# HELLP, I Can't See!
# 31-Year-Old Female with Blurry Vision OD

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## HPI:

A 31-year-old female presents with three weeks of blurry vision in her right eye. She reports her vision changes started one day after the delivery of her child. Her pregnancy was complicated by preeclampsia and HELLP (Hemolysis, Elevated Liver enzymes, Low Platelet) syndrome for which she received anti-hypertensive medications, systemic steroids and expedited delivery via C-section. She has no other ocular or medical issues and currently takes no medications. On exam her visual acuity was 20/20 in both eyes with normal intraocular pressures and pupillary exam. Her anterior segment examination was within normal limits. No vitreous cell or haze was noted. Posterior segment examination was notable for 2 hyperpigmented lesions of the parafoveal area in the right eye (Figure 1). No peripheral retinal pathology was noted. The left eye fundus was largely unremarkable. OCT through the lesions revealed a focal disruption at the level of the ellipsoid zone (Figure 2). An infrared image highlighted the extent of the lesions with sparing of the fovea (Figure 3). Autofluorescence showed mild hypoautofluoresence in the areas seen on examination (Figure 4). OCTA showed a questionable focal flow void at the level of the deep capillary plexus. Based on this constellation of findings, a diagnosis of acute macular neuroretinopathy (AMN) from preeclampsia/HELLP syndrome was our leading diagnosis. The patient was counseled on the non-progressive nature of these lesions with the possibility of a retained paracentral scotoma for the future. No treatment was initiated, and the patient was told to return in 4 weeks for repeat evaluation.

## Discussion:

Acute macular neuroretinopathy is a rare disease process with a characteristic phenotypical appearance and symptom profile.[1-4,8] Often, patients will report the appearance of sudden-onset, well delineated paracentral scotomas. Macular exam reveals reddish-brown, wedge shaped lesions of which the apices are often

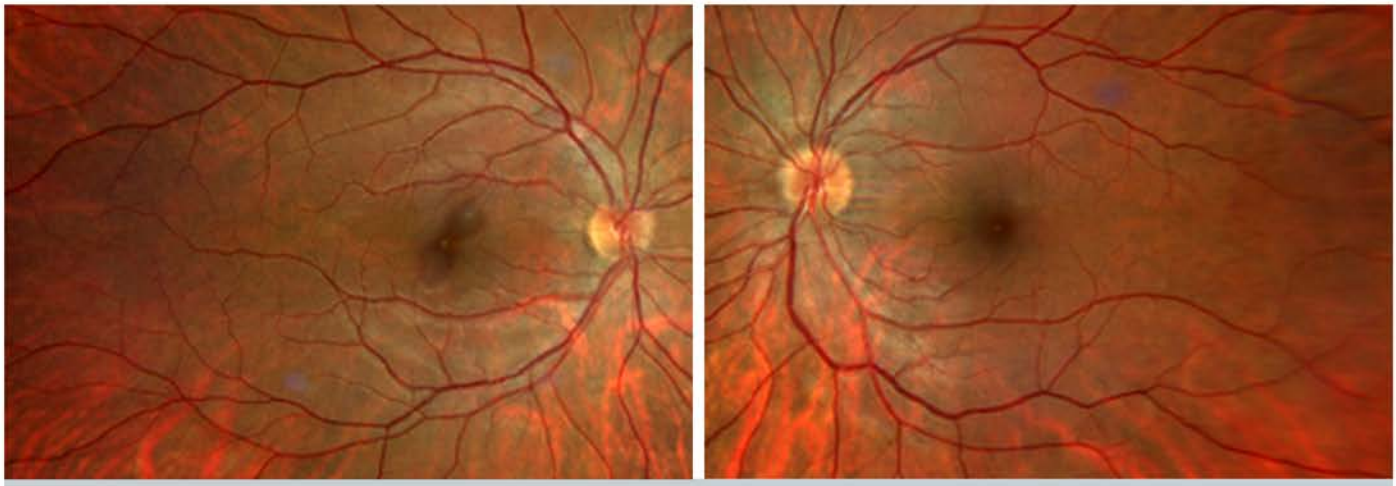

*Figure 1: Color fundus photos upon presentation. The right eye is significant for two red/brown petaloid parafoveal lesions. The left eye posterior pole was unremarkable.*

pointed towards to fovea. Patients may be able to draw these lesions exactly as seen on examination using an Amsler grid. Risk factors to developing AMN include[5]:

