What is your diagnosis?

A 68-year-old woman, affected by a vulvar tumor that was accompanied by burns, was referred to our department. The vulvo-vaginal examination revealed a voluminous blackish clitoral lesion, measuring around 30 mm, and pigmented areas on the labia majora and minora, also affecting the clitoral prepuce and the vulvar vestibule (Figures 1, 2). The patient underwent a total body exam, which did not reveal any other localization.

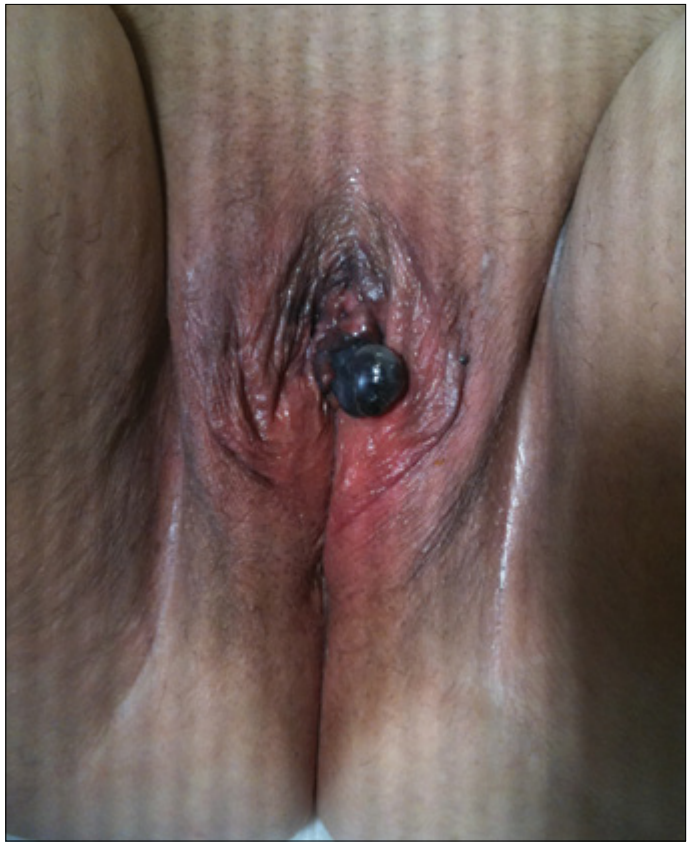

Figure 1. Macroscopic aspect of the vulvar tumor

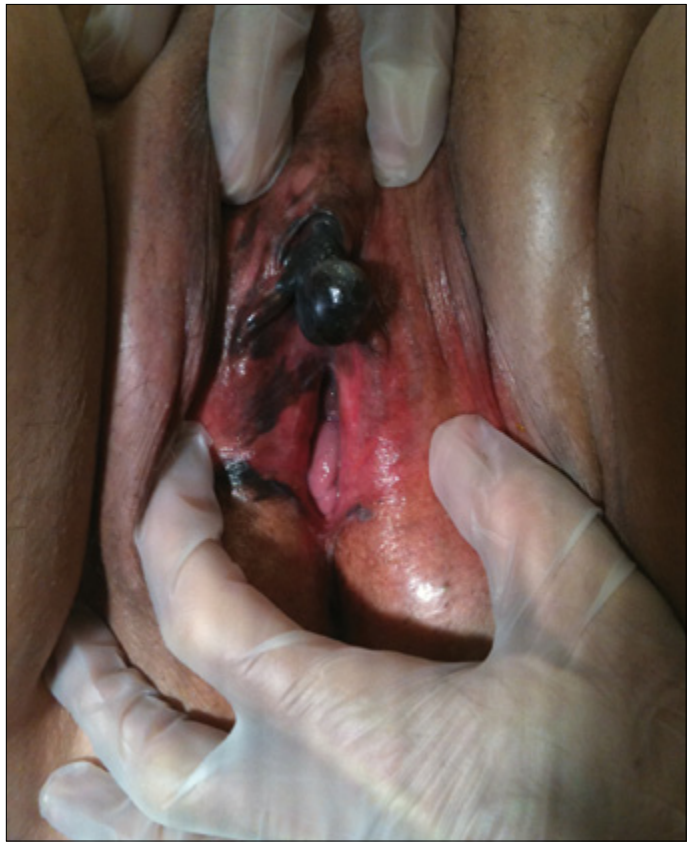

Figure 2. Genital examination findings - pigmented lesions

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Answer

An excisional biopsy on the clitoral lesion and multiple biopsies on the pigmented area were performed. The pathologic report of the tumor of the clitoris revealed melanoma with a desmoplastic component (Breslow tumor thickness of 25 mm and Clark level of invasion of 5) (Figure 3). The microscopic resection was incomplete. The other biopsies revealed an aspect of atypical lentigo. The full-body computed tomography and positron emission tomography were normal. After a multidisciplinary consultation, adjuvant radiotherapy was performed in light of the many studies that have revealed that radical surgery does not improve the survival rate with advanced tumors and can be associated with physical disabilities (1). In this case, complete surgical management would require an anterior pelvectomy, which was not justified insofar as the local extension of disease was important. Because the tumor thickness and its level of invasion were elevated, the