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A Case of Several Recurrences of an Advanced Centro-Facial Scleroderma Basal Cell Carcinoma: A Case Report

Abstract :

Basal cell carcinoma is an epithelial tumor developed at the expense of tissue epiderma. Scleroderma's variety is rare and more aggressive. We report a case of a scleroderma basal cell carcinoma of the upper white lip initially, evolving for 8 years of an 72 year-old woman, she was operated locally - for social reasons - on several occasions due to multiple recurrences and loco-regional extension of the tumor explained by non-respect of surgical margins, she was referred to our department by her surgeon with an iterative epistaxis, Computed tomography was performed showing an image of tumoral residue of the nostril cartilage with partial bone invasion. After a multi-disciplinary consultation meeting, a wide surgery by the plastic and the ENT surgeons was indicated in addition to a post Healing radiotherapy, and subsequent 3D reconstruction of the floor of the mouth and the nasal pyramid.

Keywords: Basal cell carcinoma - scleroderma - recurrence – metastasis.

INTRODUCTION

Basal cell carcinoma (BCC) is a slow-growing and mostly locally invasive tumour, its incidence is increasing worldwide [1]. Among BCCs, different histotypes with aggressive growth patterns can be distinguished, of which scleroderma form BCC (sdBCC) is one of the most important variant, presenting a higher risk of local invasiveness, perineural invasiveness and distant metastasis than subtypes with non-aggressive growth patterns, such as nodular (nBCC) and superficial BCC (sBCC), which show a significantly lower risk of such events, BCCs most often occurring de novo, localized only to the skin, never on mucosa membranes, and of local malignancy. scleroderma basal cell carcinoma is a rare variety often located near the orifices on the face. It evolves slowly in a centrifugal way and ends up ulcerating. The limits of the tumor are very difficult to specify. This kind of lesion can remain unrecognized for a long time and end up being very extensive and ulcerating. An extension to a mucosa is exceptionally rare or never seen, with only a few cases reported. We report an uncommon case of a centro-facial basal cell carcinoma of an 72-year-old woman.

CASE REPORT

An 72-year-old Mediterranean woman, of average socio-economic level, followed for scleroderma basal cell carcinoma initially in the white upper lip that recurred several times over a period of eight years, the patient refused to be operated on under general anesthesia for personal reasons, therefore the treating surgeon adapted to the patient's social circumstances, who underwent several non-oncological excisions confirmed by anatomopathological examinations. The anatomopathological examination returned in favor of a lymphatic metastasis of a basal cell carcinoma scleroderma.

The patient was referred to our consultation by her attending physician for a symptomatology made up of iterative epistaxis.



Figure 1: Patient admission photos

Computed tomography was performed, showing an image of:

- Tumoral residue of the nostril cartilage

- Partial bone invasion of the floor of the nasal fossae and the internal wall of the maxillary sinuses without cervical adenopathy

We sought the opinion of our ENT colleagues, they performed a rhinoscopy for a local extension assessment indicating a thickening of the distal part of the nasal septum, excision of the nasal pyramid, and inferior turbinates with a nasopharynx intact and a communication between the floor of the mouth and the nasal fossae, A shave biopsy was performed. Histologic examination revealed a new nostril recurrence

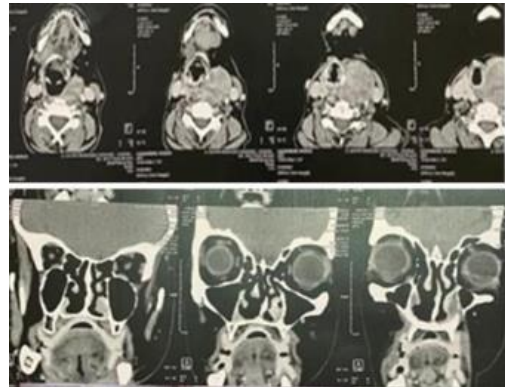


Figure 2: A facial CT scan showing an image of tumoral residue of the nostril cartilage with partial bone invasion of the floor of the nasal fossae and the internal wall of the maxillary sinuses without cervical adenopathy

After a multi-disciplinary consultation meeting with the resuscitators, the plastic surgeons, the ENT and the dental surgeon, the decision was to operate the patient with the widest cleanliness surgery possible in the presence of plastic surgeons, ENT specialists, and the dental surgeon to first take the patient's maxillary impression and then to refer the patient after regression of postoperative inflammation to oncologists for regular monitoring and radiotherapy.

Per-operatively the exeresis was wide made by both the ORL surgeons and plastic surgeons, it took all the endonasal environment, the maxillary sinus, the cartilage and the upper maxillary bone in totality, then a coverage by an X flap and closure in two planes, subcutaneous and cutaneous. With placement of a nasogastric tube and packing of the choanae. The dental surgeon was present and took the maxillary print for further reconstruction.



