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## Recurrent Cervical Lymphadenopathy with Peripheral Neuropathy – A Case Report of POEMS syndrome

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### Abstract

POEMS syndrome, which stands for polyneuropathy, organomegaly, endocrinopathy, M-protein, and skin changes, is a rare type of demyelinating neuropathy associated with plasma cell disorders and increased levels of vascular endothelial growth factor (VEGF). This syndrome is a severely debilitating and often fatal condition. The progressive nature of polyneuropathy can lead to tetraplegia, as well as significant pleural effusions and ascites, ultimately resulting in multiorgan failure. The unusual presentation of these symptoms might lead any physician to consider alternative diagnoses. We present the case of a 60-year-old man diagnosed with POEMS syndrome, initially presenting with polyneuropathy and cervical lymphadenopathy. Early confirmation of the diagnosis proved challenging since initial biochemical tests returned normal results; however, a biopsy of the cervical lymph nodes indicated Castleman's disease. Ultimately, the diagnosis was confirmed through the detection of monoclonal immunoglobulin via serum protein electrophoresis, nerve conduction studies, and the presence of additional clinical features. The patient was diagnosed with POEMS syndrome and underwent treatment with Melphalan combined with dexamethasone, resulting in a positive outcome.

**Keywords:** POEMS Syndrome, IHC, H&E, Monoclonal Gammopathy

### Introduction

POEM syndrome, which stands for Polyneuropathy, Organomegaly, Endocrinopathy, and Monoclonal Gammopathy, is a rare and complex disorders distinguished by the presence of numerous systemic symptoms [1]. POEM syndrome consists of a collection of symptoms caused by monoclonal gammopathy, which frequently results in a variety of clinical manifestations [2]. As POEM syndrome is a rare condition, national survey conducted in Japan in 2003 showed a prevalence of approximately 0.3 per 100 000 [3,4].

Among these, recurrent cervical lymphadenopathy with peripheral neuropathy is a key clinical characteristic that poses diagnostic issues for clinicians around the world [5]. The underlying cause of POEM syndrome is unresolved but it is thought to be associated with aberrant plasma cell growth and subsequent monoclonal immunoglobulin synthesis. This causes a variety of organ dysfunctions, including lymphadenopathy, hepatosplenomegaly, and endocrinopathies such as diabetes mellitus or thyroid dysfunction [6]. While diagnostic criteria for POEM syndrome have been established, the condition's rarity and variability of clinical presentations make it difficult to recognize and manage. Furthermore, the overlap of symptoms with other systemic conditions, demands a careful evaluation to appropriately distinguish between these entities.

This case report presented a difficult clinical presentation including recurrent cervical lymphadenopathy with concomitant peripheral neuropathy, which was ultimately identified as POEM syndrome following a thorough diagnostic workup. This study emphasized the importance of diagnosing and managing this unusual condition by shining light on its clinical presentation, diagnostic method, and treatment considerations. This report also addressed early diagnosis and interdisciplinary care might improve patient outcomes and quality of life.

