Transient Occlusion of the Central Retinal Artery in a Patient with Granulomatosis with Polyangiitis

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ABSTRACT

The purpose of this report is to document the occurrence of transient central retinal artery occlusion (CRAO) as a rare and serious ocular complication in a patient with granulomatosis with polyangiitis (GPA), emphasizing the potential vision-threatening nature of this condition. A 49-year-old male with a known diagnosis of GPA presented with transient CRAO. The case was analyzed to understand the ocular manifestations associated with GPA and the challenges in its management. The patient exhibited a transient CRAO, highlighting the potential for severe ocular complications in the context of GPA. Transient CRAO is a rare but serious vision-threatening complication of GPA. This case underscores the importance of early diagnosis and timely management in patients with ocular manifestations of GPA. Financial disclosure: No financial support was received for this case report. None of the authors has any proprietary interests or conflicts of interest related to this submission. It is not simultaneously being considered for publication at any other journal.

KEYWORDS

central retinal artery; granulomatosis with polyangiitis; transient occlusion

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Received: 16 November 2025
Accepted: 13 April 2026
Published online: 26 May 2026

Acta Medica (Hradec Králové) 2026; 69(1): 37–42
https://doi.org/10.14712/18059694.2026.15

INTRODUCTION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a form of vasculitis characterized by granulomatous inflammation, necrosis, and vasculitis primarily affecting small- to medium-sized vessels (1). It typically involves the respiratory tract and kidneys but can affect other organs including the eyes (2). Ocular involvement occurs in approximately 30–50% of patients with GPA, most commonly manifesting as scleritis, episcleritis, or orbital inflammation (3–5). Ocular symptoms are the first signs of GPA in only about 15% of cases (5). However, retinal vascular occlusions, particularly of the central retinal artery (CRA), are exceedingly rare but potentially sight-threatening complications, affecting less than 5% of patients (6).

The CRA includes the primary blood supply to the inner layers of the retina (7, 8). Occlusion of the CRA typically leads to acute, painless monocular vision loss and is considered a medical emergency. In most cases, CRAO is caused by embolic events or thrombosis but can also be related to vasculitic processes, particularly in autoimmune diseases like GPA (7, 8). In the context of GPA, vasculitis of the small vessels may contribute to CRAO (6).

This case report presents a rare manifestation of transient CRAO in a patient with GPA and highlights the challenges in diagnosing and managing ocular complications in patients with systemic vasculitis, and underscores the risk of inflammatory-mediated vascular occlusions in these patients.

CASE PRESENTATION

PATIENT HISTORY

A 49-year-old male with a 2-year history of GPA presented to our department with three episodes of, transient, painless monocular vision loss in his right eye, lasting a few seconds, consistent with amaurosis fugax, followed by complete recovery. There was no history of trauma, flashing lights, or floaters. He had never experienced similar symptoms in the past. His ocular history was insignificant.

At the time of presentation, the patient was hospitalized in the Pulmonology Department due to worsening of his pulmonary involvement with pleuritis. Thoracentesis revealed an exudative pleural effusion with a predominance of polymorphonuclear cells, negative cytology, and negative tuberculosis testing. Strongly positive classic Antineutrophil Cytoplasmic Antibody (c-ANCA) and elevated anti-proteinase 3 antibodies (anti-PR3) levels, in combination with renal involvement manifested by hematuria and proteinuria, as well as the patient's clinical features, were indicative of a flare of GPA. A nephrology consultation was obtained, and the renal findings were considered consistent with active c-ANCA-associated glomerulonephritis. No lung or renal biopsy was performed, as it was not deemed necessary given the characteristic clinical and serological profile strongly suggestive of pulmonary and renal involvement in a GPA flare. The patient had hypothyroidism and was a non-smoker. Lipid profile was within normal limits, and no additional cardiovascular risk factors were identified apart from arterial hypertension.

