EJPMR, 2026, 13(7), 262-267 **Case Study** **Impact Factor: 8.158**

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

ISSN (O): 2394-3211
ISSN (P): 3051-2573
Coden USA: EJPMAG

AN INTERESTING CASE OF GRANULOMATOSIS WITH POLYANGIITIS (WEGENER'S GRANULOMATOSIS) MASQUERADING AS A MULTISYSTEM DISORDER

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DOI: https://doi.org/10.5281/zenodo.21030925

How to cite this Article: Dr. Sadananth S.[1]*, Dr. Sivasubramanian MD[2], Dr. Rathnakumar MD[3]. (2026). An Interesting Case of Granulomatosis With Polyangiitis (Wegener's Granulomatosis) Masquerading As A Multisystem Disorder. European Journal of Pharmaceutical and Medical Research, 13(7), 262-267.

Article Received on 02/06/2026 **Article Revised on 23/06/2026** **Article Published on 01/07/2026**

ABSTRACT

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a small-vessel, antineutrophil cytoplasmic antibody (ANCA)-associated necrotising vasculitis that classically presents with a triad of upper respiratory, pulmonary, and renal involvement. We report a 53-year-old woman with a 15-year history of type 2 diabetes mellitus, systemic hypertension, and hypothyroidism who presented with a one-week history of bilateral ear discharge, cough, and rapidly progressive breathlessness, together with oral ulcers, decreased hearing, giddiness, and headache. Examination revealed bilateral perforated tympanic membranes with otorrhoea, oral ulceration, tachypnoea, and bilateral lower motor neuron facial palsy with absent taste sensation, together with a left-sided glossopharyngeal and vagal palsy. Laboratory evaluation showed marked leucocytosis, a strikingly elevated ESR and CRP, preserved renal function, and a negative autoimmune and infective screen, while imaging revealed bilateral paranasal sinusitis, bilateral acute-on-chronic otomastoiditis, and a thick-walled cavitating right lower lobe lesion with multiple bilateral pulmonary nodules. A bronchial wash grew Aspergillus fumigatus, presumed to represent saprophytic colonisation of a necrotic cavity, while transbronchial lung biopsy showed only an organised abscess without diagnostic granulomatous vasculitis. A strongly positive cytoplasmic ANCA (cANCA, proteinase-3 pattern), interpreted together with the ENT, pulmonary, and cranial neuropathic findings, established a diagnosis of GPA. Nerve conduction studies additionally revealed a subclinical demyelinating-axonal motor polyradiculoneuropathy despite an unremarkable bedside limb examination. The patient was treated with pulse intravenous methylprednisolone followed by an oral steroid taper; systemic symptoms improved, but bilateral facial palsy and hearing loss persisted as residual deficits at follow-up. This case illustrates the value of ANCA serology and multisystem pattern recognition in securing a diagnosis of GPA even when histopathology is non-diagnostic, and highlights cranial mononeuropathy as an under-recognised, potentially irreversible manifestation of the disease.

KEYWORDS: *Granulomatosis with polyangiitis; Wegener's granulomatosis; cytoplasmic ANCA; cranial neuropathy; facial nerve palsy; pulmonary cavitation.*

INTRODUCTION

Granulomatosis with polyangiitis (GPA) is a rare, idiopathic, necrotising small- and medium-vessel vasculitis combined with extravascular granulomatous inflammation, classified together with microscopic polyangiitis and eosinophilic granulomatosis with polyangiitis under the umbrella of ANCA-associated vasculitis (AAV). GPA is typically associated with cytoplasmic ANCA directed against proteinase-3 (PR3-ANCA), and classically involves the ear, nose and upper

respiratory tract, the lower respiratory tract, and the kidneys. The 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for GPA incorporate weighted clinical, serological, radiological, and histopathological items, including nasal/sino-nasal involvement, conductive or sensorineural hearing loss, cANCA or PR3-ANCA positivity, pulmonary nodules, masses or cavitation on imaging, and granulomatous inflammation on biopsy, reflecting the multisystem nature of the disease.[1]

While the classic pulmonary-renal pattern is well recognised, GPA may affect virtually any organ system, and its presentation is frequently protean, leading to substantial diagnostic delay. Cranial and peripheral nerve involvement, in particular, is comparatively under-recognised, occurring through granulomatous extension from adjacent structures such as the mastoid or skull base, or through vasculitic mononeuropathy, and can mimic Bell's palsy, infective skull-base disease, or malignancy.[2,3] We report a case in which a combination of otologic, pulmonary, and multiple cranial nerve findings—mimicking infective oto-sinusitis with septic pulmonary disease—was ultimately attributed to GPA on the strength of ANCA serology and the overall clinical-radiological pattern, despite a non-diagnostic lung biopsy.

