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Acute Zonal Occult Outer Retinopathy

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Abstract

Acute zonal occult outer retinopathy (AZOOR) is a rare retinal disease, which is easily missed during its early stages. We report a case of a 31 year old male who presented with acute onset scotoma associated with flashes of light within the scotoma in left eye since 3 weeks. He gave history of common cold 1 month back. His visual aquity was 6/6 in right eye & 6/6p in left eye. Fundus findings were unremarkable in both eyes. HFA revealed temporal field defect in left eye. Prior visiting us, he had multiple neurophysician consultations where he was diagnosed to be normal. OCT showed disruption of outer retinal layers around optic nerve head. Fundus autofluorescence (FAF) showed hyperautofluorescence around the disc. ERG showed mild decrease in 'a' and 'b' wave amplitude. Re-evaluation of fundus revealed very subtle changes which were missed earlier. The patient was diagnosed as a case of AZOOR & started on oral steroids 1mg/kg/day. AZOOR is commonly missed due to normal clinical findings and rarity of the disease. In cases with scintillating scotoma with normal fundus, one should suspect AZOOR as a potential diagnosis.

Keywords: AZOOR, Scintillating scotoma, OCT, FAF, ERG

Introduction

AZOOR (Acute zonal occult outer retinopathy) is a rare retinal disease of unknown etiology with a prelidiction for young women. This disease is often misdiagnosed.

Case Report

A 31 year old male presented with acute onset of scotomata in left eye associated with flashes of light within the scotoma since 3 weeks. He gave history of common cold one month back. On examination, his uncorrected visual acquity was 6/6 in right eye and 6/6p in left eye. Anterior segment evaluation was normal in both eyes. Fundus examination did not reveal any abnormality in both eyes. Intra-ocular pressure was normal in both eyes. Colour vision was normal. Neuro-ophthalmic assessment revealed normal visual field in right eye and a temporal scotoma involving the blindspot in both kinetic and static perimetry in left eye (Fig 1 and 2).

OCT of the right eye was normal, Left eye showed parafoveal disruption of outer retinal layers with loss of IS-OS junction (ellipsoid zone) & disruption of outer nuclear layer (Fig 3 and 4).

Fundus autofluorescence of right eye was normal, Left eye showed a large circumpappillary area of hyperautofluorescence with a margin of even greater hyperautoflourescence (Fig 5 and 6).

Fluorescein angiography of right eye was normal, Left eye showed circumpapillary hyperfluorescent lesion in early phase which increases in intensity in late phase. Indocyanin green angiography of right eye was normal, Left eye showed isointense picture in early phase & circumpapillary hyperfluorescent lesion in late phase (Fig 7 and 8).

ERG of right eye was normal, Left eye showed mild decrease in 'a' and 'b' wave amplitude.

Patient was treated with Tab.Wysolone 1mg/kg/day tapered over 4 weeks. On followup after 4 weeks his visual acquity was 6/6 in both eyes. He was feeling symptomatically better without further deterioration of visual field in his left eye.

