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# Sebaceous carcinoma

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# SEBACEOUS CARCINOMA

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**Abstract**
Chronic non-healing ulcers of the skin are a manifestation of many different pathologic processes, caused by local and/ or systemic factors. The causes range from vascular disease and diabetes to foreign bodies, malignancy, infections, and unknown causes. Basal cell carcinoma and squamous cell carcinoma are the most commonly encountered in the orofacial region. Sebaceous carcinoma is a rare and potentially aggressive tumor that rarely occurs in the face and scalp with a non-specific clinical presentation.
In the present paper, we describe a case of sebaceous carcinoma in a 62-year-old male patient on the right side of the face, evolving as a non-healing ulcer since six months. The etiology of the present case seems to be multifactorial; the clinical history included a trauma, ultraviolet radiation and medications intake.

**Keywords: Extra-ocular – face - non-healing ulcers - sebaceous carcinoma.**

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# CARCINOME SÉBACÉ

**Résumé**
Les ulcères chroniques de la peau sont une manifestation de nombreux processus pathologiques, causés par des facteurs locaux et/ou systémiques. Les causes englobent des maladies vasculaires, le diabète, les réactions à des corps étrangers, un processus malin ou infectueux. Le carcinome basocellulaire et le carcinome spinocellulaire sont les plus couramment rencontrés au niveau de la région orofaciale. Le carcinome sébacé est une tumeur rare et potentiellement agressive qui survient rarement au niveau du visage et du cuir chevelu avec un tableau clinique non spécifique. Dans le présent article, nous décrivons un cas de carcinome sébacé chez un homme de 62 ans, évoluant au niveau de l'hémiface droite sous forme d'un ulcère depuis six mois.
L'étiologie de la présente tumeur semble être multifactorielle; l'histoire clinique comprenait un apport traumatisme, un rayonnement ultra-violet et des médicaments.

**Mots-clés: ulcère chronique - carcinome sébacé – tumeur.**

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## Introduction

Sebaceous carcinoma is an aggressive, rare, cutaneous tumor first well-described by Allaire in 1891 [1]. The knowledge about the incidence, risk factors, and prognosis of this disease is based on isolated case reports of <200 patients [2].

Sebaceous carcinoma has been separated into two anatomic groups: ocular and extra-ocular. The ocular tumors most commonly originate from the meibomian glands (51%) and glands of Zeis (10%) on the eyelid [3, 4]. The extra-ocular lesions account for 20% of the sebaceous carcinomas and the majority are found in the head and neck region of elderly patients, including the scalp and face. Infrequently, sebaceous carcinoma may be found on the trunk, genital region, extremities, and oral cavity [5 - 7].

The cause of sebaceous gland carcinoma remains unclear. Females are more commonly affected than males and patients are usually in the seventh decade. The clinical presentation of this tumor is often inconsistent, and is not initially diagnosed [8 - 11].

Ocular sebaceous carcinomas present as slowly enlarging, painless nodules of the eyelid margins or conjunctiva that may be clinically mistaken for benign inflammatory lesions, such as chalazions, blepharitis, and conjunctivitis [12 - 15]. The extra-ocular lesions often present as larger, ulcerated, yellow-tan nodules and measure 1– 4 cm or more in diameter [16].

Reported risk factors for sebaceous carcinoma include advanced age [17], racial predisposition (Asian or South Asian race) [18], females [19], irradiation to the head and neck [20], and a genetic predisposition such as in Muir-Torre syndrome [21] or possibly familial retinoblastoma [22].

Sebaceous gland carcinoma is a destructive tumor, with a tendency for both local recurrence and distant metastasis. Standard surgical resection with wide margins and Mohs micrographic surgery are common treatments for sebaceous carcinoma [23].

Sebaceous carcinoma is commonly seen in females and usually involves the eyelids. In the present paper, we report a case of extra-ocular sebaceous carcinoma on the right side of the cheek in a 62-year-old male patient for its unusual presentation along with an added emphasis on its review.

