

# Sclerodermiform Cell Epithelioma of the Palpebromalar Region

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**Summary:** This report describes a recurrent sclerodermiform basal cell epithelioma of the malar region next to the inferior eyelid in a 57-year-old woman. Three interventions were necessary to obtain a clear margin of resection. The area of resection was closed with a local cutaneous flap. We report a rare basal cell carcinoma subtype underestimated in its aggressiveness with often inadequate medical and surgical management. This tumor, generally localized in the face, often requires aggressive surgery, and aesthetic results can be poor. The patients require close long-term follow-up even when margins are clear. General practitioners, dermatologists, and surgeons should be aware of sclerodermiform basal cell carcinoma, which is a malignant, aggressive, and recurrent tumor. (*Plast Reconstr Surg Glob Open* 2024; 12:e5796; doi: [10.1097/GOX.00000000000005796](https://doi.org/10.1097/GOX.00000000000005796); Published online 3 May 2024.)

## INTRODUCTION

Basal cell carcinoma (BCC) is the most common tumor worldwide.<sup>1</sup> Three clinical subtypes (nodular, superficial, and sclerodermiform) and four histological subtypes (nodular, superficial, infiltrative, and sclerodermiform) are differentiated.<sup>2</sup> Its sclerodermiform subtype occurs in only 2% of skin cancers.<sup>3</sup> This aggressive subtype is responsible for numerous recurrences.

The clinical case that we present describes a patient referred to our department for a tumor that recurred 3 years after the first resection. We draw attention to the aggressive surgical management necessary to reach negative margins and the poor prognosis of these tumors.

## CLINICAL CASE

A 57-year-old White woman without relevant medical history was referred to our plastic surgery department for the recurrence of a sclerodermiform basal cell carcinoma (scBCC) of the right palpebromalar region. The initial resection consisted of a 22×10 mm skin patch with direct closure. The histology showed a resected scBCC, with less than 1 mm margins. No postoperative monitoring was carried out. Three years later, the patient noticed a change in the scar, and a biopsy confirmed recurrence.

Our clinical examination, during the first consultation, revealed an erythematous, poorly demarcated, crusting lesion measuring 15×5 mm on the old scar (Fig. 1). Surgery was performed and removed a region of 50×25 mm with circumferential margins of 10 mm around the lesion according to the recommendations of the French Society for Aesthetic and Reconstructive Surgery (Table 1).<sup>3</sup> Extemporaneous examination (EE) of the upper margin only was carried out given the proximity to the lower eyelid. Frozen section examination of this margin revealed no cancerous cells, and the defect was closed with a large nasolabial advancement flap (Fig. 1). The definitive anatomopathological analysis showed positive margins in the lower internal part of the resection. Therefore, an additional revision of 42×8 mm was performed without EE, and the flap was remobilized. Again, the lower and inner margins were positive on final pathological analysis, whereas the outer margin was negative but close to tumor cells. A surgical revision was again carried out, with 10 mm additional margins. The wound was left open pending the final pathological results. This time, the excision margins were healthy, and the flap was sutured into place (Fig. 2). These three interventions were performed over 3 months.

The patient's clinical situation was discussed at multidisciplinary oncology consultations, recommending close clinical follow-up without adjuvant treatment. No sign of recurrence has been observed during the 15-month postoperative period.

## DISCUSSION

### Clinical Features

Mainly located in the cervicofacial region, scBCCs are rare tumors with varied clinical presentation. They may

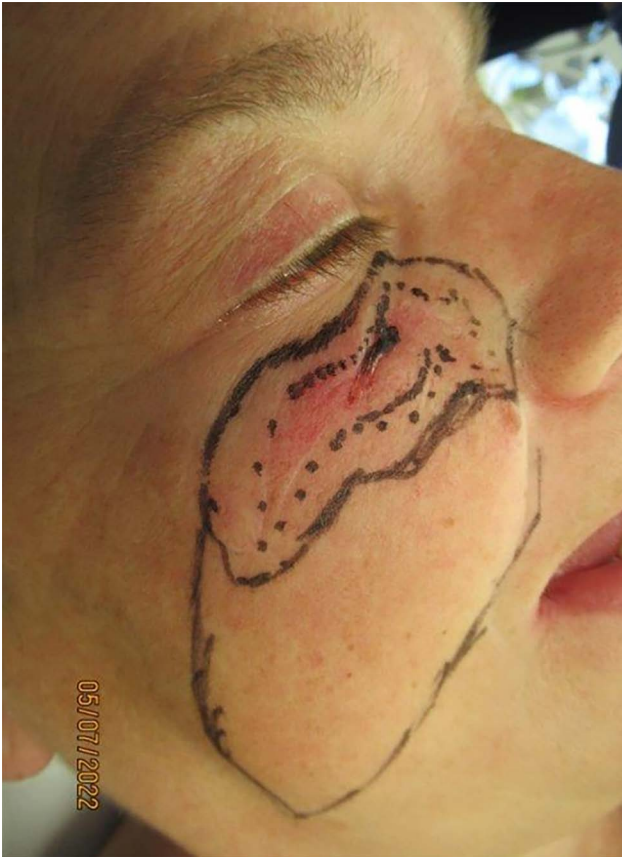
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**Fig. 1.** Photograph of recurrence of scleroderma BCC resected 3 years previously. The suspected lesion is marked with a dotted line. The resection area is indicated by a line with margins of 10 to 15 mm. Drawing of the preoperative advancement flap.

