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RESEARCH ARTICLE

A CASE OF SEVERAL RECURRENCES OF AN ADVANCED CENTRO-FACIAL SCLERODERMA BASAL CELL CARCINOMA: A CASE REPORT

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Abstract

Basal cellcarcinomais an epithelialtumordeveloped at the expense of tissue epiderma. Scleroderma'svarietyis rare and more agressive. We report a case of a scleroderma basal cellcarcinoma of the upper white lipinitially, evolving for 8 years of an 72 year-oldwoman, shewasoperatedlocally - for social reasons - on several occasions due to multiple recurrences and loco-regional extension of the tumorexplained by non-respect of surgicalmargins, shewasrefered to ourdepartment by her surgeon with an iterativeepistaxis, Computedtomographywasperformedshowing an image of tumoral residue of the nostril cartilage with partial bone invasion. After a multi-disciplinary consultation meeting, a widesurgery by the plastic and the ENT surgeons wasindicated in addition to a post Healing radiotherapy, and subsequent 3D reconstruction of the floor of the mouth and the nasal pyramid.

Introduction:-

Basal cellcarcinoma (BCC) is a slow-growing and mostlylocally invasive tumour, its incidence isincreasingworldwide [1]. AmongBCCs, differenthistotypeswithaggressivegrowth patterns canbedistinguished, of whichsclerodermaform BCC (sdBCC) is one of the most important variant, presenting a higherrisk of local invasiveness, perineuralinvasiveness and distant metastasisthansubtypeswith non-aggressivegrowth patterns, such as nodular (nBCC) and superficial BCC (sBCC), which show a significantlylowerrisk of suchevents, BCCsmostoftenoccurring de novo, localizedonly to the skin, never on mucosa membranes, and of local malignancy. scleroderma basal cellcarcinomais a rare varietyoftenlocatednear the orifices on the face. It evolvesslowly in a centrifugalway and ends up ulcerating. The limits of the tumor are verydifficult to specify. This kind of lesioncanremainunrecognized for a long time and end up beingvery extensive and ulcerating. An extension to a mucosaisexceptionally rare or neverseen, withonly a few cases reported. We report an uncommon case of a centro-facial basal cellcarcinoma of an 72-year-old woman.

Case Report

An 72-year-old Mediterraneanwoman, of averagesocio-economiclevel, followed for scleroderma basal cellcarcinomainitially in the white upperlipthatrecurredseveral times over a period of eightyears, the patient refused to beoperated on undergeneralanesthesia for personalreasons, therefore the treating surgeon adapted to the patient's social circumstances, whounderwentseveral non-oncological excisions confirmed by

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anatomopathologicalexaminations. The anatomopathologicalexaminationreturned in favor of alymphaticmetastasis of a basal cellcarcinomascleroderma.

The patient wasreferred to our consultation by herattendingphysician for a symptomatology made up of iterativeepistaxis.

Figure 1:- Patient admission photos.

Computedtomographywasperformed, showing an image of:
- Tumoral residue of the nostril cartilage
- Partial bone invasion of the floor of the nasal

fossae and the internalwall of the maxillarysinuses
without cervical adenopathy

Wesought the opinion of our ENT colleagues, theyperformed a rhinoscopy for a local extension assessmentindicating a thickening of the distal part of the nasal septum, excision of the nasal pyramid, and inferiorturbinateswith a nasopharynx intact and a communication between the floor of the mouth and the nasal fossae, A shavebiopsywasperformed. Histologicexaminationrevealed a new nostrilrecurrence

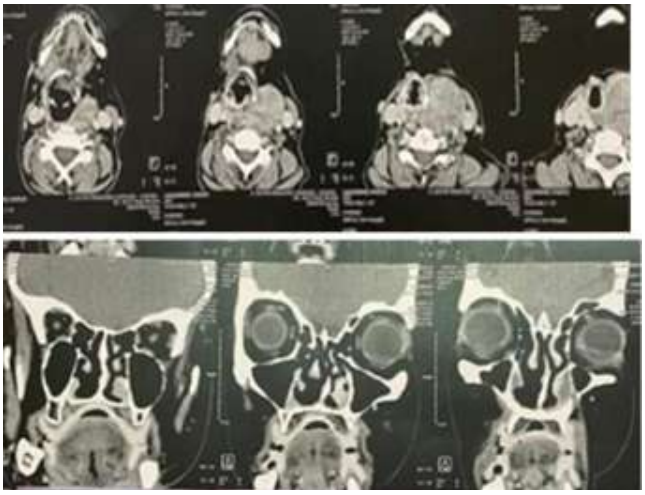

Figure 2:- A facial CT scan showing an image of tumoral residue of the nostril cartilage with partial bone invasion of the floor of the nasal fossae and the internalwall of the maxillarysinuseswithout cervical adenopathy.

