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CASE REPORT

POEMS Syndrome with Peripheral Neuropathy as the First Presentation: A Case Report

Yujie Di[1,*], Chongmei Liu[2,*]

[1]Department of Internal Medicine, Yueyang Hospital Affiliated to Hunan Normal University, Yueyang, Hunan, 414000, People's Republic of China; [2]Department of Pathology, Yueyang Hospital Affiliated to Hunan Normal University, Yueyang, Hunan, 414000, People's Republic of China

*These authors contributed equally to this work

Correspondence: Chongmei Liu, Department of Pathology, Yueyang Hospital Affiliated to Hunan Normal University, No. 263 Bayue Road, Yueyang, Hunan, 414000, People's Republic of China, Email locf79@163.com

Introduction: POEMS syndrome is a rare multisystem plasma cell disorder, often misdiagnosed when peripheral neuropathy is the initial manifestation. This report illustrates the effectiveness of autologous hematopoietic stem cell transplantation (ASCT) combined with lenalidomide that reversed neurological deficits, supported by objective electrophysiological evidence rarely documented.

Case Presentation: A 54-year-old Chinese woman presents with progressive bilateral lower extremity weakness, numbness, and edema. Initially misdiagnosed as suspected spinal metastases, POEMS syndrome was later confirmed by elevated serum IgG-λ M protein (2.57 g/L) and VEGF (556.74 pg/mL). Treatment follows a three-phase regimen: induction with lenalidomide (25 mg/day, days 1–21) plus dexamethasone (20 mg/day pulsed); ASCT with cyclophosphamide mobilization, melphalan conditioning (300 mg), and $CD34^+$ cell infusion (5.18×10^6/kg); and maintenance with lenalidomide consolidation (25 mg/day, days 1–14 monthly).

Results: Significant symptom resolution occurred post-treatment. Serum M-protein and VEGF normalized. Complete remission (CR) was sustained over 24 months. Repeat electromyography demonstrated sensory nerve amplitude increased 2–4 fold (eg, left sural nerve: 2.2 → 9.0 μV) and motor conduction velocity improved >50% in lower limbs (eg, left tibial nerve: 15.8 → 36.7 m/s).

Conclusion: This case demonstrates that ASCT-lenalidomide combination achieves dual neurological and hematological remission in POEMS syndrome. Early VEGF testing and multidisciplinary collaboration are critical to prevent diagnostic delays. Quantitative EMG improvement provides objective evidence for treatment efficacy assessment.

Keywords: POEMS syndrome, peripheral neuropathy, electromyography, autologous hematopoietic stem cell transplantation, lenalidomide

Introduction

POEMS syndrome is a monoclonal plasma cell proliferative disorder and a paraneoplastic syndrome associated with abnormal plasma cell proliferation. Due to its rarity, multisystem involvement, and high clinical heterogeneity, early diagnosis poses significant challenges, and it is frequently misdiagnosed because of its neuropathy-predominant onset. Although peripheral neuropathy as a first presentation is a recognized feature, detailed longitudinal electromyography (EMG) data documenting objective neurological recovery after treatment are rarely reported. This article reports a case of POEMS syndrome presenting with peripheral neuropathy as the initial manifestation, demonstrating how early VEGF testing and ASCT-based therapy reverse neurological deficits. We herein describe the diagnostic process to explore its clinical characteristics and management approach, followed by an analysis of prognosis, long-term management, and future directions.

Case Presentation

A 54-year-old female patient presented to an external hospital in January 2022 with intermittent claudication. PET-CT suggested possible lumbar metastasis, but lumbar biopsy revealed no malignant evidence. Symptoms subsequently

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Table 1 Key Diagnostic Findings at Presentation

Category	Key Findings
Serology	IgG-λ M-protein positive (2.57 g/L); VEGF 556.74 pg/mL
Urinalysis	Urine free kappa light chain 102.0 mg/L (Ref: 0.39–15.10)
Bone Marrow	Clonal plasma cells detected by flow cytometry (<5%)
Imaging	PET-CT: Lumbar hypermetabolic focus (biopsy negative); Chest CT: Stable left lung nodule; Abdominal Ultrasound: Hepatosplenomegaly
Neurophysiology	Bilateral lower limb peripheral neuropathy with demyelination and axonal damage
Endocrinology	Impaired glucose tolerance, primary hypothyroidism
Dermatology	Diffuse skin hyperpigmentation

