

LETTER

Giant basal cell carcinoma of the vulva successfully treated with Sonidegib

Dear Editor,

Basal cell carcinomas (BCCs) of the vulva are rare tumors, accounting for <1% of all BCCs and <5% of vulvar malignancies, that usually affect women in their 8th decade (range 20–100).¹ They are mainly located on labium majus, followed by clitoris, labium minus, perineum, and vaginal introitus.² Complaining symptoms include pruritus, pain, discomfort, and bleeding.² Histologically, nodular subtype predominates on superficial and rarest variants (infiltrative and mixed).² Available treatments comprise local or wide local excision, vulvectomy (bilateral simple, radical, and hemivulvectomy), Mohs micrographic surgery and radiation therapy.² Since diagnosis is commonly made in advanced stages due to discretion or neglect reasons, the management may be strongly demolitive and mutilative with high psychological impact, especially for younger patients.² To date, the advent of systemic target therapies, that is, smoothed inhibitors, has changed the prognosis of inoperable BCC, leading to an improvement in clinical and aesthetic outcomes.^{3,4} We herein present the case of a giant vulvar BCC in a 77-year-old woman successfully treated with 10 months of sonidegib. The patient presented with a 20-year history of an

indurated plaque of the left labia majora that progressively enlarged and became ulcerated. On clinical examination, the neoplasia reached the pubis and was intensively bleeding (Figure 1A). Dermoscopy revealed shiny white lines, polymorphic vessels, large blue ovoid nests, and ulceration (Supplementary Figure 1). Three punches from the neoplastic mass were taken, showing the presence of dermal epithelioid basal nests with high mitotic activity, focal intratumoral necrosis, focal squamous aspects, and stromal desmoplastic activity: the histopathology was compatible with focally pigmented and aggressive BCC with basosquamous and morpheaform aspects. Accordingly, immunohistochemistry was positive for p63, p40, cytokeratin 7, Ki67, and negative for cytokeratin 20, CEA, TTF1, CDX2, S100 and ENA. Since surgical or radiation treatments were not recommended due to tumor size and infiltration, the patient was administered sonidegib at label dosage (200 mg daily) resulting in a slow but progressive improvement. Complete clearance was reached at 10-month follow-up, displaying a residual scar tissue (Figure 1B) as further confirmed by a skin biopsy. Sonidegib treatment duration varies according to BCC characteristics, that is, number, dimension, and histological type (risk of recurrence),

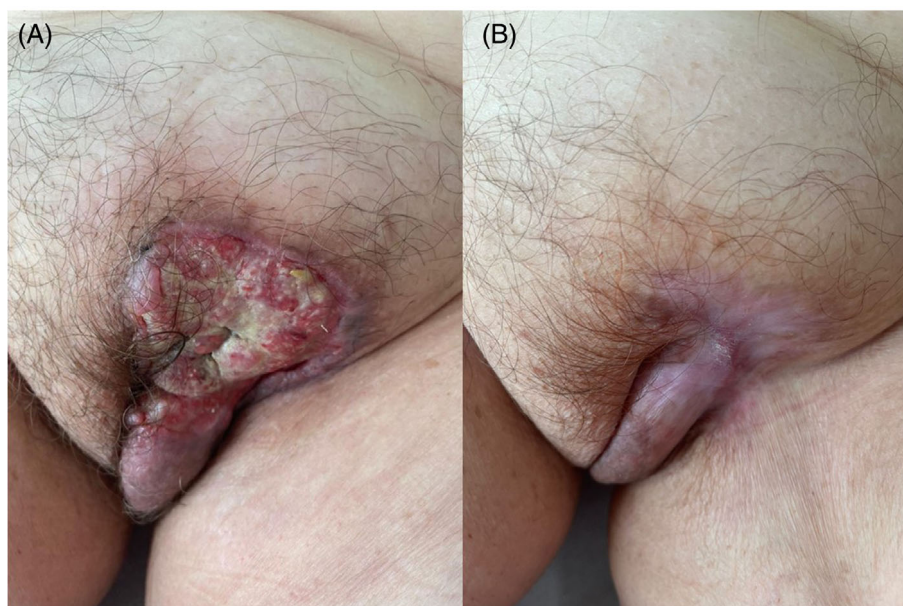


FIGURE 1 Clinical aspect of aggressive BCC of the vulva before (A) and after (B) 10 months of treatment with Sonidegib

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as well as clinical response and entity of side effects.⁵ In detail, our patient referred dysgeusia, alopecia and muscular cramps of mild entity that did not influence the therapeutic protocol nor clinical outcome. Notwithstanding clinical remission, the patient will continue the treatment progressively tapering the dosage, with the aim to prevent potential recurrency that is highly frequent in aggressive BCC.⁵

AUTHOR CONTRIBUTIONS

Conception and design: Elisa Camela, Alessia Villani, Massimiliano Scalvenzi, Claudia Costa. *Material preparation, data collection and analysis:* Elisa Camela, Alessia Villani and Claudia Costa. *First draft of the manuscript:* Elisa Camela, Alessia Villani and Claudia Costa, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript. All authors contributed equally to the manuscript and read and approved the final version of the manuscript.

ACKNOWLEDGMENT

Open Access Funding provided by Università degli Studi di Napoli Federico II within the CRUI-CARE Agreement.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST

The author declares that there is no conflict of interest.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

INFORMED CONSENT

The patient included in the article gave informed consent to publish her case details and pictures.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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