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CASE REPORT

Medicine Science International Medical Journal

Medicine Science 2026;15(1):540-3

P-ANCA positive granulomatosis with polyangiitis presented by acute motor and sensory axonal neuropathy

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Received 20 October 2025; Accepted 02 February 2026
Available online 27 February 2026 with doi: 10.5455/medscience.2025.10.289

Abstract

Acute motor and sensory axonal neuropathy (AMSAN) is a variant of Guillain-Barré syndrome (GBS) that involves motor and sensory fibers and causes axonal damage. Malignancy, vasculitis, and infectious agents may be responsible for the etiology of GBS. Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a necrotizing vasculitis affecting small-to medium-sized vessels. One of these vasculitis types is Granulomatosis with Polyangiitis (GPA). Involvement of the nervous system in GPA is at a level of 20-50%; peripheral or cranial neuropathy may be observed. It is a matter of debate whether AAV and GBS rarely occur together in patients due to autoimmune predisposition or whether vasculitic neuropathy is a result of vasculitis. In this article, a case that presented with AMSAN as a rare condition and was diagnosed with GPA is discussed. Since the prognosis depends on the correct diagnosis and treatment selection, further research is needed in this area.

Keywords: Acute motor and sensory axonal neuropathy, anti-neutrophil cytoplasmic antibody-associated vasculitis, guillain-barre syndrome, granulomatosis with polyangiitis, anti-neutrophil cytoplasmic antibody

Introduction

Acute motor and sensory axonal neuropathy (AMSAN) is a rare variant of Guillain-Barre syndrome (GBS) that involves motor and sensory fibres and causes axonal damage. GBS is an autoimmune disease characterised by ascending, symmetric, numbness and muscle weakness as a result of damage to peripheral nerves. It has been reported that before GBS, 60% of patients have a history of upper respiratory tract infection (URTI), acute gastroenteritis (AGE), an operation or vaccinations [1]. In addition, malignancy, vasculitis and other infectious agents may also be responsible for the etiology [2].

Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) comprises life-threatening autoimmune diseases that cause necrotizing vasculitis affecting small-to medium-sized vessels. They are separated into 3 groups, Granulomatosis with Polyangiitis (GPA), microscopic polyangiitis (MPA) and Churg-Strauss syndrome (CSS). Antibodies formed against c-ANCA which binds to proteinase 3 (PR3) and p-ANCA which binds to myeloperoxidase (MPO) in neutrophils play a role in the development of these diseases and their positivity can be detected at varying rates [3]. GPA is a vasculitis leading to systemic necrotising granulomatous inflammation involving the upper and lower respiratory tract and kidney [4]. Involvement of the nervous system in GPA can be seen in 20-50% of patients. Peripheral or cranial neuropathy may be observed [5]. The pathological findings of vasculitic polyneuropathy (VP) are characterized by axonal degeneration caused by vasculitis-induced ischemia [5,6]. Neurologic symptoms as the initial symptom are very rare [6].

ANCA-associated vasculitis and GBS are two diseases caused by autoimmune mechanisms and it is a matter of debate in the literature as to if they rarely develop together in patients with a predisposition to autoimmune diseases or as a result of damage to the peripheral nervous system as a result of vasculitis. In this article, a case of a patient who presented with AMSAN,

CITATION
Onur GY. P-ANCA positive granulomatosis with polyangiitis presented by acute motor and sensory axonal neuropathy. Med Science. 2026;15(1):540-3.

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DOI: 10.5455/medscience.2025.10.289

a rare condition, and was diagnosed with GPA after etiologic investigations, is mentioned.

Case Presentation

A 74-year-old woman with no known history of chronic disease, vasculitis or rheumatological disease and drug use presented to the outpatient clinic with complaints of numbness up to the level of both ankles and weakness in the right foot, for two days. She did not describe a history of previous URTI, AGE, surgery or vaccination. On neurologic examination, the right ankle dorsiflexion was 0/5, plantar flexion was 4/5, hypoesthesia was present, up to the level of both ankles, sole skin response was absent bilaterally, deep tendon reflexes were normoactive. The patient, who was thought to be GBS or one of its variant syndromes based on anamnesis and examination findings. Cranial and spinal imaging studies revealed no significant pathology to explain the complaints. A lumbar puncture was performed, cerebrospinal fluid (CSF) analysis demonstrated normal glucose and sodium levels, with an absence of leukocytes and erythrocytes. The microprotein level was measured at 54 mg/dL. These clinical findings were consistent with albuminocytologic dissociation. An electromyography (EMG) nerve conduction study and needle EMG were performed and the results were compatible with AMSAN. Table 1 demonstrates the results of the nerve conduction study (NCS). Intravenous immunoglobulin (IVIG) treatment at a dose of 0.4 g/kg/day for 5 days was given to the patient who was diagnosed with AMSAN with the current clinical and laboratory findings.

