Total upper eyelid reconstruction in the treatment of sebaceous gland carcinoma

Reconstrução total de pálpebra superior no tratamento do carcinoma de glândula sebácea

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ABSTRACT

Sebaceous gland carcinoma (SGC) is the third most frequent malignant neoplasm in the eyelids. It usually affects elderly women and can simulate blepharitis or chalazion. Upper eyelid reconstruction is always challenging. The purpose of this is to report an unusual case of SGC in the upper eyelid of a young and male patient. Total resection of the right upper eyelid was performed and histological examination revealed free margins. The reconstruction was done with two surgical procedures at different times. Due to the fact that SGC presents a high potential for metastasis, the patient continues to be followed for functional and aesthetic reevaluation.

Keywords: Carcinoma; Sebaceous gland neoplasm/surgery; Upper eyelid; Ressection; Reconstruction.

RESUMO


Descritores: Carcinoma; Neoplasias das glândulas sebáceas/cirurgia; Pálpebra superior; Ressecção; Reconstrução.

Received for publication 2/6/2019 - Accepted for publication 30/8/2019.
INTRODUCTION

Sebaceous gland carcinoma (SGC) is a rare malignant neoplasm with aggressive local and metastatic behavior. It often originates in the eyelids due to the presence of sebaceous glands in this topography. Among these, the tarsal meibomian glands are the most affected.

The incidence of SGC on the Asian continent is high, ranging from 28 to 60% of eyelid malignant neoplasms. In Westerners, this incidence is relatively low, around 4 to 5%. This carcinoma occurs in women aged 65-70 years, and is rarely found in young men. Risk factors for the development of SGC include prior local radiotherapy, immunosuppression, use of thiazide diuretics, HPV exposure, and mutations in the p53 gene.

The importance of reporting this case is based on the occurrence of SGC in a young male, with reconstruction of all upper eyelid layers after tumor exeresis.

Case report

The patient to be reported consented to the use of his images in scientific work.

A 39-year-old male patient, previously healthy, presented painless and adhered lesion in the right upper eyelid in December 2016, initially diagnosed as a hordeolus. He had topical treatment with eye ointment, but without improvement. Faced with the presence of the lesion for several months, he underwent a surgical procedure in April/2017. The total exeresis of the tumor revealed in the pathological examination a sebaceous cell carcinoma with compromised margins. At this time, the lesion was not re-approached and the patient was referred to the Ocular Plastic Sector of Hospital das Clínicas - HC / FMUSP. His medical appointment was three months later and during this period nodulation reappeared with progressive growth.

At external ocular examination, a large rounded mass was visible on the right upper eyelid, approximately 3 cm in diameter. The “mass” effect caused mechanical ptosis. The lesion was hardened, adhered to deep planes and not adhered to the adjacent skin. Eyelid eversion revealed a scar area related to previous surgery, thickening of the tarsal plate, with marked distortion of the anatomy and vascular engorgement in the periphery of the lesion. The tarsus, besides being thick, had a fibroelastic nodulation and whitish color with “fish flesh” appearance. No ducts of the meibomian glands were observed in the nodulation region. The edges were undefined and they extended beyond the upper margin of the tarsus (Figure 1). On ophthalmological examination, a superior punctate keratitis was noted in the right eye, without further alterations. Absence of palpable lymph nodes in the preauricular and submandibular regions. The orbital computed tomography exam, performed in August/2017, showed no invasion of orbital structures.

The patient underwent surgery in August/2017 for total lesion exeresis with a 5 mm margin, intraoperative freezing and upper eyelid reconstruction (Figure 2). A conjunctival mapping was performed by randomly drawing conjunctival samples around the lesion for intraoperative analysis. The surgical margins and conjunctiva samples were free in freezing (R0).

For the eyelid reconstruction, a contralateral free tarsal graft was obtained to compose the posterior lamella, associated with the sliding of a bipedicled myocutaneous flap (skin and preseptal orbicular muscle) to remake the eyelid (“bucket flap”). It was complemented by suturing a retro-auricular skin graft over the remaining preseptal orbicularis.

Pathological examination revealed moderately differentiated sebaceous carcinoma. Many atypias, pleomorphic cells, atypical mitoses and vacuolated cells were observed. The immunohistochemical profile expressed androgen receptor and loss of expression of the MSH 2 and MSH 6 genes (Figure 3). Oncology staging examination revealed no metastatic disease.

In the 10th postoperative period, he presented partial dehis-
cence in the central region of the new eyelid margin, with a small area of tissue necrosis in the adjacent skin graft region. We opted for expectant procedure, as the patient maintained good eyelid occlusion and Bell’s sign present despite the neo eyelid irregularity. However, he complained a lot about the unsightly appearance. The defect was central and had an inverted “V” appearance. It measured 3 mm vertically and 7 mm horizontally, producing a lagophthalmos of 3 mm. Absence of structures touching the cornea (Figure 4).

In July 2018 we reoperated the patient using the Cutler-Beard Technique: we revived the edges of the defect and made a myocutaneous flap of the eyelid less than 5 mm from its margin, measuring 16mm x 10mm. This flap internally crossed the eyelid border and addressed the defect, leaving the eye occluded. There was no need to obtain cartilage or palate graft (Figure 5).

The patient evolved without necrosis or dehiscence. In the fifth postoperative week, we opened the right eyelid cleft and sutured the bloody areas. After six months it remains with excellent eyelid closure and good aesthetic result (Figure 6). Currently he continues in quarterly outpatient follow-up for functional and aesthetic reevaluation.