risk of local and/or metastatic recurrence was high. Radiotherapy appeared as an alternative management. All of this information was given to the patient and her family. After 36 months, no local and metastatic clinical and radiological evolution occurred.

The density of melanocytes appears greater in the head, face, and neck but also in the genital areas (2). Malignant melanoma represents less than 5% of all skin cancers deriving from melanocytes but is nevertheless responsible for 75% of skin cancer-related mortality (3). Vulvar melanoma is a rare variant with a poor prognosis. It should be staged surgically using the American Joint Committee on Cancer (AJCC) staging system, which incorporates the Clark and Breslow scores (4). Desmoplastic melanoma is a rare entity that is more difficult to recognize insofar as the cytomorphology and the immunohistochemical features are non-specific. It is usually composed of non-pigmented fusiform melanocytes with unusual phenotypic profiles resembling fibroblasts. Its early detection and en bloc large surgical excision with safety margins and regional lymphadenectomy were considered, until recently, the standard therapeutic modality (1). Malignant melanoma appears historically to be radiation-resistant. However, adjuvant radiation therapy can be used in situations of increased risk for locoregional recurrence, like thickness, ulceration, desmoplastic or neurotropic features, and certain anatomic locations (5).

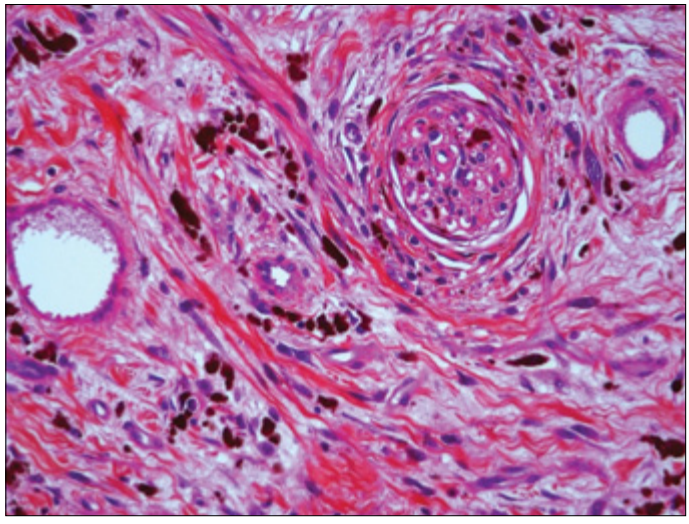

Figure 3. Microscopic view (x200) of the desmoplastic component with important fibrosis

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