Case Report

Acute zonal occult outer retinopathy misdiagnosed as giant cell arteritis: a challenging case

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ABSTRACT

Background: Acute zonal occult outer retinopathy (AZOOR) is a rare autoimmune retinopathy that is challenging to diagnose and treat. It usually presents with subtle fundus changes and severe visual symptoms. Herein, we report a challenging case of AZOOR, emphasizing that multimodal imaging could be valuable in diagnosis and monitoring of treatment response.

Case Presentation: A 53-year-old woman presented to the emergency department with a one-week history of subacute, severe, painless vision loss without photopsia in her right eye. Her best-corrected distance visual acuity was 20/800 in the right eye and 20/20 in the left eye. Slit-lamp examination findings were unremarkable, and intraocular pressure was normal in both eyes. Initially, fundus examination findings appeared normal; however, serum levels of inflammatory markers were elevated. Brain and orbital magnetic resonance imaging results were unremarkable. A relative afferent pupillary defect was present in subsequent follow-up examinations at the hospital. The patient initially received a diagnosis of posterior ischemic optic neuropathy secondary to occult giant cell arteritis, underwent steroid treatment, and was evaluated by rheumatology and neurology consultants. Both consultants concurred with the presumed diagnosis. Subsequent multimodal imaging in the ophthalmology clinic revealed a trizonal pattern of fundus autofluorescence. Corresponding to these areas, we noted a loss of the ellipsoid zone on optical coherence tomography, depression on multifocal electroretinogram, and scotoma on visual field testing. Accordingly, the diagnosis of AZOOR was made. The patient was referred back to the rheumatologist for initiation of steroid-sparing treatment, and methotrexate was administered. Five months after the initial presentation, the patient showed significant visual field improvement in both eyes.

Conclusions: Eye care practitioners should consider AZOOR in the differential diagnosis of patients with subacute painless severe unilateral vision loss and unremarkable findings on fundus examination. Multimodal imaging could be valuable in diagnosis and monitoring of treatment response, as observed in the current case. Further studies with larger sample sizes are needed to confirm the value of multimodal imaging and the available management options for AZOOR.

KEYWORDS

acute zonal occult outer retinopathy, white dot syndrome, giant cell arteritides, fundus autofluorescence imaging, electroretinographies, optical coherence tomography, visual field test

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INTRODUCTION

Acute zonal occult outer retinopathy (AZOOR) [1-3] was first described in 1992 as a rapid loss of function of one or more large zones of the outer retina, minimal fundus changes, and electroretinogram (ERG) abnormalities, with subsequent permanent vision loss due to atrophy of retinal pigment epithelial cells. AZOOR can be classified under the white dot syndromes, specifically, as an idiopathic retinal inflammatory disease described by Gass et al. [4], commonly affecting young Caucasian women, with acute unilateral photopsia, visual field defects, and myopia.

AZOOR is a rare form of autoimmune retinopathy [5], with only 131 cases reported in the English literature, according to a review published in 2011 by Monson and Smith [6]. The median patient age is 47 years, and the most common presenting symptoms are photopsia, distortion of central vision, nyctalopia, and temporal blind spots. Mrejen et al. [7] reported that the majority of patients initially presented with a normal fundus. Therefore, diagnosis of AZOOR can be challenging without ancillary testing, such as optical coherence tomography (OCT), fundus autofluorescence (FAF), and intravenous fluorescein angiography (IVFA). Barnes et al. [8], Kuo et al. [9], and Mahajan and Stone [10] reported that there is no agreement on the standard treatment for AZOOR. However, systemic and local corticosteroids, immunosuppressive medications, and antiherpetic drugs have been used to slow disease progression [8-10].

In this case report, we discuss a patient who presented to the emergency department with severe, painless, sudden-onset, unilateral vision loss. Despite the initial suspicion of giant cell arteritis (GCA), multimodal imaging confirmed the diagnosis of AZOOR.

