Case Report

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POEMS Syndrome with Biclonal Gammopathy: A Rare Association

Oncology Section

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ABSTRACT

Polyneuropathy, Organomegaly, Endocrinopathy, M protein and Skin changes (POEMS) syndrome is rare plasma cell dyscrasia with multisystem involvement. The name comes from the five characteristic features: Polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes. The presence of biclonal M band is a rare manifestation. Here, we are describing the cases of a 60-year-old lady, presented with bilateral pedal oedema and pericardial effusion and peripheral neuropathy. She also had hepatosplenomegaly, hyperpigmented rash and hypothyroidism and hyperparathyroidism. The serum protein electrophoresis and the immunofixation electrophoresis revealed two distinct monoclonal bands, immunoglobulin IgG kappa and IgA lambda. There was a mild increase in plasma cells and sclerotic bone lesion in pelvis. The POEMS syndrome is generally associated with lambda light chain restriction. The presence of biclonal gammopathy involving kappa and lambda is a rare manifestation. The pathogenic or prognostic role of different paraprotein is not known. Further studies are required to delineate such effect.

Keywords: Hepatosplenomegaly, Multiple myeloma, Protein electrophoresis

CASE REPORT

A 60-year-old, hypothyroid, nonhypertensive, nondiabetic lady, presented in Cardiology Department for bilateral pedal oedema and pericardial effusion seven months back. Gradually, she developed distension of abdomen and bilateral tingling and numbness of both lower limbs. There was no history of jaundice, bone pain, hematemesis, melena or any history of blood transfusion. There was no past history of tuberculosis or contact history with the same.

On examination, she had pallor, bilateral pedal pitting edema. Per abdomen, there was hepatosplenomegaly with moderate ascites. There were few well defined macular hyperpigmented rashes over abdominal and lower limb skin. There was no lymphadenopathy, malar rash, oral ulcer, photosensitivity or arthralgia. The neurological evaluation revealed loss of both vibration and position senses and loss of pain sensation up to ankle joint bilaterally. The power of small muscles of both feet was lost and planter reflex was absent. The knee jerks were normal while ankle jerks were reduced bilaterally. The clinical features were suggestive of peripheral neuropathy and confirmed by nerve conduction study. POEMS syndrome was suspected and Serum Protein Electrophoresis (SPEP) was done. The SPEP revealed two distinct monoclonal bands, one at the gamma globulin region and another one near beta region [Table/Fig-1]. The immunofixation electrophoresis revealed the bands as immunoglobulin (Ig)G kappa and IgA lambda, respectively [Table/Fig-2]. Both kappa and lambda light chain was raised (199 mg/dl and 255 mg/dl, respectively) and Serum Free Light Chain (SFLC) ratio was within range (0.745). The patient did not have any nephropathy. The beta-2 microglobulin was 7747 ng/ml and bone marrow aspiration and biopsy revealed a mild increase in plasma cell (8%). The hormonal study revealed the presence of hyperparathyroidism without hypercalcemia. The skeletal survey revealed sclerotic bone lesion in pelvis.

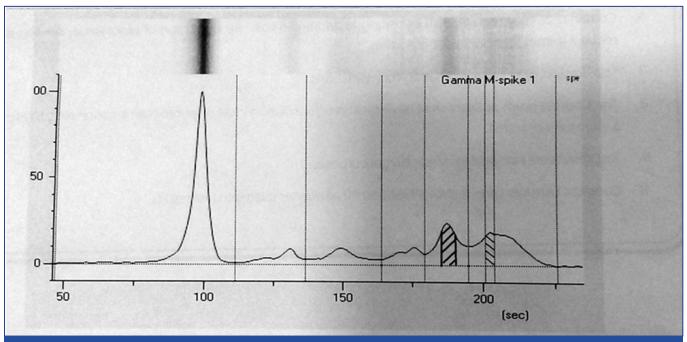
She was started on systemic chemotherapy, cyclophosphamide, lenalidomide and dexamethasone. She also received diuretics aldosterone for ascites and pregabalin for peripheral neuropathy. She was already on thyroid replacement therapy. The ascites and oedema improved and peripheral neuropathy was also symptomatically improved, though loss of function remained the same. After four cycles of chemotherapy, the SPEP showed a partial response. Patient was counseled about the need for high-dose therapy with autologous stem cell rescue but she declined and lost to follow up.

DISCUSSION

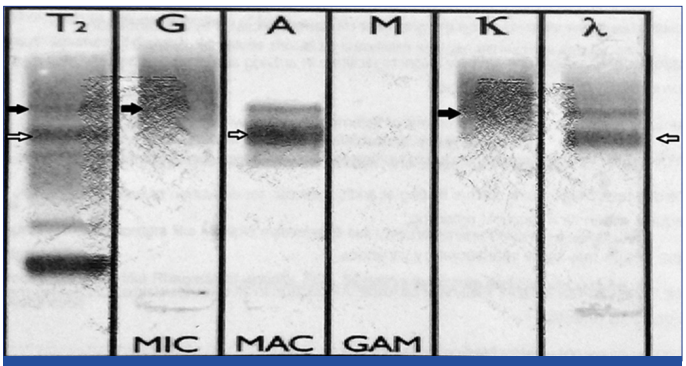
POEMS syndrome is rare multisystem disorder of plasma cell origin. It was first described by Scheinker in 1938 [1,2]. The manifestations occur because of increased secretion of vascular endothelial growth factor. The name "POEMS" syndrome comes from the following five characteristic features: Polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes. It is diagnosed by the proposed criteria laid down by Dispenzieri A and need the presence of two essential, one major criteria and one minor criterion [3].

Biclonal gammopathy is defined by the presence of two distinct M components, which is associated with either two distinct clones of plasma cells, each producing an unrelated monoclonal immunoglobulin or it may result from the production of two monoclonal proteins by a single clone of plasma cells [4,5]. The occurrence of biclonal gammopathy is a rare phenomenon and instance ranges from 0.14% to 3.2% [6-8]. Most of biclonalities in M protein were associated with multiple myeloma. The most common combination is IgG and IgA (33%) and followed by IgM and IgG combination (24%) [6]. Kyle RA et al. reported that out of the 57 patients with biclonal M protein, 30 (53%) had IgG and IgA components [4].

The POEMS syndrome is generally associated with lambda light chain restriction. The presence of biclonal gammopathy in POEMS syndrome is an extremely rare phenomenon. Very few case studies have been found in the literature documenting biclonal M protein in



[Table/Fig-1]: Serum protein electrophoresis showing two distinct monoclonal bands, one in gamma region and one in beta region.



[Table/Fig-2]: Immunofixation electrophoresis showing the types of gamma globulin in M band, the one in gamma region consists of immunoglobulin G-kappa (black arrow) and the other one in beta region consists of immunoglobulin A-lambda (white arrow).

POEMS syndrome. Ham JY et al. reported a similar case previously [9], the SFLC ratio was out of range in that patient. Raajasekar A also reported a similar case [10]. The pathogenic role or prognostic role of different paraprotein is not yet known. Although in multiple myeloma, no difference in the clinical features or treatment response have been found in case of biclonal gammopathy, but proper clinical data in a different group of plasma cell dyscrasia is lacking. Further studies are required to delineate such effect.

CONCLUSION

In summary, we described a rare case of POEMS syndrome in association with biclonal M band, involving IgG kappa and IgA lambda. Besides rarity of the disease, it also provides insight into occurrence of such phenomenon of biclonality in diverse plasma cell dyscrasia including POEMS syndrome.

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