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A case of cilioretinal artery occlusion: Diagnostic procedures

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ABSTRACT

Purpose: To evaluate characteristic imaging findings and functional outcomes of Cilioretinal Artery Occlusion (CLRAO) associated with giant cell arteritis (GCA).
Observations: We report the case of a 70-year-old woman presenting with sudden vision loss caused by a GCA-associated-CLRAO in her left eye (LE). A thorough ophthalmologic examination together with optical coherence tomography (OCT), OCT-Angiography (OCT-A), fluorescein angiography and fundus autofluorescence were performed. At presentation, the best corrected visual acuity in the LE was 20/200 and funduscopic examination revealed optic disc edema associated with retinal whitening along the area perfused by the CLRA. After 1 month, OCT and OCT-A revealed an improvement of the retinal edema and a partial reduction of the non-perfused areas in the superficial and deep capillary plexuses, as well as in the outer retina and in the choriocapillaris. Fluorescein angiography showed a reduction in the perfusion of the affected area, a delayed perfusion of the temporal sector of the optic disc, as well as areas of choroidal hypoperfusion in the peripheral temporal retina. The patient's visual acuity did not change during the follow up.
Conclusion and importance: Despite a partial recanalization of the occluded vasculature being possible after GCA-associated-CLRAO, the patient's visual prognosis remains poor.

1. Introduction

Approximately 15–30% of the world population has a cilioretinal artery partially or completely supplying the fovea.[1] While the age and sex adjusted annual incidence of central retinal artery occlusion is 1,90 per 100.000,[2] cilioretinal artery occlusion (CLRAO) is an even rarer retinal vascular phenomenon, representing about 5.3%–7.1% of all retinal artery occlusions.[3]

CLRAO has been classified by Brown et al.[4] into three subtypes: non arteritic isolated CLRAO, non arteritic CLRAO associated with central retinal vein occlusion and arteritic CLRAO associated with ischemic optic neuropathy (giant cell arteritis-associated).

Anterior ischemic optic neuropathy (AION) results from acute ischemia of the anterior portion of the optic nerve head which is predominantly supplied by the short posterior ciliary arteries (PCA) and is typically secondary to thrombotic occlusion of the PCA. As the cilioretinal artery originates from the PCA, AION can also result in non perfusion of the cilioretinal artery itself.

2. Case presentation

A 70-year-old woman presented to our clinic with sudden painless loss of vision in her left eye (LE). She had no previous ocular history, whereas her medical record was significant for type 2 diabetes, hypertension and oligoarthritis. On ophthalmic examination, best-corrected visual acuity (BCVA) was 20/32 in the right eye (RE) and 20/200 in the LE. Intraocular pressure was 14 mmHg and 13 mmHg, respectively in the RE and LE, and anterior segment slit-lamp examination was normal for both eyes. Fundoscopy of the right eye was normal, whereas the LE revealed ischemic retinal whitening in the territory of distribution of the cilioretinal artery, whitening of the optic disk and two cotton wool spots above and beneath the optic disk (Fig. 1). Also, on fundus autofluorescence (FAF), a peripapillary temporal shadow effect could be observed (Fig. 1). Accordingly, on optical coherence tomography-

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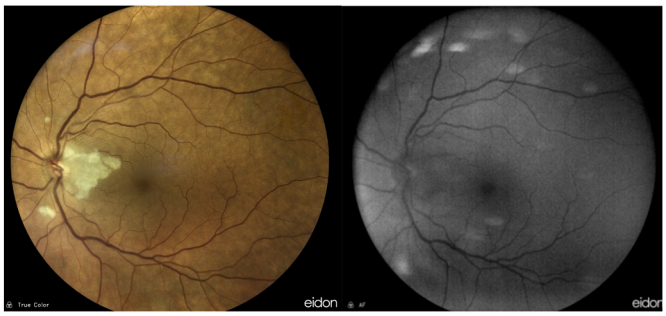

Fig. 1. Funduscopy (left) and Autofluorescence (right) of the LE at baseline.

angiography (OCT-A - Solix Full-range OCT; Optovue Inc.), there was a retinal capillary rarefaction in the superficial and deep retinal plexuses in the area of the cilioretinal artery distribution. The absence of perfusion of the superficial retinal layers and the associated edema led to a masking effect on the outer retina and choriocapillaris layers. Indeed, OCT examination revealed hyperreflectivity and thickening of the inner retinal layers in the peripapillary temporal zone.

