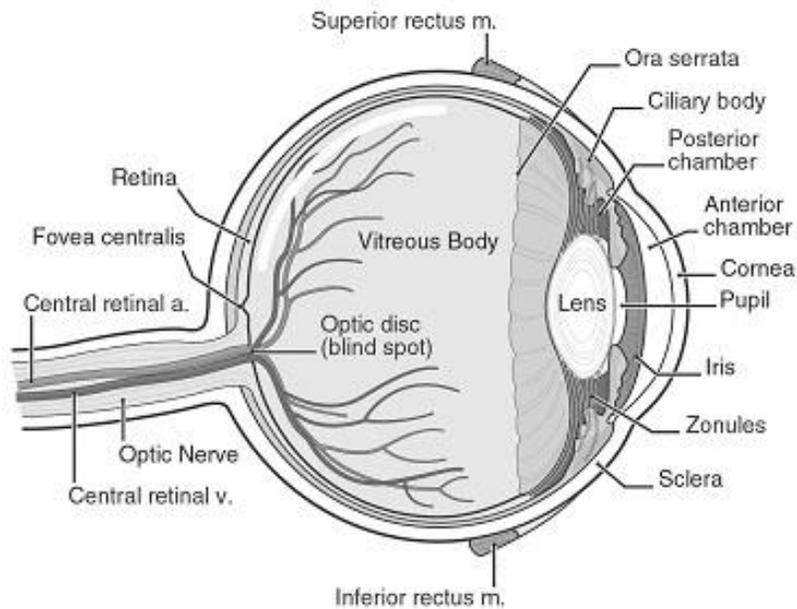


What You Should Know About Acute Zonal Occult Outer Retinopathy (AZOOR)

By David J. Browning, MD, PhD

The retina is a lining of nerve tissue on the back of the eye. It is comprised of photoreceptors which turn light into a nerve signal and other neural cells which transmit the signals from the eye to the brain via the optic nerve. Figure 1 identifies these parts of the eye.

Figure 1



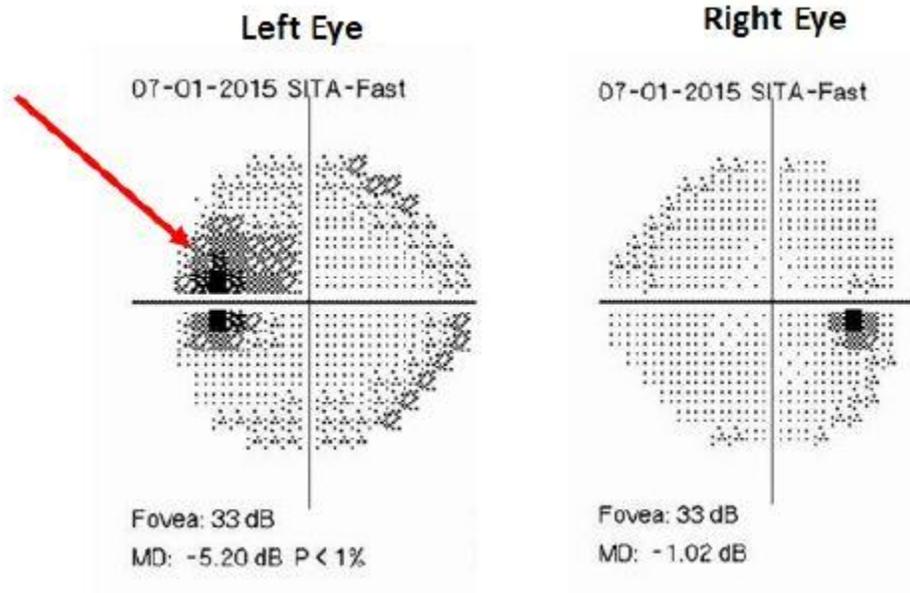
Acute zonal occult outer retinopathy (AZOOR) is a disease occurring in young to middle-aged people who have often had a preceding viral illness. Women are affected more than men in a 3:1 ratio. Patients are often myopic. Typically they lose retinal photoreceptors around the optic nerve which leads to an enlarged blind spot in the visual field. Patients often complain of shimmering lights. In two-thirds of cases one eye is affected initially. In these cases, approximately 60 per cent will go on to develop second-eye involvement.(1, 2)

Patients often describe an acute onset of a blind spot and flashing shimmering lights. When the ophthalmologist sees the patient the retina typically looks normal. Over time, pigmentary disturbances can develop around the optic nerve. Initially patients have good vision but eventually 10% of patients lose vision in affected eyes to the level of 20/200 or worse.

