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Case Report *Oculoplasty*

Masquerading malignancy

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ABSTRACT

Conjunctival melanoma is a rare yet aggressive malignancy of the ocular surface, with a global incidence of 0.02-0.5 cases/million annually. We present the case of a 65-year-old patient who exhibited a localized pigmented, bleeding protrusion on the ocular surface, initially resembling an anterior staphyloma. Further examination revealed the lesion originating from the limbus, with the adjacent cornea remaining uninvolved. Excisional biopsy confirmed the diagnosis of conjunctival melanoma. Post-operative recovery was uneventful, with no evidence of recurrence during follow-up. This case underscores the diagnostic challenges associated with atypical ocular lesions and emphasizes the importance of maintaining a high index of suspicion for malignancy. Early diagnosis and intervention are critical to minimizing morbidity and the risk of metastasis.

Keywords: Anterior segment tumors, Atypical presentations of malignancies, Conjunctival melanoma, Ocular malignancy, Rare ocular tumors

INTRODUCTION

Conjunctival melanoma is a rare but formidable ocular malignancy, accounting for approximately 1-2% of all melanomas and 5-10% of all conjunctival tumors. Although the exact etiology remains unclear,[1] several risk factors have been implicated, including chronic ultraviolet (UV) radiation exposure, pre-existing conjunctival nevi, and immunosuppressive states. It predominantly affects adults between the ages of 50 and 70, with a higher incidence in males compared to females. Despite its relatively low prevalence, conjunctival melanoma carries a significant risk of local recurrence and distant metastasis, with reports suggesting that 20-40% of affected individuals may eventually develop metastatic disease, most commonly involving regional lymph nodes.[2]

Given the potential for aggressive behavior and variable clinical presentation, early recognition and intervention are critical. This case is presented due to the rarity of the disease, its potential for atypical presentation, and its high metastatic potential, all of which necessitate increased clinical vigilance and prompt management.

CASE REPORT

A 67-year-old female presented to the ophthalmology department with a progressively enlarging mass in her left ocular surface over a three-month period [Figure 1]. She reported that the mass was initially a small brown lesion that appeared a year prior and gradually increased in size. She also noted associated pain and intermittent bleeding from the lesion's surface.

The patient provided a history of using Ayurvedic medication for cataract treatment 3 months before seeking medical attention. Following the initiation of Ayurvedic treatment, she observed a rapid increase in lesion size. However, there were no complaints of vision loss, double vision, or any features suggestive of ocular infections. There was no history of trauma or previous ocular surgeries.

Examination revealed a well-circumscribed mass, approximately 11 mm × 11 mm in size, which was pigmented, smooth, firm, and bled easily. It appeared to be arising from the inferomedial limbus, at around the 7 o'clock position [Figure 2a]. The cornea remained clear and uninvolved, there was a senile immature cataract of grade 3, and no regional lymphadenopathy was noted at presentation [Figure 2b].

Routine pre-operative investigations were conducted, and an excision biopsy was performed [Figure 3]. The excised tissue sample was sent for histopathological analysis, which revealed tumor cells arranged in sheets with a high nuclear-to-cytoplasmic (N: C) ratio, conspicuous nucleoli, coarse chromatin, and the presence of melanin pigments-findings characteristic of melanoma [Figure 4].

Postoperatively, the patient was initiated on topical mitomycin C (MMC) at a concentration of 0.2 mg/mL to reduce the risk of recurrence. Follow-up at 4 months post-surgery showed no evidence of lesion recurrence or metastasis, and the patient continued to be monitored for long-term disease control.

DISCUSSION

Conjunctival melanoma presents a diagnostic challenge due to its ability to mimic benign and other malignant ocular conditions. The initial presentation in this case closely resembled an anterior staphyloma, which could have delayed appropriate management had further evaluation not been pursued.

