EJCRIM
European Journal of Case Reports in Internal Medicine

SUDDEN BILATERAL BLINDNESS REVEALING MPO-ANCA-ASSOCIATED HYPERTROPHIC PACHYMENINGITIS IN AN 82-YEAR-OLD WOMAN

Daniela Duque Montoya[1], Luis Felipe Aguirre García[2,3], Angélica María Vallejo Portilla[3], Camila García Ávila[4], José Fernando Gómez[2]

[1] Mental Health Department, Faculty of Health Sciences, Universidad de Caldas, Manizales, Colombia
[2] Research Group on Geriatrics and Gerontology, Faculty of Health Sciences, Universidad de Caldas, Manizales, Colombia
[3] Grupo de Resistencia Antibiótica de Manizales (GRAM), Manizales, Colombia
[4] Internal Medicine Department, Faculty of Health Sciences, Universidad de Caldas, Manizales, Colombia

Corresponding author's e-mail: aguirregarcialuis@gmail.com

Received: 07/08/2026
Accepted: 12/08/2026
Published: 21/10/2026

Conflicts of Interests: The Authors declare no conflict of interest.
Patient Consent: Written informed consent for publication of the clinical history and the anonymised imaging was obtained from the patient.
Declaration of generative AI and AI-assisted technologies in the manuscript preparation process: A large language model was used to assist with language editing and manuscript formatting. All content was reviewed and verified by the authors, who take full responsibility for the accuracy and integrity of the manuscript.
This article is licensed under a CC Attribution-NonCommercial-NoDerivatives 4.0 License

How to cite this article: Duque Montoya D, Aguirre García LF, Vallejo Portilla AM, García Ávila C, Gómez JF. Sudden bilateral blindness revealing MPO-ANCA-associated hypertrophic pachymeningitis in an 82-year-old woman. EJCRIM 2026;13:doi:10.12890/2026_007403

ABSTRACT

Introduction: Hypertrophic pachymeningitis is a rare inflammatory thickening of the dura mater, complicating an estimated 1–3% of ANCA-associated vasculitis (AAV), usually PR3-ANCA disease with systemic involvement. Isolated meningeal disease with normal inflammatory markers is exceptional and easily mistaken for a non-inflammatory or ischaemic optic neuropathy.

Case description: An independent 82-year-old woman presented with 48 hours of bilateral visual loss after nine days of severe, non-postural occipital headache. She had no light perception, non-reactive pupils and optic disc pallor, without renal, pulmonary, cutaneous or sinonasal manifestations. Chest computed tomography, erythrocyte sedimentation rate and creatinine were normal. Contrast-enhanced magnetic resonance imaging showed diffuse pachymeningeal thickening with intense dural enhancement. Cerebrospinal fluid was acellular, with elevated protein and negative microbiology, including two *Mycobacterium tuberculosis* PCR assays. Anti-MPO exceeded 400 RU/ml, with negative anti-PR3, low C3 and C4, and normal IgG4, consistent with microscopic polyangiitis. Intravenous methylprednisolone 500 mg/day, started four days after blindness onset, was followed by recovery to counting fingers at 30 cm in the right eye by day 9.

Discussion: MPO specificity, isolated blindness and an absent acute-phase response made this presentation atypical. Visual loss probably reflected dural compression and hypoperfusion of the optic nerves rather than infarction, possibly explaining partial recovery. An ejection fraction of 30% favoured rituximab over cyclophosphamide.

Conclusion: MPO-ANCA-associated vasculitis may be confined to the dura, with normal acute-phase reactants. ANCA testing should accompany rather than follow the work-up for dural enhancement, and glucocorticoids should not await the result: functional reserve, not age, determines treatment intensity.

