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# Azoor Acute Zonal Ocular Outer Retinitis: A Case Report and Literature Review

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**Abstract** **Original Research Article**

Acute zonal ocular outer retinopathy (AZOOR) is a rare disease, usually manifested by persistent photopsia, reduced visual acuity, asymmetric visual field loss. A 46 years old female patient presented to our department for investigation of a slowly progressive decline in visual acuity over several years with no other associated signs. Best corrected visual acuity was 1/10 in the right eye and 8/10 in the left eye; fundoscopic examination revealed an extensive, poorly demarcated peripapillary depigmentation halo with scattered pigment clots, the oct showed atrophy of the outer retina, including interruption of the ellipsoid zone and the outer nuclear layer in the affected areas, with foveal sparing. The patient was treated with steroidal anti-inflammatory drugs, but with no obvious response.

**Keywords**: AZOOR, Visual Field, Multimodal Imaging, Peripapillary Atrophy, Case Report.



## INTRODUCTION

Acute zonal ocular outer retinopathy (AZOOR) is a rare disease of as yet unknown etiology, usually manifested by persistent photopsia, reduced visual acuity, asymmetric visual field loss and often generalized photoreceptor dysfunction on electroretinogram (ERG). The disease affects young and middle-aged patients, most of whom are female, and is in some cases preceded by a viral syndrome. Most patients recover over a period of 1-3 years, but in some situations vision loss may be permanent.

## MATERIAL AND METHODS

This is a 46-year-old female patient with a history of arterial hypertension and diabetes, who presented to our department for investigation of a slowly progressive decline in visual acuity over several years with no other associated signs. Clinical examination revealed visual acuity of 1/10 in the right eye and 8/10 in the left; anterior segment examination was normal, except an incipient corticonuclear cataract in both eyes; fundoscopic examination revealed an extensive, poorly demarcated peripapillary depigmentation halo with scattered pigment clots. The fundus examination revealed no signs of diabetic or hypertensive retinopathy. We completed the study with an autofluorescence retinophotography, which revealed a hyperautofluorescent border encircling the area of peripapillary atrophy visible on the fundus (figure 1).

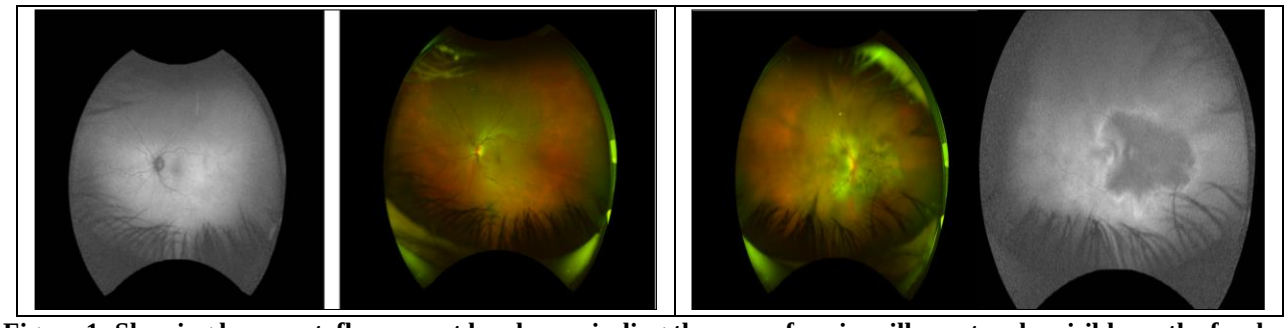

**Figure 1: Showing hyperautofluorescent border encircling the area of peripapillary atrophy visible on the fundus.**

An oct was performed, showing atrophy of the outer retina, including interruption of the ellipsoid zone and the outer nuclear layer in the affected areas, with foveal sparing in the right eye. The oct in the left eye was normal. (Figure 2)

