

INTRODUCTION

Acute zonal occult outer retinopathy (AZOOR) is a rare unilateral or bilateral disease of unknown etiology characterized by focal degeneration of photoreceptors. Currently, there is no established treatment for AZOOR. Owing to the presumed involvement of autoimmunity and inflammation in the pathogenesis of AZOOR, Patients are most commonly either observed for progression or treated with steroid therapy, non-steroidal immunomodulatory agents and less commonly, antivirals or antifungals. Overall, evidence supporting specific treatments for AZOOR remains difficult to interpret as there is lack of randomized controlled trials.

CASE REPORT

This is an observational case report. A20-year-old woman, fit and well, myopic, asymptomatic, who was reviewed in Uveitis clinic in **2018** for second opinion as abnormal visual fields which showed enlarged blind spots were found. There was no family history of Retinitis pigmentosa and positive family history of myopia. Other possible causes of the observed retinal lesions were ruled out.

INVESTIGATIONS AND MANAGEMENT

Visual field demonstrates an **enlarged blind spot** in the left eye.

Fundus examination : **peripapillary atrophy with peripheral bony spicules inferiorly in both eyes. arterial attenuation with temporal optic disc pallor.**

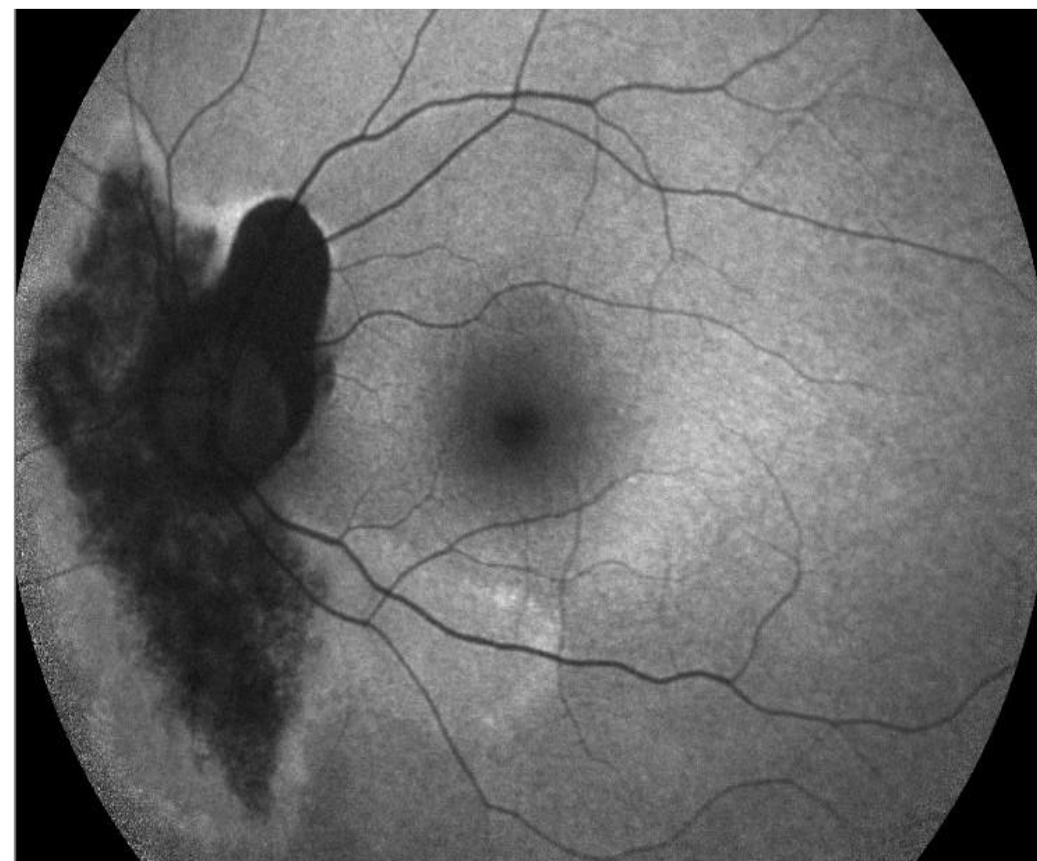
Electrodiagnostic tests : **Generalised rod and cone abnormal pathways with Normal EOG. Possible atypical RP in the peripapillary area was diagnosed.**

