**Scholars Journal of Medical Case Reports**
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

**Ophthalmology**

# Microscopic Polyangiitis Revealed by Central Retinal Vein Occlusion: Case Report

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**DOI:** https://doi.org/10.36347/sjmcr.2026.v14i04.027 | **Received:** 11.01.2026 | **Accepted:** 21.02.2026 | **Published:** 16.04.2026

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**Abstract** **Case Report**

Microscopic polyangiitis is a necrotizing vasculitis associated with antineutrophil cytoplasmic antibodies (ANCA) that primarily affects small-caliber vessels. We report the case of a 52-year-old woman in whom microscopic polyangiitis was revealed by central retinal vein occlusion. The clinical outcome under corticosteroid therapy combined with cyclophosphamide was favorable, with improvement in both renal and ocular manifestations.

**Keywords:** Microscopic Polyangiitis, Central Retinal Vein Occlusion, p-ANCA, Anti-Myeloperoxidase Antibodies.



## INTRODUCTION

Microscopic polyangiitis (MPA) is a systemic necrotizing small-vessel vasculitis belonging to the group of ANCA-associated vasculitides. It most commonly presents with a pulmonary–renal syndrome and only rarely with ocular manifestations.

We report the case of a patient in whom MPA was revealed by central retinal vein occlusion.

## CASE PRESENTATION

Mrs. L.M., a 52-year-old woman, was admitted to the ophthalmology emergency department for sudden visual loss in the right eye. She had no significant past medical history.

Ophthalmologic examination of the right eye revealed visual acuity limited to hand motion, a normal anterior segment, normal intraocular pressure, and fundus findings consistent with stage II optic disc edema associated with peripapillary flame-shaped hemorrhages and venous tortuosity (Figure 1), along with a poor macular reflex. Examination of the contralateral eye was unremarkable. General physical examination was normal.

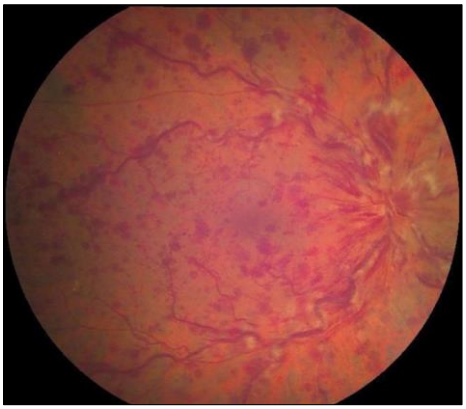

**Figure 1: Stage II papilledema with peripapillary flame-shaped hemorrhages and venous tortuosity**

Macular optical coherence tomography (OCT) demonstrated the presence of subretinal fluid (Figure2). Papillary OCT confirmed stage II optic disc edema (Figure 3). Fluorescein angiography revealed an edematous central retinal vein occlusion characterized by marked venous dilation and tortuosity, without areas of retinal ischemia (Figure 4).