CASE PRESENTATION

A 53-year-old Caucasian woman with a history of well-controlled systemic hypertension, no remarkable family, drug, surgical, and ocular history, and without a clinically significant refractive error, presented to the emergency department in July 2021 with a chief complaint of acute, painless vision loss in her right eye beginning one week before her presentation.

The patient described her symptoms as looking through dirty glasses with a large black spot in her vision. She also reported a blind spot in her left eye. A review of systems was negative, except for vision loss. Her best-corrected distance visual acuity (BCDVA) was 20/800 in the right eye and 20/20 in the left eye using a Snellen chart (Nidek Co., Ltd., Gamagori, Aichi, Japan). Slit-lamp examination findings of the anterior and posterior segments using the Haag-Streit BM 900 (Haag-Streit AG, Bern, Switzerland) were unremarkable, and intraocular pressure (Goldmann applanation tonometer, AT-900; Haag-Streit AG) was normal in both eyes. Because she had undergone dilated examination by her ophthalmologist, a relative afferent pupillary defect (RAPD) could not be detected at the initial visit. Based on the presenting symptoms and signs, she was sent to a tertiary referral hospital with concerns for posterior ischemic optic neuropathy secondary to occult GCA, given her degree of vision loss and the presence of RAPD [11] on later examination.

The patient was examined by an ophthalmologist. On fundus examination, the optic nerves were pink with sharp borders, and there was peripapillary atrophy along with discrete areas of peripheral chorioretinal atrophy and pigmentary changes in both eyes (Figure 1A, B). Laboratory tests revealed an elevated erythrocyte sedimentation rate (ESR) of 60 mm/h. Other acute phase reactants were as follows: C-reactive protein, 3.95 mg/L (normal value: < 3 mg/L [12]); fibrinogen, 265 mg/dL (reference range: 200 to 400 mg/dL [13]); and platelet count, 308 × 10^9/L (reference range: 152 × 10^9 to 371 × 10^9/L [14]).

Brain and orbital magnetic resonance imaging results were unremarkable. An RAPD was revealed (+2) during follow-up examinations at the hospital. Based on clinical findings and laboratory results, the presumed diagnosis of posterior ischemic optic neuropathy secondary to occult GCA was supported. The patient was admitted to the hospital for high-dose intravenous corticosteroid administration [15, 16] and was evaluated by rheumatology and neurology consultants. Both consultants concurred with the presumed diagnosis, and the patient was discharged with a tapering regimen of oral prednisone and scheduled for a temporal artery biopsy (TAB) [15, 16].

At the one-week outpatient follow-up, her visual acuity remained unchanged. The Humphrey visual field (HVF) 30-2 examination (Humphrey Field Analyzer II 750; Swedish interactive threshold algorithm; Carl Zeiss Meditec, Dublin, CA, USA) revealed a generalized depressed visual field in the right eye and an inferotemporal scotoma in the left eye (Figure 2A, B). OCT images (Heidelberg Spectralis SD-OCT; Spectralis software version 5.3.2; Heidelberg Engineering, Inc., Dossenheim, Germany) of the retinal nerve fiber layer were within normal limits in both eyes. Macular OCT showed atrophy of the outer retina in both eyes, but more extensive in the

