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Case Report

Multiple Cranial Neuropathies and Pachymeningitis as Initial Manifestations in Two Patients with Granulomatosis with Polyangiitis

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Abstract

Introduction: Granulomatosis with polyangiitis (GPA) is an ANCA-associated vasculitis that only exceptionally presents with pachymeningitis and multiple cranial neuropathies, which may hinder the early recognition of these atypical manifestations. ***Cases:*** We report two male patients, aged 61 and 67 years, who presented with headache, diplopia, and eyelid ptosis. Both showed pachymeningeal thickening on magnetic resonance imaging and involvement of multiple cranial nerves. They exhibited positive PR3-ANCA and marked cerebrospinal fluid hyperproteinorrachia without evidence of infection; additionally, in one case, biopsy ruled out neoplasia and IgG4-related disease. Both patients were treated with methylprednisolone and cyclophosphamide, achieving favorable outcomes with progressive neurological improvement. ***Conclusion:*** Although rare, GPA-associated pachymeningitis should be considered in patients with headache and cranial neuropathy. This case series provides novel Hispanic American evidence of a poorly characterized manifestation of the disease.

Keywords: ***Granulomatosis with Polyangiitis, Antibodies, Antineutrophil Cytoplasmic, Pachymeningitis, Cranial Neuropathies, Meningeal Diseases.***

Introduction

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a necrotizing vasculitis of small- and medium-sized vessels characterized by granulomatous inflammation and associated with antineutrophil cytoplasmic antibodies (ANCA), particularly those with specificity for proteinase 3 (PR3-ANCA) [1-2]. Although respiratory and renal involvement are its classic manifestations, between 22% and 54% of patients develop neurological complications, including peripheral neuropathy, mononeuritis multiplex, and, less frequently, central nervous system (CNS) involvement [3]. Pachymeningitis and multiple cranial neuropathy represent unusual and underrecognized presentations, described mainly in isolated series from Europe and Asia, and scarcely documented in Hispanic America. Their identification is clinically important, as they may mimic infections, neoplasms, or inflammatory diseases such as IgG4-related disease, delaying the initiation of specific treatment. In this context, we present two cases of GPA with unusual neurological involvement, managed at a Colombian university hospital.

Case Presentation

Case 1

A 67-year-old male with a medical history of hypertension and chronic inflammatory demyelinating polyneuropathy (CIDP), diagnosed one year earlier by electromyography and nerve conduction studies, as well as a prior suspicion of inflammatory myopathy supported by elevated ESR, aldolase, and CPK during a hospitalization in 2024. Since then, he had been receiving immunomodulatory therapy with azathioprine 100 mg/day and prednisolone 15 mg/day, with notable clinical improvement characterized by recovery of muscle strength to 5/5 in all four extremities and weight gain, although with persistent distal paresthesias and electrophysiological evidence of residual chronic polyneuropathy.

He was admitted due to a 3-month history of diplopia, progressive left eyelid ptosis, occasional frontal headache, and marked reduction in visual acuity of the left eye.

Physical examination revealed left ptosis, restriction of ocular movements in all directions, a relative afferent pupillary defect, and abnormal pupillary reflexes. Visual acuity was 20/25 in the right eye and counting fingers at 1 meter in the left. Ishihara testing showed 11/11 plates correct in the right eye and 0/11 in the left. Fundoscopy demonstrated increased optic disc cupping, consistent with left optic neuropathy. No motor or sensory deficits were found in the extremities.

A complete blood count and renal and liver function tests were within normal limits. Systemic inflammation was evidenced by an ESR of 49 mm/h and CRP of 2.59 mg/dL. Antinuclear antibodies (ANA), anti-dsDNA, anti-Sm, anti-RNP, anti-Ro, and anti-La were negative. A markedly positive anti-PR3 ELISA (70.6 U/mL; normal <20 U/mL) was reported, consistent with a c-ANCA pattern by indirect immunofluorescence. Anti-MPO antibodies, rheumatoid factor, and anti-TPO antibodies were negative. Complement levels were normal (C3 152 mg/dL, C4 30 mg/dL).

