

Conjunctival malignant melanoma mimicking scleromalacia perforans: a case report

Mulatu DG¹, Adamu Y², Ayalew M²

¹Ophthalmology Unit, School of Medicine and Health Sciences Wolaita Sodo University, Wolaita Sodo, Ethiopia

²Department of Ophthalmology, School of Medicine, College of Health Sciences Addis Ababa University, Addis Ababa, Ethiopia

Corresponding author: Dr DG Mulatu, Ophthalmology Unit, School of Medicine and Health Sciences, Wolaita Sodo University, Wolaita Sodo, Ethiopia. Email: findgetaneh@gmail.com

ABSTRACT

Malignant melanoma of the conjunctiva is a relatively infrequent neoplasm that can be associated with significant morbidity and cause diagnostic difficulty to both the ophthalmologist and pathologist. We herein describe a case of malignant conjunctival melanoma which clinically simulated scleromalacia perforans causing sclerouveal staphyloma or huge conjunctival cyst. Being rare but potentially lethal tumour, conjunctival melanoma should be included in the differential diagnosis of slowly growing conjunctival masses or sclerouveal staphylomas for early detection and management.

Key words: Malignant melanoma, Scleromalacia perforans, Staphyloma, Conjunctival cyst

INTRODUCTION

Conjunctival melanoma is a relatively rare condition, occurring only 1/40th as often as choroidal melanoma and approximately 500 times less often than cutaneous melanoma^{1,2}. Its incidence is 0.2 to 0.8 per million in white populations and it tends to present in adulthood with the median age at diagnosis being 60 years and with no sexual predilection¹⁻⁴. Conjunctival melanoma is a potentially lethal neoplasm, with an average 10 year mortality rate of 30%¹. It can arise *denovo* (12%), from an existing nevus (20-30%), or more frequently from an area of primary acquired melanosis (75%)^{5,6}. The lesion is identified most frequently in the peri-limbal interpalpebral bulbar conjunctiva. Tumours located in the palpebral, forniceal and caruncular conjunctival regions are infrequent and are associated with poor prognosis^{1,7}.

Classically, conjunctival melanoma presents as a darkly pigmented mass of variable duration (often short) in the interpalpebral region of the bulbar surface that would cause little diagnostic difficulty for the ophthalmologist⁷.

However, other several lesions may simulate conjunctival melanoma including extraocular extension of uveal malignant melanoma, metastatic malignant melanoma to the conjunctiva (usually from a cutaneous origin), melanosis oculi and oculodermal melanocytosis

(nevus of Ota), sclerouveal staphyloma, foreign body, polymerized epinephrine, nerve loop axenfeld, pigmented epithelial tumours including papilloma, pigmented pinguecula and ocular surface squamous neoplasia^{5,8}.

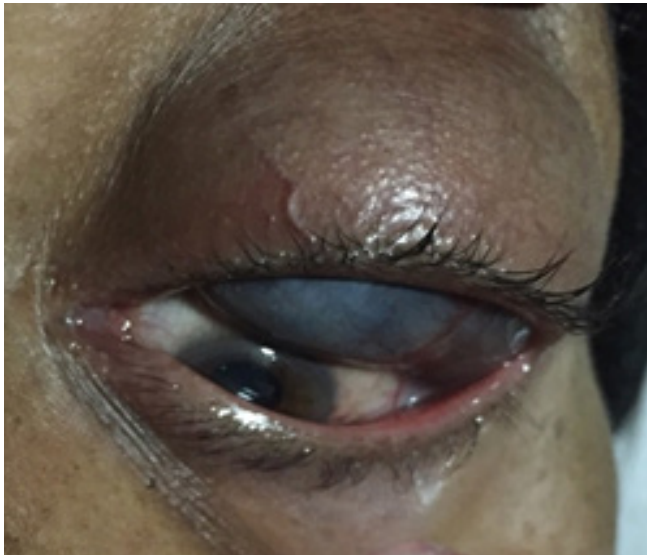
We describe a case of malignant conjunctival melanoma which simulated scleromalacia perforans with sclerouveal staphyloma.