Figure 3: Per-operative photos with the nasal print on the second one

The anatomopathological was in favor of an osteo-cartilaginous tissue with tumoral mucosa, muscle and subcutaneous tissue showing a scleroderma basal cell carcinoma, as well as tumoral bone resections.

The immediate postoperative follow-up was marked by a rejection of the shutter, the patient didn't tolerate it, we recontacted the dental surgeon who gave her an appointment after leaving our department bringing with her post-operative facial CT scan with 3D cross-sections.

During her fifteen days of hospitalization, the patient received local and general care, a pure liquid diet, and biological check-ups. She was declared discharged with an appointment at our consultation for a regular follow-up, an appointment with a speech therapist, his attending dental surgeon and the radiotherapy department.

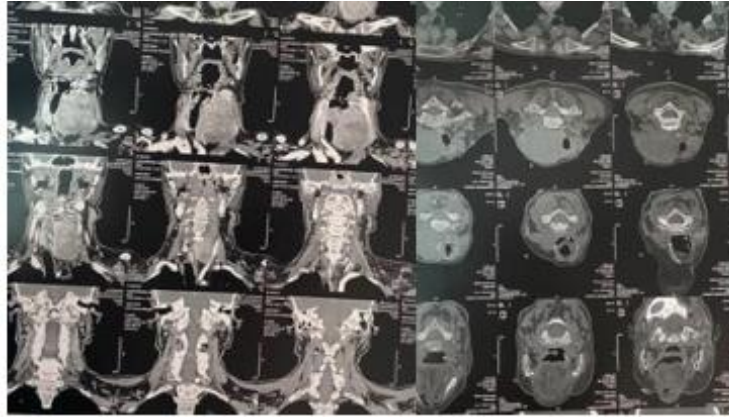


Figure 4: Cervio-facial CT scan showing thickening of the mucosa of the right maxillary sinus with a small residue at the level of the upper orifice of the left lacrimo-nasal duct and behind the proper bone of the left nose without cervical lymphadenopathy or a pulmonary metastasis

The oncologists were in favor of a local postoperative radiotherapy after healing and reduction of inflammation, they also requested a cervico-facial and thoracic CT scan.

In this sense, a CT scan was performed showing a thickening of the mucosa of the right maxillary sinus with a small residue at the level of the upper orifice of the left lacrimo-nasal duct and behind the proper bone of the left nose without cervical lymphadenopathy or a pulmonary metastasis

Currently the patient is followed in our training for control and local care, she is satisfied, she presents a symptomatology made of epiphora accentuated on the right side explained by the resection of the tear duct on this side and also in oncology for a radiotherapy cure, she is awaiting maxillary and nasal reconstruction at the end of her cure.



Figure 5: Photos taken 25 days post-operative



Figure 6: Photos taken three month days post-operative

DISCUSSION

Basal cell carcinoma of the skin is the most common malignancy in the head and neck area. Regional and distant metastases rarely occur with this type of tumour. Advanced BCC are defined as tumors of stage III and IV. Often these tumors develop over many years but are neglected by patients and relatives [2].

Advanced BCC are defined as tumors of stage III and IV. Often these tumors develop over many years but are neglected by patients and relatives [2]. There is an overlap of high-risk BCC and advanced BCC. High-risk BCC are defined as tumors of long duration, located in mid-face or on ears, diameter > 2 cm, aggressive histopathologic subtype, with perivascular or perineural infiltration, history of radiation exposure, or previous treatment failure [3, 4].

The majority of advanced BCC belong to stage III. If these tumors are 5 cm in diameter or larger they are called giant BCCs. The overall cure rate drops to about 60% with 40% of patients developing recurrences or metastatic spread within 2 years of follow-up [5]. The first treatment option is surgery including Mohs or micrographically controlled procedures.

This case allows us first of all to understand the local invasion and the gravity of this tumor, and especially the interest of an early consultation and the respect of the margins. It also highlights the importance of obtaining adequate tissue for histologic evaluation, as partial biopsies can lead to confusion. In this case, immunohistochemistry was consistent with an adnexal tumor. However, the H&E morphology rather than immunoprofile distinguished the malignant tumor from its benign counterpart. Long-term follow-up is still recommended because of the risk of local recurrence and invasion.

The particularity of our patient is that she refused surgery under general anesthesia, which made the respect of the margins impossible each time, after eight years of evolution the patient found herself faced with a submaxillary lymphatic metastasis but also a local invasion reaching the muscles, mucous membrane and also bone in whom the excision could not be complete, we reached the base of the skull during the gesture.

CONCLUSION

Advanced BCC has a presentation and course that is more aggressive than that seen in the majority of stage I and stage II BCCs. Surgery and radiotherapy are the cornerstone of therapeutic management, long-term follow-up is required. Prevention and patient awareness remains the most effective way to avoid any unmanageable development.

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