American Journal of Ophthalmology Case Reports 26 (2022) 101558

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

Coarsening of choriocapillaris on optical coherence tomography angiography as a sign of acute idiopathic blind spot enlargement

Hema Lattha Gunasagaran [a,*], Anna Waldie [a], William Xiao [b], Phoebe Moore [a]

[a] North West Eyes, Tamworth, 110 Carthage Street, East Tamworth, NSW, Australia, 2340
[b] OPSM Tamworth, 335 Peel Street, Tamworth, NSW, Australia, 2340

ARTICLE INFO

Keywords:
Acute idiopathic blind spot enlargement
Imaging

ABSTRACT

Purpose: Acute idiopathic blind spot enlargement is a rare syndrome that is classified within a spectrum of primary inflammatory choriocapillaropathies with circumscribed outer retinal dysfunction.
Observations: We observed coarsening of the choriocapillaris on en-face optical coherence tomography angiography when compared to the fellow eye in a patient with suspected acute idiopathic blind spot enlargement.
Conclusions and Importance: Increased granularity of the choriocapillaris as imaged by optical coherence tomography angiography may assist in the diagnosis of acute idiopathic blind spot enlargement, particularly during a global pandemic when access to electrodiagnostics is limited. This finding supports the current evidence of choriocapillaris hypoperfusion in the pathogenesis of acute idiopathic blind spot enlargement.

1. Introduction

Acute idiopathic blind spot enlargement (AIBSE) syndrome is a rare, peripapillary retinopathy first described by Fletcher et al., in 1988 as a temporary steep edged scotoma centred on the blind spot in the presence of normal fundoscopic findings.[1] It was later classified within the Acute Zonal Occult Outer Retinopathy (AZOOR) spectrum of diseases that also comprised of Multiple Evanescent White Dot Syndrome (MEWDS) and Acute Macular Neuroretinopathy (AMN).[1]

AIBSE typically occurs in the third decade of life, with a female predominance. Clinical features include photopsia, and acute unilateral onset of scotoma or enlarged blind spot in the absence of optic disc oedema. It may cause a relative afferent pupillary defect as the disease affects the peripapillary retina.[1] Fundus fluorescein angiography (FFA) may be normal, or may show peripapillary hyperfluorescence.[1] Fundus autofluorescence (FAF) discloses peripapillary hyperautofluoroscence.[1] Optical coherence tomography (OCT) features of AIBSE include ellipsoid zone disruption during the acute phase of the disease[1] and discontinuation of the external limiting membrane temporal to the macula.[2] Multifocal electroretinography (mERG) demonstrates attenuated responses in the peripapillary area corresponding with the enlarged blind spot on automated perimetry.[1] The natural history of AIBSE is variable, however visual acuity may improve over several months, however blind spot enlargement generally persists.[1,3]

To date, there have been limited studies on the features of AIBSE on OCT angiography (OCTA). In 2020, a Spanish study reported choriocapillaris changes in a patient with AIBSE using OCTA.[4] Numerous studies have described OCTA findings in MEWDS. More recently, Tang et al. reported changes in the retinal and choriocapillaris layer during the acute phase of MEWDS. We present a rare case that shows coarsening of the choriocapillaris on OCTA as a sign of AIBSE.[5]

Case report

A 31-year-old female presented with a 1-week history of left sided blurred vision, photopsia and headache, on a background of a mild respiratory illness 3 weeks prior. At presentation, left visual acuity (VA) was 6/7.5, with normal intraocular pressure and full Ishihara colour vision testing. Bilateral anterior segment examination was healthy. There was mild vitritis, disc hyperaemia, and a granular macular appearance in the left eye. The right posterior segment was normal.

FAF revealed patchy peripapillary, and temporal macular hyperautofluorescence (Fig. 1), corresponding to areas of ellipsoid zone disruption on OCT (Fig. 2A and B). OCTA showed a coarsening of the choriocapillaris when compared to the fellow eye (Fig. 3A and B).