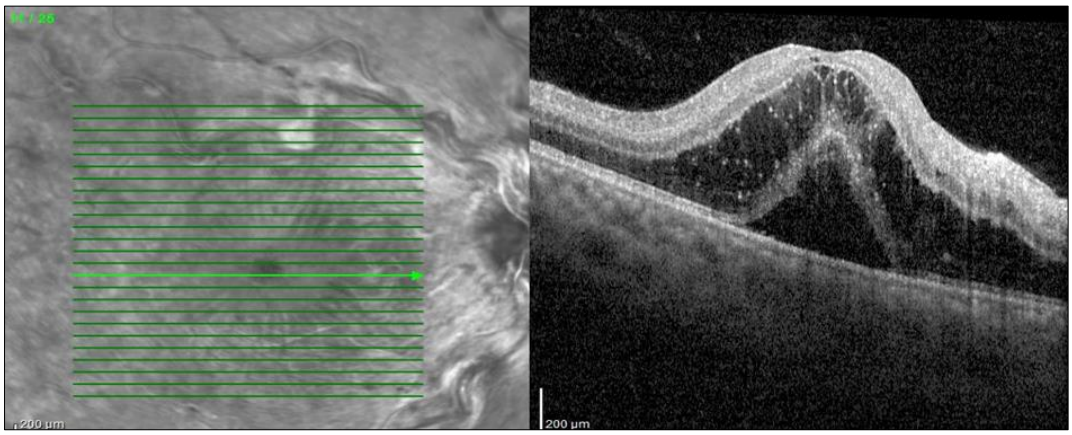


**Figure 2: Macular optical coherence tomography (OCT) showing the presence of subretinal fluid**

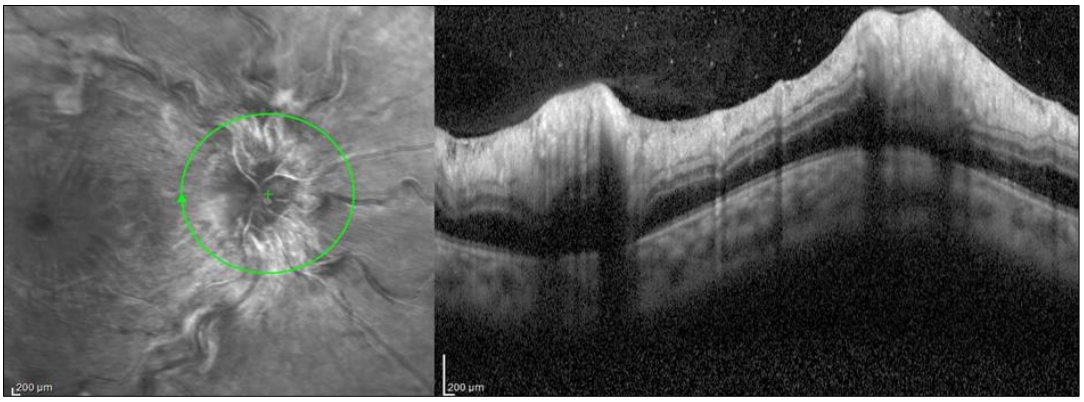


**Figure 3: Optic nerve head OCT demonstrating stage II papilledema**

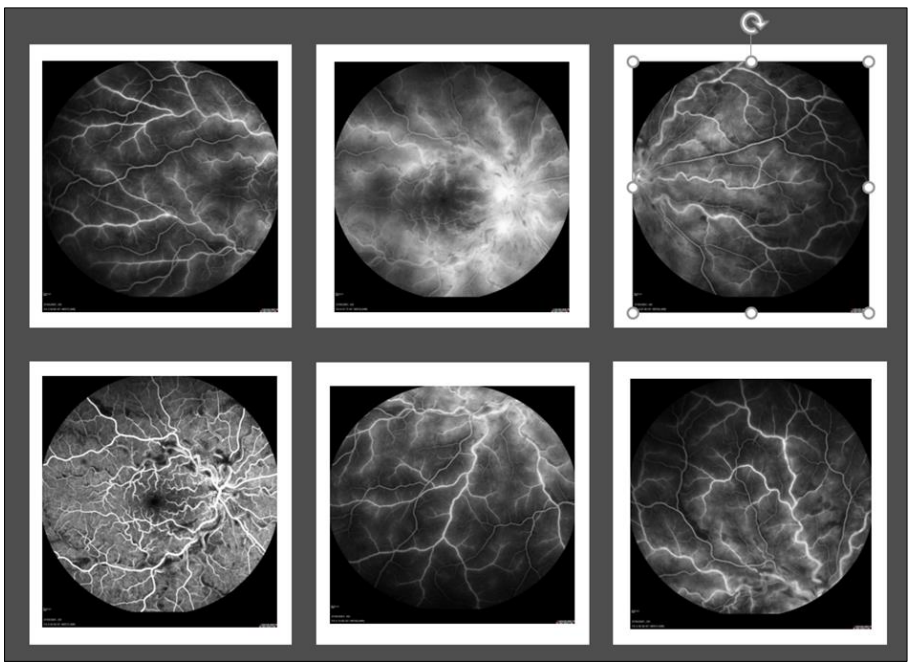

**Figure 4: Fluorescein angiography showing edematous central retinal vein occlusion with marked venous dilation and tortuosity, without areas of ischemia**

The etiological workup revealed no cardiovascular abnormalities, including 24-hour blood pressure monitoring, electrocardiography, transthoracic echocardiography, and supra-aortic trunk ultrasonography. The coagulation profile was also within normal limits (fibrinogen, factor V, proteins C and S, antithrombin III, factor XII, homocysteine levels, and antiphospholipid antibodies).

Laboratory investigations demonstrated normocytic normochromic anemia (hemoglobin 10.5 g/dL), a marked inflammatory syndrome (C-reactive protein 50 mg/L; erythrocyte sedimentation rate 60 mm at 1 hour), and elevated serum creatinine (20 mg/L). Immunological testing revealed p-ANCA with anti-myeloperoxidase specificity (MPO = 50), while antinuclear antibodies and anti–double-stranded DNA antibodies were negative.

