Malignant vulvar melanoma: Colposcopic evaluation and review of the literature. A case report

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Summary

Because of the rarity of this kind of vulvar neoplasia, the diagnostic difficulties (clinical and histopathological) conditioning the therapeutic approach and the missing iconographical material, we report a case of vulvar melanoma seen at the Department of Obstetrics and Gynecology of the University of L'Aquila in April 2001, together with a review of the literature. Owing to radical vulvectomy and bilateral inguinal lymphadenectomy with the Byron three-incision approach the histological report was: epithelioid cell apigmented melanoma radially spreading (MMSS), a tumor-free margin of at least 1.7 cm with sufficient lympholitic infiltration.

Key words: Vulvar melanoma; Colposcopy; Lymphadenectomy.

Introduction

About 4% of all gynaecological neoplasias affect the vulva and although only 1-2% of the corporeal surface is affected, this area is involved in about 3-7% of malignant melanomas.

Therefore, malignant melanoma represents the second vulvar neoplasia in frequency (8-10%), after squamocellular cancer (90-95%) [1, 2].

The incidence of vulvar melanoma is strongly related to age and it appears most frequently in women around 60-70 years old; neoplasias have also been diagnosed in premenopausal women and in women with fair complexions [3].

However, other organs (vagina, cervix, clitoris, ovary) [4] are rarely involved in malignant melanoma. The clinical aspect of malignant vulvar melanoma is rather difuse: 1) superficial spreading malignant melanoma (SSMM) with planar lesions, a superficial spreading tendency associated with poor penetration and frequency (55%); 2) nodular and isolated melanoma (NMM) often associated with satellite nodes, both pigmented and achromatic.

NMM represents the most aggressive tumoral typology with the highest metastatic tendency. Both the first and the second melanoma are generally ulcerated and can be completely apigmented. Acromial lentiginous superficial spreading melanoma is very rare (10%) [5].

Vulvar melanoma symptoms, initially absent, are successively no different from other vulvar neoplasias [6].

The subjective sensation of discomfort (caused by the new lesion), associated with vulvar itching and/or burning, plus very poor bleeding needs immediate and accurate examination.

Morrow and Rutledge [7] have reported this clinical aspect in more than one-third of patients affected by vulvar melanoma and about 60% of them were evaluated via a careful medical history and specialist examination, and four months later clinical exordium.

Due to the rarity of this kind of vulvar neoplasia, and the diagnostic difficulties (clinical and histopathological) conditioning the therapeutic approach and the missing iconographical material, we report a case of vulvar melanoma treated at the Department of Obstetrics and Gynaecology of the University of L'Aquila in April 2001 together with a review of the literature

Case Report

The persistence of vulvar symptoms (Table 1) resistant to the therapy given by the family doctor induced a 73-year-old woman to undergo a specialist examination. In April 2001, she was referred to the cervical-vaginal pathology center of the Department of Obstetrics and Gynaecology at the University of L'Aquila.

The patient underwent several clinical examinations (bacteriological examination and vaginal tampon) to study the phlogistic pathology, with conseguently negative results and then she underwent vulvoscopy. The woman, gravida 3, caucasian, married, non smoker, had previously undergone a total colpohysterectomy without bilateral salpingo-oophorectomy for uterine haemorrhagic fibromatosis in 1962.

She had been taking the following drugs for about seven years: cardioaspirin, 100 mg/daily and tiamazolo, 5 mg/daily as prophylactic treatment for a previous ictus and hyperthyroidism.

The colposcopic examination (Colposcope Zeiss-Germany with a photographic device contax 167 MT) both in the vaginal

Table 1. — *Clinical condition (vulvar symptoms).*

– Sensation of discomfort caused by the mass
– Vulvar itching and or pain
– Poor bleeding
– Ulceration
– Lesion like a "scratch"

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and vulvar site showed integral vaginal mucosa, rettocele 3° and hyperchromic areas (Figures 1 and 2) located in the external genitalia with pluristratified keratinised undamaged areas.

Using the traditional clinical criteria to diagnose pigmented lesions ABCS (aspect, borders, colouring, size), in the genitalia site as well, the colposcopic examination showed an erythematous raised area like a "contour map" in the borderline area between the 3rd superior and middle level of the of the left minus labium, 1.6 cm in diameter encircled by a pigmented lesion.

Another smaller (0.7 cm in diameter), whitish-yellow, partly raised mass with pigmented borders was located in the upper part. Other small planar and dark areas, 0.4-0.6 cm in diameter, were located bilaterally in the vestibulum, about 1 cm from the external urinary meatus.

In conclusion, in the third inferior level of the labia minora, a squamous cell hyperplasia was evident; it was associated with a reduction of the epidermis appearing whitish, dry and unelastic with poor solution continuity and microhaemorrhage, mostly on the right of the interlabia site.

According to the Breslow index (Table 2) [8], to obtain an accurate evaluation, the woman underwent a superficial colposcopic-guided biopsy in the borders, with extirpation of a 2 mm thick specimen.

