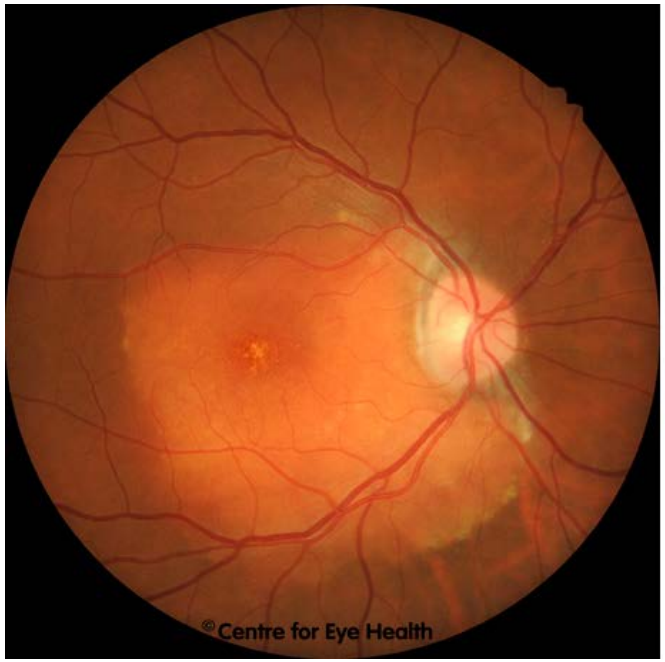
Centre for Eye Health

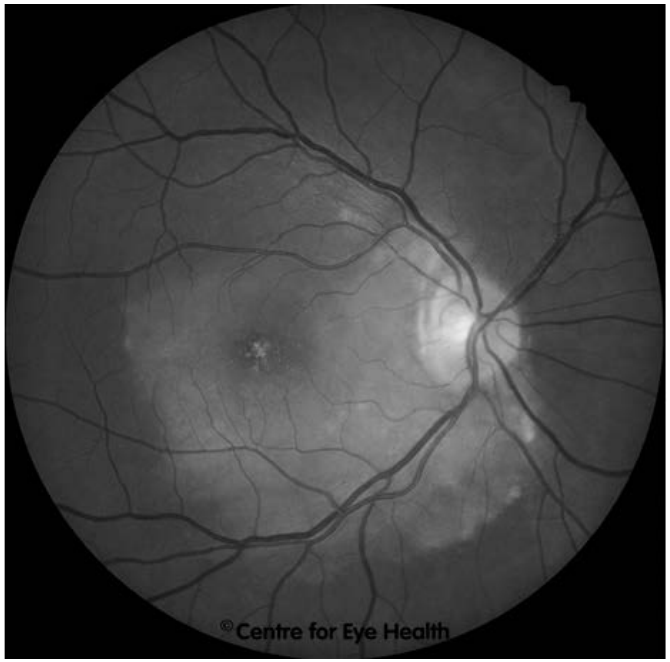
CFEH facebook Cases

CFEH Facebook Case #8

A 46 year old Caucasian female was referred to the Centre for investigation changes at the right macula. Her medical history includes a double ureter previously investigated by a nephrologist 8 years previously. BCVA is 6/3.8 OD, 6/4.8 OS. Amsler grid was unremarkable in both eyes and contrast sensitivity was normal at 1.76 log units in the right eye and 1.72 log units in the left (normal range 1.72 to 1.92 log units). A fundus photograph, red-free image, Optomap image, fundus autofluorescence and OCT images for the right eye are below. As the left eye appeared normal these images were omitted.

What is your diagnosis for this patient? How would you manage it?





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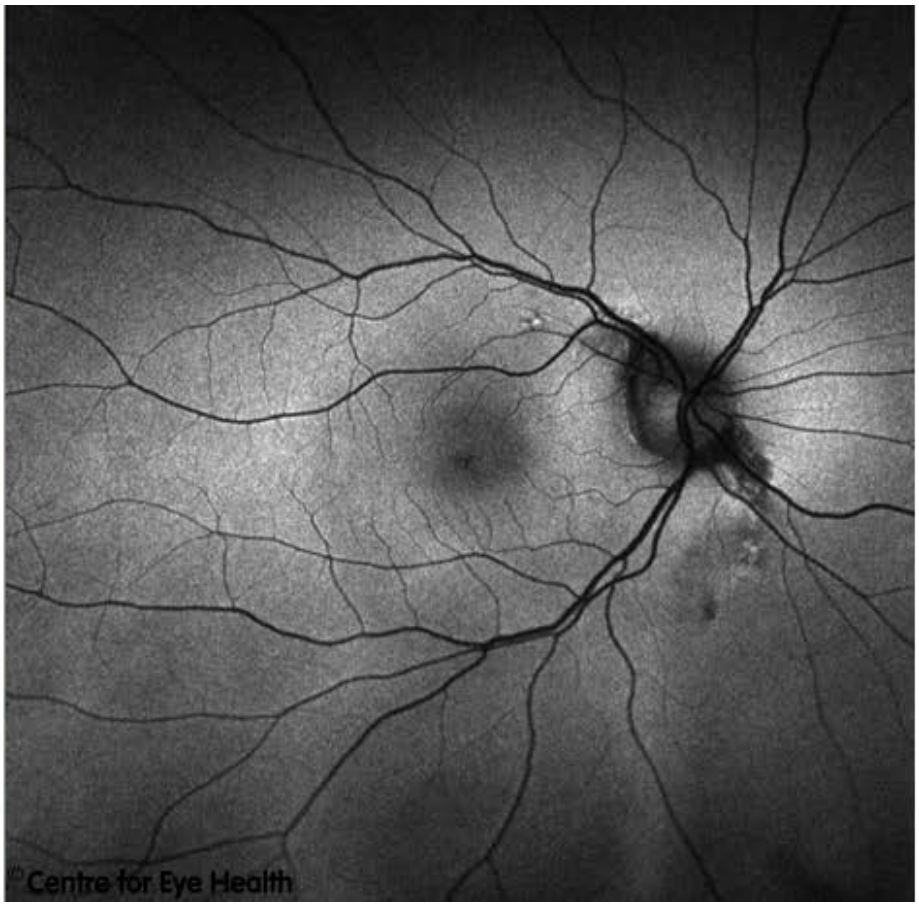