Humphrey visual field testing was unreliable due to high false negatives, but showed diffuse field loss with a visual field index of 23%. FFA showed non-specific changes of mild disc stain and mild, patchy capillary leak with no angiographic evidence of vasculitis and no focal

* Corresponding author. 110 Carthage Street, East Tamworth, NSW, 2340,
E-mail address: hl.gunasagaran@gmail.com (H.L. Gunasagaran).

https://doi.org/10.1016/j.ajoc.2022.101558
Received 4 December 2021; Received in revised form 25 April 2022; Accepted 25 April 2022
Available online 2 May 2022

Abbreviations

AIBSE	Acute idiopathic blind spot enlargement
AMN	Acute macular neuroretinopathy
APMPPE	Acute posterior multifocal placoid pigment epitheliopathy
AZOOR	Acute zonal occult outer retinopathy
CRP	C-reactive protein
ESR	Erythrocyte sedimentation rate
mERG	Multifocal electroretinography
MEWDS	Multiple evanescent white dot syndrome
MRI	Magnetic resonance imaging
OCT	Optical coherence tomography
OCTA	Optical coherence tomography angiography
VA	Visual acuity

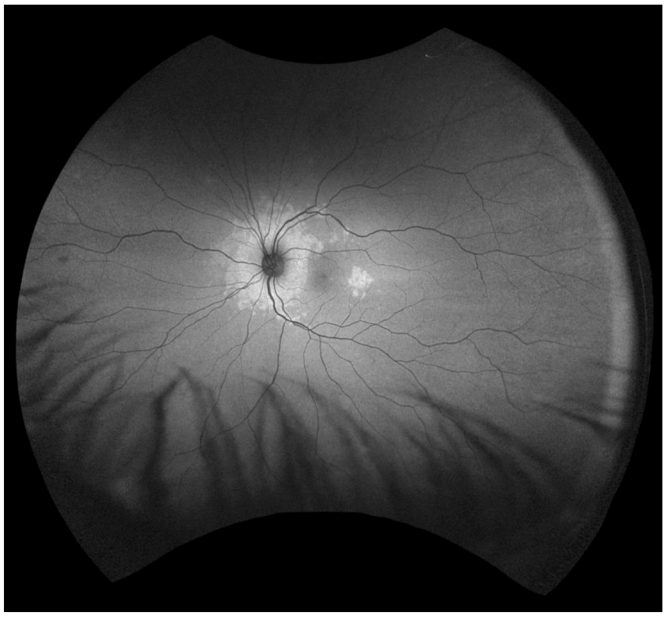

Fig. 1. Left fundus autofluorescence demonstrating peripapillary and temporal foveal hyperautofluorescence.

lesions.

The patient underwent systemic workup, yielding negative syphilis and Epstein Barr virus serology, normal angiotensin-converting enzyme, and a negative vasculitis screen. A full blood count and electrolytes were within normal limits. Inflammatory markers were elevated including Erythrocyte sedimentation rate (ESR) 47mm/hr [0–12] and C-reactive protein (CRP) 10.9 μg/mL [<3]. MRI/MRA brain was undertaken given the headaches and was normal.

The patient was initially diagnosed with MEWDS and a period of observation recommended, however, visual acuity reduced to 6/30 and her symptoms subjectively deteriorated with increased blurring and photopsias. Her OCT remained stable.

The patient was commenced on oral prednisolone 60mg daily with a weekly taper; however, this was self-ceased after 2 weeks due to intolerable side effects. No response to treatment could be demonstrated after the 2 weeks of treatment.

A presumed diagnosis of AIBSE was made and the patient was referred for electrophysiology. Unfortunately, the referral coincided with increasing cases of COVID-19, and statewide lockdown. The electrophysiology was deferred until statewide restrictions eased, however, clinical improvement commenced prior to lifting of restrictions.

No further deterioration was observed, and the clinical picture remained constant over several months with VA stabilising at 6/30. Four months following onset of symptoms, spontaneous recovery was observed and the ellipsoid zone integrity was re-established on OCT. At 5 months, while the patient reported persisting light sensitivity and photopsias and the blind spot remained enlarged, VA improved to 6/6 (Fig. 4A and B).