At initial GPA presentation two years earlier, the patient exhibited lower respiratory tract involvement characterized by pleural effusion, accompanied by migratory arthralgias and arthritis, vasculitic rash, febrile episodes, weight loss, and pericardial effusion with signs of right ventricular pressure overload without wall motion abnormalities; pulmonary embolism was excluded by Computed Tomography Pulmonary Angiography (CTPA). The diagnosis of GPA was established based on positive c-ANCA and elevated anti-PR3 levels in conjunction with characteristic clinical features, which obviated the need for histologic confirmation; thus, biopsy of the lung or other affected organs was not performed. No major relapses were reported before the current flare, and remission was achieved with low-dose glucocorticoids and methotrexate.

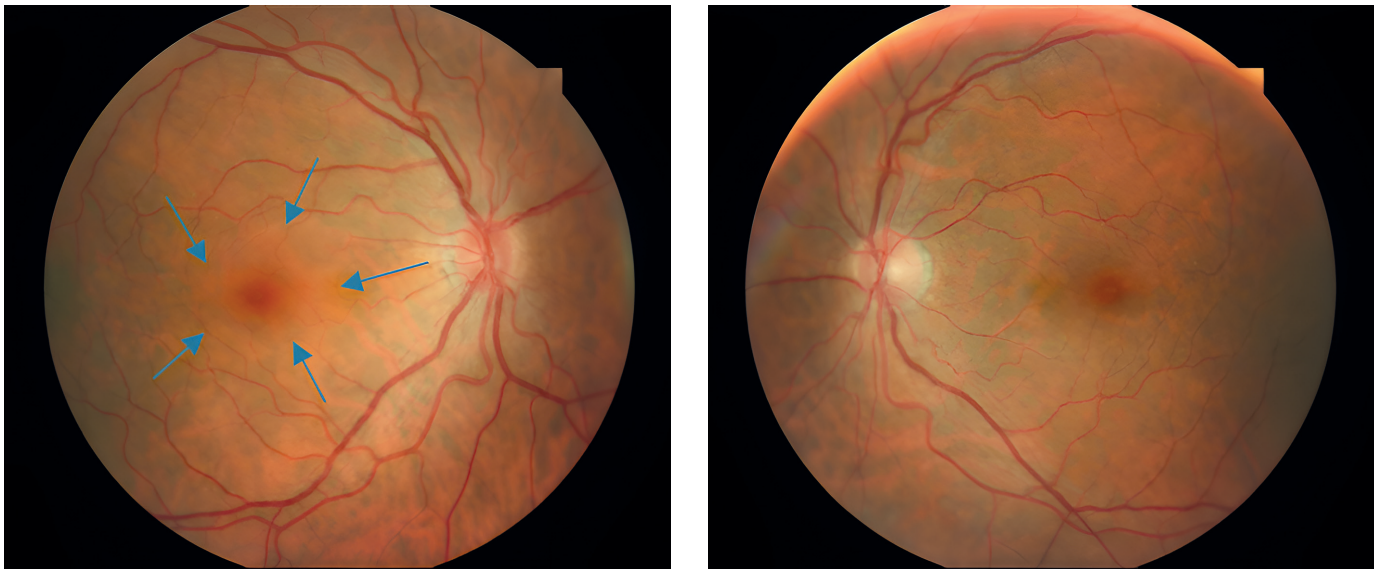

Fig. 1 Mild whitish hue of the macula in the right eye.

