Case study example

Quick title
CMO or CSCR?

Full title
Choroidal melanoma with cystoid macular oedema (CMO) presenting as suspected central serous chorioretinopathy (CSCR)

Author(s) name(s), job title(s) and brief one line description
Clinical Editor Kieran Loft MCOptom and Consultant Eye Surgeon Peter Simcock FRCOphth discuss how to discriminate between CMO and CSCR and the importance of careful fundus examination.

Presentation
A 35-year-old male presented to his optometrist for describing reduced vision in his right eye over the last two years, with intermittent photopsia over the last 2 weeks.

Medical and ocular history
Ocular history: Last eye examination 6 months ago at another practice, referred routinely for suspected central serous chorioretinopathy (CSCR) but did not attend as felt it would improve spontaneously
General health and allergies: Period of stress six to seven years ago but no recent episodes. Fit and well. No allergies.
Medication: Nil, denies history of steroid use.
Relevant family history: Father with borderline type 2 diabetes.

Clinical examination

- BCVA: 6/9 (NIPH) RE; 6/5 LE
- Near VA at 40cm: N8 RE, N5 LE
- Relevant findings:

Right	Structure	Left
Normal	Lids & lashes	Normal
Normal	Conjunctiva	Normal
Normal	Sclera	Normal
Clear	Cornea	Clear
Clear and quiet, Van Herick grade 4	Anterior chamber	Clear and quiet, Van Herick grade 4
Normal direct, consensual and near responses; no anisocoria	Iris and pupils	Normal direct, consensual and near responses; no anisocoria
Clear	Lens	Clear
Clear	Vitreous	Clear
Well defined, healthy neuro-retinal rims and moderate cupping; c:d 0.35	Optic disc	Well defined, healthy neuro-retinal rims and moderate cupping; c:d 0.30

Dull reflex and raised; cystoid macular oedema visible on OCT	Macula	
Normal, a:v = 2:3	Vessels	Normal, a:v = 2:3
Large raised non-pigmented lesion temporal retina (see figure 1); with collar stud configuration originating from the choroid on OCT (see figure 2)	Peripheral retina	Flat, unremarkable

- IOP: 15.3mmHg RE, 16.0mmHg LE at 12:35pm (NCT)
- Visual fields: corresponding visual field defect to the lesion RE, normal LE (supra-threshold perimetry)

Management

Suspected choroidal melanoma RE, so an urgent referral was made to the local HES and was seen 2 days later where they were referred onward to the Liverpool Ocular Oncology Centre. The diagnosis of choroidal melanoma was confirmed. Due to the size of the lesion, enucleation was considered the best option. Histopathology confirmed malignant spindle cell melanoma, but with no extra-ocular extension, and no reduction in life expectancy as a result. The patient was reassured and is in the process of having an artificial eye fitted

Discussion

This case highlights how CMO is not always caused solely by a problem at the macula. OCT is a useful tool to discriminate between CMO and CSCR. CSCR typically shows a puddle of sub-retinal fluid without cysts, often with a ‘volcano-like’ configuration, while CMO has classic cystoid spaces within the retinal layers. Otherwise, clinical differentiation can be difficult because both conditions can cause distortion on the Amsler grid, loss of macular reflex and colour vision abnormality. The history can offer important clues and should be carefully considered. CSCR should only be a diagnosis of exclusion, and the importance of correct HES follow-up should be made clear to the patient, especially if there is any diagnostic doubt.

CSCR typically presents in middle-aged males, causing mild unilateral blurred vision with positive scotoma, distortion and/or micropsia. ‘Type A’ personality – high ambition, work-driven, competitive (Friedman and Rosenman, 1974) – stress and steroid use are often cited as contributory factors. CSCR normally spontaneously resolve with corresponding improvement in VA within one to four months, although recurrences are common. In some cases, CSCR can affect the fellow eye or can become chronic where visual function may be permanently affected in the latter (Nicholson et al, 2013).

Choroidal melanoma is thankfully rare, with incidence of around five per million per year (Kaliki and Shields, 2017). Median age of diagnosis is around 60, although patients of any age can be affected (Kaliki and Shields, 2017). Blurred vision is the most common symptom

(38 per cent of patients), but as many as 30 per cent of patients are asymptomatic (Damato and Damato, 2012). Photopsia, floaters and field loss are often-quoted symptoms, but affect less than 10 per cent of cases (Damato and Damato, 2012). Risk factors include pale skin/ocular pigmentation, inability to tan and pre-existing ocular or cutaneous naevi (Kaliki and Shields, 2017).

Dilated fundal examination is essential for diagnosis. Malignant melanoma in its early stages can be seen as a raised dome-shaped mass, usually pigmented (55 per cent) or mixed in colour (30 per cent), but may be non-pigmented (15 per cent) (Shields et al, 2009). If the lesion ruptures Bruch's membrane, it may have a 'mushroom' or 'collar-stud' configuration. Diffuse tumours are less common. Urgent referral is indicated, and ultrasonography may be used to help confirm diagnosis. Treatment ranges from observation – for small, slow-growing tumours, or if the patient already has poor life expectancy – to enucleation. Metastasis is common, with 32 per cent undergoing metastasis by five years, rising to 62 per cent by 35 years (Kujala et al, 2003).

Figures with explanation

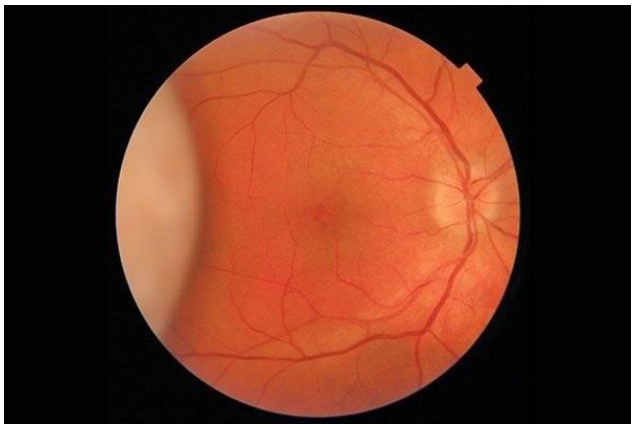

Figure 1: Fundus photograph of the collar-stud melanoma (RE temporal mid-periphery, with macula visible to right of image)

Figure 2: etc

Learning points

This case demonstrates the importance of thorough examination of the peripheral retina and not relying on macular OCT/imaging for retinal examination. Macular fluid can come from peripheral sources including rhegmatogenous retinal detachment, retinal vascular abnormalities such as Coats' disease and uveitis, retinal dystrophies such as retinitis pigmentosa, as well as choroidal lesions such as naevi and, importantly, choroidal melanoma.

- Macular oedema is not always caused by a problem at the macula; always search for other causes of oedema outside the macular area.
- Dilated fundus examinations should be performed if a cause is not obvious.
- Careful interpretation of OCT can be a useful tool in differential diagnosis.
- CSCR is a diagnosis of exclusion.

- Always ensure your patient understands the reason for and importance of referral.

References

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