Table 1. Nerve Conduction Study (NCS) Data

Nerve Motor Studies	Site / Segment	Latency (ms)	Amplitude	NCV (m/s)	F-wave (ms)
Median (R)	Wrist	3.76	10.17 mV	-	-
	Elbow	7.38	8.82 mV	58.0	
Ulnar (R)	Wrist	2.40	9.97 mV	-	-
	Elbow	5.00	9.79 mV	65.4	
	Axilla	6.34	9.16 mV	78.4	56.0
Tibial (L)	Ankle	3.85	2.87 mV	-	
	Popliteal	13.00	1.86 mV		54.0
Tibial (R)	Ankle	4.40	1.85 mV	40.4	
	Popliteal	13.80	1.59 mV	-	58.0
Peroneal (L)	Ankle	3.25	1.80 mV	40.7	
	Head of Fibula	12.35	1.46 mV		60.0
Peroneal (R)	Ankle	4.10	2.41 mV	-	
	Head of Fibula	11.30	1.40 mV	35.4	
	Popliteal	14.25	0.20 mV	20.3	
Sensory Studies					
Median (R)	Wrist	2.64	14.40 uV	53.0	-
Ulnar (R)	Wrist	2.42	14.40 uV	57.9	-
Peroneal Sup. (L)	Distal	-	Not Obtainable	-	-
Peroneal Sup. (R)	Distal	-	Not Obtainable	-	-
Sural (L)	Distal	-	Not Obtainable	-	-
Sural (R)	Distal	-	Not Obtainable	-	-

ms: Milliseconds, mV: Millivolts, µV: Microvolts, m/s: Meters per second, L: Left, R: Right

Biochemistry, hemogram, thyroid autoantibodies, tumour markers, vasculitic markers, B12 level, viral serology, syphilis antibodies were sent for the etiology of AMSAN. Among the test results, only the patient's CRP level was found to be elevated. A thoracic and abdominal computed tomography (CT) and echocardiography were performed to investigate the focus in the patient who had high CRP level, but no infection clinic. The thorax CT showed multiple cavitary lesions as the pathologic findings. Tests for tuberculosis, fungus, malignancy and vasculitic events that may have caused the multiple cavitary lesions in the lung were requested. p-ANCA, which was negative among the vasculitis markers sent for the first time, was positive when sent for the second time. In the light of the present findings, the patient was diagnosed as having GPA. The patient, whose clinical findings improved following IVIG therapy with no observed progression or additional treatment, was discharged with a scheduled outpatient follow-up. Additionally, the patient was referred to the rheumatology department for the management and follow-up of GPA. Due to the patient's rheumatological follow-up at an external facility, the exact immunosuppressive regimen initiated was unavailable.

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Discussion

Acute motor and sensory axonal neuropathy is a form of GBS that causes motor and sensory symptoms and is characterised by axonal damage rather than demyelination of peripheral nerves. Albuminocytologic dissociation can be observed in the cerebrospinal fluid (CSF) of patients. Nerve conduction studies may show conduction block, slowing of nerve conduction velocity and prolongation of latency, and acute denervation findings in needle EMGs. In the early stage of the disease, laboratory findings may be completely normal. In addition to infections, operations, malignancies and some rheumatologic diseases are also seen as etiologic causes of GBS. Although it has been reported in the literature that GBS may occur in the course of vasculitis, it is controversial as to whether vasculitis causes GBS or whether common immune mechanisms trigger both diseases [7-10].

Involvement of the nervous system in GPA is 20-50% and peripheral or cranial neuropathy may be observed during the course of the disease. The presence of neurologic symptoms as an initial symptom is quite rare. Peripheral nervous system involvement in GPA may be manifested as polyneuropathy, radiculopathy and mononeuritis multiplex. The pathological findings of vasculitic polyneuropathy (VP) are characterized by seconder axonal degeneration caused by vasculitis-induced ischemia [5,6]. Mononeuritis multiplex is more common than symmetric polyneuropathy (12-15%), with predominant involvement of the lower extremities [5,6].

