



PTOSIS MASKING MALIGNANT MELANOMA OF CHOROID

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ABSTRACT

Malignant melanoma or melanocarcinoma arising from melanocytes is one of the most rapidly spreading malignant tumour of the skin that can occur at all ages but is rare before puberty. Malignant melanoma is a relatively common neoplasm that not long ago was considered almost uniformly deadly. The great preponderance of melanomas arises in the skin; other sites of origin include the oral, anogenital mucosal surfaces, esophagus and meninges. Eyeball is an very uncommon site for Malignant melanoma, here we present a case of Malignant melanoma of choroid - left eye which is a rare presentation.

INTRODUCTION

Melanoma originates from melanocytes, which arise from neural crest and migrate, to uvea, meninges, epidermis and ectodermal mucosa. Melanomas may develop in healthy appearing skin or near a previously existing precursor lesion. A malignant melanoma developing in healthy skin is said to arise de novo. Many of the melanomas are induced by solar irradiation. The greatest risk of sun exposure induced melanoma is associated with acute, intense and intermittent blistering sunburns. The risk is different from squamous and basal cell skin cancers, which are associated with, prolonged long term sun exposure. Certain lesions are considered precursor lesions of melanoma including dysplastic nevus, common acquired nevus, congenital nevus and cellular blue nevus. Here we present a case of malignant melanoma of choroid of left eye in a sixty five years female patient which is a rare presentation.

CASE REPORT

A 65 years female, house wife by occupation came with complaints of drooping of left eyelid from past three years, diminished vision in left eye since three months and pain in left eye since two days. Patient was apparently normal three years back then she developed drooping of both upper eye lids, which was more in the left eye. It gradually progressed to present stage. [Figure 1]

Then she developed gradual loss of vision in left eye three months back which was progressive in nature since two days she is complaining of pain in the left eye which is dragging type and radiating to the left side of face. No history of redness, watering, nausea and vomiting. There is no history of trauma also. Patient is a known diabetic and hypertensive from past three years. There is no significant family history, drug history or any previous surgeries. Head posture was normal. Facial symmetry was maintained but ocular symmetry is lost due to ptosis. Hirschberg Corneal Reflex Test (HCRT) on lifting the left eye lid shows exotropia > 45degree [Figure 2].

Fundoscopy examination

Right eye was normal whereas left eye on direct and indirect ophthalmoscopy show hazily seen due to nuclear sclerosis. Greyish white reflex seen in the pupillary area on the temporal side suggestive of retinal detachment, adjacent to the reflex a black mass is seen. The extent of the margins could not be appreciated [Figure 3]. Left eye B scan show a mushroom shaped mass of about 14mm and 18mm in size arising from the posterior choroid extending upto the anterior vitreous with secondary retinal detachment. The mass suggestive of choroidal melanoma [Figure 4]. Provisional diagnosis of right eye grade 1 NS with mild ptosis and left eye melanoma of choroid with



secondary retinal detachment with complete ptosis with immature cataract or Retinoblastoma was made.

Enucleation of the left eye was done under local anesthesia and sent for biopsy.

Macroscopic examination:- eye ball measuring 2.5cms, 2cms, and 2cms. Cut section shows dark brown to black area, firm to hard in consistency [Figure 5a].

Microscopically show sclera consisting of collagen tissue and a neoplastic lesion with no defined pattern. The cells are round to polygonal are in diffuse sheets, aggregates and compact masses interepted by collagen tissue, cytoplasm is moderate with large vesicular nucleus contains prominent inclusion like nucleoli. There is prominent intra cytoplasmic and extra cellular melanin pigment identified [Figure 5b]. Foci of macrophages containing melanin pigment seen. At places, ciliary process showing double layer of pigmented and non-pigmented epithelial cells seen.

Retina very well appreciated and contains ganglion cells, internal nuclear layer, external nuclear layer and choroid plexus. The neoplastic lesion extended upto retina but not beyond the retina. There is focal angio invasion by tumor cells seen in the tumor tissue. The cornea differentiation was not made out. Retinal vasculature was not involved by tumor tissue. We diagnosed it as malignant melanoma of choroid.

DISCUSSION AND CONCLUSION

Malignant melanoma is a neoplasm of melanocytes or of the cells that develop from melanocytes. Once considered uncommon disease the annual incidence is dramatically increasing over last few decades. Early stage melanoma is curable but if melanoma has metastasized prognosis is grim with median survival of 6-9 months. Prognosis is also related to type of melanoma [2]. The general incidence of malignant melanoma of the eye

as per Duke Elder is 0.02 to 0.06% of all patients i.e. from 2 to 6 per 10,000; with regard to the part of the eye affected, the vast majority occurs in the choroid of about 85%². Only one in 20 is situated within the conjunctiva [3]. Amongst the non-invasive procedures, in addition to direct ophthalmoscopic visualization, ultrasonography remains an important diagnostic tool. The hallmark [4] of malignant melanoma on A and B scan is low to medium reflectivity of its inner tumor spikes at tissue sensitivity (5-60% in small and up to 80% in large tumors) combined with solid consistency (no after movement of the surface tissue spikes during kinetic echography) and regular structure. A number of ocular lesions or pseudomelanomas can clinically simulate malignant melanomas like aphakic cyst of ciliary body, melanocytoma of ciliary body, tumours of non-pigmented ciliary epithelium, conjunctival nevi and melanoma and ciliary staphyloma [5].