Suspecting a case of GCA-related CLRAO, erythrocyte sedimentation rate (ESR), C-reactive protein (PCR) and a color doppler ultrasound of the temporal artery were performed. These examinations resulted highly suggestive for GCA. Therefore, therapy was started immediately.

The patient was prescribed with a loading dose of intravenous corticosteroids, i.e., 1000 mg of methylprednisolone daily for 3 days, followed by oral prednisolone at an initial dose of 1 mg/kg/day which was then progressively tapered. Later on the diagnosis was confirmed by a temporal artery biopsy.

One month after the beginning of treatment, fundus examination showed a less pronounced retinal ischemic whitening and a subtle pallor in the temporal sector of the optic disk, result of CLRAO (Fig. 2). On OCT, there still was retinal hyper reflectivity in the temporal peripapillary zone, but the retinal thickness significantly decreased and the inner retinal layers were disrupted (Fig. 2).

At this time, fluorescein angiography (FA) demonstrated the absence of perfusion in the territory of the cilioretinal artery and a delayed filling of the temporal sector of the optic disk (Figs. 3 and 4). Furthermore, areas of choroidal hypoperfusion appeared in the peripheral temporal retinal sectors (Fig. 5).

Interestingly, on OCT-A there was a partial recanalization of the affected peripapillary areas, with reduction of the non-perfused area in the superficial capillary plexus (Figs. 6 and 7), in the deep capillary plexus (Fig. 6) and, as a result of the reduction of the previous masking effect, also in the outer retina and the choriocapillaris (Fig. 6). Unfortunately, the patient's visual acuity remained low (i.e. 20/200) as the area of infarction involved the papillomacular bundle.

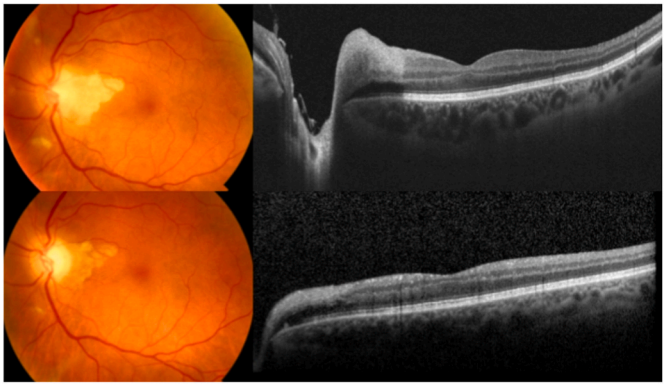

Fig. 2. Fundoscopy (left images) and OCT (right images) at baseline (upper two images) and 1 month after treatment (lower two images).

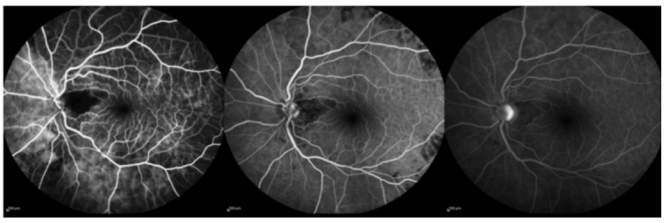

Fig. 3. FA of the posterior pole (early, intermediate and late phases, respectively from left to right) show absence of perfusion in the interpapillary macular region and hyperpermeability of the temporal sectors of the disc.

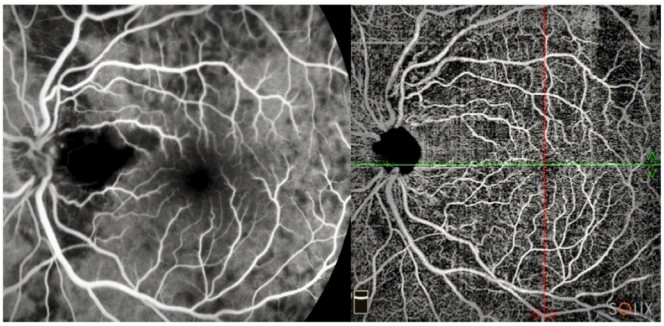

Fig. 4. Magnified figures of the perimacular region on FA (early phase) and OCT-A (Superficial Plexus) after 1 month.