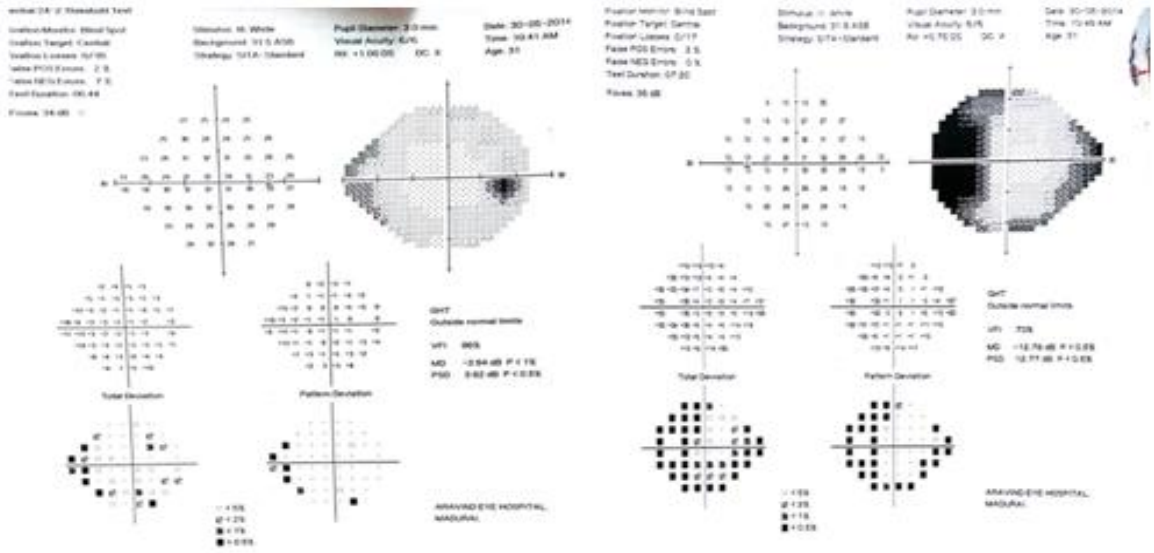


Fig 1 & 2- RE- HFA- normal ; LE- HFA- temporal scotoma involving the blindspot

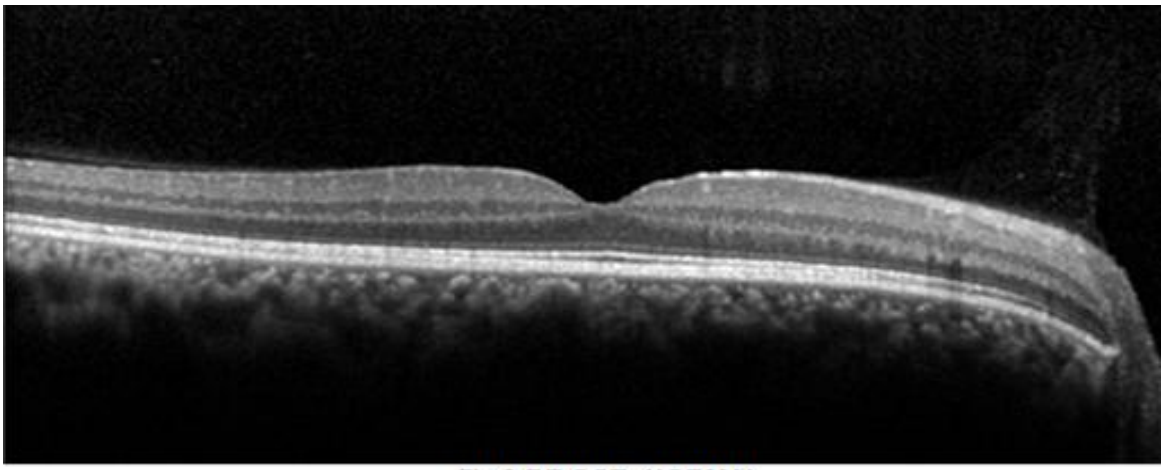


Fig 3.RE OCT- NORMAL

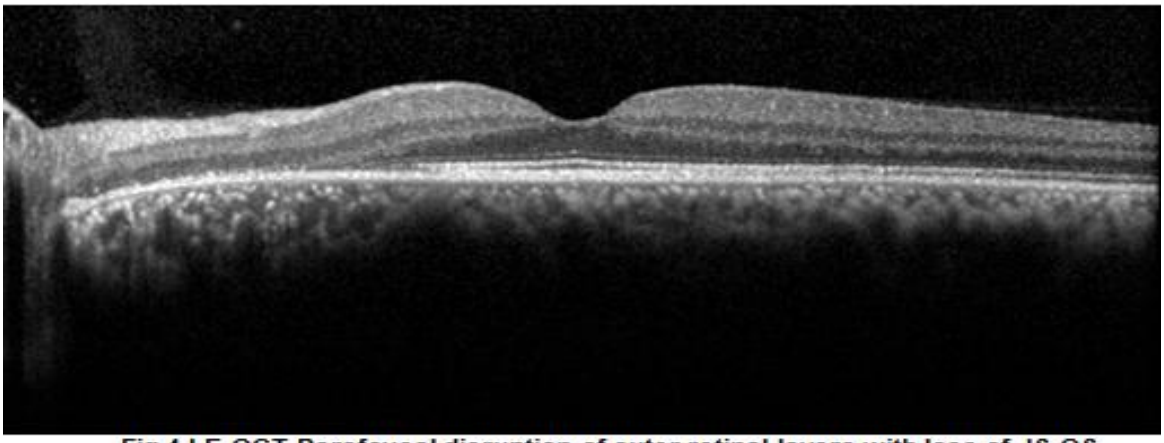


Fig 4.LE OCT-Parafoveal disruption of outer retinal layers with loss of IS-OS junction(ellipsoid zone) & disruption of outer nuclear layer.