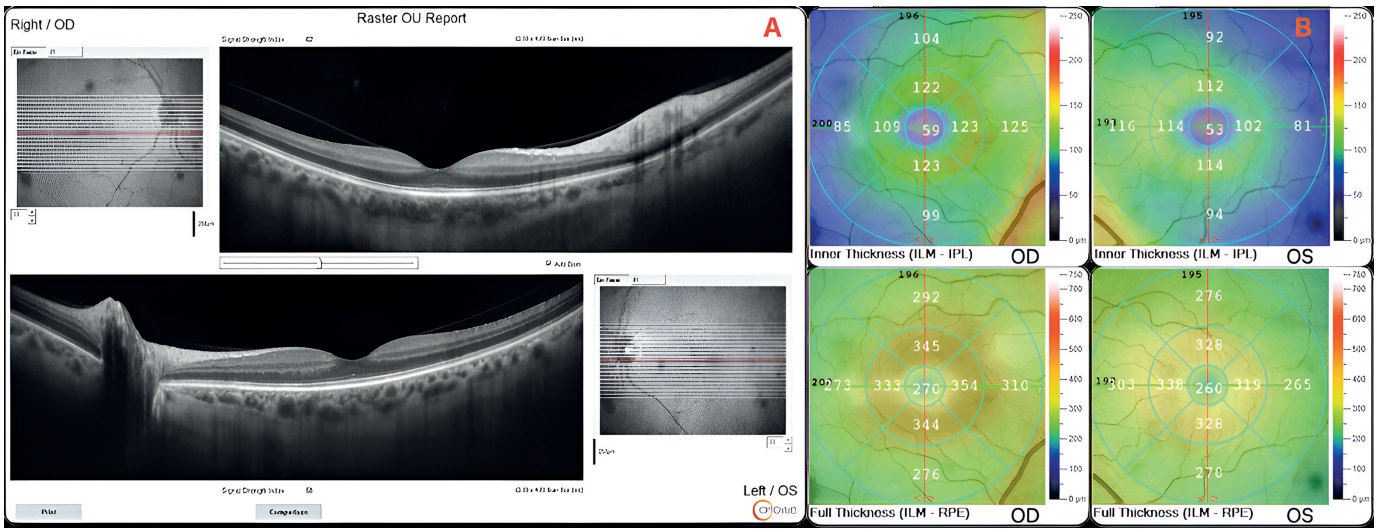

Fig. 2 A) OCT of the patient and B) Full and inner thickness maps showing thickening in the right eye.

PHYSICAL EXAMINATION

On presentation, the patient's vital signs were within normal limits. Best-corrected visual acuity (BCVA) in each eye was 20/20 (logMAR 0.0). Ocular motility was normal, and no RAPD was noted. Visual fields by confrontation were normal in both eyes.

Slit-lamp examination of the anterior segment was unremarkable. Dilated fundus examination of the right eye revealed areas of diffuse retinal ischemia with vascular attenuation and "box-carring" of flow in arteries and veins. The macula and optic disc showed no remarkable pathological signs. Left eye examination was unremarkable.

Shortly before the angiography, the patient experienced another episode of amaurosis fugax, which persisted for more than a minute, longer than previous episodes. Visual acuity in the right eye deteriorated to hand movements. Fundus examination revealed a mild whitish hue, which, in combination with the increased thickness of the inner layers in the macula, is indicative of ischemia due to CRAO (Figures 1 and 2).

DIAGNOSTIC WORKUP

Given the patient's known history of GPA, CRAO secondary to vasculitis was considered. However, other potential causes, such as embolic events, had to be ruled out. The patient underwent immediate investigations, including:

- **Optical Coherence Tomography (OCT):** There were no significant findings on b-scan but thickness maps showed full and inner retinal thickening in the right eye, probably due to ischemia secondary to CRAO (Figure 2).
- **Optical Coherence Tomography – Angiography (OCT-A):** OCT-A and enface OCT showed areas of non-perfusion in the superficial and deep capillary

