Acute Macular Neuroretinopathy Presenting as Bitemporal Defects on Humphrey Visual Field

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Abstract

A 25-year-old woman presented with blurry vision, headache, nausea, and syncope. Humphrey visual field testing revealed bitemporal defects, but magnetic resonance imaging was negative for chiasmal pathology. Macular optical coherence tomography showed focal parafoveal disruption of the photoreceptor inner segment/outer segment junction and infrared imaging showed hyporeflective macular lesions in both eyes. Our case demonstrates a diagnosis of acute macular neuroretinopathy that presented with bitemporal visual field defects. To our knowledge, bitemporal visual field loss, mimicking chiasmal pathology, has not been reported previously in association with acute macular neuroretinopathy.

Keywords: retinal diseases, optical coherence tomography, visual fields, paracentral scotoma, retinal inner segment

Introduction

Acute macular neuroretinopathy (AMNR) is a rare disorder initially described by Bos and Deutman in 1975 as a disease of the inner retina.1 Contrary to early beliefs, newer imaging modalities show that the pathology of AMNR involves the outer retina.2 Acute macular neuroretinopathy primarily affects young women and is characterized by acute onset of mild visual impairment, distinctive macular lesions, and paracentral scotomas. The characteristic macular lesions may take days or weeks to develop, often making diagnosis by biomicroscopy challenging early on.3 Visual loss can be unilateral or bilateral, and is commonly transient, but may be permanent.4 We report a patient with AMNR who initially presented with headache, syncope, normal fundus exam, and bitemporal visual field defects, suggesting a posterior chiasmal lesion. Our case provides an example of retinal pathology mimicking a central nervous system lesion, and underscores the utility of optical coherence tomography (OCT) and infrared photography in the diagnosis of AMNR.

Case Report

A 25-year-old woman with no past medical history was referred to our office for a one-day history of sudden onset blurry vision in both eyes. Three days prior to presentation she developed headache, nausea, and post-nasal drip. The following day she was admitted to an outside hospital for evaluation of a syncopal episode. She was diagnosed with dehydration and treated with intravenous fluids. No neuro-imaging was performed. The day prior to our visit, she woke up with blurry vision in both eyes. She saw an outside ophthalmologist who reported a normal dilated fundus exam (DFE), but Humphrey visual field (HVF) testing revealed bitemporal visual field defects prompting immediate referral to our office for neuro-ophthalmologic evaluation.

She denied any history of menstrual cycle irregularity or galactorrhea. Her medications included daily oral contraceptives and over-the-counter acetaminophen with phenylephrine HCl as needed for sinus headache and pain. Visual acuity was 20/20 OU. Review of her Humphrey visual field (HVF) testing revealed bitemporal visual field defects prompting immediate referral to our office for neuro-ophthalmologic evaluation.

She was sent for emergent MRI of the brain and sella to rule out pituitary apoplexy. The imaging results were negative.
The patient returned to clinic two days later where repeat visual field testing again showed bitemporal paracentral scotomas, although slightly improved. There was no thinning of the retinal nerve fiber layer or ganglion cell layer in either eye on OCT, but disruption of the inner segment/outer segment (IS/OS) junction was noted on the horizontal B scan portion of the ganglion cell layer analysis. This prompted us to obtain a dedicated macular OCT, which clearly showed focal parafoveal disruption of the photoreceptor IS/OS junction (Figure 3). These parafoveal lesions, although not detected on fundus exam, were easily visualized with infrared imaging leading to a diagnosis of acute macular neuroretinopathy.