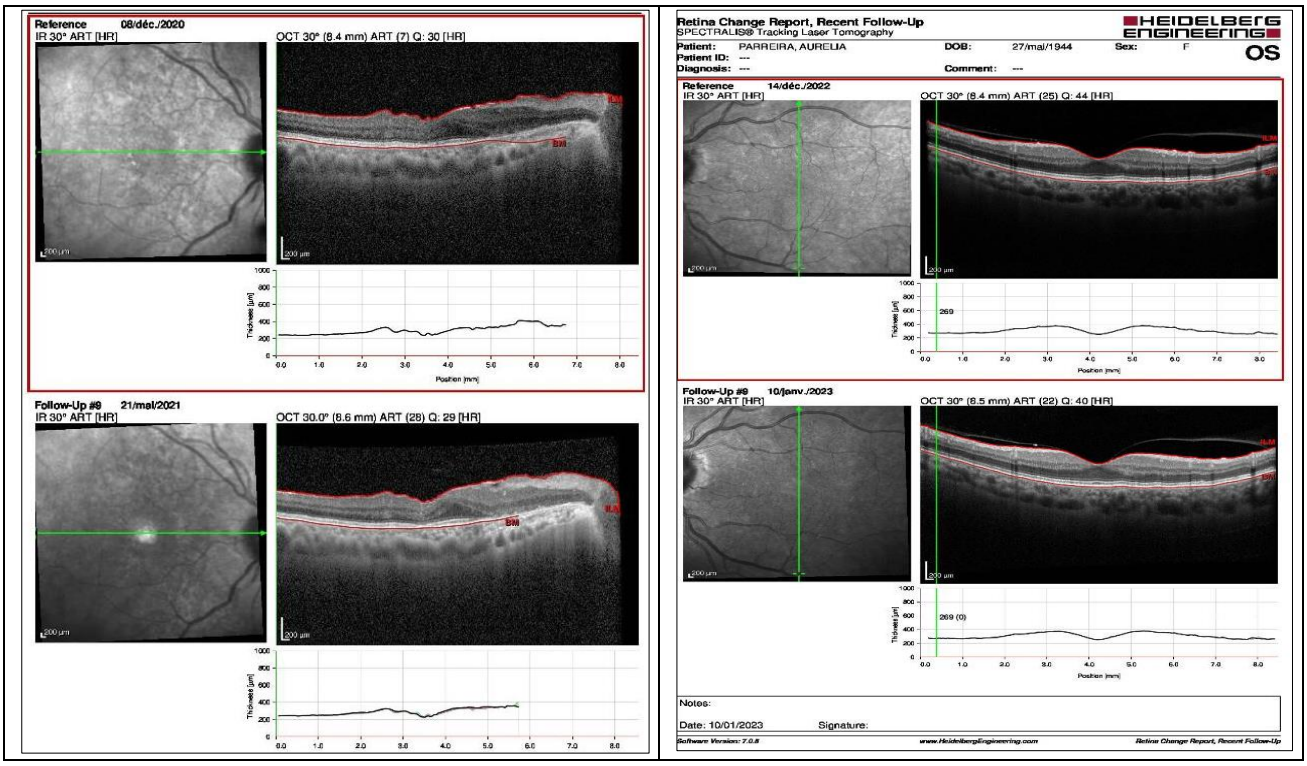


**Figure 2 : Showing atrophy of the outer retina, including interruption of the ellipsoid zone and the outer nuclear layer in the affected areas, with foveal sparing (right eye). The oct is normal (left eye).**

Indocyanine green angiography (Icg) shows peripapillary hypercyanescence without signs of frank activity, with small hypocyanescent areas in the lower periphery, and fluorescein angiography (AGF) reveals a peripapillary hyperfluorescent ring. (Figure 3)