Brain and orbital MRI revealed pachymeningeal thickening adjacent to the left orbital apex with marked enhancement on contrast-enhanced sequences (**Figure 1A**). Paranasal sinus CT showed inflammatory thickening of the maxillary sinuses, septal deviation, and concha bullosa (**Figure 1A-B**).

Lumbar puncture revealed clear cerebrospinal fluid with an opening pressure of 16 cmH$_2$O, 2 leukocytes/µL, markedly elevated protein (161 mg/dL; normal 15–45), and normal glucose (63 mg/dL). Oligoclonal bands were negative. Microbiological studies—including the meningitis/encephalitis FilmArray panel and molecular testing for Mycobacterium tuberculosis (TB-PCR)—detected no infectious agents.

The clinical presentation and medical history, together with marked hyperproteinorrhachia, elevated PR3-ANCA, and characteristic radiological findings, were highly suggestive of inflammatory pachymeningitis secondary to GPA, with orbital and upper airway involvement.

Although the patient declined meningeal biopsy, he fulfilled clinical and serological criteria for granulomatosis with polyangiitis. Therefore, induction therapy was initiated with methylprednisolone 1 g IV/day for 3 days, followed by cyclophosphamide 1125 mg IV (15 mg/kg) according to vasculitis protocol. This regimen was continued with tapering oral prednisolone (initial dose 40–50 mg/day). The patient exhibited a favorable clinical response, with marked improvement in visual acuity, ptosis, and ocular motility, as well as progressive recovery of neurological symptoms.

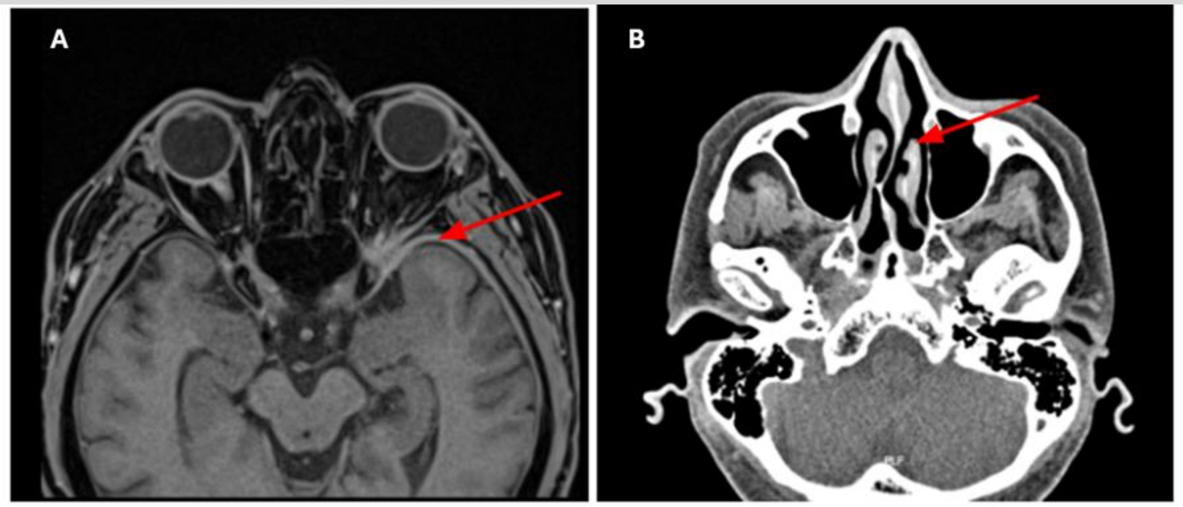

Figure 1. A. Contrast-enhanced T1-weighted brain MRI showing diffuse, irregular thickening of the dura mater with intense, heterogeneous enhancement (red arrow) after gadolinium administration, a finding consistent with pachymeningitis. The involvement predominates at the skull base, particularly in the right lateral aspect of the middle cranial fossa near the cavernous/petrous region, which explains the multiple cranial neuropathies. B. Paranasal sinus computed tomography (bone window) demonstrating mucosal opacification of the left anterior ethmoidal air cells with inflammatory mucosal thickening (red arrow), along with additional changes compatible with chronic sinusitis.