Given the elevated creatinine level, a nephrology consultation was obtained. Further investigations supported the diagnosis of nephrotic syndrome (proteinuria 3 g/24 h, serum albumin 28 g/L, total protein 59 g/L). Renal biopsy showed focal and segmental necrotizing glomerulonephritis with extracapillary crescents involving approximately 55% of the glomeruli, along with fibrinoid necrosis of the glomerular capillaries. The tubulointerstitial compartment demonstrated moderate lymphoplasmacytic inflammatory infiltrates associated with 25% interstitial fibrosis and tubular atrophy. Direct immunofluorescence did not reveal significant immune deposits.

The final diagnosis was microscopic polyangiitis with renal and ocular involvement.

Induction therapy consisted of intravenous methylprednisolone pulses (15 mg/kg/day for 3 days), followed by oral corticosteroids (1 mg/kg/day) and monthly cyclophosphamide pulses for six months. Maintenance therapy was initiated with azathioprine (2 mg/kg/day).

The patient also received three monthly intravitreal anti-VEGF (ranibizumab) injections.

Serum creatinine levels normalized after 10 days of treatment, and the nephrotic syndrome resolved by the sixth week. Ophthalmologically, subretinal fluid resolved after the third intravitreal injection (Figure 5), and visual acuity improved from hand motion to counting fingers at 5 meters.

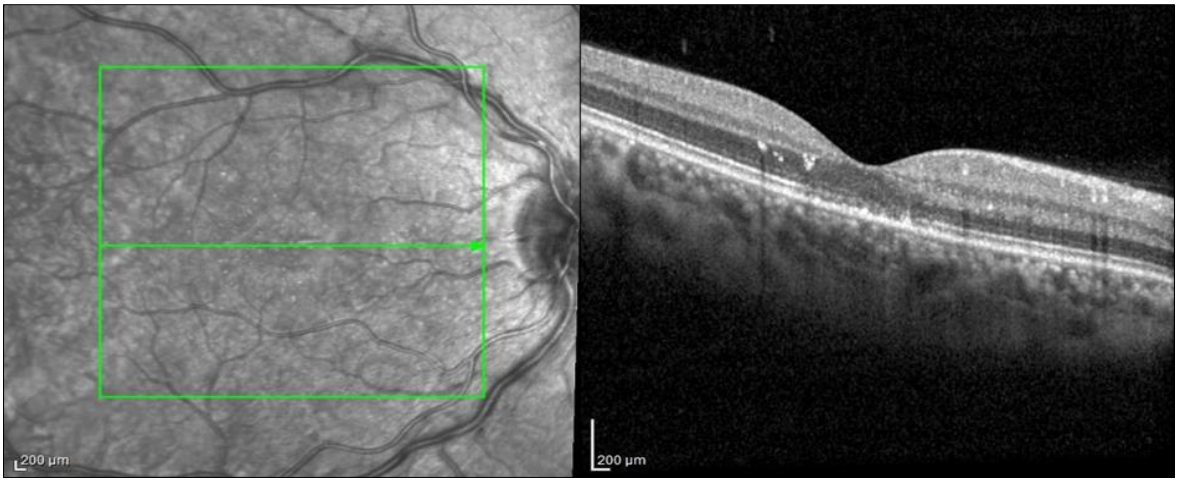


**Figure 5: Resolution of subretinal fluid**

## DISCUSSION

Microscopic polyangiitis (MPA) is a necrotizing systemic vasculitis affecting small vessels and is associated with antineutrophil cytoplasmic antibodies (ANCA) in approximately 75% of cases. These are predominantly anti–myeloperoxidase (MPO) antibodies (58%), although anti–proteinase 3 (PR3) ANCA may be detected in approximately 26% of patients [1]. In our patient, p-ANCA with anti-MPO specificity was positive.

MPA is a rare disease that can affect individuals of all ethnic backgrounds, although published series report that 85–100% of patients are Caucasian [2]. It is distributed worldwide, but its geographic distribution is not uniform. In Europe, a north–south gradient has been described, with a higher frequency reported in southern countries compared to northern regions [3, 4]. The disease most commonly affects individuals over 50 years of age. In France, the prevalence of MPA has been estimated at 25 cases per million inhabitants in the Seine-Saint-Denis district [5].

MPA belongs to the group of ANCA-associated vasculitides and is characterized in 75–80% of cases by circulating ANCA, most frequently of the perinuclear type (p-ANCA). On indirect immunofluorescence, p-ANCA produces a perinuclear staining pattern of ethanol-fixed neutrophils. Antigenically, these

antibodies are most often directed against myeloperoxidase (anti-MPO), identified by ELISA techniques [2].

Anti-MPO and anti-PR3 antibodies are capable of activating primed neutrophils, particularly in the presence of pro-inflammatory cytokines such as tumor necrosis factor-alpha (TNF-α) and interleukin-8 (IL-8). This activation leads to the generation of reactive oxygen species and the release of cytotoxic enzymes, contributing to vascular injury. The central pathogenic role of neutrophils has been demonstrated in vivo, notably in the experimental model described by Xiao *et al*., [6].