worsened, manifesting as bilateral lower limb weakness, numbness, and edema, exacerbated by activity, accompanied by night sweats. Lower limb edema was worse in the evening. EMG performed externally revealed: bilateral lower limb peripheral neuropathy, primarily demyelinating changes with concomitant axonal involvement, affecting both proximal and distal segments, with involvement of the deep sensory pathway from the lower limbs to the cerebral cortex. Further investigations, including bone marrow aspiration and biopsy, detected a minor population of plasma cells by flow cytometry. Immunofixation electrophoresis was positive for IgG λ-type M-protein. A plasma cell dyscrasia-related disorder was suspected. The patient was subsequently referred to our hospital for further evaluation.

Key Investigation Results (Table 1): Upon referral, a detailed clinical examination revealed additional findings consistent with POEMS syndrome. The patient had diffuse skin hyperpigmentation, most pronounced on the lower extremities and trunk. Endocrine evaluation demonstrated abnormalities in multiple axes: impaired glucose tolerance (fasting glucose 6.3 mmol/L, 2-hour postprandial glucose 11.2 mmol/L) and primary hypothyroidism (TSH 8.5 mIU/L, free T4 0.7 ng/dL). Abdominal ultrasound revealed mild hepatomegaly (span 16 cm) and moderate splenomegaly (span 14 cm). There was no evidence of lymphadenopathy.

Based on the presence of polyneuropathy (mandatory), monoclonal plasma cell proliferation (mandatory), elevated VEGF (major), endocrinopathy (minor), organomegaly (minor), skin changes (minor), and extravascular volume overload (edema, minor), the patient fulfilled the diagnostic criteria for POEMS syndrome.

After obtaining informed consent, systemic treatment was initiated as follows:

Induction Therapy (April 4, 2022): Lenalidomide 25 mg/day (days 1–21) + Dexamethasone 20 mg/day (days 1, 2, 8, 9, 15, 16, 21, 22), supported by gastroprotective and hepatoprotective agents. Partial symptomatic relief was achieved.

Mobilization/Collection & ASCT (May 31 - July 8, 2022): Mobilization with Cyclophosphamide (3100 mg total, 50mg/kg/day on days 1–2). Peripheral blood stem cell collection yielded CD34+ cells: 5.18×10^6/kg. The patient was admitted to a laminar flow room. Conditioning with Melphalan (300 mg) was followed by autologous stem cell infusion. The procedure was uneventful.

Maintenance Therapy (Starting October 2022): Lenalidomide (25 mg/day, days 1–14) combined with Dexamethasone (20.25 mg/day on days 1, 8, 15) to consolidate efficacy.

Follow-up: Assessments in October 2022, April 2023, and April 2024 consistently showed CR with sustained symptomatic improvement. VEGF and M-protein became undetectable The skin hyperpigmentation gradually faded, and the hepatosplenomegaly resolved on follow-up imaging. The patient's endocrine function partially improved, with normalization of blood glucose levels and a reduced requirement for thyroid hormone replacement.

Discussion

POEMS syndrome is a complex and rare hematological disorder. The acronym, proposed by Bardwick et al,[1] originates from five key clinical features: Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein (M-protein), and Skin changes. It is characterized by peripheral nerve demyelination and monoclonal plasma cell proliferation, with an estimated prevalence of approximately 0.3 per 100,000 individuals.[2] A definitive diagnosis requires meeting two

mandatory criteria (polyneuropathy and monoclonal plasma cell disorder), at least one major criterion (including elevated VEGF), and at least one minor criterion (organomegaly, endocrinopathy, skin changes, etc).[3]

Overproduction of vascular endothelial growth factor (VEGF) is a central feature of POEMS syndrome, contributing to peripheral nerve damage through disruption of the blood-nerve barrier.[4,5] In our patient, the initially elevated VEGF level (556.74 pg/mL) normalized after treatment, correlating with significant clinical and electrophysiological improvement.

The detailed quantitative EMG findings in this case (Table 2 and Figures 1–4) provide objective evidence of neurological recovery following ASCT and lenalidomide therapy. Improvements were observed not only in distal lower limb nerves but also in proximal segments, suggesting a global reversal of neuropathic processes. This degree of electrophysiological documentation is uncommon in case reports and underscores the potential for nerve regeneration even in severe neuropathy.

