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Case Report

Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal Protein, and Skin Changes (POEMS) Syndrome in the Middle East: A Case Report

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ABSTRACT

POEMS syndrome is a rare paraneoplastic syndrome characterized by polyneuropathy, organomegaly, endocrinopathy, monoclonal protein gammopathy, and skin changes. We report the first case of POEMS syndrome in Egypt. We report a 37-year-old Egyptian male who presented with left lower limb pain, numbness, weight loss, and urinary incontinence. Laboratory evaluation revealed monoclonal IgA lambda bands and 10% plasma cells on bone marrow aspiration. Nerve conduction studies confirmed severe sensory-motor polyneuropathy with axonal degeneration. Bone scan showed sclerotic changes, and imaging revealed hepatosplenomegaly. The patient met the diagnostic criteria for POEMS syndrome and was treated with the VRD protocol. Follow-up over 23 months showed declining M-band levels with eventual normalization. However, 2-microglobulin levels rose. The patient achieved a partial response and was referred for autologous bone marrow transplantation. This represents the first reported case of POEMS syndrome in Egypt, emphasizing the need for clinical awareness in non-endemic regions. A multidisciplinary approach is essential for optimal management. Further studies are needed to explore the presentation and management of POEMS syndrome in the Middle East and North Africa region.

1. Introduction

POEMS syndrome is a rare paraneoplastic syndrome characterized by polyneuropathy, organomegaly, endocrinopathy, monoclonal protein gammopathy, and skin changes [1]. Alternative names for this condition include osteosclerotic myeloma and PEP (polyneuropathy, endocrinopathy, plasma cell dyscrasia) syndrome [2, 3]. POEMS syndrome was first described by Crow in 1956, and Bardwick coined the acronym in 1980 [4, 5]. It typically presents in the fifth to sixth decade [2, 3, 6].

Although the exact cause remains unknown, proinflammatory cytokines and growth factors, such as vascular endothelial growth factor (VEGF), appear to play a central role in its pathogenesis [7, 8, 9]. The International Myeloma Working Group (IMWG) diagnostic criteria require two mandatory criteria (polyneuropathy and monoclonal plasma cell proliferative disorder), one major criterion (sclerotic bone lesions, Castleman's disease, or elevated VEGF), and one minor criterion (organomegaly, extravascular volume overload, endocrinopathy, skin changes, thrombocytosis, or papilledema) [10].

Clinical manifestations develop over weeks to months, with neuropathy, endocrine dysfunction, and extravascular volume overload being dominant, helping differentiate it from multiple myeloma [10]. Treatment primarily targets the underlying plasma cell disorder: radiation therapy for localized disease, and systemic therapy with or without hematopoietic cell transplantation for disseminated disease with bone marrow involvement [10, 11]. Patients should be monitored every three months to assess symptom progression compared to baseline. Laboratory markers, such as VEGF and M-protein, do not always correlate with disease activity, so clinical trends are essential in guiding treatment [10].

POEMS syndrome appears to be particularly rare in the Middle East and North Africa (MENA) region, with only one previously published case report from Morocco [12] and two patients with POEMS syndrome identified in a retrospective study on monoclonal gammopathies in a Moroccan hospital [13]. Herein, we report the first case report of POEMS syndrome in Egypt and the second in the MENA region.

2. Case presentation

A 37-year-old male presented with progressive distal left lower limb pain and numbness for one year, accompanied by significant weight loss (>10 kg). He had urinary incontinence for 14 months before admission. His medical history included a right below-knee amputation and a left toe amputation due to trauma. Given the constellation of symptoms, an extensive diagnostic workup was initiated to uncover an underlying systemic disorder.

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Table 1: Diagnostic laboratory panel at admission

Diagnostic criteria	Result	Reference	Significance
Platelet count	737 × 10^9/L	(150–400 × 10^9/L)	Thrombocytosis
TSH	6.97 mIU/L	(0.4–4 mIU/L)	Hypothyroidism
HbA1c	10.2%	< 5.7%	Diabetes
Liver span	20 cm	(6–12 cm)	Hepatomegaly
Spleen size	16 cm	< 12 cm	Splenomegaly
Papilledema	No	–	Absent
Extravascular overload	No	–	Absent
Skin changes	No	–	Absent

HbA1c, glycated hemoglobin; TSH, thyroid-stimulating hormone.

Laboratory evaluation revealed a monoclonal plasma cell disorder. Serum protein electrophoresis (SPEP) demonstrated an M-band (0.56 g/dL) in the beta-2 globulin region, while immunofixation confirmed two monoclonal IgA lambda bands. Additionally, bone marrow aspiration showed 10% plasma cells, supporting the presence of a plasma cell dyscrasia.

A nerve conduction study confirmed severe sensory-motor polyneuropathy with axonal degeneration, particularly affecting the left lower limb. Poor motor and sensory responses in the posterior tibial, peroneal, and sural nerves revealed polyneuropathy.

