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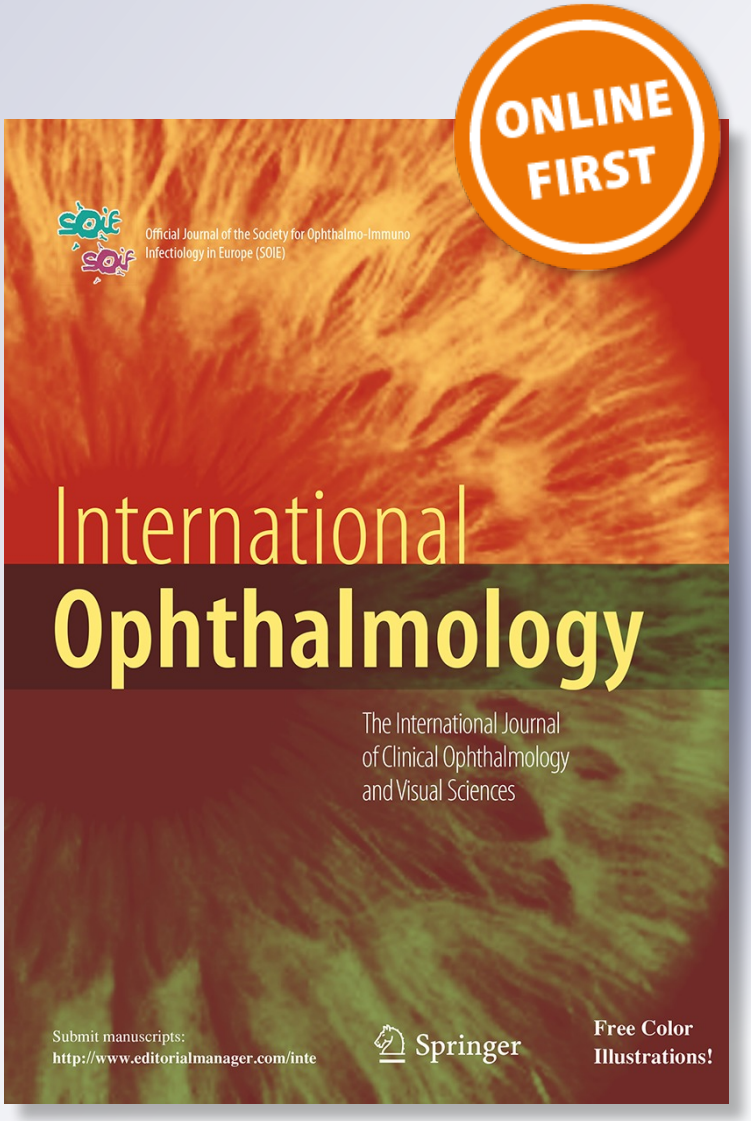

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CASE REPORT

Intraocular malignant melanoma of the choroid presenting as orbital cellulitis

Akshay Gopinathan Nair · Swathi Kaliki · Mohammed Javed Ali · Milind N. Naik · Geeta K. Vemuganti

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Abstract We report a case of choroidal melanoma with features suggestive of orbital cellulitis. A 24-year-old Asian Indian male presented with a 20-day history of sudden loss of vision in the right eye. Edematous eyelids with complete mechanical ptosis, complete ophthalmoplegia, gross proptosis accompanied by massive chemosis, and prolapse of the inferior forniceal conjunctiva were noted. He denied perception of light in the right eye. The left eye was unremarkable. B-scan ultrasonography of the right eye showed a large dome-shaped mass filling the posterior segment suggestive of choroidal melanoma. Computed tomography confirmed those findings and showed no extraocular tumor extension. The patient was conservatively treated with systemic steroids following which the inflammation subsided. He underwent enucleation of the right eye and a diagnosis of spindle A cell choroidal melanoma was confirmed by histopathological examination. Although rare, orbital cellulitis is one of the presenting features of choroidal melanoma. Uveal melanoma-associated orbital cellulitis can be well controlled with systemic steroids, does not always indicate extraocular tumor extension, and can occur in spindle A cell melanomas.