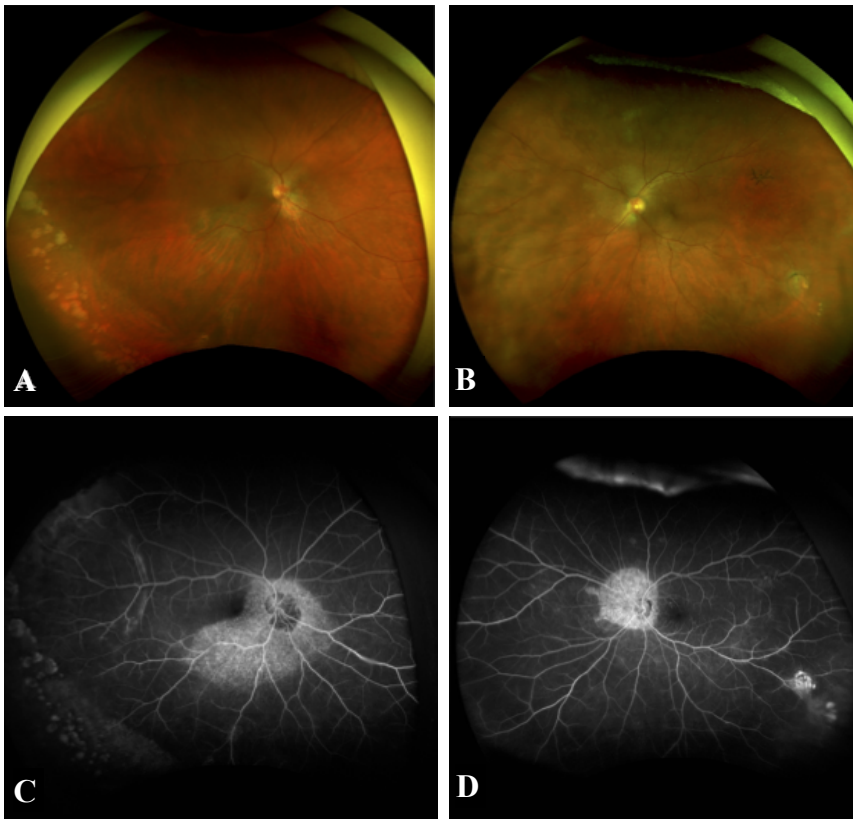

Figure 1. (A and B) Fundus photos (Topcon TRC50LX, Topcon, Tokyo, Japan) of the (A) right and (B) left eyes show peripapillary and peripheral chorioretinal atrophy and pigmentary changes, while the optic nerves have sharp borders with no pallid or chalky white disc edema. (C and D) Intravenous fluorescein angiography (Heidelberg Retinal Angiograph, Heidelberg Engineering, Dossenheim, Germany) of the (C) right and (D) left eyes shows window defects compatible with the peripapillary and peripheral atrophic lesions, as well as some faint background vascular leakage, especially in (C) the right eye.

