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A Case Report of Advanced Neglected Basal Cell Carcinoma

Loren W. Bennett OD, MPH, FAAO

James H. Quillen VA Medical Center, loren.bennett@va.gov

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A Case Report of Advanced Neglected Basal Cell Carcinoma

Abstract

Background: Basal cell carcinoma is the most common skin cancer in humans and is frequently found in the periocular region. It is generally characterized by slow growth with rare metastasis and has an excellent prognosis with current surgical techniques. However, if neglected or inadequately treated it can lead to substantial localized tissue destruction and morbidity.

Case Report: This case presents an advanced basal cell carcinoma involving the left side of the nose and medial eyelids which had been neglected by the patient, causing significant functional disability and cosmetic disfigurement. The tumor was removed with a wide surgical excision which required skin grafts and eyelid reconstruction.

Conclusion: Early diagnosis and management of suspicious lesions is important to ensure the best clinical result. In extreme cases psychosocial or economic factors may delay treatment, resulting in more aggressive growth which requires more extensive multidisciplinary intervention to achieve a satisfactory outcome for the patient.

Keywords

basal cell carcinoma, periocular carcinoma, neglected skin cancer

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INTRODUCTION

Basal cell carcinoma (BCC) is the most common periocular malignancy found in humans.^{1,2} While it is usually slow growing and rarely metastasizes, if treatment is delayed it can be highly locally invasive and cause destruction of adjacent tissue of the eye, orbit, nose, sinuses, and face. Early recognition and intervention is important to minimize local complications and disfigurement, and with appropriate management the prognosis is usually good. This case report describes an advanced basal cell carcinoma involving the left side of the nose and eyelids which had been neglected by the patient, leading to significant localized growth and disfigurement. The tumor was successfully removed with a wide surgical excision requiring skin grafts and eyelid reconstruction.

CASE REPORT

A 73-year-old male presented for care at the James H. Quillen Veterans Affairs Medical Center (VAMC) with a large growth that started in the corner of his left eye three years previously. The patient noted that it had grown rapidly with occasional associated purulent drainage and bleeding for at least the past six months, and he was having trouble breathing out of the left side of his nose. His history included not having seen a physician for 50 years, although he did see a private doctor for this tumor about 4 months before coming to the VA. The patient said he was told it was skin cancer, but he had not gone back for any follow up visits. He was alert and oriented, did not report being on any medications, and was not being followed for any systemic conditions. A plastic surgeon and an Ear, Nose, and Throat (ENT) specialist were consulted, and examination described a baseball- size fungating, exophytic, necrotic mass approximately 12 cm in diameter involving the left side of the nose with collapse of the left nasal airway. A subsequent biopsy diagnosed the tumor as an ulcerated basal cell carcinoma.

A computed tomography (CT) scan without contrast (the patient declined contrast testing) showed extension of the tumor into the left preseptal space and anterior medial orbit with involvement of the medial eyelids (Figure 1). There was mucoperiosteal thickening of the left maxillary sinus with preservation of the paranasal sinuses, but no associated bony lesion, and no focal lesion of the brain. Management options that were initially considered included discussion of a rhinectomy with possible exenteration of the orbital contents based on the degree of infiltration of the tumor, or a wide surgical excision with radiation therapy.



Figure 1: Axial CT scan without contrast showing a large basal cell carcinoma involving the left side of the nose with collapse of the left nasal airway (red arrow), and extension of the tumor into the left anterior medial orbit and medial eyelids.

The patient was then referred to the VAMC Eye Clinic to assess the degree of ophthalmic involvement. A large necrotic mass with an unpleasant odor was observed as previously described on the left side of the nose and face, extending into the medial canthus and involving the medial left upper and lower eyelids (Figure 2). The patient's affect seemed depressed; he was alert and oriented, but minimally communicative. He described a lifelong history of decreased vision in the right eye, but he hadn't had an eye exam for several years and had stopped wearing glasses because they were not improving his vision. He did note that his left eye had always been his "good eye," and stated that the vision was getting worse, but he did not complain of visual disability. When questioned as to why he did not seek care earlier, he did not provide an answer. Family members explained that he continually refused medical care until the tumor grew to the point where he finally agreed to be evaluated.



Figure 2: Color photos of a large, 12 cm diameter, necrotic basal cell carcinoma involving the left side of the nose, the medial eyelids, and lacrimal apparatus.

Uncorrected visual acuities were 20/400 OD and 20/200 OS. The pupils were round, regular, and reactive, with no afferent defect, and there was a full range of ocular motility without restriction. An adequate refraction could not be completed due to the size of the tumor and poor response from the patient. Examination of ophthalmic structures was difficult due to physical obstruction by the tumor; what could be seen of the fundus on dilated exam was grossly intact OU, with advanced nuclear cataracts OU further restricting internal examination.