CASE REPORT

A 50 year old female from the rural part of Arsi, Oromia region in Ethiopia, presented with a painless mass on the left eye of 5 years duration which progressively enlarged in size. She also complained of markedly reduced vision but denied any history of redness, ocular trauma or known systemic illnesses like diabetes, hypertension or rheumatoid arthritis.

On examination her vision on the left eye was reduced to counting fingers at 1meter. There was a bluish, cystic, fluctuant non-tender mass over the supero-temporal area of the orbit, filling the palpebral fissure and displacing the globe infero nasally. No pulsations or bruit were heard over the mass and its posterior limit was not visible (Figure 1). There was no significant lymphadenopathy.

A



B

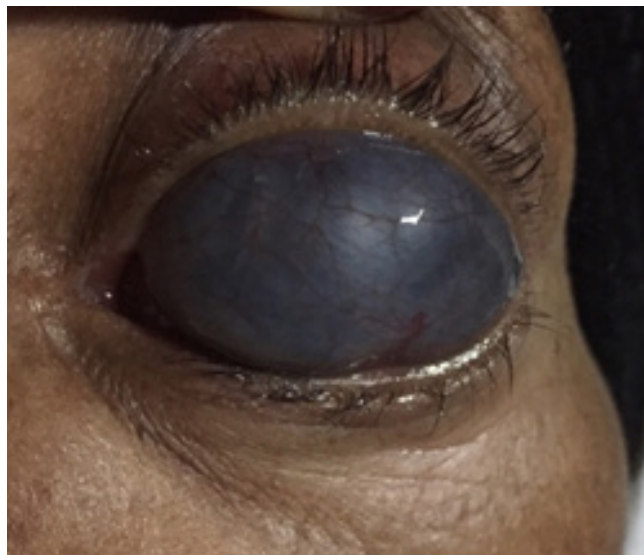


Figure 1: A & B – mass on the left eye

Slit lamp biomicroscope examination of the anterior segment was normal. Examination of the fundus using slit lamp biomicroscope and 90D Volk lens revealed diffuse chorioretinal scar involving the inferior half of the retina and macula (Figure 2).

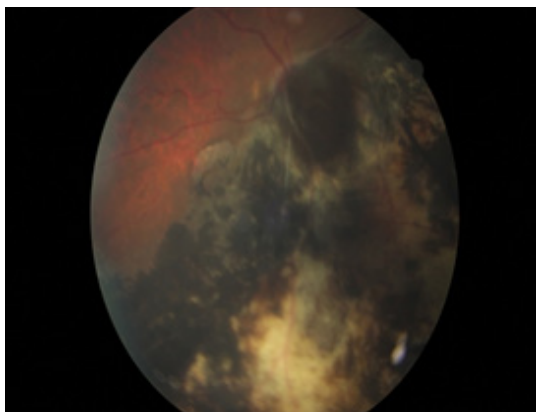


Figure 2: Fundus picture of the left eye

With a presumed diagnosis of scleromalacia perforans resulting in sclerouveal staphyloma, the patient was investigated with blood tests including Complete Blood Count (CBC); serologic tests including Rheumatoid Factor (RF), Venereal Disease Research Laboratory (VDRL), serology test for HIV 1 and HIV 2; organ function tests including liver and renal function tests all of which turned out to be normal. Chest X-ray and abdomino-pelvic ultrasound were also normal.

Ocular ultrasound showed a hypoechoic mass over the supero-temporal area of the left orbit compressing the globe but with questionable communication with the globe. Orbital CT scan showed heterogeneous orbital mass involving the supero-temporal orbit and had little enhancement with contrast.

A



B



Figure 3: CT scan of the orbit and brain, coronal section (A) & sagittal section (B)

The case was presented to attending staff and residents at the Department of Ophthalmology, Addis Ababa University as scleromalacia perforans. Upon discussion, the diagnosis was challenged and agreed to be in need of further work-up including Cybersight® consultation. Doppler ultrasound was done and showed a highly vascularized mass with no communication to the globe.

