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Leiomyosarcoma of the Penis, an Exceptional Entity

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ABSTRACT

In tumors of the penis, mesenchymal tumors are extremely rare and within them, sarcomas are exceptional. We report a patient with a sarcomatous lesion treated with conservative surgery with good surgical outcome and the review of the literature, to present the latest advances in the treatment of this unusual entity.

Introduction

In tumors of the penis, squamous cell carcinoma is the most common, accounting for 95% of the total.[1] On the contrary, mesenchymal tumors are extremely rare and within them, sarcomas are exceptional. The pathologic classification of these tumors is divided as superficial and deep lesions.[2] The superficial type usually appears in tegumentary support areas, in the distal surfaces of the penis or in the smooth muscle of the glans; forming slow-growing subcutaneous nodules,[3] with a tendency to recur, but with a better prognosis. The deep type are born of the supporting structures of the corpora cavernosa and spongiosum. The latter ones metastasize prematurely invading the urethra; therefore the outcome is much worse.[4]

Case report

39-year-old patient, smoker, frenuloplastia intervention in 1990, comes to the urology clinic in December 2009 due to presenting an enlargement of a tumor in the ventral area of the penis of 1 year's evolution, accompanied by pruritus. Physical examination revealed a palpable tumor of 1 cm in diameter in preputial frenulum area, adjacent to the urethra, mobile, painless and without inguinal lymphadenopathy.

Excision of the lesion is performed under local anesthesia, respecting urethra and aiming macroscopically that remains its indemnity.

The anatomopathology result reports: leiomyosarcoma composed of long spindle cells. Proliferating cells showed nuclear atypia and mitotic index of 2/10 with high power field. With Ac. Ki67 proliferation index was 25% (Figs. 1 and 2).

Extension study was conducted by CT toracoabdominopelvic, with no evidence of pathological findings.

Given this situation, it was decided to reoperate with conservative intention, extending surgical margins of the lesion to ventral urethra and glans sending intraoperative sample which reported the absence of tumor cells.

The patient had a postoperative evolution without complications. Currently the patient is asymptomatic, with uro-oncologic controls and no signs of local recurrence for 3 years and 6 months of follow-up.

Discussion

The non-squamous malignancies are rare in cancer of the penis. Sarcomas are uncommon tumors whose malignant lesions show a greater predilection for the proximal region of the penis while for the benign, it is the distal. Myogenic lesions are rare in this subgroup, the third in frequency.

The origin of leiomyosarcomas of the penis is related to the following anatomical structures: dartos muscle, arrector pili

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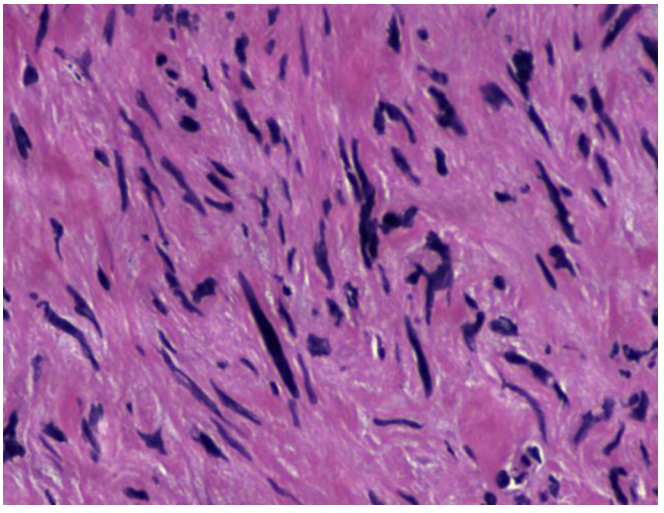

Figure 1. Spindle cells with clear nuclear pleomorphism and high mitotic index.

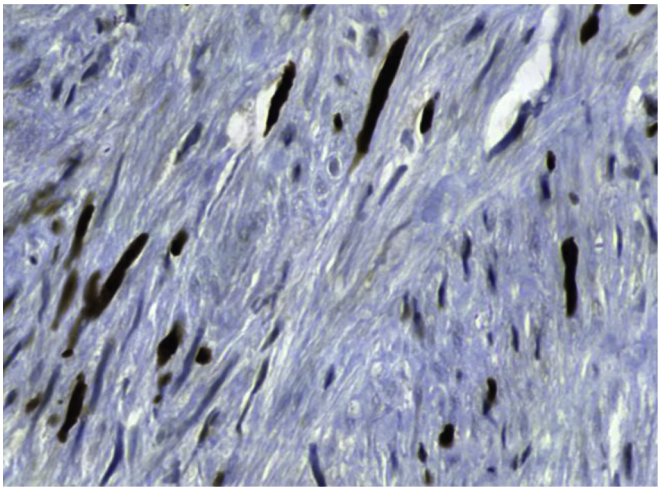

Figure 2. Immunohistochemistry Ac.Ki767 demonstrating the high proliferation index.

muscle, muscle layer of superficial vessels outside the tunica albuginea and muscular layer of deep vessels within the cavernous and spongy bodies.[5]

Leiomyosarcomas are divided into superficial and deep. The superficial tumors tend to appear in tegumentary areas of support in the proximal surfaces of the penis or glans smooth muscle, forming subcutaneous nodules that are slow growing, with little tendency to invade deeper structures but with a high relapse rate. For this reason, after local tumor resection, the treatment of choice is partial penectomy with negative margins of resection. In contrast, deep tumors arise from the supporting structures of the corpora cavernosa and spongiosum, invading nearby structures such as the urethra, with early metastases, so the prognosis is much worse. In these cases, total penectomy was the standard procedure chosen.

Fetsch et al.[6] studied 38 penile leiomyosarcoma, only 8% reported metastases in patients with superficial tumors, while 50% of metastases in deep tumors. In terms of size, no metastases were found in tumors less than or equal to 2 cm, whereas in tumors from 2 to 5 cm, metastases were found in 29% and 50% in those over 5 cm, 36% of patients with deep tumors died of them, whereas none of the patients with superficial tumors died. The differential diagnosis includes spindle cell neoplasms such as sarcoma sarcomatoid, Kaposi sarcoma, desmoplastic and spindle cell melanoma, malignant fibrous histiocytoma and leiomyoma, which on the surface must be distinguished from dermatofibrosarcoma protuberans.[7]

Lymphadenopathy on clinical presentation, even in deep tumors with a late onset, with rare metastases at the start of the clinic. The most common site of spread are the lungs and less frequently the pericardium, stomach and kidney.[8,9]

Chemotherapy and radiation therapy would not be indicated as primary and even its role as a therapy, whether adjuvant or neoadjuvant, has no scientific evidence yet to support it.

Regarding prognosis should take into account tumor size, presentation (superficial or deep) and the treatment of choice; partial penectomy in the superficial, initial total penectomy in the deep. In highly selected cases of superficial tumors there is the option of an extended tumorectomy of the lesion, closely followed up given the high rate of local recurrence.[10]

Conflict of interest

The authors declare that they do not have any conflict of interest.

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