In addition to neutrophil activation, anti-MPO antibodies may directly interact with their target antigen. Myeloperoxidase is a 118 kDa cationic enzyme that generates oxidant species, particularly hypochlorous acid (HOCl), in the presence of hydrogen peroxide and chloride ions. Increased production of these oxidants under the influence of anti-MPO antibodies may represent an additional cytotoxic mechanism, independent of cellular activation, contributing to endothelial damage [7].

Furthermore, autoantibodies directed against endothelial cells—whose precise antigenic targets remain incompletely characterized—have been identified in patients with MPA [8]. Their serum levels appear to correlate with disease activity [9]. These antibodies may promote leukocyte adhesion to the endothelium, thereby amplifying vascular inflammation.

Retrospective analyses of European cohorts have shown that most patients with MPA present with renal involvement. Alveolar hemorrhage occurs in up to 30% of cases, musculoskeletal manifestations in approximately two-thirds, cutaneous lesions in about half, gastrointestinal involvement in 30–50%, and peripheral neuropathy in up to one-third of patients [10].

Ocular manifestations are uncommon but may include episcleritis, scleritis, iridocyclitis, and choroidal or retinal vasculitis [11]. Recent reports have described hypopyon iridocyclitis, retinal cotton-wool spots, conjunctival involvement, and optic neuropathy [12, 13]. Rarely, ocular involvement may precede systemic manifestations. Central retinal vein occlusion (CRVO) as the presenting feature of MPA is exceptionally rare.

As in other systemic vasculitides, a nonspecific inflammatory syndrome is usually present. Significant anemia may be observed, either inflammatory in origin or secondary to alveolar hemorrhage. In patients with glomerulonephritis, renal function impairment is common and is frequently preceded or accompanied by microscopic hematuria and proteinuria. p-ANCA are detected in 75–80% of cases, predominantly with anti-MPO specificity [10-16]. Double positivity (anti-PR3 and anti-MPO) or isolated c-ANCA positivity is uncommon. Renal biopsy is almost indispensable for diagnosis, particularly when it demonstrates necrotizing crescentic glomerulonephritis. It also provides important prognostic information. Involvement of more than 60% of glomeruli is associated with a poorer renal prognosis, as are marked interstitial inflammation, tubular damage, and glomerular sclerosis.

The natural course of untreated MPA is severe, with mortality rates approaching 90% at one year. However, the introduction of corticosteroids and immunosuppressive therapy has dramatically improved outcomes [14]. Mortality predominantly occurs in patients with severe disease, particularly those presenting with poor prognostic factors according to the Five Factor Score (FFS) [15], or with alveolar hemorrhage [16]. Although MPA is generally considered a severe disease, milder forms have been reported [10].

Management of ANCA-associated vasculitis includes high-dose corticosteroids combined with immunosuppressive therapy according to disease severity, assessed using the Five Factor Score (FFS) [17, 18]. When the FFS is ≥1, immunosuppressive treatment is recommended.

Cyclophosphamide or rituximab is considered first-line induction therapy. The RAVE and RITUXVAS trials demonstrated that rituximab is not inferior to cyclophosphamide [19-21]. In cases of relapse, rituximab appears to achieve higher remission rates (67% vs. 42% in the study by Stone *et al*.,) [19]. Plasma exchange is reserved for severe cases, particularly those with alveolar hemorrhage or persistent deterioration of renal function despite appropriate immunosuppressive therapy [15].

The MAINRITSAN and RITAZAREM trials demonstrated the superiority of rituximab over azathioprine for maintenance therapy in ANCA-associated vasculitis [20, 21]. Additionally, the WEGENT trial showed that methotrexate is an alternative to azathioprine when renal function permits [22]. In our patient, azathioprine was chosen for maintenance therapy due to its availability in our country. The patient's FFS was 1, owing to renal impairment, which justified induction therapy with cyclophosphamide combined with corticosteroids, followed by maintenance therapy with azathioprine.

## CONCLUSION

Microscopic polyangiitis is a potentially life-threatening ANCA-associated small-vessel vasculitis. It most commonly presents with renal and pulmonary involvement, whereas ocular manifestations are rare. Central retinal vein occlusion as the initial presentation is exceptional and may delay diagnosis if a comprehensive systemic evaluation is not performed.

This case highlights the importance of considering systemic vasculitis in cases of atypical or unexplained retinal vascular occlusion, particularly when associated with inflammatory or renal abnormalities. Early diagnosis, prompt initiation of immunosuppressive therapy, and multidisciplinary management are essential to improve both renal and visual outcomes.

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