Case Report: An Atypical Presentation of Acute Zonal Occult Outer Retinopathy (AZOOR)

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Abstract

Acute Zonal Occult Outer Retinopathy (AZOOR) is a rare inflammatory condition that is considered to be part of the white dot disease spectrum and may be due to viral or autoimmune causes. Signs and symptoms can include scotomas, persistent photopsias, and potential vision loss. The condition typically presents unilaterally in young Caucasian females with fundus signs being minimal or absent, leading to a delayed or missed diagnosis. This report details an atypical presentation of AZOOR with bilateral appearance with focal retinal deposits in a middle-aged Black female. A comprehensive review of the condition is presented, including pathophysiology, treatment, and multimodal imaging results.

KEY WORDS:
acute zonal occult outer retinopathy; trizonal, inflammation

INTRODUCTION

Acute zonal occult outer retinopathy (AZOOR), an infrequently encountered inflammatory disorder, was originally defined by Gass in 13 patients who presented with sudden-onset photopsias and scotomas. He assumed the condition was due to damaged outer retinal function.[1] The condition classically presents unilaterally in young females with fundus signs being minimal or absent, leading to a delayed or missed diagnosis.[2-4]While visual acuity can be relatively preserved, most cases will eventually show retinal and choroidal atrophy.[5-7] Initially, this condition was grouped together with other conditions under the white dot spectrum, since they all presented in a similar manner and had no known etiology; the so-called acute zonal occult outer retinopathy complex.[4,6] With the introduction of multimodal imaging, such as spectral domain optical coherence tomography and fundus auto fluorescence, the diagnosis and classification have become clearer.[6] Unfortunately, the etiology of the condition is still currently unknown, with theories pointing to a possible viral or autoimmune cause.[8] As the etiology is still elusive, various treatment options have been attempted with limited success. This case report details a rare and atypical presentation of chronic and bilateral AZOOR with focal retinal deposits in a middle-aged Black female that was diagnosed with the aid of multimodal imaging.

CASE REPORT

A 60-year-old Black female presented for a routine eye exam. Her medical history was positive for sleep apnea, hyperthyroidism, high cholesterol, and hypertension. Her current medications included atorvastatin, hydralazine, methimazole and carvedilol. Her ocular history included chronic dry eyes and photopsias in both eyes, specifically shimmering red lights that had started two years earlier but then subsided after a year. There was no viral illness associated with the onset of the photopsias. An examination by an outside eye care physician, at the time of the initial onset, had found no obvious ocular pathology to account for her photopsias (Figure 1).

Distance visual acuity, best-corrected, was 20/20 in both the right and left eyes. Entrance testing, including extraocular muscle movement, visual fields, and pupil testing, was unremarkable. Mild proptosis and eyelid retraction were noted in both eyes, which corresponded with the patient's history of thyroid-

associated orbitopathy (TAO). Slit lamp examination revealed capped meibomian glands and trace nuclear sclerotic cataracts in both eyes. Intraocular pressures were 14 mmHg in both the right and left eyes by Goldmann tonometry. Blood pressure (right arm, sitting) was 125/66mmHg. Cup-to-disc ratios were 0.25 round in both the right and left eyes. Focal yellow deposits were noted in the superior and inferior arcades as well as nasal to the optic nerve in each eye. A white gray demarcation line separated the normal retina from the affected retina in each eye. (Figure 1). Fundus autofluorescence (FAF) (California; Optos Inc, Marlborough, MA) revealed circumpapillary hyper-auto fluorescence with extension into the superior and inferior arcades along with hypo autofluorescence inferior to the nerves in both eyes, within the area of hyper autofluorescence. A demarcation line separated the areas of hyper auto fluorescence from the normal retina (Figure 2). Visual field testing showed superior temporal defects in both the right and left eyes extending from the blind spot that corresponded to areas noted on FAF (Figure 3). Juxtapapillary scan with spectral domain optical coherence tomography (Cirrus; Zeiss Inc, Dublin, CA) showed loss of the photoreceptor integrity line in both eyes, which corresponded to the hyper autofluorescence areas on fundus auto fluorescence (Figure 4). Spectral domain optical coherence tomography scanning also showed retinal pigment epithelium atrophy inferior to the nerve which corresponded to the hypo auto fluorescence noted on fundus auto fluorescence (Figure 5). The spectral domain optical coherence tomography scanning also revealed deposits or precipitates in the outer retina (Figure 5). Lab testing was ordered including rapid plasma reagin, anti-nuclear antibodies, fluorescent treponemal antibody absorption test, angiotensin converting enzyme levels, chest X-ray, serum lysozyme, and QuantiFERON Gold, and all test results returned normal. Based on the examination findings, ancillary testing, and the patient's previous history of photopsias, she was diagnosed with bilateral AZOOR. A retina consult was sought with a retina specialist who confirmed the diagnosis and recommended intra-vitreal steroid injections. The patient declined and continues to be followed semi-annually.

