Case Report

A rare case of conjunctival malignant melanoma with orbital invasion

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Abstract

Background: Conjunctival malignant melanoma is a rare ocular malignancy most commonly affecting mostly the elderly population. It is a pigmented lesion which can arise from primary acquired melanosis, de novo or from preexisting nevus.
Case: A 63 year old male presented with a chief complaints rapidly increasing mass in the left eye for two months following trauma with a wooden stick. He had preexisting nevus in the same eye. His best corrected visual acuity was 6/12 in right eye and 1/60 left eye respectively. Slit lamp biomicroscopy examination showed 4 cm x 4 cm pigmented mass in the nasal bulbar and inferior palpebral conjunctiva causing mechanical ectropion of the lower lid with keratinization of palpebral conjunctiva. Incisional biopsy of conjunctiva showed malignant melanoma. On computed tomography, there was expansion of bony orbit. Considering all the findings, exenteration of the left orbit was done. Histopathological report of exenterated mass was suggestive of malignant melanoma of conjunctiva.
Conclusion: Conjunctival melanoma is a rare malignant tumor of eye which has high metastasis rate and the treatment option is surgery with adjuvant therapy.

Key words: Conjunctival malignant melanoma, De novo, Incisional biopsy

Introduction

Conjunctival malignant melanoma is a rare pigmented lesion of the ocular surface with high recurrence. There is direct extension to the globe and orbit in local metastasis cases (H. GökmenSoysal et al. 2008). Mostly the tumor is unilateral. It comprises about 2% of all eye tumors and 5% of melanomas in the ocular region. It arises from primary acquired melanosis (57-76%), de novo (16-25%) and from nevi (1-6%) (Shields CL et al. 2011). Most common site of presentation is bulbar conjunctiva but may involve other parts including limbal, forniceal, palpebral conjunctiva, and/or cornea (Shields CL et al. 2009). The distant metastasis is mostly seen in lungs, skin, liver, and brain. Orbital extension of the conjunctival melanoma is less common comprising about 2.6% (H. GökmenSoysal et al. 2008). The conjunctival melanoma can be treated with surgical excision, excision

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combined with cryotherapy, radiotherapy, and topical chemotherapy (Salazar Méndez R et al. 2012). The recurrence rate of 50% and tumor mortality rate of 32% is reported (Anastassiou G et al. 2002).

Case report

A 63 years old male presented to OPD with the chief complaints of a rapidly increasing mass in his left eye. He had a small elevated reddish lesion in the inner white part since two years which suddenly increased to the present size since last two months following trauma with a wooden stick. On examination, the best corrected visual acuity in the right eye was 6/12 while 1/60 in the left eye on Snellen's chart. There was presence of pigmented mass of about 4 cm x 4 cm in the nasal bulbar and inferior palpebral conjunctiva of left eye causing mechanical ectropion of the lower lid with keratinized surface. The mass was firm in consistency with the central area of necrosis (Fig: 1). There was proptosis with the eyeball displaced superotemporally with restriction of extraocular motility in all cardinal gazes. Pterygium was present nasally which was 3mm from the limbus. Cornea was clear without any pigmentation (Fig: 2). Anterior chamber was normal in depth and quiet with nuclear sclerosis grade II. The fundal glow was present in the left eye. The regional lymph nodes were not palpable. Right eye examination was normal except nuclear sclerotic cataract grade II. The intraocular pressure was 13 mmHg in the right eye while it was digitally soft in the left eye.

USG B-scan of the left eye showed the ocular coats were intact with no intraocular extension of the mass but the eyeball was tented inferiorly most probably due to the mass effect (Fig: 3). CT-scan of the head and orbit showed homogenously enhancing soft tissue lesion about 6.3x 4.3x 4.3 cm in the superomedial, inferomedial and inferior aspect of left orbit. There was expansion of bony orbit, thinning of inferior and medial wall. The eyeball was compressed and displaced anterolateral (Fig: 4).