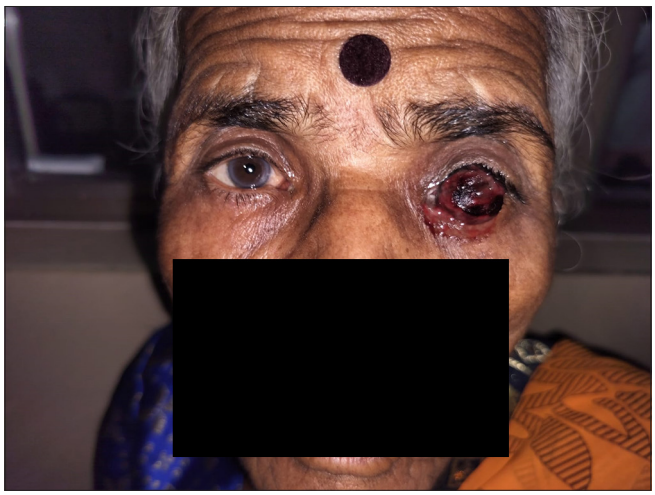

Figure 1: Picture of the left ocular surface showing a conjunctival melanoma which is masquerading an anterior staphyloma.

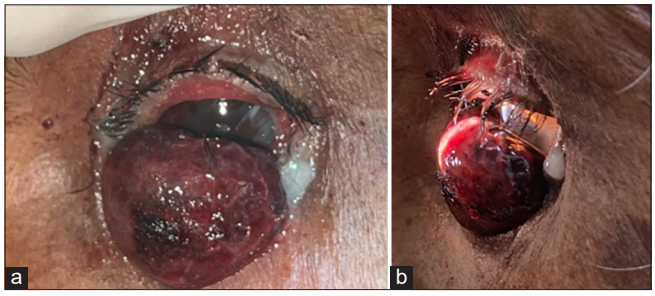

Figure 2: (a) Picture of the left ocular surface with the patient looking down. The uninvolved cornea can be clearly visualized. (b) Picture of the left ocular surface from the side. The melanoma can be clearly seen to be arising from the limbus.

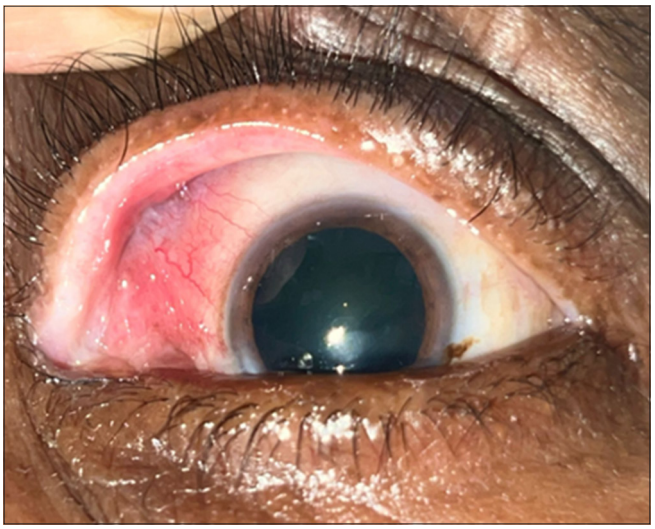

Figure 3: Picture of left ocular surface status post excision biopsy. See the uninvolved clear cornea.

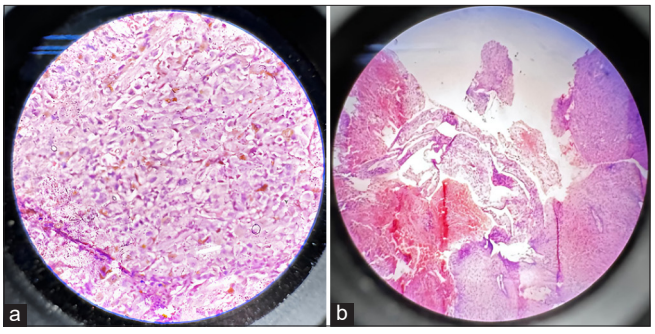

Figure 4: (a) Histopathology: Low magnification image showing tumor cells arranged in sheets (H&E, ×100). (b) Histopathology: High magnification imaging showing tumor cells arranged in sheets with individual cells having a high nucleo-cytoplasmic ratio and dense coarse chromatin suggestive of melanoma (H&E, ×400). H&E: Hematoxylin and eosin.