Figure 1: *Fundus photo of the right eye (Photo A) and left eye (Photo B) revealing focal yellow deposits in the superior and inferior arcades as well as nasal to the nerve. An area of atrophy is also noted inferior to the nerve in each eye. A demarcation line can be seen separating the normal retina from the affected retina in either eye.*

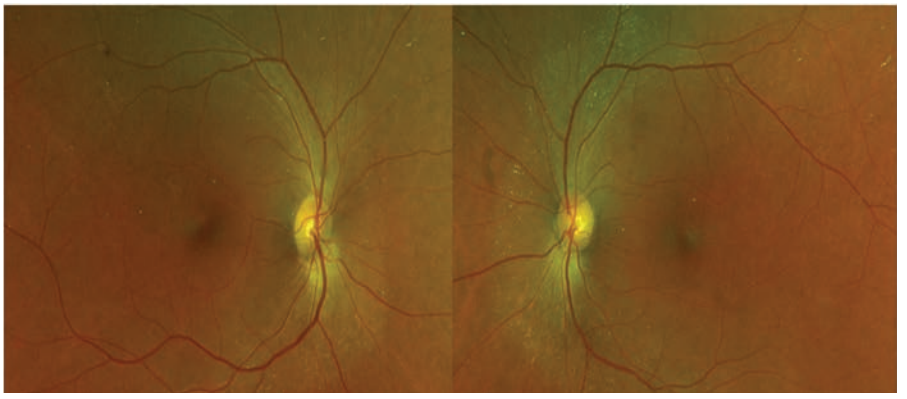


Figure 2: *Circumpapillary hyper-auto fluorescence with extension into the superior and inferior arcades along with hypo autofluorescence inferior to the nerve right eye (Photo A) and left eye (Photo B) within the area of hyper autofluorescence. A demarcation line separated the areas of hyper auto fluorescence from the normal retina in both eyes.*

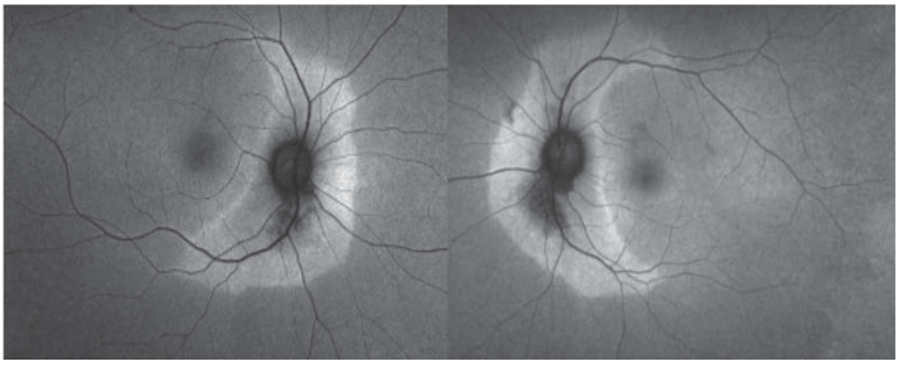


Figure 3: *Visual field testing showing superior temporal defects in the right and left eyes, extending from the blind spot that corresponded to areas noted on fundus auto fluorescence.*