KEYWORDS

Vasculitis, central nervous system, ANCA-associated vasculitis, myeloperoxidase, bilateral blindness, glucocorticoids

EFIM
European Federation of Internal Medicine

Publisher
smc media srl

LEARNING POINTS

- MPO-ANCA-associated vasculitis may present with complete bilateral blindness as the sole initial manifestation, even in the absence of renal, pulmonary, cutaneous or sinonasal involvement. Normal acute-phase reactants, including a normal erythrocyte sedimentation rate, and an otherwise unrevealing systemic evaluation do not exclude the diagnosis.
- Diffuse dural enhancement is non-specific; therefore, ANCA testing with anti-MPO and anti-PR3 assays should be performed alongside the evaluation for infectious, IgG4-related, neoplastic and intracranial hypotension–related causes. Hypertrophic pachymeningitis remains a diagnosis of exclusion.
- Suspected inflammatory pachymeningitis with visual loss should be treated as an ophthalmological emergency, as in giant cell arteritis; frailty and functional reserve, not chronological age, should determine treatment intensity, even when comorbidity constrains the choice of immunosuppressant.

INTRODUCTION

The spectrum of ANCA-associated vasculitis (AAV) encompasses granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis[1]. Hypertrophic pachymeningitis (HP), defined as focal or diffuse inflammatory thickening of the dura mater on contrast-enhanced magnetic resonance imaging, is among its rarest manifestations, affecting an estimated 1–3% of patients. It may also result from IgG4-related disease, sarcoidosis, tuberculosis, neoplastic infiltration or spontaneous intracranial hypotension[2,3]. Most reported AAV-related HP occurs in PR3-ANCA GPA, while MPO-ANCA cases remain a minority described mainly in East Asian cohorts[4-6].

Neurological involvement affects 20–50% of patients with AAV, usually as peripheral neuropathy; central nervous system disease is uncommon, and when HP occurs the picture is dominated by a headache and cranial neuropathy. Complete bilateral blindness as the initial manifestation of AAV, in the absence of systemic vasculitic features and despite normal acute-phase reactants, is exceptionally rare and may be misdiagnosed as a non-inflammatory optic neuropathy. In this case, corticosteroid pulse therapy, initiated four days after the onset of blindness, was followed by partial visual recovery.

CASE DESCRIPTION

An 82-year-old woman with hypertension, prediabetes and no autoimmune history presented with 48 hours of bilateral visual loss. Nine days earlier, she had developed a severe non-postural occipital headache. She was fully independent and not frail (Clinical Frailty Scale 2/9).

She was afebrile, with a blood pressure of 150/72 mmHg. There was no purpura, arthritis, sinonasal symptoms, haemoptysis or peripheral neuropathy. Neurological examination was normal except for the visual system: no light perception bilaterally, anisocoric non-reactive pupils and bilateral optic disc pallor, with full extraocular movements and no meningeal signs.

Acute-phase reactants and renal function were normal, arguing against systemic vasculitis *(Table 1)*. Non-contrast brain computed tomography (CT) showed only involutional changes, but contrast-enhanced magnetic resonance imaging on day 2 demonstrated diffuse pachymeningeal thickening with intense homogeneous dural enhancement of the falx and tentorium, without parenchymal lesions, brain sagging or subdural collections *(Fig. 1)*.

Cerebrospinal fluid (CSF) was acellular, with raised protein and negative microbiology, including *Mycobacterium tuberculosis* polymerase chain reaction (PCR); repeat puncture on day 13 confirmed this pattern. Chest CT excluded pulmonary and mediastinal disease, but suggested dilated cardiomyopathy, confirmed by echocardiography with a left ventricular ejection fraction of 30%. Urinalysis showed microscopic haematuria without

