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Periocular basal cell carcinoma with orbital invasion: a case report

Carcinoma basocelular periocular com invasão orbitária: relato de caso

Case Report

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ABSTRACT
Basal cell carcinoma is the most frequent periocular neoplasm. Although mortality is low, orbital invasion represents a severe complication associated with high morbidity. We report the case of a 49-year-old male smoker with an aggressively evolving periocular lesion culminating in amaurosis. Treatment included orbital exenteration and complex reconstruction using local and regional flaps, complicated by partial necrosis and bone exposure, requiring multiple debridements. Histopathological examination revealed a sclerodermiform pattern with squamous differentiation. This case highlights the devastating consequences of locally advanced basal cell carcinoma as well as the importance of early diagnosis and a multidisciplinary approach to prevent deformity and loss of function.
Keywords: Carcinoma, Basal Cell; Orbital Exenteration; Skin Neoplasms

RESUMO
O carcinoma basocelular é a neoplasia periocular mais frequente. Embora a mortalidade seja baixa, a invasão orbitária representa uma complicação grave associada a alta morbidade. Relata-se o caso de um paciente do sexo masculino, 49 anos, tabagista, com lesão periocular de evolução agressiva, culminando em amaurose. O tratamento incluiu exenteração orbitária e reconstrução complexa com retalhos locais e regionais, complicada por necrose parcial e exposição óssea, exigindo múltiplos debridamentos. A histopatologia evidenciou padrão esclerodermiforme e diferenciação escamosa. O caso destaca as consequências devastadoras do carcinoma basocelular localmente avançado e a importância do diagnóstico precoce e da abordagem multidisciplinar para evitar deformidades e perda funcional.
Palavras-chave: *Carcinoma Basocelular; Exenteração Orbitária; Neoplasias Cutâneas*

INTRODUCTION

Basal cell carcinoma (BCC) is the most common malignant neoplasm of the periocular region, accounting for approximately 80% to 96% of tumors in this location. Although often considered a tumor with indolent behavior and low metastatic potential, periocular BCC may exhibit locally aggressive behavior. Orbital invasion is an uncommon event, with an estimated incidence ranging from 0.8% to 5.5% in developed countries, reaching alarming rates of up to 17% in developing countries, often due to delays in diagnosis and treatment.[1]

Clinical progression may be silent, requiring constant vigilance for signs of high-risk tumors. The main factors associated with orbital invasion include multiple recurrences, large tumor size, positive surgical margins, location in the canthal region (medial or lateral), and aggressive histological subtypes, such as morpheaform/sclerodermiform, infiltrative, and basosquamous variants. Perineural invasion, although present in less than 1% of cases, is a marker of poorer prognosis.[2]

Although the mortality rate associated with periocular BCC remains low, morbidity is high. Tumor progression may result in severe facial disfigurement and vision loss, dramatically affecting patient quality of life.[3]

The objective of this study is to report a case of periocular BCC with aggressive progression, resulting in visual loss and the need for orbital exenteration, highlighting the serious consequences of delayed specialized care and the importance of early treatment.

CASE REPORT

A 49-year-old male patient, smoker, with no comorbidities, was referred to a specialized service with a history of a periocular lesion that had first appeared in 2021. The condition began as a small papule on the lower eyelid and progressed over two years with ulceration, intense pain, and expansive growth, culminating in ipsilateral loss of visual acuity (Figure 1).

Dermatological examination revealed extensive ulceration in the left orbital region, with pearly borders and large-scale tissue destruction. Ophthalmological examination found ocular globe atrophy and corneal ulceration (Figure 2).

An incisional biopsy was performed, confirming the diagnosis of BCC. Imaging studies found orbital involvement without intracranial invasion. Given the extent of the tumor and the functional loss, orbital exenteration was performed with negative frozen-section margins (Figure 3).

Histopathological examination revealed sclerodermiform BCC with squamous differentiation (Figures 4 and 5). There was no optic nerve invasion.

Reconstruction included a mid-forehead flap associated with a cervicofacial advancement flap (Figure 6). The postoperative course was marked by partial necrosis (Figure 7), orbital bone exposure, serial debridements, and subsequent reconstruction using a temporalis muscle flap. Conservative management was adopted, allowing healing by secondary intention (Figure 8). Following these interventions, the patient was lost to clinical follow-up, and resuming it was not possible despite repeated attempts at contact by the medical team.

Figure 1: Ulceration in the left orbital region

Figure 2: Lesion with pearly borders and hyperpigmentation, ocular globe atrophy, corneal leukoma, and corneal ulceration

DISCUSSION

This case illustrates the concept of locally advanced BCC, a challenging condition in which conventional surgery or radiotherapy alone may not be curative or may result in ma-

Figure 3: Intraoperative view of orbital exenteration and tumor resection with frozen-section histological margin control.