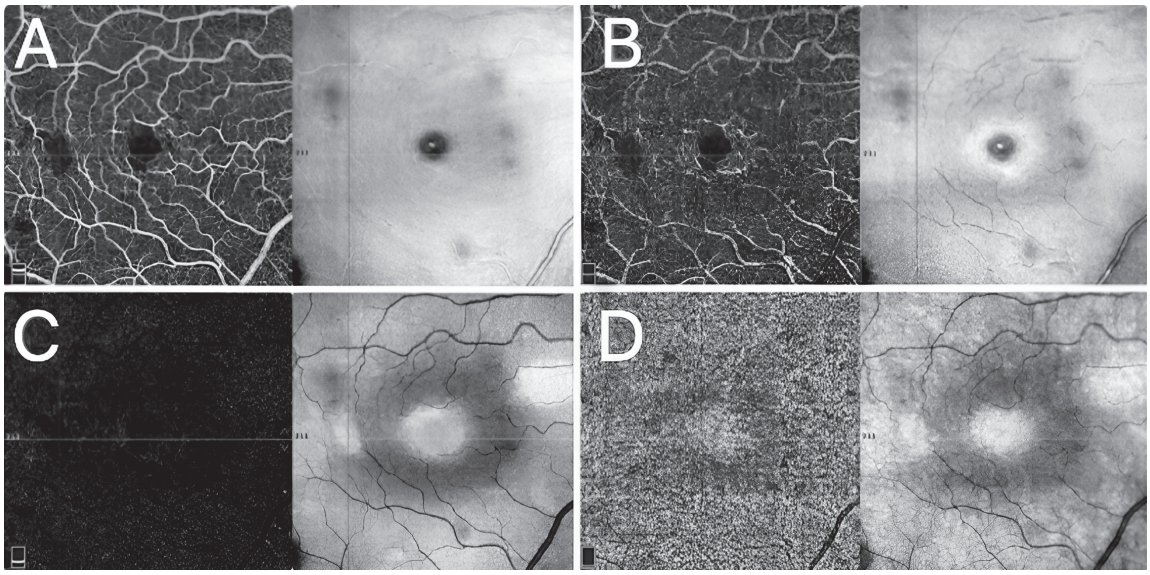

Fig. 3 OCT-A findings. A) Superficial and B) Deep capillary plexus abnormalities. C) Outer retina and D) choriocapillaris layers shadowing.

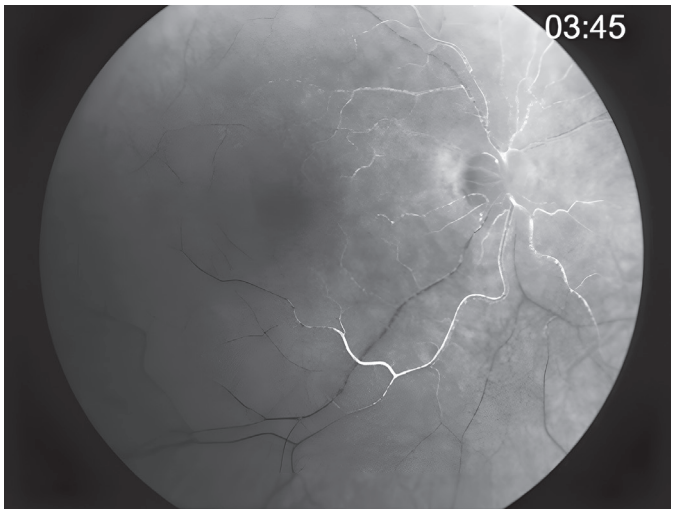

Fig. 4 CRAO signs in FA.

plexus (Figures 3A and 3B). Furthermore, shadowing to the outer retina and choriocapillaris layers due to edema in the inner retinal layers was revealed (Figures 3C and 3D).