CASE REPORT

History

A 53-year-old woman presented with a one-week history of bilateral ear discharge that was initially serous and became mucopurulent, non-foul-smelling and non-blood-stained, accompanied by decreased hearing on both sides. She had a concurrent one-week history of cough with scanty white mucoid expectoration, and breathlessness of three days' duration that was acute in onset and rapidly progressive (MMRC grade 2 to grade 4), such that she was breathless at rest by the time of admission. She additionally reported oral ulcers for three days, giddiness for one week, and headache for one week. There was no fever, chest pain, palpitations, haematuria, haemoptysis, significant weight loss, vertigo, seizures, or limb weakness.

She was a known case of type 2 diabetes mellitus, systemic hypertension, and hypothyroidism, each of 5–15 years' duration and on regular treatment, with no other comorbidities, prior similar episodes, or surgical history. Personal and family history were non-contributory.

Examination Findings

On general examination the patient was conscious, oriented, afebrile and tachypnoeic, without pallor, icterus, cyanosis, clubbing, pedal oedema, or lymphadenopathy. Multiple crusted oral ulcerations were present, along with bilateral ear discharge with perforated tympanic membranes. Vitals revealed a blood pressure of 130/80 mmHg (right arm, sitting, without radio-radial or radio-femoral delay), a regular pulse of 89/min of good volume, a respiratory rate of 25/min, and oxygen saturation of 99% on room air.

ENT examination showed bilateral yellowish nasal crusting without septal deviation. Respiratory examination revealed normal vesicular breath sounds with bilateral interscapular and infrascapular wheeze; the cardiovascular and abdominal examinations were unremarkable. Higher mental functions were normal. Cranial nerve examination revealed bilateral lower motor neuron facial palsy and additional lower cranial nerve involvement, summarised in Table 1.

Table 1: Cranial nerve examination findings.

Cranial Nerve	Right	Left
I – Olfactory	Smell perception intact	Smell perception intact
II – Optic	Acuity 6/6; fields, colour vision, reflexes, fundus normal	Acuity 6/6; fields, colour vision, reflexes, fundus normal
III, IV, VI	Full eye movements, no ptosis, pupil 3 mm reactive, no nystagmus	Same as right
V – Trigeminal	Facial sensation and corneal reflex normal	Facial sensation and corneal reflex normal
VII – Facial	LMN-type weakness: reduced forehead wrinkling, incomplete eye closure (Bell's phenomenon +), reduced nasolabial fold, absent anterior 2/3 taste, absent hyperacusis	Same findings (bilateral)
VIII – Vestibulocochlear	Conductive hearing loss (Rinne's AC<BC)	Conductive hearing loss (Rinne's AC<BC); Weber's not lateralised
IX – Glossopharyngeal	Gag reflex present	Gag reflex absent
X – Vagus	–	Uvula deviated to the right (left vagal weakness)
XI – Accessory	Normal shoulder shrug / SCM power	Normal shoulder shrug / SCM power
XII – Hypoglossal	Tongue midline, no fasciculations	Tongue midline, no fasciculations

Bedside examination of the limbs showed normal bulk, tone, and power (5/5) in all muscle groups, with deep tendon reflexes of 2+ throughout, bilaterally flexor plantar responses, normal coordination and sensation, no peripheral nerve thickening, a normal spine examination, and an intact autonomic system (no orthostatic hypotension, abnormal heart-rate/blood-pressure variability, or excessive sweating).

INVESTIGATIONS

Haematology

Table 2: Serial complete blood counts, showing persistent leucocytosis without thrombocytopenia.

Parameter	Day 1	Day 8	Day 17	Day 24	Day 27
Total count (/µL)	20,600	17,300	20,600	19,300	26,800
Hb (g/dL)	11.0	10.0	11.0	10.8	11.8
PCV (%)	37	35	33	37	41
MCV (fL)	95	98	101	100	106
Platelets (lakh/µL)	2.81	2.93	3.61	3.30	3.81

Renal and Liver Function

Table 3: Serial renal function tests; creatinine remained within normal limits throughout.

Parameter	Reading 1	Reading 2	Reading 3
Urea (mg/dL)	38	23	29
Creatinine (mg/dL)	0.70	0.58	0.90
Sodium (mEq/L)	139	138	145
Potassium (mEq/L)	4.0	2.7	4.0
Random blood sugar (mg/dL)	87	105	185

Table 4: Liver function tests, showing initial hypoalbuminaemia consistent with active systemic inflammation, with improvement on follow-up.

