Our Dermatology Online

Case Letter

A mutilate form of a metatypical carcinoma of the face

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Sir,

Metatypical carcinomas are rare, aggressive, non-melanoma skin cancers, combining both basaloid and squamous cell proliferation with metastatic potential [1]. Herein, we report a case of metatypical carcinoma mutilating the face responsible for the invasion of the homolateral eye with blindness.

A 94-year-old patient presented to our clinic with an ulceration on the face evolving for four years and gradually increasing in size. A clinical examination revealed a well-limited ulceration with regular contours, a clean surface, a budding background with infiltrated and pigmented border measuring 15 cm in the left frontotemporal area with the invasion of the homolateral eye responsible for its complete destruction with blindness, as well as multiple BCCs on the rest of the face (Fig. 1a).

A dermoscopic examination revealed bluish-gray pigmented border and hemorrhagic crusts (Fig. 1b).

A histological examination revealed basaloid tumor cells and signs of dyskeratotic maturation in favor of a metatypical carcinoma.

Extension workup with ultrasound of the lymph nodes and parotid area was normal, and a craniofacial CT scan revealed infiltration of fronto-temporal and orbital tissue, infiltration of the temporal and masseter muscles, destruction of the lacrimal gland, and bone lysis without endocranial extension. The patient was discussed in RCP and was a candidate for treatment with radiotherapy.

Metatypical carcinoma is a rare entity with an incidence ranging from 1.7% to 2.7% [2]. As defined by the WHO, "basosquamous carcinoma is a term used to describe basal cell carcinomas that are associated with squamous differentiation" [3].

They have a metastatic capacity more similar to SCCs than BCCs [4].

The tumor usually develops in elderly individuals with a strong male preponderance [2].

The clinical presentation of metatypical carcinomas is non-specific. The most common clinical presentation is a long-standing nodule that gradually becomes ulcerated in sun-exposed areas [1].

Due to the rarity of this entity, very few studies evaluating the dermoscopic features of these tumors have been conducted until now, with the most common dermoscopic criteria for metatypical carcinomas being arborescent vessels, yellowish keratin parches, white areas without structures, scales, hemorrhages, and ovoid nests [5].

Histologic examination remains the gold standard diagnostic method for metatypical carcinomas. Numerous authors have described the presence of histological features of basal cell carcinoma as well as those of squamous cell carcinoma with a transition zone between them [2].

Metatypical carcinomas have an aggressiveness comparable to that of squamous cell carcinoma. Indeed, the tumor is characterized by a rapid increase

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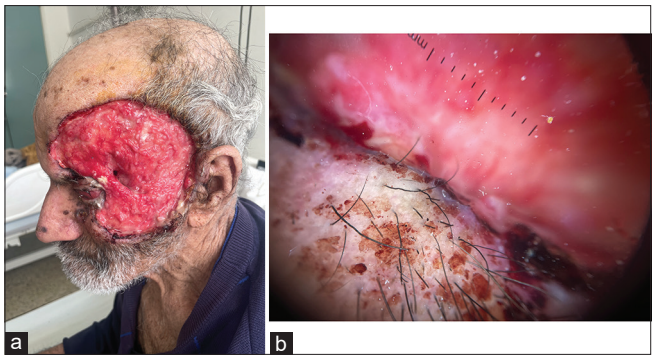

Figure 1: (a) Clinical picture of the ulceration of the face with the destruction of the eye. (b) Dermoscopic image showing a bluish-gray, pigmented border and hemorrhagic crusts.

in size, a high risk of recurrence, and a high metastatic potential.

The bad prognostic factors of these tumors are the male sex, invasive excision margins, lymphatic invasion, and peri-nervous involvement [6].

To date, there have been no established standard therapeutic guidelines for the treatment of metatypical carcinomas.

Several therapeutic modalities may be proposed [1]:

- Wide surgical excision yet with a high risk of recurrence;
- Mohs micrographic surgery as the best treatment option for metatypical carcinomas, with a 8.9% recurrence rate;
- Radiotherapy and chemotherapy in adjuvant or palliative therapy;
- New emerging therapies: Hedgehog pathway inhibitor (vismodegib).

Metatypical carcinomas are highly rare, aggressive, non-melanoma skin tumors that combine features of basal cell carcinoma and squamous cell carcinoma.

Histology is the gold standard of diagnosis.

Early recognition of these tumors allows for rapid management and a better long-term prognosis.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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