Discussion

Acute macular neuroretinopathy is a rare disorder with no known treatment that primarily affects young women of reproductive age. Since its description in 1975 to December 2014, 101 cases of AMNR have been reported. Patients typically present with paracentral scotomas which correspond to reddish-brown wedge shaped macular lesions, better seen on red-free imaging. Patients report sudden unilateral or bilateral visual impairment that may improve, resolve, or persist. Though the etiology and pathophysiology remain uncertain, AMNR is thought to be caused by retinal ischemia, and may be associated with a variety of conditions including viral illness, migraines, trauma, hypotension, cocaine, caffeine, oral contraceptives, leukemia, dengue fever, ulcerative colitis, chronic kidney disease, systemic lupus erythematosus, and adrenergic agonists. To our knowledge, there have been no prior reports of AMNR presenting with bitemporal visual field defects mimicking a lesion of the optic chiasm.

Bitemporal visual field defects that respect the vertical meridian are characteristic of chiasmal pathology. This is due to crossing of the nasal fibers from each eye, which carry visual information from the temporal visual field. The macular fibers cross in the posterior chiasm and thus posterior chiasmal lesions may cause central bitemporal hemianopic scotomas, as seen in our patient. Pituitary adenomas are the most common cause of chiasmal compression and generally produce slowly progressive vision loss. Hemorrhage or infarction of the tumor is a potentially life threatening condition known as pituitary apoplexy, which classically presents with headache, nausea, altered consciousness and acute vision loss or double vision. Thus we initially did not suspect macular pathology when our patient presented with sudden bitemporal vision loss associated with headache, nausea, syncope, and a normal-appearing fundus. Although chiasmal compression causes bitemporal visual field defects, bitemporal paracentral scotomas are rare, and should prompt careful evaluation of the retina.

Acute macular neuroretinopathy often poses a diagnostic challenge, as it may present with no detectable macular lesions on retinal biomicroscopy making diagnosis more difficult in the absence of additional imaging. One review found that only 24.4% of eyes had the characteristic wedge shaped macular lesions seen on DFE and 5.8% had no clinically identifiable changes. Patients may undergo extensive neurologic workup with repeat
MRIs and even lumbar punctures before reaching the correct diagnosis. Spectral domain OCT, infrared reflectance (IR), autofluorescence, OCT angiography, and multifocal electroretinography have all been used to diagnose and monitor AMNR. Spectral domain OCT is a highly sensitive technology for use in AMNR, detecting one or more abnormalities in 98.7% of cases. AMNR initially manifests as hyperreflectivity of the outer plexiform layer on OCT, which may precede the appearance of macular lesions on DFE. This is followed by progressive disruption of the IS/OS junction. Outer nuclear layer thinning gradually develops, and is a chronic finding of AMNR.

Sarraf et al. proposed a new variant called paracentral acute middle maculopathy or Type 1 AMNR, suggesting there are two variations of this disease distinguishable on OCT: Type 1 (above the outer plexiform layer (OPL)), with a hyperreflective band in the outer plexiform layer/inner nuclear layer (INL) region and subsequent INL thinning, and Type 2 (below the OPL), as previously described above. Infrared imaging reveals hyperreflective lesions more definitively than those seen on DFE. The edges of the IR lesions are found to correspond most strongly to the outer segment/retinal pigment epithelium line, as opposed to the IS/OS junction. Fundus autofluorescence may be normal or show hyperautofluorescent lesions or hypoautofluorescent lesions surrounded by hyperautofluorescence. Fluorescein angiography (FA) is typically unremarkable, despite the suspected vascular pathophysiology. It may, however, help to differentiate AMNR from conditions that usually show abnormal FA findings, such as multiple evanescent white dot syndrome, acute posterior multifocal placoid pigment epitheliopathy, and choroidal neovascularization. Optical coherence tomography angiography has been shown to visualize capillary perfusion defects of the retina, though remains an evolving modality. Multifocal electroretinography may show localised areas of depressed waveform tracings, indicative of photoreceptor dysfunction.

In conclusion, AMNR is a rare primarily outer retinal disorder that can mimic the bitemporal visual field cuts seen in optic chiasm disease. AMNR is best detected by macular OCT and infrared photography as DFE may appear entirely normal.

References