Malignant melanomas spread [6] by direct extension, local metastasis, or generalised metastasis. Current management can range from periodic observation and fundus photography of selected small lesions that appear dormant to photocoagulation, radiotherapy, or local resections that is, iridocyclectomy in case of growing tumors in eyes with useful or salvageable vision. In cases where the tumor is far advanced and there is no hope for useful vision, enucleation is often inevitable as it was done in our case. Prognosis also depends on the type of melanoma as it is 5 year survival rate in epitheloid type.

The role of postoperatively adjuvant chemotherapy or radiotherapy either singly or in combination is not known. Radiotherapy has been tried in mucosal melanoma of the head and neck with disappointing results. Chemotherapy has been used mainly for palliation only. To conclude it is very important to follow elderly patients with ptosis regularly even in absence of any complaints.

Table 1. Showing Right and Left eye examinations

Right Eye	Left Eye
Vision – 6/18	NOPL
BCVA – 6/12	NOPL
NV – N/18	Absent
Colour vision - Normal	Absent
Ptosis present upto pupillary margin	Complete
Palpebral fissure – 8mm	1mm
Levator function - poor	Absent
Eyelid crease - present	Absent
MRD1 – 2 mm	0
MRD2 – 6mm	0
Lid margin - Normal	Normal
Lacrimal apparatus - Normal	Normal
Conjunctiva - Normal	Normal
Eyeball movements - Normal	Normal

NOPL –No Perception of Light ,BCVA –Best Corrected Visual Acuity,NV –Near Vision,MRD- Margin Reflex Distance.



Table 2. Showing Right and Left eye examinations

Right Eye	Left Eye
Cornea - Normal	Normal
Anterior chamber - Normal	Normal
IRIS - normal	Normal
Pupil shape - circular reacting to light	Greyish mass seen in the papillary area posterolateral to the lens
Lens -NS grade 1	NS grade 2
IOP - 14mm Hg	14mm Hg

NS- Nuclear Sclerosis,IOP-Intra Ocular Pressure.

Figure 1. Photograph showing drooping of both eyelids which is more on left side, with normal facial symmetry



Figure 2. Photograph showing various ocular movements of both eyes



Figure 3. Fundoscopy of right eye [Figure a] and left eye [Figure b]

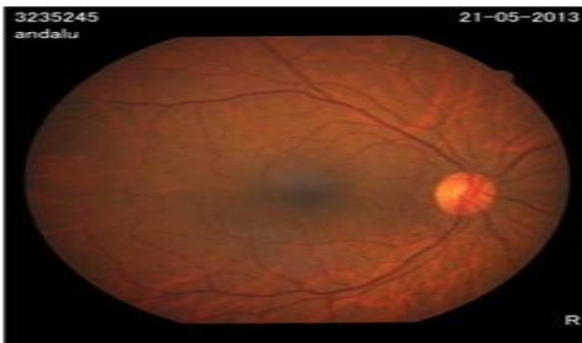


Figure 4. Left eye B scan show a mushroom shaped mass of about 14mm and 18mm in size arising from the posterior choroid extending upto the anterior vitreous with secondary retinal detachment

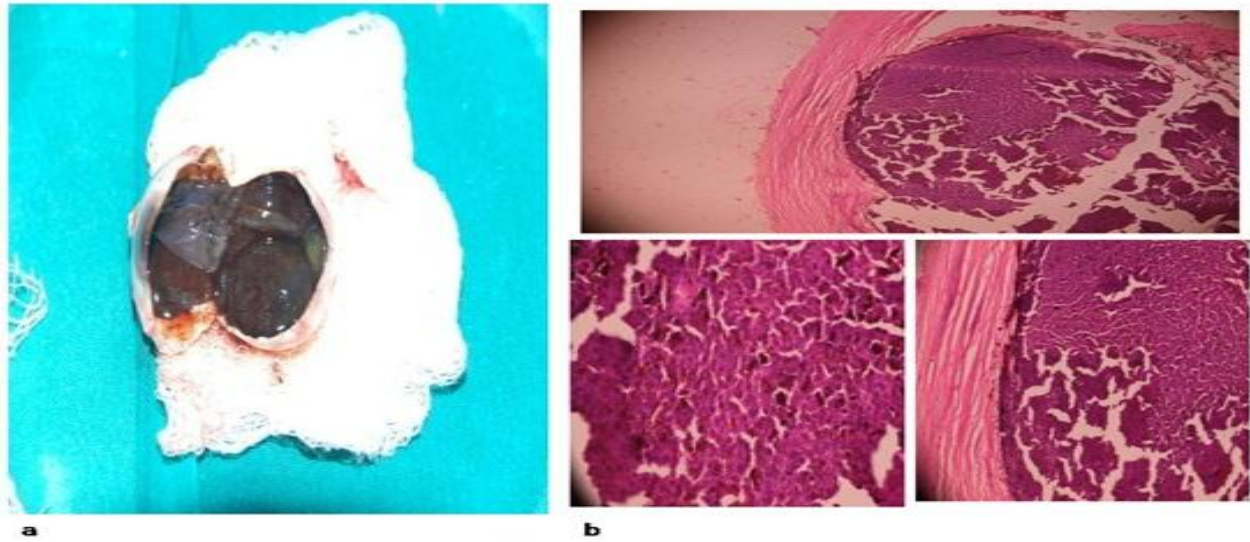


Figure 5. Gross photograph of the tumour [Figure a].Microscopic picture showing tumour cells along with melanin pigment [Figure b]



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