Sebaceous Carcinoma of the Eyelid: A Case Report and Review of the Literature

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ABSTRACT

Palpebral sebaceous carcinoma (PSC) is a rare malignant disease. Despite the possibility of extraocular location, the ocular region is the most described site in the literature. This tumor is poorly known and can mimic various benign conditions, explaining the frequent diagnostic delays. We report a new observation of sebaceous carcinoma in a 59-year-old patient and a review of the literature. It mainly affects the elderly with a female predominance. Its location in the upper eyelid remains the most common site, unlike our patient who presented with a lower eyelid disease. Surgery seems to be the treatment of choice for local sebaceous cell carcinoma, the role of radiotherapy is still to be clearly defined but the studies detailed in our paper have confirmed its effectiveness in inoperable patients or patients with advanced local disease, the use of chemotherapy during or before radiotherapy remains to be clarified.

Keywords: Sebaceous carcinoma, radiation therapy, eyelid tumor, oncology.

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I. INTRODUCTION

Palpebral sebaceous carcinoma (PSC) is a malignant disease originating from the sebaceous glands of the eyelids [1]. Despite the possibility of extraocular location, the ocular region is the most described site in the literature [1]. This tumor is rather rare and poorly known and can mimic various benign conditions, explaining the frequent diagnostic delays [2].

We report a new observation of a sebaceous carcinoma in a 59-year-old patient and a review of the literature.

II. CASE DESCRIPTION

Our patient was a 59-year-old male with no prior medical history and an ECOG (Eastern Cooperative Oncology Group) performance status of 1. He presented with a painful right lower eyelid mass that had been evolving for more than a year (Fig. 1). Magnetic resonance imaging of the facial region showed a 35 x 29 mm tissue formation in the lower eyelid with extraconical development enhanced after gadolinium injection without associated adenopathy (Fig. 2). A biopsy of the palpebral lesion was performed and demonstrated a sebaceous carcinoma. An extension workup was carried out (Thoracic CT scan and abdominal ultrasound) without finding secondary localizations.

Due to the advanced nature of the tumor and the inability to perform a wide local excision, a right total orbital exenteration with homolateral jugulo-carotid lymphadenectomy was performed.

Clinical examination revealed a clean orbital cavity, without any palpable cervical node.

The rest of the general examination was unremarkable.

The pathology (Fig. 3) confirmed the sebaceous carcinoma measuring 3.5 cm in the long axis, the deep resection limit was less than 1 mm (R1), with the presence of perineural and lymphovascular space invasion. The jugulo-carotid lymph node dissection included 10 nodes free of tumor proliferation.

The tumor was classified pT3bN0M0 R1 of the American Joint Committee on Cancer (AJCC) TNM classification of eyelid tumors in its 7th edition.

After discussion of the case in a multidisciplinary team it was decided to proceed with adjuvant concomitant chemoradiation because of the deep resection margin involvement (less than 1 mm). The patient underwent a dosimetric scanner with fitting of a 5-point thermoformed mask.

The treatment was delivered by intensity modulated radiotherapy with the integrated boost technique (Fig. 4). The delivered dose was 66 Gy on the tumor bed and 56 Gy on the homolateral cervical lymph nodes. The concomitant chemotherapy was based on cisplatin 40 mg per m² per week.

The treatment sessions were followed regularly by the patient without incident except for a grade 1 radiodermatitis that appeared around the edge of the operating bed in the right eye.

Follow-up for 18 months showed no local or metastatic recurrence.
III. DISCUSSION

Sebaceous carcinoma is a rare tumor that often affects the eyelids. Sebaceous carcinoma constitutes 0.2 - 0.8% of eyelid tumors, 1 - 3.2% of all eyelid malignancies, and 2.8% of orbital neoplasms [3]. It originates from the Meibomian glands of the tarsus, the Zeis glands of the palpebral skin or the sebaceous glands of the wattle. It mainly affects the elderly with a female predominance. [1] Its location in the upper eyelid remains the most common [4].

Palpebral sebaceous carcinoma usually presents as a hard nodule [5], a painless papule, or a rapidly progressing cystic nodule. [4] Diagnosis is often delayed because early periorcular sebaceous carcinoma may appear similarly to chalazion, conjunctivitis, blepharitis, or other inflammatory conditions of the eye unit [6].

The mean time reported from lesion onset to diagnosis ranges from 1.0 to 2.9 years. Our patient consulted 1 year after the onset of symptoms.

Biopsy is essential to establish the diagnosis [2]. There is no pathognomonic clinical sign differentiating sebaceous carcinoma from other tumors such as basal cell carcinoma or squamous cell carcinoma. The definitive diagnosis is histological [5].

