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Raised vulvar lesions: be aware!

Fernanda S. Resende[1], Claudio Conforti[1], Roberta Giuffrida[2], Mayara Hamilko de Barros[3], Iris Zalaudek[1]

1 Dermatology Clinic, University of Trieste, Hospital Maggiore, Trieste, Italy
2 Department of Clinical and Experimental Medicine, Section of Dermatology, University of Messina, Messina, Italy
3 Professor Rubem David Azulay Institute, Charity Hospital of Rio de Janeiro, Rio de Janeiro, Brazil

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Corresponding author: Claudio Conforti, MD, Università Campus Biomedico (UCBM), Via Alvaro del Portillo 200. 00128 Rome, Italy. Email: claudioconforti@yahoo.com

ABSTRACT Vulvar melanoma is a rare and deadly cancer in women, and the prognosis is often poor. There are limited studies on the dermoscopic features of vulvar melanoma. Described criteria include the presence of blue, gray, or white colors. Herein we present the clinical and dermoscopic characteristics of a hypopigmented and heavily pigmented nodule in a 92-year-old and an 80-year-old woman. Dermoscopy in the former revealed structureless milky-red to white areas, remnants of brown pigmentation at the base and polymorphic vessels, while the latter displayed structureless blue-gray areas with black dots and peripheral lines at the base. In both cases, histopathology revealed a stage III melanoma. Our two cases along with a review of the literature suggest that the dermoscopic features described for diagnosing cutaneous nodular melanoma, apply also for vulvar melanoma. Clinicians should always raise the suspicion if observing plaques or nodules with a dermoscopic polymorphic vascular pattern and blue-black color on the genitals of postmenopausal women.

Introduction

Vulvar melanoma (VM) is the second most common vulvar malignancy, but represents less than 1% of all melanomas and 1.0% to 2.3% of all melanomas in women [1]. It typically affects postmenopausal women with a peak incidence in the seventh decade of life [1,2]. VM is associated with a poor prognosis. The reported five-year survival rates are less than 60% [3].

It is unclear if the poor prognosis of VM is due to delayed detection or a highly aggressive biological behavior, but early identification and intervention may improve patient outcomes.

Dermoscopy improved the early diagnosis of melanoma, but little is known about the dermoscopy features of melanoma of the vulva [4]. Given the low incidence of VM, most of the information is derived from small retrospective case series and single case reports [1,5].

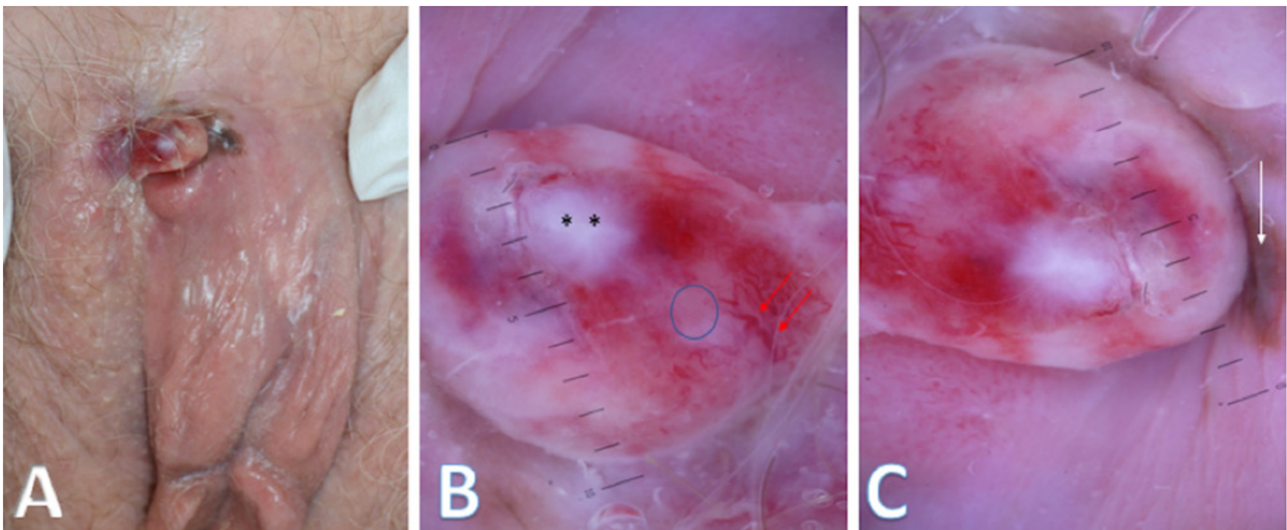

Figure 1. Vulvar melanoma, clinical and dermoscopic presentation. (A) Ill-defined, brownish red colored tumor with smooth surface, 2 cm in diameter. (B) Dermoscopy shows milky-red (blue circle) and white structureless areas (black asterisk), unfocused arborizing vessels (red arrows), with a © structureless brown zone around the nodule (white arrows). [Copyright: ©2018 Resende et al.]

Herein we report the clinical and dermoscopic characteristics of two cases of VM seen in a routine dermatology service and review of the literature on dermoscopy of this rare but highly aggressive melanoma.

Case 1

A 92-year-old Caucasian woman was referred to our skin cancer unit because of a bleeding nodule on the right periclitoral region. Physical examination showed an ill-defined, reddish nodule with a flat, pigmented base measuring 2 cm in diameter (Figure 1A). Dermoscopy revealed milky-red and white structureless areas and polymorphic vessels. At the base of the nodule, a small brown pigmentation was seen (Figure 1B, C). Histopathology revealed a nodular melanoma measuring > 5 mm thickness. At time of diagnosis, the patient had lymph node metastases and died shortly thereafter from widespread metastatic disease.