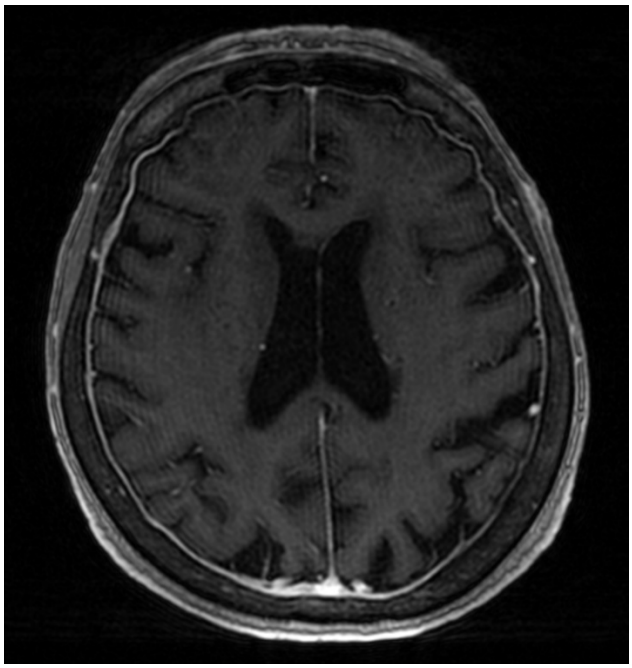


Figure 1. *Gadolinium-enhanced brain MRI, post-contrast T1-weighted axial image, showing diffuse pachymeningeal thickening with intense, homogeneous dural enhancement involving the falx cerebri and the tentorium cerebelli, consistent with hypertrophic pachymeningitis. There is no parenchymal lesion, hydrocephalus, brain sagging or subdural collection. Background global cortical atrophy and Fazekas grade 2 periventricular white matter changes are also present.*

Investigation	Result	Reference range
Immunology		
Anti-MPO (RU/ml)	>400	<20
Anti-PR3 (RU/ml)	<2	<20
p-ANCA (indirect immunofluorescence)	Positive, 1:20	Negative
c-ANCA (indirect immunofluorescence)	Negative	Negative
Complement C3 (mg/dl)	67.3 (low)	90–170
Complement C4 (mg/dl)	13.7 (low)	17.4–52.2
Serum IgG4 (mg/dl)	17.1 (normal)	3.92–86.4
Antinuclear antibodies (HEp-2)	Reactive, speckled 1:320	Negative
Anti-dsDNA	Non-reactive	Negative
Extractable nuclear antigens, total (RU/ml)	23.18 (weakly positive)	<20
Anti-RNP, anti-Sm, anti-SSA/Ro, anti-SSB/La	All negative	Negative
Rheumatoid factor (IU/ml)	153.1	<20
Anti-CCP (RU/ml)	2.1	<20
Cerebrospinal fluid (two lumbar punctures, days 2 and 13)		
Protein (mg/dl)	Raised; 46.1 on repeat	15–45
Leucocytes (/mm^3)	0	0–5
Glucose (mg/dl)	54	45–80
Gram stain, fungal preparations, cultures	Negative	Negative
Mycobacterium tuberculosis PCR	Not detected (×2)	Not detected
VDRL	Non-reactive	Non-reactive
Systemic screen (arguing against, but not excluding, vasculitis)		
Erythrocyte sedimentation rate (mm/h)	20	0–20
Leucocytes (/mm^3)	5,550	4,000–10,000
Haemoglobin (g/dl)	12.8	12–16
Creatinine (mg/dl)	0.81	0.50–1.10
Urinalysis	>40 red cells/high-power field, no proteinuria	Normal
N-terminal pro-B-type natriuretic peptide (pg/ml)	1,101	<125
Chest computed tomography	No infiltrate, granuloma or adenopathy	Normal

Table 1. *Key investigations.*
Abbreviations: Anti-CCP, anti-cyclic citrullinated peptide; ANCA, antineutrophil cytoplasmic antibodies; MPO, myeloperoxidase; PCR, polymerase chain reaction; PR3, proteinase 3; VDRL, Venereal Disease Research Laboratory.

proteinuria. Inflammatory pachymeningitis was considered most likely. After baseline immunological sampling, intravenous methylprednisolone 500 mg/day was started on day 3, four days after blindness onset, and continued for five days. Subsequent serology showed perinuclear ANCA at 1:20, anti-MPO above 400 RU/ml (reference <20), undetectable anti-PR3, low C3 and C4, and normal serum IgG4 *(Table 1)*.

The erythrocyte sedimentation rate was also normal by the age-adjusted upper limit for women (age + 10)/2 = 46 mm/h.

