Sebaceous carcinoma: A rare cutaneous tumor

S. Dhambri, N. Halila, M. Dhaha, S. Kedous, S. Jebali, S.Touati, S. Gritli

Otorhinolaryngology-Head and Neck Surgery Department, Salah Azaiez Institute of Oncology, Tunis, Tunisia

ABSTRACT

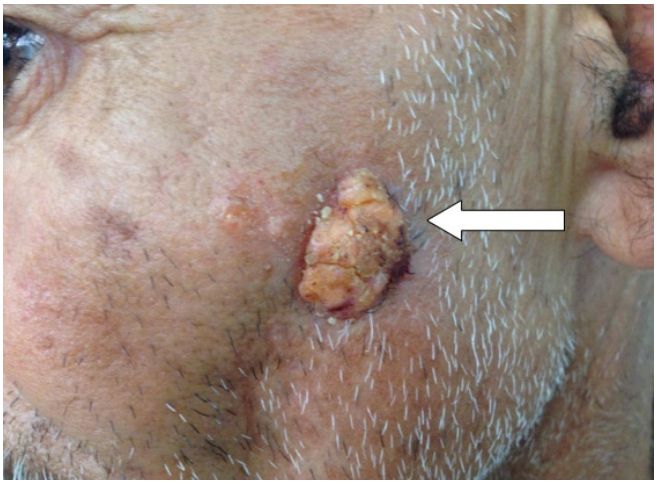
INTRODUCTION: Cutaneous sebaceous carcinoma is a rare disease that makes up an estimated 0.05% of all cutaneous malignancies. The ocular region accounts for nearly 75% of all reported cases. It's is a potentially aggressive cutaneous tumor with high recurrence rate, substantial potential for metastases and significant mortality rate.
OBJECTIVES: We aim to highlight the potential aggressive nature of sebaceous carcinoma and review the clinical features of the tumor as well as the different possible treatment modalities reported in the literature.
RESULTS: We report three cases of sebaceous carcinoma, two occurring on the eyelid and one on the face. The search of visceral tumors and distant metastasis was negative. Surgical resection was performed with tumor free margins. The two patients with orbital tumor has metastasized to regional lymph nodes. They underwent neck dissection with parotidectomy followed by adjuvant radiotherapy.
CONCLUSION: Whenever sebaceous tumors are identified, clinicians should consider the presence of internal malignancies. Early recognition aids in the prompt treatment of patients, as well as screening of family members for early intervention.
Keywords: Adenocarcinoma, Sebaceous, Muir -Torre Syndrome

INTRODUCTION

Cutaneous sebaceous carcinoma is a rare disease that makes up an estimated 0.05% of all cutaneous malignancies [1]. Classically, it occurs in females and the older population at sixth to a seventh decade, and arises in the ocular region from the meibomian gland of the tarsal plate and upper eyelid [2]. It, clinically, mimics other diseases and is difficult to diagnose. Hence, an accurate and prompt diagnosis is crucial because of its fulminant course, serious associations with Muir-Torre syndrome and high potential for regional and distant metastasis [2].
We report three cases of sebaceous carcinoma, two occurring on the eyelid and one on the face. We aim to highlight the potential aggressive nature of sebaceous carcinoma and review the clinical features of the tumor as well as the different possible treatment modalities reported in the literature.

OBSERVATIONS

Case 1:
A 53-year-old man, with a medical history of hypertension, have presented for 10 years with a protruding mass of the left cheek that grew in size in the past 2 years. Clinical examination showed a well-defined budding tumor of the left cheek measuring 2 cm that bleeds at the touch (figure 1). There were no enlarged cervical lymph nodes. Further investigations in search of visceral tumors and distant metastasis were normal (digestive endoscopy, colonoscopy and cervical thoracic and abdomino-pelvic CT scan). The patient underwent a surgical excision considering a 5 mm margin with direct suture since there was not an important skin defect. The margins were tumor free in the definitive histopathology and the diagnosis of sebaceous carcinoma was confirmed. The patient was cancer free after five years of follow up.



Figures 1 : Budding lesion of the left cheek (arrow)

Case 2:
A 51-year-old woman, with no medical history, presented with a nodular tumor of the lower eyelid of the right eye with conjunctival tarsal plate invasion. The patient underwent surgical resection at the ophthalmology department considering a 4 mm margin and repairing with a cutaneo-muscular transfer flap. The histopathology report showed that the margins were tumor free. The search of visceral tumors and distant metastasis was negative.
The patient was transferred to our department 2 years later presenting with enlarged lymph nodes of the right tragus region (2 cm) and the right submandibular region (3 cm). She underwent a right exofacial parotidectomy along with