## Case report

A 62-year-old, Indian male, a farmer by occupation, reported to the Department of Oral Medicine and Radiology with the chief complaint of deep non-healing ulcerative lesions over the right mid region of the face, evolving in the last six months and swelling of four months duration. The patient's anamnesis revealed a history of trauma to the right cheek from a wooden object; the trauma caused erosion that evolved into ulcers around the wound. The wound and ulcers failed to heal and gradually expanded. The patient also consulted a local doctor for the same injury. He was prescribed oral and topical medications but no relief was felt to treatment. The rest of the physical examination was normal. Patient was a known case of hypertension for last 10 years and was under treatment (Lasix tablet, 40 mg per day).

### Clinical examination

Extra-oral examination revealed two ulcers of about 3x2 cm in size with erythematous floor seen at the junction of the cheek and the nose with mild serous exudation from both. The edges were raised and the ulcers were found fixed to the underlying structures.

The ulcers were surrounded by a diffuse mild erythematous swelling measuring approximately 3x4 cm in size involving the right mid-face region, extending medio-laterally from the ala of the nose to the zygomatic bone; supero-inferiorly, they extend from the lower eyelid till 1 cm below the ala, and elevating it. The swelling was erythematous and had an irregular surface with diffuse borders. Surrounding areas appeared normal.

Other findings included a nodule of about 4x3 cm in size over the lower eyelid and multiple, flat to elevated erythematous plaques and patches with focal crusting of varying sized over the cheek (Fig. 1). On palpation, the swelling and the nodule were hard in consistency, and non-tender. No lymphadenopathy was detected.

Intraoral examination revealed obliteration of the right side of the vestibule in the region of second premolar and first molar in the first quadrant of the oral cavity (Fig. 2). The overlying mucosa was not fixed. On palpation, the swelling was firm, non-tender, non-fluctuant, and non-compressible.

Other dental findings included completely edentulous maxillary arch, missing mandibular anterior teeth, and right and left first premolars with moderate amount of stains and calculus.

Based on the history and clinical examination a provisional clinical diagnosis of chronic granulomatous infection due to foreign body was made.

Basal cell carcinoma, squamous cell carcinoma, chronic suppurative osteomyelitis secondary to trauma, deep fungal infections, and metastatic carcinoma of the skin were considered as differential diagnosis.

The patient was subjected to the following investigations to reach a probable diagnosis: KOH test and acid-fast bacillus were negative for organisms. Gram staining of biopsy specimens from the right cheek was positive for Cocci in clusters, consistent with Staphylococcus aureus. A complete hemogram was performed and all the values were in the normal range.

The patient underwent radiographic examination. Panoramic radiograph revealed slight opacification of right maxillary sinus. Other dental findings noted were completely edentulous maxillary arch, missing mandibular anterior teeth, right and left premolars (Fig. 3). Paranasal sinus view revealed well-defined radio-opaque

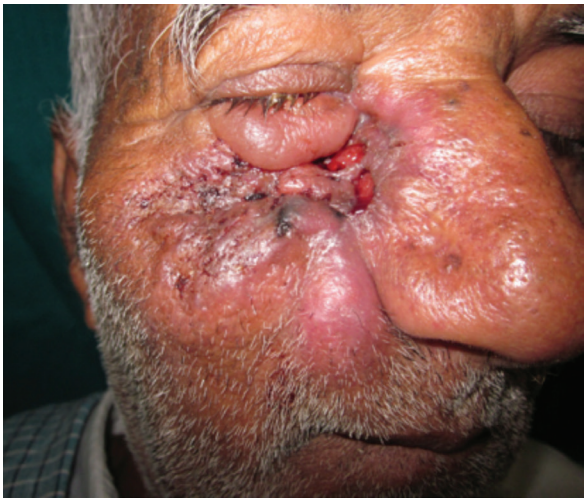

Fig. 1: Extra oral photograph of the patient with the lesion on the right side of the cheek.

