CASE REPORT

Optical Coherence Tomography Angiography of Macula in Chronic Anterior Ischaemic Optic Neuropathy Associated with Giant Cell Arteritis

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ABSTRACT

A 74-year-old man presented with bilateral optic nerve atrophy related to arteritic anterior ischaemic optic neuropathy (AION) which started 22 years ago. Atrophic papilla was noted in both eyes and optical coherence tomography showed thinning of the retinal nerve fibres layer as well as the macular ganglion cell complex. Optical coherence tomography angiography of the macula found a decreased density of the superficial capillary complex, whereas deep capillary complex was well-preserved. Inner retinal layer atrophy in chronic arteritic AION is followed by a loss of vascularisation in these layers as a consequence of the neurodegeneration.

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Introduction

Giant cell arteritis (GCA) is a granulomatous vasculitis which generally affects the large blood vessels of people over 50 years of age such as the aorta, external and internal territory of the carotid or vertebral arteries. Therefore, ophthalmic vascularisation can be involved which can lead to loss of vision through anterior ischaemic optic neuropathy (AION) and/or retinal ischaemia which are the consequence of cilio-retinal and/or central artery occlusions.^{1,2} However, small blood vessels and capillaries can be affected despite the absence of large artery involvement.³

A previous report about chronic arteritic AION has showed a peripapillary capillary dropout in optical coherence tomography angiography (OCT-A).⁴ However, as the chronic AION is accompanied by an alteration of macular inner layers, we aimed to assess the modifications of the vascularisation of the macula in a case of optic nerve atrophy due to arteritic AION.

Case report

A 74-year-old man presented with bilateral optic nerve atrophy related to acute AION which had a 22-year history and was likely due to GCA confirmed

with a biopsy. Visual acuity was limited to competing finger 2 m in the right eye and to light perception in left one. Intraocular lens was normally placed in the posterior chamber and intraocular pressure was 12 mmHg in both eyes. In the fundus, we noted a bilateral white and atrophic papilla associated with an atrophic and thinning aspect of the macula, whereas no abnormalities were seen in the periphery. In both eyes, the Spectral Domain-OCT (SD-OCT; Spectralis, HRA+OCT, Heidelberg, Germany) showed a bilateral thinning of the retinal nerve fibre layer (RNFL) as well as of the macular ganglion cell complex (mGCC) (Figure 1). Furthermore, in a 3 × 3-mm macular cube of OCT-A (Spectralis, HRA+OCT), we noted a decreased density of the superficial capillary complex (SCC) while the density of the deep capillary complex (DCC) and choriocapillaris were well-preserved in both eyes as well as the integrity of the foveal avascular zone (Figure 2).

Discussion

In this case, there was a long history of AION leading to a well-established visual impairment. The optic nerve atrophy was the consequence of a significant optical fibre loss leading to an

Figure 1. In fundus of both eyes, the papilla has an atrophic white aspect while macula was thin. In the SD-OCT of the right and left eyes, both RNFL and mGCC were deeply altered with a severe optic fibre loss.

atrophic papilla in fundus transposed on the SD-OCT by a deep thinning of RNFL. In addition, we noted an important reduction of mGCC thickness and which was accompanied by a low visualisation of SCC in OCT-A which normally corresponds to the capillaries located in the inner retinal layers. At the same time, there was a normal thickness of the intermediate retinal layers with a well-preserved visibility of DCC located in the retinal layers.

In GCA, cotton-wool spots can be found in the fundus which are probably caused by focal retinal ischaemia due to microembolisation of capillaries associated with vasculitis of large vessels.⁵ It is less likely to be a consequence of local small vessels vasculitis.⁶ Generally, this focal ischaemia is not homogeneously spread providing a spot-like appearance.⁷ In our case, the decreased density of SCC was homogeneously spread and well-established and cannot be explained by only a small artery focal occlusion. Furthermore, some authors have already demonstrated in the OCT-A that the focal non-perfusion in arteritic AION was inconsistent and was located only in peripapillary the region with normal macular perfusion.⁸ The likely hypothesis of this decreased SCC density may be the collapse of the capillary as a consequence of low oxygen demands in the superficial atrophic layers which contained only a few remaining optic fibres. The other explanation is the definitive

and complete atrophy of the capillaries of SCC accompanying by the neurodegeneration. These hypotheses are strengthened by the observation of an early structural damage in glaucoma without vascular damage in the OCT-A. This suggests that the appearance of neurodegeneration was prior to the capillary dropout which was probably only the consequence of this optic fibre atrophy.⁹

Additionally, the persistence of the normal density of DCC suggests that the absence of SCC was not linked to the occlusion of large or medium blood vessels supplying both superficial and deep capillaries nor to vasculitis of small vessels which can involve superficial and/or deep capillaries. In summary, the visualisation of this DCC can be explained by the model of retinal vascularisation proposed by Paques et al. in which vessels supplying the DCC emerge from underside of large vessels. Provided that OCT-A gives an enface visualisation, these feeding vessels of DCC are invisible most probably because they were only masked by the large vascular trunks and were still present.¹⁰ Lastly, the only capillaries really lost were probably only those supplying the inner retinal layers while those intended to outer layers remained despite their emergence from the same vessels.

Finally, in a case study of non-arteritic AION, the authors have reported a peripapillary capillary dropout that correlated to the topography of peripapillary

Figure 2. In OCT-A, the visibility of the SCC was deeply altered with large and homogenous areas of capillary dropout while the DCC was normally visible with a good preservation of the integrity of the foveal avascular zone.

and macular optic fibre loss found in SD-OCT.¹¹ To best of our knowledge, this report is the first case of the OCT-A imaging of the macula in a patient who presented an optic nerve atrophy due to GCA. We show that the optic nerve atrophy was accompanied by the atrophy of both ganglion cell and their corresponding capillaries. The capillary inflammation seemed did not appear to be implicated in this capillary loss which was probably the consequence of GCC atrophy rather its cause.

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