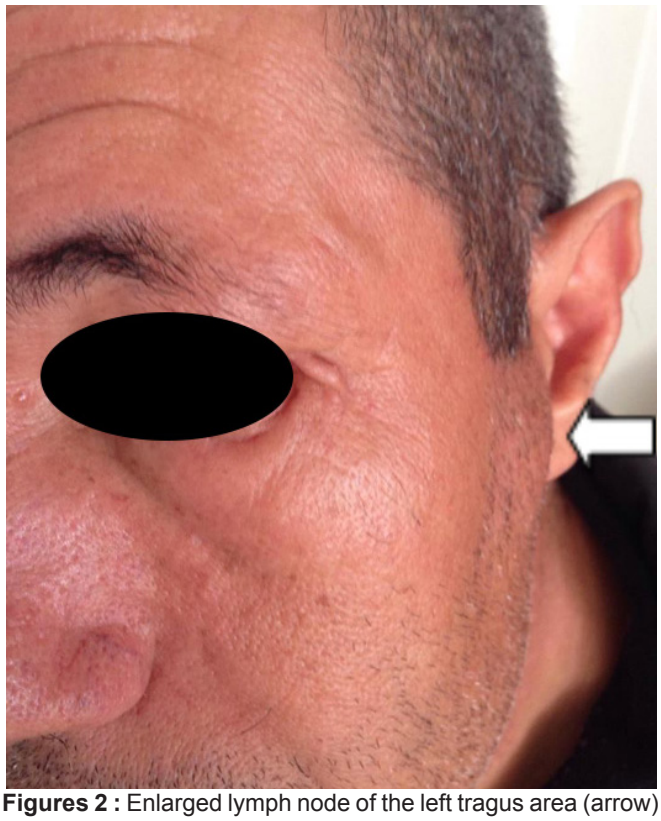
Corresponding Author : DHAMBRI Sawsen
E-mail adress: sawsendhambri@gmail.com

a right functional neck dissection removing lymph nodes of levels I, II, III and IV. Histopathology concluded to lymph node metastasis (17 N+/27N) with capsular rupture. The patient underwent four cycles of chemotherapy and 66 Gy external beam radiotherapy. She was cancer free after five years of follow- up.

Case 3:

A 55-year-old man, with no medical history, presented ten years ago with a nodular tumor of the left lower eyelid, treated surgically at the ophthalmology department. He presented, 10 years later a local recurrence treated surgically at the ophthalmology department and repaired using a transfer flap. The histopathology report showed that margins were tumor free. The search of visceral tumors and distant metastasis was negative.

The patient was referred to our department 7 months later for enlarged lymph nodes of the left tragus region (4 cm) along with local recurrence and conjunctival invasion (figure 2). He then underwent a left exofacial parotidectomy along with a left functional neck dissection removing lymph nodes of levels II, III and IV. Histopathology concluded to lymph node metastasis of the parotid. The conjunctival biopsy concluded to recurrence of sebaceous carcinoma. The patient get external beam radiotherapy focused on the parotid and cervical areas and was operated a second time at the ophthalmology department. The patient was cancer free after three years of follow up.



Figures 2 : Enlarged lymph node of the left tragus area (arrow)

DISCUSSION

Sebaceous carcinoma is a malignant tumor with sebaceous differentiation that was first described by Allaire in 1891 [3]. It is a rare and aggressive malignant neoplasm usually occurring in the head and neck region, arising, in 75 % of cases, in the periocular region, particularly the upper eyelid, due to its abundance in meibomian glands [4]. It constitutes less than 5% of eyelid malignancies and has been considered to be the third most common eyelid malignancy [5].

Sebaceous carcinoma is traditionally divided into 2 groups: Tumor arising from the ocular adnexa, and those arising in extra ocular sites. Latest one most commonly involves the head and neck region [2]. It affects mainly women in the ocular form, and usually manifests between the sixth and the seventh decades [6].

It may occur in Muir-Torre syndrome (MTS). MTS is a rare autosomal dominant genetic disease that is a phenotypic variant of hereditary non-polyposis colorectal cancer or Lynch syndrome. Germline mutations in hMSH2 and hMLH1 genes are often associated with this disorder [7]. Criteria for diagnosis of MTS include the presence of at least one sebaceous adenoma, sebaceous epithelioma, or sebaceous carcinoma and at least one visceral cancer [7]. Sebaceous tumors precede visceral malignancies in 22% [8]. The most common visceral malignancies occurring in MTS are colorectal and genitourinary cancers. Therefore, some authors advocated genitourinary surveillance in addition to upper and lower gastrointestinal endoscopy for all patients with sebaceous tumors [9]. None of our patients had visceral tumors associated to the cutaneous sebaceous carcinoma.

Sebaceous carcinoma is a potentially aggressive cutaneous tumor. The overall mortality rate is 6–11% because of delay in the diagnosis and treatment [5]. Regional lymph node metastasis occured in 20–30% of cases, systemic metastasis in 8–67%, and disease-related mortality in 3–41%[6]. The two patients with orbital sebaceous carcinomas had metastasized to regional lymph nodes and the parotid.

Authors have reported predictive factors of poor prognosis: duration of symptoms more than 46 months, tumor diameter exceeding 10 mm, involvement of both upper and lower eyelids, orbital invasion, multicentric origin, poor differentiation, high infiltrative pattern, vascular invasion, lymphatic invasion, and pagetoid invasion by the tumor [6].

Most authors recommended radical surgical excision with a 4mm tumor free margin. Moh's micrographic surgery is a more effective method of treatment followed by eyelid reconstruction [5, 6].

Conjunctival map biopsies are recommended before start of the surgery as there is always a chance of intraepithelial pagetoid spread. If the tumor is very large or recurrent, if there is pagetoid spread, or if it has spread to the other eyelid or to orbital tissues, a subtotal or complete exenteration is required [6]. Local nodal disease without distant metastasis is treated by radical neck dissection [10]. Radiation is indicated for poor surgical outcome candidates due to advanced age, for palliation, and in patients who refuse exenteration for advanced local disease [5].

CONCLUSION

Sebaceous carcinoma is an uncommon aggressive malignancy originating from sebaceous glands. The distinction from other benign entities is important and challenging. Predisposing cancer syndromes should always be excluded. Treatment of these tumors as illustrated in these 3 cases, is based on a combination of surgery and adjuvant RT or Chemoradiation therapy. Though the low mortality rate, management of the disease should be early to avoid mutilating interventions.

Conflicts of interest: No conflicts of interest to declare in connection with the article.

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