Case 2

A 61-year-old male with no significant past medical history presented with a 23-day history of persistent sudden-onset headache associated with diplopia, right eyelid ptosis, blurred vision, dysphagia, sialorrhea, and aphonia, without motor deficits in the extremities. Neurological examination revealed multiple right cranial nerve involvement, including complete ophthalmoplegia (III, IV, and VI), ptosis with fixed mydriasis, hypoesthesia in the V1 distribution, and bulbar palsy (IX and X) causing severe dysphagia, dysphonia, and poor secretion control, in addition to XII nerve paresis resulting in limited tongue protrusion.

Laboratory findings showed leukocytosis (15,090/µL) with neutrophilia, thrombocytosis (771,000/µL), elevated C-reactive protein (82.4 mg/L), and normal ESR, with preserved renal and hepatic function. The autoimmune panel (ANA, ENA, rheumatoid factor, and anti-CCP) was negative, as were HIV, syphilis, and hepatitis B and C screening tests. Complement levels (C3 and C4) were within normal limits. In the vasculitis workup, indirect immunofluorescence (IIF) for ANCA was negative; however, subsequent ELISA testing revealed a clearly positive anti-PR3 result (67.4 U/mL), with a negative anti-MPO (0.9 U/mL), providing serologic evidence of ANCA-associated vasculitis.

Brain CT and CT angiography showed no acute ischemia or hemorrhage. Neck CTA identified an incidental thyroid nodule without vascular involvement. Cranio-cervical MRI demonstrated a hypointense infiltrative lesion with heterogeneous enhancement in the left parapharyngeal space, extending to the clivus and intracranially (left temporal epidural and subdural spaces), with circumferential vascular involvement but without luminal obliteration (**Figure 2A-C**). Additionally, a second mass-like lesion in the right cavernous sinus extended into the superior orbital fissure, compressing the optic nerve. The findings were interpreted as a chronic granulomatous inflammatory process. Chest CT revealed several cavitated pulmonary nodules in the right upper lobe (**Figure 2D**).

A biopsy of the left subtemporal lesion was performed, showing fibrous tissue with mononuclear lymphocytic infiltrate, necrosis, and reactive gliosis, without evidence of neoplasia (**Figure 3A**). These findings were compatible with chronic pachymeningitis. Immunohistochemistry demonstrated lymphocytic infiltrates and scarce CD38+/CD138+ plasma cells, with negative IgG4 staining, effectively ruling out IgG4-related disease (**Figure 3B**).

The correlation of clinical presentation, imaging findings, serology (positive anti-PR3), and histopathology supported the diagnosis of granulomatosis with polyangiitis (GPA) with multiple cranial neuropathies and pachymeningitis.

Treatment was initiated with methylprednisolone pulses (500 mg IV every 24 hours for 3 days), followed by oral prednisolone 50 mg/day with gradual tapering. Induction therapy with cyclophosphamide 1.5 g IV monthly (dose calculated as 750 mg/m²) was administered, with the first dose on 08/31/2024 and the second on 10/01/2024. The patient experienced progressive improvement in ocular and bulbar involvement, recovery of swallowing function, and a decrease in inflammatory markers, requiring swallowing and ophthalmologic rehabilitation. He was discharged 102 days after initial admission.

