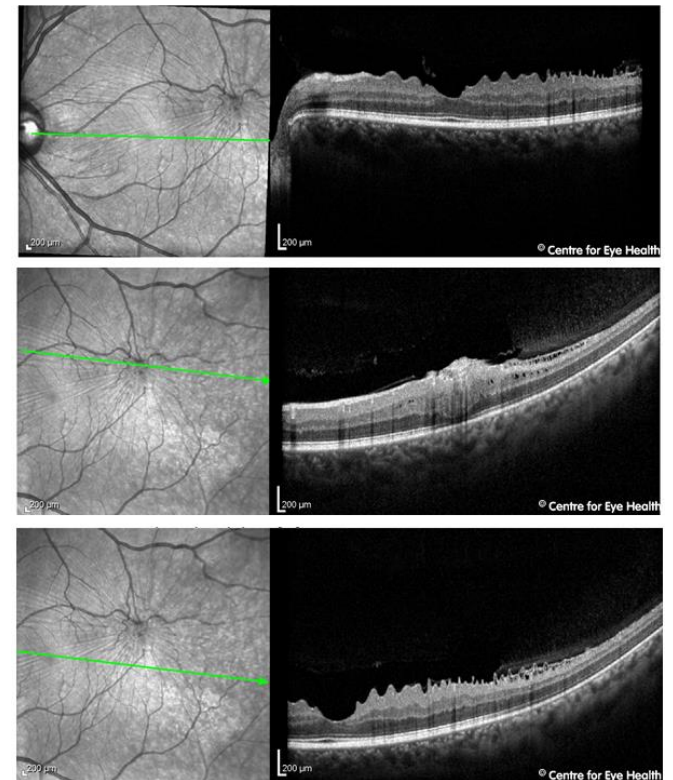
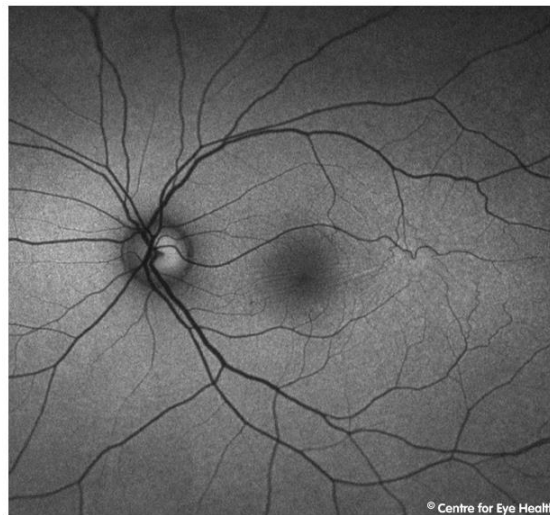




CFEH Facebook Case #97

A pregnant 36 year old female presented for a macular assessment after noticing central distortions in her left eye. She is a migraine sufferer but her history is otherwise unremarkable. What is the most likely diagnosis for this patient?



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ANSWER

Imaging shows a lesion temporal to the macula with associated retinal striations and traction. This traction has caused an ILM detachment and the formation of cystic spaces in the inner nuclear layer (as seen on OCT). These findings are consistent with a diagnosis of combined hamartoma of the retinal and RPE (CHRRPE) which is causing retinal traction in the left eye.

CHRRPE are benign elevated tumours and the level of the sensory retina and RPE. Pigmentation can be variable and the lesions are often associated with epiretinal membrane formation (as in this case here). Lesions can cause significant visual disturbance in some cases, depending on their location. They may also progress with time causing complications such as vitreous haemorrhage, neovascularization, macular hole, and peripheral hole formation.

Recently Dedania et al. (2018) devised a classification system for CHRRPE which takes into account the location of the lesion, the retinal layers affected and the level of traction noted. Based on the classification derived, they also developed recommendations for follow up intervals ranging from 2 to 12 months.

The literature does report an association with Neurofibromatosis 1 and 2 and a bilateral presentation of CHRRPE should increase suspicion of these conditions.

Reference

Dedania, V., Ozgonul, C., Zacks, D., Besirli, C. (2018) Novel classification system for combined hamartoma of the retina and the retinal pigment epithelium. *Retina* 38:12–19, 2018