Vision improved from the fourth treatment day. By day 9, the right eye had recovered from no light perception to counting fingers at 30 cm and the left to light perception, with residual dyschromatopsia. She was discharged on day 15 with MPO-ANCA-associated vasculitis consistent with MPA presenting as isolated HP, for rituximab induction.

DISCUSSION

Hypertrophic pachymeningitis is a recognised manifestation of AAV, but the reported phenotype is narrow: predominantly PR3-ANCA GPA, presenting with headache and cranial neuropathy, and accompanied by renal, pulmonary or upper airway disease pointing towards the underlying vasculitis[4-6]. This case differed on every axis. The specificity was MPO; the presenting deficit was complete bilateral blindness rather than a headache; there was no extradural organ involvement other than microscopic haematuria to raise suspicion. Most misleading was the absent acute-phase response. A normal erythrocyte sedimentation rate in an older adult is commonly interpreted as evidence against vasculitis, and here it was normal even by age-adjusted criteria *(Table 1)*. The diagnosis therefore rested on serology.

Why inflammation localises to the dura remains unresolved.

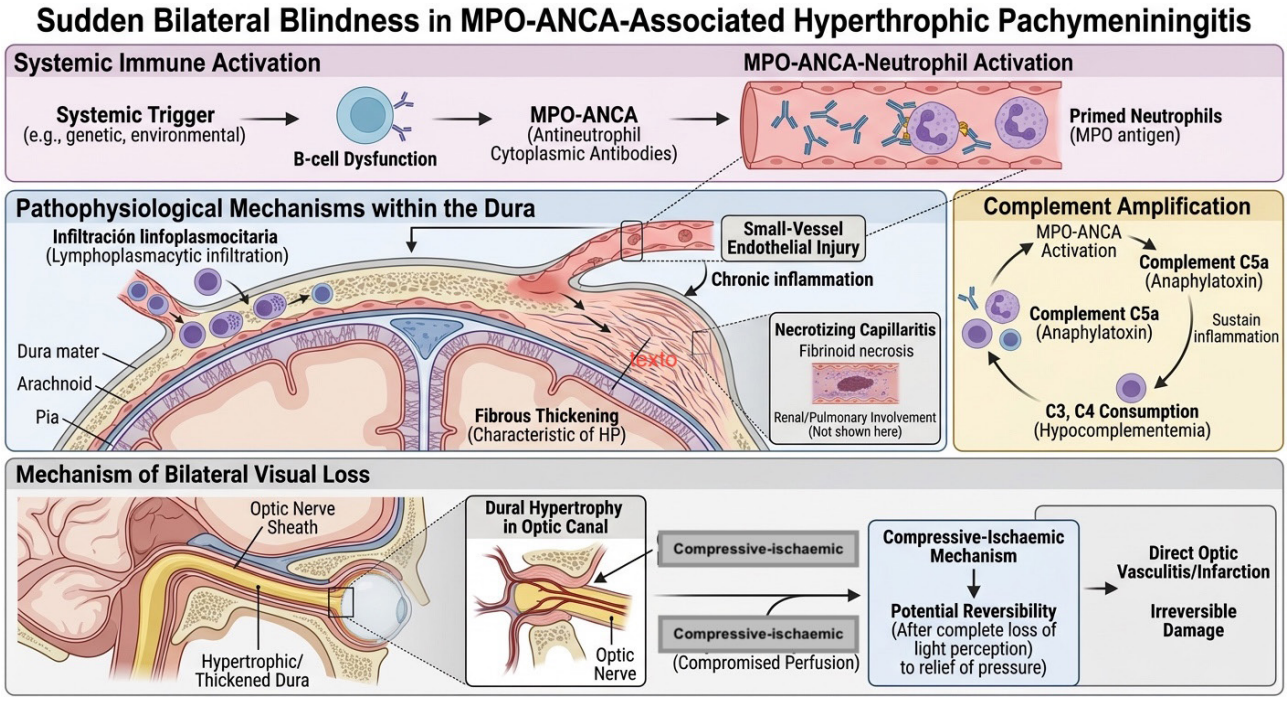


Figure 2. Proposed schematic of the pathophysiology of hypertrophic pachymeningitis in MPO-ANCA-associated vasculitis: 1) circulating anti-MPO antibodies bind primed neutrophils aberrantly expressing myeloperoxidase; 2) degranulation with release of reactive oxygen species and proteolytic enzymes; 3) complement C5a amplifies neutrophil priming through a positive feedback loop; 4) endothelial injury in small dural vessels with local lymphoplasmacytic infiltration; 5) progressive dural thickening compressing adjacent neurovascular structures and impairing optic nerve perfusion; 6) visual loss.

Cause	Discriminating investigations	Findings in this patient
ANCA-associated vasculitis	ANCA by indirect immunofluorescence; antigen-specific anti-MPO and anti-PR3; urinalysis; creatinine; chest CT	p-ANCA 1:20; anti-MPO >400 RU/ml; anti-PR3 <2 RU/ml; microscopic haematuria; diagnostic
IgG4-related disease	Serum IgG4; dural biopsy (IgG4+/IgG ratio, storiform fibrosis, obliterative phlebitis)	IgG4 17.1 mg/dl (normal); biopsy not performed, so seronegative disease not formally excluded
Tuberculous and other infection	CSF PCR, acid-fast smear and culture, fungal and standard cultures, VDRL; chest CT	All negative on two lumbar punctures; chest CT normal
Neoplastic infiltration (lymphoma, carcinomatous meningitis, meningioma en plaque)	CSF cytology and flow cytometry; whole-body imaging; dural biopsy	Chest CT normal; CSF cytology and flow cytometry pending at discharge