Three months after his initial presentation, the patient agreed to undergo a surgical resection of the tumor at another VA facility. A wide local excision with eyelid reconstruction and skin grafts was successfully accomplished under general anesthesia, with frozen section margins determined to be tumor-free. While the cancer was estimated to have invaded 50% of the lower eyelid and 25% of the upper eyelid, the eye itself was able to be preserved; however, the lacrimal canaliculi were involved with the tumor and removed during the surgery. The final skin defect on the nasal dorsum measured 5.5 cm x 4.5 cm.

A 2-month post-operative examination at the Eye Clinic showed significant residual medial ectropion of the left lower lid, loss of the left lower punctum with chronic epiphora, and partial left upper lid ptosis (Figure 3). Ophthalmic examination was considerably easier at this stage, and the patient's affect was much improved. A refraction found -10.00 DS OD and -0.50 DS OS, with best corrected distance acuities of 20/400 OD and 20/150 OS. Based on the refractive anisometropia and the patient's history of longstanding decreased vision since childhood in that eye, a diagnosis of refractive amblyopia OD was made. The dilated fundus examination was unremarkable, while slit lamp evaluation was significant for grade 3+ nuclear cataracts OU.



Figure 3: Post-operative color photo showing the patient after a wide surgical excision of the basal cell carcinoma with skin graft and eyelid reconstruction.

Cataract surgery was recommended, but the patient declined any further surgical intervention. Glasses and low vision aids were offered, which he also refused, stating that his vision was good enough for his needs. The patient continued to be followed periodically without recurrence of the tumor, and he passed away seven years after his initial presentation.

DISCUSSION

Basal cell carcinoma is the most common cutaneous tumor in humans, accounting for 80% of all nonmelanoma skin cancers, and is the most frequent cancer of the eyelid.¹⁻³ It arises from the neoplastic proliferation of basal cells in the epidermis of the skin, which can then invade the dermal layer and surrounding tissue. Since ultraviolet (UV) exposure is a known primary risk factor, these tumors characteristically develop on areas of the body which are exposed to the sun, with 80% developing on the head and neck, including the eyelids.³ There is also a geographic and occupational correlation as the incidence of BCC is higher with closer proximity to the equator and in workers with outdoor jobs due to increased UV exposure.⁴ Other risk factors for BCC include phenotypic traits such as fair skin or light colored hair, use of tanning beds, immunosuppression, trauma such as burns leading to cutaneous scarring, smoking, and exposure to environmental contaminants such as arsenic, coal tar, and ionizing radiation.^{1,3,5} Periocular BCC is more commonly found on the lower eyelid (44%-56%), followed by the medial canthus (27%-37%), upper eyelid (8%-9%), and lateral canthus (6%-8%).^{6,7} Eyelid BCC is more prevalent in older adults in their sixth to eighth decade of life, but up to 15% may develop in children or young adults.⁸

Clinically, periocular BCC typically manifests initially as asymptomatic or mildly symptomatic eyelid lesions, with slow growth and minimal invasion of adjacent tissue. Metastasis is extremely rare, with incidence rates reported from 0.0028% to 0.55% of all BCCs, depending on the size, location, and histological subtype.^{9,10} While several clinical and histological subtypes have been described, the most common morphological form is nodular, accounting for 50% – 79% of all BCC presentations.^{1,11} Nodular BCC manifests with pearly, shiny papules or nodules with branching telangiectasias. As it grows, crusting may appear overlying a central depression, and over time this depression may ulcerate, which has sometimes been referred to as a “rodent” ulcer.³ Bleeding of the lesion is common with minor trauma. Superficial BCC is the second most common clinical subtype (up to 15% of cases), typically appearing as a well circumscribed, scaly, thin reddish macule or plaque.¹ Nodular and superficial subtypes tend to be less aggressive in their clinical course. Morpheaform (sclerosing, infiltrative) is the least common presentation but the most aggressive form, occurring in 5% - 10% of cases. Lesions are pink to white in color, with a shiny, smooth, scar-like appearance, possibly with indurated plaques, crusting and erosions, and indistinct margins.¹ Because of this variability in presentation, BCC may be difficult to diagnose from clinical appearance alone; a wide range from 10%-40% of periocular BCCs are reported to be initially misdiagnosed.^{3,6,12} A biopsy is therefore recommended to ensure accurate identification of growths which are suspicious for malignancy.¹²

