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# A Case with Poems Syndrome

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**Abstract:** POEMS (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, skin changes) syndrome is a rare disorder of plasma cell. A 57 years old man admitted to our clinic with complaints of legs pain and weakness. He had sclerodactly and hypopigmented lesions was on the fingers and legs. The patient's laboratory data were indicative of anemia, high sedimentation rate and elevated plasma IgG level. M spike was determined in protein electropheresis. A monoclonal band of IgG-kappa was detected in serum and urine immunoelectropheresis. Electromyography showed sensorio-motor polyneuropathy in bilatheral lower extremity. A computed tomography scan showed sclerotic bone lesion in left femur. Combined chemotherapy (lenalidomide 25 mg once daily oral on days 1-21 and dexamethosone 40 mg/day days 1-4 of repeated 28-day cycles) was started to our patient for POEMS syndrome. The symptoms of neuropathy decreased after 2 months. M spike was decreased in protein electropheresis. The patient with POEMS syndrome generally are diagnosed late because of this syndrome is rare. We should think POEMS syndrome in patient with neuropathy and clonal plasma disorder.
**Keywords:** POEMS syndrome, neuropathy, monoclonal gammopathy.

## INTRODUCTION

POEMS (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, skin changes) syndrome is a rare multisystem disorder, also known as Crow–Fukase syndrome, Shimpo syndrome, Takatsuki disease [1]. The mandotory criterias are the presence of peripheral neuropathy, a monoclonal plasma cell disorder. Castleman's disease, increased levels of serum vascular endothelial growth factor (VEGF) and osteosclerotic bone lesions are majör criteria. Minor features include organomegaly, endocrinopathy, Extravascular volume overload, typical skin changes, thrombocytosis/ erythrocytosis and papilledema. According to the revised POEMS syndrome criteria, two mandatory criteria, including one major criterion and one minor criterion are required to confirm the diagnosis.

POEMS syndrome is a rapidly progressive disease and can cause death and life-threatening conditions. Because pleural effusion, acites, and/or cardiac effusion are common in patients with POEMS syndrome. Therefore early diagnosis is very important. We reported a case with POEMS syndrome because of rarity.

## CASE REPORT

A 57 years old man admitted to our clinic with legs pain and weakness. His past medical history included of chronic obstructive pulmonary disease and coronary artery disease for 5 years. On physcial examination his blood pressure was 120/80 mmHg, pulse was 90 beats/min, no fever, pale skin, sclerodactly, hypopigmented lesions on fingers, legs and without any other significant findings.

Laboratory findings were as follows; level of hemoglobin was 11.9 gr/dl, hematocrit 40.1%, white cell count 5500/mm³, platelets 330000 µ/l, eerythrocyte sedimentation rate 63 mm/hour. Level of serum urea was 14 mg/dl, creatinine 0.5 mg/dl, sodium 130 mEq/L, potassium 4.1 mEq/L, uric acid 4.3 mg/dl, total protein 9.6 mg/dl, albumin 2.4 mg/dl, C-reactive protein 13.5 mg/l, aspartate amino transferase 27 UI/L, alanine amino transferase 31 UI7L, alkaline phosphatase 33 UI/L, gamma glutamyl transferase 68 U/L, alkaline phosphatase 68 U/L, lactate dehydrogenase 230 U/L. IgG 53 g/l , IgA 1.36 g/l , IgM 0.6 g/l. M spike was determined in protein electrophoresis. Serum and urine immunoelectrophoresis showed the presence of a monoclonal band of IgG-kappa. The bone marrow aspiration showed normal cellularity and 2% infiltration by plasma cells. Therefore multipl myeloma was excluded in this patient. Level of serum cortizol was 5.86 µg/dl, testesteron 0.52 ng/ml (normal range: 1.75-7.81 ng/ml) idi Normal response was detected to ACTH stimulation test. The electromyogram (EMG) revealed sensorio-motor polyneuropathy in bilatheral lower extremity. Sclerotic bone lesion in left femur was detected with computerize tomograhy. Thereby, the diagnostic criteria for POEMS syndrome were fulfilled because one element of the other three major criteria

(sclerotic bone lesions) and two of the minor criteria (endocrinopathy, skin changes) had already been met. The patient began receiving chemotherapy based on a regimen for multiple myeloma (lenalidomide 25 mg once daily orally on days 1-21 and dexamethosone 40 mg/day days 1-4 of repeated 28-day cycles). After two courses of chemotherapy, the symptoms of neuropathy improved. M spike was decreased from 42% to 31%.

## DISCUSSION

POEMS syndrome was first reported by Crow in 1956 [2]. In 1980, the acronym POEMS was coined by Bardwick *et al.;* based on the 5 main features of the disease, namely, polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes.The mechanism of the POEMS syndrome is not well understood. Various cytokines such as interleukin 1β, interleukin 6 and TNFα appears to be a major feature of this disorder. Elevation of plasma VEGF levels is an important feature of the POEMS syndrome and is useful for monitoring therapy [3]. We could not measure level of serum VEGF of our patient. Osteosclerotic lesions occur in 95% of patients. Some lesions are densely sclerotic, while others are lytic with a sclerotic rim, while still others have a mixed soap-bubble appearance. We determined sclerotic lesion in our patient with with computerize tomograhy.

Peripheral neuropathy is the clinical feature of POEMS syndrome and it is is typically distal, symmetric, and slowly progressive with demyelinating changes. It involves both motor and sensory nerves. Symptoms begin in the feet and consist of tingling, feelings of coldness, paresthesias. The neuropathy is seldom painful, and autonomic involvement is rare [4,5]. Our patient has sensoriomotor polineuropathy in lower extremites. The presence of a monoclonal plasma cell disorder is another mandatory criteria of POEMS syndrome. Clonal immunoglobulin is usually IgG or IgA and almost always the monoclonal λ type [6]. M spike was determined in protein electrophoresis and immunoelectrophoresis (serum and urine) showed the presence of a monoclonal band of IgG-kappa.

The most common skin changes in patients with POEMS is hyperpigmentation. Hyperpigmentation is may be because of adrenocortical hypofunction. Rubor and flushing, clubbing, hypertrichosis, white nails, acrocyanosis and plethora are another skin lesions. Hypopigmented lesions were detected in our patient. Diabetes mellitus and hypogonadism are the most common endocrinopathies. Thyroid and parathyroid abnormalities, glucose metabolism abnormalities, and adrenal insufficiency are another endocrine abnormality. Hypogonadism was detected in our patient.

## CONCLUSION

POEMS syndrome is a very rare disorder. But POEMS syndrome can cause death ve life-threatening conditions. Therofore we should think of POEMS syndrome in patients with neuropathy and monoclonal gammopathy.

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