

CASE REPORT

CHOROIDAL MELANOMA WITH SECONDARY GLAUCOMA

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Abstract. The case of a 40-year-old female patient with choroidal melanoma with secondary glaucoma presenting as a painful blind right eye is reported. Liver metastasis was detected by ultrasonography. The choroidal tumor measured 2 x 2.1 x 1.5 cm; histopathology showed that it was of the spindle cell (spindle A) variety. Such tumors are rare in non-white races and secondary glaucoma is an uncommon presentation.

Choroidal melanoma is rare in non-white races. Only two reports have been documented in India and none presented with glaucoma (Manohar *et al*, 1991; Biswas *et al*, 2000). Only 2% of eyes affected by choroidal melanoma develop secondary glaucoma (Shields *et al*, 1987). To the best of our knowledge this is the first case report from the Indian subcontinent of choroidal melanoma presenting with secondary glaucoma.

A 40-year-old female presented with a one-month history of right eye ache and headache; she also gave a ten-day history of redness of the right eye. She had been seen one week earlier by another ophthalmologist who had prescribed topical timolol 0.5%, flurbiprofen drops and oral acetazolamide tablets - a regimen that had failed to relieve the patients symptoms.

On examination, visual acuity was no perception of light (NPL) in the right eye and 6/9 in the left eye; the visual acuity of the left eye after refraction was 6/6. The right eye had circumcorneal congestion, a very shallow anterior chamber with +2 flare and +1 cells, a fixed, dilated, non-reacting pupil with a clear lens. A brownish mass in the retrolental space (nasally) and a detached retina were shown by slit lamp

examination. Indirect ophthalmoscopy revealed a brownish, elevated, oval-shaped mass nasally and exudative retinal detachment. Ocular examination of the left eye revealed no abnormalities. The intraocular pressure in the right eye was 50 mmHg; in the left eye it was 17 mmHg.

B-scan ultrasonography showed a large dome-shaped mass with acoustic hollowness, choroidal excavation and orbital shadowing, along with retinal detachment. A-scan revealed low to medium reflectivity, negative angle kappa and spontaneous vascular movements in the intraocular mass. A chest X-ray was normal. Ultrasonography of the liver showed metastasis.

A clinical diagnosis of malignant melanoma of the right choroid with secondary glaucoma and metastasis to the liver was made. An explanation of the prognosis of malignant melanoma was given to the patient. Enucleation of the eye was subsequently performed. The post-operative period was uneventful and the patient was referred to the Bharatpur Cancer Hospital for chemotherapy.

Dissection of the enucleated eye revealed a uniformly black tumor, measuring 2.0 x 2.1 x 1.5 cm, and localized retinal detachment (Fig 1). Histologic sections showed a heavily pigmented tumor, with black pigment involving the angle region. Melanin bleach with potassium permanganate showed that the cells

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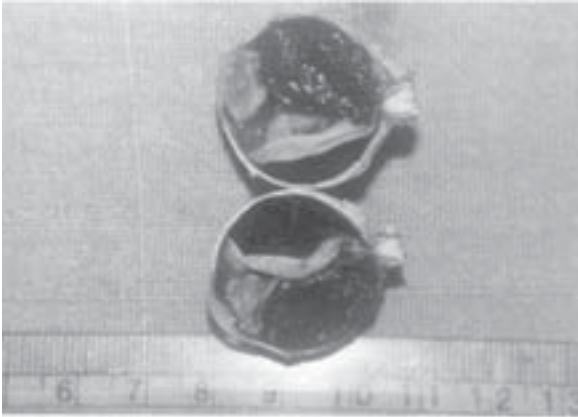


Fig 1—Section of the eyeball showing a dome-shaped dark colored tumor occupying the vitreous cavity; note the associated retinal detachment.

were arranged in a fascicular pattern. The cells were of a cohesive spindle A type, having poorly defined cell borders, small spindle nuclei with a dark stripe but no distinct nucleoli and only occasional mitoses. There was no invasion of the Schlemm canals or vortex veins; the resected end of the optic nerve was also free of tumor.

Though choroidal melanoma is the commonest primary intraocular malignancy, it has a very low incidence, only 6 cases per million population per year, in the US. Moreover, black patients have less than eight times the risk of developing disease compared with white patients. The ratio of white patients to dark skinned patients who have this neoplasm is over 50:1 (Seddon *et al*, 1989).

In most cases, the median age at diagnosis is about 55 years, and there is slight preponderance of males (Seddon *et al*, 1989). In New England, rates were uniformly higher in males, except among those aged 20-39 years, whose rates were 2.3 times higher in females (Egan *et al*, 1987).

In a survey of 2,111 eyes in Philadelphia, it was found that only 2% of the eyes with choroidal melanoma had secondary glaucoma at the time of diagnosis (Shields *et al*, 1987). The commonest mechanism in the cases of choroidal melanoma was neovascularization of the

iris (Shields *et al*, 1987). The second most common mechanism of secondary glaucoma is anterior displacement of the lens iris diaphragm by the tumor mass, with the development of PAS and closed-angle glaucoma (Spierer *et al*, 1983; Shields *et al*, 1987): our case had glaucoma caused by this mechanism. Other mechanisms that can lead to glaucoma are the direct extension of the tumor into the trabecular meshwork, the seeding of tumor cells into outflow channels, pigment dispersion, inflammation, hemorrhage inducing hemolytic glaucoma, and melanolytic glaucoma.

Uveal melanoma can lead to visual loss and death. Studies of patients treated for uveal melanoma have shown tumor-related mortality rates of the order of 50% within 10-15 years after enucleation. Survival from the time of the development of systemic metastasis ranges between one and thirty-one months (median 7 months), with a one-year survival rate of 29% (Bedikian *et al*, 1981). The liver is the commonest site of metastasis.

The four important factors affecting the prognosis of choroidal malignant melanoma are size, cell type, scleral extension, and mitotic activity (Yanoff and Fine, 1996). Malignant melanomas over 1 cm³ have a poor prognosis. Slightly less than 50% of ciliary body and choroidal malignant melanomas are of the spindle cell variety: these have a better prognosis than the non-spindle cell variety (epithelial, mixed or necrotic). In our patient the tumor measured more than 1 cm³ and was of the spindle A type with occasional mitoses and no scleral extension.

Cases of primary choroidal malignant melanoma presenting with secondary glaucoma in non white patients are extremely rare. This case is all the more interesting, given its rarity and its unusual clinical presentation.

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