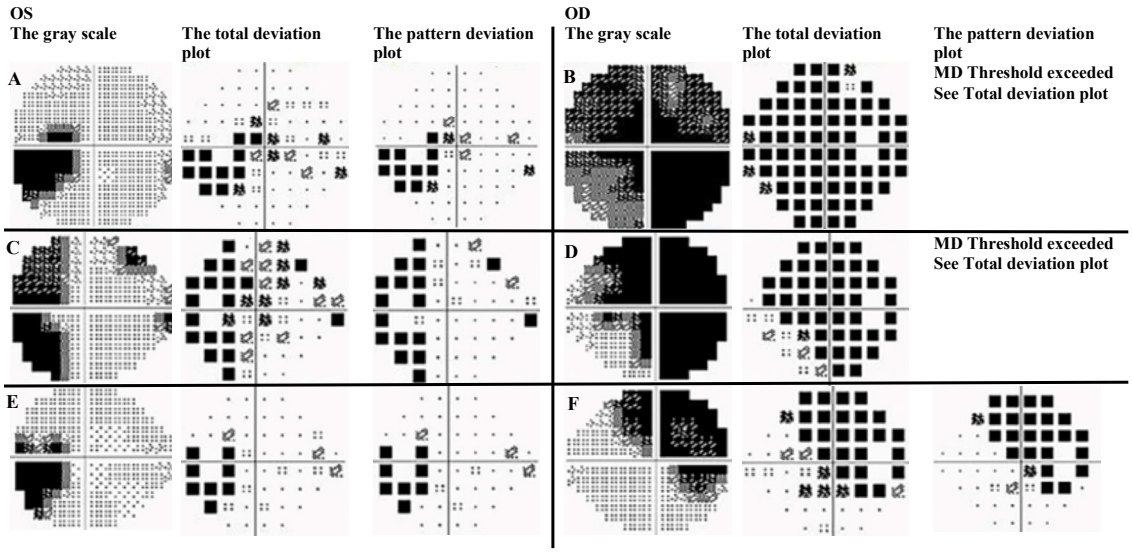

Figure 2. (A and B) Humphrey visual field (HVF) 30-2 examination (Humphrey Field Analyzer II 750; Swedish interactive threshold algorithm; Carl Zeiss Meditec, Dublin, CA, USA) of the (B) right and (A) left eyes on the patient's first visual field assessment. In (B) the right eye, there is generalized depression of the entire field with involvement of central fixation. In (A) the left eye, an enlarged blind spot with an inferotemporal scotoma is evident. (C and D) HVF 24-2 of (C) the left eye two months after presentation, when worsening visual symptoms were reported by the patient. The temporal scotoma progressed to a temporal hemifield defect with a superior defect crossing the vertical midline, while (D) the right eye revealed a subtle improvement. (E and F) Five months after presentation, HVF 24-2 in both eyes (E, left eye, and F, right eye) revealed significant improvement. Abbreviations: OS, left eye; OD, right eye, MD, mean deviation.

right eye (Figure 3A). IVFA (Heidelberg Retinal Angiograph; Heidelberg Engineering, Inc.) demonstrated faint retinal vascular leakage in the posterior pole and periphery, which was more extensive in the right eye. Window defects corresponded to the peripapillary atrophy (Figure 1C, D). A uveitis workup (lupus, sarcoidosis, and syphilis) and tests for anti-recoverin and anti-enolase antibodies were negative. A TAB specimen acquired 16 days after the initiation of corticosteroid therapy showed no evidence of temporal arteritis. However, she continued the tapering prednisone regimen [16] at the discretion of the treating rheumatologist.

Two months after her initial presentation, the patient experienced worsening vision in the left eye. Although the BCDVA in this eye was still 20/20, the HVF of this eye revealed scotoma progression (Figure 2C). The patient was evaluated by a neuro-ophthalmologist and referred to the emergency department for management of a possible exacerbation of GCA. Additional studies were recommended, including routine computed tomography of the chest, abdomen, and pelvis, to assess for a possible occult malignancy with cancer-associated retinopathy [17]. All study results were normal. She was discharged and prescribed an increased dosage of oral prednisone [15, 16].

When the patient returned to the ophthalmology clinic for follow-up, we believed there were multiple features that were inconsistent with GCA, including the patient's relatively young age, lack of other GCA symptoms, a significant HVF defect in the contralateral eye, and macular OCT changes at the time of presentation to the clinic. An FAF (Heidelberg Engineering, Inc.) examination at this visit revealed a classic trizonal pattern [7] (Figure 3C, D). A hypoautofluorescent ring surrounded by a hyperautofluorescent area was present around the optic nerve in both eyes. The hyperautofluorescence extended toward the macula in the right eye (Figure 3C), and some patchy temporal hyperautofluorescence was noted in the left eye (Figure 3D). Areas with normal autofluorescence were observed in both eyes (Figure 3C, D). These areas corresponded with both scotomas