Keywords Malignant melanoma · Choroidal melanoma · Orbital cellulitis · Enucleation

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A 24-year-old Asian Indian male presented with a 20-day history of sudden loss of vision in the right eye. 5 days prior to examination, he had developed painful, progressive proptosis of the right eye. On visual acuity testing, the patient denied perception of light in the right eye and the vision was 20/20 N6 in the left eye. While the left eye was normal, examination of the right eye revealed edematous eyelids with complete mechanical ptosis (Fig. 1), complete ophthalmoplegia, gross proptosis accompanied by massive chemosis, and prolapse of the inferior forniceal conjunctiva. Hyphema precluded any view of the anterior and posterior segments of the right eye. Intraocular pressure in the right eye was 36 mmHg. B-scan ultrasonography of the right eye disclosed an acoustically dense dome-shaped mass measuring $16 \times 14 \times 15$ mm^3 filling the posterior segment with choroidal excavation and overlying total retinal detachment (Fig. 2c). A-scan ultrasonography of the mass revealed low to medium internal reflectivity. Based on these findings, a clinical diagnosis of necrotic choroidal melanoma with secondary aseptic orbital cellulitis in the right eye was established. In

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Fig. 1 A 24-year-old male with eyelid edema, complete mechanical ptosis, proptosis, chemosis and conjunctival prolapse of the right eye at presentation (**a**, **b**)

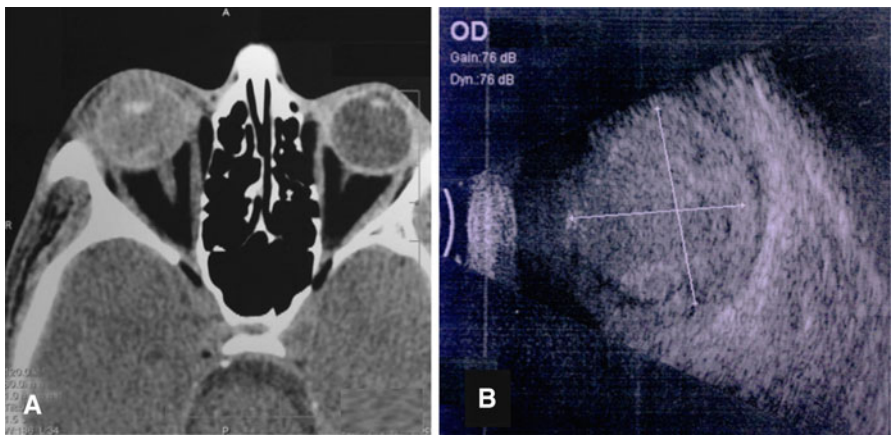

Fig. 2 Axial section of computed tomography orbit performed after 3-day course of systemic steroids showing crystalline lens dislocation and a moderately hyperdense mass with irregular margins occupying two-thirds of the posterior segment with no obvious extraocular tumor extension (**a**). B-scan ultrasonography showing an acoustically dense dome-shaped mass filling the posterior segment (**b**)

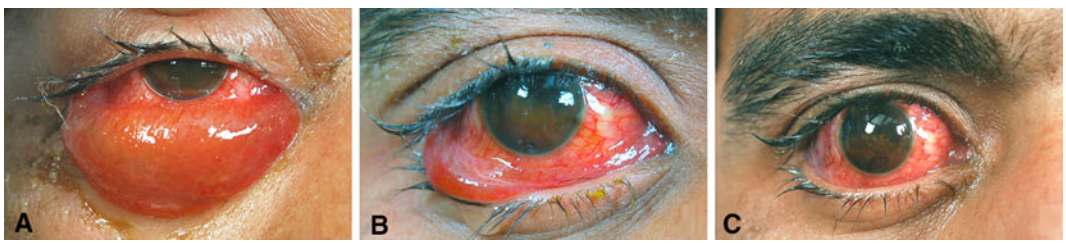

Fig. 3 Eyelid edema, inferior conjunctival chemosis, severe congestion, forniceal prolapse, and hyphema of the right eye at presentation (**a**). After 2 days (**b**) and 3 days (**c**) of systemic steroids showing good response to conservative treatment with near complete resolution of inflammation

view of the severe inflammation, the patient was managed conservatively for 3 days with intravenous dexamethasone (8 mg 3 times a day), intravenous antibiotics, topical steroids and cycloplegic eye drops. After 3 days, eyelid edema and conjunctival chemosis resolved with residual conjunctival congestion and a resolving hyphema (Fig. 3). Computed tomography of the right orbit revealed resolved preseptal inflammation, dislocation of crystalline lens, and an irregular, moderately hyperdense mass filling the posterior segment. There was no evidence of extraocular tumor extension (Fig. 2a, b). With a clinical diagnosis of a necrotic choroidal melanoma in the right eye, the patient underwent enucleation with implantation of a polymethyl methacrylate implant in the right eye. Gross examination of the sections of the eyeball showed a large pigmented necrotic tumor arising from the choroid, a detached retina and a displaced crystalline lens (Fig. 4a). The tumor was composed of spindle A melanoma cells with >50 % necrosis of the tumor (Fig. 4c). There was no extraocular tumor extension. Histopathology confirmed a diagnosis of

