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Plaque radiation for ciliary body medulloepithelioma presenting with neovascular glaucoma and vitreous hemorrhage in 13-year-old Asian girl

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1. Case report

A 13-year-old female child presented with complaints of diminution of vision and pain in left eye of 15 days duration. She had undergone pan-retinal laser photocoagulation elsewhere before presenting to us. The visual acuity was 6/12 in the left eye and 6/6 in the right eye. Biomicroscopic examination of left eye revealed neovascularization of iris, ectropion uvea and cells in the anterior vitreous cavity. Intraocular pressure (IOP) was 42 mmHg despite being on topical anti-glaucoma medications dorzolamide, timolol and brimonidine. Gonioscopy revealed closed angles with neovascularization of angles in all quadrants and grayish-yellow mass lesion originating from the ciliary body (CB) in the inferior quadrant (Fig. 1b). Fundus examination showed a mass lesion arising from the CB extending on to the retina with laser marks around the lesion. Neovascularization of the disc and neovascularization elsewhere with vitreous hemorrhage was noted. (Fig. 1a).

On ultrasound biomicroscopy (UBM) the mass measured 4.8 × 2.6 mm (width x thickness), extending from 4 to 10 clock hour position with intralesional cysts (Fig. 1c). A retrolenticular cyclitic membrane was noted attached to its apex. Magnetic resonance imaging (MRI) brain and orbit showed a small irregular mass in left CB measuring 4.3 × 2 mm. High-resolution computed tomography of the chest done to rule out pleuropulmonary blastoma did not reveal any pathology. A diagnosis of medulloepithelioma with neovascular glaucoma (NVG) was made.

Initial fine needle aspiration biopsy (FNAB) of the lesion was inconclusive. A repeat FNAB revealed cells arranged in chords, sheets and tubules in a myxoid stroma suggestive of medulloepithelioma (Fig. 1d). Enucleation of the eye was advised. As the parents were not willing for enucleation, I-125 plaque brachytherapy was done. Two months following brachytherapy the tumor size reduced to 3 mm in basal diameter and 1.3 mm in thickness. As the IOP was 36 mmHg despite topical dorzolamide, timolol, brimonidine and oral sustained release acetazolamide, diode laser cyclophotocoagulation was done. Two years later, the lens haze worsened. No tumor recurrence was noted on UBM. Phacoemulsification with posterior chamber intraocular lens implantation was done. The IOP was 30 mmHg at follow-up and the patient continued to be on topical dorzolamide, timolol and brimonidine.

In our reported case, plaque brachytherapy was successful for tumor regression and globe salvage (Fig. 1e and f). However, due to glaucomatous optic atrophy, the visual acuity dropped to hand movements close to face at the last follow-up, 5 years from surgery.

2. Discussion

CB medulloepithelioma is a rare intraocular tumor arising from the nonpigmented ciliary epithelium.[1] Median age at diagnosis reported in literature is two to five years, although the patient in our report was in her early teens.[2] Kaliki et al. reported a large case series of 41 patients with CB medulloepithelioma.[3] Tumor associated clinical features reported by them included iris neovascularization (51%), secondary glaucoma (44%), retrolental neoplastic cyclitic membrane (51%) and intratumoral cysts (61%). Similar features were seen in our patient. Medulloepithelioma has been classified as benign or malignant and