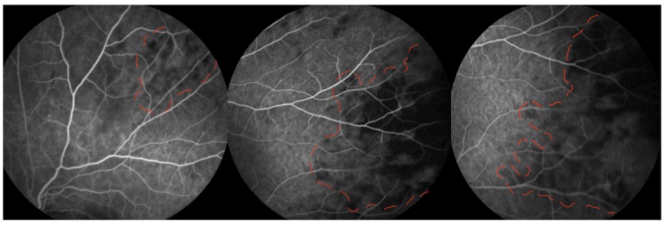

Fig. 5. FA at 1 month follow up showing focal choroidal hypoperfusion areas in the temporal retinal sectors (red dotted area). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

3. Discussion

Cilioretinal artery occlusion has already been reported to be associated with anterior ischemic optic neuropathy.[5] In our case the correlation to GCA was insidious as our patient did not present any of the characteristic symptoms of GCA, i.e., scalp tenderness, headache, or jaw claudication. The risk factors that suggested an association with GCA were the patient's age (over 50-year-old) and the fact that she suffered from autoimmune oligoarthritis. Also, the hypothesis of GCA was supported by the chalky white aspect of the optic disk and by the optic disk perfusion defect on FA. In addition, on FA we noticed areas of choroidal peripheral hypofluorescence compatible with areas of choroidal ischemia caused by hypoperfusion of the posterior ciliary artery in the temporal periphery. Hayreh et al.,[6] noted that choroidal non-perfusion from occluded posterior ciliary arteries is a cardinal angiographic sign of giant cell arteritis and stated that this is "almost diagnostic" of an arteritic etiology. Indeed, the diagnosis of CLRAO associated with GCA can be based on the combination of three findings[6]: a chalky white optic disc, retinal infarct in the region of the occluded cilioretinal artery and the presence of posterior ciliary artery occlusion on FA.

Overall, in the literature there actually is a very small number of studies about CLRAO associated with GCA. Most of the reported cases manifest a poor presenting visual acuity, attributed to both a

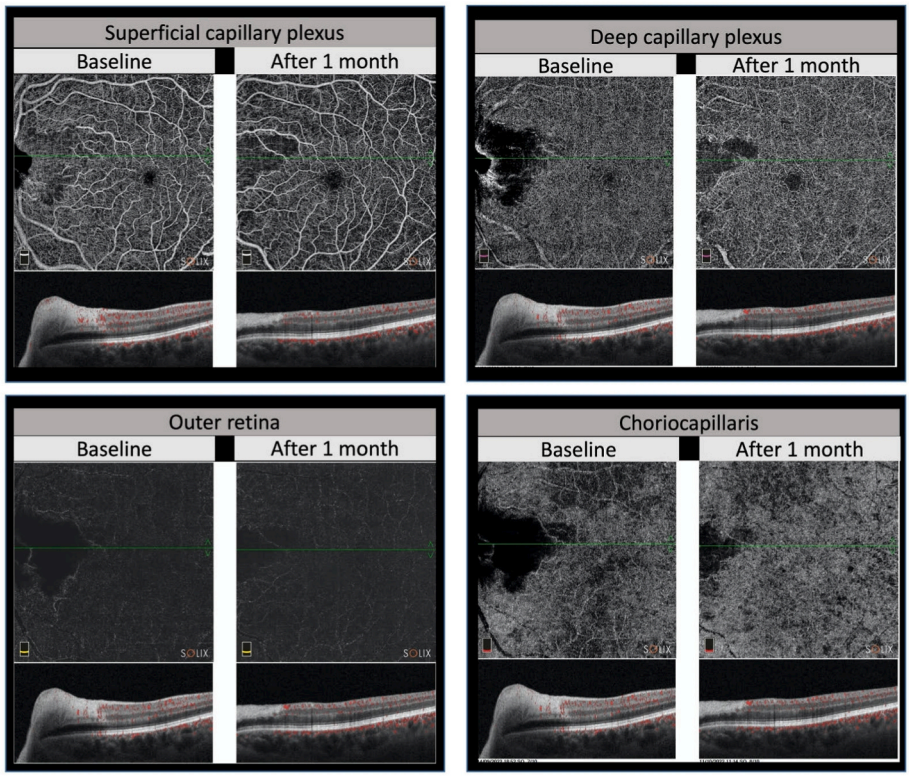

Fig. 6. OCT-A of superficial and deep capillary plexus, outer retina and choricapillaries at baseline and after 1 month. At baseline, the superficial and deep capillary plexuses, show absence of perfusion due to the combination of ischemia and temporal iuxta-papillary edema. After 1 month, resolution of edema results in the reduction of non-perfusion area with persistence of ischemical area and correspective focal fiber atrophy (B-scan).