Histopathological evaluation plays a crucial role in differentiating conjunctival melanoma from other pigmented ocular lesions. While clinical examination can provide significant clues, definitive diagnosis requires histological analysis. Key microscopic features include a high N: C ratio, nuclear pleomorphism, conspicuous nucleoli, and melanin pigment deposits. Immunohistochemical staining can further aid in diagnosis by detecting markers such as HMB-45, S-100, and Melan-A, which are commonly expressed in melanoma cells.

The development of conjunctival melanoma has been linked to several predisposing factors, including UV radiation exposure (similar to cutaneous melanoma), pre-existing nevi (transformation occurring in approximately 20% of cases), and immunosuppression (conditions such as human immunodeficiency virus/acquired immunodeficiency syndrome and post-transplant cases).

Conjunctival melanoma has a high potential for metastasis, primarily spreading through the lymphatic system. The most common sites of metastasis include the regional lymph nodes (preauricular and submandibular nodes), lungs, liver, and brain.

Studies indicate that 20-40% of patients eventually develop metastatic disease, significantly impacting prognosis and survival outcomes. Thus, early detection and intervention are paramount.

Several case reports have described conjunctival melanoma presenting in unusual forms. Shields *et al*.[2] documented a series of 150 cases of conjunctival melanoma, emphasizing the importance of early detection and surgical excision with adjunctive cryotherapy to reduce recurrence rates. Another study by Wong *et al*.[3-5] reported cases where melanomas originated from pre-existing nevi, highlighting the importance of regular monitoring of pigmented ocular lesions. Shildkrot and Wilson[4] reviewed cases of conjunctival melanoma with atypical presentations, noting instances where lesions were misdiagnosed initially, leading to delayed intervention and increased metastatic risk. These reports reinforce the need for heightened clinical suspicion and multidisciplinary management.

The cornerstone of treatment for conjunctival melanoma is complete surgical excision with histopathological margin control. Additional therapeutic strategies include:

Adjuvant Therapy - Topical chemotherapy agents such as MMC are frequently used postoperatively to minimize recurrence. MMC acts as an alkylating agent, inhibiting the deoxyribonucleic acid synthesis in rapidly dividing cells.[5,6]

Cryotherapy - This adjunctive treatment involves freezing the surgical margins to destroy residual tumor cells and reduce recurrence risk

Radiation Therapy - In cases of incomplete excision or high-risk features, brachytherapy or external beam radiotherapy may be considered

Systemic therapy - Advanced or metastatic disease may require systemic immunotherapy or targeted therapy, although such interventions are still being explored in clinical trials.

Prognosis depends on several factors, including tumor size, histopathological features, and the presence of metastasis at diagnosis.[7] Regular follow-up with periodic slit-lamp examination and regional lymph node assessment is crucial in detecting early recurrences. Additional imaging, such as ultrasound biomicroscopy or magnetic resonance imaging, may be employed for deeper tissue evaluation.

In the present case, post-operative use of MMC[8] and vigilant follow-up resulted in no recurrence at four months. However, long-term surveillance remains essential, given the high recurrence rates reported in the literature.

CONCLUSION

This case underscores the diagnostic challenges associated with conjunctival melanoma, particularly when it mimics benign conditions such as anterior staphyloma. A review of previous cases demonstrates the importance of early intervention, multidisciplinary treatment approaches, and long-term follow-up to optimize patient outcomes. Given the high risk of recurrence and metastasis, adjunctive therapies such as MMC and regular monitoring play a crucial role in long-term disease control.

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