In **2022** fundus autofluorescence photographs showed **increased hypo autofluorescence at the margin of the RP atrophy**. AZOOR was identified and initial treatment of **systemic Mycophenolate** was started. since then, close follow ups has not shown any new progressions and peripapillary RPE atrophy remained unchanged.

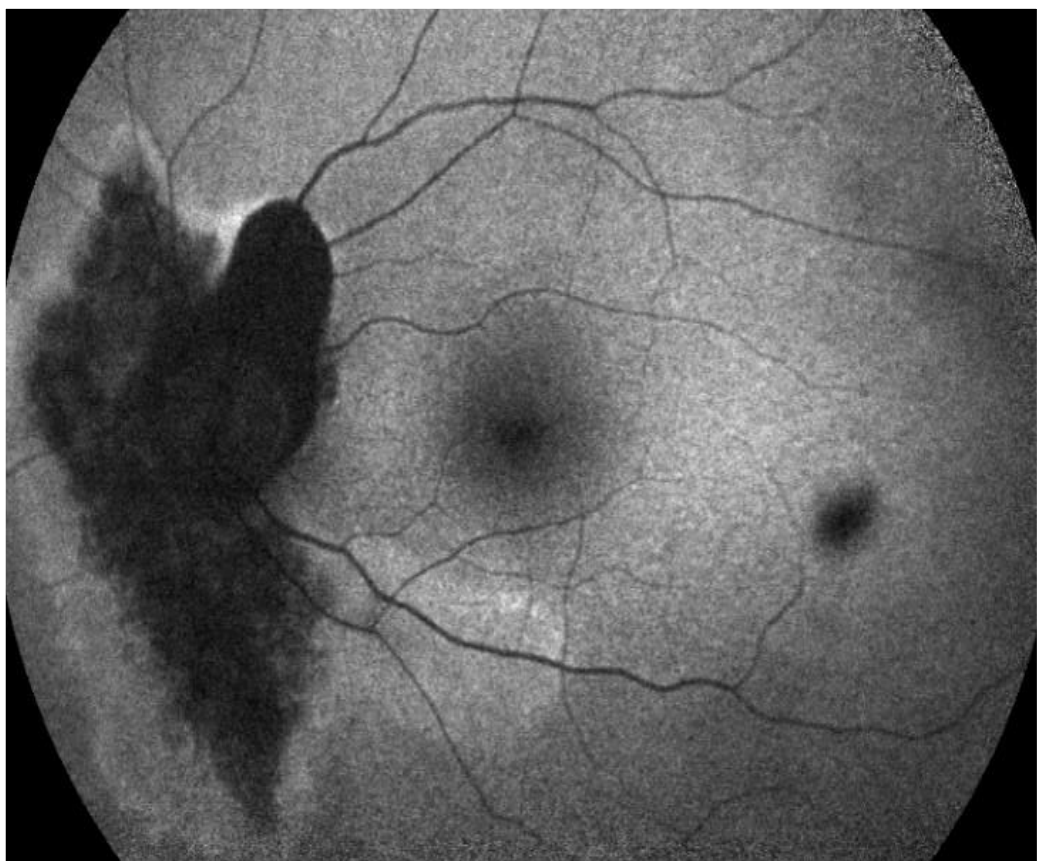
Left eye 2018



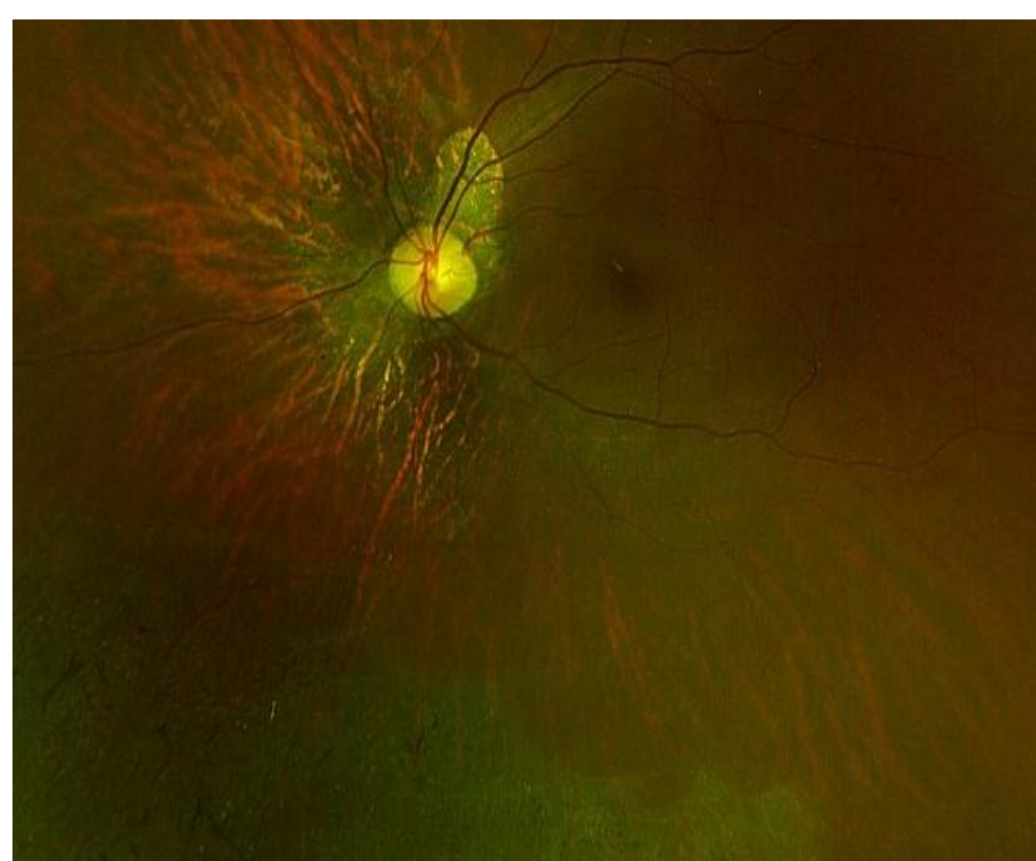
Left eye 2022



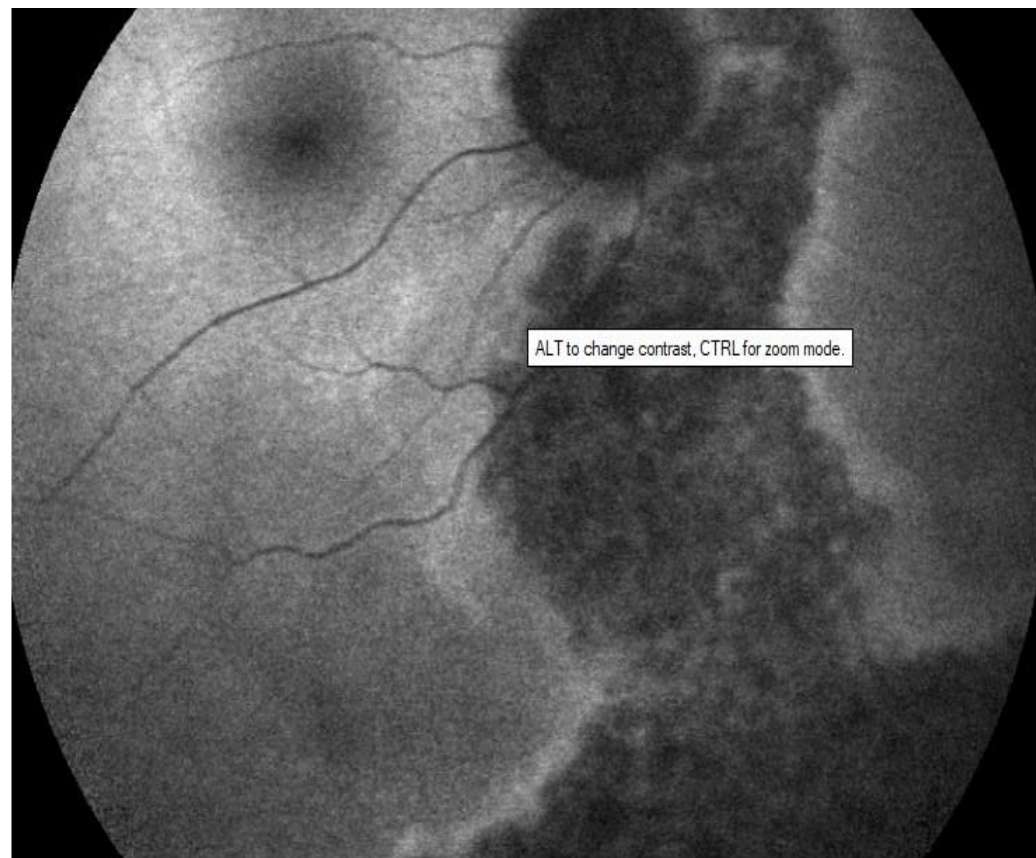
Left eye 2023