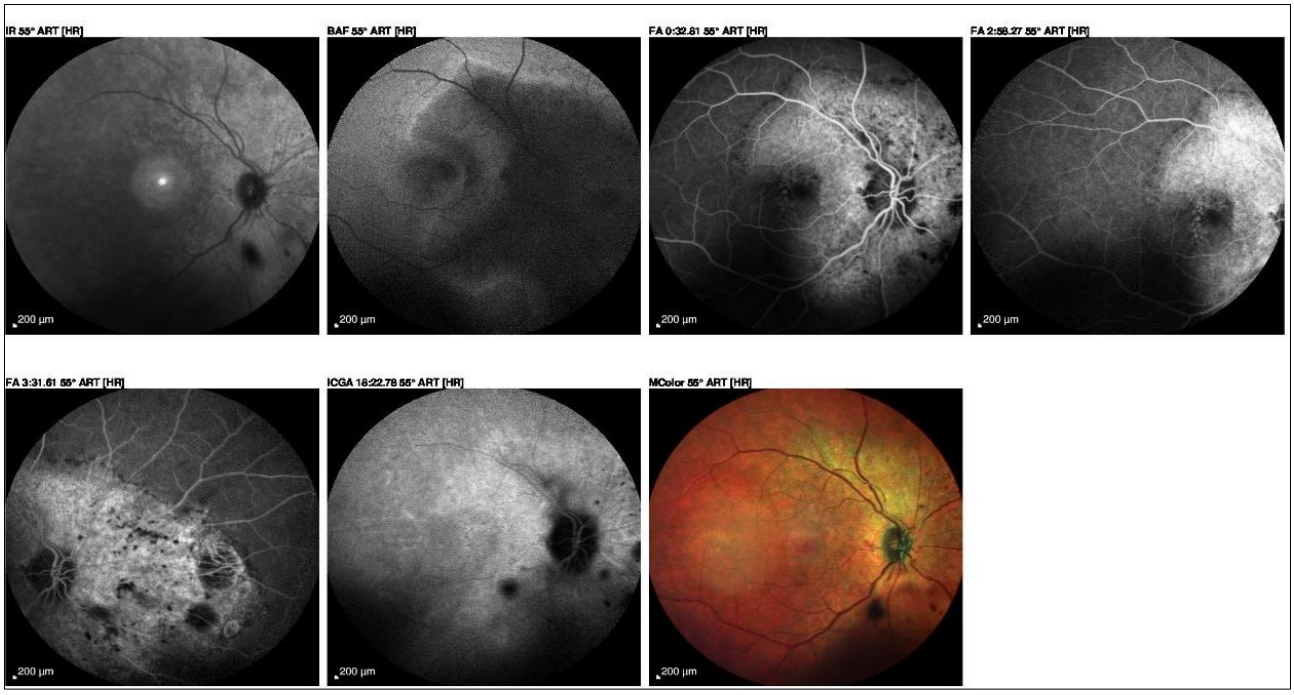


**Figure 3: Showing different aspects of AZOOR in infra red, autofluorescence, fluorescein angiography and ICG**

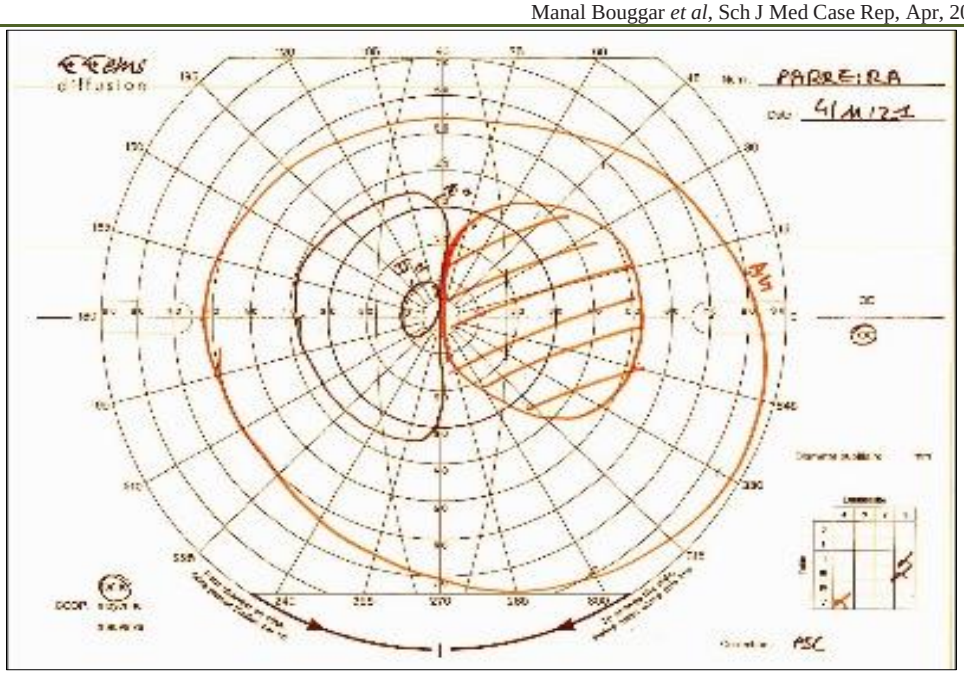


**Figure 4: Goldmann visual field with isopter narrowing and paracentral scotoma**

The Global ERG electroretinogram shows a normal Scotopic pattern and asymmetry due to peripapillary rod damage; in photopic condition asymmetry in relation to right-sided damage, with correct activity in left-sided areas (figure5). On the multifocal ERG, the peak was present in the right eye, but the amplitude was not optimal, with a reduction in overall sensitivity; the peak was perfect and overall sensitivity was preserved on the left side (figure 6).