**Discussion**

The patient in this report was young. The average age of patients affected in the literature is 67.5 years.(5) No justification was found for the early onset of the disease such as Muir-Torre Syndrome, HIV or retinoblastoma.(3,6) In addition, the patient was male, which also contradicts the studies, in which the majority are women.(5,7)

The clinical picture began painlessly and the appearance of the lesion also resembled what is described in the literature: usually a deeper, yellowish, painless lump in the upper eyelid. Moreover, this carcinoma may have a diffuse inflammatory process (5,8) and the delay in diagnosis is frequent. There is great similarity of the neoplasia to benign lesions such as blepharitis, recurrent chalazion, upper limbic keratoconjunctivitis, and ocular pemphigoid, (8,9) as in our report, which was confused with hordeolus and chalazion.

When suspecting SGC, a biopsy should be performed and the pathologist alerted to the potential diagnosis.(7) Shields et al. reported that even after biopsy, the diagnosis of SGC was made in only 50% of cases and 18% were misdiagnosed as squamous cell carcinoma (SCC). This suggests the difficulty in diagnosing SGC.(10)

SGC has a tendency towards pagetoid growth, making surgical management difficult as well.(7) The main therapeutic option is extensive exeresis of the lesion with freezing or micrographic surgery (Mohs), followed by reconstruction.(4) We used freezing and two reconstructions at different times.

Regardless of the type of reconstruction and tissue appearance, in the same surgical procedure, small bulbar, fornical and eyelid conjunctival mapping biopsies should be performed 360°, as in our report. Thus, the extent of conjunctival and corneal pagetoid invasion can be determined and the surgical procedure planned according to these findings. (10)

No perineural invasion was detected in our patient. When this invasion is found in the surgical specimen, even if the margins are negative, most authors advocate additional treatments such as postoperative adjuvant radiotherapy.(5)

Total reconstruction of the upper eyelid is always challenging. If performed improperly causes serious problems such...
as conjunctivitis, dry eye, keratitis and aesthetic deformities. The main function of the upper eyelid is protection and corneal lubrication. If this function is impaired, the eye surface and visual quality will be compromised. The purpose of reconstructing this ocular annex is to eliminate entirely the neoplasia while maintaining anatomical and physiological functions as well as eyelid aesthetic appearance. The tissues used for upper eyelid reconstruction should be well vascularized and are usually neighboring structures. A smooth internal surface must be maintained so as not to damage eye structures. Auricular cartilage grafts are not a good alternative due to fragility in surgical handling and high memory, although some authors use them. Part of the contralateral tarsus was used in our patient, replacing the same structure. Superiorly to the posterior lamella, a “Bucket” flap consisting of sliding skin and orbicular muscle was held medially and laterally, maintaining vascularization. Unfortunately there was a necrosis in the central area, possibly due to the thin flap or section of one of the vascular pedicles.

Our patient was reoperated using the Cutler-Beard flap, which is advisable for defects that occupy more than 50% of the upper eyelids and with total levator aponeurosis. Complications of this reconstruction include entropion, eyelid margin irregularity, eyelash loss, lower eyelid retraction, and flap necrosis. Occasionally it may spread tumor cells to the lower eyelid, however it has been employed in a second/third phase and the chances of this occurring are extremely low.

In addition to reconstructive exeresis, cryotherapy, topical Mitomycin-c, neoadjuvant and adjuvant chemotherapy, external beam radiotherapy and exenteration can also be used as treatment. It is important to remember that local control is the key to preventing the spread of this tumor. As our patient had adequate local control with surgical removal, we did not use the other treatments mentioned above. However, even if the surgery is successful and with free margins, systemic follow-up with an oncologist is recommended, as was the case with our patient.

Mitomycin-C topical chemotherapy or cryotherapy may benefit patients with confirmed intraepithelial invasion on mapping biopsies. On the other hand, neoadjuvant chemotherapy is a promising and safe treatment strategy, providing adequate tumor volume reduction and tumor locoregional control after 3 to 6 cycles. This is followed by local external radiotherapy if excision margins are suspected and by 3 to 6 cycles of adjuvant chemotherapy.

Approximately 38% of recurrent SGC tumors will spread to orbit and their direct invasion is treated with exenteration. Metastases to regional lymph nodes occur in 8% to 20% of patients with SGC. Haematogenic dissemination to distant sites, including lung, liver, bones and brain, occurs in 1% to 6% of patients.

The patient reported here had a large lesion and eight months of evolution until complete exeresis. Clinical and histological features that have been associated in the literature with increased mortality in patients with SGC include larger tumor size; vascular, lymphatic or orbital invasion; duration of symptoms for more than six months; poor differentiation; previous radiation and multicenter origin. Reported overall mortality rates range from 3% to 22%.

In patients unable to tolerate or refusing to undergo surgical excision, modern radiotherapy may be an acceptable treatment alternative for this malignant eyelid tumor. At least 55 Gy of radiation must be given to the tumor.

Our patient had a severe carcinoma involving the entire right upper eyelid and with divergent epidemiology in the literature, but the evolution has been favorable so far, only with complete surgical exeresis of the lesion. The sooner the diagnosis and treatment are done, the greater the chances of cure. In-depth investigation of suspected cases and dialogue between surgeon and pathologist should be encouraged. Always perform freezing or micrographic Mohs surgery intraoperatively. Systemic assessment with oncologist is fundamental in follow-up.

References


