ELECTRONIC CLINICAL CASE

Circumscribed choroidal haemangioma mimicking chronic central serous chorioretinopathy

Hémangiome choroidien circonscrit imitant une chorioretinopathie séreuse centrale

W. Rahman*, N. Horgan, J. Hungerford

Moorfields eye hospital, 162, city road, London EC1V 2PD, United Kingdom

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Summary We describe a rare case of bilateral circumscribed choroidal haemangioma in an otherwise healthy male, which mimicked chronic central serous chorioretinopathy (CSCR). A 52-year-old Asian man presented with a one-year history of visual decline in his left eye. The vision in the right eye had been reduced for 15 years. Visual acuity was 6/60 in the right eye and 6/18 in the left eye. Fundus examination of the right eye revealed an area of discolouration with overlying retinal pigment epithelial changes in the macula and evidence of prior surrounding argon laser photocoagulation. The left macula showed a raised choroidal lesion with overlying retinal pigment epithelial changes and associated subretinal fluid. This appearance illustrates how chronic retinal pigment epithelial alterations associated with longstanding subretinal fluid exudation from circumscribed choroidal haemangiomas may mimic the appearance of chronic central serous chorioretinopathy. B-scan ultrasonography, fluorescein angiography, indocyanine green angiography and optical coherence tomography helped to establish the diagnosis. The active lesion in the left eye was treated with verteporfin photodynamic therapy with improvement in vision.

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* Corresponding author.
E-mail address: waheeda.rahman@moorfields.nhs.uk (W. Rahman).

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Introduction

Circumscribed choroidal haemangioma is an uncommon benign hamartomatous tumour of the uvea, which was first described by Leber [1]. It occurs sporadically, posterior to the equator and is usually unilateral. Bilateral cases are extremely rare. We describe a case of bilateral circumscribed choroidal haemangioma in a healthy Asian male, which mimicked chronic central serous chorioretinopathy (CSCR).

Case description

A 52-year-old Asian man presented with a one-year history of gradual deterioration of vision in his left eye. He had undergone prior laser treatment to his right eye 15 years earlier at another institution. The patient was otherwise systemically well.

Best-corrected Snellen visual acuity at presentation was 6/60 in the right eye and 6/18 in the left eye. The anterior segments and intraocular pressures were normal. Fundus examination revealed an area of orange discoloration temporal to the right fovea with overlying retinal pigment epithelial (RPE) changes, and evidence of prior surrounding argon laser photocoagulation (Fig. 1A). Examination of the left macula showed a raised choroidal lesion with overlying RPE alteration and associated subretinal fluid (Fig. 1B).

A B-scan ultrasound confirmed bilateral macular dome-shaped choroidal lesions, which were acoustically solid. The lesion in the right eye measured 3.1 × 3.4 mm in base and 0.7 mm in thickness and the lesion in the left eye measured 6.3 × 5.8 mm in base and 1.7 mm in thickness, with an associated shallow retinal detachment (Figs. 1C and D). Fluorescein angiography (FA) demonstrated a window defect in the region of the right macular lesion, with evidence of a prior subretinal fluid track extending inferiorly, mimicking the appearance of RPE change often associated with chronic CSCR (Fig. 1E). On FA the left macular lesion showed marked hyperfluorescence (Fig. 1F). Indocyanine green angiography of the left macular lesion showed early hyperfluorescence and reduced fluorescence late in the angiogram. Time-domain optical coherence tomography (OCT) (Stratus; Carl Zeiss Meditec, Dublin, CA) confirmed a raised choroidal lesion at the macula in the left eye with overlying subretinal fluid and significant cystoid intraretinal fluid (Fig. 2A).

The findings were consistent with a diagnosis of bilateral circumscribed choroidal haemangiomas. The patient had no cutaneous haemangiomas. Computerised tomography scan of the head demonstrated no intracranial abnormalities. Based on the recent visual deterioration in the left eye related to the active subretinal fluid exudation from the haemangioma, the lesion in the left eye was treated with verteporfin photodynamic therapy (vPDT, Visudyne, Novartis Ophthalmics, Basel, Switzerland; 50 J/cm², single spot). As the lesion in the right eye appeared clinically inactive no treatment was recommended for that eye.

Follow-up 3 months later revealed subjective improvement of vision in the left eye, with best-corrected Snellen visual acuity improving to 6/12. OCT demonstrated almost complete resolution of the subretinal fluid, but persistence of some overlying cystoid intraretinal fluid (Fig. 2B). The thickness of the lesion on ultrasound remained stable.

Discussion

Circumscribed choroidal haemangiomas usually present in Caucasian patients in their fourth to sixth decade of life but occur infrequently in Asians [2]. Characteristic features on fluorescein and indocyanine green angiography and ultrasonography, as described in this case, can help to establish the correct diagnosis.

Bilateral circumscribed tumours have been rarely reported in association with Sturge-Weber syndrome (SWS) [3,4]; diffuse choroidal haemangiomas are more commonly associated with this condition. Only three cases in the literature describe bilateral circumscribed choroidal haemangiomas in otherwise healthy individuals [5–7]. Tran et al. suggested that this may represent a low penetrance phenotype of SWS [5]. Our patient had no associated systemic anomalies.
The appearance of the lesion in this patient’s right eye at the time of presentation (Figs. 1A and E) illustrates how chronic RPE alterations associated with longstanding subretinal fluid exudation from circumscribed choroidal haemangiomas can mimic the appearance of chronic CSCR. In this case in particular, the haemangioma in the patient’s right eye was clinically inactive and may have regressed at the time of presentation, and ultrasound revealed only minimal thickening within the choroid at the site of the haemangioma. Ultrasound and angiographic evaluation of the left eye, however, allowed confirmation of the diagnosis of an active circumscribed choroidal haemangioma in that eye, illustrating the value of these ancillary investigations in establishing the diagnosis.

The treatment of subfoveal haemangiomas remains a challenge. Chronic subretinal and intraretinal fluid associated with these lesions can cause significant visual impairment, especially when there is macular involvement. Various treatment methods have been advocated. Argon laser photocoagulation has previously been employed for the treatment of these lesions [2,8] although more recently photodynamic therapy with verteporfin has evolved as the
preferred first line treatment [9,10]. Visual outcomes with treatment are poorer in patients with subfoveal haemangiomas [8] and in those with a longer duration of symptoms prior to treatment [2], and the number of vPDT treatment sessions required may be inversely related to the final visual acuity [11].

Disclosure of interest

The authors declare that they have no conflict of interest concerning this article.

References


Figure 2. A. OCT scan showing subretinal and intraretinal fluid over choroidal haemangioma in left eye before treatment. B. OCT scan 3 months after photodynamic therapy showing significant reduction of fluid over lesion in left eye.