Table 2 Comparison of Pre-Treatment (2022) and Post-Treatment (2024) Electromyography (EMG) Findings

Nerve & Measurement	2022 Left	2024 Left	2022 Right	2024 Right	Distance (mm)	2022 CV (m/s)	2024 CV (m/s)
Sensory Nerve Conduction							
Median S. (Digit II - Wrist)							
Latency (ms)	2.88	2.53	2.77	2.58	130		
Amplitude (μV)	27.5	37.0	17.3	28.4		45.1	51.4
Ulnar S. (Digit V - Wrist)							
Latency (ms)	2.19	1.95	2.06	1.96	100		
Amplitude (μV)	23.0	31.3	24.3	31.2		45.7	51.3
Superficial Peroneal S. (Mid/Low Leg Lat. - Ankle)							
Latency (ms)	2.36	2.10	2.29	1.95	85.0		
Amplitude (μV)	5.8	7.4	5.1	8.8		36.0	40.5
Sural S. (Mid/Low Leg Post. - Lat. Malleolus)							
Latency (ms)	3.46	2.95	3.10	2.87	130		
Amplitude (μV)	2.2	9.0	3.1	8.6		37.6	44.1
Motor Nerve Conduction							
Median M. (Wrist - APB)							
Distal Latency (ms)	3.60	3.38	4.26	4.29			
Amplitude (mV)	9.4	11.2	10.9	11.3			
Median M. (Elbow - Wrist)							
Proximal Latency (ms)	7.88	7.19	8.60	8.57	220		
Amplitude (mV)	9.3	10.3	9.9	11.2		51.4	57.7
Ulnar M. (Wrist - ADM)							
Distal Latency (ms)	2.29	2.40	2.48	2.25			
Amplitude (mV)	8.7	8.7	9.1	8.3			
Ulnar M. (Elbow - Wrist)							
Proximal Latency (ms)	6.72	6.35	6.88	6.31	230		
Amplitude (mV)	8.3	8.4	8.4	8.8		51.9	58.2
Common Peroneal M. (Ankle - EDB)							
Distal Latency (ms)	5.36	4.26	5.72	3.65			
Amplitude (mV)	0.11	0.83	0.26	1.16			
Common Peroneal M. (Fib Head - Ankle)							
Proximal Latency (ms)	18.5	13.8	16.2	12.0	300		
Amplitude (mV)	0.031	0.57	0.14	0.66		22.8	31.4
Tibial M. (Ankle - AH)							
Distal Latency (ms)	8.95	5.60	7.76	5.72			
Amplitude (mV)	0.013	0.25	0.015	0.20			

(*Continued*)

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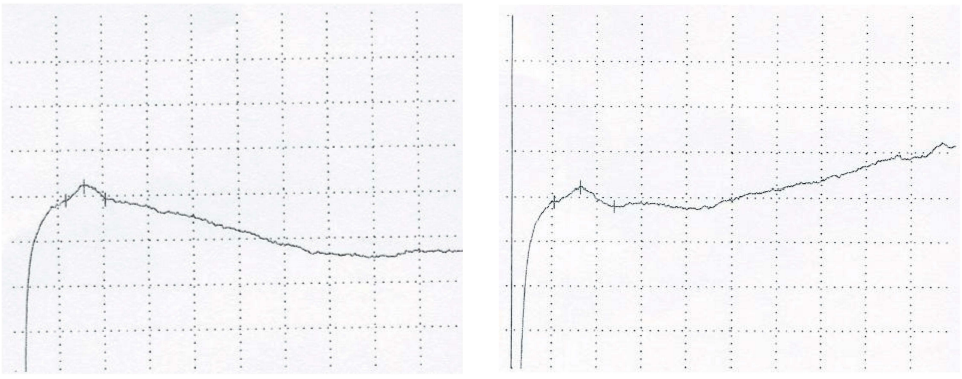
Table 2 (Continued).