2. Discussion

The specific pathogenesis of inflammatory choriocapillaritis, including AIBSE is still unknown but two hypotheses have been postulated. Firstly, photoreceptor impairment can be secondary to a primary choriocapillaritis. Secondly, a primary inflammation of the photoreceptors also known as “photoreceptoritis” leads to outer segments attrition.[6]

There has been limited research on the features of AIBSE on OCTA. A Spanish study published in 2020 reported well-defined foci of hypoperfusion at the choriocapillaris layer in a patient with AIBSE using OCTA.[4] The authors also observed peripapillary hyporeflectivity with adjacent hyperreflective points at the level of the ellipsoid zone on the en-face image of the OCTA.[4] OCTA features in MEWDS and AZOOR have also been described. Two initial studies reported unremarkable flow changes on OCTA in patients with MEWDS.[7,8] In 2020, Tang et al. described features of OCTA in MEWDS using quantitative analysis. They described a diffuse, significant reduction in flow densities in the retinal deep capillary plexus and choriocapillaris, which recovered during the convalescent phase.[5]

Burke et al. described the use of OCTA to characterise chorioretinal lesions in acute posterior multifocal placoid pigment epitheliopathy (APMPPE). They found significant hypoperfusion at the level of the choriocapillaris in all active lesions, supporting growing evidence that choriocapillaritis is the primary process in the pathogenesis of APMPPE, followed by the development of retinal pigment epithelium and outer retinal changes secondary to the hypoperfusion.[9]

Our study describes a novel finding of asymmetry of en-face OCTA in the choriocapillaris layer. This can be best described as “coarsening” of this image on the affected side. This finding is consistent with the postulated mechanism of choriocapillaritis with choriocapillary ischaemia and compensatory dilation. This study supports the findings of the previously published Spanish study, however is the first study to describe changes objectively as “coarsening”. Our paper adds to the clinical utility of OCTA in the evaluation of AIBSE.

We suggest the utility of OCTA in assisting diagnosis of AIBSE, particularly in a pandemic era, when access to electrodiagnostics is reduced due to lockdown restrictions.

3. Conclusion

OCTA can play an important role in diagnosing as well as providing clinical information to aid in the understanding of the pathogenesis of inflammatory choriocapillaritis, in particular AIBSE.

Patient consent

The patient consented to publication of the case in writing.

Funding

No funding or grant support

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

Research ethics

We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

IRB approval was obtained (required for studies and series of 3 or more cases).

Written consent to publish potentially identifying information, such as details or the case and photographs, was obtained from the patient(s) or their legal guardian(s).

Authorship

The International Committee of Medical Journal Editors (ICMJE) recommends that authorship be based on the following four criteria:

1. Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; AND
2. Drafting the work or revising it critically for important intellectual content; AND
3. Final approval of the version to be published; AND
4. Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

All those designated as authors should meet all four criteria for authorship, and all who meet the four criteria should be identified as authors. For more information on authorship, please see http://www.icmje.org/recommendations/browse/roles-and-responsibilities/defining-the-role-of-authors-and-contributors.html#two.

All listed authors meet the ICMJE criteria.

