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Leiomyosarcoma of the vulva: a case report and review of the literature

Giuseppe Comerci[1], Venelia Picarelli[1], Emilia Crisanti[2], Giandomenico Raulli[2]

[1] Gynecology Oncology Service, Department of Obstetrics, Gynecology & Pediatrician
[2] Department of Pathology, "Santa Maria delle Croci" Hospital, Ravenna, Italy

ABSTRACT

Leiomyosarcoma of the vulva is very uncommon and it represents 1% of the vulval neoplasms. A case of 44-years old lady affected by leiomyosarcoma of the vulva is presented.
She was admitted to our unit because of complaining painful nodule localized on the left labia majora. It was supposed to be a vulval abscess and under local anesthesia it was excised. Histology report was consistent with leiomyosarcoma of the vulva. Imaging did not reveal any other lesion. A second surgery has been done in order to guarantee optimal free margins. She did not require adjuvant treatment. After five years of follow-up there is no evidence of local recurrence and distant metastases.
Most vulval lesion are benign but the possibility of leiomyosarcoma should always be considered if a rapidly growing hard vulvar mass is found. The surgical excision with widely free margins is recommended. Long term careful follow-up is necessary.

Keywords: Gynecologic oncology; surgery in GYN cancers; cancer of the vulva; gynecologic imaging; epidemiology of GYN cancers.

SOMMARIO

Il leiomiosarcoma della vulva è una neoplasia maligna molto rara e rappresenta l'1% dei tumori vulvari.
In quasto articolo si illustra il caso clinico di una paziente di 44 anni ricoverata presso la nostra unità di ginecologia per la comparsa di un nodulo dolente a carico del grande labbro vulvare di sinistra.
La diagnosi di ammissione è stata di ascesso vulvare che, in anestesia locale, è stato escisso. La diagnosi istologica è stata di leiomiosarcoma della vulva.
In seguito a tale diagnosi è stata fatta una stadiazione per immagini sottoponendo la paziente ad una TC total-body che non ha rivelato secondarismi.
Una chirurgia di seconda istanza è stata successivamente eseguita in funzione di garantire sufficienti margini di tessuto indenne. Non è stato necessaria alcuna terapia adiuvante.
Dopo cinque anni di controlli clinici e strumentali la paziente non ha sviluppato lesioni locali o a distanza.
Gran parte delle lesioni vulvari sono benigne ma la possibilità che possa trattarsi di una neoplasia invasiva dovrebbe essere sempre considerata qualora si noti una lesione di consistenza dura e a rapida crescita. E' raccomandata fortemente una chirurgia radicale con ampi margini di tessuto sano. E', inoltre, opportuno eseguire un attento e duraturo follow-up.

INTRODUCTION

Malignant tumors of the vulvar soft tissue are very rare. Leiomyosarcoma is the most frequent histological type, representing 1% of the malignant neoplasms of the vulva[1-3]. However, it is an aggressive disease which usually present as slowly growing nodule and it can often mistaken for a benign lesion causing a delay in diagnosis. The major incidence is seen in women between 40 and 55 years old. These tumors are thought to originate from smooth muscle within erectile tissue or blood vessel walls, the round ligament, the dartos muscle, or the erectorpili muscle[4]. Primary therapy is surgical and prognosis is difficult to estimate based on the rareness of these tumors[1]. The case of a patient who had a leiomyosarcoma growth in the left labia majora is reported.

Correspondence to: giuseppe_comerci@yahoo.com

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CASE

Forty-four years old, para 1 was admitted to our unit because of complaining painful 2 cm nodule localized on the left labia majora. The patient reported that the lesion was present since years but during the last four months increased rapidally in size. It was thought to be a possible vulvar abscess but white cell count with neutrophil count was normal and reactive protein C was negative. Anyway, the following day, under local anesthesia it was found to be a 2 x 2 cm hard mass on the left labia majora. It was incompletely excised. Histology confirmed leiomyosarcoma. The tumor greatest dimension was 1,5 cm. Grossly it has white cut surface and ill-defined margins. It is composed primarily of spindle cell arranged in fascicles with area showing pleomorphism and mitotic figures (average 18 per 10 HPF), with infiltrative margins and area of necrosis (**Figure 1**). It