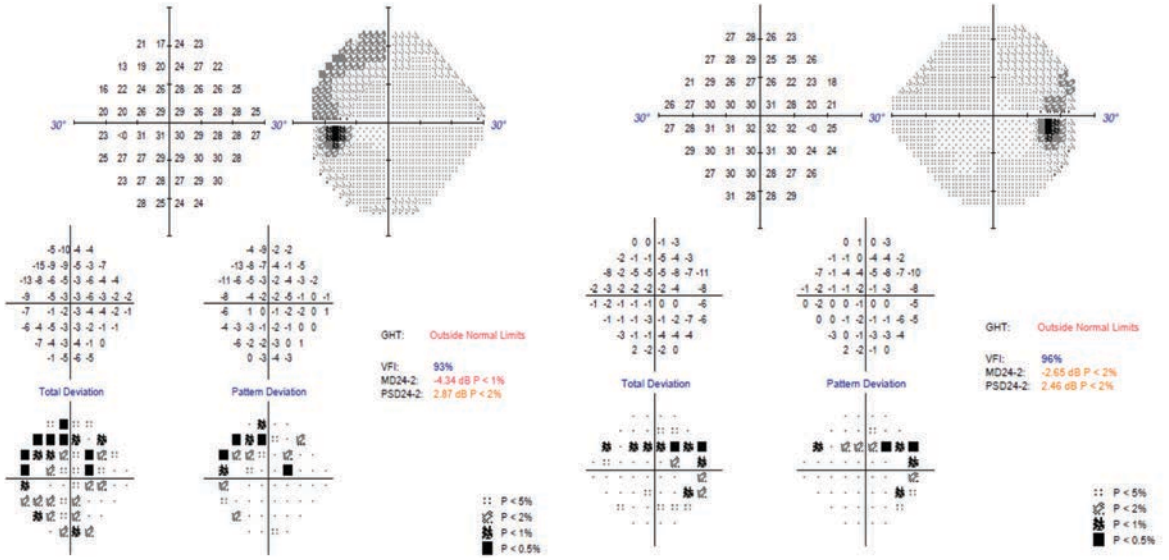


Figure 4: *Spectral domain optical coherence tomography of the right eye (above) and left eye (below) showing a trizonal pattern of normal retina followed by photoreceptor atrophy (blue squares) and retinal pigment epithelium atrophy (yellow squares).*

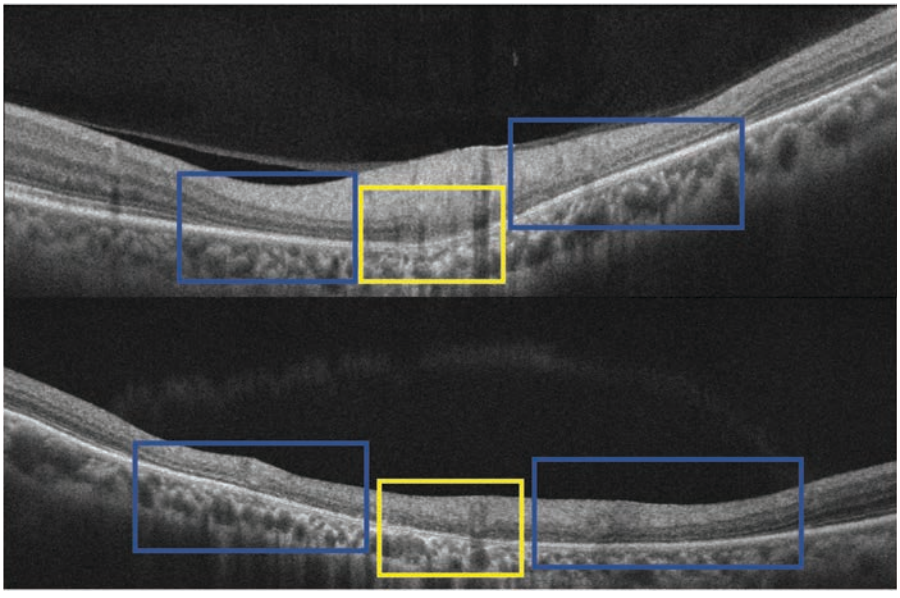
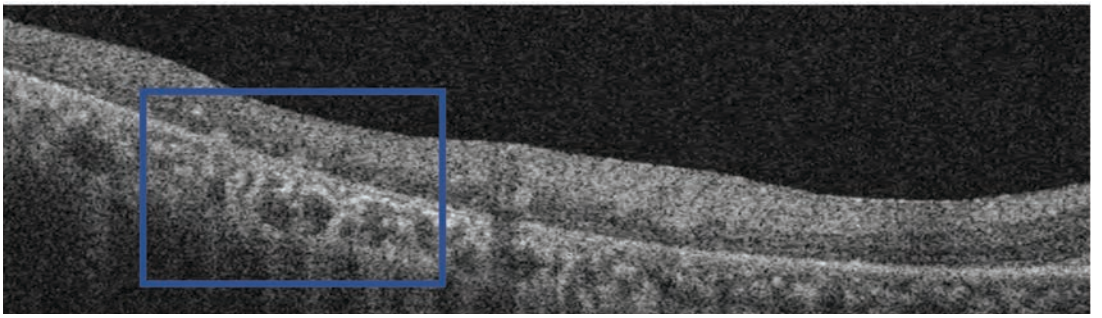


