived from the ectoderm. They are less than 1% of all cancers diagnosed annually in the United States. Histopathologically, mesenchymal cells can mature into striated and smooth muscle, cartilage, bone, adipose and fibrous tissue, giving a varied spectrum.

Mesenchymal tumors represent only the 0,2% of the malignant prostate tumors what makes them an unknown disease with only small groups of cases published in the literature. The rarity of sarcomas, and the wide number of histological subtypes and locations, is the main reason why the natural history and the different patterns of response to the treatments are still unknown.

A multidisciplinary approach optimizes the treatment of soft tissues sarcomas. If it is possible, it is recommended to send the patient to an experts center. Usually there can be found specialist centers in bone and soft tissue sarcomas, but due to the broad field of presentation, some as the prostate sarcoma don’t have a standard treatment established.

**CASE REPORT**

Our first from case from 1976 is a 21 year old patient with no medical history who came to the emergency room for acute urine retention. In the rectal examination there was found an indurated irregular prostate. The remaining physical examination was totally normal. The blood test showed renal failure with a creatinine of 3.5 mg / dl. We performed a chest x-ray finding multiple lung and rib metastases. In these days US study and CT scan were not an available imaging test in our hospital. The patient was admitted in our department undergoing urography and TURP.

Final pathology was prostate rhabdomyosarcoma. The microscopic description of the specimen described a normal prostatic epithelium infiltrated by eosinophilic tumor cells with prominent nucleoli and high mitosis, which according to immunohistochemical techniques were consistent with striated muscle cells.

The patient was discharged the 7th after surgery, deciding palliative treatment because of the advanced
The patient died at home one month after being discharged.

The second is a much more recent case. A 33 year old male, with no drug allergies, or relevant medical history, who goes to the emergency room twice due to intense proctalgia since 3 months, with worsening progression and accompanied by tenesmus, hematochezia, dysuria, and loss of 20 kg.

The digital rectal examination was very painful, with a hardened rectum and a very big prostate.

CT abdominal scan described a large 11x10x9 cm mass with liquefactive encapsulated necrosis, located in the lower pelvis that displaced the bladder and rectum. No exact origin of the tumour could be defined. There were suspicious inguinal lymph nodes with no other systemic involvement.

MRI was requested suggesting prostate sarcoma (Figures 1 and 2). The blood tests and markers CA19.9, CEA and PSA were normal.

The patient developed a bowel obstruction, deciding to reject the biopsy and opted for radical surgery scheduled. During the surgery there was found a large tumor occupying the entire pelvis, displacing and infiltrating the rectum and the bladder. It was necessary an enbloc resection of the tumor which included rectum, prostate, seminal vesicles and bladder, leaving a terminal colostomy in left lower abdominal quadrant with derivation of both ureters to the pouch.

The pathologist description was a large mass that occupied the central region of the lower pelvis, poorly demarcated and non encapsulated, measuring 11x10x9 cm (Figure 3). The surgical specimen included prostate, bladder and anterior rectum, showing large areas of necrosis and loose fragments. Macroscopically there was a clear infiltration of the rectal wall and prostate. Under the microscope we see a highly cellular tumour population, very atypical, with predominant spindle cell morphology and counting at least 18 mitosis in some of the optical fields. The neoplasm infiltrated nerves and vascular structures (Figures 4, 5, 6).

Immunohistochemical study showed staining of tumour cells for Cd99, Cd31 and focal Cd43, Factor XIII. Hormonal markers (oestrogen and progesterone) were negative, as well as the S-100 and Cd117 and cytokeratins AE1-AE3. The conclusions obtained with the IHC studies were that it couldn’t be a leiomyosarcoma because actin was focally positive, defining it as a high grade (because of the mitosis), spindle cell (cells aspect) sarcoma (mesenchymal).

The final diagnosis was a high-grade spindle cell sarcoma of prostate not clearly identified.

