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DILEMMAS IN MANAGEMENT OF OCULAR SURFACE SQUAMOUS NEOPLASIA (OSSN) – A RETROSPECTIVE INTERVENTIONAL CASE REPORT

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ABSTRACT
A 70 years old male with history of being diagnosed as carcinoma-in-situ in left eye (OS) at nasal limbus was operated for mass excision with bare sclera 3 months back – histopathologically – squamous cell carcinoma (SCC) was diagnosed and patient was on topical steroids. Then, patient developed scleral melt and was referred. On presentation, there was a scleral perforation at nasal limbus and uveal prolapse, shallow anterior chamber and decentered posterior chamber intraocular lens (PCIOL) with nasal choroidal detachment on ultrasound (USG). Rheumat work up was done to rule out autoimmune etiology for scleral melt but it was within normal limits. Patient was operated for OS granuloma excision with cryopexy on free margins with scleral patch graft and conjunctival autograft. Histopathology revealed it to be moderately differentiated SCC with scleral invasion with free margins. Patient was given 2 cycles of interferon α eye drops. But, conjunctival autograft retracted after 3 days, so resuturing was done. Then there was sterile scleral melt with corneal edema and drop in vision to light perception 1 month later. Patient was started on oral steroids but melt persisted and again patient developed scleral perforation. So, we have:
Dilemma 1 – Was scleral melt due to autoimmune condition / recurrence / interferon toxicity?
Dilemma 2 – Management – Re-sclero-corneal lamellar patch graft / enucleation / lid sparing exenteration?

KEYWORDS: OSSN, Dilemma, Recurrence

INTRODUCTION

Ocular surface squamous neoplasia (OSSN) represents a spectrum of dysplastic and neoplastic squamous epithelial lesions involving the conjunctiva and cornea, ranging from conjunctival intraepithelial neoplasia (CIN) and carcinoma in situ (CIS) to invasive squamous cell carcinoma (SCC). It is the most common non-pigmented malignancy of the ocular surface, with incidence varying geographically and being particularly higher in regions with increased ultraviolet-B exposure. [1,11] Advanced age, male sex, chronic solar exposure, immunosuppression, human papillomavirus (HPV) infection, and human immunodeficiency virus (HIV) infection have been identified as important risk factors for its development. [2,10,11]

Although OSSN is generally considered a low-grade malignancy with favorable outcomes when diagnosed early, local recurrence remains a significant concern. Recurrence rates have been associated with larger tumor size, positive surgical margins, advanced histopathological grade, and inadequate primary treatment. [4,8] Surgical excision using a "no-touch" technique with cryotherapy to surgical margins remains a widely accepted treatment modality, often supplemented by topical chemotherapeutic agents such as interferon alfa-2b, mitomycin-C, or 5-fluorouracil. [6-8,12] Nevertheless, recurrent and invasive disease can pose considerable diagnostic and therapeutic challenges.

Intraocular and scleral invasion by conjunctival SCC is uncommon, occurring in only 2–8% of reported cases, but may result in severe ocular morbidity and difficult management decisions. [2,3,9] Furthermore, postoperative scleral thinning or melt may arise from diverse etiologies, including tumor recurrence, surgical ischemia, inflammatory or autoimmune processes, infection, or drug-related toxicity. Distinguishing among these entities is often challenging because clinical manifestations may overlap, while treatment strategies differ substantially. [10,11] Delay or error in diagnosis may lead either to undertreatment of persistent malignancy or to unnecessary radical surgery.

We report a case of recurrent OSSN with histopathologically confirmed scleral invasion that presented with progressive scleral melt and recurrent perforation following previous excision and adjunctive interferon alfa-2b

therapy. The case highlights the diagnostic uncertainty in differentiating tumor recurrence from non-neoplastic causes of scleral necrosis and illustrates the complex management dilemmas encountered in advanced OSSN.