Figure 5: *Spectral domain optical coherence tomography scan of the right eye showing focal retinal deposits above the retinal pigment epithelium (blue square).*



DISCUSSION

Acute zonal occult outer retinopathy (AZOOR) is a condition of the outer retina that was initially noted by Gass in 13 patients; it includes features such as sudden loss of one or more areas of outer retina function, absent or very minimally noticeable posterior segment changes with late progression to noticeable atrophic changes in the retina, electroretinographic changes, and chronic visual defects.[1] Most of the subjects in his and future studies were noted to be young healthy females in their mid-thirties (76% of cases), with males being affected to a lesser extent.[9,10] Another study noted that the average age of onset was 47 years.[6] In addition, Caucasian females were affected more than other ethnicities. [4,10]

Although imaging technology has contributed to the diagnosis of AZOOR and has solidified the notion that damage to the photoreceptors leads to the known signs and symptoms, the exact etiology and pathogenesis are still unclear.[1,3,4,10] Two main hypotheses are suggested in the literature. Gass suggested that, since the condition resembled and presented similar (similar demographic features and localization to the outer retinal layers) to white dot diseases, an underlying viral process was responsible. This includes diseases such as multiple evanescent white dot syndrome, acute idiopathic blind spot syndrome, and multifocal choroiditis, which are considered to be differentials for acute zonal occult outer retinopathy.[1] The conditions are briefly summarized below.

- Multiple evanescent white dot syndrome typically presents in young females, with many having a preceding viral prodrome. Symptoms can include painless vision loss, photopsias, and scotomas.[11] Most cases are unilateral with retinal presentation showing white lesions at the level of the RPE. An enlarged blind spot can also be noted on visual field testing.[12] A distinct finding is foveal granularity, indicating RPE and photoreceptor involvement, which can be confirmed by optical coherence tomography.[12,13] These retinal findings and location are not seen in cases of acute zonal outer retinopathy.

- Acute idiopathic blind spot enlargement syndrome is another rare outer retinopathy and has been proposed to be a late variant of multiple evanescent white dot syndrome, albeit without the dots.[14] It typically presents with scotomas and photopsias in young women with the aforementioned blind spot enlargement noted on visual field testing.[15] The fundus presentation is usually unremarkable until later stages. Compared to acute zonal occult outer retinopathy, this condition is typically unilateral and lacks the trizonal pattern mentioned later in this paper.[6]

- Multifocal choroiditis is an inflammatory condition that presents with yellow gray lesions in both the retina and choroid and tends to present bilaterally.[16] The condition is truly inflammatory as active disease shows an anterior chamber reaction along with a vitritis.[16,17] Symptoms can also include decreased vision, floaters representative of the vitritis, and photopsia.[17] Other findings with multifocal choroiditis can include optic nerve head edema, cystoid macular edema, and choroidal neovascular membranes.[18] The condition tends to be chronic with multiple relapses and can lead to significant visual disability.[16,17,19]

