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Acute Macular Neuroretinopathy Associated With Idiopathic Intracranial Hypertension

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Acute macular neuroretinopathy (AMN) has recently been described in association with optic neuritis (1). Hence, we describe a case of AMN associated with idiopathic intracranial hypertension (IIH).

An 18-year-old woman presented in emergency with complaints of blurred vision with moderate headache and tinnitus. Her medical history was unremarkable apart from asthma treated with salbutamol and use of an estroprogestative contraceptive pill. She smoked 1 pack per day for 3 years. No alcohol use was reported. She did not report a recent viral illness, particularly no COVID-19 infection nor recent vaccination.

The visual acuity was 20/20 in the right and left eyes. Intraocular pressure was normal. There was no relative afferent pupillary defect. Extraocular motility was intact and full. Anterior segment examination was unremarkable, but funduscopic examination revealed bilateral Frisen grade 3 papilledema. Macular spectral domain optical coherence tomography (SD-OCT) (Heidelberg SPECTRALIS; Heidelberg Engineering, Germany) revealed normal foveal profile and peripapillary intraretinal fluid (Fig. 1A, F). General examination was unremarkable, with a body weight of 55 kg for a height of 160 cm (body mass index: 21.5 km/m^2), and no recent weight gain. Blood pressure was 132/88 mm Hg. Injected brain CT scan and injected brain MRI were normal. Blood tests, including antinuclear antibodies, procoagulant profile, and C-reactive protein, and serological test for CMV, EBV, HSV1, HSV2, HHV-6, HPV, VZV, syphilis, and SARS-CoV-2 were normal. Lumbar puncture found an opening pressure of 30 cmH2O, with no associated biochemical, cytological, or microbial abnormalities leading to the diagnosis of IIH. Treatment with acetazolamide 250 mg 6 tablets/day was started.

We reviewed the patient 3 days later as she reported a worsening of her visual symptoms, with a feeling of paracentral scotoma. Goldman visual field revealed only bilateral blind spot enlargement, while static visual field 24-2 testing (MonCV3, MetroVision, Perenchies, France) revealed bilateral paracentral temporal scotoma associated with blind spot enlargement. Optic disc examination was unchanged, but fundus examination revealed a wedge-shaped paracentral lesion pointing to the fovea in both eyes. Near-infrared reflectance showed hyporeflective paracentral lesions and macular OCT showed corresponding disruption of the ellipsoid zone (EZ) and interdigitation zone (IZ) with outer nuclear layer (ONL) hyperreflectivity, leading to a diagnosis of AMN (Fig. 1B, G).

Complete resolution of the papilledema was obtained after 2 months of treatment with acetazolamide, with no damage to the ganglion cell layer in both eyes. The visual acuity remained 20/20 in both eyes. ONL hyperreflectivity resolved in both eyes within 1 month (Fig. 1C, H). EZ loss of signal resolved in 6 months in the right eye (Fig. 1J) and in 1 month in the left eye (Fig. 1C). However, the loss of signal corresponding to the outer segment (IZ) of the photoreceptors (cones) partially resolved for the left eye, but not for the right eye, 6 months after disease onset (Fig. 1E, J). As a result, static visual field of

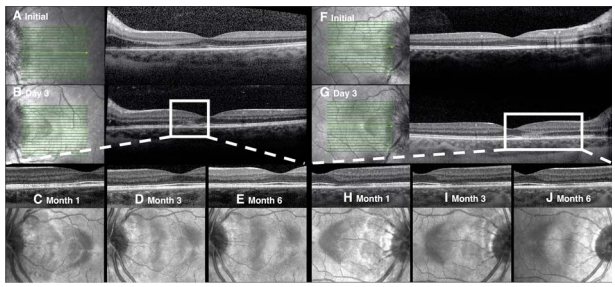

FIG. 1. A and F. Initial spectral domain optical coherence tomography (SD-OCT) and corresponding near-infrared reflectance (NIR) images of the right eye and left eye demonstrating normal foveal profile, no subretinal fluid and peripapillary intraretinal fluid. **B and G.** NIR reflectance of the right and left eyes obtained 3 days after presentation showing arrowhead-shaped hyporeflectance pointing toward fovea. Corresponding macular SD-OCT revealing disruption of the ellipsoid zone (EZ) and interdigitation zone (IZ) and ONL hyperreflectivity. **C, D, and E.** Left eye EZ and IZ disruption resolved spontaneously over months so as NIR hyporeflectance. **H, I, and J.** Right eye EZ disruption resolved within 6 months in contrast to the IZ which partially resolved so as NIR hyporeflectance. ONL, outer nuclear layer.

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The authors report no conflicts of interest.