### Case Summary

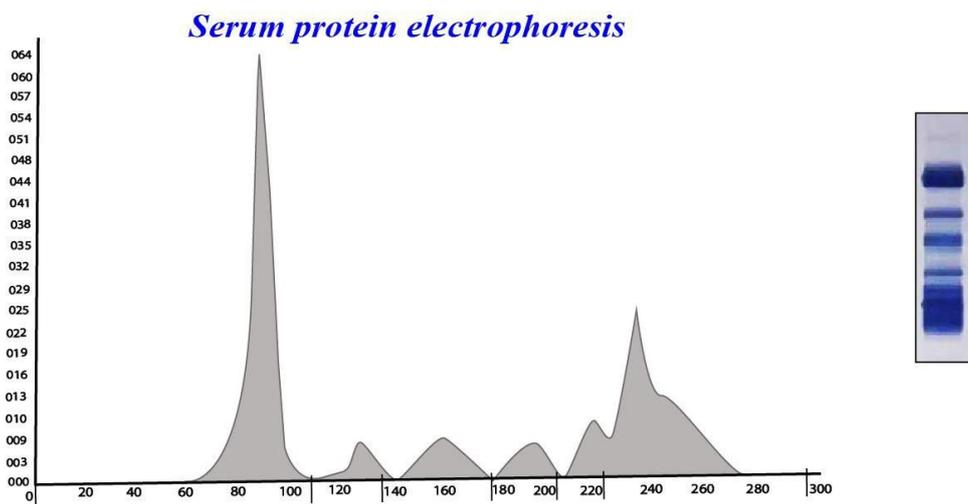
Our patient is a 60-year-old male with a past medical history of ischemic heart disease, hypertension, who complained of tingling, numbness of lower limb since about 2 weeks prior. He also complained of fever and malaise to be associated with it. Examination revealed diminished sensation up to L1, vibration and position sense was intact. Lymphadenopathy was noted in cervical region in the clinical exam. The patient's family history was unremarkable.

As the patients complained of fever and malaise, hematological assessment was done. Biochemical results did not show any specific findings.

Sl. number	Name of the test	Result	Reference Value
1	RBS	4.0 mmol/L	< 7.8 mmol/L
2	S. Creatinine	0.9 mg/dL	< 1.2 mg/dl
3	Hemoglobin	16.2 g/dL	M=12-16 F= 10-14 gm/dl
4	Total platelet count	297000/Cumm	150000-450000 / Cumm

**Table 1: Biochemical Examination**

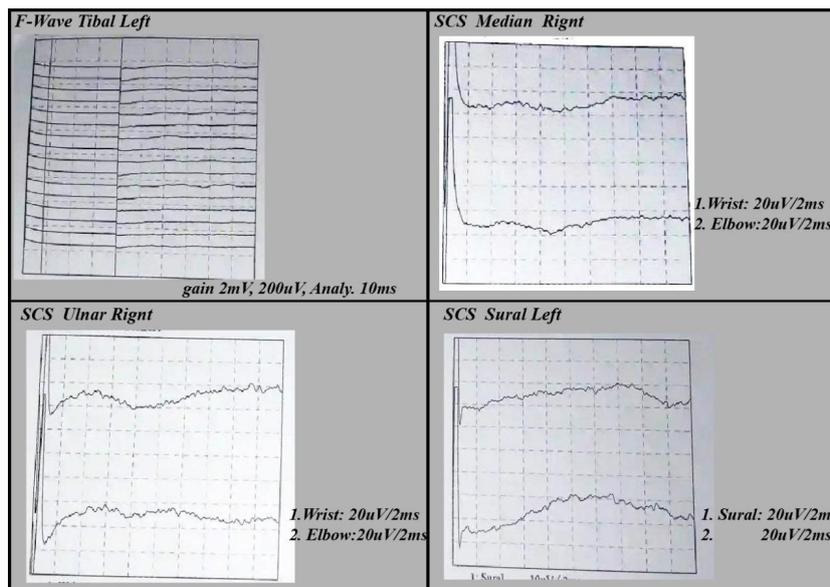
Serum protein electrophoresis was done. Which showed there was a moderate band in the gamma region which most likely represent monoclonal band. Also, Electrophysiological findings were suggestive of demyelinating sensory-motor polyneuropathy.



