

# **What You Should Know About Acute Macular Neuroretinopathy**

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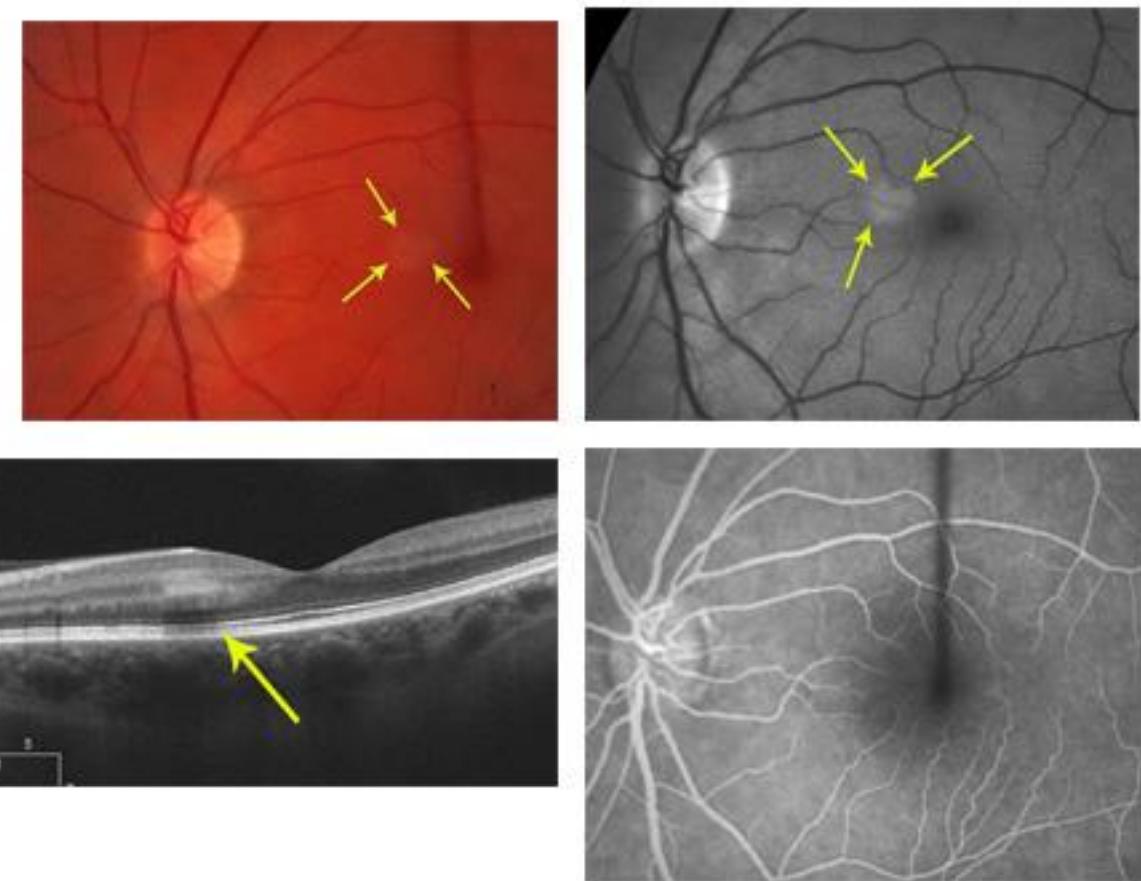
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Acute macular neuroretinopathy is a condition characterized by the sudden, painless onset of paracentral blind spots or blur spots in one or both eyes. Patients frequently give a history of an antecedent flu-like illness or exposure to intravenous epinephrine or ephedrine. Occasionally, there may be a history of exposure to intravenous contrast agents, taking birth control pills, or excessive coffee drinking. Rare cases have been associated with blood loss, hypotension, previous trauma, or chiropractic manipulation. Women are affected six times as often as men. The median age of the patients who have been reported is 26, with a range of 12 to 64. In 54% of cases, both eyes are involved. Eighty per cent of patients have better than 20/40 vision, but all have bothersome scotomas adjacent to fixation.<sup>1,2</sup>

On examination, in the early stages, tear drop-shaped, reddish brown or slightly gray areas are seen in the macula pointing toward the fovea. These are best imaged with near infrared reflectance photography. This modality is mediated with light having a longer wavelength than conventional photography

which penetrates the retina better and reveals the lesions located in the outer retina. Fluorescein angiography may show faint hyperfluorescence in the early frames of the study, but is not impressive. Electrooculography and global electroretinography are normal, but the multifocal electroretinogram indicates that there is an outer retinal abnormality in the area of the scotomas. This shows up as reduced voltage responses in these areas. Optical coherence tomography shows hyperreflectivity in the outer retina sometimes with disruption of the ellipsoid zone. Later in the course of the condition, there will be thinning of the outer nuclear layer. Optical coherence tomography angiography shows defective flow in the choriocapillaris, which could represent a shadow artifact from overlying hyperreflectivity in the outer retina. An example is shown in the figure.

## Figure



**Legend:** Fundus images from a 42 year old female who complained of decreased vision in the left eye with a grayish paracentral scotoma for 5 days. She was taking gabapentin for chronic back pain and an oral contraceptive. Her visual acuity was 20/25 in the left eye. She had no afferent papillary defect. Her confrontation fields were full. The slit lamp examination was normal. The fundus exam on the right was normal. On the left she had a nasal parafoveal grayish patch (top left panel). The lesion is better shown with near infrared reflectance imaging (top

right panel). The OCT (bottom left panel) shows hyperreflectivity in the inner nuclear layer in the same area (yellow arrow). The fluorescein angiogram (bottom right panel) showed no leakage in the involved area. She was asked to discontinue all the pain medications and return in 6 weeks at which time the lesion had resolved and her visual acuity had returned to 20/20, but her scotoma was unchanged.

The course of the disease features gradual improvement in most cases, although some residual scotomas are often present. It is important to rule out optic nerve disease in these.

The best synthesis we can make for the cause of the disease is ischemia of either the deep capillary plexus or the choriocapillaris.<sup>3</sup> Many of these patients have symptoms and signs of autoimmune disturbance suggesting that this is an autoimmune disease perhaps triggered by previous infection.

## **Reference List**

1. Bhavsar KV, Lin S, Rahimy E, et al. Acute macular neuroretinopathy: a comprehensive review of the literature. *Survey of Ophthalmology* 2017; <http://dx.doi.org/10.1016/j.survophthal.2016.03.003>.
2. Pellegrini F, Interlandi E, Pavesio C, Ferreyra HA. We cannot see what she cannot ignore. *Survey of Ophthalmology* 2017.
3. Sarraf D, Rahimy E, Fawzi AA, et al. Paracentral acute middle maculopathy. A new variant of acute macular neuroretinopathy associated with retinal capillary ischemia. *JAMA Ophthalmol* 2013;131:1275-87.