In a very interesting study [2] a group of pathologists identified risk factors for lymph node metastases and developed and validated a new predictive nomogram using various methods to offer accurate personalized estimates for sebaceous carcinoma eyelid lymph node metastasis. The proposed nomogram included clinicopathologic factors in addition to the T category of TNM staging system and has shown great clinical potential application.

Treatment is essentially based on extensive early surgical excision. Micrographic surgery according to Mohs may be indicated for a better evaluation of the resection margins. During Mohs surgery, the surgeon removes the tumor along with a very small amount of healthy tissue surrounding the tumor. The surgeon uses a microscope to examine what has been removed.

If the surgeon finds cancer cells at the edge of the removed tissue, they remove another amount of tissue and examine it under a microscope. This process continues until the surgeon sees no more cancer cells on the edge of the removed tissue [8].

Exenteration should be considered in cases of extensive growth into the orbit or recurrent orbital disease after globe-sparing surgery.

Cryotherapy and Mitomycin C can be used in case of superficial pagetoid lesion of the epithelium [9].

Few studies have reported the use of Radiotherapy (RT) in the treatment of sebaceous carcinoma.

Definitive Radiotherapy is used in non-surgical candidates or as an adjuvant for incompletely excised tumors or in case of perineural invasion and can be used palliatively for metastatic or incurable diseases [10].

The dose varies from 50 to 70 Gy in fractions of 2 Gy. However, it should be noted that the risk of some undesirable effects occur when the patient undergo a conservative approach surgery such as dryness of the eye, retinopathy, secondary tumors. Eye drops of local anesthesia with steroids are applied before insertion of eye shields and patients wear an eye patch for 1-2 hours after RT each day [11].

A study of patients with periorcular sebaceous carcinoma treated with 39 Gy RT gave a 100% success rate. 39 Gy in 6 fractions of RT resulted in complete response and 57% disease-free survival at 5 years [12].

In another study, patients treated with RT from 50 to 66 Gy had an overall 5-year survival rate of 80% and a free local progression rate of 93% [13].

Radiation therapy as primary treatment for patients who were inoperable or refused exenteration showed surprisingly good results when the radiation dose was 50-60 Gy. In a series published by [13], the survival was at 100% after 5 years in patients who received radiation therapy as their primary treatment.

Postoperative irradiation after exenteration of T3 tumors (AJCC 7th edition) significantly reduces the risk of recurrent disease. Radiation therapy has also been recommended for recurrent disease after exenteration and postoperatively if insufficient margins or perineural extension is detected. Reference [14] recommend radiotherapy in case of T3 or more (AJCC 7th edition), lymph nodes metastases.

Neoadjuvant systemic chemotherapy is often reserved for advanced tumors. In rare cases, neoadjuvant chemotherapy may allow local resection of advanced tumors that would otherwise have higher morbidity. A very interesting recent study [15] demonstrated that neoadjuvant chemotherapy reduced tumor size remarkably and spared patients exenteration. The follow-up was only 18 months. However, chemoradiation probably seems a promising treatment method.

Evidence for the treatment of sebaceous carcinoma with chemotherapy or concomitant chemoradiotherapy with curative or palliative intent is limited to case reports.

Most regimens include the antimetabolite 5-fluorouracil or cisplatin-based chemotherapy given experience with these drugs in head and neck cancers [15].

Due to the rarity of this malignancy, little literature exists to quantify prognostic factors associated with disease prognosis, mortality, or treatment success.

A study by [16] tried to identify these settings and found that elderly patients and patients with poorly differentiated tumors and/or distant disease at presentation have the highest risk of disease-specific mortality. Lymph node metastasis does not appear to be an independent prognostic factor in sebaceous cell carcinoma.

IV. CONCLUSION

In conclusion, surgery seems to be the treatment of choice for local sebaceous cell carcinoma, the role of radiotherapy is still to be clearly defined but the studies detailed above have confirmed its effectiveness in inoperable patients or patients with advanced local disease, the use of chemotherapy during or before radiotherapy remains to be clarified.

PATIENT CONSENT

Written informed consent for publication of their clinical details and clinical images was obtained from the patient.
AUTHOR CONTRIBUTION
Hatim Talib: corresponding author and writing the paper; Tarik Chekrine: writing the paper and study concept; Mouna Bourhafour: correction of the paper; Hassan Jouhadi: Correction of the paper; Zineb Bouchbika: correction of the paper; Nadia Benchekroun: correction of the paper; Nezha Tawfiq: correction of the paper; Souha Sahraoui correction of the paper.

CONFLICT OF INTEREST
Authors declare that they do not have any conflict of interest.

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