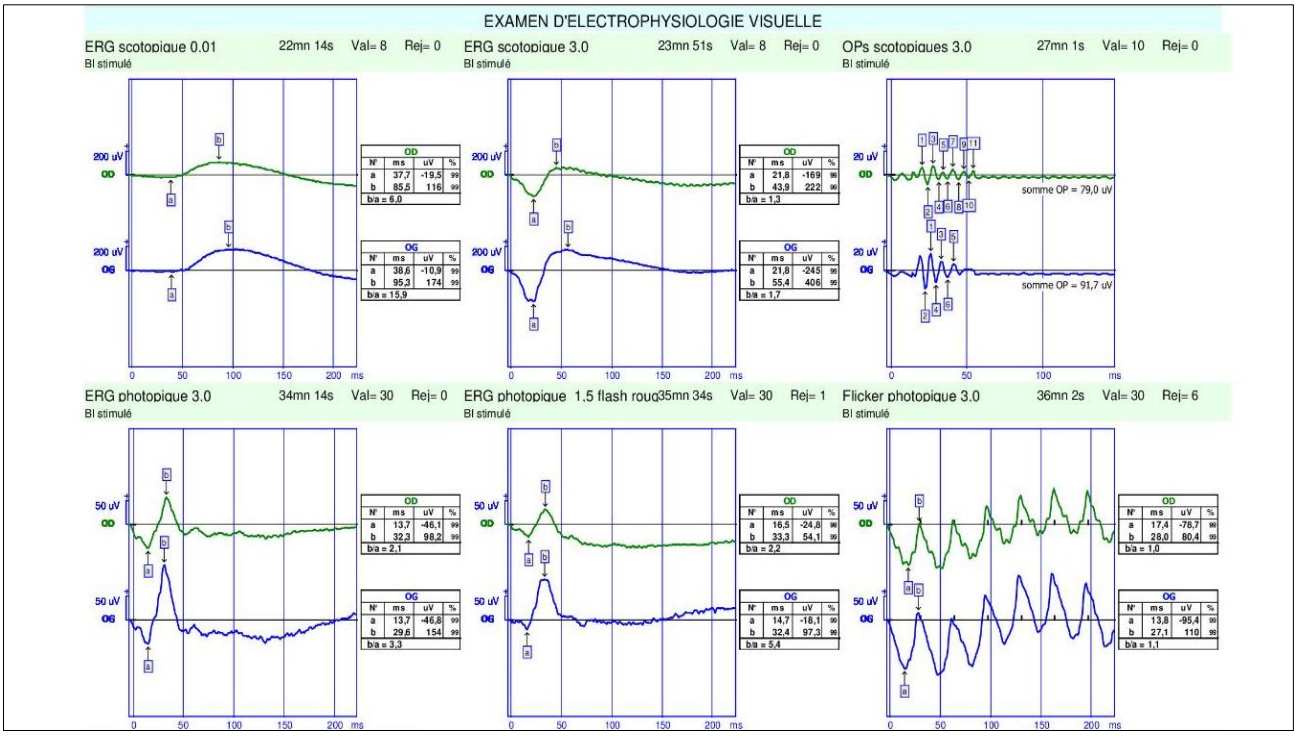


**Figure 5: Showing global ERG in scotopic and photopic condition (right eye in green, left eye in blue)**

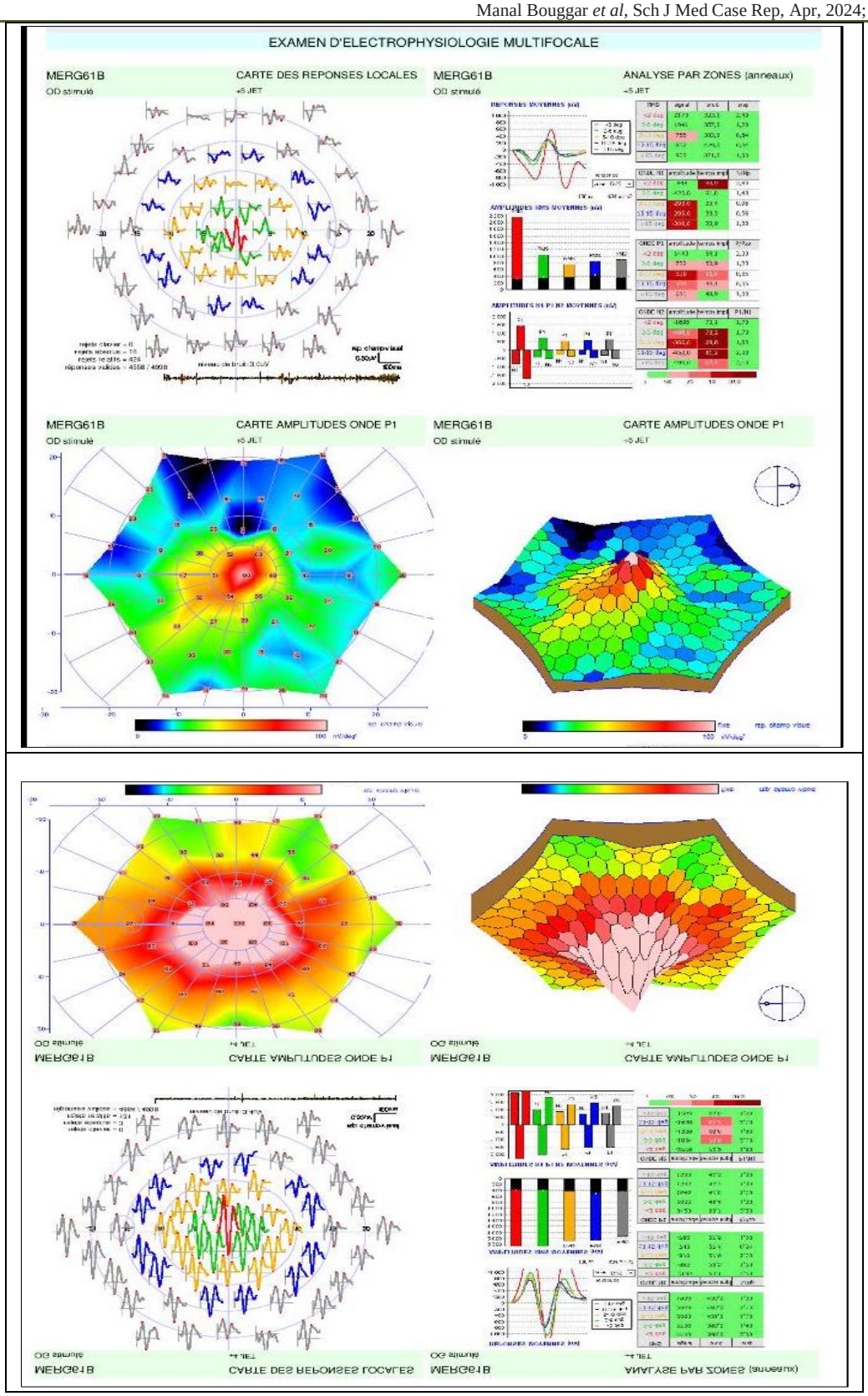


**Figure 6: Showing multifocal ERG in right then left eye**

The patient was treated with steroidal anti-inflammatory drugs, but with no obvious response. At present, our attitude consists of therapeutic abstention and monitoring only. The disease progressed slowly.

## DISCUSSION

Acute zonal occult outer retinopathy (AZOOR) is a condition of the outer retina that was initially described by Gass in 13 patients; it includes features such

as sudden loss of one or more areas of outer retinal function, absent or minimally perceptible posterior segment changes with late progression to perceptible atrophic retinal changes, electroretinographic changes and chronic visual defects [1].

There is a predilection for young, healthy women in their mid-thirties (76% of cases), with men affected to a lesser extent. Another study noted that the average age of onset was 47. In addition, Caucasian women were more affected than other ethnic groups [2]. The disease is often unilateral, but secondary bilateralization is not uncommon. Two main hypotheses are suggested in the literature: Gass suggested that, since the condition resembled and presented similarly to diseases of the white dot syndrome spectrum, an underlying viral process was responsible.