**Figure 1: Serum Protein Electrophoresis**

Fractions	%	Ref. %	g/l	Ref. g/l
Albumin	42.0	55.8-66.1	31.9	40.2-47.6
Alpha 1	4.5	2.9-4.9	3.4	2.1-3.5
Alpha 2	8.5	7.1-11.6	6.5	2.1-3.5
Beta 1	4.7	4.7-7.2	3.6	3.4-5.2
Beta 2	6.5	3.2-6.5	4.9	2.3-4.7
Gumma	33.8	11.1-18.8	25.7	8.0-13.5

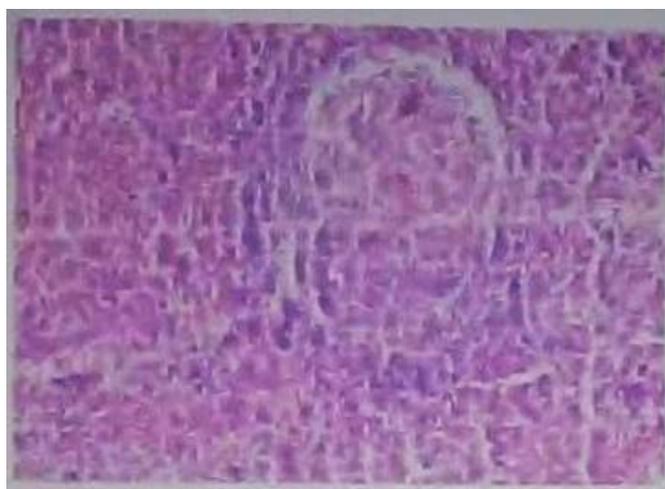
These findings lead to CT scan of abdomen, where the reports showed multiple enlarge lymph nodes (~2.0X1.8 cm & smaller) in both inguinal regions. After I/V contrast, lymph nodes showed significant contrast enhancement.



**Figure 2: Electrophysiological Findings**

Also, as cervical lymphadenopathy was found during examination, cervical lymph node biopsy was done. The report revealed an enlarged lymph node section with marked hyperplasia of lymph follicles with proliferation of blood vessels within the follicles. Hyalinization within the follicles were also seen. The interfollicular area showed marked proliferation of plasma cells with many Russel bodies. The interfollicular area also showed proliferation of blood vessels and some of the proliferative blood vessels have entered into the follicles. No malignancy was found.

In Immunohistochemical study, Castleman’s disease was confirmed along by strong positivity of CD21, CD4 and CD8 antibodies.



**Figure 3: Photomicrograph of Lymph Node Biopsy (H&E, 40X)**



**Figure 4: Photomicrograph of IHC Study of Excised Lymph Node Showing Strong Positivity of CD4, CD21, CD8 (a,b,c) Antibodies Respectively (40X)**

For the confirmation of the diagnosis, patient had 3 times radical neck dissection for cervical lymphadenopathy and was diagnosed as Castleman's Disease in histopathological examination. As there was economical constrain on the patients' side, we started Melphalan plus dexamethasone as per treatment protocol. And after getting 2 cycle treatment patient showed significant improvement.

## Discussion

The precise mechanism underlying the pathogenesis of POEMS syndrome remains unclear, but cytokines could be a significant factor. The elevated levels of proangiogenic and proinflammatory cytokines, including interleukin-1 (IL-1), IL-6, IL-12, tumor necrosis factor (TNF), and particularly vascular endothelial growth factor 165 (VEGF165), are fundamental components in its pathophysiology. VEGF, produced by plasma cells and various other cells such as macrophages, osteoblasts, and megakaryocytes, targets endothelial cells and is responsible for a rapid and reversible increase in vascular permeability. The elevation of serum VEGF levels may contribute to certain clinical features of POEMS, including edema, neuropathy, organomegaly, and increased microvascular permeability of blood vessels leading to endometrial edema. Research indicates that VEGF levels are typically 5 to 10 times higher in individuals with POEMS syndrome [7-9].

The diagnosis of the POEMS syndrome is based on a combination of clinical and biological features. It requires the presence of one of the two mandatory major criteria, one of the three other major criteria and at least one of the six minor criteria [1].

### Criteria of the Diagnosis of POEMS Syndrome (2014) are- Mandatory Major Criteria

- Polyneuropathy (typically demyelinating)
  - Monoclonal plasma cell-proliferative disorder (almost always  $\lambda$ )
- Other major criteria (One required)
- Castleman disease.
  - Sclerotic bone lesions.
  - Vascular endothelial growth factor elevation.

