

CENTRAL SEROUS CHORIORETNIOPATHY AS A MASKING SYNDROME OF CHOROIDAL HAEMANGIOMA

SUMMARY

Introduction: The finding of the neuroretinal ablation in fovea centralis area on Optic Coherence Tomography (OCT) examination in young men is a typical finding for central serous chorioretinopathy. This finding may masquerade the presence of the choroidal hemangioma.

Case report: The author presents case reports of two young men, in which, according to the finding of neuroretinal ablation on OCT, the diagnosis of central serous chorioretinopathy was established. Using following examinations, especially fluorescence angiography, the correct diagnosis of choroidal hemangioma was set. In both patients, the photodynamic therapy with Visudyne was consequently indicated.

Conclusion: In young patients with slightly decreased visual acuity, neuroretinal ablation finding in the macula on OCT, and without the tendency to spontaneous improvement, it is necessary to keep in the mind the possible presence of choroidal hemangioma.

Key words: hemangioma, central serous chorioretinopathy, misdiagnosis, ablation of the neuroretina, macula, choroid

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INTRODUCTION

Choroidal haemangioma is a vascular benign intraocular tumour. It is considered to be a congenital hamartoma, but usually appears in young adults. We differentiate between two types of haemangioma – bordered and diffuse. Diffuse haemangioma is typically present in patients with Sturge-Weber syndrome. Bordered haemangioma is isolated and not connected with any systemic pathology. The unilateral form appears in the 3rd-5th decade of life [5, 7]. In up to 80% of patients the first manifestation of the presence of haemangioma is a deterioration of visual acuity. In 81% of patients with choroidal haemangioma, ablation of the neuroretina in the macula is present [10].

Diagnostic methods include ophthalmoscopy, ultrasonic examination, fluorescence angiography and indocyanine green angiography. Bordered haemangioma is manifested on the ocular fundus as an oval orange mass projecting on the posterior pole of the eye. Sometimes induced hyperplasia or fibrous metaplasia of the retinal pigment epithelium. On ultrasound a characteristic finding is high internal reflexivity, since a haemangioma is composed of blood vessels and from a range of various different cells. Upon examination by fluorescence angiography slight hyperfluorescence is evident in the early phase, which progressively grows. In the case of a bordered haemangioma, indocyanine green angiography displays a lesion with high reflexivity in the incipient phases and progressively a reduction of fluorescence is evident (“wash out” phenomenon) [1, 5].

OBJECTIVE:

The objective of this study is to draw attention to the potential misdiagnosis of bordered choroidal haemangioma

as central serous chorioretinopathy (CSCR), primarily in young patients.

CASE REPORT 1

A thirty three year old patient reported to the Department of Ophthalmology at Brno University Hospital due to a deterioration of visual acuity in the right eye persisting for several days. Best corrected visual acuity in the right eye was 5/10, in the left eye 5/5. The personal and ocular anamnesis was negative, the patient stated increased psychological stress at work (as the owner of a private company). The finding was physiological on the anterior segment of both eyes, on the fundus of the right eye we observed ablation of the neuroretina in the macula, in the left eye the finding was physiological. The patient underwent an examination by optical coherence tomography (OCT), in the right eye there was evident high ablation of the neuroretina (fig. 1). A diagnosis of central serous chorioretinopathy was determined in the patient, and auxiliary antiedematous therapy was applied (escinum alfa tbl). Within the course of 3 months there was an improvement in the local finding, but the ablation of the neuroretina in the centre on OCT persists (fig. 2). Upon biomicroscopy of the fundus a flat orange deposit is visible above the macula by the upper arcade (fig. 3). We re-evaluated the diagnosis of the patient as suspected choroidal haemangioma, and indicated the performance of fluorescence angiography (fig. 4, 5, 6,7) and indocyanine green angiography (fig. 8, 9, 10). On the basis of the performed examinations a diagnosis of choroidal haemangioma was confirmed. We indicated photodynamic therapy (PDT) with verteporfin (Visudyne, Novartis). At a follow-up examination 3 months later there was an im-

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provement of the patient's visual acuity to 5/5, on OCT a foveolar depression was formed (fig. 11), and this condition persisted also for one year after the performance of PDT (fig. 12). During treatment we indicated a sonographic examination of the kidneys and magnetic resonance imaging of the brain for the patient in order to exclude the possibility of further haemangiomas, both examinations were negative.