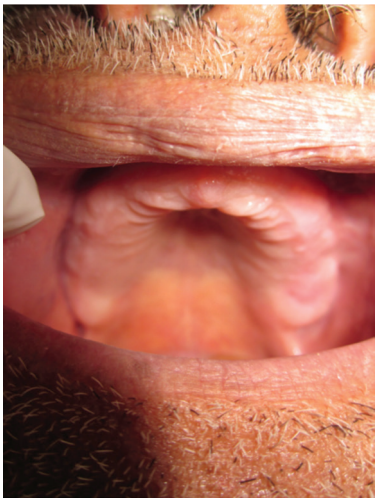

Fig. 2: Intraoral photograph showing obliteration of the buccal vestibule in the region of teeth #15 and 16.

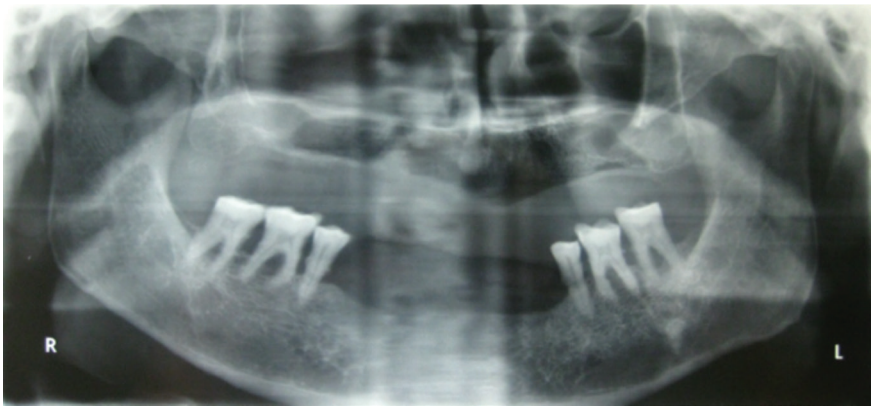


Fig. 3: Panoramic radiograph revealing slight opacification of right maxillary sinus.

shadow occupying right maxillary sinus (Fig. 4).

CT scan of maxilla revealed complete opacification of the right maxillary sinus. There was an ill-defined hyperdense soft tissue lesion, measuring approximately 5.8 x 2.7 cm, in the anterior aspect of the right maxillary antrum, extending anteriorly into the buccal region and infiltrating the skin. The lesion extended into the inferomedial aspect of the orbit, abutting the globe. The lesion involved the medial rectus and the inferior oblique muscles.

On contrast study, there was heterogenous enhancement of the lesion. An erosion of the anterior, medial, postero-lateral walls and roof of the right maxillary sinus, the maxilla and the nasal process of the maxilla on the right side (Figs. 5 and 6). There was no evidence of extension into the infratemporal fossa and the ethmoidal sinus on the right side or even an intracranial extension.

Based on the extent of destruction, the CT diagnosis (combined with other radiographs) of a neoplastic lesion was considered.

The lesion was then subjected for incisional biopsy and the biopsy specimen was taken from the right infraorbital region.

Histopathology revealed skin epithelium with pilosebaceous follicle showing solid sheets, cords and nests of clear cells with interspersed connective tissue stroma. These islands were lined by outer layer of columnar cells with clear cells in the centre. Few tumour giant cells were seen in within these proliferating islands along with thick bundles of collagen fibres with proliferating plump to spindle shaped fibroblasts in streaming pattern. Few lymphocytes were seen in connective tissue septa (Fig. 7). Histologically, the lesion was diagnosed as sebaceous carcinoma.