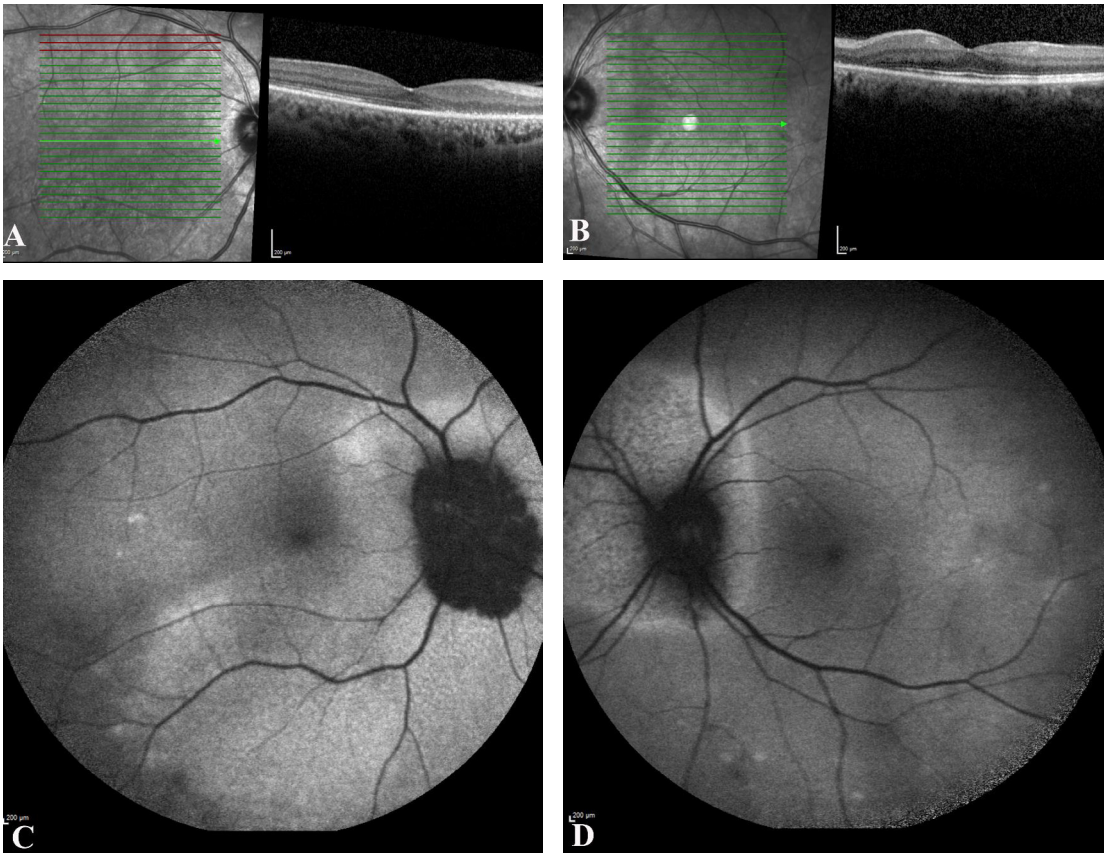

Figure 3. (A and B) Baseline spectral domain optical coherence tomography (Heidelberg Spectralis SD-OCT; Spectralis software version 5.3.2; Heidelberg Engineering, Inc., Dossenheim, Germany) images of the macula showing outer retinal atrophy in the papillomacular bundle of both eyes, which is more extensive in (A) the right eye. (C and D) Fundus autofluorescence (Heidelberg Engineering) of both eyes shows the classic trizonal pattern of acute zonal occult outer retinopathy (AZOOR). A hypoautofluorescent ring can be seen around the optic nerves, surrounded by a hyperautofluorescent area. This hyperautofluorescence extends toward the macula in (C) the right eye, with some patchy temporal hyperautofluorescence in (D) the left eye. Areas with normal autofluorescence can also be seen in (C and D) both eyes. (C and D) Hypo- and hyperautofluorescent areas around the optic nerves signify areas of retinal pigment epithelial cell and/or ellipsoid zone loss, which is evident in (A and B) corresponding optical coherence tomography images.

