DOI 10.5935/0034-7280.20200087

Azoor: Acute Zonal Occute Outer retinopathy associated with autoimmune disease

Azoor: Retinopatia externa oculta zonal aguda associada com doença autoimune

Carolina Correa Leal Lima[1] https://orcid.org/ 0000-0003-3921-3532
Maria Luisa Gois da Fonsêca[1] https://orcid.org/ 0000-0002-3800-5136
Raul Nunes Galvarro Vianna[1] https://orcid.org/0000-0003-2045-466X

Abstract

Acute zonal occult external retinopathy (AZOOR) was first described by Gass in 1993 as a syndrome with rapid loss of one or more large areas of the external retinal segments. Male, 35 years, with Crohn's disease complaining of occasional eye pain and right eye nictalopia since childhood. In regular use of azathioprine and mesalazine. better visual acuity 20/20 OU. At fundoscopy, hyperpigmented lesions in the right temporal arcade of the right eye, in trellis, accompanying local vasculature. After exclusion of differential diagnoses, Azoor's diagnosis was reached. Azoor is an idiopathic syndrome characterized by an acute onset of photopsy, scotoma or both and is typically associated with a persistent loss of visual function involving one or more areas of the external retina. Despite classic photopsy symptoms, the patient in question had an atypical clinical presentation. We describe a case of a peripheral male with choroidal thinning and associated autoimmune disease. Thus, we believe that further investigation is necessary to verify the etiology of choroidal alteration and its association with the specific disease.

Keywords: *Azoor; Autoimmune diseases; Crohn disease; Tomography, optical coherence; Angiography; Retina*

Resumo

Retinopatia externa oculta zonal aguda (AZOOR) foi descrita pela primeira vez por Gass em 1993 como uma síndrome com perda rápida de uma ou mais zonas extensas dos segmentos externos da retina. Paciente masculino, 35 anos, portador de doença de Crohn, queixando-se de dor ocular eventual e nictalopia em olho direito desde infância. Em uso regular de azatioprina e mesalazina. melhor acuidade visual 20/20 AO. À fundoscopia, lesões hiperpigmentadas em arcada temporal inferior de olho direito, em treliça, acompanhando vasculatura local. Após exclusão de diagnósticos diferenciais chegou-se ao diagnóstico de Azoor. Azoor é uma síndrome idiopática caracterizada por um quadro agudo início de fotopsia, escotoma ou ambos e é tipicamente associado a uma perda persistente de função visual que envolve uma ou mais zonas da retina externa. Apesar dos sintomas clássicos de fotopsia, o paciente em questão teve uma apresentação clínica atípica. Descrevemos um caso ocorrido em indivíduo do sexo masculino em região periférica apresentando afinamento coroidiano e com doença autoimune associada. Dessa forma, acreditamos que é necessária maior investigação para verificar a etiologia da alteração coroideana e da associação com a doença específica.

Descritores: Azoor; Doença autoimune; Doença de Crohn; Tomografia de coerência óptica; Angiografia; Retina

[1]Department of Retina and Vitreous, Hospital Universitário Antonio Pedro, Medical School, Universidade Federal Fluminense, Rio de Janeiro, RJ, Brazil.

The work was submitted and approved by the ethics committee HUAP -UFF under the number: CAAE 12125619.9.0000.5243

The authors declare no conflict of interest

Received for publication 13/10/2019 - Accepted for publication 26/11/2019.

Introduction

Acute zonal occult external retinopathy (AZOOR) was first described by Gass in 1993 as a syndrome with rapid loss of one or more large areas of the external retinal segments. [1]

It affects predominantly young women, and is characterized by photopsy, fundoscopy alterations, and electroretinographic abnormalities affecting one or both eyes. [1,2]

The etiology of AZOOR remains controversial. Gass suggested an infectious viral process of the external retina. Autoimmune and inflammatory hypotheses have been proposed by Jampol and Becker.[3] Other possible mechanisms include fungal infiltration, polycythemia vera, toxic retinopathy, antirretin antibody. [1,2]

The present paper aims to describe an atypical case of AZOOR.

Case report

A 35-year-old male patient with Crohn's disease with occasional ocular pain and right eye nictalopia since childhood in regular use of azathioprine and mesalazine.

Patient had better visual acuity 20/20 J1. Biomicroscopic examination without relevant alterations. Normotensive intraocular pressure.