Left eye 2024



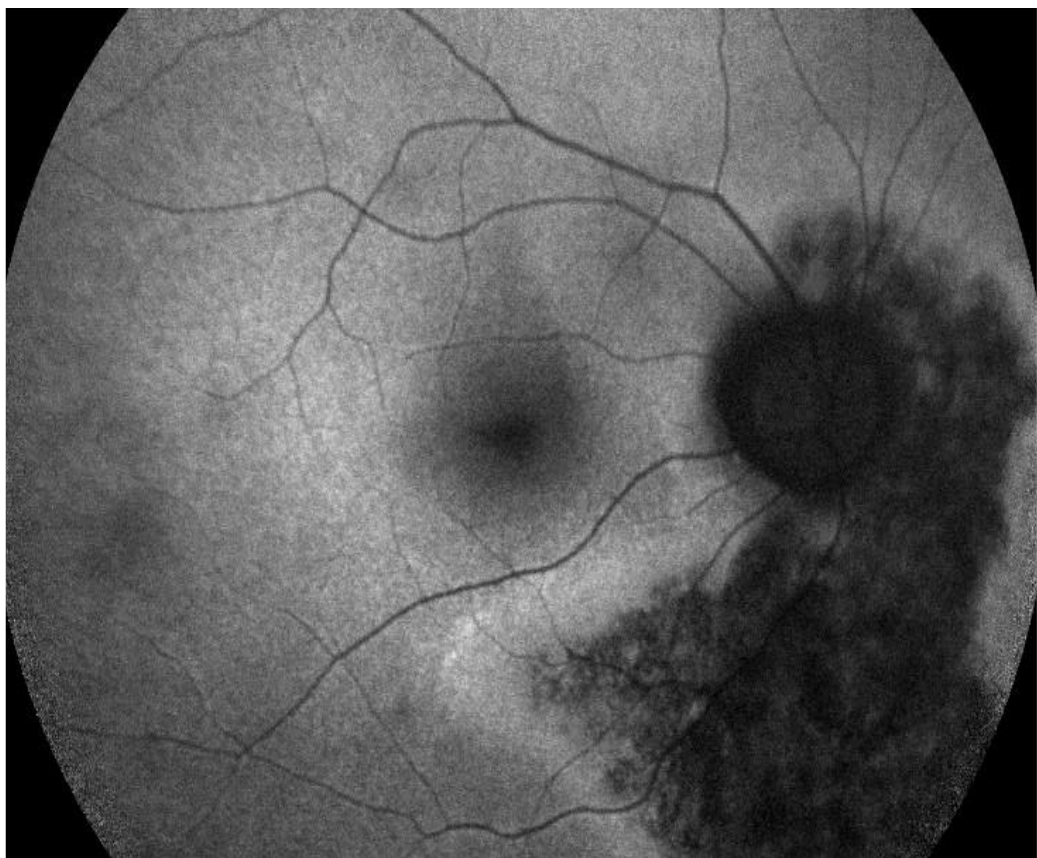
Right eye 2018



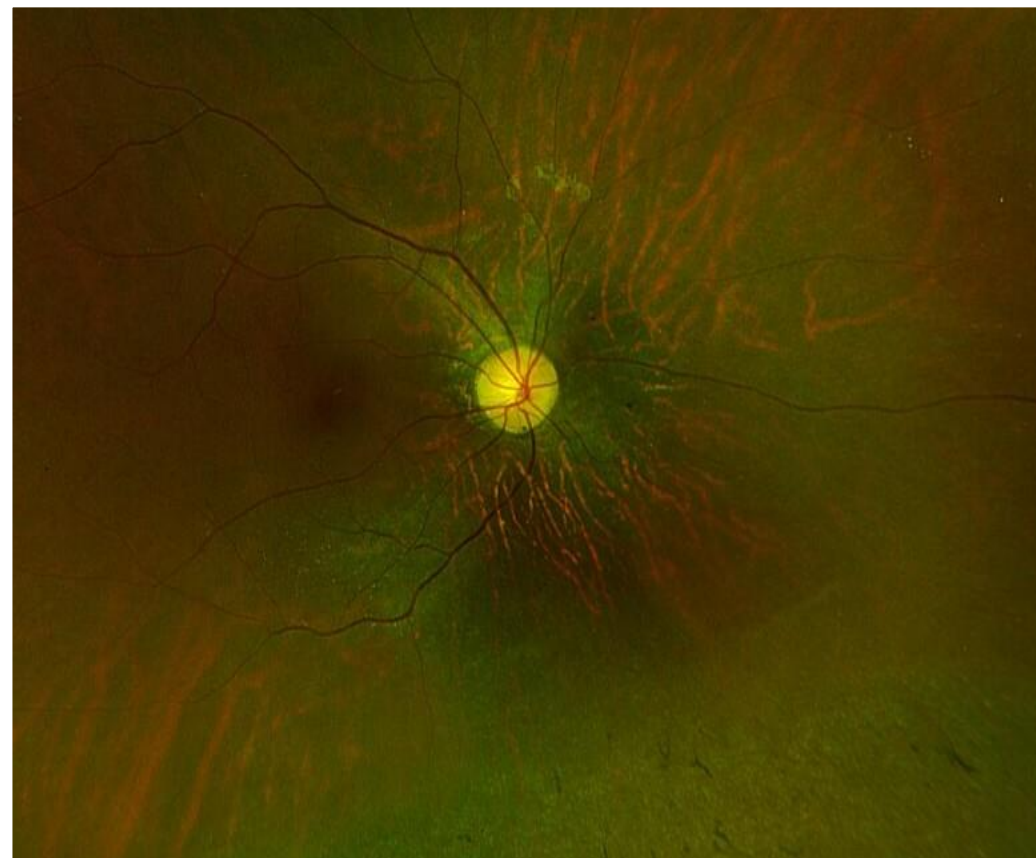
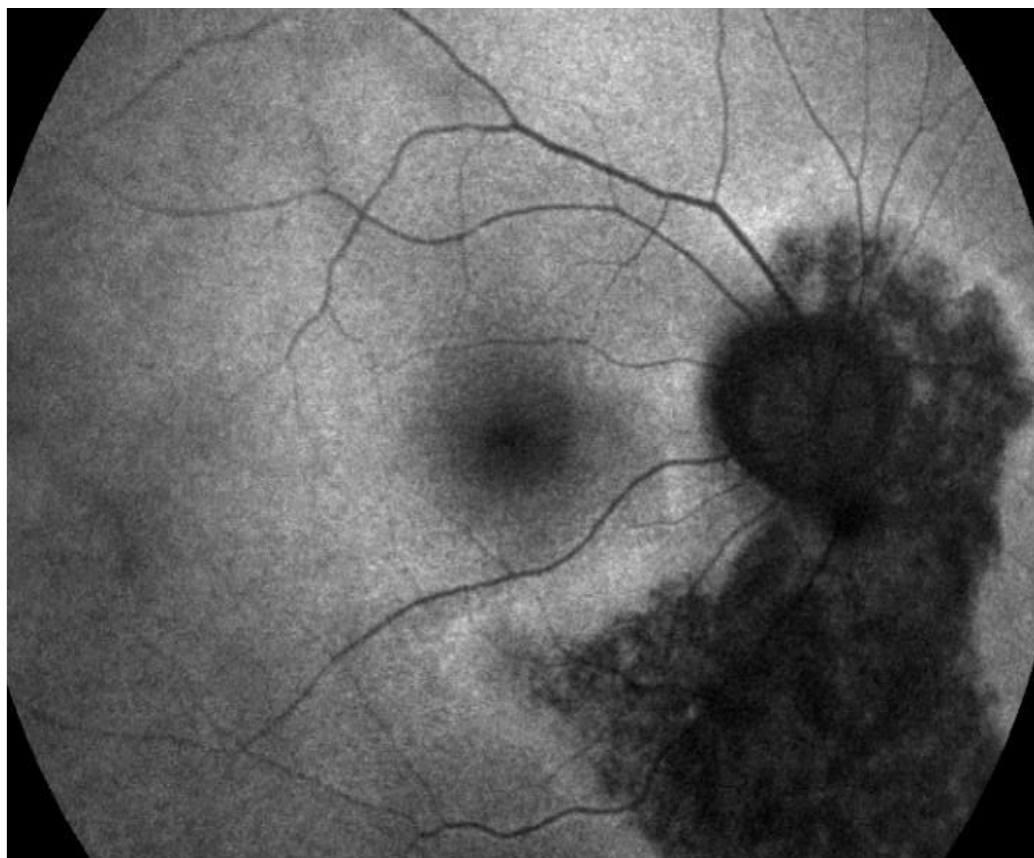
Right eye 2022