### Minor Criteria

- Organomegaly (splenomegaly, hepatomegaly, or lymphadenopathy).
- Extravascular volume overloads (edema, pleural effusion, or ascites).
- Endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, pancreatic).
- Skin changes (hyperpigmentation, hypertrichosis, glomeruloid hemangiomas, plethora, acrocyanosis, and flushing, white nails).
- Papilledema.
- Thrombocytosis/ polycythemia.

### Other Symptoms and Signs

Clubbing, weight loss, hyperhidrosis, pulmonary hypertension/restrictive lung disease, thrombotic diatheses, diarrhea, low vitamin B12 values [1].

The polyneuropathy is the main feature of the syndrome. Patients usually present with a subacute, symmetrical, distal sensorimotor neuropathy, often painful. It usually begins as a sensory neuropathy and progresses to motor symptoms. The lower limbs are affected earlier, and more severely, than the upper limbs. Clinical examination may reveal distal wasting, weakness and sensory loss affecting both large and small fiber sensory modalities [1].

Electrodiagnostic studies show a length-dependent sensorimotor neuropathy, typically demyelinating, with axonal degeneration. Any or all of the liver, spleen, and lymph nodes can be enlarged. The most commonly found M protein is an immunoglobulin G (IgG) or IgA and almost always of the lambda type. Nearly 20% of all patients experience thromboembolic complications, such as stroke, myocardial infarction, and Budd-Chiari syndrome [1].

The common misdiagnoses of POEMS syndrome include CIDP, MGUS, and multiple myeloma. The outlook for POEMS is favorable, with a median life expectancy of 13.8 years based on the Mayo Clinic series. The occurrence of events does not have a correlation with survival rates. Fatality is not primarily due to the advancement of plasma cell dyscrasia but rather attributed to cardiac, respiratory, or infectious factors [1].

In our case, the patient presented with polyneuropathy (mandatory major criteria), Castleman's disease (major criteria) and lymphadenopathy (minor criteria), which strongly support our diagnosis. The diagnosis was confirmed by nerve conduction study and biopsy of cervical lymph node followed by abdominal CT scan. Moreover, in protein electrophoresis, monoclonal gammopathy was present, which further adds to our suspicion. Bone marrow study was not done, as finding Castleman's disease was enough to exclude multiple myeloma from our differentials.

In this particular case, it took less than 6 months from the onset of the first symptoms of peripheral neuropathy to reach a conclusive diagnosis. Our diagnosis was comparatively prompt, and the patient demonstrated a satisfactory outcome after treatment.

## Conclusion

POEMS syndrome is a rare and deadly condition in which individuals appear with a variety of early symptoms as a result of multisystem involvement. A thorough medical history, physical examination, and comprehensive knowledge of this rare condition are required for an accurate diagnosis and therapy. We have reported this case with the aim of reminding clinicians about this rare disorder and so prevent misdiagnosis.

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## Author's Contributions:

Dr. Hafeja Akhter Hima: Conceptualization, writing, editing, clinical information of the patient, patient treatment, investigation

Dr. Raihan Rotap Khan: Conceptualization

Dr. Sayedatus saba: Corresponding author, Conceptualization, writing, editing, final draft, histopathological and immunohistochemical study

Prof. Dr. Habibur Rahman Tarek: Investigation, patient treatment, supervision

Dr. Mohiuddin Mohammad Alamgir: Proof reading

Dr. Md. Rezaur Rahman: Proof reading

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## Declaration of Conflicting Interests

There are no conflicting interests.

## Research Ethics Statement

- Approval of the research protocol by an Institutional Reviewer Board: N/A.
- Informed Consent: Informed written consent was taken from the patient
- Registry and the Registration No. of the study/trial: N/A.
- Animal Studies: N/A.

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