Gass further postulated that the presumed virus entered the fundus through the peripapillary zone, which is the area of retina that is predominantly affected in cases of acute zonal occult outer retinopathy. In contrast, Jampol and Becker suggested that the observed inflammation was secondary to a genetic predisposition in combination with

an autoimmune disease and environmental factors. This could possibly be due to the greater number of affected females with the condition and therefore susceptibility to develop a possible autoimmune condition.[20] Other theories also include fungal infections,[21] polycythemia vera,[22] and anti-retinal antibodies.[23] Anti-retinal antibodies have received particular attention as one study suggested that antiretinal antibody leakage around the optic nerve resulted in AZOOR.[23] However, this study was countered by Forooghian, who argued that the simple presence of antiretinal antibody does not imply pathogenicity.[24] Further research is still required to elucidate the exact cause of AZOOR.

As mentioned earlier, symptoms of the condition can include photopsia and scotomas, and can also include photophobia and nyctalopia. Earlier reports on signs of AZOOR included no fundus abnormalities to mild fundus changes including retinal pigment epithelium atrophy or pigment clumping with possible arteriole attenuation.[3,4,9,10,25,26] However, with improved multimodal retinal imaging, a demarcating line that separates the un-involved and involved retina (AZOOR lesion) can also be seen.[2,11,27] Mrejen et al.[6] reported that this line was orange in color and its presentation was either continuous, interrupted, or scalloped. At times, the line can also be white/gray in appearance early in the disease course. These cases were originally classified as acute annular outer retinopathy, which is now considered to be an early form of AZOOR, as the line was visible ophthalmoscopically.[28,29] Mrejen et al. postulated that this represents an early form of acute zonal outer retinopathy as the line can diminish but can also turn into the previously mentioned orange line. They also proposed a trizonal pattern of chorioretinal degeneration for the AZOOR lesion based on fundus photography, fluorescein angiography, fundus auto-fluorescence, and spectral domain optical coherence tomography. The trizonal pattern represents normal retina, photoreceptor and retinal pigment epithelium degeneration, and chorioretinal atrophy. Specifically, zone 1 is normal retina outside the demarcation line; therefore, multimodal imaging testing is unremarkable here. Zone 2 presents a flecked hyper autofluorescence on fundus auto fluorescence, which corresponds to photoreceptor and retinal pigment epithelium disruption on spectral domain optical coherence tomography.[6] Although indocyanine green angiography was not performed on this patient, it likely would have shown extra choroidal leakage, which is the typical finding of AZOOR lesions in zone 2. Finally, zone 3 represents photoreceptor, retinal pigment epithelium, and choroidal atrophy and is easily visualized on spectral domain optical coherence tomography. This corresponds to fundus autofluorescence hypo-autofluorescence and hypo fluorescence on indocyanine green angiography.

Our patient had this typical trizonal pattern of abnormalities. Specifically, the patient had normal fundus auto fluorescence and spectral domain optical coherence tomography findings outside the demarcating line. Within the demarcation line, hyper autofluorescence was noted on fundus auto fluorescence which correlated with photoreceptor and retinal pigment epithelium disruption on spectral domain optical coherence tomography. Finally, the patient had areas of hypo-auto fluorescence inferior to the optic nerves in each eye, which correlated with photoreceptor and retinal pigment epithelium atrophy.

Due to the scarcity of the condition, possible improper classification of previous AZOOR cases due to a lack of multimodal imaging, and to the lack of long-term follow-up in the literature, it can be difficult to determine the prognosis.[1,4,5,6,10] The studies that have been published have shown that the prognosis for patients with AZOOR can be favorable depending on potential foveal involvement. According to one study on 205 eyes diagnosed with AZOOR, 74% of patients had acuities of 20/40 or better.[10] Another study revealed that 68% of patients had final visual acuities of 20/40 or better.[4] Stabilization or a burning out of the condition after 6 months has also been reported after the initial onset. Although the data show that many patients preserve visual function and therefore treatment is not essential, cases in which there is foveal involvement or progression to potential foveal involvement may justify treatment.[3,8,9,25,26] This, however, is complicated by the fact that the pathogenic process has not been determined and therefore no precise guidelines exist on how to manage and treat patients. Treatment efforts have included systemic steroids, intra-vitreal steroids, antibiotics, and antiviral agents, but with limited success. Recently, however, studies have shown that steroids may have a beneficial effect if given early in the disease course.[25,30] These studies have shown visual field recovery, in addition to improved or stable acuity, or improvement based on patient response and improved and enhanced multimodal testing results. The authors proposed that steroids may have worked in their series because treatment was initiated earlier due to improved diagnostic detection with previously absent multimodal imaging.[3] Clinicians should keep in mind the side effects associated with systemic steroid use such as increased intra-ocular pressure and cataract formation. Immunosuppressants have also been used as a treatment option, however they provide little efficacy.[31] A newer finding, although rare in terms of AZOOR presentation, has also been noted. Cases have documented the development of choroidal neovascular membranes which were subsequently treated with anti-Vegf agents or photo dynamic therapy.[6,9,32,33]

