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Rare POEMS Syndrome With Multicentric Castleman Disease Presenting As Demyelinating Polyneuropathy In Young Male: A Case Report

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Abstract
POEMS syndrome is a rare paraneoplastic disorder characterized by polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder, and skin changes. Its association with multicentric Castleman disease can worsen prognosis and complicate diagnosis

Keywords: POEMS syndrome, Castleman disease12, polyneuropathy5, demyelination, paraneoplastic syndrome

Introduction

POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal plasma cell disorder, and Skin changes) is a rare paraneoplastic syndrome linked to plasma cell dyscrasias. Its clinical heterogeneity often leads to delayed diagnosis. Coexisting multicentric Castleman disease (MCD) is seen in a subset of patients and contributes to systemic manifestations via cytokine dysregulation. Early recognition is crucial for initiating timely treatment.[1]

Case:

A 37-year-old male presented with one month of difficulty passing urine, constipation, bleeding oral ulcers, low-grade fever, and progressive lower limb weakness with gait imbalance. Initial urinary retention required catheterization. Over the next two weeks, he developed worsening constipation, mouth ulcers with bleeding, and gradual weakness of both lower limbs. Subsequently, he experienced gait imbalance and stool incontinence.

On examination, he had palpable bilateral axillary lymphadenopathy, fever (101°F), tachycardia (110/min), and multiple bleeding oral ulcers. Neurological examination showed power of 5/5 in upper limbs and 2/5 in lower limbs without cranial nerve involvement.

Laboratory investigations revealed progressive anemia (Hb 9.2→8.4 g/dL), thrombocytopenia (platelets 80,000→68,000), elevated ESR (95 mm/hr), ferritin (2000 ng/mL), and CRP (4.7 mg/L). Electrolytes, renal and liver function tests were within normal limits. HIV, HBsAg, and HCV was negative.

MRI brain and lumbar spine showed demyelinating lesions; nerve conduction studies revealed moderate-to-severe sensory-motor axonal polyneuropathy in both lower limbs. CSF analysis was unremarkable with normal protein and glucose, and infectious workup was negative. PET-CT demonstrated multiple hypermetabolic lymph nodes. Right axillary lymph node biopsy and sural nerve biopsy confirmed multicentric Castleman disease and demyelinating features consistent with POEMS syndrome. Bone marrow biopsy supported monoclonal plasma cell proliferation.

A final diagnosis of POEMS syndrome with multicentric Castleman disease was established. Treatment was initiated with IV meropenem, doxycycline, and pulse methylprednisolone (1 g daily for 5 days), followed by IV tocilizumab. The patient showed significant clinical improvement, with gradual recovery of lower limb strength and resolution of systemic symptoms.

3. Discussion:

POEMS syndrome is a rare multisystem disorder with heterogeneous presentation. Neuropathy, often the first manifestation, can progress rapidly to severe weakness. The presence of lymphadenopathy and biopsy-proven Castleman disease suggested systemic involvement. Elevated cytokines, including VEGF and IL-6, play a pivotal role in pathogenesis, contributing to polyneuropathy and systemic features. [4,5,7]

This case illustrates that early consideration of POEMS syndrome in patients with unexplained polyneuropathy, systemic inflammatory signs, and organomegaly—even in young adults—can facilitate timely diagnosis. Prompt initiation of corticosteroids and targeted anti-IL-6 therapy (tocilizumab) led to substantial symptomatic improvement, consistent with emerging treatment paradigms. [2,8]

This case emphasizes the importance of recognizing POEMS syndrome in patients presenting with polyneuropathy and systemic features. Coexisting multicentric Castleman disease should raise suspicion and prompt further evaluation. Early diagnosis and appropriate immunosuppressive therapy can improve outcomes in this rare but serious condition.

Conclusion: This case highlights the need to consider POEMS syndrome in patients with unexplained neuropathy and systemic features, even in young adults.

Early diagnosis and timely initiation of immunosuppressive therapy are crucial for improving clinical symptoms

Figures

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Figure 1. MRI Brain-Few Small Hyperintense Signal in the Bilateral Periventricular White Matter