Hematological investigation, USG of abdomen and Chest X-ray were within normal limit. Incisional biopsy of the conjunctival mass was taken which was suggestive of malignant melanoma (Fig: 5). Exenteration of the left orbit was done. The exenterated eyeball with the mass was sent for histopathological examination that revealed conjunctival malignant melanoma with AJCC staging of pT3Nx **(Fig: 6).** He was referred to the oncology department but didn't follow up.

Figure 1: Left eye showing proptosis with pigmented mass causing mechanical ectropion and keratisation of palprebral conjunctiva.

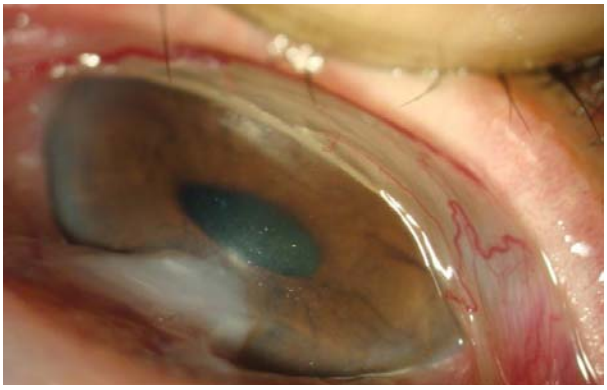

Figure 2: Left eye showing pterygium nasally with clear cornea.

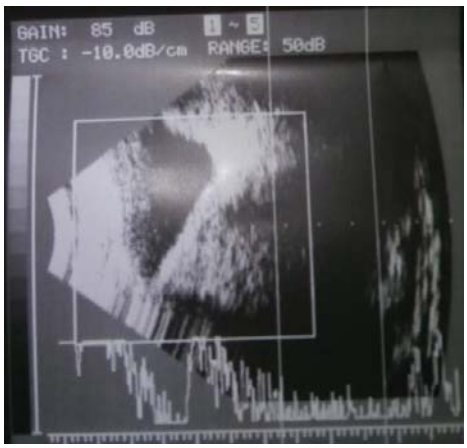

Figure 3: USG B Scan of left eye showsinferioly tented eye ball mainly due to mass effect with no intraocular extension.

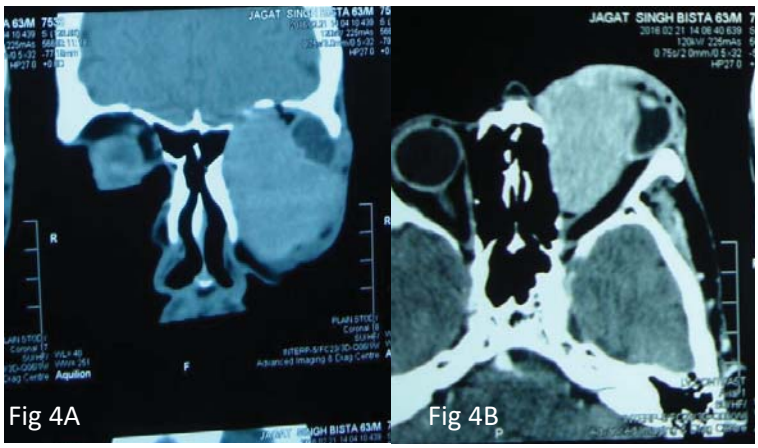

Figure 4A: CT scan coronal veiw of left orbit showed homogenously enhancing soft tissue lesion about 6.3 x 4.3 x 4.3 cm in the superomedial, inferomedial and inferior aspect with expansion of bony orbit. Figure 4B: CT scan axial view of left orbit showed homogenous mass compressing and displacing the eyeball anterolaterally with thinning of medial wall

