

## CASE LETTER

### Extraocular sebaceous carcinoma: tumor presentation of rapid evolution<sup>☆</sup>

Dear Editor,

Sebaceous carcinoma (SC) is a rare malignant neoplasm derived from the adnexal epithelium of the sebaceous glands, with a higher incidence in the ocular region, particularly in the eyelid region, and has a potentially aggressive behavior.<sup>1-3</sup> Older age, previous radiotherapy, and association with Muir-Torre syndrome are predisposing conditions.<sup>1-3</sup>

This is the case report of a 75-year-old white male patient with a history of squamous cell carcinoma (SCC), referred for treatment of facial lesion noted three months before, with rapid growth and bleeding episodes associated with trauma. Upon examination, a 5-cm, rounded, erythematous-violaceous, pre-auricular tumor was observed, with friable and necrotic areas associated with a 1-cm satellite lesion with similar characteristics and a post-SCC excision skin graft scar (Fig. 1). No regional lymph node enlargement was detected. The hypotheses were SCC, SCC metastasis, and angiosarcoma. Histopathological examination (Figs. 2 and 3) showed a dermal neoplasm with polygonal clear cells, evident nuclear pleomorphism, cell debris, and frequent mitoses. Immunohistochemistry disclosed positivity for epithelial markers AE1/AE3 and epithelial membrane antigen (EMA) which, associated with histopathological findings, allowed the diagnosis of sebaceous carcinoma (SC), and

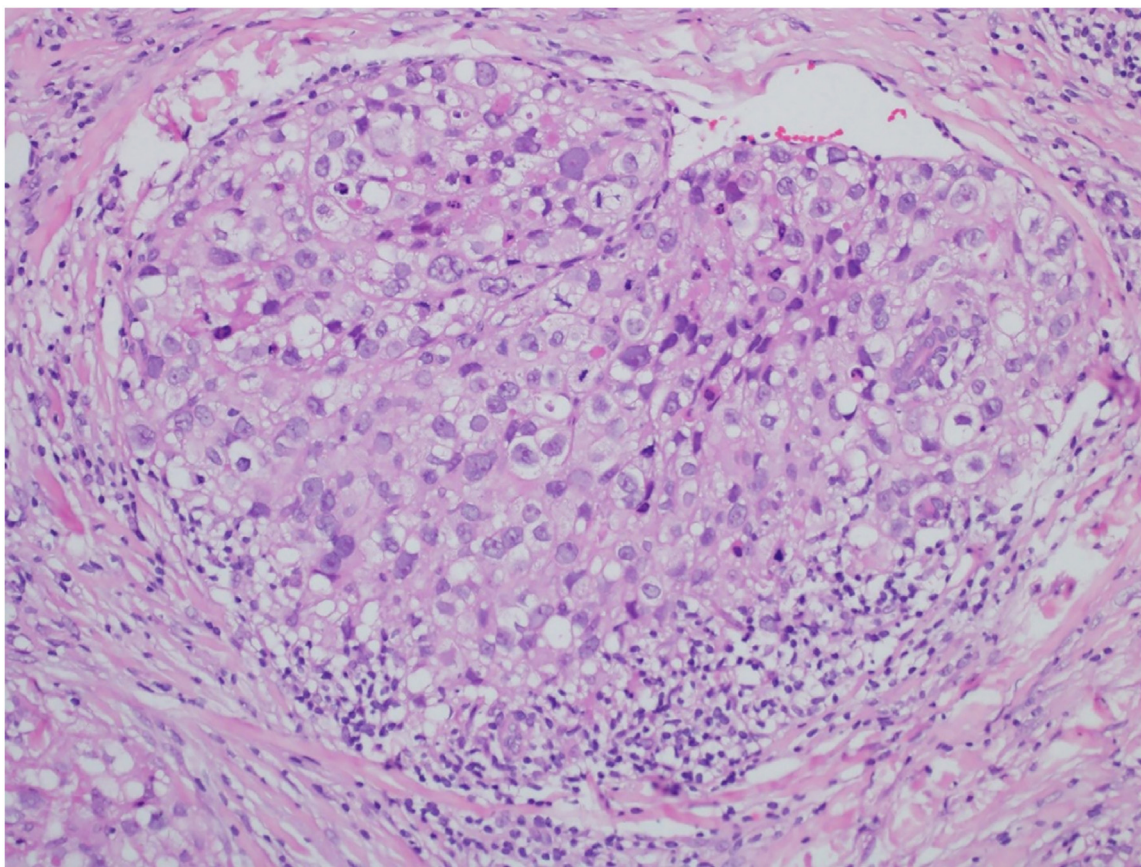


**Figure 1** Extraocular sebaceous carcinoma. Tumor measuring 5 cm, rounded, with an erythematous-violaceous color, located in the right preauricular region, with friable and necrotic areas. Satellite lesion measuring 1 cm with similar features besides a SCC excision scar.

<sup>☆</sup> Study conducted at the Department of Infectology, Dermatology, Diagnostic Imaging and Radiotherapy, Faculdade de Medicina, Universidade Estadual Paulista, Campus de Botucatu, SP, Brazil.

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**Figure 2** Extraocular sebaceous carcinoma. Neoplasm characterized by polygonal clear cells, nuclear pleomorphism, cell debris, and frequent mitoses (Hematoxylin & eosin,  $\times 40$ ).

thus, the patient was referred to the Head and Neck Surgery Division of the institution.

