

Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal Gammopathy, and Skin Changes (POEMS) Syndrome

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Mr. Y, a 60-year-old Caucasian male, presented to the clinic with complaints of fatigue, headache, and dry eyes but no vision changes. He stated that his symptoms started five months earlier but was only seeking medical attention because the headaches had increased in severity and frequency. He stated that he recently had undergone an extensive ophthalmology workup, the results of which were unremarkable. Mr. Y also reported reddening of his skin and purplish discoloration of his palms and soles of his feet. He lost about 10 pounds in three months and had poor appetite, early satiety, and gastric reflux, but denied nausea, vomiting, diarrhea, or constipation. He reported feeling "short winded" during periods of activity. Mr. Y reported having generalized pain, numbness, and tingling from his knees down to his feet and increasing abdominal girth with associated leg edema. He denied syncope, palpitations, chest pain, cough, fever, or chills or any head or spinal trauma or history of hypertension, congestive heart failure, diabetes, or stroke.

A physical examination revealed a man in a wheelchair appearing to be chronically ill with no acute distress. Vital signs were stable and within normal limits. He had alopecia and increased hair growth on his upper and lower extremities. His abdomen was distended, mildly firm, and nontender with a positive fluid wave. The edge of the liver was palpable with deep inspiration at the right costal margin; the spleen was palpable 12 cm below the left costal margin. Mr. Y's lower extremities had bilateral two-plus pitting edema and hyperpigmentation, his palms and soles had

purplish discoloration, and the remainder of his skin had a bronze appearance.

Laboratory data revealed the following abnormalities: hematocrit 30.2% (range 40%–52%), serum creatinine 1.6 mg/dl (range 0.5–1.4 mg/dl), blood urea nitrogen 30 mg/dl (range 7–21 mg/dl), calcium 7.9 mg/dl (range 8.9–10.4 mg/dl), and albumin 3 units/L (range 3.5–4.8 units/L). Thyroid function test revealed a thyroid stimulating hormone of 10.97 units/ml (range 0.4–4.5 units/ml) and free T4 of 0.5 ng/dl (range 0.8–1.5 ng/dl). A vascular endothelial growth factor (VEGF) level of 3,010 pg/ml (range 31–86 pg/ml) was elevated. The hepatitis panel was negative. A computed tomography scan of the abdomen revealed hepatosplenomegaly and ascites. Bone marrow aspiration and biopsy revealed 3% plasma cells without clonality. Serum protein electrophoresis revealed an immunoglobulin-A λ (IgA λ) protein but no quantifiable monoclonal protein. X-ray bone survey showed no lytic lesions. An echocardiogram showed an ejection fraction of 60% with mild tricuspid regurgitation. A lumbar puncture was performed because of Mr. Y's complaints of headache. Cerebrospinal fluid obtained from the lumbar puncture revealed no white blood cells, one red blood cell, glucose of 89 mg/dl (range 50–80 mg/dl or about two-thirds of the blood glucose level), and protein of 55 mg/dl (range 15–45 mg/dl). Cerebrospinal fluid cytology and cultures were negative as was the computed tomography scan of the head. The rheumatoid panel and nerve conduction tests were unremarkable. Mr. Y was diagnosed with polyneuropathy, organomegaly,

endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome.

What is polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes syndrome?

POEMS syndrome is a rare paraneoplastic syndrome secondary to a plasma cell dyscrasia. The acronym POEMS syndrome was first coined by Bardwick et al. in 1980. POEMS syndrome also is known as osteosclerotic myeloma, Crow-Fukase syndrome, Takatsuki syndrome, and PEP (plasma cell dyscrasia, endocrinopathy, polyneuropathy) syndrome. Important traits not included in the acronym include elevated levels of VEGF, sclerotic bone lesions, Castleman disease (a rare disorder characterized by benign lymph node tumors), papilledema, peripheral edema, ascites, effusions, thrombocytosis, polycythemia, fatigue, and clubbing (Dispenzieri, 2007). The prevalence and incidence rates of the syndrome are difficult to determine because of misdiagnosis and under reporting. The peak incidence of the POEMS syndrome is in patients aged 40–60, unlike multiple myeloma which has a peak incidence in patients aged 60–80 (Chan, 2006). The course of POEMS syndrome is chronic and patients typically survive for more than a decade in contrast to multiple myeloma, where life expectancy may be measured in months or a few years.

What is the pathogenesis?

The pathogenesis of this multisystem disease is complex. The cause of POEMS syndrome is unknown, although chronic overproduction of proinflammatory and other

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