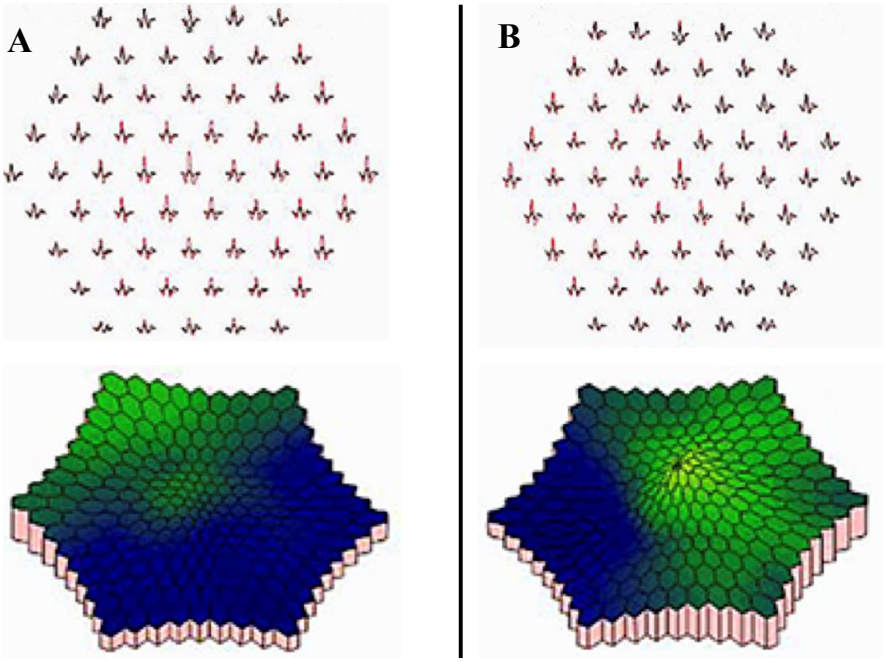

Figure 4. Multifocal electroretinogram of the (A) right and (B) left eyes shows decreased amplitude corresponding to the areas of abnormality on optical coherence tomography and autofluorescence observed in Figure 3.

present on the HVF (Figure 2) and the defects found on multifocal ERG (mfERG) (Figure 4). Visual evoked potential results were incompatible with a sole diagnosis of optic neuropathy. A diagnosis of AZOOR was made with the aid of multimodal imaging. The patient was referred back to the rheumatologist for initiation of corticosteroid-sparing treatment and was prescribed a methotrexate regimen [18].

Five months after the initial presentation, the patient showed a significant improvement in the visual field of both eyes (Figure 2E, F); however, no changes were observed in the follow-up OCT and FAF images. She was advised to continue methotrexate with slow tapering of oral prednisone under expert supervision and regular laboratory monitoring, and follow-up has continued under both the treating rheumatologist and ophthalmologist.

This case report received ethical approval at the department level. Informed consent was obtained from the patient for the publication of this case, including the publication of all images. This article follows the tenets of the Declaration of Helsinki and complies with the Health Insurance Portability and Accountability Act. Information revealing the patient's identity was excluded.

DISCUSSION

We have presented a challenging case of AZOOR that was initially misdiagnosed as GCA. Ultimately, we exploited multimodal imaging features to confirm the diagnosis of AZOOR.

Diagnosis of AZOOR is difficult, and there is often a delay between the onset of symptoms and the diagnosis [19]—as noted in the current case—especially in the emergency department setting with limited ophthalmic testing resources. With visual symptoms in both eyes, an elevated ESR, an inconclusive TAB result, and an apparently normal initial eye examination, a diagnosis of posterior ischemic optic neuropathy due to GCA was suspected despite a lack of symptoms commonly found in GCA, such as jaw claudication, scalp tenderness, headache, diplopia, fever, recent weight loss, and soreness in the shoulders or thighs [20]. The managing clinician initially considered the possibility of occult GCA [21] in the absence of systemic symptoms; however, severe unilateral vision loss, an RAPD, and elevated acute phase reactants were observed in this case.

Although not impossible, GCA was less likely in this patient; however, given the risk of missing a GCA diagnosis [22], treatment was pursued by the on-call team. In this case, the suspected diagnosis was reinforced by multiple specialties, the true diagnosis was delayed, and the patient's hospitalization was prolonged. This misdiagnosis is not uncommon and was reported by Fletcher and Imes [23]. In a patient with vision loss and unremarkable slit-lamp and fundus examinations, an optic nerve or intracranial etiology tends to be more probable [19].