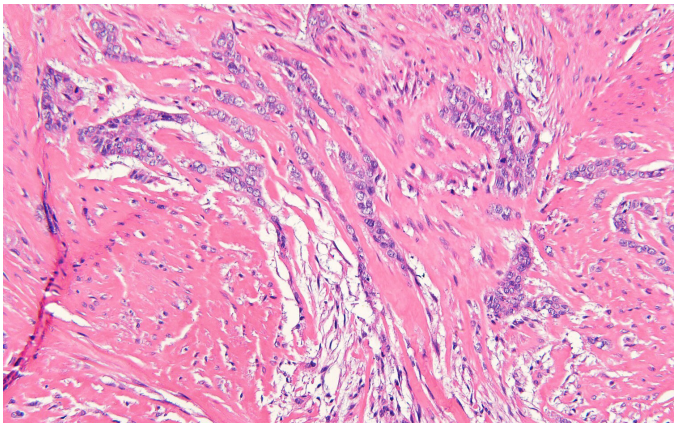

Figure 4: Histology showing basal cell carcinoma composed of thin cords of basaloid cells embedded in a desmoplastic stroma, characteristic of the sclerodermiform pattern

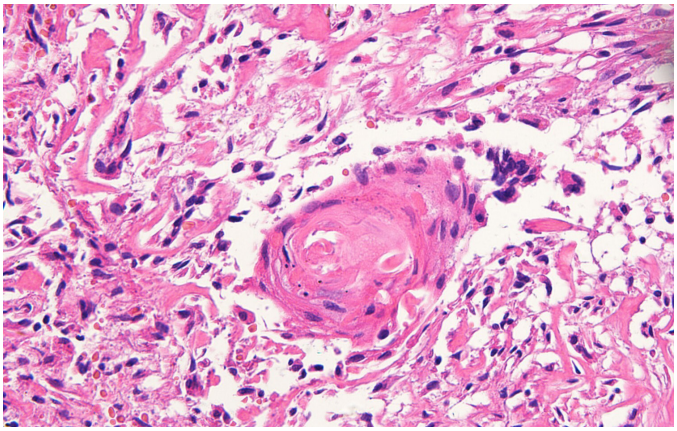

Figure 5: Evidence of squamous differentiation.

Figure 6: Immediate postoperative appearance showing the mid-forehead flap based on the supratrochlear artery and a cervicofacial advancement flap

Figure 7: Partial flap necrosis with bone exposure, requiring serial debridements

jor deformities. The 2023 European consensus-based guideline emphasizes that BCCs should not be underestimated; neglected or recurrent tumors may invade deep tissues, making treatment complex.[7]

The classification proposed by the European Association of Dermato-Oncology (EADO) and the guidelines of the National Comprehensive Cancer Network (NCCN) (version 2.2024) recommend a multidisciplinary approach for these cases. The first line of treatment for locally advanced BCC remains surgery (with strict margin control) and/or radiotherapy.[4]

In the present case, radical surgery (orbital exenteration) was required because of orbital invasion and prior visual loss. Histopathological examination revealed aggressive subtypes (sclerodermiform and squamous differentiation), corroborating

Figure 8: Wound closure by secondary intention after partial flap necrosis, 10 months after the initial tumor surgical procedure.

the rapid and destructive clinical course observed.[5,6] The sclerodermiform pattern is known for its poorly defined margins and high recurrence rate, whereas squamous differentiation (basosquamous pattern) confers a more aggressive biological behavior, approaching that of squamous cell carcinoma.

When surgery and radiotherapy are contraindicated or unsuccessful, current guidelines recommend systemic therapy. Hedgehog pathway inhibitors (such as vismodegib and sonidegib) and immunotherapy with anti-PD-1 checkpoint inhibitors (such as cemiplimab) have emerged as valuable treatment options.[4,7] For locally advanced periocular BCCs and metastatic orbital disease, vismodegib (150 mg/day) has demonstrated efficacy and may even be used as neoadjuvant therapy to reduce tumor mass and potentially decrease the rate of orbital exenteration.

Regarding prognosis, periocular BCC with negative surgical margins presents recurrence rates ranging from 1.5% to 5.6%, depending on the anatomical location. The medial canthus is the site most prone to recurrence and deep invasion because of embryologic cleavage planes that facilitate tumor spread.[4] The literature documents that recurrence rates are significantly higher for periorbital malignancies than for BCCs arising at other cutaneous sites.[9]

CONCLUSION

This case report highlights the importance of early diagnosis and regular follow-up for periocular lesions. Although BCC is often curable, it may progress to locally advanced forms with orbital invasion, resulting in blindness and mutilation, including the need for orbital exenteration. A multidisciplinary approach involving dermatologists, ophthalmologists, head and neck surgeons, and plastic surgeons is essential for appropriate management, aiming to achieve oncologic cure while minimizing functional and aesthetic sequelae.

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