Table 2. — *Breslow index (infiltration level)*

Stage I:	invasion < 0.75 mm
Stage II:	invasion between 0.76 and 1.50 mm
Stage III:	invasion between 1.51 and 2.25 mm
Stage IV:	invasion between 2.26 and 3 mm
Stage V:	invasion > 3 mm

In Figures 3, 4 and 5 the histopathological characteristics of malignant melanoma after hematoxylin and eosin staining and S100 immunohistochemical protein are evident.

Microscopic report of the superficial colposcopic-guided biopsy: epithelioid cells, amelanotic melanoma, Breslow index 1.7 mm (Stage III). The woman presurgically underwent clinical-haematologic examinations and a computed tomography scan to obtain an accurate evaluation of inguinal and pelvic lymph node involvement and distant metastases.

Following radical vulvectomy and bilateral inguinal lymphadenectomy with the Byron three-incision approach the histological report was: epithelioid cell apigmented melanoma radially spreading (MMSS), a tumor free margin of at least 1.7 cm with sufficient lympholitic infiltration. The melanomatous tissue was found only in the specimens immediately next to the previously biopsied area.

Infiltration thickness was between 1.7 mm and 2.6 mm and there was no nodal metastasis in the 23 lymph nodes surgically extirpated.

The postoperative period was unremarkable except for a vulvar dehiscence restored without needing resuturing.

Discussion

This case report shows several important problems still under consideration by the authors. In the management of this neoplasia, diagnosis, clinical stage, therapy and prognosis can be considered the most important and, at the same time, the most difficult. As for diagnosis, an excisional biopsy of the pigmented vulvar area associated with a local tumor-free excision of at least 2-3 mm should be performed. Furthermore it is necessary to include a thickness extending through the subcutaneous and/or mucous fat: thus the biopsy can be considered adeguate for the diagnosis.

In our experience, this diagnostic approach seems to be a very stereotypical one, because as shown by our decisive histological report, not all pigmented lesions should be considered melanomas.

Thus we consider that radical procedures in the management of the lower female genital tract should not be the rule because the diagnostic criteria can be affected by the diversity of melanosis: apigmented lesions can appear frequently.

On the contrary, we consider that, if necessary, superficial colposcopic-guided biopsy, with a 1.5-2 mm thickness, leads to a certain diagnosis without compromising the therapeutic management (radical or conservative surgery), prognosis and psychological condition of the patient.

Colposcopic vulvar examinations associated with traditional clinical criteria (ABCS) are to be considered fundamental in the diagnosis and especially in the follow-up of pigmented and acromial vulvar lesions.

FIGO criteria adequate in staging squamous vulvar cancer, do not seem accurate in staging vulvar melanoma because FIGO parameters include the tumor size and its superficial spreading, but not the infiltration levels or the lymph node status which are the most important prognostic factors in vulvar malignancy.

In our case report, Breslow's index was usually correlated with the clinical prognosis [8, 9] (2.6 mm vertical infiltration level) and the lymph node status [10] (23 metastasis-free lymph nodes) can not explain the death of the woman 16 months and 10 days after surgery.

Like other authors, we believe that systematic bilateral inguinal lymphadenectomy in patients without lymph node involvement should not be the rule [11].

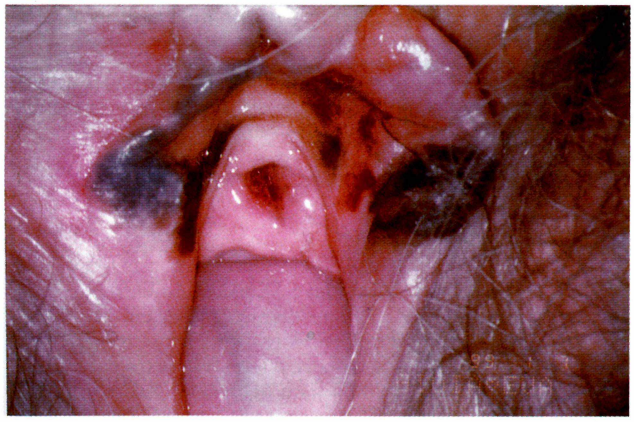
Today it is not clear if malignant melanoma gives rise to sequential metastases through vascular lynphatic spaces. According to Morton and others [12-14], radionuclide intraoperative lymphoscintigraphy associated with the sentinel lymph node technectium-99 m-linked biopsy is an easy and useful approach.

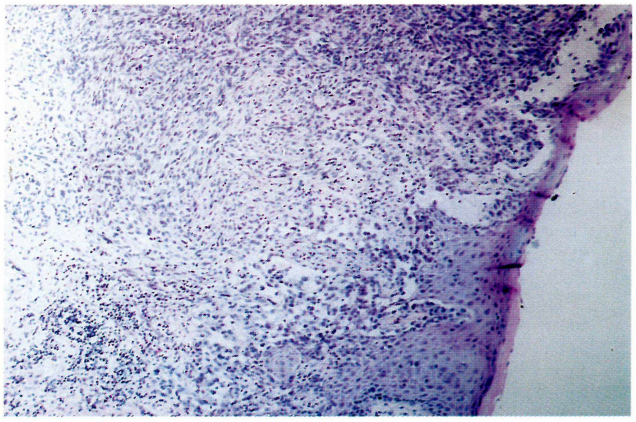
Lymphoscintigraphy could be an easy and reliable method for the detection of patients who have to undergo pelvic-femoral-inguinal lymphadenectomy. This is true if the time of surgery is considered as a prognostic and therapeutic factor.