Our case of AZOOR is, in relation to previous studies, atypical regarding its age of presentation, bilateral involvement, patient ethnicity, and most uniquely, the presentation of outer retinal deposits. The patient age at presentation is later than those in most cases in the literature, which have ranged from 14 years to 86 years.[2,6] However, as alluded to earlier, most patients are in their mid-thirties to late-forties.[4,6,10,26]

Although the condition typically presents in a unilateral fashion, fellow eye involvement can be seen. One study showed that 61% of patients had unilateral presentation at the initial onset and diagnosis (51 patients for an average of 8 years); however, by the final examination, 76% had advanced to bilateral presentation. Most notably, advancement of the other eye was not seen until 50 months after the initial presentation.[4] This is likely what happened in our patient, as the damage seen on fundus auto fluorescence seems to be more significant in the left eye than in the right eye.

Most patients with AZOOR are Caucasians, however other races can be affected, but to a much lesser extent. A study by Gass and Agrawal included 30 patients, of which 91% were Caucasian, 7 % were Hispanic, and 2% were Asian.[4] Our patient was Black, and a review of the literature found that they are under-represented compared to other races. [1] It is currently unknown why the condition affects Caucasians more than other ethnicities.[10] In accordance with Jampol and Becker's theory that AZOOR may be the result of an interplay between autoimmunity and genetics, it is conceivable that certain ethnicities are predisposed to acquire AZOOR. Some ethnic groups are more likely to exhibit various autoimmune conditions; for example, lupus is more common in Blacks, while multiple sclerosis is more common in Caucasians.[34,35] Furthermore, our patient had a history of auto-immune disease with her thyroid-associated orbitopathy and hyperthyroidism, which may have put her at additional risk for developing AZOOR. One survey noted that most common auto-immune diseases linked to AZOOR were associated with the thyroid gland. This included seven patients with Hashimoto thyroiditis with one patient presenting with Grave disease.[10] However, due to the rarity of the condition, smaller study sample sizes, and the lack of case-controlled studies, causation cannot be inferred.[36] Further research is essential at this time to explain the exact cause(s) of AZOOR and possible links to genetics/autoimmune conditions and ethnicity. Lastly, the patient presented with diffuse and focal yellow deposits within the AZOOR lesion in each eye. To the best of our knowledge, this is the first case of AZOOR that has presented with retinal deposits. These deposits were hyper-reflective on spectral domain optical coherence tomography and were localized to the outer retina, specifically in areas with concurrent photoreceptor damage. It is possible that they represent degenerated photoreceptor outer segments that migrated and deposited in the outer retina secondary to chronic underlying inflammation. Similar deposits have been noted in conditions such as vitamin A retinopathy. Specifically in these cases, the deposits were localized as damaged photoreceptors segments and are thought to be due to a deficiency of vitamin A, which plays an important role in the formation of rhodopsin in the phototransduction pathway.[37,38]

This case report highlights an atypical presentation of AZOOR. Although rare, patients can present outside the norms reported in the literature. Clinicians should keep these findings in mind when thinking of a possible diagnosis for a patient who does not match the traditional definitions of AZOOR. Patients should be monitored on an appropriate basis, with multimodal imaging, to gauge for possible progression. Further research and study are vital to clarify the exact cause(s) of AZOOR. Once this has been established, possibly with advances in multimodal imaging, innovative treatment options may be developed for future use.

Recognizable health information was not used in this manuscript. ●

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