Right eye 2023



Right eye 2024



DISCUSSION

Diagnosis of AZOOR is frequently complicated by the unspecific symptoms of the disease and significant number of differential diagnoses. Complete diagnostic work-up is important to rule out other etiologies. Therapeutic decisions obligatorily rely on empiric treatment decisions in combination with frequent follow-up examinations. The natural course of AZOOR is variable. Bilateral involvement is often asymmetrical, and recurrences of inflammation can occur. According to Gass, the majority of cases completely or partially stabilize within 6 months, while a few patients (4%) experience gradual deterioration with the progression of the disease. Patients with AZOOR are most commonly either observed for progression or treated with steroid therapy or other non-steroidal immunomodulatory agents. Less commonly, antivirals, antifungals, and other therapies have also been used with variable success. Several studies suggest that Adalimumab may be beneficial in managing progressive cases of AZOOR. However, only one case report indicates that, after monitoring disease progression and treatment response, adding mycophenolate may be effective in recurrent cases. There are no documented cases of stable AZOOR being maintained solely on mycophenolate monotherapy. Overall, evidence supporting specific treatments for AZOOR remains difficult to interpret given the favourable natural history of the disease as well as a paucity of prospective trials and complete lack of randomized controlled trials.

CONCLUSIONS

This is a typical case of AZOOR, . Fundus autofluorescence shows a characteristic trizonal appearance of peripapillary hypo autofluorescence, followed by a speckled hyper autofluorescence, and finally by a border of bright hypo autofluorescence. suggesting that the Mycophenolate mofetil may benefit and stop the progression of this condition as this has been stable for 2 years. There is only one case report in 2023 which suggests that Mycophenolate mofetil combined with Adalimumab showed improvement and stabilization of the disease in a 4-year follow-up.

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