Nerve & Measurement	2022 Left	2024 Left	2022 Right	2024 Right	Distance (mm)	2022 CV (m/s)	2024 CV (m/s)
Tibial M. (Knee - Ankle)							
Proximal Latency (ms)	31.8	15.4	21.1	15.4	360		
Amplitude (mV)	0.048	0.20	0.057	0.43		15.8	36.7
Radial M. (Above Elbow - EDC)							
Latency (ms)	2.90	3.29	2.78	3.35			
Amplitude (mV)	5.5	6.9	5.1	5.9			
Musculocutaneous M. (Erb's - Biceps)							
Latency (ms)	5.29	4.15	5.21	4.25			
Amplitude (mV)	3.0	6.0	3.4	6.9			
Axillary M. (Erb's - Deltoid)							
Latency (ms)	3.85	3.17	3.81	3.59			
Amplitude (mV)	8.3	14.3	8.5	15.4			
Femoral M. (Groin - Vastus Med.)							
Latency (ms)	6.02	4.44	5.05	4.46			
Amplitude (mV)	3.3	5.8	4.4	6.0			

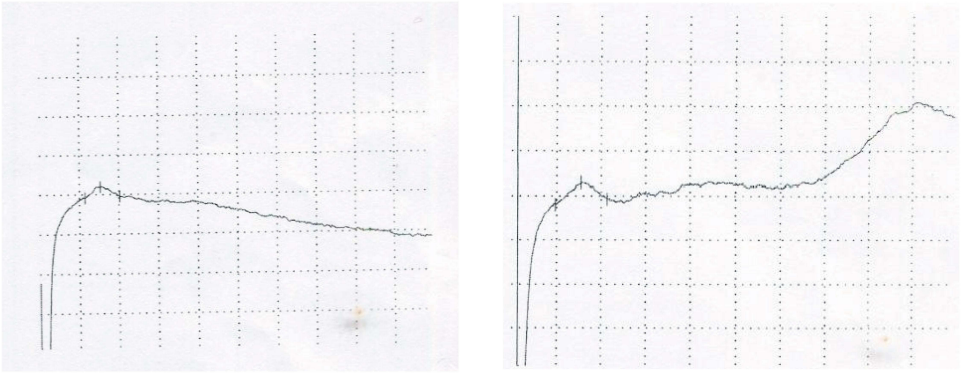
While the combination of ASCT and lenalidomide is not novel as a concept, this case highlights the importance of a sequential, well-documented approach. ASCT serves as a potent debulking therapy, while lenalidomide maintenance may help suppress residual clones and prevent relapse.[6,7] Recent reviews have further supported the efficacy of this treatment paradigm in POEMS syndrome.[8,9] However, lenalidomide, while considered less neurotoxic than thalidomide, still carries a potential risk of peripheral neuropathy, necessitating close neurological monitoring.[10] In this patient, no worsening of neuropathy was observed, and instead, continued improvement was noted during maintenance, suggesting a favorable risk-benefit profile. Long-term follow-up studies indicate that sustained remission is achievable with such approaches, though late relapse remains a consideration.[11]

This report has several limitations. As a single case study, the findings are not generalizable to all POEMS patients. The follow-up period of 24 months, while showing sustained remission, is insufficient to assess long-term outcomes such as late relapse or secondary malignancies. Additionally, the relative contributions of ASCT versus lenalidomide maintenance to the observed neurological recovery cannot be definitively separated. Standardized neuropathy scales and minimal residual disease assessment were not employed, which would have provided additional granularity. As a single case, the findings require validation in larger cohorts.



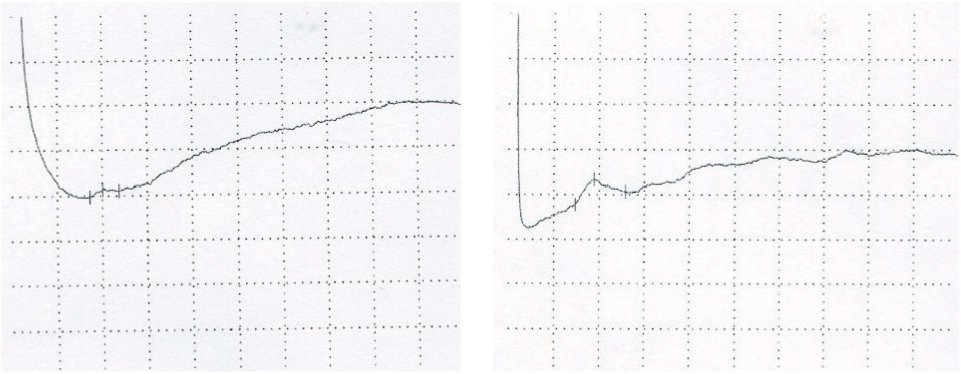
2022 Left superficial peroneal sensory nerve 2024Left superficial peroneal sensory nerve

Figure 1 Left superficial peroneal sensory nerve (Mid/Low lateral leg - Ankle; 20 μV/division, 2 ms/division).