The patient was discharged from General Surgery Department the 16th postoperative day, with oral tolerance and functional colostomy.

Subsequently, the patient received chemotherapy with Adriamycin until March 2011, presenting in this period of time recurrent urinary infections that required admission in two occasions.

Three months after the surgery a control CT scan showed a 4 cm mass in the prostatic field compatible with local recurrence, a 10 cm mass in the right iliac fossa, bilateral inguinal lymph nodes, and metastases in both lungs. Supportive care treatment was decided. Patient passed away after 4 months of the initial diagnosis.

Figure 3. Specimen.  Figure 4. Necrosis.
DISCUSSION

Soft tissue sarcoma can arise from any site as they come from the mesenchymal tissue derived from the ectoderm.

The most frequent sarcomas of the retroperitoneum are the liposarcomas (composed mainly with fat) and leiomyosarcomas (composed mainly with smooth muscle). From the genitourinary tract, the leiomyosarcoma is the most common type and may arise in the bladder, kidney or prostate. Rhabdomiosarcomas are more frequent in children and young patients (2) and apart from the retroperitoneum, they have also been described as paratesticular masses. There are also published a few cases of liposarcomas of the spermatic cord.

Sarcomas arising from the prostate are extremely rare, up to the date there are almost 100 cases published, with different types of histological patterns. They are only the 0,2% of all prostate tumors. Usually prostate sarcomas are published as an isolated case or a small series of cases. Janet et al. described in their series of 10 patients with prostate sarcoma that the most frequent type was the rhabdomyosarcoma (with 5 cases), followed by the carcinosarcoma (with 2 cases), high grade sarcomas not clearly identified (2 cases) and leiomyosarcoma (with 1 case) (1). Sexton et al. and Cheville et al. published a series of 21 and 23 cases each with similar percentage of the different subtypes (3, 4).

The range of age is wide (between 30 and 90 years) with a peak of incidence between 60 and 70. Rhabdomyosarcoma seems to appear during childhood (2) meanwhile leiomyosarcoma affects young adults (less than 50 years old). Prostate sarcomas generally, have a bimodal distribution with 15% of the cases before the first decade and a 30% after the seventh (5).

The ethiopathological mechanism is still unknown, although there are some papers describing higher incidence of prostate sarcoma after treatment with radiotherapy for prostate adenocarcinoma (5).

The most frequent symptom of prostate sarcoma are LUTS caused by the obstruction created by the occupying space mass. Less frequently it can cause rectal pain, abdominal or rectal palpable mass with constipation, rectal bleeding or tenesmus.

If we refer to sarcomas, biopsy is a critical step. An open biopsy if needed must be planned carefully to avoid complications, as it is shown in two series where they conclude that 20% of patients had to receive more surgeries, irradiation or chemotherapy because of a bad performance of the biopsy (6). Core needle biopsy, which is the method most used for diagnose of prostate sarcoma, is highly accurate in the diagnosis of malignant soft tissue sarcomas, with minimal morbidity and low cost. Transurethral resection of the prostate can be the way of diagnose in some cases that had LUTS, but the transrectal ultrasound guided core biopsy remains the main tool of diagnose. Usually the patients don’t present elevation of the plasmatic levels of PSA, only if they had any component of epithelial tissue like in the carcinosarcomas, what usually delays the indication of a prostate biopsy.

Grading in low, moderate and high grade, should be used for untreated primary sarcomas and along with the histological pattern will prove its benign or malignant behavior.

Immunohistochemistry studies are always needed in any sarcomatous tumor to identify the origin, been the vimentin, the keratin, desmin, leukocyte common antigen and S-100 the most used antigens. Leiomyosarcoma expresses...
actin and desmin. Carcinosarcoma is a biphasic tumor with malignant epithelial and mesenchymal component. In spite of the use of immunohistochemistry techniques there are some sarcomas that remain unclassifiable. High grade sarcoma doesn't present any benign epithelial component (6).