MATERIALS AND METHODS

Retrospective, Interventional Case Report

RESULTS

Written, informed consent has been taken from the patient for writing this case report. A 70-year-old male was referred to our tertiary eye care centre with complaints of decreased vision, ocular discomfort, and a scleral perforation with uveal tissue prolapse in the left eye. Three months earlier, he had been diagnosed elsewhere with carcinoma in situ involving the nasal limbal conjunctiva of the left eye and underwent surgical excision using a bare sclera technique. Histopathological examination of the excised specimen revealed squamous cell carcinoma (SCC). Following surgery, the patient was treated with topical corticosteroids for approximately two months.

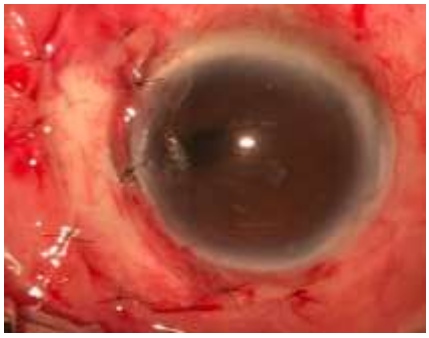
Subsequently, he developed progressive scleral thinning, culminating in scleral perforation associated with uveal prolapse and surrounding granulomatous tissue formation, for which he was referred to our institute. On presentation, the best-corrected visual acuity in the left eye was counting fingers at 4 meters. Slit-lamp examination revealed a scleral perforation at the nasal limbus with prolapsed uveal tissue and adjacent granulomatous lesion. (Figure 1)

(Figure 1: Left Eye showing scleral perforation with uveal prolapse at the site of OSSN)

The anterior chamber was shallow, and a decentered posterior chamber intraocular lens was noted. Ultrasonography demonstrated nasal choroidal detachment. The right eye examination was unremarkable.

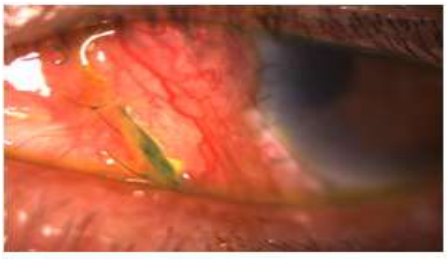
Considering the possibility of recurrent OSSN with associated scleral involvement, the patient underwent excision of the granulomatous lesion with a 3-mm tumor-free margin. Adjunctive double freeze-thaw cryotherapy was applied to the surgical margins. The resultant scleral defect was reconstructed using a scleral patch graft covered with a conjunctival autograft.

On the first postoperative day, both the scleral patch graft and conjunctival autograft were well positioned. (Figure 2)



(Figure 2: Well apposed scleral and conjunctival autograft on 1st post-operative day)

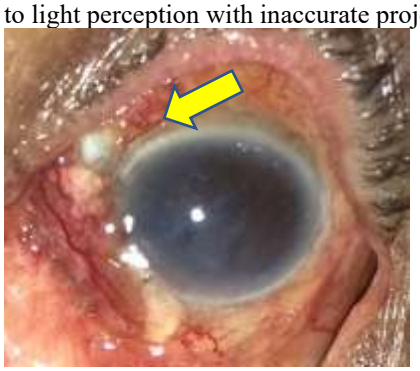
However, by the third postoperative day, graft retraction was observed, necessitating resuturing of both the scleral patch graft and conjunctival autograft. (Figure 3)



(Figure 3: Graft retraction and broken sutures on 3rd post-operative day)

Histopathological examination of the excised tissue demonstrated moderately differentiated squamous cell carcinoma with scleral invasion. Surgical margins were free of tumour.

In view of the invasive nature of the lesion, adjunctive topical interferon alfa-2b therapy (1 million IU/mL) was initiated. The patient received two cycles of treatment administered four times daily for one week followed by a one-week drug-free interval, along with lubricants and topical corticosteroids.
Approximately one month after surgery, the patient developed progressive corneal edema, sterile scleral melt, and marked deterioration in visual acuity to light perception with inaccurate projection of rays. (Figure 4)



(Figure 4 showing scleral melt (yellow arrow) with corneal edema)