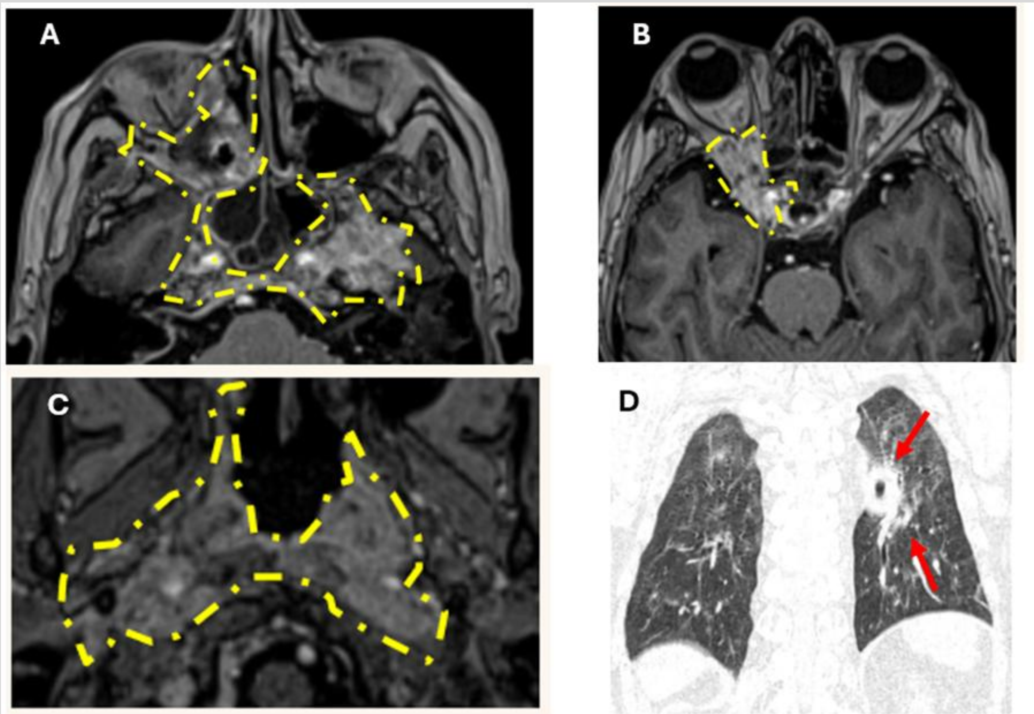

Figure 2. Brain MRI. (A) Axial view demonstrating heterogeneous and diffuse enhancement at the skull base, involving the bilateral petroclival regions, Meckel's cave, and the cavernous sinuses (yellow dashed lines). (B) Post-gadolinium axial T1-weighted image showing an expansile lesion with intense enhancement in the right petroclival region (yellow dashed lines), in close association with the course of the trigeminal nerve. (C) Post-gadolinium axial T1-weighted image revealing irregular infiltrative enhancement along the clivus, with bone destruction and granulomatous inflammation (yellow dashed lines). (D) Coronal reconstruction of chest CT demonstrating two cavitated left pulmonary nodules (red arrows), a characteristic finding of granulomatosis with polyangiitis.

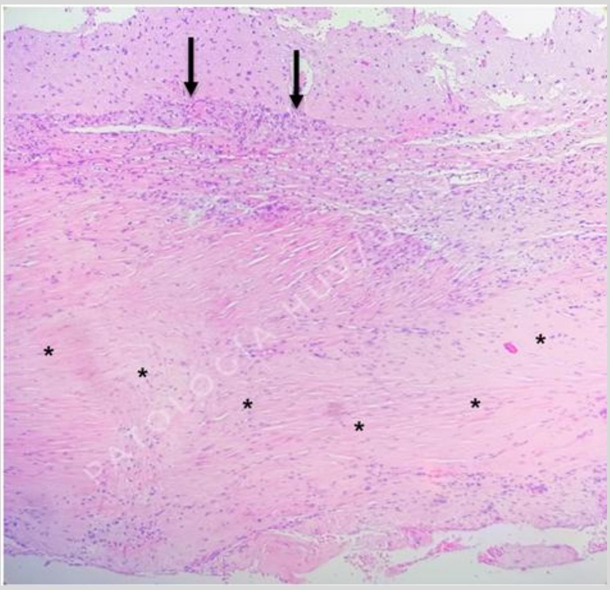

Figure 3. Biopsy of the left subtemporal lesion. H&E 100×. Fibrous tissue with predominance of dense, organized pink collagen fibers (*). Compact distribution indicative of lymphocytic inflammatory infiltrate (arrows).