Tumor size and grade are the main risk factors for local, distant recurrence and survival (7). Location of primary tumor, age over 50 years, size over 10 cm and positive margins are the main predictors for local recurrence. The most common local progression is the infiltration of the bladder, rectum and pubis. In these past years some nomograms have been developed to predict the postoperative survival of retroperitoneal sarcomas (8).

CT scan is the most used imaging study for initial staging if there is any suspect or diagnose of sarcoma. A chest scan is recommended for all newly diagnosed patients to discard lung metastases, despite the low risk for patients with tumors less than 5 cm in diameter. Magnetic resonance is the method of choice for the extremities sarcomas and for local staging but not for abdominal and retroperitoneal cases, where it should be perform in case of difficulties in the CT scan to know its origin.

Distant metastases are present in the 7% of the newly diagnosed patients and 39% will develop them during the disease (3). The most frequent places of metastases in retroperitoneal and prostate sarcomas are the lung, bone and liver. With this data it is recommended an aggressive local and systemic treatment.

Regional nodes are only frequently affected in rhabdomyosarcomas, being the grade of differentiation an important predictor. We can say that nodal involvement is uncommon with primary tumors less than 5 cm, and do not carry the same poor prognosis as distal metastases.

Surgical resection is the most effective potentially curative therapy for soft tissue sarcomas regardless the site of origin. Depending on the places, the complete resection will or will not be possible. In the case of prostate sarcoma usually the complete resection is impossible because of the locally advance stage. It's recommended in many publications to have a sequential treatment with neoadjuvant chemotherapy followed by radical surgery and adjuvant radiotherapy (3).

Some patients, especially with sarcomas <5 cm of length, can be cured with high dose radiation therapy with or without chemotherapy (9). Neoadjuvant radiotherapy is not recommended, and neoadjuvant chemotherapy with doxorubicin and ifosfamide has been suggested as a beneficial in large tumors (>8 cm). Adjuvant radiation therapy is advised in patients with high grade resected sarcomas, what may improve local control but no impact on survival has been shown (10). Adjuvant chemo-

therapy, except for childhood sarcomas (2), has no clear benefit. Brachytherapy has utility in children sarcomas in order to avoid mutilant resections (11).

Patients with prostate sarcoma need long term follow up because there have been described cases with recurrence even 20 years after the primary tumor has appeared (12). When a recurrence appears and if it is possible, salvage surgery can improve survival (13).

Janet et al. described in a single institutional review of 10 cases with prostate sarcoma, that histology type seems to be an important prognosis factor. The overall survival was poor [24 months] except in the rhabdomiosarcoma [140 months] (1). In their study, with the same results as Sexton et al., no difference was found in the survival between different grades or size, but the presence of metastases was a poor outcome predictor.

**CONCLUSIONS**

Prostate sarcomas in adults are very infrequent tumors what make their natural history unknown. The prognosis is poor even with a multimodal treatment. Usually at the moment of diagnosis they are locally advance and are spread systemically. The different histological types and the possibility of total resection of the tumor are the main prognosis factors. Local recurrences are frequent; if a recurrence appears is recommended to try to do a radical surgery. There is a need of more randomized studies that could suggest better protocols of neoadjuvant and adjuvant therapies to improve the survival of these patients.

**REFERENCES AND RECOMMENDED READINGS**

(*of special interest, **of outstanding interest)

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Case Reports

PRIMARY AND METASTATIC RENAL HEMANGIOPERICYTOMA

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Summary.- OBJECTIVE: Haemangiopericytoma is an uncommon perivascular tumor that occurs more frequently in soft tissues and is extremely rare in the kidney.

METHODS: We report two cases: The first one is the case of a 57-year-old man with bilateral metastatic renal haemangiopericytoma which appeared 18 years after removal of a meningeal haemangiopericytoma. The second is a 29-year-old woman with a primary kidney

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