Parameter	Initial	Follow-up
Total bilirubin (mg/dL)	0.7	0.6
Direct bilirubin (mg/dL)	0.4	0.3
Indirect bilirubin (mg/dL)	0.3	0.3
SGOT (U/L)	63	46
SGPT (U/L)	24	27
ALP (U/L)	235	230
Total protein (g/dL)	4.4	5.2
Albumin (g/dL)	2.4	3.2
Globulin (g/dL)	2.0	2.0

Inflammatory Markers, Serology and Urinalysis.

Table 5. The strongly positive cANCA (PR3 pattern) with negative pANCA, ANA, and viral markers, alongside near-normal renal parameters, was pivotal in distinguishing GPA from its mimics.

Test	Result	Reference / Remarks
ESR	125 mm/hr	Markedly elevated
CRP	92 mg/L	Markedly elevated
ANA profile	Negative	–
cANCA	Positive (16)	>3 considered positive
pANCA	Negative (0.2)	>5 considered positive
HBsAg / HCV / HIV	Negative / Negative / Negative	–
Free T3 / Free T4 / TSH	1.14 / 2.07 / 1.06	Euthyroid
Urine albumin / sugar / RBCs	Trace / Nil / Nil	No active urinary sediment
24-hour urinary protein	190 mg/day	Within normal limits

Microbiology

Table 6: Sterile cultures and negative tuberculosis workup argued against an infective process; biopsy did not show diagnostic granulomatous vasculitis.

Specimen / Test	Result
Sputum, urine and blood culture	No growth
Sputum CBNAAT	Negative
Sputum AFB	Negative

Aural swab culture	No growth
Bronchial wash fungal culture	Aspergillus fumigatus
Bronchial wash cytology	Inflammatory smear
Bronchial wash CBNAAT	Negative
Transbronchial lung biopsy	Organised lung abscess; no granulomas or vasculitis identified

Imaging, Electrophysiology and Cardiac Evaluation

Contrast-enhanced CT of the paranasal sinuses showed mucosal thickening in the right maxillary and ethmoidal sinuses and the left maxillary sinus, consistent with sinusitis. CT of the temporal bones demonstrated bilateral mastoid and middle-ear fluid density with sclerosis and paucity of air cells, in keeping with bilateral acute-on-chronic otomastoiditis. CT of the chest revealed a thick-walled cavitating lesion in the mediobasal segment of the right lower lobe, with multiple fairly well-defined nodular lesions scattered bilaterally—predominantly in the lower lobes—and consolidative change in the right upper lobe and right middle lobe/lingula. CT of the brain and abdomen were normal. Electrocardiography showed sinus tachycardia (rate approximately 104/min) with a prolonged corrected QT interval and a normal axis. Two-dimensional echocardiography showed an ejection fraction of 60% with grade I diastolic dysfunction and no regional wall motion abnormality. Nerve conduction studies of the bilateral upper limbs, lower limbs, and facial nerves revealed a motor polyradiculoneuropathy, predominantly demyelinating with secondary axonal features—an electrophysiological abnormality not apparent on bedside limb examination.

DIFFERENTIAL DIAGNOSIS

The combination of otologic, pulmonary and constitutional features initially suggested several alternative diagnoses, each of which was systematically reconsidered as additional data became available (Table 7).

Table 7: Differential diagnoses considered and the basis for their exclusion.

Differential Diagnosis	Supportive Cues	Exclusion Criteria	Reality in this Case
Infective oto-sinusitis with septic pulmonary emboli/abscess	Bilateral otorrhoea, perforated tympanic membranes, productive cough, cavitating lung lesion	Aural swab, blood, sputum and urine cultures sterile	Sterile granulomatous/vasculitic inflammation rather than true bacterial infection
Pulmonary tuberculosis	Cavitating right lower lobe lesion, multiple nodules, constitutional decline	Sputum AFB and CBNAAT negative	GPA cavitatory nodules can closely mimic tuberculosis radiologically
Pulmonary aspergillosis / aspergilloma	Bronchial wash fungal culture grew Aspergillus fumigatus	Biopsy showed no fungal invasion or hyphae; cytology only inflammatory	Saprophytic colonisation of a pre-existing necrotic GPA cavity
Bell's palsy (idiopathic LMN facial palsy)	Bilateral LMN facial weakness	Bilateral, simultaneous onset with concurrent IX/X and ENT involvement, atypical for idiopathic Bell's palsy	Cranial neuropathy from skull-base/temporal-bone granulomatous inflammation and/or vasculitic mononeuropathy
Systemic lupus erythematosus / other ANA-associated disease	Multisystem involvement, elevated inflammatory markers	ANA profile negative	cANCA/PR3 pattern supports GPA rather than an ANA-associated connective tissue disease

DIAGNOSIS

The combination of granulomatous sino-nasal and otomastoid disease, multiple cranial neuropathies (VII, IX, X), pulmonary nodularity with cavitation, and strongly positive cANCA/PR3 serology—together with the systematic exclusion of infective, mycobacterial, fungal and other autoimmune aetiologies—satisfied multiple weighted items of the 2022 ACR/EULAR classification criteria for GPA, and a diagnosis of granulomatosis with polyangiitis was established despite a non-diagnostic transbronchial lung biopsy.