Discussion

Granulomatosis with polyangiitis (GPA) is a necrotizing vasculitis of small- and medium-sized vessels characterized by granulomatous inflammation, predominantly affecting the respiratory tract and kidneys, and strongly associated with antineutrophil cytoplasmic antibodies (ANCA), particularly PR3-ANCA [1].

GPA is a rare condition. Recently, Redondo-Rodríguez et al. (2022) reported a combined global incidence of ANCA-associated vasculitis of 17.2 cases per million person-years (95% CI: 13.3–21.6), with GPA being the most common (≈9 cases/million person-years) [2]. Neurological involvement in ANCA-associated vasculitis occurs in 22–54% of patients with GPA and 24–72% of those with microscopic polyangiitis (MPA) [3]. The Japanese multicenter J-CANVAS study, which included 663 patients with ANCA-associated vasculitis, reported pachymeningitis in approximately 5% of cases, predominantly in GPA, with cranial neuropathy present in 46% of those cases, multiple cranial neuropathies in 15%, and orbital involvement in 31% [4]. The most frequently affected cranial nerve was the trigeminal nerve (23%) [4].

In Hispanic America, epidemiological data on GPA with neurological involvement are scarce. In Argentina, a retrospective cohort of 37 patients with severe GPA (1994–2011) reported neurological involvement in 43%, although cranial neuropathy was infrequent (10.8%) [5]. In Mexico, Cervera-Castillo et al. (2017) reported a series of 28 cases in which 28.6% of patients with ANCA-associated vasculitis exhibited neuropsychiatric involvement, mostly peripheral neuropathy (21.4%) and, to a lesser extent, CNS manifestations (7.1%), while cranial neuropathies were exceptional [6]. Another Mexican series by Higuera-Ortiz et al. included 13 patients with GPA and pachymeningitis; 46% presented cranial neuropathies, with the trigeminal nerve being the most frequently involved, as well as multiple cranial nerve deficits in some cases [7]. These findings are consistent with our series, where multiple cranial nerve involvement also predominated in the context of pachymeningitis.

In Colombia, Santacruz-Sandoval et al. (2022) described 67 patients with ANCA-associated vasculitis: 32.8% presented CNS involvement and 23.9% peripheral neuropathy, more commonly in eosinophilic granulomatosis with polyangiitis (EGPA) and MPA, but also in GPA (28.6%) [8]. Hurtado-Uriarte et al. (2024) evaluated 23 hospitalized patients with severe ANCA-associated vasculitis and documented CNS involvement in 17.4% [9]. Although there are Hispanic American reports of ANCA-associated pachymeningitis [10-16], few describe simultaneous involvement of multiple cranial nerves and orbital disease as in our series (see comparative table).

Table 1: Comparison of ANCA-associated vasculitis cases with pachymeningitis reported in Latin America

