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CASE REPORT
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A Case Report of a Child Suffering from Combined Hamartoma of Retina and Retinal Pigment Epithelium

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Abstract

A 2½ years old male baby presented to us with complain of not following light both eyes. Anterior segment of both eyes were normal. Fundus examination revealed CHRRPE lesions in both eyes. Diagnosis of Combined hamartoma of the retina and retinal pigment epithelium mainly depends on fundus manifestations, FFA and OCT. B ultrasound scan is useful for differential diagnosis. Surgical dissection can be effective in the treatment of vitreomacular traction associated with CHRRPE with a good functional and anatomical outcome.

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Introduction

Combined hamartoma of the retina and retinal pigment epithelium (CHRRPE) is an uncommon and benign ocular tumor. Typically single and unilateral, which may compromise the optic disc, macula and less frequently the periphery. This congenital lesion consists of glial cells, vascular tissue, and sheets of pigment epithelial cells.[1] It is commonly found in healthy young children; however, some cases associated with systemic diseases have been described. We report a case of a CHRRPE.

Clinical Case

A 2½ yrs old male baby who has been followed from December 2017 to till date. There was a history of late preterm, i.e., 7 ½ months twin delivery, out of which 1st twin died. The patient's birth weight was 1.8 kg and was admitted to NICU for 1½ month. The parents brought the baby for ROP screening when this lesion was found out. Fundus examination of the right eye showed a yellow-grey mass, slightly elevated above the optic disc and peripapillary retina, extending to the macula and hyper-pigmented areas around the lesion, also tortuous vessels (Figure 1).

On USG B-scan- there was mild thickening of retina of right eye (Figure 2). OCT-demonstrated an elevated lesion with loss of normal architecture of optic nerve and gradual transition from this lesion to normal retinal layers, the retinal pigment epithelium is intact. Macula demonstrates loss of foveal depression and presence of hyper-reflecting nodules (Figure 3).

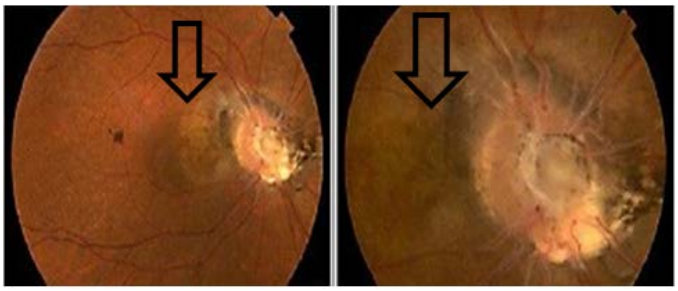

Figure 1: BE fundus photo showing elevated mass above optic disc extending to macula -CHRRPE lesion (black arrows).

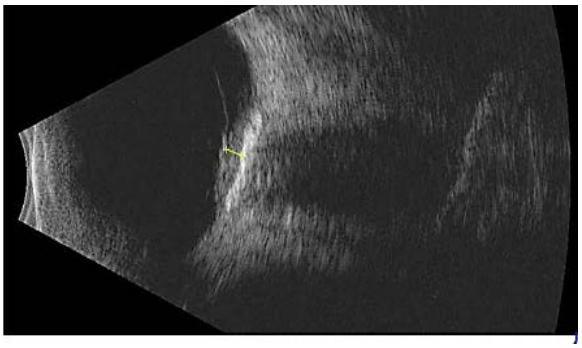

Figure 2: USG B-scan showing thickening of retina

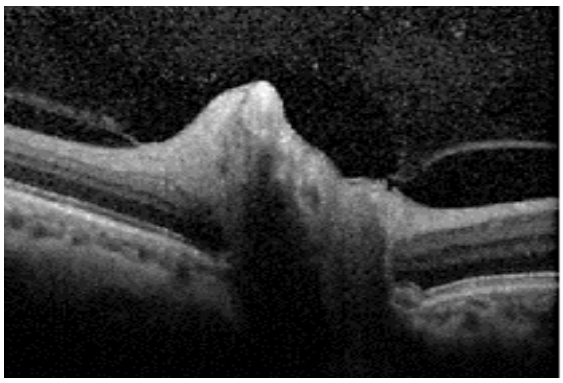

Figure 3: OCT showing loss of architecture of optic nerve

Treatment

The patient underwent right eye PPV with ILM peeling with traction release under GA. PPV with membrane peeling for ERM associated with CHRRPE can result in improved retinal architecture and visual acuity. Now the baby is able to follow the light.[2]

Conclusion

Combined hamartoma of the retina and retinal pigment epithelium (RPE) is a rare, congenital, and benign tumor that may be mistaken with a choroid melanoma or retinoblastoma. These tumors often show an epiretinal membrane, tangential traction, disorganization of the retinal layers, and underlying uniform choroidal thinning.[3] Diagnosis of CHRRPE mainly depends on fundus manifestations, FFA and OCT. USG B-scan is useful for differential diagnosis.[4] Surgery aims to reduce the vitreous traction and establish surface regularity. Timely surgical interventions may yield gratifying functional and anatomical success.

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