In a review of 1349 SC cases, a predominance of males (54%) was observed, as well as mean age of 73 years, 86% whites, 38.7% on the palpebra, with a survival rate of 91.9%, and 79.2% in 5 and 10 years respectively.<sup>1</sup> The most frequent metastases were found in the lymph nodes.<sup>1,3</sup> Most cases occur *de novo*, although it may originate from benign sebaceous lesions and, when located in the upper or lower eyelid, it is associated with the Meibomian and Zeis glands.<sup>1-3</sup> Clinical presentation is variable; it is usually painless and slow-growing, but it can be rapid-growing and aggressive.<sup>2</sup>

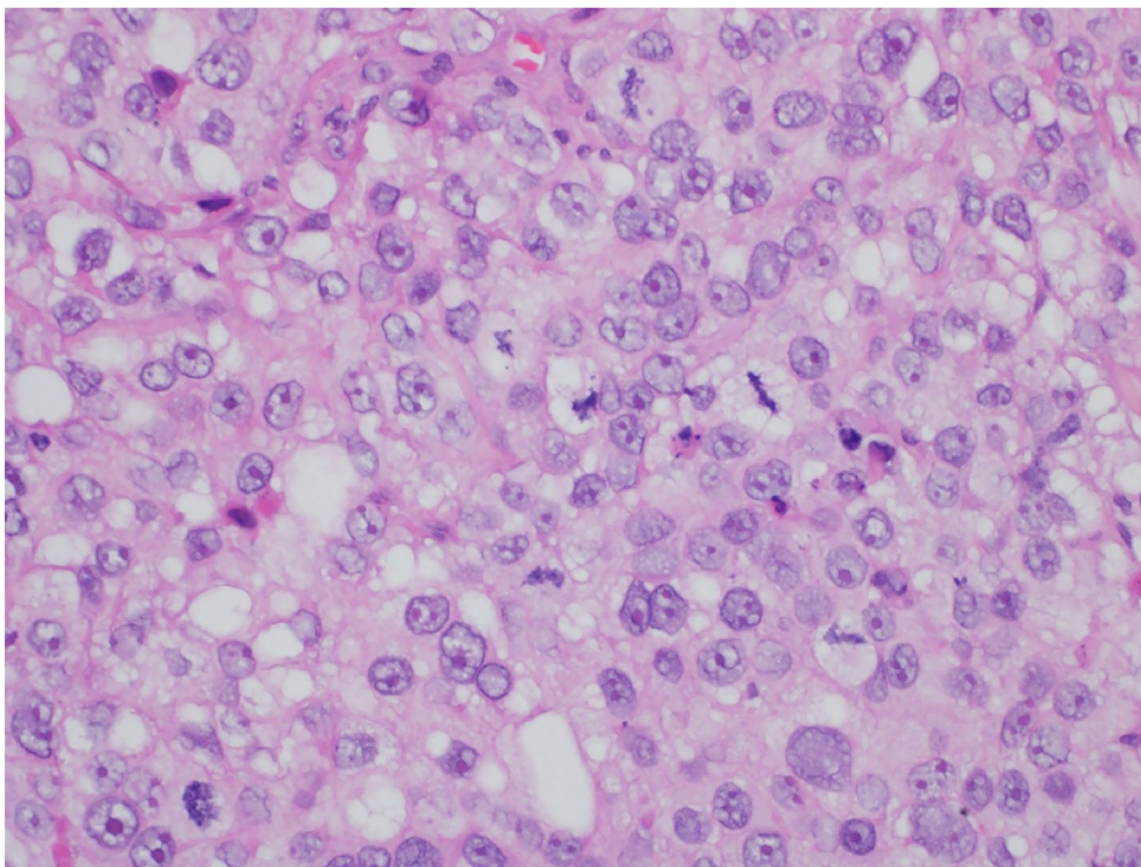
It is the third or fourth most frequent malignant neoplasm of the eyelids, depending on the reference.<sup>1-3</sup> The most frequent extraocular location is the cephalic segment, especially the face. The diagnosis of SC should be a warning sign, as it is a possible marker of Muir-Torre syndrome, a genodermatosis characterized by the presence of skin tumors of sebaceous origin associated with systemic malignancies, particularly of the gastrointestinal tract.<sup>1</sup> A subcutaneous nodule is usually observed in SC, which is normochromic;

however, it may disclose different morphologies, colors and behavior, depending on its place of origin.<sup>1-3</sup>

The differential diagnosis of extraocular SC includes basal cell carcinoma (BCC), SCC, amelanotic melanoma, Merkel cell carcinoma, and cutaneous lymphoma.<sup>1,3</sup> The immunohistochemical use of markers for BerEP4, EMA (negative in BCC), AE1 and AE3 (negative in melanoma, lymphomas), adipophilin (negative in SCC, Merkel), p53 and Ki-67, will aid in the diagnosis and prognosis.<sup>1-4</sup> The treatment comprises surgical resection with a 1-cm margin or the use of the micrographic surgical technique.<sup>2</sup> A therapeutic option in cases of metastatic SC to the lungs and CNS is immunotherapy with pembrolizumab, which belongs to the class of inhibitors of anti-PD1 immunological checkpoints (programmed death-1), and is also used in metastatic melanoma and Merkel cell carcinoma.<sup>5</sup>

This case report exemplifies a case of an atypical presentation of extraocular SC, especially due to the rapid growth and aggressiveness of the tumor, which when diagnosed had a specific satellite lesion and tumorous clinical aspect.





**Figure 3** Extraocular sebaceous carcinoma. Detail of neoplastic cells showing nuclear pleomorphism, prominent nucleoli, and multilobular cytoplasm (Hematoxylin & eosin,  $\times 400$ ).

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### Authors' contributions

Luana Moraes Campos: Approval of the final version of the manuscript; design and planning of the study; critical review of the literature.

Joana Alexandria Ferreira Dias: Approval of the final version of the manuscript; critical review of the manuscript.

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### Conflicts of interest

None declared.

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