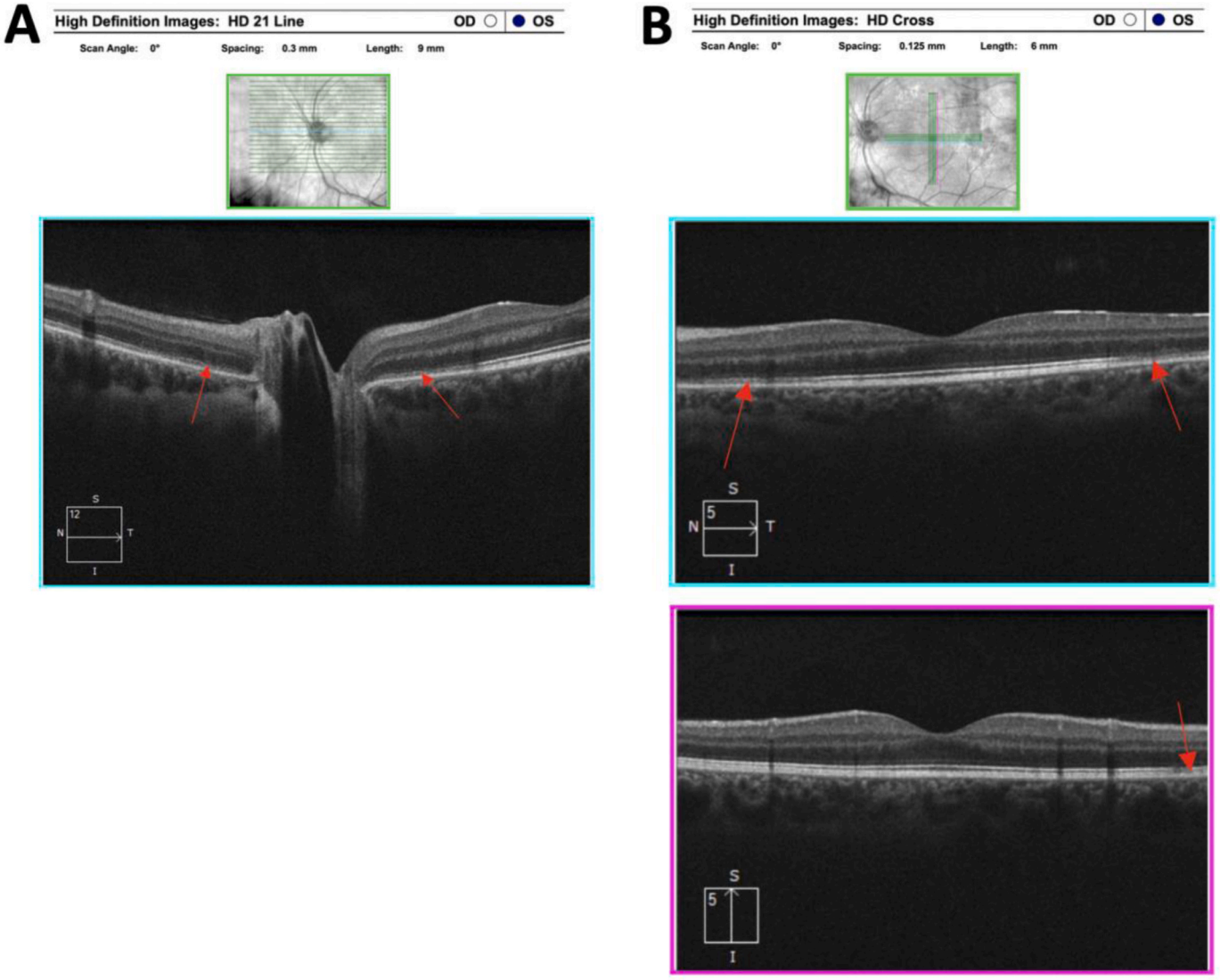

Fig. 2. (A) and (B) Left optical coherence tomography. A is papillary and B is a macula-centred image. Note areas of ellipsoid zone disruption that corresponds to hyperreflective changes around the optic disc and temporal to the fovea (red arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

We attest that all authors contributed significantly to the creation of this manuscript, each having fulfilled criteria as established by the ICMJE.

One or more listed authors do(es) not meet the ICMJE criteria.

We believe these individuals should be listed as authors because:

We confirm that the manuscript has been read and approved by all named authors.

We confirm that the order of authors listed in the manuscript has been approved by all named authors.