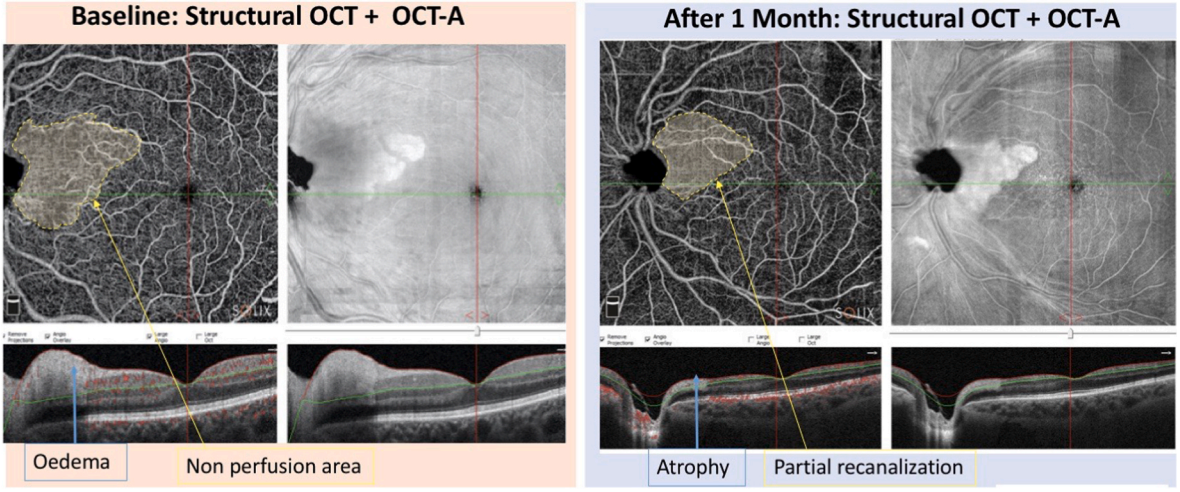

Fig. 7. Details of OCT-A, enface and B-scan structural OCT (superficial capillary plexus) at baseline and after 1 month of treatment. The area of non-perfusion (yellow dotted) was reduced after 1 month due to a partial recanalization of the peripheral capillaries. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

malfunction of the optic nerve and to a suspected additional involvement of the choroidal circulation typically affected in these cases.[7]

Our case is one of the few providing a 1 month follow up of CRLAO associated with GCA. In contrast to other studies,[8] despite the partial improvements noted on OCT-A, in our patient immediate treatment with corticosteroids didn't improve the visual acuity during the follow up. In support of this, Hayreh et al.[8] found that only 4% of eyes affected by Retinal Vascular Occlusion related to GCA improved vision after steroid treatment.

Optical coherence tomography angiography was able to analyze separately each of the singular vascular layers,[9] showing a partial reperfusion of the involved areas. A decrease in the retinal capillary density on OCT-A in eyes affected by CRLAO associated with GCA has already been reported,[10] however, our study is among the first to demonstrate a partial recanalization of the superficial and deep plexuses, the outer retina and the choriocapillaris after 30 days of follow up. These results might suggest that some sort of repairing mechanism involves all four layers during the process of recanalization of neighboring tissue.

4. Conclusion

Giant cell arteritis-associated-CLRAO is a rare event that should be considered as an ocular emergency. Despite a partial recanalization of the involved areas being possible, the visual prognosis of affected patients remains poor, even with early treatment.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Author contributions

Conceptualization, M.C.S. and A.S.; Data curation, C.R and R.K.; Formal analysis, C.F.; Methodology, C.F. and M.C.S.; Project administration, S.R.; Supervision, M.C.S., and S.R.; Writing—original draft, C.R. and R.K.; Writing—review and editing, M.C.S. and R.K. All authors have read and agreed to the published version of the manuscript.

Patient consent

A written informed consent to publish this case report has been obtained from the patient. This report does not contain any personal identifying information.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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