At fundoscopy, he presented hyperpigmented lesions in the lower temporal arch of the right eye in trellis following local vasculature. (Figure 1)

In OCT, both eyes showed preserved retinal layers, foveal anatomy, and vitreoretinal interface in the macular region of both eyes associated with thinning and areas of irregularity in the submacular choroid of the affected eye. In the area of the lesion we observed thinning of all retinal layers, besides subretinal hyperreflexivities. (Figure 2)

Autofluorescence examination reveals preservation of retinal structures in the macular region, and hypoautofluorescence in the region of pigment blockade surrounded by hyperautofluorescence. (Figure 1)

The OCT-a was examined and showed preservation of the macular vascular plexuses in addition to absence of lesions in all layers studied. (Figure 3)

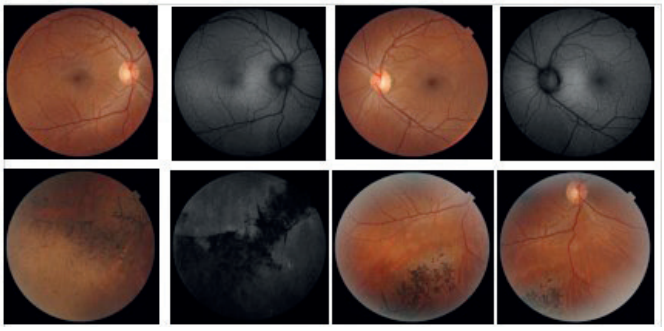

Figure 1: Retinography and autofluorescence

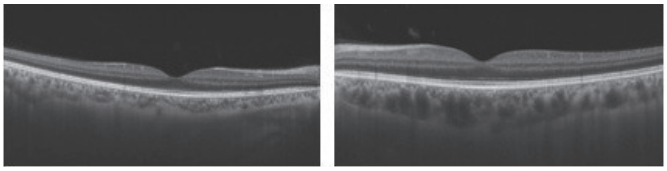

Figure 2: Optical coherence

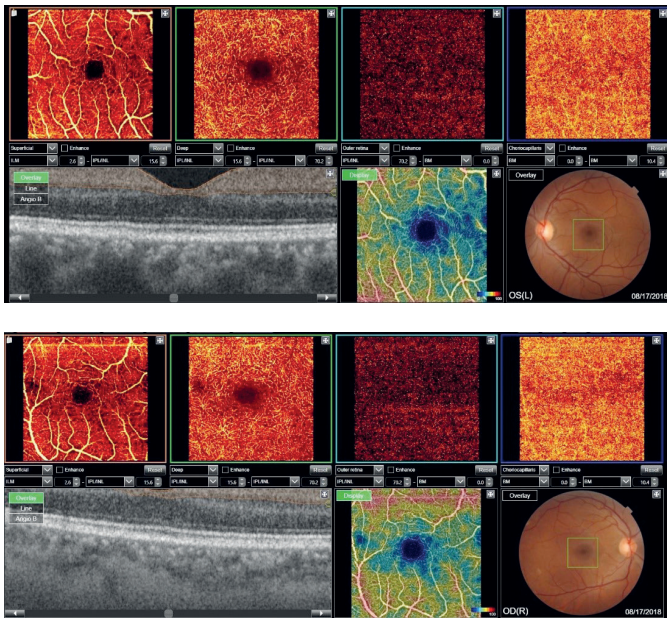

Figure 3: Optical coherence tomography - angiography

ERG and CVC showed no alterations especially due to size and location of the lesion.

The present study was approved by the Research Ethics Committee of Universidade Federal Fluminense under number 12125619.9.0000.5243.

Discussion

Acute zonal occult outer retinopathy (AZOOR) is an idiopathic syndrome characterized by an acute condition of photopsy, scotoma or both, and is typically associated with a persistent loss of visual function involving one or more areas of the external retina.[1,4]

In 1992, Donald Gass reported the first cases of AZOOR during a lecture by the Netherlands Ophthalmological Society. At that time, he had a series of cases in thirteen patients.[1]

AZOOR may involve one or both eyes, and the condition usually stabilizes within 6 months of the onset of symptoms, although some cases continue to progress. In our case, the onset of symptoms is not well established, but the lesion remained stable throughout the follow-up. [1,4]

It is a rare disease characterized by acute visual field loss with photopsy, minimal or no presence of vitreous cells, minimal alterations of the eye fundus, normal fluorescein angiography, and decreased amplitude of electroretinographic waves. The case described had no alterations in the computerized visual campimetry nor in the electroretinogram due to the unusual and unilateral lesion location.[5]

Despite classic photopsy symptoms, the patient in question had an atypical clinical presentation. This rare diagnosis was corroborated in the patient in question by first excluding retinal effects caused by Crohn's disease (major ones: uveitis, episcleritis, scleritis, optic neuritis, retinal serous detachment, retinal vascular occlusion) and the medication in use.[6]

And associated with lesions characteristic of fundoscopy and retinography, as well as OCT with typical subretinal hyperre-

flexivity and thinning of all retinal layers in the lesion. In addition, hyperautofluorescence was observed around the lesion area.

AZOOR lesion is a degeneration of the external segment of the photoreceptor. The pathology and vision impairment are present in a unilateral presentation. However, concomitant involvement of the other eye is observed in some individuals. [5]

The pathogenesis of this disease is still controversial, and theories have been proposed including immunologically mediated virus or even fungi. After a detailed anamnesis, it was not possible to establish the etiology of our patient.[7]

There are reports of AZOOR associated with demyelinating white matter lesions, transverse myelitis, and clinically confirmed multiple sclerosis.[8] These entities may share an underlying autoimmune etiology. The patient in question had an autoimmune disease that may be related to the development of retinal lesions.