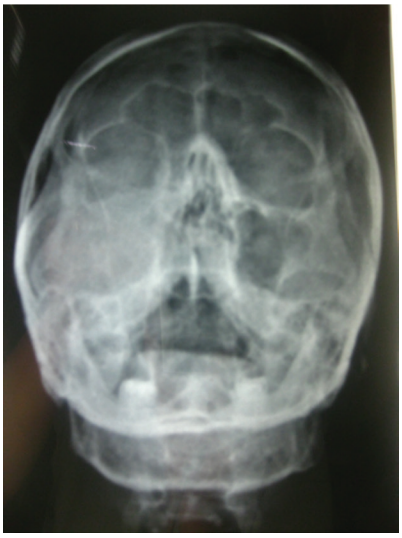

Fig. 4: PNS radiograph revealing well defined radio-opaque shadow occupying the right maxillary sinus.

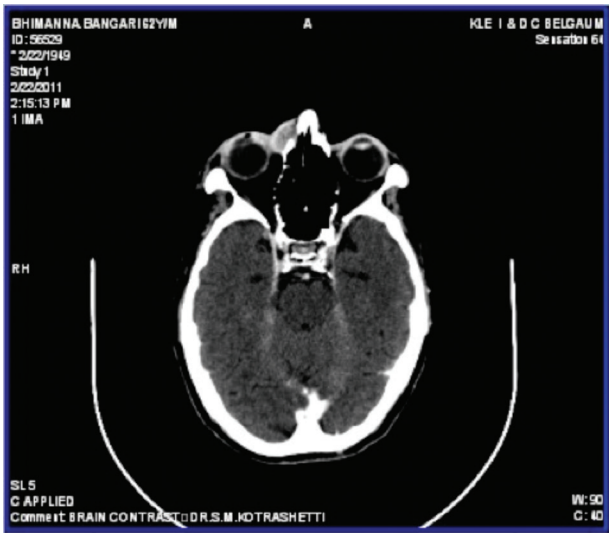


Fig. 5: CT scan revealing complete opacification of the right maxillary sinus.

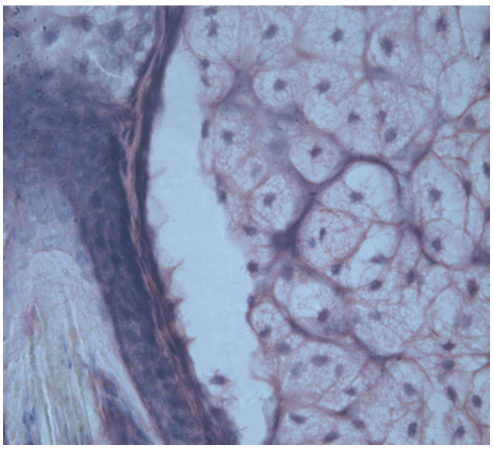

Fig. 6: Photomicrograph of the lesion (10 X).

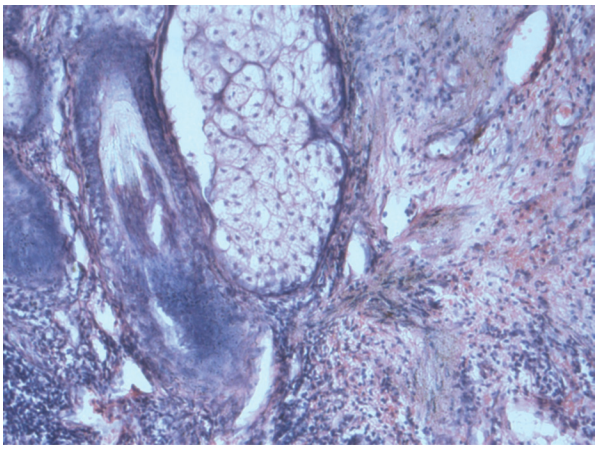

Fig. 7: Photomicrograph of the lesion (40X).

Subsequent to histological diagnosis, surgical excision with wide margins and skin grafting was planned for the patient. In his next visit, the patient was also scheduled for a biopsy of a nodule present over the right lower eyelid and referral to ophthalmologists, dermatologists and gastroenterologists for the opinion but he failed to report back for further investigations and treatment.