The patient was subjected to surgical exploration for possible pathologic diagnosis by either excisional or incisional biopsy by an orbital surgeon. Intraoperative, a double walled cystic, dark brown vascularized mass was identified, and the posterior limit of the mass was unreachable. Incisional biopsy was done and specimen was sent for pathologic evaluation (Figure 4).

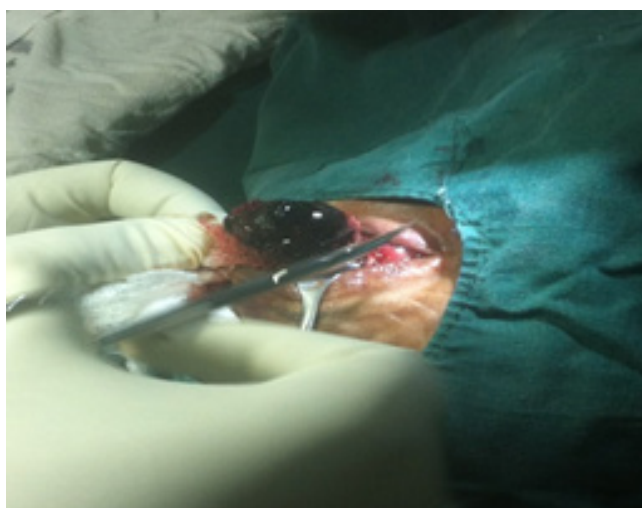


Figure 4: Excision of mass

Pathologic section showed well-delineated tissue with solid sheets of discohesive cells having vesicular nuclei, prominent eosinophilic nucleoli and moderate cytoplasm containing dark brownish pigment which was suggestive of the diagnosis of malignant melanoma of the left conjunctiva (Figure 5).

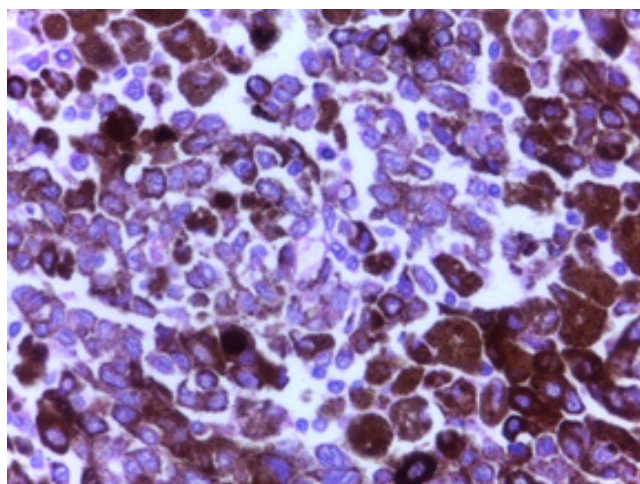


Figure 5: Histologic section



Figure 6: 1 month post operation

Post-operatively the patient was followed at the oculoplasty clinic, counseled for possible subsequent exenteration of the left orbit and also referred to the oncology department for secondary opinion and follow up (Figure 6).

DISCUSSION

Given the nature and the duration of the lesion, no malignant condition was initially considered as a differential diagnosis. Instead, the possibility of scleromalacia perforans causing sclerouveal staphyloma or huge conjunctival cyst was entertained. The diagnosis of conjunctival malignant melanoma confirmed with the histopathologic study.

Conjunctival melanoma is a relatively rare condition carrying high lethal potential¹⁻⁴. It spreads through the lymphatics, primarily to the preauricular lymph nodes and sequentially to the submandibular and cervical nodes. Excisional biopsy is the only way to differentiate melanoma from a benign lesion⁴.

The primary treatment is surgical excision of the entire tumour with wide surgical margins of 3-5mm. Exenteration is indicated once the orbit is involved. The prognosis is poor and life expectancy is short for those associated with regional lymph node metastases^{9,10}.

CONCLUSION

Although conjunctival melanoma is a rare tumour, it should be included in the differential diagnosis of slowly growing conjunctival masses or sclerouveal staphylomas. Early detection and treatment is crucial as conjunctival malignant melanoma is associated with a high mortality and metastases rate.

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