Multimodal imaging can assist in a more accurate diagnosis. Fletcher and Imes [23] emphasized the importance of considering retinal pathology such as AZOOR in one's differential diagnosis in cases with a normal-appearing fundus [23]. This is especially true in the emergency department or hospital, where ancillary ophthalmologic testing may be inaccessible. The diagnosis of AZOOR involves multimodal imaging, as described by Monson and Smith [6], using OCT, FAF, and sometimes IVFA. IVFA findings usually range from subtle leakage of the retinal vasculature, as observed in the current case (Figure 1C, D), to more extensive leakage [24].

Usually, there are focal areas of inflammation of the outer retina with autofluorescence changes in those areas and corresponding defects in the HVF and mfERG, as well as loss of the outer retina on OCT in those areas. Patients usually report photopsia with preservation of central vision until the late stages of the disease [24, 25]. We did not perform OCT angiography [26] in this case, which might be a limitation of our report. However, we could redirect the diagnostic workup by considering that the optic nerve in GCA cannot have a pink appearance during exacerbation and is usually observed as pallid or chalky white disc edema [27, 28].

As in its diagnosis, the treatment of AZOOR is challenging for ophthalmologists. Due to the rarity of the disease, no randomized clinical trials have investigated treatment options. Barnes et al. [8] found that intravitreal corticosteroids may play a role in treating AZOOR, as they reported improvement in disease stability in all nine observed eyes. However, this treatment carries significant risks, such as the development of central serous chorioretinopathy, ocular hypertension, and cataracts. A small case series by Mahajan et al. [10] reported three patients with AZOOR who experienced rapid improvement with valacyclovir treatment; however, this benefit was not confirmed in other studies. Our patient responded to systemic methotrexate treatment while taking a slowly tapering dose of oral prednisone.

Treatment with systemic corticosteroids has also been investigated, and a case series of nine patients by Chen et al. showed benefits in treatment with high-dose intravenous corticosteroids [29]. Moreover, Guijarro et al. [30] reported azathioprine treatment in a 42-year-old woman with AZOOR, who unfortunately experienced progression of the disease despite the subsequent administration of intravenous immunoglobulins and subcutaneous abatacept. Most treatments aim to stabilize the disease, as it usually progresses and visual function deteriorates [30]. Therefore, a timely diagnosis may halt extension of the scotoma and preserve central vision.

Our case report could have educational value for eye care practitioners who encounter challenging cases of painless, subacute, unilateral vision loss in the emergency department and seek multimodal imaging assistance. However, the limitations of our case report are an atypical presentation of a rare ocular entity (limiting generalization), the likelihood of over-interpretation, and the retrospective case report design. Future studies with larger sample sizes are required to determine the value of multimodal imaging assistance, various treatment options, and especially, more intensive management of AZOOR. These studies could identify the optimal diagnostic approach and treatment regimens for saving or partially restoring vision.

CONCLUSIONS

AZOOR is a rare disease that presents as acute vision loss and is challenging to diagnose and treat. The unremarkable examination findings in our case reinforced the presumed diagnosis of GCA. Clinicians should consider AZOOR in the setting of severe unilateral vision loss, a near-normal fundus examination, and normal brain imaging so that patients can receive timely, appropriate imaging and avoid unnecessary interventions. Multimodal imaging can assist in diagnosis and monitoring of treatment response, as we observed in the current case. However, studies with larger sample sizes are required to determine the value of multimodal imaging and available options for managing AZOOR. Such studies could identify the optimal diagnostic approach and treatment regimens for saving or partially restoring vision.

ETHICAL DECLARATIONS

Ethical approval: This case report received ethical approval at the department level. Informed consent was obtained from the patient for the publication of this case, including the publication of all images. This article follows the tenets of the Declaration of Helsinki and complies with the Health Insurance Portability and Accountability Act. Information revealing the patient's identity was excluded.

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