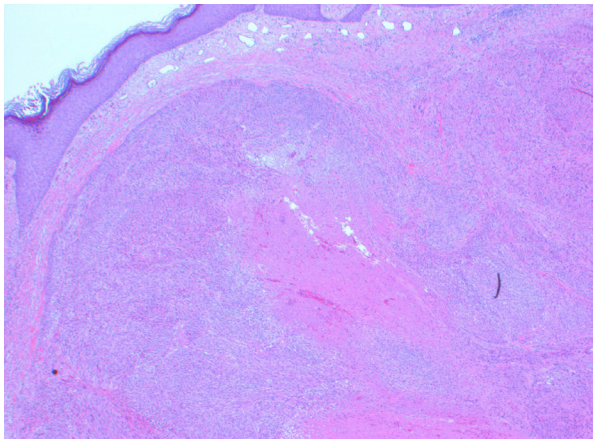

Figure 1.
The tumor is composed primarily of spindle cells arranged in fascicles with necrosis, infiltrative margins and mitotic figures (18 x 10 Hpf).

stains for smooth-muscle actin and desmin, there is no staining for keratin or S-100 (**Figure 2**). A CT scan of the thorax, abdomen and pelvis was performed, revealing no additional lesions. A second surgery was scheduled and a wide local excision of the left vulva, deeper up to the fascia, was done. Microscopic examination showed a 5 mm residual tumor with free margin greater than 1 cm. Five years after the diagnosis, patient remains well with no clinical evidence of recurrence.

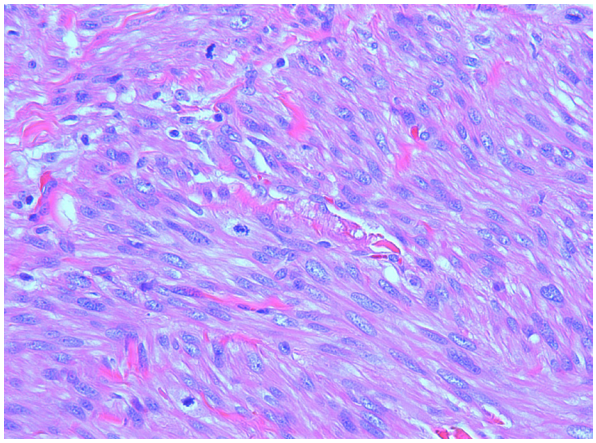

Figure 2.
The cells stain for smooth-muscle actin and desmin; no staining is observed for keratin or S-100 (40x).

DISCUSSION

This is the first case of vulvar leiomyosarcoma reported in our unit as, already mentioned, the disease is rare. As in this case, an enlarging painless mass, located in labia majora or minora is usually the only symptom of vulvar leiomyosarcoma. Its biological behavior in the vulva is similar to that in other subcutaneous tissue locations, characterized by high rate of local recurrence and frequency of metastasis by hematogenous route. So these tumors have an insidious evolution and usually reach huge dimensions before accurate diagnosis is done. Because of the rarity of these neoplasms our knowledge is limited.

Differentiation between benign and malignant lesion in the vulva is difficult and it causes diagnostic problems. To overcome this problem, Nielsen et al. in 1996 proposed an useful scoring system in order to differentiate leiomyomas and leiomyosarcomas of the vulva. The most important pathologic findings are: tumor diameter greater than 5 cm, infiltrative margins, more than five mitotic figures per 10 HPF and moderate to severe nuclear atypia. Tumors with three or more of these characteristics are considered sarcomas; those that have only one of these characteristics should be diagnosed as leiomyomas and those with only two characteristics should be considered benign but atypical leiomyomas.

A review of the literature shows a wide variety of treatment options due to a small number of cases but most of the authors feel that a radical vulvectomy is only mandatory if a wide local excision should not guarantee a pathological free margin greater than 1 cm[5]. In our case, because

the lesion was 2 cm in greater dimension and localized in the labia majora we have had the possibility to preserve the clitoris in order to decrease the consequences for psychosexual function. In our case there were no clinically and radiologically enlarged lymph nodes at the moment of diagnosis; we decided not to perform inguinal lymphadenectomy. As reported in the literature, this disease behave aggressive in general, with a high rate of local recurrence and distant metastases by haematogenous route (liver and lung mainly). In the review analysed by Aartsen et al, the authors found that the disease may present with possible late recurrence[6]. For this reason we have planned to follow-up our patient for at least ten years.

In conclusion, any vulval lesion with unusual characteristics or insidious evolution should be investigated rapidally, in order to make an accurate diagnosis, and due to the rarity of the disease referral to a gynecological cancer centre is mandatory[7-9].

DISCLOSURE

The authors declare that they have no conflict of interest.

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