Reference	Age/Sex	ANCA	Diagnosis	Clinical manifestations	Imaging findings	Treatment	Outcome
Rodríguez et al. 2020 (Colombia) [10]	57F	MPO+	ANCA-associated vasculitis (MPA) with hypertrophic pachymeningitis	Hemicranial headache, otalgia, facial paresthesias, chronic IgA nephropathy	MRI: diffuse pachymeningitis in tentorium, posterior fossa and left hemicranium	Prednisolone + azathioprine (no cyclophosphamide due to chronic renal disease)	Clinical improvement after immunosuppression
Huaringa et al. 2020 (Peru) [13]	60M	PR3- and MPO- (ANCA negative)	ANCA-negative GPA with hypertrophic pachymeningitis	Progressive headache, vomiting, unilateral papilledema, sinusitis, otomastoiditis, severe anemia	MRI: sinusitis, otomastoiditis and diffuse pachymeningitis; meningeal biopsy confirmed necrotizing granulomatous vasculitis	Corticosteroids only (no cyclophosphamide due to malnutrition)	Initial improvement, death due to septic shock 7 days post-discharge
Restrepo et al. 2016 (Colombia) [11]	65M	MPO+, PR3-	GPA with pachymeningitis, aortitis, and glomerulonephritis	Chronic headache, elevated ESR, aortitis, nephritis; initially suspected temporal arteritis	MRI: diffuse supra- and infratentorial pachymeningitis; MR angiography showed aortitis; temporal biopsy normal	Methylprednisolone + IV cyclophosphamide → clinical improvement	Clinical improvement and normalization of labs after induction
Contreras et al. 2018 (Colombia) [12]	67M	p-ANCA+	p-ANCA vasculitis with pachymeningitis and rapidly progressive glomerulonephritis	Proptosis, severe dysphagia, multiple cranial neuropathies, renal failure requiring hemodialysis	MRI: frontal, temporal, posterior fossa and cavernous sinus dural enhancement; biopsy ruled out infection/neoplasm	Methylprednisolone, IV cyclophosphamide, plasmapheresis → partial remission	Partial remission, discontinuation of dialysis
Benavides-Burbano et al. 2020 (Colombia) [15]	39F	PR3+	GPA with posterior fossa involvement and pachymeningitis	Headache, nausea, tinnitus, vertigo, photophobia, chronic sinusitis, subglottic stenosis	MRI: intra- and extra-axial infratentorial lesion suggestive of meningioma; pathology: necrotizing granulomatous inflammation	Surgery (dural resection), methotrexate, azathioprine; later corticosteroids and antibiotics for meningitis	Post-surgical infectious complications (Klebsiella), later sustained improvement
De Deus-Silva et al. 2003 (Brazil) [16]	45F	Negative	Idiopathic hypertrophic pachymeningitis vs neurosarcoidosis	Psychosis, progressive visual loss, optic atrophy, nasal septum destruction	MRI: basal frontal dural thickening with enhancement; biopsy: fibrosis and chronic inflammation with early granulomas	Corticosteroids, azathioprine	Partial improvement of visual and psychiatric symptoms; relapses treated with steroid pulses
Argüello et al. 2021 (Argentina) [14]	65M	PR3+, c-ANCA	Long-standing GPA with pachymeningitis and orbital mass	Chronic headache, left ocular proptosis, orbital pain, elevated acute phase reactants	MRI: cerebral and tentorial dural enhancement; left orbital mass; biopsy: necrotizing granuloma with vasculitis	Methylprednisolone, IV cyclophosphamide, later rituximab; prior azathioprine maintenance	Favorable response with rituximab after relapses; clinical improvement with corticosteroid tapering
Berrocal et al. Case series (Colombia)	61M and 67M	PR3+	PR3+ GPA with pachymeningitis and multiple cranial neuropathies	Headache, diplopia, eyelid ptosis, multiple cranial nerve involvement	MRI: pachymeningeal thickening with inflammatory skull base lesions; CSF hyperproteinorrachia	Methylprednisolone + IV cyclophosphamide → clinical improvement	Favorable evolution with progressive neurological recovery

Abbreviations: *HP: Hypertrophic Pachymeningitis; GPA: Granulomatosis with Polyangiitis; MPA: Microscopic Polyangiitis; MRI: Magnetic Resonance Imaging; RPGN: Rapidly Progressive Glomerulonephritis; CSF: Cerebrospinal Fluid.*

Both of our patients presented the characteristic clinical and radiologic triad of GPA with meningeal and orbital involvement. They developed subacute to chronic cranial neuropathies including ptosis, diplopia, visual loss, dysphagia, and dysphonia–accompanied by MRI findings of pachymeningeal thickening and enhancement, skull-base and cavernous sinus lesions, as well as cerebrospinal fluid hyperproteinorrachia and positive anti-PR3 serology. In one case, chest CT also demonstrated characteristic pulmonary abnormalities–bilateral nodules, some cavitated (**Figure 2**) supporting systemic involvement. This combination of clinical, serological, imaging, and when possible, histopathological findings enabled diagnostic confirmation even in the absence of pulmonary or renal biopsy. Histologic confirmation remains the gold standard, but it may be negative in localized forms or when the affected structures (e.g., skull base) are difficult to access.