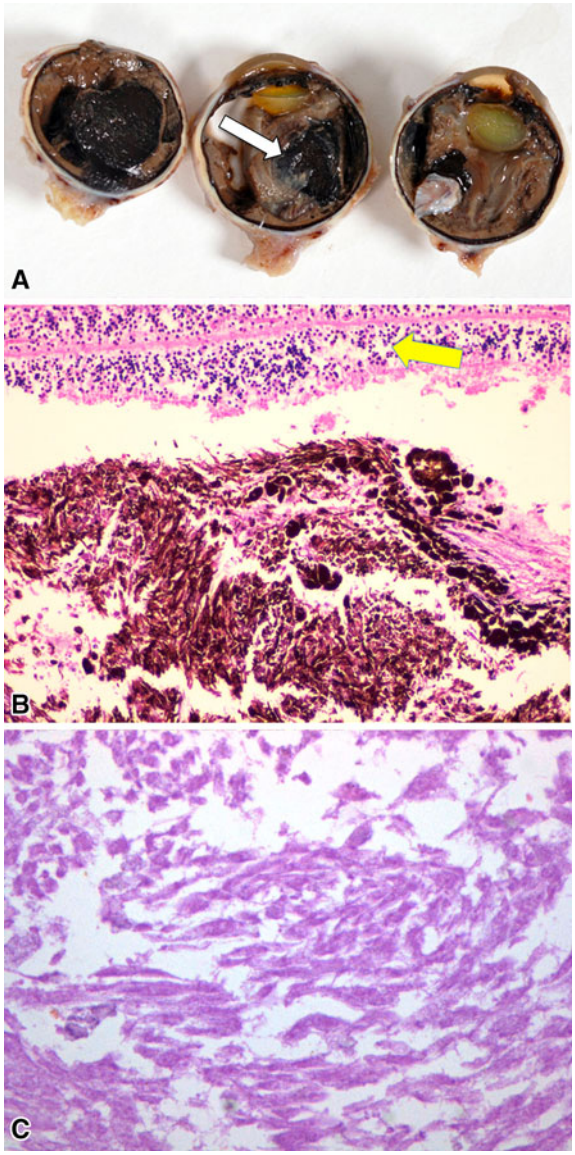

Fig. 4 Gross examination of the sections of the eyeball showed a large pigmented necrotic tumor (*white arrow*) arising from the choroid, a detached retina and a displaced crystalline lens (**a**). Pigmented spindle-shaped choroidal melanoma cells with scanty cytoplasm, elongated nuclei and indistinct nucleoli. The overlying retina is disintegrated suggestive of necrosis (*yellow arrow*) Pigment-laden macrophages and areas of necrosis are noted within the choroidal melanoma (**b**) (H&E staining, ×20 magnification). Spindle A cells with a darker slender nuclei and lacking visible nucleoli are seen in a background of necrosis and dense inflammatory infiltration (**c**) (H&E staining, bleached with potassium permanganate, ×40 magnification)

necrotic malignant melanoma of the choroid. There was no evidence of systemic metastasis at 3-month follow-up.

Choroidal melanoma, the most common intraocular tumor in adults, is usually detected incidentally during routine fundus examination [1]. 41 % of patients are asymptomatic, and 59 % of patients present with symptoms of decreased visual acuity, visual field defect, photopsia, and/or floaters [1]. Painful proptosis secondary to orbital cellulitis is an uncommon presenting feature of choroidal melanoma, and is known to occur in cases with extraocular tumor extension [2–5]. However, orbital cellulitis can also occur in cases of choroidal melanoma with no evidence of extraocular tumor extension, as seen in our case. The pathogenesis of choroidal melanoma-associated orbital cellulitis is probably related to tumor necrosis [2]. A rapidly growing intraocular tumor can outgrow its vascular supply and subsequently undergo necrosis. This in turn releases mediators of inflammation leading to secretion of inflammatory cytokines causing orbital inflammation which respond favorably to systemic steroids [2].

In an analysis of 1,527 globes with uveal melanoma by the Collaborative Ocular Melanoma Study group, >50 % tumor necrosis was noted in 1 % of medium-sized and 2 % of large-sized melanomas [6]. Severe tumor necrosis is more commonly associated with large tumor size, increased tumor pigmentation, epithelioid cell type, and iris neovascularization [6]. In our case, the patient had a large pigmented tumor with iris neovascularization but with spindle A cell-type melanoma. In summary, uveal melanoma-associated orbital cellulitis can be well controlled with systemic steroids, does not always indicate extraocular tumor extension, and can occur in spindle A cell melanomas.

Conflict of interest The authors have no financial or conflicting interests to declare.

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