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BRIEF COMMUNICATION

Optical coherence tomography angiography in nonarteritic anterior ischemic optic neuropathy due to optic nerve head drusen

Gilda Cennamo[1] · Daniela Montorio[2] · Piera Giunta[2] · Fausto Tranfa[2]

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Abstract

Optic nerve head drusen (ONHD) are typically benign hyaline and calcified concretions located within the optic nerve. Initially asymptomatic, they can increase slowly in size and number over time, and they can cause mechanical stress at the axons of the optic nerve and progressively lead to a loss of the visual field and papillary vascular changes. Here, we reported a rare case of a young patient with bilateral ONHD complicated by a nonarteritic anterior ischemic optic neuropathy (NAION) in the right eye. The aim of this case was to evaluate, using optical coherence tomography angiography (OCTA), the vascular network of the papillary region in both eyes in the presence of this ischemic event associated to ONHD. At OCTA examination, the whole papillary region revealed a general rarefaction of the vascular network, but it turned out to be greater in the right eye affected by nonarteritic anterior ischemic optic neuropathy than in fellow eye. Therefore, these findings demonstrated that ONHD may determine significant ischemic events of the optic nerve and OCTA represents a valid and noninvasive tool in the diagnosis and in the pathogenesis of these rare cases of NAION associated to ONHD in young patients.

Keywords Optical coherence tomography angiography · Nonarteritic anterior ischemic optic neuropathy · Optic nerve head drusen

Optic nerve head drusen (ONHD) are typically benign hyaline and calcified concretions located within the optic nerve and are more prevalent in Caucasians and women [1]. They appear in childhood and are usually asymptomatic; indeed, the diagnosis is accidentally made during a routine eye examination [2].

The pathophysiology underlying ONHD may be associated to an inherited dysplasia of the disc, and based on anatomical location within the optic nerve, they can be superficial and deep [3].

ONHD may occur alone, without any other ocular disease, or in association with other syndromes such as retinitis pigmentosa, Usher, Joubert, and Alagille [4].

The drusen can increase slowly in size and number over time, and they can cause mechanical stress at the axons of the optic nerve leading progressively to a loss of visual field. Cases of optic nerve ischemia, such as anterior ischemic optic neuropathy, occur rarely [2].

A detailed analysis of the retinal vascular network can be performed by the optical coherence tomography angiography (OCTA) that allows to detect the papillary vascular network alterations in the presence of ONHD in noninvasive way [5].

Here, we reported a case of bilateral ONHD complicated by a nonarteritic anterior ischemic optic.

A 15-year-old woman with previous diagnosis presented a sudden decreased visual acuity in the right eye. Best corrected visual acuity (BCVA) was 0.22 logMAR in the right eye and 0.0 logMAR in the left eye, and the anterior segment did not show alteration.

Fundus examination revealed in the right eye, the presence of an anterior ischemic optic neuropathy characterized by the optic disc with edema, blurred margin and peripapillary retinal hemorrhages nasally to the macula. The fellow eye showed normal visual acuity and blurred margin optic disc due to the presence of ONHD but without peripapillary retinal hemorrhages at fundus examination.

The magnetic resonance imaging did not show any optic chiasm/nerve compression due to intracranial lesion.

✉ Gilda Cennamo
xgilda@hotmail.com

[1] Department of Public Health, Eye Clinic, University of Naples Federico II, Via S. Pansini 5, 80133 Naples, Italy

[2] Department of Neurosciences, Reproductive Sciences and Dentistry, University of Naples Federico II, Naples, Italy

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Laboratory exams including blood cell counts, erythrocyte sedimentation rate, and C-reactive protein level presented normal values.

Spectral domain (SD)-OCT (Spectralis, Heidelberg Engineering, Heidelberg, Germany) displayed in both eyes the presence of ONHD that appeared as hyperreflective round structures located anterior to the lamina cribrosa.

Moreover in right eye, the subretinal and intraretinal fluid were observed in papillary and peripapillary regions involving also the macular region, while the left eye revealed no alteration in macular region (Fig. 1A, A1).

The patient underwent XR Avanti AngioVue OCT Angiography (software ReVue version 2017.1.0.151, Optovue Inc., Fremont, CA, USA), a device based on split-spectrum amplitude de-correlation algorithm (SSADA) [6]. The AngioVue disc mode automatically segmented the radial peripapillary capillary (RPC) plexus analyzing the whole papillary region with an area scan of 4.5 × 4.5 mm in the superficial retinal layers that extended from the inner limiting membrane to the retinal nerve fiber layer posterior boundary [7].

Moreover, the software AngioAnalytic™ automatically calculated in the RPC plexus the vessel density (VD), defined as the percentage area occupied by vessels in the analyzed region on the OCTA en-face images [8].

At OCTA examination, the right eye revealed a greater rarefaction of the vascular network with an impaired VD in the whole papillary region with respect to fellow eye. The VD of the RPC whole region was 36.1% for the right eye and 40.4% for the left eye (Fig. 1B, B1).