A bone scan revealed a deformed hip joint with sclerotic changes and flattening of the right femoral head. Ultrasound imaging identified hepatosplenomegaly. His CBC demonstrated thrombocytosis (737 × 10/L). Additionally, he had endocrinopathy, evidenced by elevated TSH, as well as elevated HbA1c (10.2%). Detailed laboratory values and diagnostic findings that met these criteria are summarized in **(Table 1)**. Based on the diagnostic framework for POEMS syndrome, this patient met the mandatory major criteria of both polyneuropathy and a monoclonal plasma cell disorder while also fulfilling one additional major criterion (sclerotic bone lesions) and three minor criteria (organomegaly, thrombocytosis, and endocrinopathy). These findings collectively confirmed the diagnosis of POEMS syndrome, guiding the initiation of targeted therapy.

The patient was started on the VRD protocol, as per the current guidelines for disseminated POEMS syndrome [10]. This 22-day cycle regimen included Velcade (bortezomib) at 1.3 mg/m² on days 1, 8, 15, and 22; Lenalidomide at 25 mg on days 1-21; and Dexamethasone at 40 mg administered weekly. Given the prothrombotic risk associated with POEMS and immunomodulatory therapy, anticoagulation was initiated with Rivaroxaban alongside Aspirin (81 mg daily).

Follow-up assessments tracked treatment response through serial laboratory monitoring **(Table 2)**. The patient achieved a partial response, with normalization of the monoclonal protein by January 2025. However, the plasma cell burden remained stable at 15%, and 2-microglobulin levels showed a concerning rise over time. Despite this, the patient's clinical course and symptoms remained the key markers of disease progression. In light of the biochemical follow-up assessments and the discordant response to treatment, the patient was subsequently referred to the bone marrow transplant unit for autologous bone marrow transplantation.

3. Discussion

POEMS syndrome is a rare disease characterized by multisystem affection that requires clinical awareness and a high index of suspicion in cases presenting with peripheral neuropathy, organomegaly, refractory ascites, and skin changes of unknown origin. This is particularly important in non-endemic regions where country-specific data remains limited.

In our case, a 37-year-old male presented with sensory-motor polyneuropathy with axonal degeneration. Initial evaluation showed elevated IgA lambda bands, and bone marrow aspiration showed 10% plasma cells, fulfilling the mandatory major diagnostic criteria of POEMS syndrome [10]. Imaging revealed sclerotic bone lesions, fulfilling another major criterion. The patient also fulfilled three minor criteria: hepatosplenomegaly, thrombocytosis, and endocrinopathy (diabetes mellitus and hypothyroidism). Although the existing literature features many cases of POEMS syndrome, this case report represents the first published case of POEMS syndrome in Egypt and the second in the MENA region, after Benbella et al.'s case report on a 41-year-old Moroccan female patient [12]. We conducted a targeted literature search using PubMed and Google Scholar to identify previously reported cases of POEMS syndrome in the MENA region. However, we acknowledge the possibility that additional cases may exist in non-indexed or non-English regional journals, which our search may not have captured.

POEMS syndrome has a unique geographical distribution, characterized by a significantly higher prevalence in Asian populations, particularly in Japan, compared to Western countries. Many of the early case reports originating from Japan in the 1980s [3, 14], and a national Japanese survey indicated a prevalence of approximately 0.3 per 100,000 persons [15], notably higher than reported rates in other regions. Over the years, many case reports have been published from China [16], the United States [1], India [17], and France [18].

In comparing our case with the case reported by Benbella et al. [12], both patients initially presented with polyneuropathy, which is the dominant clinical feature of POEMS syndrome in nearly 100% of cases, as reported in large retrospective studies [3, 18, 14]. Both cases featured systemic symptoms, including weight loss and sphincteric disorders. Although both patients demonstrated an IgA lambda monoclonal gammopathy, it is worth noting that our patient exhibited a slightly higher M-band level (0.56 g/dL vs. 0.35 g/dL in Benbella et al.), despite a modestly lower total serum IgA level (673 mg/dL vs. 722 mg/dL in Benbella et al.) [12]. The neuropathy subtype differed between the two cases: our patient exhibited severe sensory-motor polyneuropathy with secondary axonal degeneration, whereas Benbella et al.'s case involved primary demyelinating polyradiculoneuropathy. Since axonal regeneration is typically less complete than remyelination, the presence of substantial axonal degeneration in our case may indicate slower neurological recovery and a more severe disease course. This highlights the importance of early diagnosis and prompt initiation of therapy to prevent significant axonal damage. The Moroccan case presented with additional manifestations that were not seen in our case, including papilledema, bilateral temporal-occipital headaches, paroxysmal facial erythema, and low vitamin B12 levels.