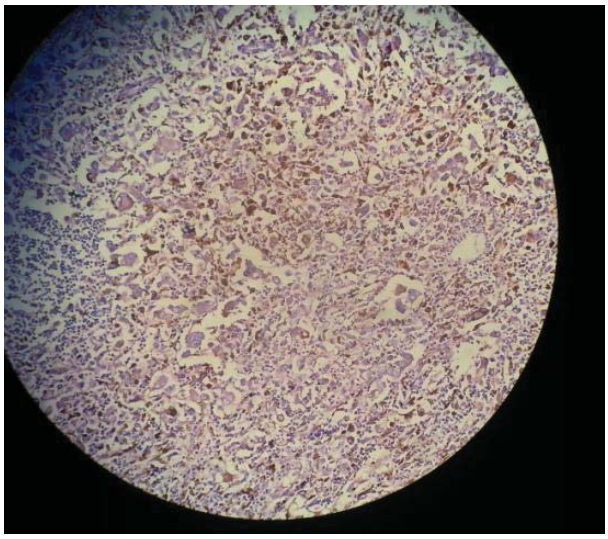

Figure 5: Incisional biopsy of conjunctival mass showed scattered tumor cells with enlarged round to oval hyperchromatic nuclei having prominent eosinophilic nucleoli with moderate amount of cytoplasm withdense intracytoplasmic melanin pigments suggestive of malignant melanoma of conjunctiva.

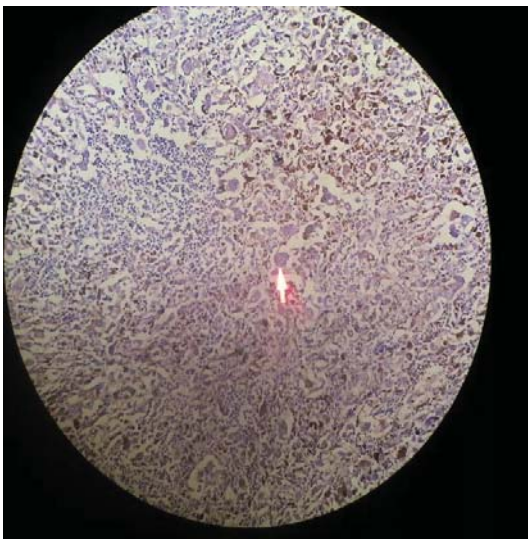

Figure 6: Histopathology of exenterated mass – arrow indicates melanoma cell suggestive of malignant melanoma of conjunctiva.

Discussion

Conjunctiva malignant melanoma is uncommon tumor, accounting for less than 2% of ocular melanomas and less than 1% of malignant tumor of the eye (Scotto J et al. 1976). Most commonly it arises from PAM (57-76%) (Sheilds CL et al. 2011) but in our patient it developed from pre-existing nevus following trauma with a wooden stick. Most common site of presentation is interpalpebral region of bulbar conjunctiva same as in our case but may involve other parts including limbal, forniceal, palpebral conjunctiva, and/or cornea.The conjunctival malignant melanoma usually spread first through lymphatic drainage (Sheilds CL et al. 2011) and invade the secondary structures through direct extension or hematogenous spread (Seregard S. et al. 1998). Most common distant metastasis is seen in the lungs, skin, liver, and brain.The tumor may extend to the secondary structures like orbit (2%), eyelids (1%), paranasal sinuses (1-5%) (Sheilds CL et al. 2011). The regional preauricular and submandibular lymph nodes were not palpable (Browstein S. 2004) and no distant metastasis was present in our case. CT scan of brain and orbit showed expansion of the bony orbit with thinning of inferior and medial wall of left orbit. Intra-operatively the orbital roof, maxillary sinus and the ethmoidal sinus and inferior orbital wall were thinned out. Because of the massive lesion (4 cm x 4 cm), incisional biopsy was performed which showed conjunctival malignant melanoma. Because of chances of seeding of tumor cells incisional biopsy should not be performed. (Shields CL. 2000). So a wide excisional biopsy is preferred. Because of the massive lesion in our patient, incisional biopsy was performed for the diagnosis (Jovanovic P et al. 2013). Enucleation and exenteration should be performed only in cases of diffuse conjunctival melanoma because of disfigurement, blindness and no improvement in survival rate (Paridaens AD et al. 1994). Incisional biopsy was suggestive of malignant melanoma of conjunctiva and CT scan showed homogenously enhancing soft tissue lesion in the superomedial, inferomedial and inferior aspect of left orbit with expansion of bony orbit. With above findings, exenteration of the left orbit was done.The exenterated eyeball with the mass was sent for histopathological examination that revealed conjunctival malignant melanoma with AJCC staging of pT3Nx. Patient was referred to oncologist but he didn't follow up.

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