A case of POEMS syndrome in a 53-year old male: A diagnostic challenge

Niki Thakur, Gopi Aryal, Anuj Poudel, Reena Rana, Kricha Pandey
Department of Laboratory Medicine and Pathology, Nepal Mediciti Hospital, Lalitpur, Nepal

ABSTRACT

BACKGROUND

POEMS syndrome is a rare paraneoplastic disorder associated with plasma cell dyscrasia and characterized by polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes. Due to its protein manifestations, diagnosis is often delayed.

CASE PRESENTATION

We report a case of 53-year old male who presented with polyneuropathy, weight loss, burning sensation in bilateral sole and night sweats. Serum immunofixation electrophoresis and histopathological evaluation demonstrated a monoclonal plasma cell disorder. Imaging revealed multiple osteosclerotic bone lesion, mild hepatomegaly, pleural and pericardial effusion and lymphadenopathy.

CONCLUSIONS

This case highlights the diagnostic challenge posed by POEMS syndrome and the importance of a multidisciplinary approach.

KEY WORDS

POEMS syndrome, plasma cells dyscrasia, polyneuropathy, case report

*Corresponding Author |
Dr Niki Thakur
Department of Laboratory Medicine and Pathology
Nepal Mediciti Hospital, Sainbu, Lalitpur, Nepal.
Email: thakurniki277@gmail.com

INTRODUCTION

POEMS syndrome is a rare paraneoplastic multisystem disorder characterized by the constellation of polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin chnages.(1) It is associated with plasma cell neoplasm, most often exhibiting osteosclerotic bone lesions and varying degree of bone marrow fibrosis. (2) The syndrome typically affects individuals in the sixth decade of life and shows a slight male predominance.

The diagnosis of POEMS syndrome is based on established criteria requiring the presence of both mandatory features-polyneuropathy and a monoclonal plasma cells disorder along with at least on major and one minor criteria include castleman disease, osteosclerotic bone lesions and elevated vascular endothelial factor (VEGF) levels, while minor criteria encompasses organomegaly, endocrinopathy, skin changes, papilloedema, thrombocytosis and extravascular volume overload. Owing to its rarity and heterogeneous clinical presentation, POEMS syndrome remains a diagnostic challenge, often leading to delayed recognition. (3)

CASE REPORT

A 53-year old male presented with history of progressive weight loss of approx 20kg over three years, including a 5 kg loss during the preceding two months. This was associated with night sweats and intermittent burning sensation over the bilateral soles for the past two years, with worsening of neuropathic symptoms in the two months prior to reservation. There was no history of fever or anorexia.

Initial laboratory evaluation revealed a normocytic normochromic blood picture with mild thrombocytosis (hemoglobin 14.5 g/dl, mean corpuscular volume 87 fl, total leucocyte count 6,000/cumm, platelet count 1,54,000 cells/mm. Biochemical investigations showed low serum calcium and albumin levels with an elevated lactate dehydrogenase (LDH) of 186 IU/L. Serum magnesium, vitamin B12, blood urea, nitrogen and other routine renal parameters were within normal limits.

Magnetic resonance imaging of the lumbosacral spine demonstrated degenerative changes with central canal stenosis at the L3-L4 and L4-L5 levels. High resolution computed tomography (HRCT) of the chest revealed multiple osteosclerotic lesions involving the right 5th ribs, with additional sclerotic foci in the left clavicle and scapular blade. Computed tomography of the thorax and whole abdomen showed minimal left sided pleural effusion, mild pericardial effusion, mild hepatomegaly, mild prostatomegaly, enlarged para aortic lymph nodes and sub centimetric mediastinal lymphadenopathy.

Serum protein electrophoresis and immunofixation, performed using high resolution agarose gel electrophoresis, demonstrated a discrete monoclonal band in the beta-globuin region, consistent with an IgA lambda monoclonal gammopathy.

BONE MARROW ASPIRATION FINDINGS

(Figure 1A) Bone marrow aspirate smears showed a normocellular marrow for age with megaloblastic changes in erythroblasts and 06 % plasma cells. (Figure 1B) Few binucleated plasma cells noted.

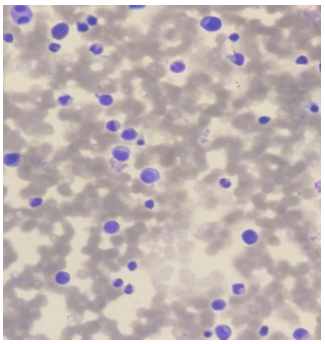

Figure 1A

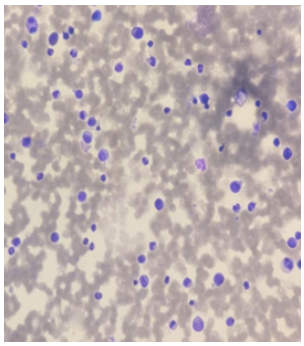

Figure 1B

BONE MARROW BIOPSY REPORT

Figure 2A: Low power view shows bone marrow biopsy shows normocellular marrow for age with focally thickened trabecular bone. Figure 2B: Intertrabecular bone shows paratrabecular fibrosis with trilineage normal hematopoiesis and entrapped few plasma cells.