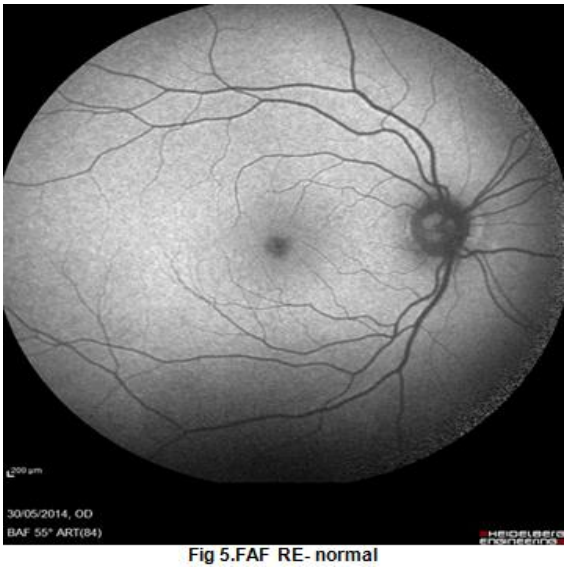


Fig 5.FAF RE- normal

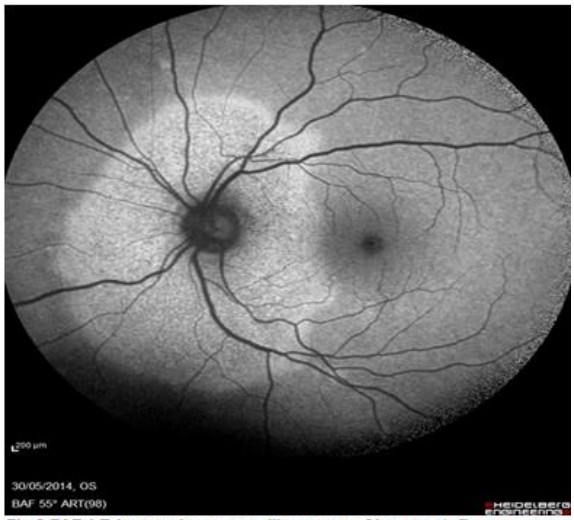
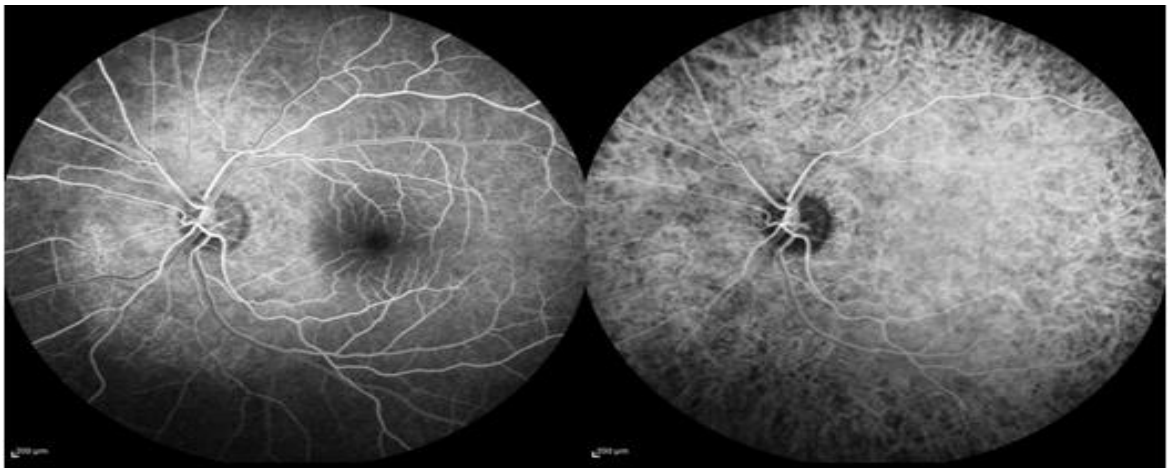