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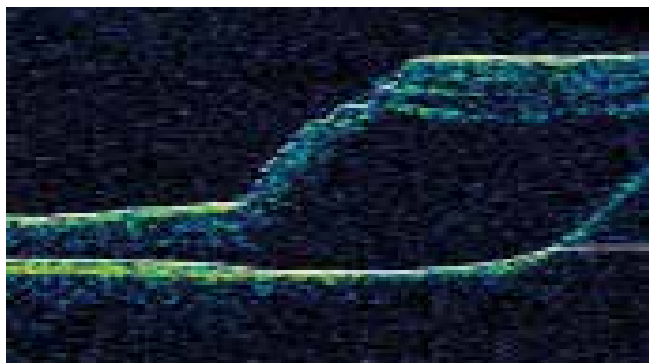


Fig. 1 OCT findings in the right eye at the first examination (Stratus OCT)

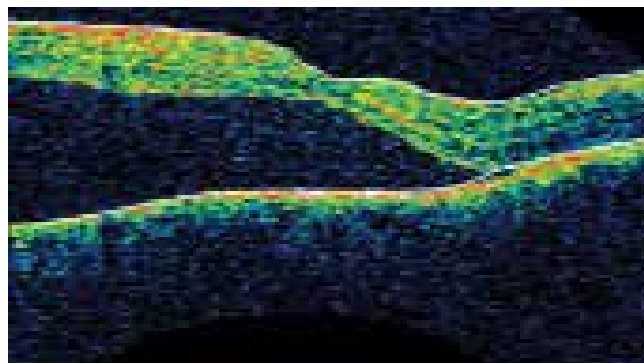


Fig. 2 OCT findings in his right eye three months after initiation of supportive therapy (Stratus OCT)