It is important to differentiate between vasculitic polyneuropathy, which may involve the peripheral nervous system commonly affected by vasculitis, and GBS. In addition to typical EMG findings suggestive of GBS, the clinical presentation of the patient with acute/subacute ascending sensorimotor weakness, and the absence of systemic involvement characteristic of vasculitis, play a crucial role in the differential diagnosis between GBS and vasculitic neuropathy. While axonal degeneration in vasculitic neuropathy is secondary to vasculitis-induced ischemia, the axonal damage in AMSAN is directly caused by the autoimmune response itself. [11].

The patient presented with acute/subacute ascending sensorimotor symptoms originating in the lower extremities. This clinical picture, combined with EMG findings of sensorimotor axonal damage and conduction block in the peroneal nerve, as well as the detection of albuminocytologic dissociation in the CSF, primarily suggested AMSAN. Notably, the patient's medical history was negative for any neuropathic complaints prior to admission.

During the etiological workup, elevated sedimentation rates and acute-phase reactants, along with cavitary lesions in the lungs, raised clinical suspicion for malignancy and vasculitis. Following a positive result on a repeat p-ANCA test, the patient was diagnosed with GPA. A positive clinical response was observed, with improvement in neurological symptoms following IVIG treatment. In light of these findings, it was considered that AMSAN and GPA may have coexisted, rather than the presentation being a manifestation of vasculitic neuropathy alone. A review of the literature revealed only a very limited number of cases where these two conditions occurred concurrently.

Gutzler et al. stated that they did not see granulomatous formation around the neurons in the pathological examination they performed in their 49-year-old patient who had GPA and GBS association, but that there may be a pathogenetic connection between the two diseases [12]. While membranous glomerulonephritis may be observed in GBS, whereas Kenan et al. reported a case with p-ANCA-associated nephrotic syndrome and GBS. The association of both diseases suggests the presence of a common immunologic mechanism, but further studies are needed to evaluate the importance of p-ANCA [11]. On the other hand, Nagashima et al. described a case of p-ANCA positive GPA characterised with central nervous system involvement characterised with granulomatous pachymeningitis in the pathological materials of a 53-year-old female patient who presented with various neurological symptoms for two years and was found to have hypertrophic pachymeningitis and multiple cranial neuropathy [13]. Nakamura et al. performed a study on 6 patients with Churg-Strauss syndrome and neuropathy and found that two patients had GBS and a sural nerve biopsy revealed eosinophilic lymphocytic infiltration in the small artery and extravascular connective tissue. In the same study, it was shown that patients with vasculitic neuropathy had necrotising vasculitis findings and the presence of secondary demyelination after vasculitis-related ischemia [14]. Ueda et al. reported that they detected c-ANCA in a 57-year-old patient with a history of asthma and pansinusitis after GBS was detected, they diagnosed CSS in this patient and found perivascular lymphocytic infiltration with myelin loss in the sural nerve biopsy, and they improved the clinical symptoms of the patient with steroid, IVIG and immunosuppressant combined treatment [15].

Vasculitis should always be part of the differential diagnosis of GBS because first-line treatments may differ. Steroids are not beneficial in GBS and may even worsen the clinical symptoms, but they are the mainstay of vasculitis treatment. On the other hand, IVIG and plasmapheresis may be effective in both vasculitis and GBS (18). The first-line treatment of choice in AAV is high-dose corticosteroids, not IVIG. Immunosuppressive therapy (cyclophosphamide) combined with steroids should be considered when corticosteroids are ineffective or in cases with severe symptoms or antibodies. IVIG may be a treatment option for patients with severe disability in the chronic phase [14].

Conclusion

In this case, GPA and GBS coexistence is presented as a very rare case; the clinical features and pathogenesis of GPA and GBS coexistence have not yet been clarified in the literature. Since the prognosis depends on the correct diagnosis and appropriate choice of treatment, there is a need for further research to investigate the clinical and experimental aspects of these patients.

DOI: 10.5455/medscience.2025.10.289

Conflict of Interests
The authors declare that there is no conflict of interest in the study.

Financial Disclosure
The authors declare that they have received no financial support for the study.

Patient Informed Consent
A signed consent form was obtained from the patient on 10.06.25.

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