POEMS is a complex disease; challenges in early recognition and diagnosis play a critical role in the morbidity and mortality of the disease. There is an average delay of 13 to 18 months between symptom onset and diagnosis [19]. In our case, the delay in diagnosis was 13 months, in line with the previously reported

Table 2: Serial laboratory values (treatment response monitoring)

Parameter	Month post-baseline	M-protein (g/dL)	Plasma cells (%)	β2-microglobulin (mg/L)
Baseline	0	0.56	10	–
Oct 2023	6 months	0.28	–	4.5
Apr 2024	12 months	–	15	–
Aug 2024	16 months	–	–	10.9
Jan 2025	21 months	Normal	–	–
Mar 2025	23 months	Normal	4	–

HbA1c, glycated hemoglobin; TSH, thyroid-stimulating hormone

average. The rarity of the disease, limited awareness of hematological conditions, and the affordability of diagnostic investigations all contributed to this delay. This case highlights the importance of maintaining clinical suspicion for POEMS syndrome in patients with multisystem involvement. This is particularly important in non-endemic regions, where diagnostic delay may be partially due to a decreased index of suspicion among practitioners, given the rarity of the condition in the region. Low socioeconomic status, limited health literacy, and non-adherence to treatment and follow-up among patients may further exacerbate the delay. In addition, healthcare system-related factors such as prolonged waiting times and the availability and affordability of diagnostic tools may also contribute. Further studies are needed to address the diagnostic delay in patients with POEMS syndrome, especially in non-endemic areas. This can help detect the factors that contribute significantly so that they can be eliminated.

The differential diagnoses of POEMS syndrome include chronic inflammatory demyelinating polyneuropathy (CIDP), Monoclonal Gammopathy of Undetermined Significance (MGUS), multiple myeloma (MM), and amyloidosis (AL), among others. Historically, the diagnosis of POEMS was made in cases of subacute motor-dominant demyelinating polyradiculoneuropathy by exclusion of CIDP in refractory cases not responding to IVIG therapy [10]. Mauermann ML et al. compared a group of 138 POEMS patients to a control group of 69 CIDP patients through nerve conduction studies (NCS) and electromyography (EMG). POEMS patients demonstrated: greater axonal loss, greater slowing of the intermediate nerve segments due to demyelination, less temporal dispersion and conduction blocks, and absent sural sparing [20]. Polyneuropathy can differentiate POEMS syndrome from classical MM. On the other hand, MGUS typically presents with no other systemic findings.

Our patient had an extensive history of diabetes mellitus with an HbA1c level of 10.2% at the time of admission. Interestingly, this is similar to a case previously described by Zhou et al., which highlighted how the presentation of POEMS syndrome can mimic that of other conditions, such as diabetes [21].

The treatment of POEMS syndrome is based on the extent of plasma cell infiltration. The patient was started on the standard VRD protocol. Treatment response monitoring **(Table 2)** revealed a critical challenge in managing POEMS syndrome: biomarker discordance. While M-protein achieved complete normalization, suggesting hematologic response, persistent plasma cell burden (15%) and rising 2-microglobulin levels (4.5→10.9 mg/L) indicated ongoing disease activity. This phenomenon in POEMS syndrome strongly justifies transplant referral. VEGF levels testing was not available at our facility. While our patient met diagnostic criteria through an alternative major criterion (sclerotic bone lesions), the absence of VEGF measurements represents a limitation for comprehensive disease monitoring, as VEGF serves as both a diagnostic marker and a treatment response indicator in POEMS syndrome.

Given the complexity of the disease and the presence of barriers to its management, a highly coordinated, multidisciplinary approach is necessary to optimize patient outcomes. The management of our patient involved collaboration among the hematology, neurology, and endocrinology departments, as well as a referral to the bone marrow transplant unit. This enabled timely diagnosis and comprehensive management. Although this case report offers valuable insights, it is challenging to conclude from a single case report. The lack of long-term follow-up data post-transplantation to assess sustained remission is another limitation to this case, making it impossible to determine if our therapy approach (VRD induction followed by autologous stem cell transplantation) was ultimately successful. The lack of data on the prevalence and outcomes of POEMS syndrome in the MENA region also limited our understanding of this case. Thus, further studies are needed to establish prevalence patterns, identify diagnostic barriers, and develop standardized treatment protocols in the MENA region.

4. Conclusion

This case represents the first documented patient with POEMS syndrome in Egypt, highlighting the diagnostic challenges and the 13-month delay typical in non-endemic regions. The successful multidisciplinary approach involving hematology, neurology, and endocrinology departments was crucial for comprehensive management, though partial response to VRD therapy necessitated transplant referral. Enhanced clinical awareness and improved access to diagnostic tools are essential to reduce diagnostic delays and improve outcomes for POEMS syndrome patients in the MENA region.

Conflicts of Interest

The authors declare that they have no competing interests.

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Informed consent

We obtained written informed consent for publication from the patient.

Large Language Model

None

Authors Contribution

MAH, RNE, IA, MS, and MMM contributed to writing the original draft. MS, AZ, and AE were responsible for supervision. MAH and MMM handled project administration. MAH, RNE, IS, MS, MMM, AZ, and AE participated in reviewing and editing. All authors read and approved the final content.

Data Availability

Clinical and laboratory data are available.

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