A 58-Year-Old Man with Progressive, Monocular Outer Retinal Loss

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Introduction:

A 58-year-old white man initially presented in the fall of 2015 with "macular changes" noticed by his referring optometrist. The patient was asymptomatic at the time. He had a history of hypertension and non-Hodgkin's lymphoma, last treated in 2012 without evidence of recurrence. The vision was 20/25 in both eyes and neither fundus was seen as abnormal. The vitreous was clear in both eyes. Optical coherence tomography (OCT) showed vitreo-macular adhesion in the right eye with a macular posterior vitreous detachment in the left. The patient was told to follow-up as needed.

About 2 months later, the patient called with new onset blurry vision and photopsias in the right eye that started about 2 days prior. He denied any recent viral illness. On exam, he was still 20/25 in both eyes. The anterior segment and anterior vitreous were still clear. However, on examination of the posterior pole of the right eye, there appeared a light gray, deep subretinal/choroidal ring surrounding the optic nerve, with smaller rings around the edge of the larger ring (Figure 1a). The left eye still appeared normal (Figure 1b). OCT of the right eye showed significant loss of the outer retina (more nasal than temporal to the fovea), while the left eye remained normal (Figure 2a-b). Fluorescein angiography and fundus autofluorescence were both relatively unremarkable (Figure 3a-e). A limited laboratory workup was conducted (FTA, RPR, HIV, CBC), all of which was normal.

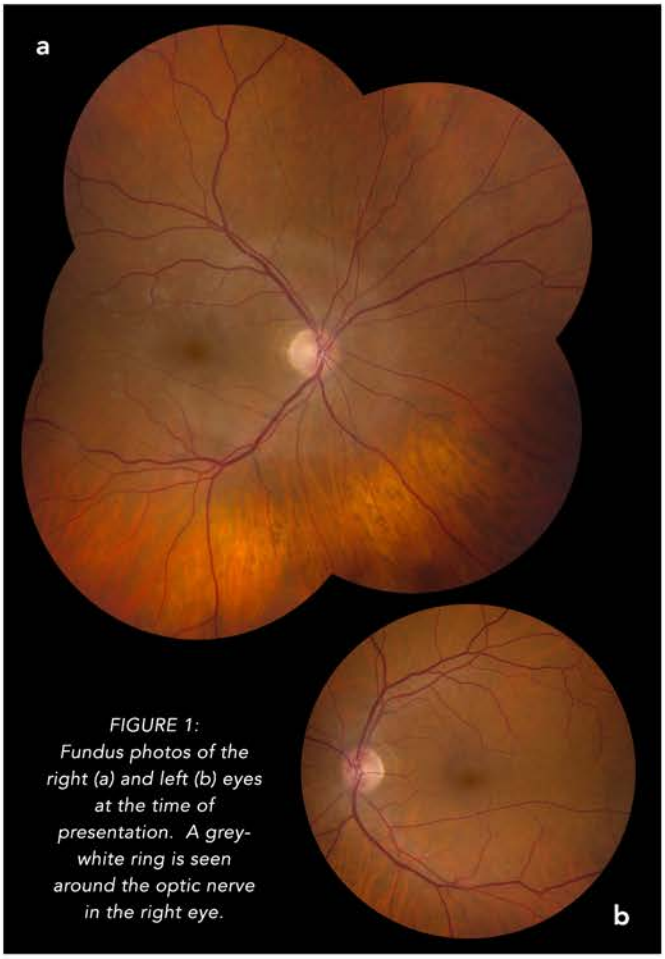

FIGURE 1: Fundus photos of the right (a) and left (b) eyes at the time of presentation. A grey-white ring is seen around the optic nerve in the right eye.