These include conditions such as multiple evanescent white dot syndrome and multifocal choroiditis, which are considered differential diagnoses for acute zonal occult external retinopathy. Other authors have suggested that the inflammation observed is secondary to a genetic predisposition in combination with autoimmune disease and environmental factors [3, 4]. Further research is still needed to elucidate the exact cause of AZOOR.

Symptoms of the disease may include photopsia and scotomas, as well as photophobia and nyctalopia. There may be no fundus abnormalities or only slight fundus changes, including atrophy of the retinal pigment epithelium or pigment agglutination with possible attenuation of the arterioles. However, thanks to improved multimodal retinal imaging, a line of demarcation separating the uninvolved retina from the involved retina can also be observed [5]. The trizonal model represents normal retina, photoreceptor and retinal pigment epithelium degeneration, and chorioretinal atrophy. More specifically, zone 1 is normal retina outside the line of demarcation; consequently, multimodal imaging tests here are normal. Zone 2 shows speckled hyperautofluorescence, consistent with disruption of photoreceptors and retinal pigment epithelium on spectral-domain optical coherence tomography [6]. Although ICG angiography was not performed in this patient, it would probably have shown additional choroidal leakage, which is the typical finding of AZOOR lesions in zone 2. Finally, zone 3 represents atrophy of the photoreceptors, retinal pigment epithelium and choroid, and is easily visualized on spectral-domain optical coherence tomography. It corresponds to hypoautofluorescence and hypofluorescence on ICG angiography [6].

Fluorescein angiography may show non-pathognomonic areas of pigment epithelial atrophy.

Examination of the Goldmann visual field reveals deficits of varying type and location, the most frequent being widening of the blind spot or central scotoma. During follow-up, areas of pigment epithelial atrophy or pigment migration may appear in the area of the deficient visual field, leading to a misdiagnosis of retinitis pigmentosa. Although the data show that many patients retain their visual function and that treatment is therefore not essential, cases of foveal damage or progression to potential foveal damage may warrant initiation of treatment.

Major therapeutics have included systemic steroids, intravitreal steroids, antibiotics and antiviral agents, but with limited success.However, recent studies have shown that steroids can have a beneficial effect if administered early in the course of the disease [7].

## CONCLUSION

AZOOR can have several presentations. Patients should be monitored appropriately, with multimodal imaging, to assess possible progression. Further research and studies are essential to establish the exact causes of AZOOR, so that innovative treatment options can be developed for future use.

## REFERENCES

1. Gass, J. D. M. (1993). Acute zonal occult outer retinopathy. Donders lecture: The Netherlands ophthalmological society, Maastricht, Holland, June 19, 1992. *Journal of Neuro-Ophthalmology*, *13*(2), 79-97.
2. Monson, D. M., & Smith, J. R. (2011). Acute zonal occult outer retinopathy. *Survey of ophthalmology*, *56*(1), 23-35.
3. Barnes, A. C., Lowder, C. Y., Bessette, A. P., Baynes, K., & Srivastava, S. K. (2018). Treatment of acute zonal occult outer retinopathy with intravitreal steroids. *Ophthalmic Surgery, Lasers and Imaging Retina*, *49*(7), 504-509.
4. Jampol, L. M., & Becker, K. G. (2003). White spot syndromes of the retina: a hypothesis based on the common genetic hypothesis of autoimmune/inflammatory disease. *American journal of ophthalmology*, *135*(3), 376-379.
5. Tavallali, A., & Yannuzzi, L. A. (2017). MEWDS, common cold of the retina. *Journal of Ophthalmic & Vision Research*, *12*(2), 132.
6. Mrejen, S., Khan, S., Gallego-Pinazo, R., Jampol, L. M., & Yannuzzi, L. A. (2014). Acute zonal occult outer retinopathy: a classification based on multimodal imaging. *JAMA ophthalmology*, *132*(9), 1089-1098.
7. Vadboncoeur, J., Jampol, L. M., & Goldstein, D. A. (2020). Acute Zonal Occult Outer Retinopathy (AZOOR): a case report of regression following intravitreal dexamethasone (Ozurdex) implant. *Retin Cases Brief Rep*.