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Vulvar Leiomyosarcome Rare Pathology: A Case Report and Review of the Literature

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Abstract: *Leiomyosarcoma of the vulva is a rare malignant tumour with smooth muscle differentiation, occurring in the 4th or 5th decade. Its malignancy is difficult to assess. The rarity of the disease is a challenge for prognostic classification. We report the case of a 49 years old patient, followed and operated in the University Hospital Center HASSAN 2 of Fez Morocco.*

Keywords: sarcoma, soft tissue tumors, vulva

Introduction

Vulvar cancer is a rare disease and accounts for about 5% of gynaecological cancers. Vulvar leiomyosarcoma accounts for only 1% of vulvar cancers[1] and usually occurs in the 4th or 5th decade. Its malignancy is difficult to assess and the rarity of the disease is a challenge for prognostic classification [2].

Our case adds to the literature on leiomyosarcoma of the vulva and through it we will emphasize the diagnostic modalities and management of this rare pathology.

Observation :

We report the case of a 49 year old female patient, multiparous, with a family history of a sister followed for breast cancer, who consulted for a vulvar tumour evolving for one year and progressively increasing in volume. The clinical examination revealed a budding tumour of the right labia minora classified as T1bN0Mx (Figure 1). Tumour biopsy with immunohistochemical study was in favour of a melanoma or a peripheral sheath tumour. Pelvic MRI showed a vulvar lesion process with irregular contours and a central necrotic area with no locoregional extension of 26 mm in length. The patient underwent a total vulvectomy with bilateral inguinal curage with definitive anatomopathological and immunohistochemical examination in favour of leiomyosarcoma with a healthy resection limit and a single positive inguinal lymph node. The treatment was complete with radiotherapy.

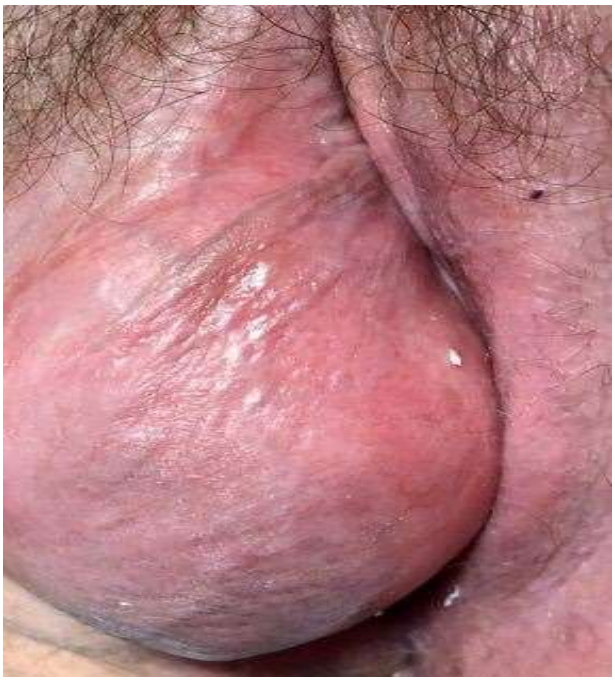

Figure 1: Budding tumour of the right labia minora

Discussion:

Smooth muscle tumours are rarely located in the vulva, they are much more common in the uterus. Uterine smooth muscle tumours have their own criteria for malignancy that do not apply to vulvar smooth muscle tumours [3].

The average age of diagnosis is 50 years and the diagnosis is usually delayed because the vulvar mass is mistakenly considered to be a bartholin cyst or an abcess and the presenting signs are discomfort associated with a vulvar mass, dyspareunia or both [4]. The prognosis and risk of recurrence is a function of tumour size, extension to adjacent tissue and mitotic activity [5,6]. In our patient the tumour size was less than 5cm with healthy resection limits but with significant mitotic activity with frank atypia and the complement of radiotherapy after surgical treatment was adopted knowing that radiotherapy did not show any survival benefit [7].

In our case the initial diagnosis on biopsy was either melanoma or peripheral sheath tumour but on definitive pathology the diagnosis of leiomyosarcoma was made by an immunohistochemical study showing tumour cells intensely and diffusely expressing anti-AML antibodies, anti-HcalDesmone and anti-Ps100 antibodies and lesser and focal anti-CD10 antibodies and the other diagnoses were eliminated carcinomatous nature, melanoma by MelanA and HMB45 negative and also vascular tumours, although the CNGOF requires three diagnostic criteria including the size of more than 5cm which was not found in our case (2)

Conclusion

Vulvar leimyosarcoma is a very rare pathology that must be clinically suspected in front of vulvar masses with slow progression, the treatment is surgical and the prognosis depends on the degree of local invasion and the adjuvant treatment has not shown any benefit in terms of survival

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