The patient returned 2 weeks later. He had noted slow progression of symptoms. However, his visual acuity remained at 20/25 in both eyes. The OCT showed continued outer retinal loss (Figure 4a-b). Multifocal electroretinogram showed significantly reduced signal in

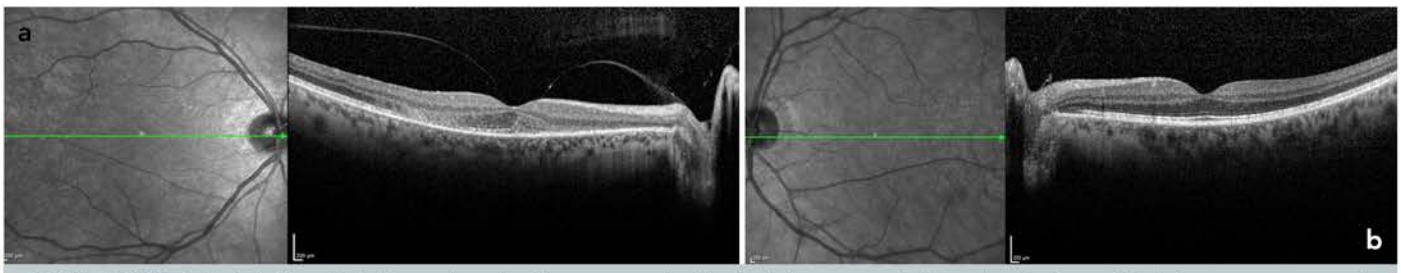

FIGURE 2: OCT of the right (a) and left (b) eye show significant outer retinal loss in right eye, especially nasal to the fovea. The left eye appears normal.

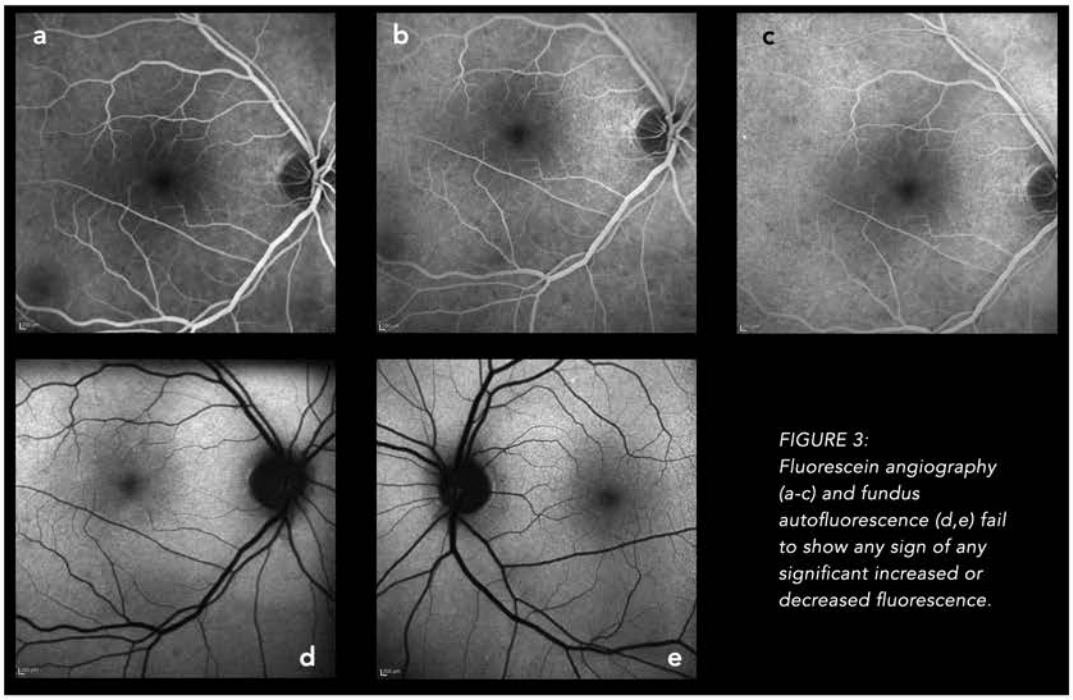

FIGURE 3: Fluorescein angiography (a-c) and fundus autofluorescence (d,e) fail to show any sign of any significant increased or decreased fluorescence.

and photopsias, with minimal, if any, fundoscopic changes[1]. Only later did patients show significant loss of the outer retina and retinal pigment epithelium (RPE). Our patient presented with a rare variant of AZOOR called acute annular outer retinopathy (AAOR), which was first described in 1994. It is similar to AZOOR in imaging, exam and follow-up, but presents with a large, thin, gray-white ring which disappears over several weeks[2].