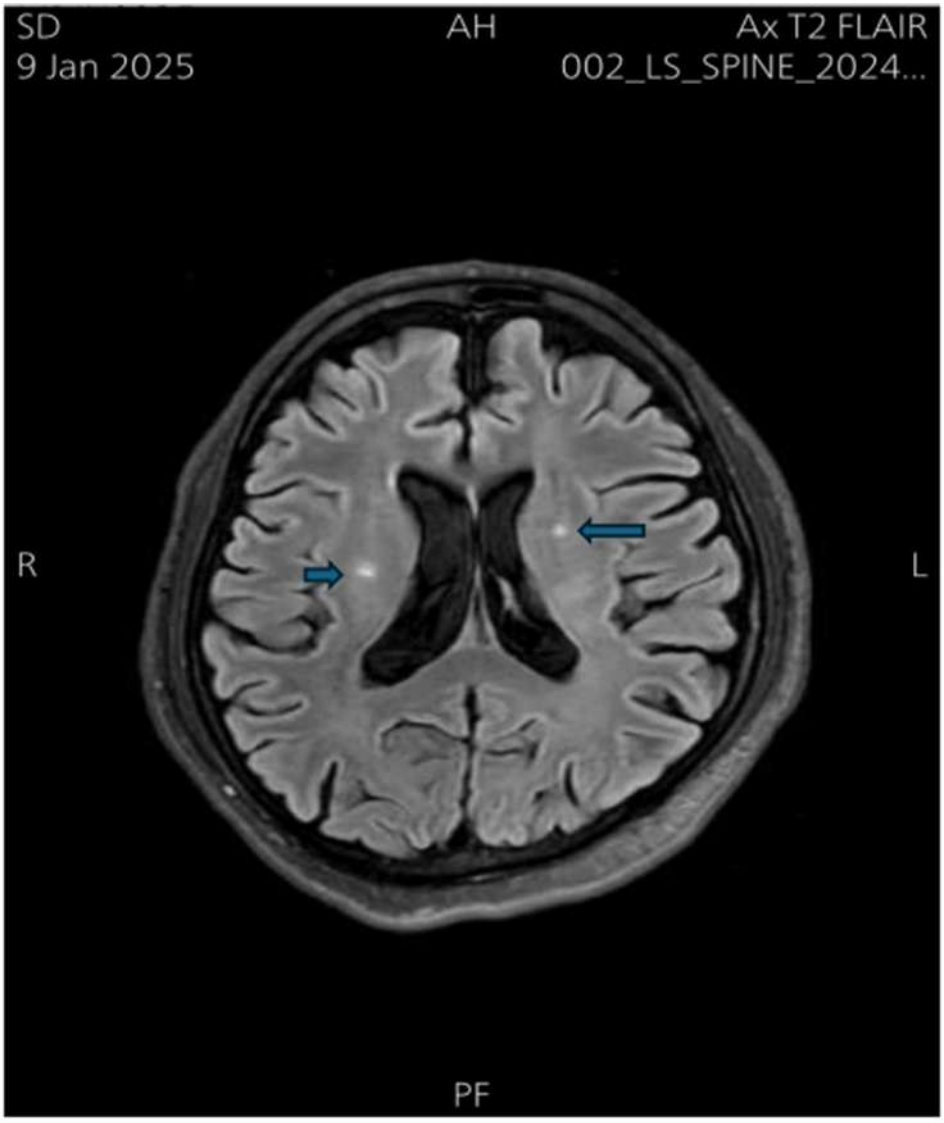

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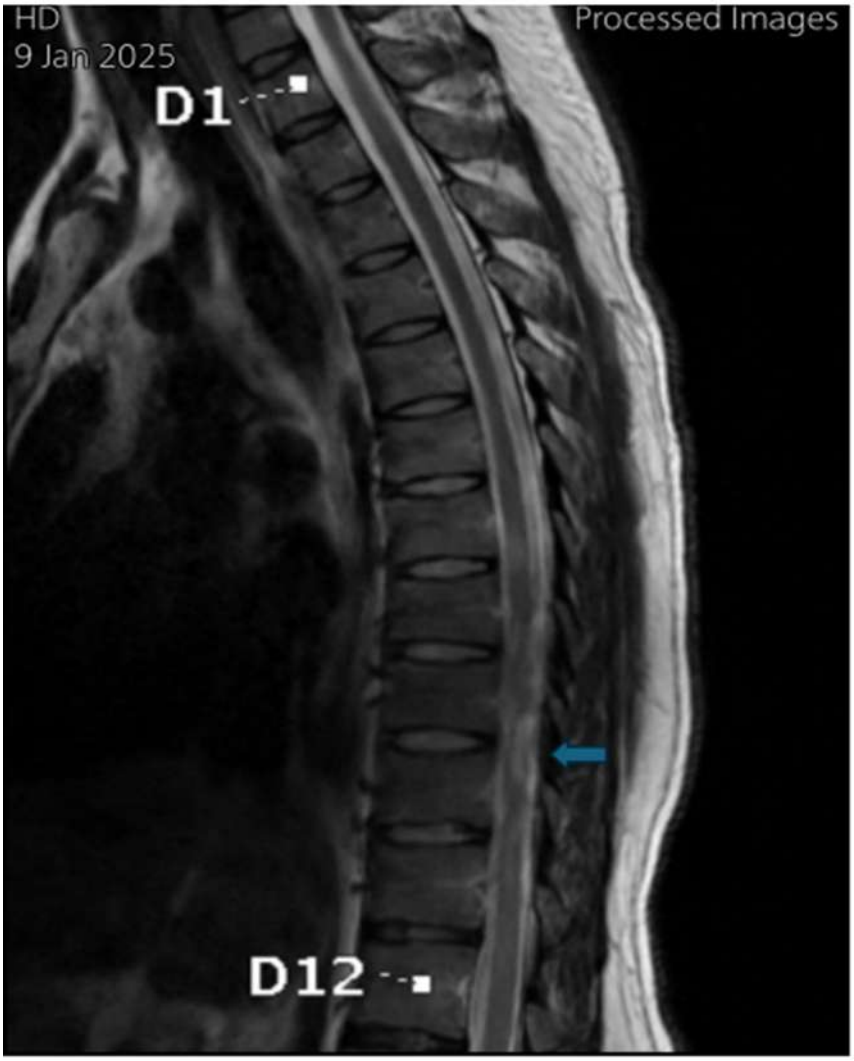
Figure 2.MRI Lumbosacral Spine -Focal Intramedullary Hyperintense Signal in the Spinal Cord at D7 Vertebral Level. Possibility Of Demylination Is Likely



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