- **Fluorescein Angiography (FA):** FA showed delayed filling of the central retinal artery, with normal choroidal circulation (Figure 4). No evidence of vascular leakage or emboli was observed, further supporting the diagnosis of CRAO due to vasculitis rather than an embolic event.
- **Complete Blood Count (CBC) and Erythrocyte Sedimentation Rate (ESR):** CBC was within normal limits, but ESR was elevated at 88 mm/hr, suggesting active inflammation.
- **C-reactive Protein (CRP):** CRP was also elevated, consistent with systemic inflammation (24 mg/L).
- **Serum Creatinine and Urinalysis:** Serum creatinine was elevated at 1.3 mg/dL, and urinalysis showed hematuria (10–20 red blood cells per high power field) and proteinuria (1.2 g/24h), raising concerns about renal involvement in the context of a GPA flare.
- **Serological Testing:** The patient was positive for c-ANCA and anti-PR3 with a high titer, which is characteristic of GPA. This finding, in combination with elevated inflammatory markers, suggested a GPA flare. A longitudinal representation of c-ANCA titers demonstrated elevated levels (1:160) at initial diagnosis, followed by a period of low titers after initiation of immunosuppressive therapy, consistent with disease remission. A subsequent rise in c-ANCA levels was observed at the time of clinical exacerbation (1:80) (Figure 5), supporting the association between systemic disease activity and retinal vascular occlusion. Anti-PR3 levels followed a similar trend, with elevated values at diagnosis (216 IU/mL) and relapse (244 IU/mL).

DIAGNOSIS

The patient was diagnosed with transient central retinal artery occlusion secondary to a flare of GPA-related vasculitis. The diagnosis was based on clinical presentation, history of GPA, elevated inflammatory markers and the absence of emboli on fluorescein angiography.

MANAGEMENT

Given the acute nature of CRAO and the risk of permanent vision loss, immediate treatment was initiated. The patient was admitted for aggressive immunosuppressive therapy to control the GPA flare and prevent further vascular complications.

First Line Therapy

- **Anticoagulation and Vasodilators:** Although embolic causes were ruled out, low-dose aspirin was prescribed as a preventive measure. Topical vasodilators and ocular massage were attempted to improve retinal

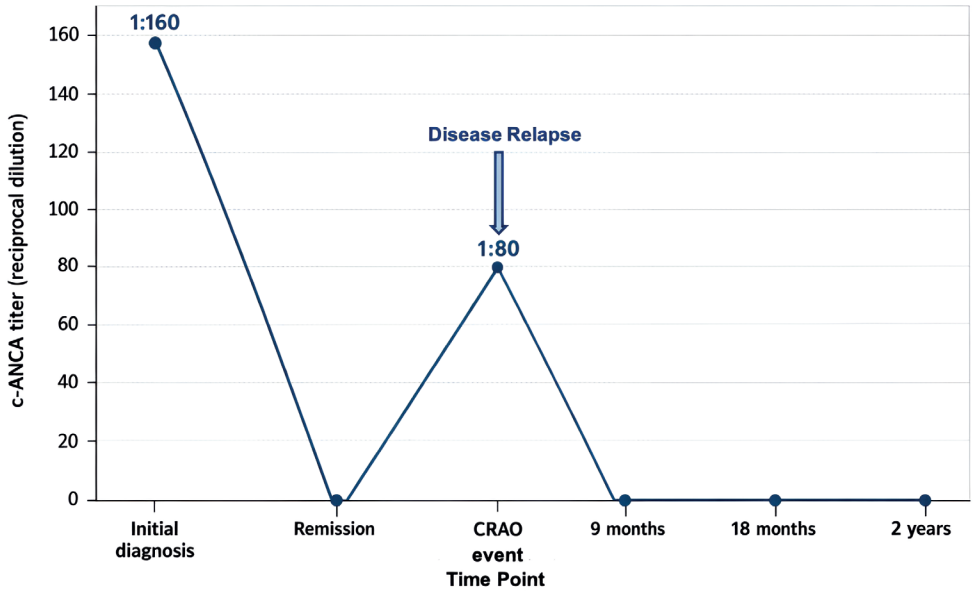

Fig. 5 Longitudinal changes in c-ANCA titers. c-ANCA titers decreased from 1:160 at initial diagnosis to undetectable levels during remission, followed by a rise to 1:80 at the time of disease relapse associated with CRAO. Subsequent measurements remained undetectable throughout follow-up. Titers are expressed as reciprocal dilution values.

perfusion, although these measures have limited efficacy in CRAO (8).