the right eye throughout the macula, while normal in the left (Figure 5). When the patient came back 2 months later, his vision declined to 20/30 and an afferent pupillary defect was now present. The right optic nerve now had some mild pallor and the retinal arterioles were noticeably attenuated (Figure 6a-b). The grayish white ring had disappeared. OCT showed further loss of the outer retina, with only the fovea with an intact ellipsoid band (Figure 7a-b). Repeat fluorescein angiography demonstrated a window defect in a ring around the optic nerve, only visible on widefield imaging (Figure 8a, b).

Discussion:

Acute zonal occult outer retinopathy (AZOOR) was first described by Dr. Gass in 1993. He described 13 patients, mostly young women, who presented with rapid loss of one or more zones of outer retinal function

By far, the most consistent symptom with AZOOR is photopsias, especially flashes that appear to have movement. Unlike symptoms suggestive of a posterior vitreous separation, the photopsias are often more noticeable in light settings. The photopsias can precede, appear concurrently, or succeed visual field loss. The field loss is highly variable, sometimes large and other times small, often having a connection with the blind spot. Progression of the field loss can occur for several weeks to months after initial presentation. Although patients report persistent visual field defects, some patients have been shown to have improvement. The disease affects one eye about 60% of the time, with an occasional delayed presentation in the other eye, sometimes as long as 2 years later[3].

At the time of presentation of AZOOR the fundus exam shows minimal, if any, signs of disease. There are reports describing variable degrees of vitritis,

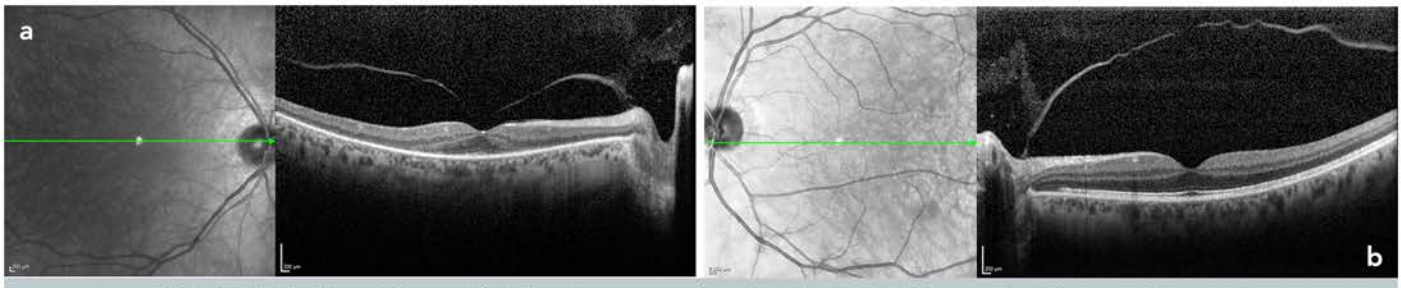

FIGURE 4: OCT of the right (a) and left (b) eye show progressive outer retinal loss in right eye. The left eye remains normal.

perivascular exudation and disc edema, all of which develop several weeks after onset of symptoms and are thought to be an inflammatory response to the dead retinal cells[4]. Weeks to months later, patients can develop retinal vascular narrowing and retinal pigment migration in the abnormal areas[3].