MANAGEMENT AND OUTCOME

Multidisciplinary opinions were obtained from neurology, ENT, thoracic medicine, nephrology, ophthalmology, and rheumatology. The patient was treated with pulse intravenous methylprednisolone followed by a tapering course of oral corticosteroid. Systemic and respiratory symptoms improved with treatment; however, the bilateral facial nerve palsy and hearing loss persisted as residual deficits at the time of follow-up.

DISCUSSION

GPA is a PR3-ANCA-predominant, pauci-immune necrotising vasculitis with extravascular granulomatous inflammation that most often involves the upper and lower respiratory tracts and kidneys, though virtually any organ can be affected. Biopsy, while strongly recommended, is frequently non-diagnostic because of

the patchy distribution of granulomatous vasculitis, particularly in transbronchial or percutaneous lung samples; consequently, current guidance emphasises an integrated clinical, serological and radiological assessment rather than reliance on histopathology alone.[4] This pattern was borne out in the present case, in which lung biopsy yielded only an organised abscess, and the diagnosis instead rested on the convergence of ENT, pulmonary, and serological findings.

The clinical picture here closely mimicked an infective process—bilateral otomastoiditis, a cavitating pulmonary lesion, and a positive fungal culture together raised concern for bacterial sepsis, tuberculosis, or primary pulmonary aspergillosis. Cavities in GPA are well documented to become secondarily colonised by Aspergillus fumigatus, radiographically and microbiologically mimicking aspergilloma or invasive aspergillosis; recognising this as colonisation of a vasculitic cavity, rather than the primary pathology, was essential to avoid a purely anti-infective approach that would not address the underlying vasculitis.[4,5]

Cranial neuropathy is a comparatively under-recognised manifestation of GPA, occurring through direct granulomatous extension from the paranasal sinuses, mastoid, or skull base, or through vasculitic mononeuropathy of the vasa nervorum. Bilateral facial palsy with additional lower cranial nerve involvement, as seen in this patient, has been described but remains uncommon and is easily mistaken for idiopathic Bell's palsy when ENT and systemic features are not actively sought.[2,3] Delayed recognition of GPA as the cause of cranial neuropathy has been associated with incomplete neurological recovery despite eventual immunosuppression, consistent with the residual facial and auditory deficits observed in this patient even after symptomatic systemic improvement.[2]

Peripheral nerve involvement in GPA is reported in a minority of patients and most often manifests as a mononeuritis multiplex or motor-predominant polyradiculoneuropathy; electrophysiological abnormalities can precede or occur without overt clinical deficits, underscoring the value of nerve conduction studies as a sensitive surveillance tool even when the bedside neurological examination of the limbs is unremarkable, as in this case.[6]

The near-normal renal parameters and modest proteinuria in this patient place her within the subset of GPA with limited renal involvement at presentation, which is generally considered more favourable prognostically, though ongoing surveillance for evolving glomerulonephritis remains important. Current EULAR recommendations advocate high-dose glucocorticoids in combination with rituximab or cyclophosphamide for remission induction in organ- or life-threatening AAV; in this patient, pulse glucocorticoid therapy alone achieved systemic and pulmonary improvement, although the persistence of cranial neuropathy raises the question of whether earlier or more intensive induction therapy might have improved neurological outcome.[7]

CONCLUSION

This case underscores the importance of maintaining a high index of suspicion for granulomatosis with polyangiitis in patients presenting with combined ENT, pulmonary, and cranial-neurological features, even when renal function is preserved and lung histopathology is non-diagnostic. cANCA/PR3 serology, interpreted alongside the overall clinical and radiological pattern, was central to establishing the diagnosis in this patient. Cranial mononeuropathy should be recognised as a potentially irreversible manifestation of GPA, reinforcing the rationale for prompt and adequately intensive immunosuppressive therapy once the diagnosis is suspected.

DECLARATIONS

Conflict of Interest: The authors declare no conflict of interest.

Patient Consent: Obtained.

Funding: None declared.

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