Contact with the editorial office

The Corresponding Author declared on the title page of the manuscript is:

Coarsening of choriocapillaris on optical coherence tomography

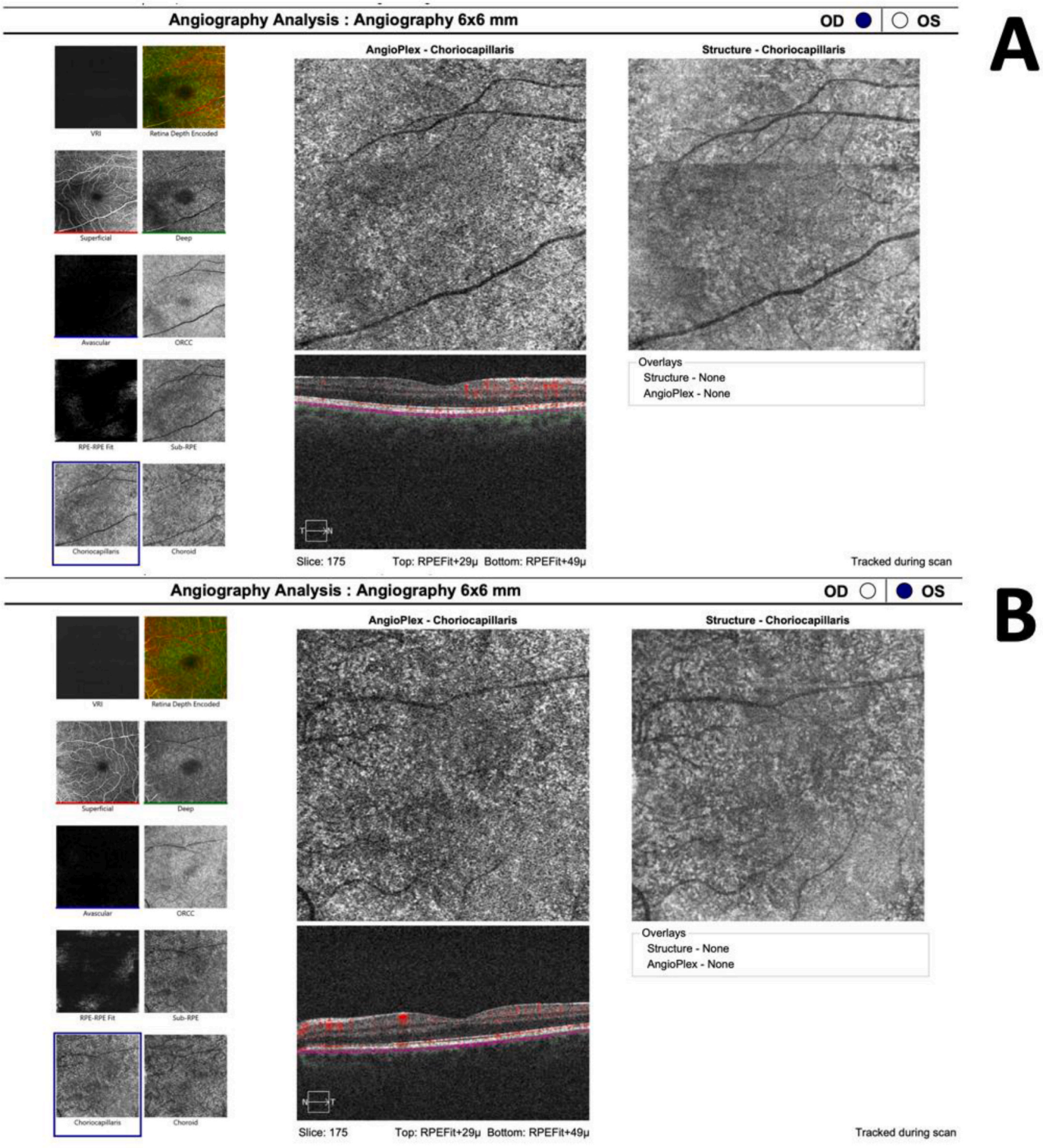

Fig. 3. Optical coherence tomography angiography (OCTA) of the right (A) and left (B) macula. There is coarsening of the choriocapillaris in the left eye relative to the fellow eye.