Systematic Therapy

- **High-Dose Glucocorticoids:** The patient was started on intravenous methylprednisolone (1 g/day) for three days, followed by oral prednisone and taper off.
- **Immunosuppressive Therapy:** Methotrexate was discontinued, and rituximab was introduced as a more effective long-term therapy for GPA management.

OUTCOME AND FOLLOW-UP

Within 30 minutes of the aforementioned treatment (anticoagulation and vasodilators), the patient reported full recovery of vision in his right eye.

Within 24 hours of initiating treatment, BCVA remained stable at 20/20 (logMAR 0.0), and fundus examination revealed regression of CRAO signs. No further amaurosis fugax episodes were reported.

After one week of corticosteroid therapy, the patient's BCVA remained stable at 20/20 (logMAR 0.0). The patient was discharged on a maintenance dose of oral prednisone and rituximab infusions every six months.

At the three-month follow-up, no further evidence of vasculitis was noted, and ANCA levels had decreased. Repeat OCT showed normal thickness of the inner retina. (Figure 6). FA showed normal filling of the retinal vessels. Fundus examination was unremarkable.

At the two-year follow-up, the patient remained clinically stable with no signs of recurrent vasculitis. c-ANCA levels were negative (Figure 5), and no further episodes of amaurosis had been reported.

DISCUSSION

This case highlights a rare but serious ocular complication of GPA, transient CRAO. Retinal artery occlusions in the context of GPA are exceedingly rare and are typically associated with active vasculitis, as demonstrated in this case report. Early recognition and aggressive immunosuppressive treatment are crucial for avoiding vascular occlusive events.

Ocular involvement in GPA most commonly manifests as scleritis or orbital inflammation, but retinal vascular occlusion can occur in severe cases of vasculitis (3–5). The pathophysiology involves immune-mediated inflammation of small blood vessels, leading to vessel wall damage, inflammatory thrombus formation, and subsequent ischemia. In this case, the patient's elevated inflammatory markers and positive c-ANCA and anti-PR3 titers suggested a systemic vasculitis flare, which likely contributed to the transient CRAO (9, 10). Indeed, despite initial treatment with methotrexate, the patient experienced disease relapse, highlighting that this agent may be insufficient to maintain remission in certain cases of granulomatosis with polyangiitis. Consequently, therapy was escalated to