Sarcoidosis	Chest CT, serum and CSF angiotensin-converting enzyme, PET-CT, biopsy	No hilar or mediastinal adenopathy, no granulomas on chest CT
Spontaneous intracranial hypotension	Orthostatic headache; brain sagging, venous engorgement or subdural collections on MRI; spinal imaging	Non-postural headache; no brain sagging or subdural collection
Rheumatoid arthritis and other connective tissue disease	Rheumatoid factor, anti-CCP, antinuclear antibodies, extractable nuclear antigen panel; joint examination	Rheumatoid factor 153.1 IU/ml but anti-CCP negative, no synovitis; specific ENA panel negative
Idiopathic hypertrophic pachymeningitis	Diagnosis of exclusion, ideally after dural biopsy	Excluded by markedly positive anti-MPO

Table 2. Parallel diagnostic framework for unexplained diffuse pachymeningeal enhancement, with the findings in this patient. Abbreviations: ANCA, antineutrophil cytoplasmic antibodies; anti-CCP, anti-cyclic citrullinated peptide; CSF, cerebrospinal fluid; CT, computed tomography; ENA, extractable nuclear antigens; MPO, myeloperoxidase; MRI, magnetic resonance imaging; PCR, polymerase chain reaction; PET, positron emission tomography; PR3, proteinase 3; VDRL, Venereal Disease Research Laboratory.

MPO-ANCA activates primed neutrophils, causing small-vessel endothelial injury and lymphoplasmacytic infiltration, which in the dura produces fibrous thickening rather than the necrotising capillaritis characteristic of renal or pulmonary disease[4]. Complement amplification through the alternative pathway and C5a may sustain this process *(Fig. 2)*. Complement is usually normal or raised in AAV, so the concurrently low C3 and C4 were atypical and prompted an extended search for an immune-complex-mediated process. Visual loss probably resulted not from optic nerve vasculitis itself, but from thickened dura compressing the nerve and compromising its perfusion. This distinction matters because a compressive-ischaemic mechanism may be reversible, whereas infarction is not, potentially explaining recovery after complete loss of light perception.

The practical implication is to change the diagnostic sequence. A serial work-up – infection, then IgG4-related disease, malignancy and autoimmunity – may delay treatment. Early antigen-specific ANCA testing, alongside CSF microbiology and chest imaging *(Table 2)*, may identify treatable vasculitis within 48 hours. Glucocorticoids were started on clinical and radiological grounds before serology returned; the 2022 ACR/EULAR criteria for MPA were met once anti-MPO became available[7].