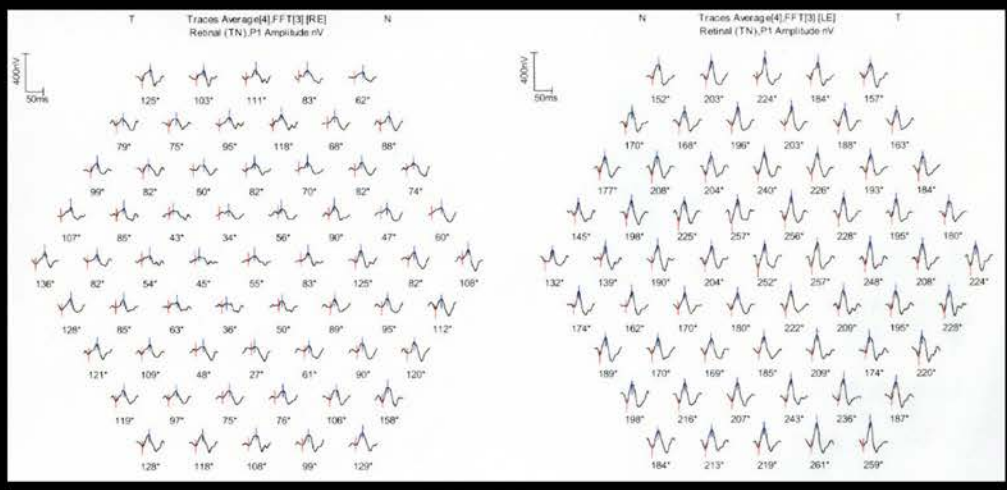

FIGURE 5: Multifocal electroretinogram of the right eye shows a reduced signal throughout the entire macula, while normal in the left eye.

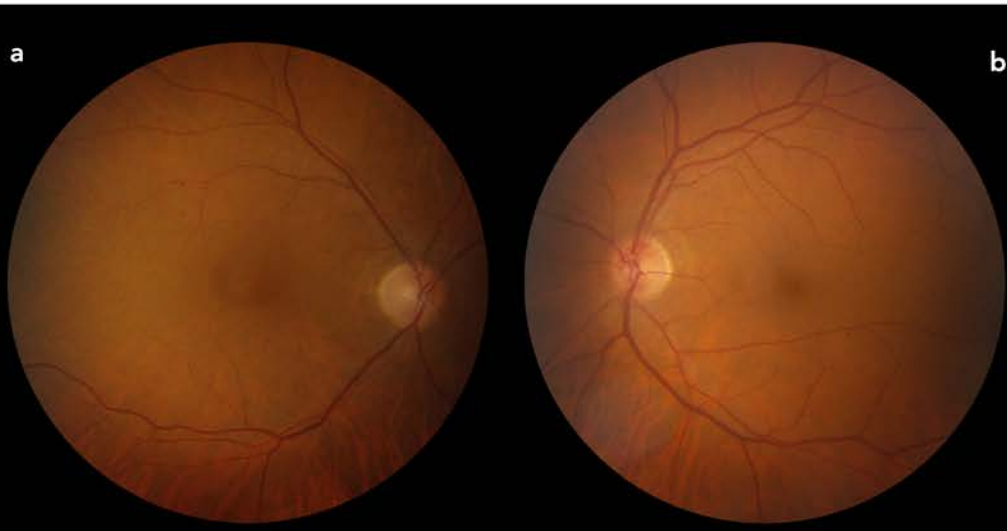

FIGURE 6: Several months after presentation, the right eye (a) now shows vascular attenuation and mild disc pallor, while the left eye (b) remains normal.

With regards to fluorescein angiography, initial testing can be normal. Only later, as patients develop significant outer retinal and RPE loss, will patients develop hyperfluorescence from transmission defects. Autofluorescence often shows hypoautofluorescence in the areas of RPE dysfunction and occasionally a ring of hyperautofluorescence in the rim of the defect[5].

Although anatomic changes can be subtle or absent early, electrophysiology is very useful in making a diagnosis in the early phases of the disease. Full field electroretinograms (ERG) show significant photoreceptor dysfunction in the affected eye as compared to the normal eye. Multifocal ERG is also helpful in documenting the extent and localization of the abnormality[6].