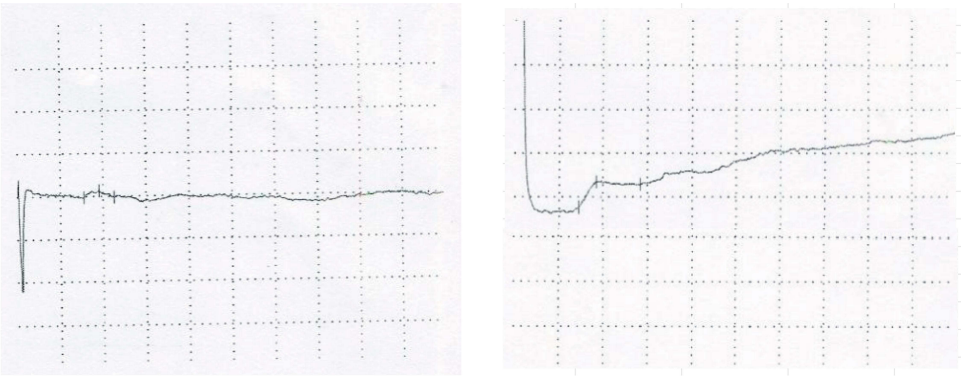
2022 Right superficial peroneal sensory nerve 2024Right superficial peroneal sensory nerve

Figure 2 Right superficial peroneal sensory nerve (Mid/Low lateral leg - Ankle; 20 μV/division, 2 ms/division).



2022Left sural sensory nerve 2024Left sural sensory nerve

Figure 3 Left sural sensory nerve (Mid/Low posterior leg - Lateral malleolus; 20 μV/division, 2 ms/division).



2022Right sural sensory nerve 2024Right sural sensory nerve

Figure 4 Right sural sensory nerve (Mid/Low posterior leg - Lateral malleolus; 20 μV/division, 2 ms/division).

Differential diagnosis is crucial for POEMS syndrome. Its peripheral neuropathy must be distinguished from chronic inflammatory demyelinating polyneuropathy (CIDP), which typically lacks M-protein and multisystem involvement. Sclerotic bone lesions need differentiation from multiple myeloma, which primarily causes lytic bone destruction and is often associated with hypercalcemia and renal insufficiency.[12] This patient was initially suspected of having metastatic

bone disease, highlighting the diagnostic challenge and the value of a multidisciplinary approach in recognizing the constellation of symptoms.

Conclusion

This case illustrates a favorable outcome in a POEMS syndrome patient presenting with peripheral neuropathy, treated effectively with ASCT and lenalidomide induction/maintenance. The detailed EMG data provide valuable objective evidence of neurological recovery. However, clinicians should recognize that such outcomes may not be representative of all patients. The case underscores the need for a high index of suspicion for POEMS in patients with unexplained peripheral neuropathy, especially when accompanied by seemingly unrelated systemic findings. In particular, in patients with unexplained demyelinating polyneuropathy and systemic features, serum VEGF screening should be considered as a valuable diagnostic tool. Early VEGF testing and multidisciplinary collaboration are critical to prevent diagnostic delays. Future research should focus on optimizing patient selection for ASCT and defining the role of maintenance therapy, with careful consideration of individual patient risk factors.

Abbreviations

APB, Abductor Pollicis Brevis; ADM, Abductor Digiti Minimi; EDB, Extensor Digitorum Brevis; AH, Abductor Hallucis; EDC, Extensor Digitorum Communis; Lat., Latency; Amp., Amplitude; Dist., Distance; CV, Conduction Velocity; S, Sensory; M, Motor; Post., Posterior; Lat., Lateral/Lateralis; Med., Medialis; Ref., Reference.

Data Sharing Statement

The original contributions presented in the study are included in the article. Further inquiries can be directed to the corresponding author.

Ethical Approval Statement

Approval was obtained from the Ethics Committee of Yueyang Hospital Affiliated to Hunan Normal University for the publication of this case report. Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Consent for Publication

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

The authors declare that they have no conflicts of interest in this work.

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