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Histological and Surgical Features of Sclerodermiform Basal Cell Carcinoma

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Abstract | **Original Research Article**

Basal cell carcinoma scleroderma is a scarce form of basal cell carcinoma. It is special in its architecture and its aggressive evolution as well as high frequency of recidivism. Treatment remains primarily surgical with a centimeter of margin regardless of location. Micrographic surgery remains the only means of local control. The objective of our study was to assess the severity of this clinico-histological form on the local level and to determine the risk factors for tumor recurrence.

Keywords: Sclerodermiform basal cell carcinoma, morpheiform basal cell carcinoma, tumor of the face, excisional biopsy.

INTRODUCTION

Basal cell carcinoma sclerodermiform (CBCs) or morpheaform is an uncommon form of basal cell carcinoma (BCC). It is one of three histological subtypes of aggressive CBC (sclerodermiform, infiltrating and micronodular) [1]. Sclerodermiform CBC was originally described by Radcliffe-Crocker in 1895 [2]. Usually located in the face and neck, it appears as a yellowish or whitish infiltrated plaque, a smooth surface sometimes covered with a squama, or in evolved lesions, a depressed center, telangiectasia, with imprecise boundaries, slowly extensive. The lack of beaded or raised margins and the absence of ulceration and crust may simulate scarring [2, 3]. The positive diagnosis remains histological. Although these CBCs are considered to be benign tumors, they are unique in their invasive architecture and potentially aggressive evolution warranting radical surgical treatment [4]. The objective of our study was to evaluate the severity of this clinico-histological form at the local level and to determine the risk factors for tumor recurrence.

PATIENTS AND METHODS

This was a retrospective study of 31 patients (13 women, 18 men) with face and neck CBCs operated on between January 2013 and October 2018. The average age at surgery was 67 with extremes ranging from 34 to 80 years old.

The distribution of the lesions was as follows: 9 involved the nasal region, 7 the orbital region, 5 the frontotemporal region, 3 at the jugal level, 4 at the periauricular, 2 at the cervical level and one mandibular case. The delay between appearance and consultation was extremely variable, ranging from 6 months to 10 years. The average size of the lesions was 5.8 cm (1-15cm).

All the patients were taken in charge after histological confirmation of CBCs by biopsy or excisional biopsy. This punch or incisional biopsy was deep enough to include the reticular dermis to detect an infiltrative component and to specify at best the histological type.

The excision or recovery margins conformed to a centimeter or more margin of safety, the lesions were first delimited with a dermographic pen, then the margin of safety was achieved. All the excision was performed in a cold-knife way. The closure was carried out according to the size, the depth of the loss of substance and according to the therapeutic scale with means that allow local control. All the excisions were oriented with a marker wire placed at noon or on a cork. A clinical information sheet including a schema of the lesion and the surgical margins used was systematically completed and sent to the pathological anatomy laboratory. Macroscopically, the lesion is characterized by sclerosing, cicatricial, badly well circumscribed with

white or yellow color, which looks like scleroderma. The histopathological examination showed a neoplasm composed of malignant epithelial cells. Architecturally, the tumor was arranged in an angulated narrow tumor growing in an infiltrative manner at the leading edge of the tumor (Figure-1). It consists of small-sized basaloid cells with homogeneous hyperchromatic nuclei and scant cytoplasm. Mitotic figures were present within sclerotic stroma. The perineural and vascular invasion was found in 4 cases. Resection margins were healthy in all patients. Adjuvant treatment was not systematic. Depending on the localization and the size of the tumor, a local and loco-regional radiological assessment was carried out and according to the indication this assessment was completed by magnetic resonance imaging (MRI). The evolution was marked by recurrence in 5 patients and the appearance of an epidermoid contingent in 6 other patients.

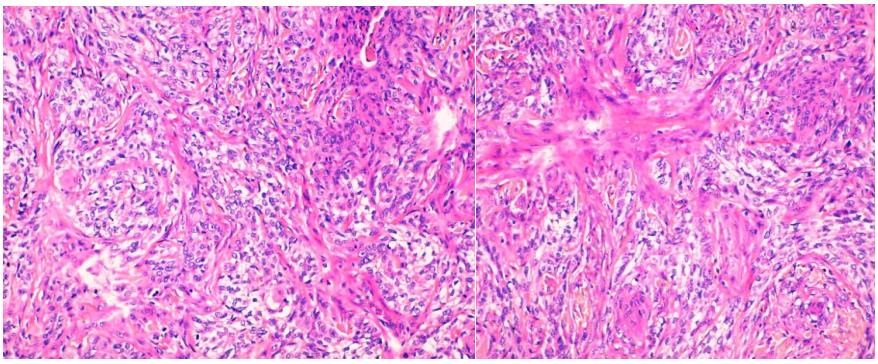

Fig-1: A+B: Histologically, the tumor was arranged in angulated narrow tumor nests growing in an infiltrative manner at the leading edge of the tumor

