CLINICAL CASE

Primary leiomyosarcoma of the penis

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KEYWORDS
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Abstract Primary leiomyosarcoma of the penis is an extremely rare tumor. There are only 46 cases reported in the international medical literature. We present herein the case of a 36-year-old man with a tumor at the base of the penis. The histopathologic study showed fusiform cells consistent with “primary leiomyosarcoma of the penis”. We are adding a case to the medical literature and also providing a brief literature review.

PALABRAS CLAVE
Leiomiosarcoma primario de pene

Resumen El leiomiosarcoma primario del pene es un tumor extremadamente raro, existen sólo 46 casos reportados en la literatura médica mundial. Nosotros presentamos el caso de un paciente masculino de 36 años de edad, con un tumor en la base del pene, el estudio histopatológico muestra células fusocelulares compatible con “leiomiosarcoma primario de pene”. Agregamos un caso a la literatura médica, y hacemos una breve revisión del mismo.
Primary leiomyosarcoma of the penis is an extremely rare tumor. There are only 46 cases reported in the international medical literature. This type of malignant tumor has a very low incidence and varies demographically with a rate of 0.1 to 0.9/100 000 men, and in the developed countries of Europe and the United States, up to 19/100 000 men. In the poorer countries of Africa and South America, there is a 10% incidence of malignant tumors in men.

The age range varies and extends from six to 80 years. Only three cases have been described in individuals under the age of eighteen; the majority of cases present in the third and fifth decades of life. Patients can be asymptomatic and treatment is surgical, from lesion excision to partial or total penectomy. It has a poor response to chemotherapy and radiotherapy.

Case presentation
A 36-year-old man sought medical attention for a subcutaneous nodule at the base of the penis of 19-month progression that was associated with occasional pain. Physical examination revealed a 5.5 x 4 x 3.5 cm mobile tumor that was not adhered to the deep planes and that had even, smooth edges. There were no inguinal adenopathies, and the rest of the examination was unremarkable. Routine laboratory workup was normal. No evidence of metastatic disease was seen on the chest x-ray. A penile ultrasound (US) image was taken and showed an increase in volume at the base and right lateral wall of the penis and three predominantly echoic, heterogeneous, poorly-defined images with an ovoid shape and lobulated contours, with no interface between them. The largest of the three measured approximately 40 x 28 x 30 mm with a volume of 18cc; the other two were adjacent to it and had the same characteristics, measuring 28 x 16 x 22 mm. A Doppler study showed an increase in vascularity, apparently dependent on the right corpus cavernosum; at the level of the base there was engorgement of the innermost part of the left corpus cavernosum, suggesting extension into it; the testes were normal in shape and size.

The biopsy reported: high grade fusocellular sarcoma, conclusive for “primary leiomyosarcoma of the penis”. The patient was managed as follows: he initially underwent wide local resection surgery in which margins were negative; however, there was superficial location 1 mm from the surgical site and so conservative surgery was performed. Surgery is the criterion standard for patients with sarcoma of the penis, but there is no standard management for patients with leiomyosarcoma of the penis, and so all treatment is based on the experience of the cases that have been reported worldwide. There are no management protocols in relation to adjuvant treatment and its benefits. In our case, the patient was given adjuvant radiotherapy, due to neoplastic positivity found 1 mm from the surgical site.

Macroscopic description
A nodule of tissue partially covered by an excess of skin, ovoid in shape, irregular, of medium consistency, and measuring 5.5 x 4 x 3.5 cm, that upon cutting, displayed a solid surface due to nodular, pale pink, tissue with a neoplastic aspect.

Microscopic description
Neoplastic tissue made up of fusiform cells arranged in irregular bundles, with zones of tumor necrosis (Figure 1). The cells had a pale cytoplasm, moderate in quantity, with a fibrillar aspect; the nuclei were elongated with blunt ends, hyperchromatism, and an apparent nucleolus; up to 20 mitotic figures were observed per 10 fields at 40x, and in some fields there was pleomorphism (Figure 2).

Diagnosis: High grade fusocellular sarcoma with 30% tumor necrosis. Immunomarking: Cytokeratin E 1/3 and desmin were negative in neoplastic cells and smooth muscle actin and vimentin were positive in neoplastic cells (Figures 3 and 4). With these results the definitive diagnosis was “primary leiomyosarcoma of the penis”.

