Case Report

Acute Macular Neuroretinopathy in a Patient with Retinal Vascular Tortuosity

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Keywords

Acute macular neuroretinopathy · OCT angiography · Retinal vascular tortuosity · Retinal capillary plexus

Abstract

We report a rare case of a young woman with acute macular neuroretinopathy (AMN) in the right eye and concomitant retinal vascular tortuosity in both eyes. A 19-years-old woman presented with a sudden loss of central vision in the right eye. Apart from flu-like infection 2 weeks before the onset of symptoms, she reported overall good health. She used oral contraceptive pills. Multimodal imaging techniques including color fundus photography, fundus autofluorescence, infrared reflectance imaging, fluorescein angiography, swept-source optical coherence tomography (SS-OCT), and visual field assessment were used for the diagnosis of AMN as well as disease monitoring during follow-up. At presentation, ophthalmoscopy revealed a reddish parafoveal lesion, while SS-OCT showed hyper-reflectivity in the outer plexiform and outer nuclear layers with a slightly disrupted inner segment/outer segment junction. All these imaging findings indicated AMN, but the interpretation was slightly difficult due to the presence of tortuous retinal arteries in both eyes. During the disease course, functional and morphological recovery was documented at 1- and 6-month follow-up. However, as the abnormal appearance of the retinal vessels did not change, congenital retinal vascular tortuosity was diagnosed. Since the pathogenesis of AMN has not been fully elucidated, there is currently no effective treatment. Numerous studies have emphasized a vascular origin and the key role of ischemia in AMN. Our rare case suggests that congenital tortuosity of the retinal vessels, although constituting a common finding in healthy individuals, may be involved in the pathophysiology of the disease.

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Introduction

Acute macular neuroretinopathy (AMN) is a rare condition that typically affects young women and presents with 1 or more sudden-onset paracentral scotomas as well as transient or permanent visual impairment [1]. On clinical examination, reddish-brown, wedge-shaped lesions are observed around the center of the macula, corresponding to scotomas in vision. The etiology of AMN remains unclear although microvascular abnormalities in the deep retinal capillary plexus have been implicated [2]. Some factors have also been associated with a higher risk of AMN, such as flu-like symptoms (e.g., fever) [3], use of oral contraceptive pills [4], and use of sympathomimetic drugs (e.g., adrenaline and epinephrine) [3]. The differential diagnosis should include paracentral acute middle maculopathy, which has a similar clinical presentation to that of AMN. However, in the case of paracentral acute middle maculopathy, swept-source optical coherence tomography (SS-OCT) demonstrates lesions in the inner plexiform and inner nuclear layers of the retina [5]. We report an atypical case of AMN that developed in an eye with tortuous retinal vessels.

Case Report

A 19-years-old female patient complained of a sudden loss of central vision in the right eye. Best-corrected visual acuity was 20/200 in the right and 20/20 in the left eye. Intra-ocular pressure was normal. Slit-lamp biomicroscopy of the anterior segment of the eye was unremarkable. Multicolor ophthalmoscopy revealed a reddish-brown parafoveal lesion in the nasal region of the right macula and normal left macular morphology (Fig. 1a, b). Additionally, infrared reflectance images showed tortuous retinal arteries in both eyes, with a parafoveal hyporeflective area in the right eye (Fig. 1c, d). The patient complained of flu-like infection 2 weeks earlier but was otherwise generally healthy. Her laboratory tests showed D-dimer levels of 0.3 mg/L (reference range <0.55 mg/L); fibrinogen, 2.3 g/L (reference range, 1.8–3.5 g/L); and C-reactive protein, 15 mg/L (reference range <3 mg/L). An SS-OCT (Topcon DRI OCT Atlantis, Japan) scan of the right eye showed hyper-reflectivity primarily in the outer plexiform and outer nuclear layers, with subsequent involvement of the inner segment/outer segment junction (Fig. 2a, b). On OCT angiography (OCTA), we observed areas of lower capillary density in the nasal parafoveal quadrant (Fig. 3; white arrow) with hyperdensity (Fig. 3; red arrow) due to projection artifacts cast from superficial blood vessels and a hyper-reflective lesion (Fig. 3). On fluorescein angiography, no leakage or ischemia was shown. Moreover, as a cilioretinal artery was absent, the location of the intraretinal hyper-reflectivity did not correspond to its supply area. However, tortuosity of the retinal vessels in the right and left eye was revealed. The only administered treatment was topical dexamethasone (0.1%, 4 times/day). The patient underwent an ophthalmological examination at 1- and 6-month follow-up, which showed resolution of pathological changes on SS-OCT and OCTA, improvement of best-corrected visual acuity up to 1.0 in the right eye, as well as resolution of scotoma in the central visual field. This outcome is in line with another report describing that lesions usually resolved after 4 weeks to few months [6].