Systemic lupus erythematosus warranted consideration given the low C3 and C4, and antinuclear antibodies at 1:320, but anti-dsDNA and the specific extractable nuclear antigen panel were negative, with no other clinical or haematological criteria. The effect of the four-day interval cannot be established from one patient, while the absence of dural histology, pending CSF cytology and short follow-up limit the conclusions.

Comorbidity, not age, constrained treatment. With an ejection fraction of 30%, cyclophosphamide was unattractive, and rituximab is non-inferior for induction[8]. PEXIVAS supports reduced-dose glucocorticoids, which matters when serious infection competes with the disease as a source of harm[9]. Full independence and a Clinical Frailty Scale score of 2 made an active strategy defensible in a way chronological age never could.

Visual loss with unexplained dural enhancement warrants giant cell arteritis-level urgency; in Latin America, where MPO-ANCA MPA appears to predominate, this phenotype may be under-recognised[10].

REFERENCES

1. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al. 2012 revised International Chapel Hill Consensus Conference nomenclature of vasculitides. Arthritis Rheum. 2013;65:1–11. doi: 10.1002/art.37715.
2. Abrantes FF, Moraes MPM, Rezende Filho FM, Pedroso JL, Barsottini OGP. A clinical approach to hypertrophic pachymeningitis. Arq Neuropsiquiatr. 2020;78:797–804. doi: 10.1590/0004-282X20200073.
3. Yonekawa T, Murai H, Utsuki S, Matsushita T, Masaki K, Isobe N, et al. A nationwide survey of hypertrophic pachymeningitis in Japan. J Neurol Neurosurg Psychiatry. 2014;85(7):732-9. doi: 10.1136/jnnp-2013-306410.
4. Shimojima Y, Sekijima Y. Hypertrophic pachymeningitis in ANCA-associated vasculitis: clinical and immunopathological features and insights. Autoimmun Rev. 2023;22:103338. doi: 10.1016/j.autrev.2023.103338.
5. Shimojima Y, Kishida D, Ichikawa T, Kida T, Yajima N, Omura S, et al. Hypertrophic pachymeningitis in ANCA-associated vasculitis: a cross-sectional and multi-institutional study in Japan (J-CANVAS). Arthritis Res Ther. 2022;24(1):204. doi: 10.1186/s13075-022-02898-4.
6. Sakairi T, Sakurai N, Nakasatomi M, Ikeuchi H, Kaneko Y, Maeshima A, et al. Hypertrophic pachymeningitis associated with antineutrophil cytoplasmic antibody-associated vasculitis: a case series of 15 patients. Scand J Rheumatol. 2019;48:218–224. doi: 10.1080/03009742.2018.1498916.
7. Suppiah R, Robson JC, Grayson PC, Ponte C, Craven A, Khalid S, et al. 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for microscopic polyangiitis. Arthritis Rheumatol. 2022;74:400–406. doi: 10.1002/art.41983.
8. Stone JH, Merkel PA, Spiera R, Seo P, Langford CA, Hoffman GS, et al. Rituximab versus cyclophosphamide for ANCA-associated vasculitis. N Engl J Med. 2010;363:221–232. doi: 10.1056/NEJMoa0909905.
9. Walsh M, Merkel PA, Peh CA, Szpirt WM, Puechal X, Fujimoto S, et al. Plasma exchange and glucocorticoids in severe ANCA-associated vasculitis. N Engl J Med. 2020;382:622–631. doi: 10.1056/NEJMoa1803537.
10. Santacruz-Sandoval E, Lopez-Bonilla J, Guevara-Calderon LA, Nieto-Aristizabal I, Ruiz-Ordonez I, Canas CA, et al. Clinical characteristics and outcomes of patients with ANCA-associated vasculitides in a Colombian hospital. J Clin Rheumatol. 2022;28:e491–e497. doi: 10.1097/RHU.0000000000001777.