After a multi-disciplinary consultation meeting with the resuscitators, the plastic surgeons, the ENT and the dental surgeon, the decisionwas to operate the patient with the widestcleanlinesssurgery possible in the presence of plastic surgeons, ENT specialists, and the dental surgeon to first take the patient'smaxillary impression and then to refer the patient afterregression of postoperative inflammation to oncologists for regular monitoring and radiotherapy.

Per-operatively the exeresiswaswide made by both the ORL surgeons and plastic surgeons, ittook all the endonasalenvironment, the maxillary sinus, the cartilage and the uppermaxillarybone in totality, then a coverage by

an X flap and closure in two planes, subcutaneous and cutaneous. With placement of anasogastric tube and packing of the choanae. The dental surgeon waspresent and took the maxillaryprint for further reconstruction.

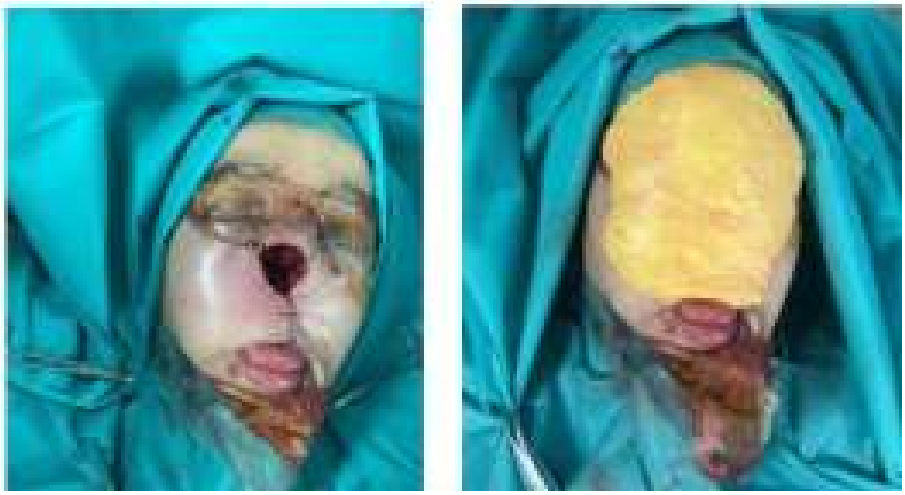

Figure 3:- Per-operative photos with the nasal print on the second one.

The anatomopathologicalwas in favor of an osteo-cartilaginous tissue with tumoral mucosa, muscle and subcutaneous tissue showing a scleroderma basal cellcarcinoma, as well as tumoral boneresections.

The immediatepostoperativefollow-up wasmarked by a rejection of the shutter, the patient didn'ttolerateit, werecontacted the dental surgeon who gave her an appointmentafterleavingourdepartmentbringingwithher post-operative facial CT scan with 3D cross-sections.

Duringherfifteendays of hospitalization, the patient received local and general care, a pure liquiddiet, and biologicalcheck-ups. Shewasdeclareddischargedwith an appointment at our consultation for a regularfollow-up, an appointmentwith a speech therapist, hisattending dental surgeon and the radiotherapydepartment.

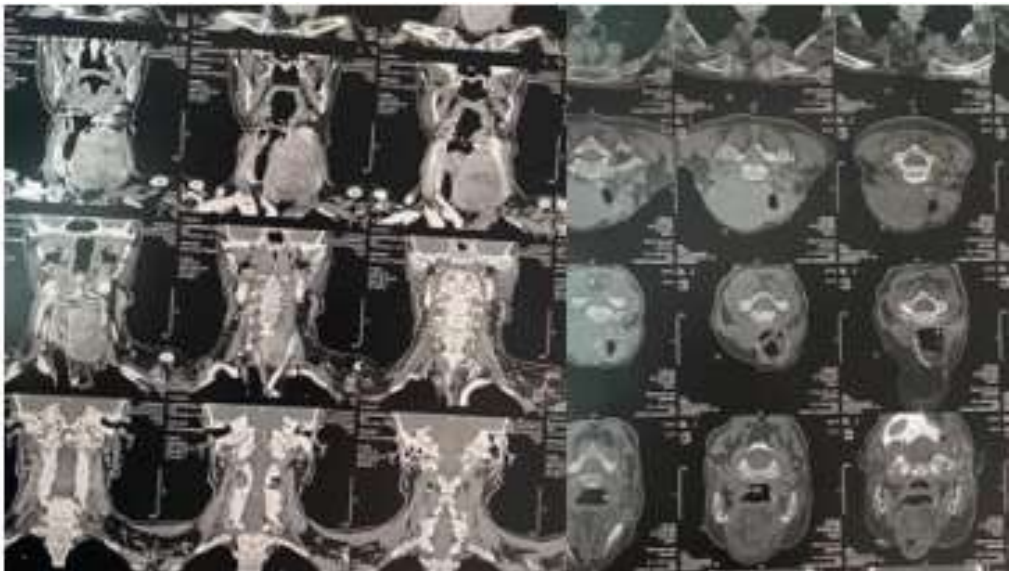

Figure 4:- Cervio-faciall CT scan showingthickening of the mucosa of the right maxillary sinus with a smallresidue at the level of the upper orifice of the left lacrymo-nasal duct and behind the properbone of the leftnosewithout cervical lymphadenopathy or a pulmonarymetastasis.