Clinical evaluation raised several differential diagnoses, including recurrent tumour infiltration, interferon-induced scleral toxicity, autoimmune scleral necrosis, graft-related complications, or ischemic scleral melt. Comprehensive systemic investigations for autoimmune disease were performed and were non-contributory. Following physician clearance, oral corticosteroid therapy was initiated along with topical antibiotics and corticosteroids.
Despite intensive medical management, the scleral melt progressed, ultimately resulting in recurrent scleral perforation with uveal tissue prolapse and corneal decompensation. At this stage, significant uncertainty remained regarding the underlying etiology of the scleral necrosis and the possibility of residual or recurrent malignancy. Various treatment options, including repeat scleral patch grafting with corneal transplantation, enucleation, and lid-sparing orbital exenteration, were considered. However, the patient declined further surgical intervention and was subsequently lost to follow up.

DISCUSSION

Ocular Surface squamous neoplasia comprises a spectrum of conjunctival and corneal epithelial malignancies and represents the most common ocular surface tumor of epithelial origin. [1,10,11] Although the majority of cases are diagnosed at an early stage and respond favorably to treatment, recurrence remains an important clinical challenge. Recurrence has been associated with increasing age, larger tumor size, positive surgical margins, higher histopathological grade, and inadequate initial treatment. [4,5,8]
The present case illustrates an unusual and aggressive clinical course characterized by recurrent disease, histopathologically proven scleral invasion, progressive scleral melt, and recurrent perforation. The patient had initially undergone excision using a bare sclera technique, after which SCC was diagnosed on histopathological examination. Bare sclera excision has largely fallen out of favor because of its association with increased recurrence rates and ocular surface complications. Contemporary management favors complete tumor excision using a “no-touch” technique combined with cryotherapy and adjunctive topical therapy when indicated. [6,8]
Scleral and intraocular invasion are rare manifestations of OSSN but may significantly worsen prognosis and complicate management. Previous studies have reported intraocular extension in only a small proportion of patients, usually in association with delayed diagnosis, recurrent disease, or inadequate treatment. [2,3,9] Histopathological confirmation of scleral invasion in our patient indicated aggressive local behaviour despite prior intervention and highlighted the need for careful long-term surveillance.
A particularly challenging aspect of this case was the development of progressive scleral melt following surgery and adjunctive interferon alfa-2b therapy. Scleral necrosis after treatment of OSSN may result from multiple mechanisms, including recurrent tumor infiltration, surgically induced ischemia, autoimmune-mediated inflammation, infection, graft-related complications, or medication-associated toxicity. [10,11] Distinguishing among these possibilities can be difficult because clinical findings often overlap and no single feature is pathognomonic.
Tumour recurrence was an important consideration because recurrent SCC may mimic chronic inflammatory or necrotizing scleral disease. However, the absence of clinically evident recurrent tumour and the presence of histologically tumour-free margins following repeat excision argued against obvious residual disease. Autoimmune scleral necrosis was also considered; however, systemic investigations failed to reveal evidence of an underlying autoimmune disorder. Although topical interferon alfa-2b is generally regarded as a safe and effective treatment for OSSN, the temporal relationship between interferon administration and progression of scleral melt in our patient raised the possibility of treatment-related toxicity, although a definitive causal association could not be established. [7,11,12]
The management of advanced OSSN with scleral invasion presents a significant therapeutic dilemma. Globe-preserving procedures such as repeat scleral patch grafting and corneoscleral reconstruction may maintain ocular

integrity but risk undertreating residual invasive disease. Conversely, more radical procedures such as enucleation or lid-sparing exenteration may provide superior oncological control but result in substantial functional and cosmetic morbidity. Similar challenges have been described in advanced OSSN, where treatment decisions must be individualized based on tumor extent, histopathological findings, visual potential, and patient preference. [8,9,12]

CONCLUSION

This case emphasizes the complexity of diagnosing progressive scleral melt in a patient with previously treated OSSN. Histopathologically proven scleral invasion, recurrent perforation, and the inability to definitively distinguish between tumor recurrence and non-neoplastic scleral necrosis created substantial diagnostic and therapeutic uncertainty. Such cases underscore the importance of multidisciplinary evaluation, meticulous follow-up, and individualized management strategies in advanced OSSN.

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