Ancillary testing can help to establish or exclude the diagnosis. The most common abnormality involves the electroretinogram (ERG), which measures voltages of the photoreceptors. (1,2) ERG amplitudes are usually reduced in affected eyes. Formal

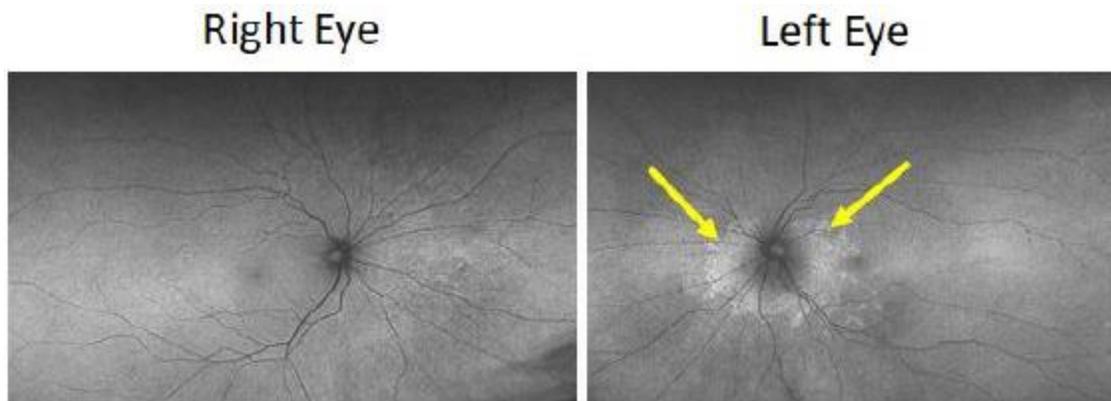
visual field testing usually shows an enlarged blind spot (figure 2). Abnormalities of fundus autofluorescence imaging (figure 3), near infrared reflectance imaging (figure 4A), and optical coherence tomography (figure 4B) are also typical. (2,3,4)

Figure 2 Enlarged blind spot in a case of AZOOR involving the left eye



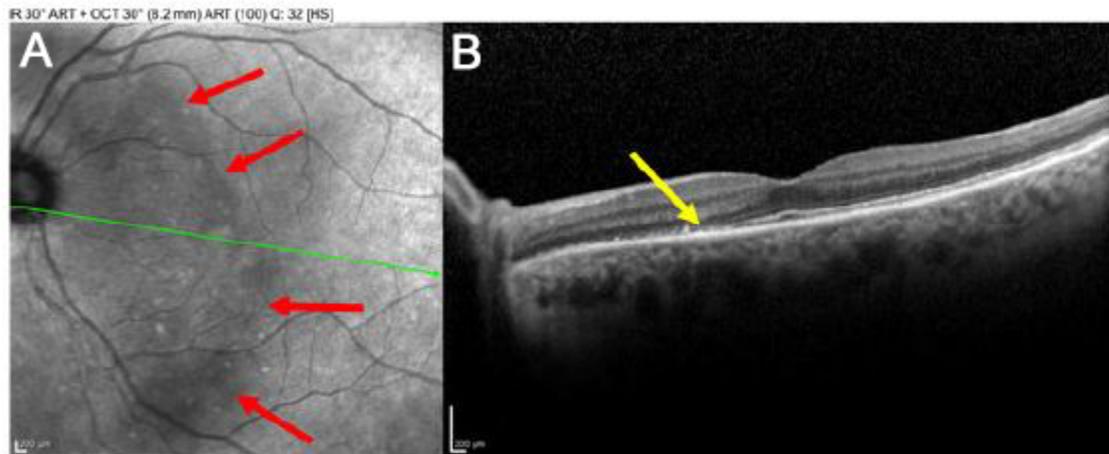
Legend: The visual field of the right eye is normal and should be used for comparison. The blind spot of the visual field in the left eye is enlarged (red arrow).

Figure 3. Abnormal fundus autofluorescence imaging in a case of AZOOR of the left eye



Legend: The autofluorescent image of the right eye is normal. Use this for comparison. The autofluorescent image of the left eye show hyperautofluorescence around the optic disc (yellow arrows).

Figure 4 Optical coherence tomography in AZOOR



Legend: The photoreceptors have been lost from the optic disc to a location two-thirds of the way to the center of the macula. This region is shown in the near infrared reflectance image on the left with the boundary indicated by the red arrows. The loss of photoreceptors is shown on the SD-OCT image on the right (the yellow arrow points to the missing ellipsoid zone line).

What causes AZOOR?

The cause of AZOOR is unknown. The two leading hypotheses are that it is a response to a viral infection or that it is an autoimmune reaction in which autoantibodies attack retinal cells leading to their demise. It is possible to have retinal autoantibodies as an epiphenomenon to another process. Their presence does not mean that they are the cause of the damage. Therefore, the significance of having retinal autoantibodies in AZOOR remains controversial.

What is the prognosis?

Recurrent attacks of disease can be seen in 15% of patients. Episodes can occur years after the initial attack. Three quarters of patients stabilize within 6 months of the initial episode, but one quarter will go on to have recurrences and further loss of visual field over years of follow-up. Recovery of visual function is rare.

Is there any treatment for AZOOR?

There is no known effective treatment for AZOOR. Various researchers have tried antibiotics, antiviral drugs, steroids, and immunomodulating drugs such as methotrexate to treat AZOOR. Nothing yet has shown convincing effectiveness.

Final Comments

AZOOOR is a rare disease of unknown cause and no known effective treatment. The best management plan is to exclude treatable infections, document the extent of the disease, and follow the patient carefully over time to avoid missing treatable unrelated conditions.

After you read this document, if you have further questions, please visit my website at www.retinareference.com. Another excellent resource for medical literature is Pubmed, on the National Library of Medicine website, accessible at www.pubmed.com.

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