RESULTS

The average follow-up was 2.1 ± 2.8 (6 months -5) (Table-1). Five cases of recurrence were found in our patients at follow-up, 16.13% (two patients recurred twice despite surgical treatment and adjuvant radiotherapy).

These cases of recurrence were localized in the areas at risk: 3 cases at the internal canthal level and two cases at the nasal level. Two cases were lost sight of. The average time to recidivism was 2 years (1-3 and a half). We found three main and significant factors of the risk of recidivism:

- The location of the tumor
- Tumor Size
- The quality of initial surgical management
- The passage of a basal cell carcinoma nodular or others in sclerodermiform

The Repair of the loss of substance was a challenge for our surgical team in front of the localization (the face) and the size of the CBCs, we were able to follow in Directed cicatrization 9 patients, a case of direct suture, 9 cases of skin graft and 12 patients were covered with flap (Figure-2).

Table-: Different results

Number of patients	31
Average age (years)	67
Mean decline (years)	2
Recurrences (years)	5
Time to recurrence (years)	2

DISCUSSION

The Infiltrative / Morpheaform variants of basal cell carcinoma represents 10% of all BCC, including non-sclerosing and sclerosing histological variants with an infiltrating rather than expansible growth pattern, where long, thin strands of tumor cells deeply penetrate the collagen fascicles. The superficial layers of the tumor often have a solid growth pattern and the infiltrative type is present in the lower or proximal layers of the tumor. This variant of infiltrative BCC is characterized by an increased number of fibroblasts and the presence of fibrotic desmoplastic stroma, which gives the tumor a characteristic clinical picture of morphea or keloid scar.

Most studies on the epidemiological profile, the risk factors, and the efficiency of CBC treatments have to do mostly with low-risk CBCs and are not extrapolable to scleroderma-like CBCs [5]. Sclerodermiform CBCs are associated with an increasing risk of subclinical extension in several prospective and retrospective studies [6, 7]. The retrospective study of Breuninger found a complete excision rate for sclerodermiform CBCs of more than 95% at 13 to 15 mm while margins of 3 to 5 mm were at a rate between 66% and 82% [8]. Burg recommended a margin of more than 5 mm for Scleroderma CBCs due

to a subclinical extension significantly greater than that of nodular CBCs [9].

Fig-2: A patient operated on for recurrence of CBCs at internal canthal level

A: Recurrence on a medial frontal flap after a delay of two and a half years; **B:** The limits of exenteration; **C:** The exenteration cavity; **D** and **E:** The cover by an ipsilateral temporal muscle flap of the exenteration cavity; **F:** Result after thin skin graft

The concern of the use of the Dermoscope in the determination of the macroscopic margins was strongly defended and objectified in the nonrandomized study of Carducci [10].

The current recommendations of the scholarly societies are not consensual on the lateral margins to be respected, but what is recommended is margins of at least 10 mm for the primitive CBCs and the recidivants, by privileging the extemporaneous examination or the surgery in two times [11].

Concerning the deep margins, the recommendations are more unequivocal for margins carrying the hypodermis, except if there is deep invasion of the structures (to take away) the fascia, the cartilage (perichondrium) the periosteum or even the hard mother [3, 11-13]

The Mohs technique has been strongly recommended if the practical conditions of its implementation are met before the prospective and retrospective studies that compared the techniques leading the authors to conclude that CMM was the treatment of choice for CBCs with an aggressive histological subtype (sclerodermiform, micronodular, trabecular, infiltrating, metatypic), as well as recurrent CBC [14].

The studies do not mention the means of reconstruction in front of the complexity of the medical supervision: the local and general state, the quality of the resection, the structure of the surroundings, the neoadjuvant treatment and especially the concern for the need for a simple way to watch for a possible recurrence.

Conclusion

The treatment of CBCs often leads to severe mutilations especially at an advanced stage. Their evolutionary prognosis is unfavorable. Hence the question about the real malignancy of these tumors which depend exclusively on the quality of initial surgical supervision.

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