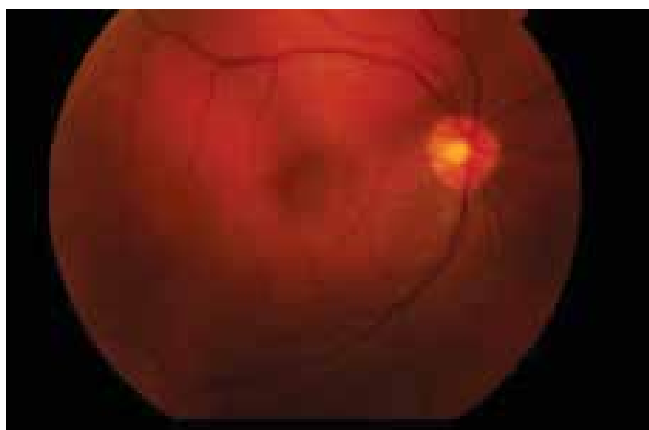


Fig. 3 The finding on the fundus of the right eye – color photo-

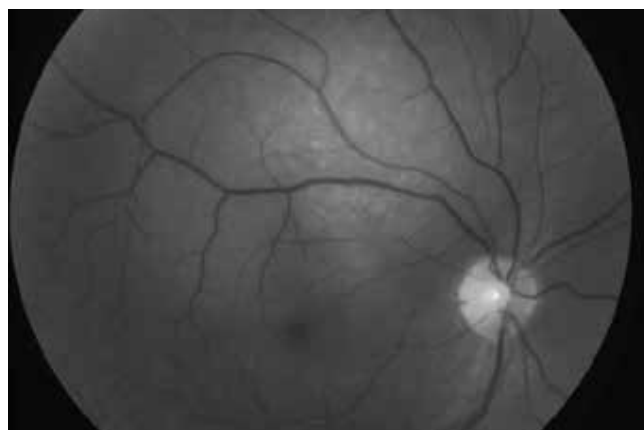


Fig. 4 The finding on the fundus of the right eye - red free photography

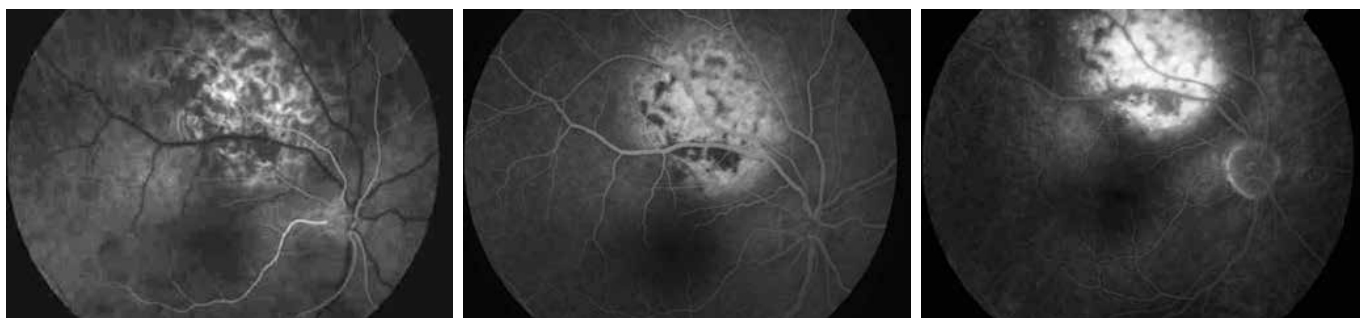


Fig. 5, 6, 7 The finding on the fundus of the right eye on fluorescein angiography



Fig. 8, 9, 10 The findings on the fundus of the right eye on indocyanine angiography

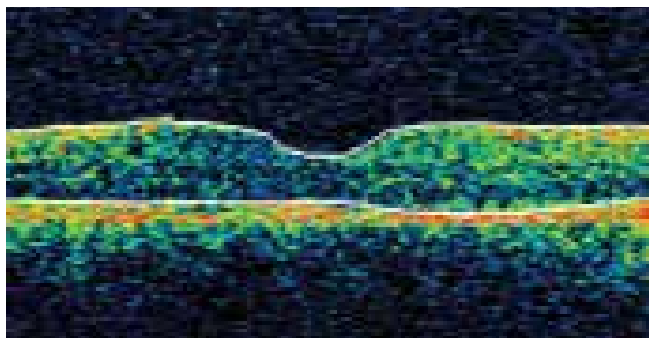


Fig. 11 OCT findings in the right eye three months after PDT (Stratus OCT)



Fig. 12 OCT findings in his right eye – one year follow up after PDT (Cirrus OCT)

CASE REPORT 2

A patient aged 40 years reported to our outpatient clinic due to sudden deterioration of visual acuity in the left eye. The personal anamnesis was negative, the ocular anamnesis contained only information about myopia. The patient was highly anxious, with a pronounced tendency towards self-examination, a university graduate with a technical background. BCVA in the right eye was s -2.0 Dsf 4/4, BCVA in the left eye s -2.5 Dsf 4/6. The finding was physiological on the anterior segment of both eyes, on the fundus of the right eye and also on the fundus of the left eye was manifest ablation of the neuroretina (fig. 13). On OCT we confirmed this ablation of the neuroretina (fig. 14). The patient was recommended psychological rest and antiedematous therapy (escinum alfa tbl). At a follow-up examination one month later there was a slight improvement in the finding on OCT (fig. 15), the patient stated persistent pronounced complaints and fluorescence angiography was indicated (fig. 16, 17, 18, 19). With regard to the fact that we expected a hot spot upon central serous chorioretinopathy in the centre, we photographed the central field, and only in the venous phase we also photographed the upper temporal region, where a deposit was shown upon hyperfluorescence. After a time interval we repeated the examination, focusing on the upper temporal region, where a rounded orange deposit was evident (fig. 20). On the basis of fluorescence angiography (fig. 21, 22, 23) we expressed a diagnosis of choroidal haemangioma. With regard to the fact that the use of indocyanine green is off label in the ocular pathology, and due to the patient's conflictual character, we did not indicate this



Fig. 13 The finding on the fundus of the left eye – color photography

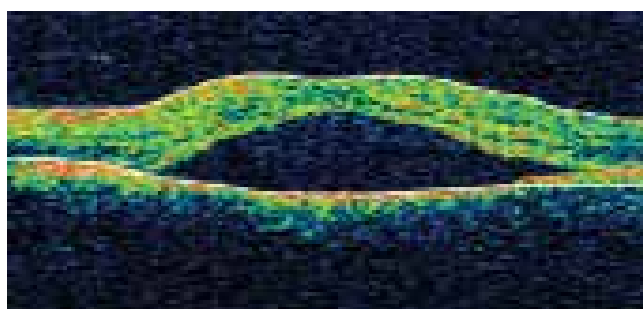


Fig. 14 OCT findings in the left eye at the first examination (Stratus OCT)