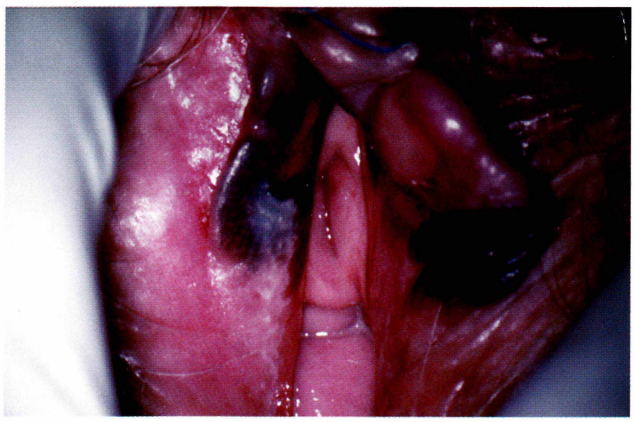
According to several studies the overall 5-year survival rate in patients affected by malignant vulvar melanoma is strongly related to tumoral clinical and pathologic features.

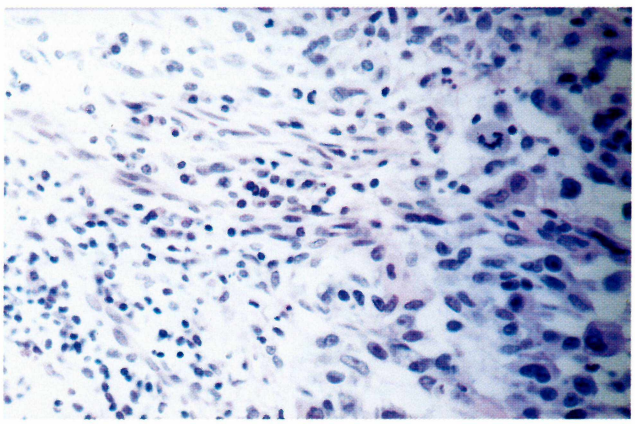
The most important prognostic factors are lymph node involvement, central anatomic site, depth invasion, advanced patient age and ulceration of the lesion.

In our case report no loco-regional lymph node metastases, no organic metastases (brain, liver, lung, kidney) shown by a total body TAC, Breslow index of 2.6 mm (Stage IV), an unulcerated lesion, superficial spreading melanoma, single lesion and good clinical status of the









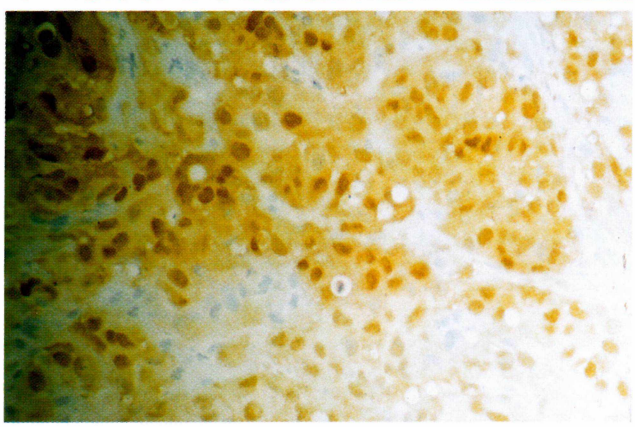


Figure 1. — Retocele III and hyperchromic areas.
Figure 2. — Hyperchromic areas located in the external genitalia with pluristratified keratinised undamaged areas.
Figure 3. — Epithelioid cell apigmented melanoma radially spreading with undamaged skin (Hematoxylin and eosin x 200).
Figure 4. — Lympholytic infiltration of the neoplastic cells, prominent nucleoli, nuclear inclusions and mithoses (Hematoxylin and eosin x 400).
Figure 5. — Neoplastic cells strongly positive for immunohistochemical protein S100 x 400.

patient upon discharge from hospital, can not explain the death of the woman 16 months and 10 days after surgery. Probably, we believe, as confirmed by autopsy, that vulvar melanoma is at variance with cutaneous melanoma; it presents a high clinical variability because of a still unknown lymphatic drainage. Histological reports of the examined specimens showed pelvic and retroperitoneum lymph node involvement consisting of micro- and macro-metastases and also splanchnic metastases. Vulvar melanoma can be considered as a not easily standardized tumor, thus individualized management is needed. However, other and more specific studies are necessary to have a better knowledge about the surgical anatomy and the individualized tumoral spreading pattern in order to improve the patient's quality of life.

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