Discussion
The incidence of malignant neoplasia of the penis is very low and varies demographically from 0.1 to 0.9/100 000 men.
men; in the developed countries of Europe and the U.S. the figure goes up to 19/100 000 hombres; the poorer countries of Africa and South America present with a 10% incidence of malignant tumors in men. However, tumors of mesenchymal origin are extremely rare and make up less than 5% of all malignant tumors of the penis. The majority of penile tumors of epidermoid and mesenchymal origin are exceptional, and only a few cases are reported in the international medical literature; just 46 cases appear between 1930 and 2006. Dehner and Smith carried out a review of primary sarcoma of the penis, in which leiomyosarcoma represented 13.5% of all penile sarcomas. The first case was reported in 1930 by Levi; Kreiberg and Meller Mc Kenzie et al. classified them as superficial and deep; Pratt and Ross later classified them as superficial, deep, and of the prepuce. Depth is of prognostic importance for the patient. Up until the present, only 20 cases of deep location had been reported, and 26 cases in general. Because of the few cases reported in the medical literature, treatment modality has not yet been established, and varies between local surgery, wide resection, and adjuvant radiotherapy or chemotherapy.

Age also varies widely, ranging from six to 80 years of age. Only three cases have been reported in under age patients; the majority of cases present between the third and fifth decades of life. Symptoms are indolent, and present as a nodule in the majority of cases; they are superficial or deep, depending on their location. The superficial tumors originate in the smooth muscle fibers of the dartos and generally are located in the distal portion of the penis. Those presenting in the prepuce are of slow growth, have low metastatic potential, and present at early ages. The deep tumors and those presenting in the prepuce originate in the smooth muscle of the glans of the preputial dermis or of the corpus spongiosum, and are situated around the vascular canals and in the interstitial space, forming subcutaneous nodules that grow slowly, are proximal, and present at more advanced ages; they tend to metastasize early or invade the urethra.

Differential diagnoses of the malignant fusocellular tumors are fusiform cell carcinomas, fibrosarcomas, and neurogenic sarcomas. The most common of the differential diagnoses is Kaposi sarcoma: positive PAS stain, prominent lymphoplasmacytic infiltrate, immunoreactivity for CD31-CD34, and negative desmin. Histiocytoma is rare in the penis and shows negative activity for desmin and actin. Leiomyosarcoma is distinguished from sarcomatoid carcinoma of the penis through its lack of cellular atypia on the surface of the epithelium and absence of immunoreactivity for keratin. Immunohistochemistry studies are very useful for establishing diagnosis.

Leiomyosarcomas have been described as having a tendency to recur and those of deep location present with a greater metastatic potential. Lymph node disease is very rare, and so lymphadenectomy is not recommended; if it presents there is a high rate of distant disease, principally to the lung. Other metastatic sites are the liver and brain. Recurrences are treated with re-excision or amputation, and can be accompanied with adjuvant chemotherapy or radiotherapy.

Treatment depends on tumor location, and surgery is the treatment of choice. However, radiotherapy and chemotherapy have been used as adjuvants. Superficial tumors are treated with local excision with disease-free margins; deep tumors with partial or total amputation in distal lesions, and radical penectomy in proximal lesions. However, patients with deep lesions have a greater tendency for recurrence and for developing metastatic, lymph node, or distant disease, in which case radical penectomy is recommended. Radiotherapy in sarcomas of the penis has not been shown to improve survival, and due to the small number of cases, no clinical studies have been conducted showing the usefulness of radiotherapy in cases of leiomyosarcoma of the penis. Brachytherapy is being used in superficial lesions, but its results have not yet been reported. Chemotherapy with anthracyclines and etoposide have shown few results, however these two modalities are used in patients that cannot undergo palliative surgery. Due to the high incidence of recurrence with local excision, adjuvant radiotherapy and chemotherapy are recommended in such cases, despite the fact that they have not shown an increase in overall survival.
In our case, we performed local resection with wide tumor-free margins, however there was positivity 1 mm from the surgical site, and therefore treatment was complemented with superficial external radiotherapy of 110Kv (6 400cGy) one month after surgery. This has resulted in a disease-free survival at the 38th month of follow-up and the patient continues to be under surveillance and has had no signs of disease.

The most important prognostic factor is whether the initial presentation is superficial or deep. Our patient’s lesion was superficial, which is why partial resection was carried out.

A review at 12 years of follow-up reported that 26% of the patients presented with local recurrence at three to 18 months from the first procedure, and of those that had deep lesions, eight patients died within the first month and 36 months of follow-up after surgery. Deep tumor survival was nine months and location was the main outcome factor. Despite the fact that there are no studies suggesting that adjuvant radiotherapy reduces the risk for recurrence, it is best to be radical and complement these cases with adjuvant radiotherapy, given that it is a highly recurrent disease. There should be strict surveillance in the follow-up with chest x-ray, due to the high recurrence rate and the development of distant metastasis.

Conflict of Interest
The authors declare that there is no conflict of interest.

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References