**Table 1. Margins of Recommendations from the French Society for Aesthetic and Reconstructive Surgery**

Tumor Diameter, mm	Resection Margin, mm
6	4
8	5
10	6
12	8
16	10
20	13

Data from Loddé et al.<sup>3</sup>

appear as flat, yellowish, ivory lesions, or in the form of induration with retraction of the borders mimicking a scar and are sometimes surrounded by epitheliomatous pearls.<sup>1,4</sup> The presence of telangiectasia and central ulcerations are indicative of advanced aggressiveness.<sup>4</sup>

Macroscopically, scBCCs are generally not clearly visible, with fuzzy borders; thus, they are usually diagnosed when locally advanced.<sup>1,3</sup> Dermoscopy or high-definition optical coherence tomography (HD OCT), are used to distinguish BCC subtypes.<sup>4,5</sup> High-resolution ultrasonographic imaging permits accurate observation of cutaneous structures to a 4 to 7cm depth. In this way, rapid and low-cost measurements of tumor axes and flow

### Takeaways

**Question:** What are the key points to know about the management of the scleroderma basal cell carcinoma?

**Findings:** The article shows through a clinical case the difficulties of the management of sclerodermiform basal cell carcinoma. A literature review makes it possible to learn about the surgical margins that must be adapted to the tumor size. We draw attention to the necessity for close and long-term follow-up.

**Meaning:** Sclerodermiform basal cell carcinoma is a rare and aggressive subtype of basal cell carcinomas requiring strictly surgical treatment with healthy resection margins to reduce the risk of recurrence.



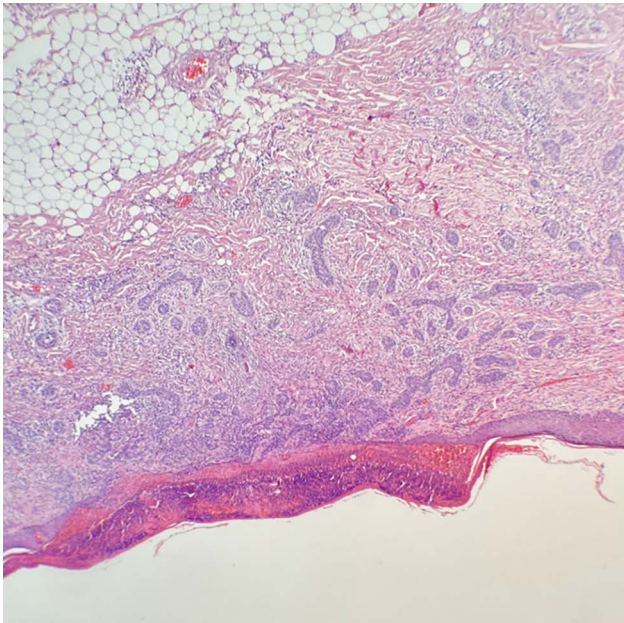
**Fig. 2.** Photograph of the wound 22 days after the last revision showing advancement of the flap with an extension of the latter under the mandible.

provide preoperative landmarks and reduce postoperative disfigurement.<sup>6</sup>

### Histological Features

Histologically, scBCC consists of an iceberg-shaped proliferation of intradermal keratinocytes (Fig. 3).<sup>1,7</sup> The tumor cells are arranged in small, anastomosed cords composed of two to three spindle-shaped cell layers (Fig. 4).<sup>2,7</sup> The nuclei are large, regular dark basophilic.<sup>1,7,8</sup> The stroma is characterized by intense sclerosis surrounding the carcinomatous elements.<sup>7</sup>





**Fig. 3.** Microscopic magnification 40× of the histopathologic specimen; note the proliferation of keratinocytes in carcinomatous trabeculae that extend like an iceberg in dermal sclerosis.