Fig 6.FAF LE-Large circumpappillary area of hyperautofluorescence with a margin of even greater hyperautoflourescence



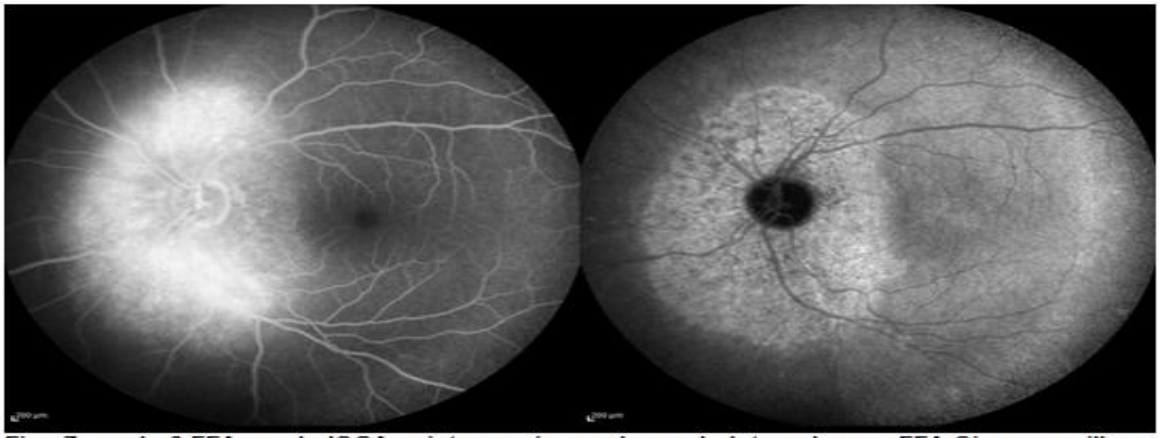


Fig 7 and 8.FFA and ICGA pictures in early and late phase. FFA-Circumpapillary hyperflourescent lesion in early phase which increases in intensity in late phase. ICGA- Isointense picture in early phase & circumpapillary hyperfluorescent lesion in late phase.

Discussion

Acute zonal occult outer retinopathy was first described by Gass in 1992 with a report of 13 patients, mostly young women, with acute loss of zones of outer retinal function, photopsia, ERG changes and minimal funduscopic changes. Since the first reports there have been more than 130 patients described in the literature. Some patients have a viral prodrome. Occasionally vitreous inflammation develops, notably in those with a large zone of visual field loss. The visual field usually stabilizes in 4-6 months and sometimes will improve. Although unilateral in presentation more than 60 percent of the time, there can be delayed involvement of the fellow eye. There can also be recurrences in the same eye.

Gass proposed the pathogenesis to be related to an infection of the retinal photoreceptors adjacent to the optic nerve, likely from a viral source. He felt that the white dot syndromes were related entities that typically occur in young women with a similar constellation of findings. Jampol and Becker have presented an alternative theory with a common genetic hypothesis of autoimmune/inflammatory diseases. They hypothesize that there is an interplay of genetics, the immune system and environmental triggers that lead to specific diseases such as AZOOR.

There has been no treatment proven to improve outcomes in AZOOR. There are several reports of attempted corticosteroid use. Gass felt that oral or intravenous steroids made no difference in outcomes. A recent report documents improved visual function in a patient with steroid pulse therapy. OCT shows disruption of the IS/OS junction in the region of the scotoma and occasionally loss of the outer nuclear layer. Abnormalities of RPE function as shown by autofluorescence have also been reported. Electroretinography is abnormal in the majority of AZOOR cases.

Conclusion

In summary, our patient has AZOOR with the classic symptoms of photopsia and visual field loss. On imaging we demonstrate some of the characteristic findings on SD-OCT and autofluorescence. ERG in our case shows mild change in the left eye consistent with his diagnosis.

AZOOR is a unique retinal disorder that is often misdiagnosed as an optic neuropathy or as hysteria / malingering because the fundus is typically normal ophthalmoscopically and hence the term ‘occult’.

Advances in imaging with SD-OCT and now with adaptive optics, scanning laser ophthalmoscopy enable us to see more of the pathology, where previously the ophthalmoscopic examination was unremarkable in these patients.

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