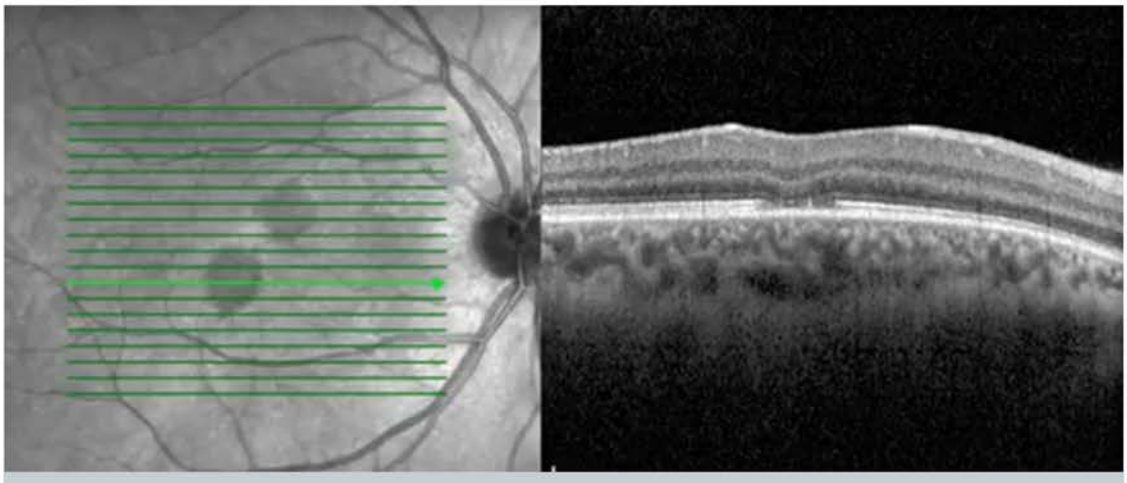

*Figure 2: Optical coherence tomography (OCT) of the right eye through the inferior lesions demonstrating focal disruption of the ellipsoid zone.*

- Recent upper respiratory infection
- Use of oral contraceptive medications
- Pre-eclampsia
- Caffeine
- Severe hypotension
- Hypercoagulable states

In a recent review of 101 cases of AMN, 55 (54.4%) cases were noted to be bilateral and scotomas were reported in the majority (72%) of cases.[6] Patients with AMN tend to be younger and more commonly female. The use of near infrared reflectance imaging especially highlights the borders of the lesions as shown in our case and can be used in counseling the patient to the extent of his or her scotoma. Optical Coherence Tomography (OCT) may demonstrate ellipsoid zone disruption, hyperreflectivity of the outer nuclear layer, or generalized outer retinal loss. In a minority of cases, these structural imaging changes may resolve over time. However, in an analysis by Fawzi et al. of eight cases with AMN, persistent structural abnormalities were seen OCT and correlated with functional defects for up to 14 months after presentation.[7] OCT angiography may also be of benefit to localize areas of vascularnon-perfusion as the pathophysiology of these lesions has been surmised to be secondary to insults to the retinal deep capillary

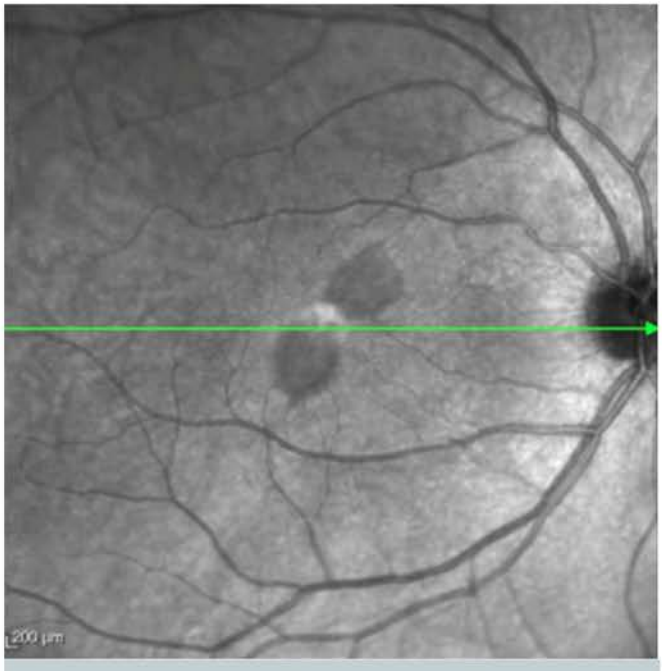

*Figure 3: Infrared image demonstrating two petaloid-shaped lesions sparing the fovea of the right eye.*

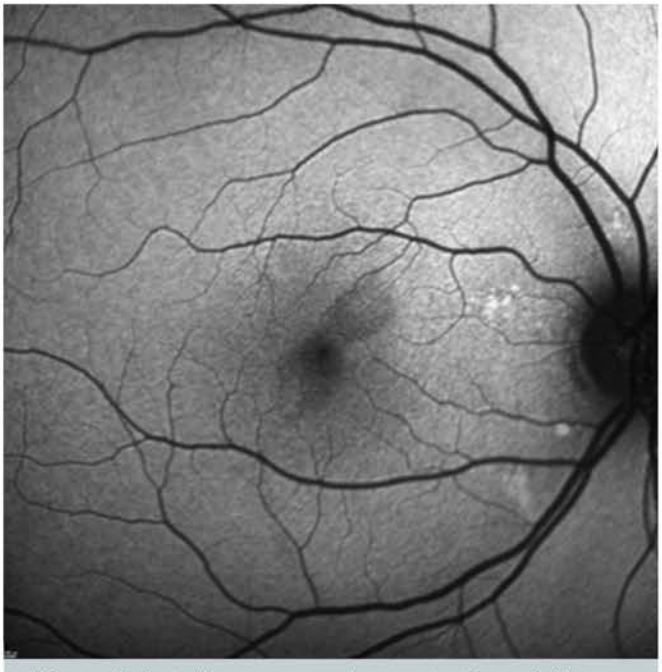

*Figure 4: Autofluorescence demonstrating mild hypo-autofluoresence in the area of the lesions seen on color fundus photography.*

plexus. Growing literature states that AMN may be a spectrum disease with its counterpart, Primary Acute Middle Maculopathy (PAMM) also thought to be caused by vascular insult to the intermediate and deep capillary plexus. No specific treatment regimen has been proven effective in hastening the resolution of these lesions. In the case of our patient, we believe this is the first report of HELLP syndrome with pre-eclampsia to be associated with AMR.

## References:


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