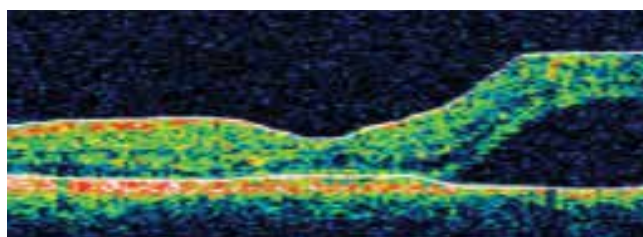


Fig. 15 OCT findings in the left eye after one month of supportive treatment (Stratus OCT)

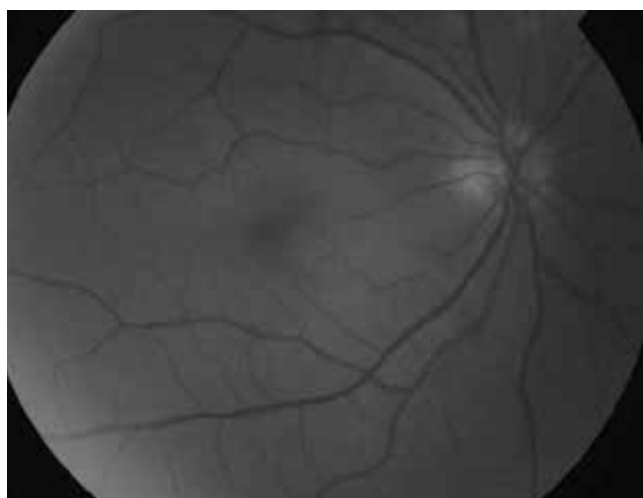


Fig. 16 The finding on the fundus of the left eye - red free photography



Fig. 17, 18 The finding on the fundus of the left eye on fluorescein angiography – posterior pole

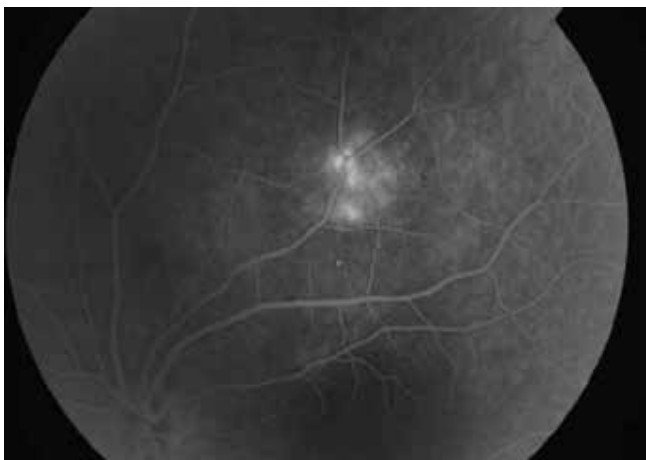


Fig. 19 The finding on the fundus of the left eye on fluorescence angiography - upper temporal quadrant

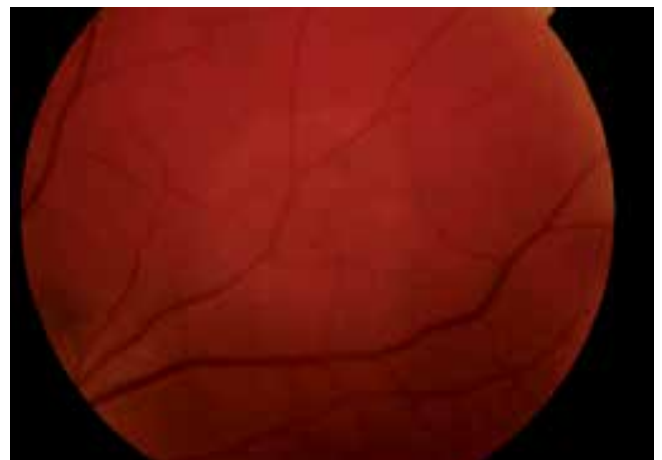


Fig. 20 The finding on the fundus of the left eye - upper temporal quadrant – color photography