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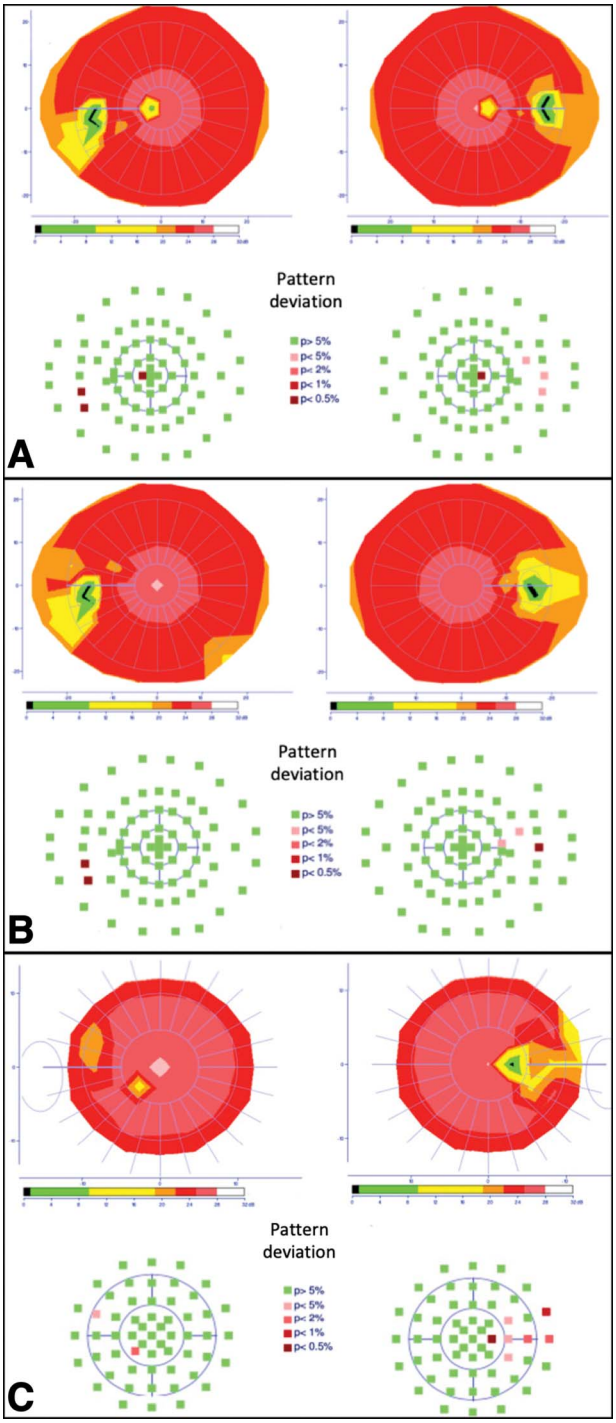

FIG. 2. A. Day 3 metrovision automated 24-2 perimetry illustrates bilateral temporal paracentral scotomas associated with bilateral blind spot enlargement. **B**. Follow-up metrovision automated 24-2 perimetry 6 months later demonstrates near complete resolution of paracentral scotoma in the left eye, with persistant paracentral scotoma in the right eye. **C**. Follow-up metrovision automated 10-2 perimetry 6 months later demonstrates near complete resolution of paracentral scotomas in the left eye with persistent arrowhead-shaped temporal scotoma pointing toward fovea in the right eye.

the right eye still showed a paracentral scotoma with a triangular shape visible on the central 10° visual field, while the left eye visual field was almost normal. The area of NIR hyporeflectivity had almost disappeared at 6 months in both eyes (Fig. 2).

We describe a case of AMN associated with IIH. In the presented case, the OCT imaging shows the absence of subretinal fluid and the typical hyperreflective lesion in the ONL with altered EZ, associated with a paracentral scotoma corresponding to the lesioned area (2).

AMN was first described by Bos and Deutman in 1975 (3). It is believed to result from an impairment of the deep capillary plexus (DCP), possibly leading to a disruption of Müller cells in the Henle fiber layer (4). However, AMN has become an umbrella term used to define any acute disruption of the photoreceptor outer segment/retinal pigment epithelium interface. Although similar on SD-OCT, it is possible that the pathophysiology of our patient's AMN—having IIH—is different from those initially described by Bos. On the other hand, our patient also shared known risk factors such as young age and use of estrogen–progestin pills, and IIH may have been a trigger for AMN in this patient. In addition, Deschamps et al showed an association between AMN and optic neuritis, suggesting a common pathophysiology pathway between AMN and optic neuropathy (1).

In conclusion, we describe a case of AMN-like lesion in the context of IIH. This case highlights that the onset of a paracentral scotoma in a patient with IIH may be related to the development of AMN and progression of the optic neuropathy related to papilledema. Further studies are needed to establish frequency, causality, and prognosis of AMN in IIH.

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