Discussion

AMN was first described by Bos and Deutman [4]. The reported risk factors include flu-like infections [3], use of oral contraceptive pills, and even nonocular trauma [7].

Fig. 1. Multicolor image of the right eye, showing a reddish-brown parafoveal lesion in the nasal quadrant of the macula (yellow arrow) with abnormal morphology of the fovea and retinal vascular tortuosity (**a**); the left eye, showing less severe retinal vascular tortuosity compared with the right eye (**b**). Infrared reflectance image of the right eye, showing abnormal morphology of the fovea (**c**); the left eye, showing normal morphology of the macula (**d**).

The present case emphasizes the usefulness of various multimodal imaging techniques, especially OCTA, in the correct diagnostic workup of such patients. Although data on the links between alterations in the deep capillary plexus and AMN are inconsistent, Sarraf et al. [2] reported AMN to be associated with retinal ischemia. To our knowledge, no reports have described a link between AMN and congenital tortuosity of the retinal vessels. Given the association between the tortuosity of superficial retinal vessels seen on ophthalmoscopy and infrared reflectance image as well as alterations in the deep retinal plexus on OCTA [8], we hypothesize that abnormalities of the retinal vessels may be involved in the etiology of AMN. However, it is important to note that the disease may be induced by flu-like symptoms in predisposed individuals. The potentially reduced blood flow in tortuous vessels and the presence of other factors that increase blood density and coagulability in outer retinal layers may contribute to the development of abnormalities. Our findings are in line with previous reports [1, 7, 9, 10].

Retinal vascular tortuosity is a group of different disease entities such as bilateral congenital vascular tortuosity [11], congenital retinal arteriovenous communication [9], or familial retinal arteriolar tortuosity [12]. Retinal vascular tortuosity itself can increase the resistance of blood flow or even lead to blood flow obstruction [9]. Moreover, tortuosity of

Fig. 2. a, b SS-OCT scans of the right eye, showing hyper-reflectivity in the outer plexiform layer and outer nuclear layers (red arrow), with a slight disruption of the inner segment/outer segment (green arrow). SS-OCT, swept-source optical coherence tomography.

Fig. 3. OCTA scans showing higher capillary density areas in the nasal macular below a hyper-reflective lesion on SS-OCT (red arrow), with a lower capillary density area nasally to the fovea (white arrow). OCT, optical coherence tomography; SS-OCT, swept-source optical coherence tomography; OCTA, OCT angiography.

retinal vessels has been postulated as an indicator of retinopathy, arterial hypertension, disease of cerebral vessel, ischemic heart disease, and stroke [10]. Ocular complications of vessel's tortuosity include retinal vein occlusion (45%), retinal hemorrhages (33%), macular edema, and neovascular glaucoma [12–14]. Kuriakose et al. [7] reported cotton wool spots and retinal hemorrhages in their patient, which highlights the role of ischemia of the deep retinal capillary plexus in AMN. Previous studies emphasized the ischemic and inflammatory etiology of AMN; therefore, we believe that the coexisting vascular alterations may predispose patients to the development of this disease. The fundoscopic findings of AMN and cotton wools may also suggest hypertensive retinopathy and choroidopathy due to hypertensive crisis [15, 16] or roller coaster retinopathy described by Patel et al. [17].

Although the exact etiology of AMN is still unclear, our case suggests that retinal vascular tortuosity may be involved in the pathophysiology of this rare disease. This is in line with the observations of Kuriakose et al. [7], who also revealed significant tortuosity of the retinal vessels in a patient with AMN. Such a hypothesis is also supported by the presence of reduced DCP flow in the macula in the course of AMN, as revealed by projection-resolved OCTA [5]. However, further studies on a larger population of patients are needed to provide definitive conclusions.

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Statement of Ethics

The authors have no ethical conflicts to disclose. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

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Author Contributions

All the authors contributed to the study concept and design, data collection and analysis, as well as the preparation of the material. The first draft of the manuscript was written by Izabella Karska-Basta, and all the authors provided their comments. All the authors read and approved the final version of the manuscript.

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