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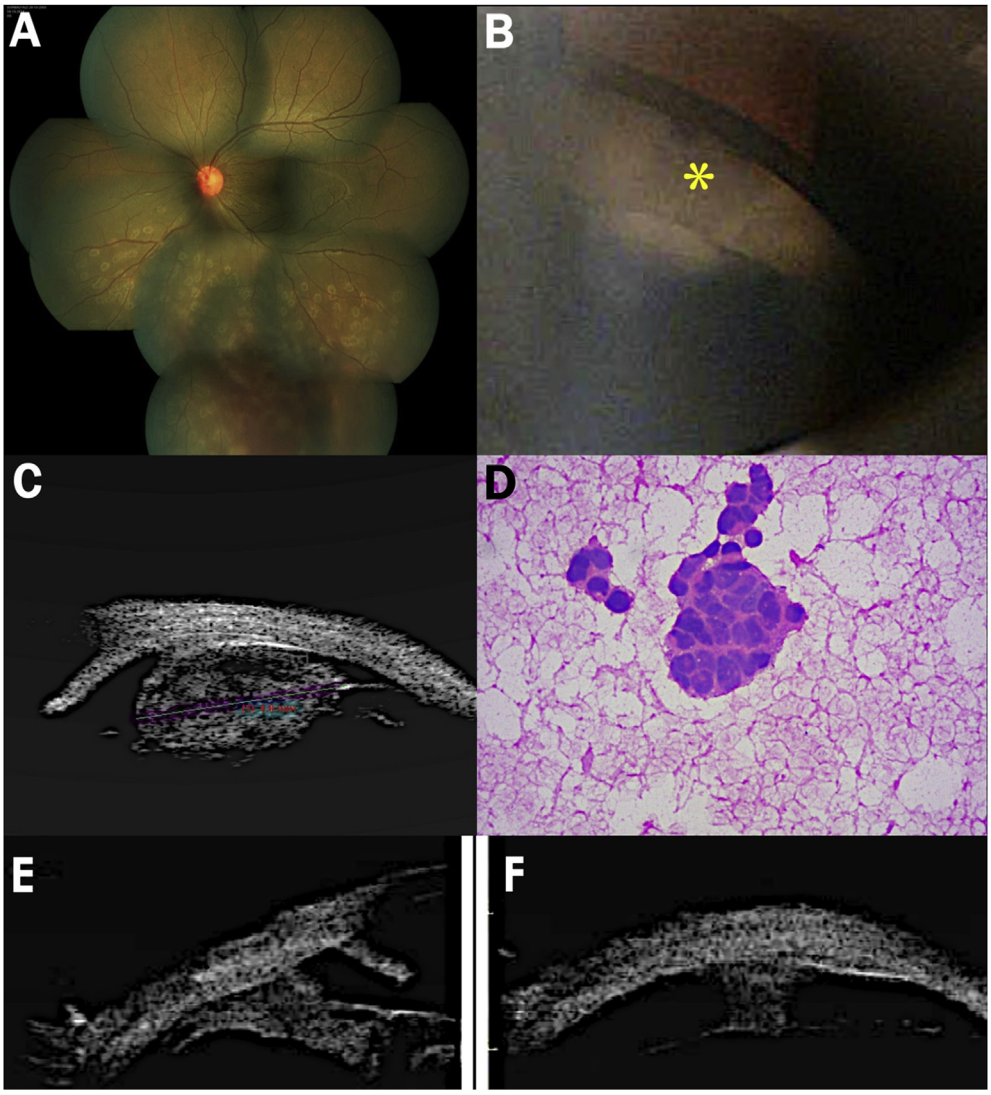

Fig. 1. (B) Gonioscopy reveals a grayish-yellow mass lesion (star) over the ciliary body with a retrolental, cyclitic membrane. (A) Fundus image shows neovascularization of the disc, fresh vitreous hemorrhage in the inferior quadrants with sectoral laser photocoagulation marks. (c) Ultrasound biomicroscopy shows ciliary body mass 4.8mm in basal diameter and 2.6mm in thickness with cystic spaces in the lesion. (d) Microphotograph (H&E, 40X) shows tumor cells arranged in tubular and cord like pattern in a myxoid stroma. (e, f): UBM reveals regressed tumor till the last follow-up 5 years following plaque radiation. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

teratoid or non-teratoid by Anderson and Broughton et al.[2,4] Our patient had a benign and non-teratoid tumor. Plaque radiotherapy has been described as a treatment modality in 9% of the eyes in the case series by Kalki et al. and in a series of 6 cases by Ang et al.[3,5] The tumor reduced in size following brachytherapy and hence globe salvage could be achieved. Due to the rarity of the tumor and lack of established guidelines for optimal management, it is worthwhile considering the following approach with respect to tumor size-surgical resection for small tumors (size upto 3 × 3 mm), plaque radiation for medium sized tumors (size > 3 × 3 mm and up to 6 × 6 mm) and enucleation for large tumors. (size > 6 × 6 mm). However, we do recognize that such recommendations can be best based on a meta-analysis of published reports.

In conclusion, vitreous hemorrhage and NVG can be the presenting signs of an intraocular tumor like medulloepithelioma in young patients. Globe salvage is possible in selected cases with plaque radiation therapy.

Patient consent

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Declaration of competing interest

There in no conflict of interest.

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