Case 2

An 80-year-old Caucasian woman was referred to our skin cancer unit with a rapidly growing nodule on the right side of the labium majus. Clinically an ulcerated, blue-black nodule measuring 3 cm in diameter was seen. Dermoscopy revealed structureless blue-gray areas, black and brown dots, and some streaks at the periphery.

Histopathology revealed a melanoma with a Breslow thickness of 1.2 cm.

At time of diagnosis, inguinal lymph node metastases were present, and she died few months thereafter from metastatic melanoma.

Discussion

Our findings are consistent with the current literature suggesting that VM typically affects woman after the seventh decade of life, is often detected at late stage and has a poor prognosis and outcome (Table 1). Among the clinical features of VM, the data consistently report a nodular polypoid shape and lack of pigmentation in up to 27% of cases [6]. This underlines the need for careful evaluation of both pigmented and non-pigmented vulvar lesions, especially if raised.

Dermoscopy is proven to increase early melanoma detection compared to the naked eye, but its impact on early diagnosis of VM remains to be defined.

A review of the literature revealed a dermoscopic description of 22 cases of VM [4,6,7-15]. In the majority of cases, patterns of advanced melanoma such as a multicomponent pattern composed by a blue-white veil, atypical network, irregular streaks, dots and atypical vessels were described (Table 2) [6,7-10].

In a study by the International Dermoscopy Society, the dermoscopic patterns of a large series of mucosal lesions were analyzed. In that study, the presence of blue, gray, or white colors with or without structureless areas yielded a 100% diagnostic sensitivity for mucosal melanoma, only two cases of VM were included [7]. Among those two, was one amelanotic and revealed polymorphous vessels [8].

Clinically, VM can present as a flat or raised lesion with irregular borders and multiple colors [4,6,7] (Table 2). A 2004 review that included 20 cases of VM demonstrated median Breslow thickness at diagnosis of 3.1 mm and the clitoris or periclitoris as the common location and reported the superficial spreading and nodular as the most common histological subtypes.

TABLE 1. Patient demographics and tumor characteristics of vulvar melanoma. Numbers of reported cases are shown in brackets.

References	N	Age	Location	Size in mm	Clinical feature	Tumor thickness in mm
Stolz et al. 2002	1	n.a.	n.a.	n.a.	n.a.	n.a.
Virgili et al. 2004	2	**79	Lab. Min. (2)	> 10 mm (1) < 10 mm (1)	Nodular (1) Flat (1)	0.25 mm (1)
De Giorgi et al. 2005	1	68	Lab. min. & maj.	10 mm	Flat	0.5 mm
Lin et al. 2009	2	n.a.	n.a.	n.a.	n.a.	n.a.
Blum et al. 2011	2	n.a.	n.a.	n.a.	n.a.	n.a.
Ferrari et al. 2011	5	36 43 53 63 67	Lab. min. (3) Clitoris (1) Multifocal (1)	> 10 mm (4) < 10 mm (1)	Nodular (4) Flat (1)	0.6 mm (range 0.5 to 4 mm) *
Ronger-Savle et al. 2011	5	n.a.	n.a.	< 10 mm (3) < 10 mm (4)	Papule (1) Papule (2) Nodular (5)	n.a. (1) 0.3 mm (2) 0.15 mm (3) 0.2 mm (4) 0.12 mm (5)
Rogers et al. 2016	1	50	Lab. min. & clitoris	> 10 mm	Flat	MIS
Oakley A 2016	3	62 67 62	Pubis (1) Lab. maj. (2) Lab. min. (3)	> 10 mm	Nodular	7.2 mm (1) 4.95 mm (2) 7 mm (3)
Blum et al. 2016	1	70	Lab. min.	<10 mm	Papule	n.a..

*mean tumor thickness; ** mean age,
MIS: melanoma in situ; n.a.: not applicable; Lab. min.: Labia minora; Lab. maj.: Labia majorum

TABLE 2. Dermoscopy of vulvar melanoma

Reference	n	Dermoscopy Patterns
Stolz et al. 2002	1	Polymorphous pattern, large blue-gray areas, irregular dots and globules
Virgili et al. 2004	2	Asymmetric darkening, whitish gray area, irregular globules, linear irregular vessels
De Giorgi et al. 2005	1	Nonhomogeneous lesion, central blue-gray area, whitish veil
Lin et al. 2009	2	Multiple colors, homogeneous regions, irregular network, blue-white veil, irregular vessels
Blum et al. 2011	2	Blue, gray, white color, structureless zones
Ferrari et al. 2011	5	Irregular brown black dots, blue-white veil, atypical vessels, reticular depigmentation
Ronger-Savle et al. 2011	5	Irregular-reticular or irregular-polycircular, blue-whitish veil, white veil, regression structures, irregular globules, irregular vessels, milky-red areas
Rogers et al. 2016	1	Asymmetric darkening, structureless areas, central blue and pink colors
Oakley A 2016	3	Asymmetry of color and structure, blue-gray structures, polymorphous vessels
Blum et al. 2016	1	Polymorphous vessels (linear, curved, hairpin-like with different diameter)

Based on this data, it appears that the majority of melanomas were at advanced stage at diagnosis. Whether this was caused by delayed self-detection of patients, low frequency of genital inspections during skin examinations or the tumor biology itself, remains unclear. It is noteworthy that we observed in both of our patients a flat, pigmented area at the base of the nodular tumor, which points towards an at least initially, horizontal growth pattern.

Conclusion

In conclusion, our cases highlight the importance of an inspection of the genital areas especially in postmenopausal woman. The dermoscopic patterns of VM do not differ from melanomas at other body sites. Any pigmented or non-pigmented nodule, dermoscopically exhibiting blue and black or blue-white colors or polymorphic vessels should be carefully evaluated with a very low threshold for biopsy [9].

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