#### Treatment

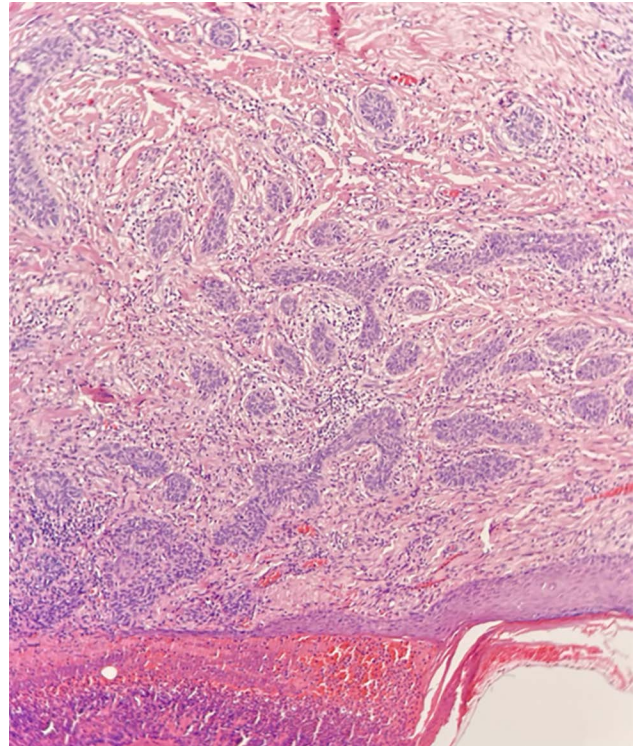
The treatment of scBCC is exclusively surgical. Local dermatologic treatments should be avoided. Radiotherapy is contraindicated because of resistance.<sup>2,3,9</sup> Circumferential safety margins must be increased due to scBCC development into an iceberg.<sup>3</sup> Breuninger and Undeutsch proposed that resection margins should be based on the diameter of the tumor lesion (Table 1).<sup>3</sup> The National Comprehensive Cancer Network offers regularly updated practical guides from evidence-based consensus. The current recommendation for treatment of scBCC is Mohs micrographic surgery with a definitive anatomicopathological examination.<sup>2,3,9</sup> EE and two-stage excision surgery have a higher recurrence rate given the infiltrative and multifocal nature of scBCC.<sup>2</sup>

If one surgical margin is positive, in contact with, or flush with the lesion, or if there is damage to the periosteum or perichondrium, reoperation must be performed.<sup>2,3</sup> Directed healing, direct closure, or total skin grafting can be done while waiting for the results.<sup>9</sup> Coverage by a local skin flap should be considered only in the case of negative margins or coverage of a cavity, or for aesthetic reasons.

The clinical case reported here illustrates the difficulties of complete removal of scBCC. Mohs micrographic surgery would have been the best option, but unfortunately, not feasible in our institution. EE associated with a two-stage surgery coverage, until negative margins were obtained, would have been more appropriate. Nevertheless, immediate flap coverage was the best choice from an aesthetic point of view and after discussion with the patient.

#### Follow-up

As aggressive tumors with possible deep extension, scBCCs have metastatic potential. The local recurrence



**Fig. 4.** Microscopic magnification 100× of the histopathological specimen; note that the tumor cells are arranged in small more or less anastomosed cords composed of two to three fusiform basal cell layers.

rate exceeds 20%, and 90% during the first 3 years.<sup>10</sup> Recommendations from the National Comprehensive Cancer Network advise lifelong surveillance.<sup>2,9</sup> If there is inflammation, induration, or the presence of a crust, a punch biopsy 3 mm deep at least is warranted.<sup>1,3</sup> A histological change during a recurrence is possible, and a nonaggressive primary BCC-type lesion recurrence with histology of scleroderma is observed in 57% of cases.<sup>10</sup>

### CONCLUSIONS

Rare and aggressive tumors, scBCCs have varied clinical presentation. Their limits are blurred and their development as an iceberg gives them a falsely reassuring appearance. The treatment is strictly surgical, often with poor aesthetic results. Mohs micrographic surgery is recommended. Long-term postoperative monitoring is essential due to the risk of recurrence. The poor prognosis requires rapid, specific care and in-depth knowledge of this entity by practitioners.

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#### DISCLOSURE

*The authors have no financial interest to declare in relation to the content of this article.*

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