In terms of etiology, no one has defiantly proven the cause of the disease. Gass initially attributed it to an inflammatory response to a viral agent within the photoreceptors[1]. It is currently thought to be a combination

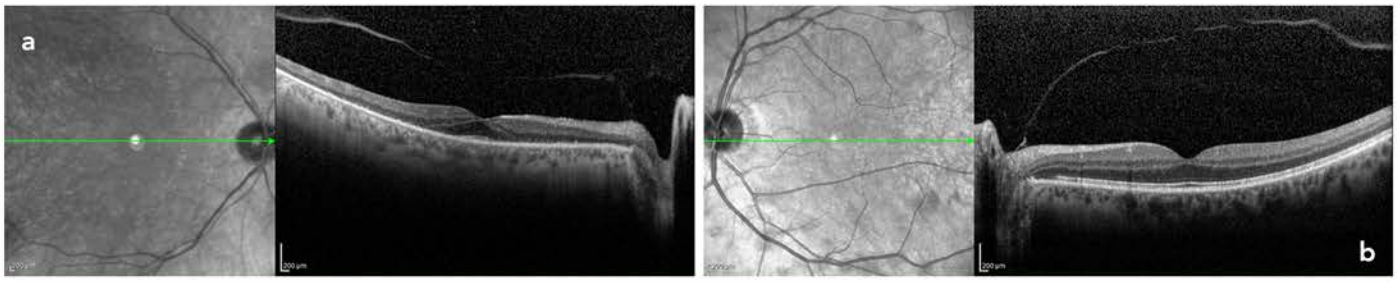

FIGURE 7: OCT of the right (a) and left (b) eye show progressive outer retinal loss in right eye, now with only an intact fovea. The left eye remains normal.

of genetics, autoimmunity and possibly environmentally triggered[3]. Steroids and antivirals have given to patients in the hope of stopping or reversing the disease. In general, the consensus is that these treatments are of limited value. There is a paucity of reports describing visual improvement with pulsed steroids[7].

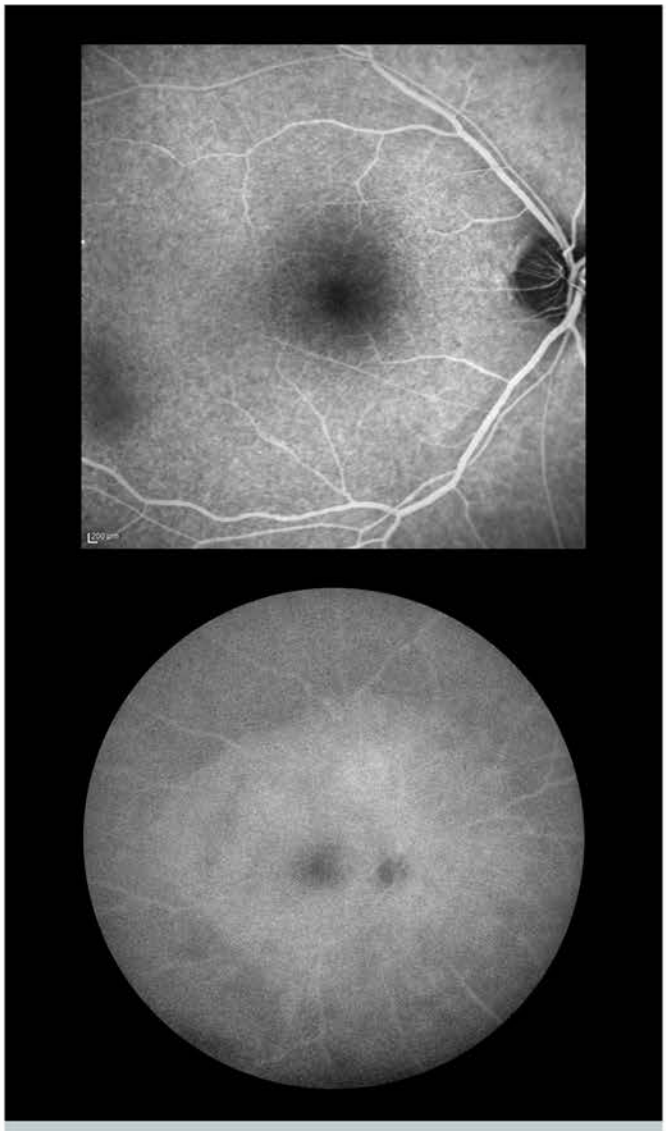

FIGURE 8: Repeat angiography of the right eye 2 and half months after presentation shows increased fluorescence in the area of dysfunction centrally, best seen on widefield imaging.

References:

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