## Discussion

Many different medical conditions can cause chronic non-healing ulcers of the skin. The conditions range from vascular disease and diabetes to foreign bodies, autoimmune diseases, malignancies, infections (mostly due to resistant bacteria) and unknown causes.

Cutaneous extra-ocular sebaceous carcinoma is a rare and potentially aggressive tumor. The etiology of this tumor is still not clear. Its occurrence in actinic keratosis indicates a possible role for ultraviolet radiation in the etiopathogenesis of the extra-ocular type. In a series of 20 patients with sebaceous gland carcinoma, 8 patients had a history of diuretic use, and a possible association was suggested [24].

Sebaceous gland carcinoma seems to be more common in Asian populations than the other populations, and involvement of human papillomavirus (HPV) has been suggested as a possible etiologic factor in these populations [25]. Genetic factors clearly play a role, because sebaceous gland carcinomas are part of the genodermatosis Muir-Torre syndrome.

In our case, the patient presented with the history of trauma. He had also hypertension and was under medications. The patient did not have the prescriptions or previous reports. He was a farmer by occupation and so was exposed to ultraviolent radia-

tion. So the etiology in the present case could be multifactorial like age, trauma, ultraviolet radiation or some medications.

Most affected patients are in their sixth or seventh decade of life and women tend to be affected somewhat more often than men, with 57-77% of patients being women in several large series [8 – 11] unlike in the present case where the patient was a 62-year-old male.

The most difficult aspect of sebaceous carcinoma is its multitude of clinical presentations, which delay diagnosis by 1 to 2.9 years and may increase mortality [26]. Since sebaceous carcinoma presents most commonly as a gradually enlarging, firm nodule, it can mimic more common ophthalmologic or dermatologic conditions. Definitive diagnosis should be only based on histopathology [27].

Sebaceous gland carcinoma is a destructive tumor, with a tendency for both local recurrence and distant metastasis. Approximately 10% of the ocular sebaceous carcinomas are multicentric [28], and they often have direct extension or intraepithelial (pagetoid) spread to adjacent organs (lacrimal drainage system and nasopharynx). There is local tumor recurrence in 40% of the affected patients, lymph node metastasis (preauricular and cervical nodes) in 25-33% of cases, and a 5-year tumor-related mortality rate of 22% [29].

Extra-ocular sebaceous carcinoma and smaller tumor sizes are associated with lower metastatic potential and, consequently, lower mortality [30, 31].

Standard surgical resection with wide margins and Mohs micrographic surgery are common treatments for sebaceous carcinoma [32]. Radiation and topical and systemic therapies also have been suggested as useful treatment of the sebaceous carcinoma, although knowledge on their effect is limited. It has been suggested, but not studied, that topical chemotherapy to the involved conjunctivae following surgical excision of the invasive tumor may be beneficial [33].

Referral to an internist and gastroenterologist is warranted in patients diagnosed with sebaceous carcinoma in order to evaluate for the presence of internal (internist) and bowel (gastroenterologist) lesions associated with Muir-Torre syndrome.

Reported clinicopathological features associated with a poor prognosis include orbital invasion, upper and lower eyelid involvement, poor differentiation [23], lacrimal gland involvement [35], tumor diameter greater than 10 mm [36], pagetoid spread, and symptom duration greater than 6 months.

Sebaceous carcinoma in our patient extended to involve orbit, medial rectus muscles, inferior oblique muscles, lower eyelid, and pagetoid spread which was confirmed by CT scan and we acquitted the patient about the diagnosis. Considering the aggressiveness of the lesion, we advised prompt treatment for the patient but he failed to report back for the treatment.

## Conclusion

Sebaceous carcinoma is a rare and aggressive malignant tumor, especially in the mid region of the face. Due to its presentation, it can be misdiagnosed. A proper clinical examination, adequate imaging, diagnosis and treatment are necessary to improve the quality of life of the patient.

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