Several medical therapies for BCC have been proposed, including radiation therapy, cryotherapy, carbon dioxide laser, photodynamic therapy, and pharmacologic agents such as intralesional interferon or topical imiquimod, but these methods are associated with higher rates of recurrence.¹³ The treatment of choice for periocular BCC is surgical excision with microscopic monitoring of the excision margins, either using a wide surgical excision with frozen section or paraffin margin control, or with a Mohs micrographic surgery technique.^{4,13,14} Standard excision without intraoperative histological margin monitoring may be appropriate for some cases of smaller nodular tumors where the clinical margins can be defined with reasonable accuracy. The primary goal is to remove the tumor along with an adequate margin of normal surrounding skin to ensure complete clearance of cancerous cells and reduce the risk of recurrence, while trying to preserve function and cosmesis at the surgical site.¹³⁻¹⁵ Surgical cure rates without recurrence for periocular BCC are reported as high as 95%.^{7,13-15} A Mohs technique has been suggested to be the preferred method for periocular BCC excision to minimize the loss of normal tissue and achieve the best functional and cosmetic results around the eyelids with the lowest recurrence rate; however, it is generally considered more expensive and is not available in every setting.^{7,16,17} A Cochrane literature review comparing these surgical techniques though could not

conclusively confirm that Mohs procedures had a clear advantage in recurrence or complication rate compared to standard surgical excision with wide margins, and also did not find any direct cost comparison of the different techniques.¹⁸ Management decisions should be individualized based on the clinical presentation, patient risk factors, and surgeon's expertise in each setting.

More aggressive or neglected periocular BCC can result in orbital and intracranial invasion, an uncommon complication occurring in less than 5% of cases.¹⁹ Signs that suggest orbital involvement include a visible or palpable mass fixed to the bone, limitation of ocular motility, and globe displacement. If left unchecked, posterior extension into the orbital apex and cavernous sinus may occur. Longstanding neglected tumors, recurrent or incompletely excised tumors, and medial canthus location are risk factors for orbital and intracranial extension.^{19,20} Radiologic imaging is necessary in cases of suspected orbital involvement to determine the extent of the infiltration, with computed tomography (CT) to look for bony destruction and magnetic resonance imaging (MRI) to demonstrate soft tissue changes. Management of more extensive orbital invasion often requires exenteration with or without adjunctive radiotherapy to achieve local control of the tumor.^{19,20} In selected cases of anterior orbital involvement or with monocular patients, local resection of the tumor may still be an option.

In patients with more locally advanced or metastatic BCC, or in cases of basal cell nevus syndrome (Gorlin syndrome) which involves numerous periocular and facial cutaneous tumors, the condition may not be amenable to surgery or radiotherapy.²¹ Vismodegib and sonidegib are oral Hedgehog signaling pathway inhibitors that have had some success in shrinking the tumor size.²²⁻²⁴ (The Hedgehog signaling pathway is critical for cellular differentiation during fetal development, with limited activity beyond this stage.) Anomalous activation of the Hedgehog pathway has been implicated in several types of malignancies including BCC. In a study of seven recurrent, locally advanced cases of periocular BCC without metastasis that were not good candidates for surgical resection or radiation, more than half which were treated with vismodegib showed clinically significant shrinkage of the tumor which was sustained for at least six months.²⁴ Emerging clinical trial data will continue to better define the role of this medical therapy for selected patients who are not good candidates for other treatment modalities.

Early identification and treatment of suspicious lesions is important to improve the outcome and lessen the chance of complications from more aggressive tumor growth. It is rare for a BCC to progress to this size and show the degree of localized invasion and destruction of contiguous tissue seen in this patient. However, instances of significant BCC growth with extensive localized morbidity and disfigurement are occasionally reported from longstanding neglected tumors.²⁵⁻³²

There is likely a complex interaction of individual psychological and socioeconomic risk factors that leads to a situation of neglect and allows these lesions to progress to such advanced stages. Older age, lower socioeconomic status, mistrust or fear of the medical system, mental illness, and lack of education concerning the significance of the diagnosis are some proposed influences that may contribute to the delay in seeking medical attention for skin cancers.^{26,27,32} It is also possible that a slow growing, asymptomatic neoplasm may not initially cause concern, and the patient can react to subsequent growth with denial of the problem or an acceptance of a slowly progressive situation without understanding its clinical significance. The patient's decision to seek medical treatment may be ultimately influenced by external factors such as a sudden change in the tumor (like bleeding, pain, or rapid growth), or the proactive intervention of a friend or family member.^{25,27} A multidisciplinary approach may be needed to address these barriers to care and get the patient to the stage where he is ready to consider treatment options.³²

CONCLUSION

BCC is generally a slow growing, indolent tumor which usually carries a high cure rate with appropriate surgical techniques. However, delays in diagnosis or in seeking medical care with more aggressive forms of BCC can lead to extensive localized tissue destruction, cosmetic disfigurement, and functional disability. Cases of extreme tumor neglect are rare and may require more complex intervention with extensive reconstructive efforts. Understanding and overcoming the multiple psychosocial factors which manifest in denial and delayed care can be challenging. This case is a reminder of the importance of early recognition and intervention to prevent significant morbidity.

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