Pachymeningitis and cranial nerve involvement in GPA, while uncommon, are well documented in case series and reports [17,18]. They have been described in patients aged 27–81 years, without sex predominance. The most frequently affected cranial nerves include II, III, VI, and VII; VIII involvement is rare, and some cases report orbital apex syndrome, bulbar dysfunction, or acute visual loss. Cranial neuropathies without pachymeningitis have also been described, usually attributed to vasculitic processes [19]. Chronic sinusitis, otitis media, or mastoiditis often coexist [18]. Most patients exhibit elevated inflammatory markers such as ESR and CRP [17]. Computed tomography has low sensitivity for meningeal disease, whereas contrast-enhanced MRI shows focal or diffuse dural thickening and enhancement [20]. Cerebrospinal fluid typically shows elevated protein and mild lymphocytic pleocytosis [17]. Histology may reveal granulomatous inflammation or lymphocytic infiltrates. Although PR3-ANCA positivity is common, cases associated with MPO-ANCA, dual ANCA positivity, or even ANCA-negative disease have been reported [13,22]. Therefore, serology alone does not rule out the diagnosis.

From a pathophysiological standpoint, meningeal involvement and orbital apex syndrome in GPA may be explained by three mechanisms: (1) contiguous granulomatous extension from paranasal sinuses or orbit; (2) primary granulomatous inflammation of the dura mater (hypertrophic pachymeningitis); and (3) small-vessel meningeal vasculitis leading to ischemia or neural infiltration. These mechanisms account for the frequent involvement of multiple cranial nerves and the coexistence of optic, motor, and bulbar neuropathies [3].

Regarding treatment, no large clinical trials have focused specifically on neurological involvement in GPA; recommendations are extrapolated from systemic disease management. Induction therapy includes high-dose glucocorticoids combined with cyclophosphamide or rituximab [23,24]. In severe cases, plasma exchange may be considered, although current evidence shows limited benefit except in diffuse alveolar hemorrhage or severe renal failure [25]. Once remission is achieved, maintenance therapy with methotrexate, azathioprine, mycophenolate mofetil, or ideally rituximab is recommended [26].

Prognosis depends on timely diagnosis and initiation of immunosuppressive therapy. Early treatment is associated with better neurological recovery and headache resolution [16], although residual visual deficits, cranial nerve dysfunction, and cognitive impairments are common. Relapse occurs in up to 40% of patients within 2 years, even with maintenance immunosuppression [26]. Neurological involvement is associated with a higher risk of recurrence and poorer quality of life. Although mortality data specific to GPA with neurological involvement are lacking, European cohorts of ANCA-associated vasculitis with severe renal disease report 5-year survival rates of 69–91% [27].

In this context, our cases provide Latin American evidence of an uncommon manifestation of GPA, involving orbital disease and multiple cranial neuropathies, with favorable response to immunosuppressive therapy. These findings underscore the importance of including GPA in the differential diagnosis of pachymeningitis and cranial neuropathy in clinical practice.

Conclusion

Neurological involvement in granulomatosis with polyangiitis (GPA) poses significant diagnostic and therapeutic challenges. Pachymeningitis and multiple cranial neuropathies, although rare, should be considered in patients presenting with persistent headache, diplopia, or focal cranial nerve deficits. Diagnosis requires a comprehensive evaluation to exclude infections and neoplasms, supported by imaging, serologic testing, and—when feasible—histologic confirmation. Early initiation of immunosuppression with glucocorticoids and cyclophosphamide or rituximab improves outcomes, although the risk of relapse and neurological sequelae remains. Our series provides novel evidence from Hispanic America and highlights the need for systematic reporting of such cases to strengthen future diagnostic and therapeutic guidelines.

Declarations

Ethical Approval

Approved by ethics committee

Consent of Publication

Taken

Conflict of Interest

None

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