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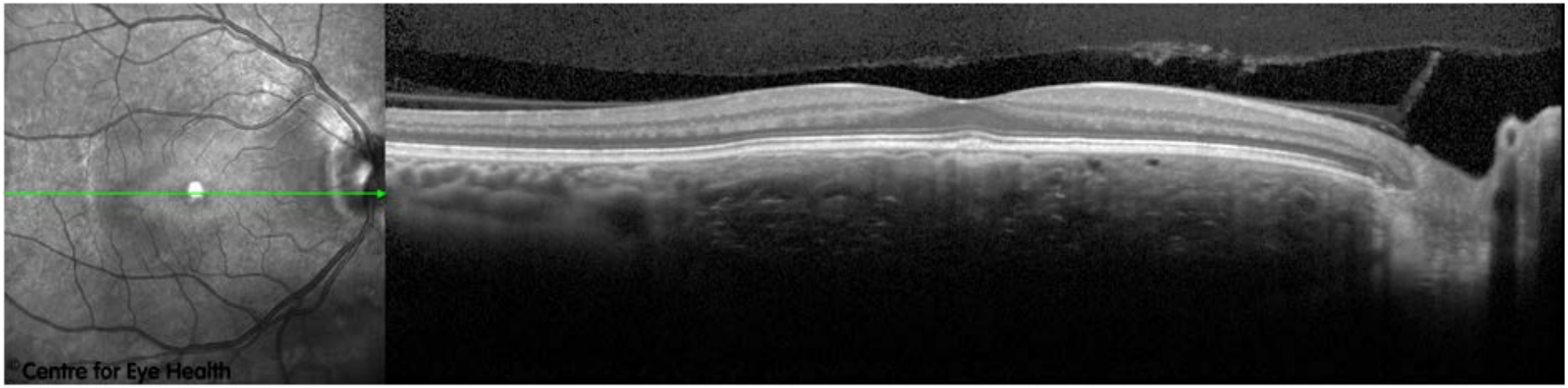

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ANSWER

Choroidal osteoma

Choroidal osteomas are rare, benign tumours of the choroid, usually affecting the juxtapapillary and macular areas. They are more prevalent in females and are usually unilateral (80% of cases). Age of onset can vary significantly, from a few months of age to late 60's, but they are predominantly found in young adults.

Osteomas are typically irregular in shape with sharply defined or scalloped borders, are slightly elevated and usually have fine vascular networks on the surface of the tumor. The colour of an osteoma is related to the level of RPE depigmentation which increases over time meaning that in early stages, they are orange-yellow in colour and in later stages more of a yellow-white colour.
Looking at the clinical picture for this patient, we can see a juxtapapillary yellow-orange lesion that involves the macula. It is well demarcated with scalloped borders and there is associated RPE mottling at the macular. Fundus autofluorescence shows a speckled hyper- and hypo-autofluorescence pattern along the inferonasal and superior margins of the lesion.

OCT images show an abrupt change in the choroidal architecture at the site of the lesion while overlying inner and outer retinal layers appear undisturbed.

In this case, OCT imaging showed there was no apparent subretinal fluid, RPE/photreceptor alterations or subretinal haemorrhage, explaining why visual acuity is still so good. This patient was referred to a retinal specialist for further assessment and the diagnosis was confirmed by B-scan ultrasound which showed a strong acoustic shadow which is characteristic of choroidal osteomas.

Although a choroidal osteoma is benign, most show slow growth over time and vision can be affected through several mechanisms:

1. Atrohpy of the RPE and choriocapillaris, producing a thin, yellow-grey area overlying the osteoma.

2. Choroidal neovascularization (CNV), the prevalence of which has been found by various studies to be between 31 and 47%. Tumors with overlying haemorrhage and an irregular surface are at highest risk of developing CNV.

3. Subretinal fluid, haemorrhage and serous retinal detachment due to a damaged RPE are often associated with osteomas in the absence of CNV. One long-term study found that 64% of eyes with sub-retinal fluid resolved spontaneously, but those that didn't had poor visual outcomes. The 10-year probability of losing visual acuity to the level of 6/60 was found to be around 56%.

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