The oncologistswere in favor of a local postoperativeradiotherapyafterhealing and reduction of inflammation, theyalsorequested a cervico-facial and thoracic CT scan.

In thissense, a CT scan wasperformedshowing a thickening of the mucosa of the right maxillary sinus with a smallresidue at the level of the upper orifice of the left lacrymo-nasal duct and behind the properbone of the leftnosewithout cervical lymphadenopathy or a pulmonarymetastasis

Currently the patient isfollowed in our training for control and local care, sheissatisfied, shepresents a symptomatology made of epiphoraaccentuated on the right sideexplained by the resection of the tearduct on thisside and also in oncology for a radiotherapy cure, sheisawaitingmaxillary and nasal reconstruction at the end of her cure.

Figure 5: Photos taken 25 days post-operative

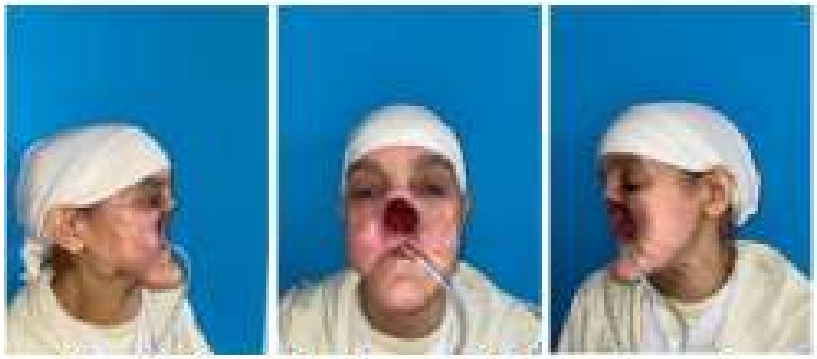

Figure 6: Photos taken three month days post-operative

Discussion:-

Basal cellcarcinoma of the skin is the mostcommonmalignancy in the head and neck area. Regiona and distant metastasesrarelyoccurwiththis type of tumour. Advanced BCC are defined as tumors of stage III and IV. Oftenthesetumorsdevelop over manyyears but are neglected by patients and relatives [2].

Advanced BCC are defined as tumors of stage III and IV. Oftenthesetumorsdevelop over manyyears but are neglected by patients and relatives [2]. There is an over-lap of high-risk BCC and advanced BCC. Highrisk BCC are defined as tumors of long duration, located in mid-face or on ears, diameter> 2 cm, aggressivehistopathologicsubtype, withperivascular or perineural infiltration, history of radiation exposure, or previoustreatmentfailure [3, 4].

The majority of advanced BCC belong to stage III. If thesetumors are 5 cm in diameter or largerthey are calledgiantBBCs. The overall cure rate drops to about 60 with 40% of patients developingrecurrences or metastatic spread within 2 years of follow-up [5]. The first treatment option issurgeryincludingMohs or micrographicallycontrolledprocedures.

This case allows us first of all to understand the local invasion and the gravity of thistumor, and especially the interest of an early consultation and the respect of the margins. It alsohighlights the importance of obtainingadequate tissue for histologicevaluation, as partial biopsies can lead to confusion. In this case, immunohistochemistrywas consistent with an adnexaltumor. However, the H&E morphologyratherthanimmunoprofiledistinguished the malignanttumorfromitsbenigncounterpart. Long-termfollow-up isstillrecommendedbecause of the risk of local recurrence and invasion.

The particularity of our patient isthatsherefusedsurgeryundergeneralanesthesia, which made the respect of the margins impossible each time, aftereightyears of evolution the patient foundherselffacedwith a submaxillarylymphaticmetastasis but also a local invasion reaching the muscles, mucous membrane and alsobony in whom the excision could not becomplete, wereached the base of the skullduring the gesture.

Conclusion:-

Advanced BCC has a presentation and course thatis more aggressivethanthatseen in the majority of stage I and stage II BCCs. Surgery and radiotherapy are the cornerstone of therapeuticmanagement., long-termfollow-up isrequired. Prevention and patient awarenessremains the most effective way to avoidanyunmanageabledevelopment.

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