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CASE REPORT

Combined central retinal vein and cilioretinal artery occlusion

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Abstract We present a case of combined central retinal vein and cilioretinal artery occlusion which, due to the absence of the temporal branch retinal artery, was initially misdiagnosed as a combined central retinal vein occlusion and temporal branch retinal artery occlusion. Given that – in contrast to cases of combined central artery and central retinal vein occlusion – the prognosis for cilioretinal artery occlusion with central retinal vein occlusion is quite good, this case illustrates the importance of suspecting an unusual condition in the presence of a combined occlusion.

Keywords Central retinal vein occlusion · Cilioretinal artery occlusion · Combined occlusion · Homocysteine · Microperimetry

Case report

A 48-year-old man was referred to our department with blurred vision in his left eye (LE). His visual acuity (VA) was 20/20 in the right eye (RE) and 20/200 in the LE. Fundus examination of the LE demonstrated a whitening of the retina along the distribution of the inferior temporal artery involving the inferior aspect of the macula. The retinal veins were mildly dilated and tortuous and accompanied by adjacent retinal haemorrhages (Fig. 1a). Based on these findings, we suspected a combined central retinal vein and branch retinal artery occlusion. The fundus of the RE was normal.

Surprisingly, fluorescein angiography (FA) demonstrated the presence of a partially occluded cilioretinal artery – and not an inferior temporal retinal artery – which did not fill before 20 s. Venous filling was delayed in all four quadrants but without any signs of capillary non-perfusion (Fig 1b–d). The patient was ultimately diagnosed with a combined central retinal vein and cilioretinal artery occlusion.

Systemic evaluation of the patient was unremarkable. The genetic screening of the 5,10-methylenetetrahydrofolate reductase (MTHFR) gene revealed a single-nucleotide polymorphism, C677T, in a homozygotic state, which was responsible for the moderate elevation of serum homocysteine (28.1 μmol/l).

Early follow-up examinations showed a central and peripheral scotoma (Fig. 2) along the distribution of the occluded abnormal cilioretinal artery. After 1-month, best corrected visual acuity (BCVA) returned to 20/20 LE, although microperimetry (MP-1 Micro-Perimeter; Nidek Technologies, Padova, Italy) revealed a persistent but reduced sensitivity loss in the inferior aspect of the macula (Fig. 3).

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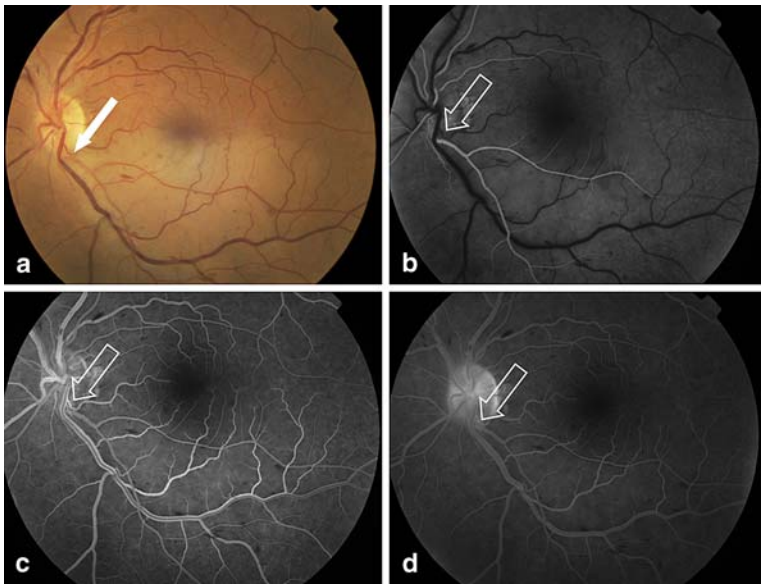

Fig. 1 Color fundus photography demonstrates a whitening of the macula along the anomalous distribution of the cilioretinal artery (*thick arrow*) and mildly dilated and tortuous retinal veins accompanied by adjacent retinal haemorrhages (**a**). Fluorescein angiography frames, from early to late phase, demonstrate the presence of the cilioretinal artery (*open arrows*) filling only after more than 20 s, and delayed venous filling with dilation and tortuosity of the retinal veins in all four quadrants (**b**, **c**, **d**)