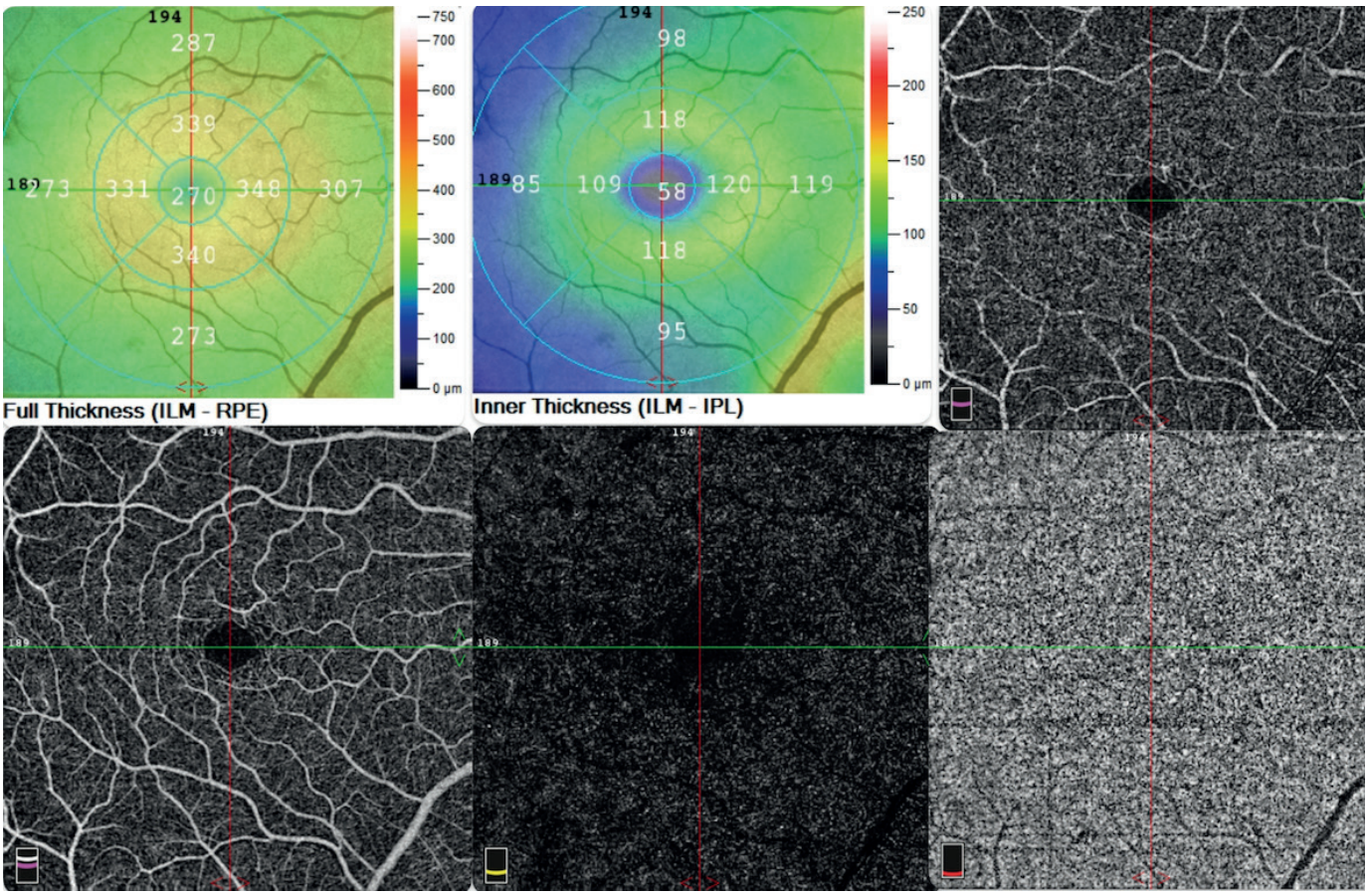

Fig. 6 Thickness maps and OCT-A at three-month follow-up.

rituximab, reflecting the need for more potent immunosuppression in the setting of disease reactivation.

In the present case, no histopathological confirmation was obtained, as biopsy was not deemed necessary at either the time of initial diagnosis or during disease relapse. This decision was based on the presence of a highly characteristic clinical presentation in combination with supportive laboratory findings, including positive c-ANCA and anti-PR3 serology, which together provided sufficient diagnostic confidence for granulomatosis with polyangiitis.

A thorough literature review revealed that cases of CRAO associated with GPA are extremely rare in the English literature. The limited number of published cases suggests that CRAO typically occurs in the setting of active, systemic disease, often accompanied by other manifestations such as renal, pulmonary, or ear, nose, and throat (ENT) involvement (11). In comparison to the existing literature, our case shares several overlapping features, including the presence of active vasculitic disease and elevated c-ANCA titers at the time of ocular presentation.

Furthermore, previously reported cases emphasize the importance of early immunosuppressive treatment, as visual prognosis following CRAO is generally poor despite prompt intervention (11). In contrast to these reports, in our case, immediate management was associated with a transient occlusive event, without recurrence during follow-up. This observation reinforces the importance of heightened clinical suspicion and early intervention when patients with known or suspected GPA present with acute visual symptoms (9).

To our knowledge, this is the first case report that includes a comprehensive diagnostic workup, including FA, OCT and OCT-A findings, and demonstrates full vision recovery (9, 12).

STATEMENT OF ETHICS

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patients for publication of this case report and any accompanying images.

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