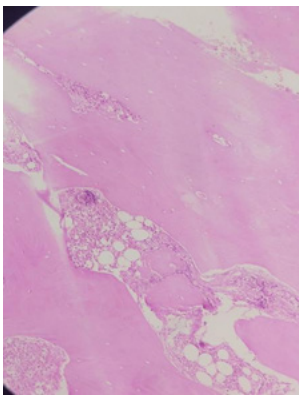

Figure 2A

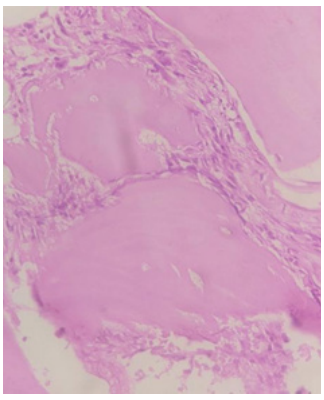

Figure 2B

DISCUSSION

POEMS syndrome is a multisystem disorder characterized by a constellation of clinical and laboratory findings, typically associated with an underlying plasma cell neoplasm, consistent with established literature (4), the diagnostic hallmark is the presence of a monoclonal plasma cell population, which is almost invariably lambda light chain restricted band of the IgG or IgA type. The characteristics bone marrow lesion is an osteosclerotic plasmacytoma,

histologically defined by focally thickened trabeculae with associated paratrabecular fibrosis containing entrapped plasma cells. While marrow plasma cell percentage away from these lesions is often below <5%, it can be markedly elevated in disseminated disease. (5)

The disease follows a chronic progressive coarse with reported 5 year survival rates ranging from 60% to 95% (6). Although no specific genetic markers reliably predict prognosis (7), disease activity exhibits a strong correlation with serum VEGF levels, a key pathogenic mediator. Due to its rarity and clinical overlap with other neuropathic conditions- most notably chronic inflammatory demyelinating polyradiculoneuropathy- diagnosis is frequently delayed. Early recognition and intervention are, therefore, critical to preventing irreversible neurological and systemic damage (9).

The findings in our patient align with the diagnostic framework for POEMS syndrome. The diagnosis meets on fulfilling the mandatory criteria, which in case includes a demonstrable monoclonal gammopathy on serum immunofixation electrophoresis and a polyneuropathy. Histopathological examination of bone marrow aspiration and biopsy further support a plasma cell dyscrasia. Major diagnostic criteria were satisfied by the identification of multiple osteosclerotic lesion involving the right 5th, 7th and 11 the ribs, left 3rd and 5th ribs, left clavicle and scapula. The clinical picture was supplemented by several minor criteria: left sided pleural effusion, mild pericardial effusion, mild hepatomegaly, mild prostatomegaly, enlarged para aortic lymph nodes, subcentimetric mediastinal lymph nodes, and hyperpigmented skin lesions. The patient was subsequently referred to abroad for specialized further management.

REFERENCES:

1. POEMS syndrome: definitions and long-term outcome. Dispenzieri A, Kyle RA, Lacy MQ, et al. Blood. 2003;101:2496–2506. doi: 10.1182/blood-2002-07-2299. [DOI] [PubMed] [Google Scholar].
2. Schey, S. "Osteosclerotic myeloma and 'POEMS' syndrome." Blood reviews vol. 10,2 (1996): 75-80. doi:10.1016/s0268-960x(96)90035-1
3. Dispenzieri A. (2007). POEMS syndrome. Blood reviews, 21(6), 285–299. https://doi.org/10.1016/j.blre.2007.07.004
4. Dispenzieri, A., Kyle, R. A., Lacy, M. Q., Rajkumar, S. V., Therneau, T. M., Larson, D. R., Greipp, P. R., Witzig, T. E., Basu, R., Suarez, G. A., Fonseca, R., Lust, J. A., & Gertz, M. A. (2003). POEMS syndrome: definitions and long-term outcome. Blood, 101(7), 2496–2506. https://doi.org/10.1182/blood-2002-07-2299
5. Dao, L. N., Hanson, C. A., Dispenzieri, A., Morice, W. G., Kurtin, P. J., & Hoyer, J. D. (2011). Bone marrow histopathology in POEMS syndrome: a distinctive combination of plasma cell, lymphoid, and myeloid findings in 87 patients. Blood, 117(24), 6438–6444. https://doi.org/10.1182/blood-2010-11-316935
6. Dispenzieri A. (2014). POEMS syndrome: 2014 update on diagnosis, risk-stratification, and management. American journal of hematology, 89(2), 214–223. https://doi.org/10.1002/ajh.23644
7. Dispenzieri, A., Kyle, R. A., Lacy, M. Q., Rajkumar, S. V., Therneau, T. M., Larson, D. R., Greipp, P. R., Witzig, T. E., Basu, R., Suarez, G. A., Fonseca, R., Lust, J. A., & Gertz, M. A. (2003). POEMS syndrome: definitions and long-term outcome. Blood, 101(7), 2496–2506. https://doi.org/10.1182/blood-2002-07-2299
8. Keddie, S., & Lunn, M. P. (2018). POEMS syndrome. Current opinion in neurology, 31(5), 551–558. https://doi.org/10.1097/WCO.0000000000000610
9. Dispenzieri A. (2019). POEMS Syndrome: 2019 Update on diagnosis, risk-stratification, and management. American journal of hematology, 94(7), 812–827. https://doi.org/10.1002/ajh.25495