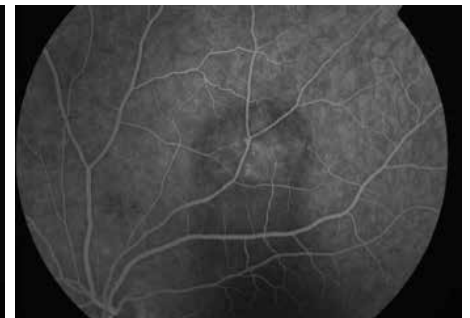


Fig. 21, 22, 23 The findings in the fundus of the left eye on fluorescein angiography - upper temporal quadrant

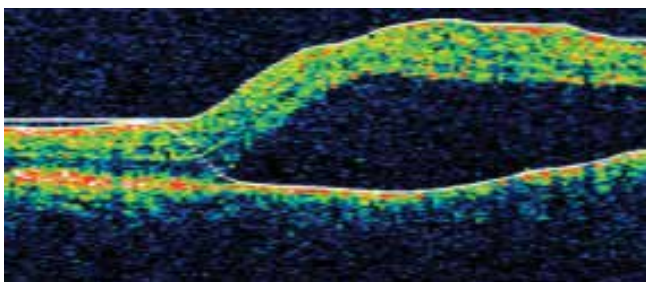


Fig. 24 OCT findings in the left eye on the day of PDT (Stratus OCT)

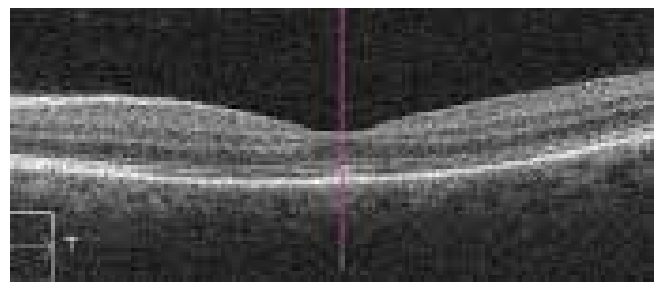


Fig. 25 OCT findings in the left eye two months after PDT (Cirrus OCT)

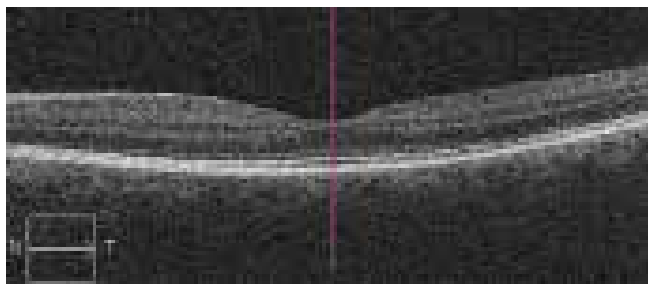


Fig. 26 OCT findings in the left eye after one year after PDT (Cirrus OCT)

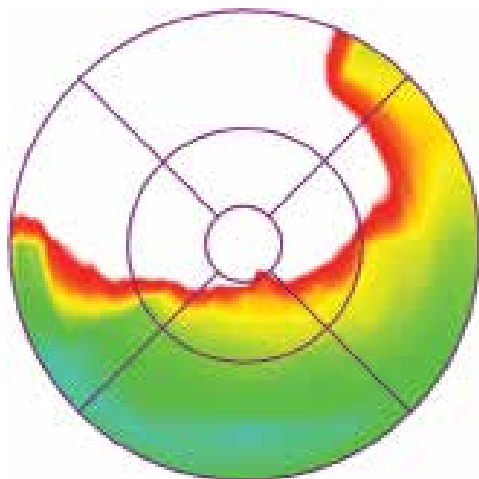


Fig. 27 OCT findings – color retinal map (Cirrus OCT)

examination. The patient underwent photodynamic therapy (PDT) with verteporfin (Visudyne, Novartis), (fig. 24). After 2 months an improvement of BCVA was achieved of s – 2.0 Dsf to 4/4, on OCT a foveolar depression was formed (fig. 25), the same finding persisted also for one year after treatment (fig. 26). Although the patient subjectively perceived an improvement in the left eye, he continued to observe an obtrusive curvature of the image in the lower part of the visual field. Examinations in order to exclude the possibility of further haemangiomas in the body (sonography of kidneys and magnetic resonance imaging of brain) were negative.