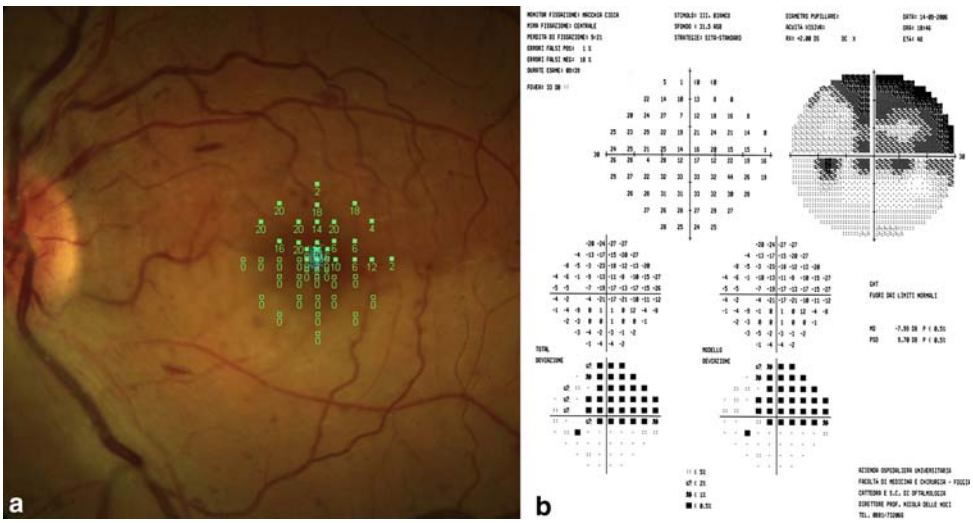

Fig. 2 Fundus-related perimetry and automated static threshold perimetry (Humphrey Visual Field Analyzer model HFA II; Carl Zeiss Meditec) reveal central and peripheral scotoma (**a**, **b**) along the distribution of the occluded cilioretinal artery

Discussion

Combined central retinal vein and cilioretinal artery occlusions were first described by Oosterhuis [1] and are reported to represent 40% of all cilioretinal artery obstructions [2]. The pathogenesis of this rare condition remains controversial. It appears that the cilioretinal artery occlusion occurs subsequent and secondary to the central retinal vein occlusion. Since the perfusion pressure in a cilioretinal artery is lower than in a retinal artery [3–5], a cilioretinal artery occlusion is more likely to be caused by optic disc swelling and/or reduced cilioretinal artery perfusion following central retinal vein occlusion.

Our case is unusual in that the cilioretinal artery has a large retinal distribution completely replacing the inferior temporal artery and supplying the inferior aspect of the macula. In general, the outcome of a combined central retinal vein and cilioretinal artery occlusion depends mainly on the sequelae of the

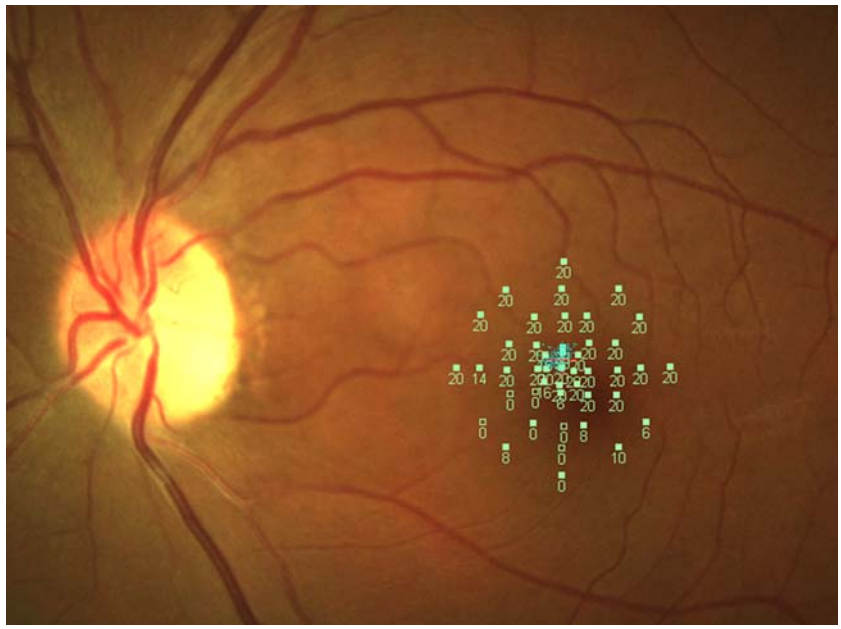

Fig. 3 Fundus-related-perimetry revealed a persistent but reduced sensitivity loss in the inferior aspect of the macula in the 1-month follow-up examination

venous occlusion. Therefore, prognosis for visual acuity is good with 70% of eyes returning to 20/40 or better [2, 6, 7]. In our case, despite vision recovery to 20/20, involvement of the macula resulted in a persisting central scotoma even after resolution of the venous stasis.

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