Several studies reported vascular occlusive diseases associated to ONHD, such as anterior ischemic optic neuropathy, central retinal artery occlusion, and central retinal vein occlusion [5, 9–11].

ONHD represent a significant cause of the alteration in axoplasmic transport of the optic nerve [2] because the accumulation of these calcified aggregates determines an axonal compression with consequent thinning of the retinal nerve fiber layer, detected by OCT, and a slow and progressive visual field loss, such as the enlargement of the blind spot and peripheral alterations [4, 5, 9, 10, 12].

The precise mechanism by which ONHD causes the anterior ischemic optic neuropathy is not yet defined; probably the drusen, narrowing the optic disc space, could increase the possibility that ischemic events occur [13–18].

Consequently, the reduced blood flow, due to increased optic disc tissue pressure, may cause a visual loss [5] and often an inferior altitudinal or arcuate visual field defect [14].

OCTA is an objective method, which is helpful in the analysis of the papillary vascular network in the presence of ONHD, as confirmed by a study that demonstrated a significant reduction in papillary blood flow and vessel density in patients with ONHD with respect to healthy eyes [15].

Indeed, Gaier et al., using OCTA, revealed a focal attenuation in the superficial capillary network of the optic disc associated to the presence of drusen [5]. Also the study conducted by Abri Aghdam et al. showed a reduced vessel density of the peripapillary region and inside disc in eyes with ONHD with respect to normal eyes [19].

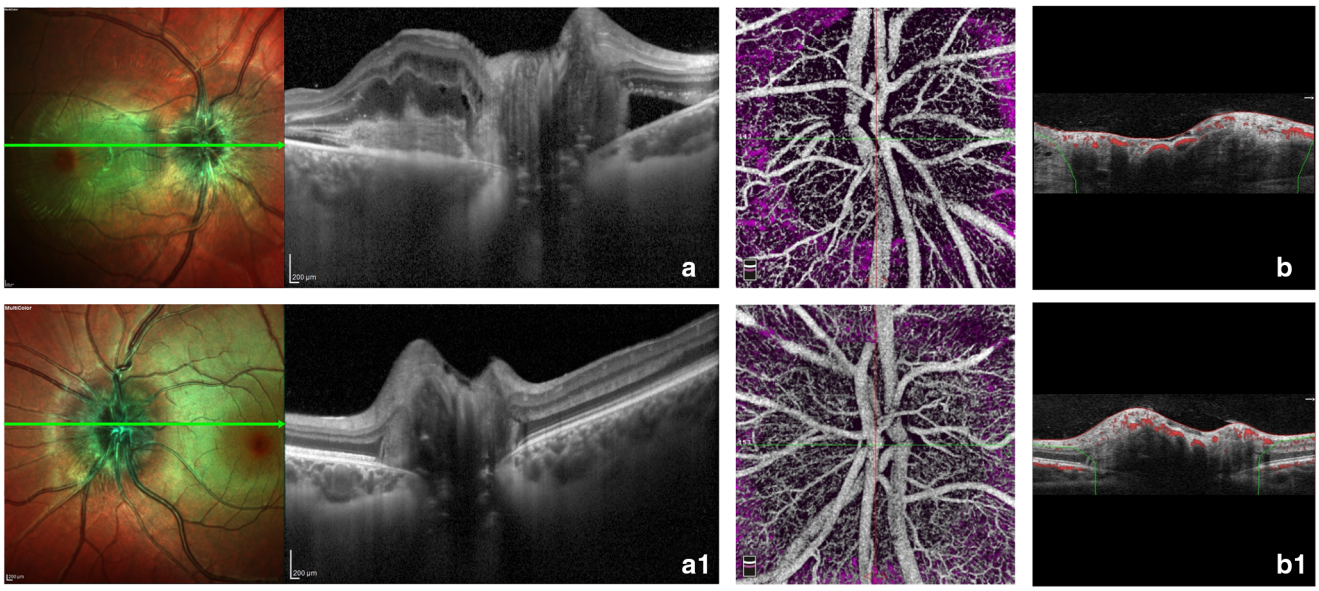

Fig. 1 A patient (female, 15 years old) with bilateral optic nerve head drusen (ONHD) complicated by nonarteritic anterior ischemic optic neuropathy in right eye. At structural spectral domain optical coherence tomography (SD-OCT), the ONHD in both eyes appear as hyperreflective round structures located anterior to the lamina cribrosa. In right eye, the subretinal and intraretinal fluid were observed in the papillary and peripapillary regions involving also the macular region (A), while no macular alteration was observed in the left eye (A1). OCT angiography (OCTA) in the right eye revealed a greater rarefaction of the papillary vascular network (B) than in fellow eye (B1)

In this case, the OCTA, in noninvasive way, allowed to detect the impairment of the papillary vascular network, confirming that the ONHD determined significant ischemic events of the optic nerve.

In conclusion, OCTA is a safe imaging technique that improves the diagnosis and the pathogenesis of these rare cases of NAION associated to ONHD.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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