Discussion:

Bordered choroidal haemangioma is most often manifested by a deterioration of visual acuity. In his study, Shields retrospectively evaluated 155 patients with an average age of 45 years with isolated choroidal haemangioma. The first symptoms included a deterioration of visual acuity (80 %), defect in the visual field (7 %), metamorphopsia (3 %), turbidities in front of the eye (2 %), progressive hypermetropia (1 %), photopsia (1 %) and pain (1 %). In 6 % of patients this concerned a chance finding and the patients did not state any complaints. Shields et al. further evaluated the primary diagnosis under which these patients were sent to a higher centre. A correct diagnosis of choroidal haemangioma was determined in 29 % of patients, among the incorrect diagnoses the most frequent was malignant melanoma (29 %), followed by diagnosis of metastasis (9 %), retinal detachment (6 %) and central serous chorioretinopathy (5 %) [10].

Isolated choroidal haemangioma is manifested on the ocular fundus as a bordered orange mass, usually one to three papillary diameters (1500 µm) from the macula. The mass of the haemangioma itself is not pigmented, but induced pigmentation may sometimes develop over it (hyperplasia of retinal pigment epithelium), which leads to incorrect diagnosis of malignant melanoma of the choroidea. This concerns a very slowly growing tumour, manifesting only a very low cell proliferation, unlike other “genuine” haemangiomas [2, 8, 10].

A very typical manifestation of haemangioma is an accumulation of subretinal fluid, according to the study by Shields et al. it is present in up to 80 % of patients. This probably concerns a consequence of breach of the haemato-retinal barrier [10].

In differential diagnostics of haemangioma, in first place is malignant melanoma of the choroidea and metastasis. An important role in determining the correct diagnosis is played by ultrasound examination, primarily evaluation of internal reflexivity on an A scan. On an A scan typical features of a melanoma of the choroidea are regular internal structure and medium to low reflexivity, which corresponds to the fact that this tumour is composed of small uniform cells. By contrast, choroidal haemangioma manifests high internal reflexivity, which is typical for a mass composed of different types of cells [1]. Upon ultrasound examination it is very difficult to differentiate choroidal metastasis from haemangioma. Another important examination is fluorescence angiography, in which haemangioma manifests slight, progressively increasing hyperfluorescence from the very beginning. In the case of melanoma we find patchy hyperfluorescence in the arterial and early venous phase, in the case of a breached Bruch's membrane pronounced hyperfluorescence is evident in the venous phase, a typical image is of dual circulation (simultaneous fluorescence of retinal and choroidal blood vessels). Metastasis is typified by a block of fluorescence in the incipient phase and subsequently mild diffuse hyperfluorescence [3]. A very typical image is of haemangioma on indocyanine green angiography – high hyperfluorescence at the beginning and progressive fading of contrast substance (wash out phenomenon). Magnetic resonance imaging is not capable of reliably differentiating choroidal haemangioma from malignant melanoma, both lesions manifest hyperintensity in a T1 weighted image and in a T2 weighted image are iso-intense in comparison with the vitreous body [8]. Differentiation of choroidal haemangioma from central serous chorioretinopathy is possible on the basis of fluorescence angiography. In the case of CSCHR spotted fluorescence is typical. In the late phases the colouring suffuses into the subretinal space and spreads vertically (chimney phenomenon) or horizontally (umbrella phenomenon) [3].

Patients with isolated choroidal haemangioma are typically in middle age, usually without a systemic disorder, state slight deterioration of vision and on the fundus have evident ablation of the neuroretina in the central region. Misdiagnosis as central serous chorioretinopathy is therefore probable, above all in patients manifesting personality

traits typical of patients with CSCHR. It is necessary to be aware of the potential presence of choroidal haemangioma primarily in the case of "non-healing" CSCHR, including in the case of partial improvement of the finding on the fundus following the application of antiedematous therapy. It is necessary to examine carefully above all the region around the temporal arcades. We should be alerted to possible diagnostic error in CSCHR by high ablation of the neuroretina or asymmetry of ablation of the neuroretina

in the macula, which is clearly evident on a colour retinal map on OCT (fig. 27). A study highlighting the same problem of misdiagnosis was published by Rahman [9], and a similar theme, though with regard to diffuse haemangioma, was dealt with by Dave et al. [4].

Conclusion: In young patients, in the case of central serous chorioretinopathy which does not heal spontaneously, it is always necessary to consider the possibility of isolated choroidal haemangioma.

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