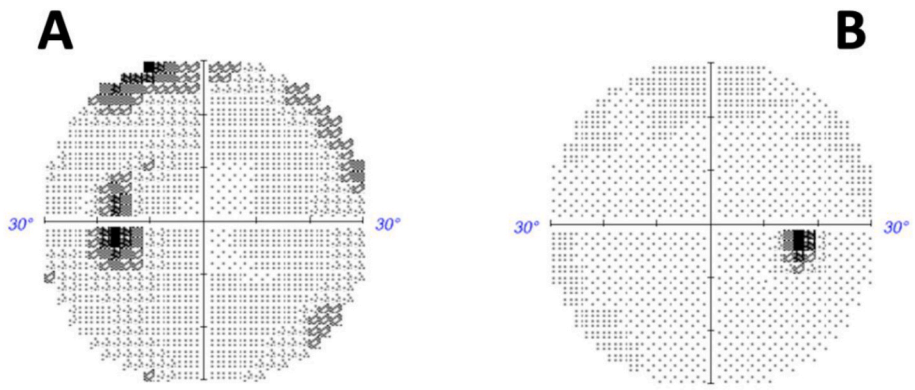

Fig. 4. (A) and (B) Left and right Humphrey visual field (30−2) respectively at 5 months. Note persistent blind spot enlargement on the left.

angiography as a sign of acute idiopathic blind spot enlargement.

This author submitted this manuscript using his/her account in EVISE.

We understand that this Corresponding Author is the sole contact for the Editorial process (including EVISE and direct communications with the office). He/she is responsible for communicating with the other authors about progress, submissions of revisions and final approval of proofs.

We confirm that the email address shown below is accessible by the Corresponding Author, is the address to which Corresponding Author's EVISE account is linked, and has been configured to accept email from the editorial office of American Journal of Ophthalmology Case Reports:

hl.gunasagaran@gmail.com.

Someone other than the Corresponding Author declared above submitted this manuscript from his/her account in EVISE:

We understand that this author is the sole contact for the Editorial process (including EVISE and direct communications with the office). He/she is responsible for communicating with the other authors, including the Corresponding Author, about progress, submissions of revisions and final approval of proofs.

We the undersigned agree with all of the above.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Acknowledgements

No other acknowledgements.

References

1. Fletcher WA, Imes RK. Acute idiopathic blind spot enlargement and acute zonal occult outer retinopathy: potential mimics of neuro-ophthalmic disease. *J Neuro Ophthalmol.* 2020;40:S43–S50.
2. Piri N, Kaplan HJ, Sigford DK, Tezel TH. High-definition optical coherence tomography findings in acute idiopathic blind spot enlargement (AIBSE) syndrome. *Ocul Immunol Inflamm.* 2014;22(6):494–496.
3. Volpe NJ, Rizzo JF, Lessell S. Acute idiopathic blind spot enlargement syndrome: a review of 27 new cases. *Arch Ophthalmol.* 2001;119(1):59–63.
4. Moll-Udina A, Hernanz I, Dotti M, Llorenç V, Adán A. *Multimodal Imaging in Acute Idiopathic Blind Spot Enlargement Syndrome.* English Edition. Archivos de la Sociedad Española de Oftalmología; 2020.
5. Tang W, Guo J, Liu W, Xu G. Quantitative analysis of retinal and choriocapillary vascular density of multiple evanescent white dot syndrome by optical coherence tomography angiography. *Graefe's Arch Clin Exp Ophthalmol.* 2020;258(8):1697–1707.
6. Pellegrini F, Cirone D, De Simone L, Marullo M, Cimino L. Acute zonal occult outer retinopathy complex disease: lessons learned about choroid, photoreceptors, and retinal function. *Eur J Ophthalmol.* 2021;31(2):817–822.
7. Pichi F, Srvivastava SK, Chexal S, et al. En face optical coherence tomography and optical coherence tomography angiography of multiple evanescent white dot syndrome: new insights into pathogenesis. *Retina.* 2016;36:S178–S188.
8. Zicarelli F, Mantovani A, Preziosa C, Staurenghi G. Multimodal imaging of multiple evanescent white dot syndrome: a new interpretation. *Ocul Immunol Inflamm.* 2020;28(5):814–820.
9. Burke TR, Chu CJ, Salvatore S, et al. Application of OCT-angiography to characterise the evolution of chorioretinal lesions in acute posterior multifocal placoid pigment epitheliopathy. *Eye.* 2017;31(10):1399–1408.