Gass speculated that AZOOR was pathologically and etiologically related to a mode spectrum called the "AZOOR complex", and included multiple evanescent white dot syndrome (MEWDS), acute idiopathic blind spot enlargement syndrome (AIBSES), acute macular neuroretinopathy (AMN), presumed ocular histoplasmosis (POHS), punctate inner choroidopathy (PIC), and multifocal choroiditis (MFC).[1,9]

Fundus autofluorescence (FAF) findings in patients with AZOOR have been described in a limited number of reports. A common finding about FAF imaging in AZOOR is a hypoautofluorescence zone surrounded by hyperautofluorescence.[10,11] Mrejen et al. classified these findings of autofluorescence as a trizonal area that can also be seen on OCT and green indokinin angiography. This trizonal lesion is characterized by AZOOR lesion having a hypoautofluorescent pattern inside with a hyperaerofluorescence granular line around it and normal autofluorescence outside this area. [10.11] Our patient presented this pattern with autofluorescence.

Optical coherence tomography (OCT) provides direct evidence of the compromised ellipsoid zone, the distinctive feature of acute retinal disturbance. In addition, OCT allows the detection of ellipsoid zone change dynamically, identifying different stages of disease progression. In the case described the disease was already well established, and we could only notice total destruction of the layers in the lesion area besides a submacular choroidal thinning in the affected eye not previously described in the literature studied. [12]

Alternatively, this characteristic change can also be revealed by the implicit delayed time of the 30 Hz cone, faster responses in full-field electroretinogram (ERG), or suppressed responses on the multifocal electroretinogram (mfERG). mfERG has its unique advantages in judging visual functions during recovery. [12]

The therapeutic effects of systemic steroid or immunosuppressive agents have been sporadically reported in the literature. We chose to keep only observation of the lesion and symptoms, since the lesion remained stable and without impairing the visual functions of our patient.

We believe that due to its appearance with a spiculated pattern and atrophic area around the lesion, it is a more advanced AZOOR lesion, as defined by Mrejen et al. [10]

Conclusion

Azoor is a rare disease with poorly understood pathophysiology. We describe a case occurred in a male patient in a peripheral area with choroidal thinning and associated autoimmune disease.

Although antibody studies have ruled out the autoimmune etiology of this disease, we believe that further investigation is needed to verify the etiology of choroidal alteration and its association with the specific disease, and may represent an association with the patient's autoimmune disease.

References

1. Gass JD. Acute zonal occult outer retinopathy. Donders Lecture: The Netherlands Ophthalmological Society, Maastricht, Holland, June 19, 1992. J Clin Neuroophthalmol. 1993;13:79–97.
2. Kuo YC, Chen N, Tsai RK. Acute Zonal Occult Outer Retinopathy (AZOOR): a case report of vision improvement after intravitreal injection of Ozurdex. BMC Ophthalmol. 2017;17(1):236.
3. Jampol LM, Becker KG. White spot syndromes of the retina: a hypothesis based on the common genetic hypothesis of autoimmune/inflammatory disease. Am J Ophthalmol. 2003; 135(3):376-9.
4. Shifera AS, Pennesi ME, Yang P, Lin P. Ultra-wide-field fundus autofluorescence findings in patients with acute zonal occult outer retinopathy. Retina. 2017;37(6):1104–19.
5. Crawford CM, Rivers BA, Nelson M. Acute zonal occult outer retinopathy: vision loss in an active duty soldier. Case Rep Med. 2013; 2013:240607.
6. Yamane IS, Reis RS, Moraes Jr HV. Oclusão venosa central de retina na remissão de doença de Crohn: relato de caso. Arq Bras Oftalmol. 2007;70(6): 1034-6.
7. Tan AC, Sherman J, Yannuzzi LA. Acute zonal occult outer retinopathy affecting the peripheral retina with centripetal progression. Retin Cases Brief Rep. 2017;11(2):134-40.
8. Wang JC, Finn AP, Grotting LA, Sobrin L. Acute Zonal Occult Outer Retinopathy Associated With Retrobulbar Optic Neuritis. J Neuroophthalmol. 2017;37(3):287-90.
9. Tavallali A, Yannuzzi LA. Acute zonal occult outer retinopathy; Revisited. J Ophthalmic Vis Res. 2015;10(3):211-3.
10. Mrejen S, Khan S, Gallego-Pinazo R, Jampol LM, Yannuzzi LA. Acute zonal occult outer retinopathy: a classification based on multimodal imaging. JAMA Ophthalmol. 2014;132(9):1089-98.
11. Shifera AS, Pennesi ME, Yang P, Lin P. Ultra-wide-field fundus autofluorescence findings in patients with acute zonal occult outer retinopathy. Retina. 2017;37(6):1104-19.
12. Si S, Song W, Song Y, Hu Y. The clinical characteristics and prognosis of acute zonal occult outer retin

Corresponding author:
Maria Luisa Gois da Fonsêca
Avenida Marques do Paraná, 303 - Centro - Niterói
Rio de Janeiro – RJ - CEP 24033900