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 areata 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 deep, and within normal limits. He had purplish discoloration, and the remainder of his skin had a bronze appearance.

Laboratory data revealed the following abnormalities: hemoglobin 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 hepatosplenic megaly and ascites. Bone marrow aspiration and biopsy revealed 3% plasma cells without monoclonal protein. Serum protein electrophoresis revealed an immunoglobulin-A3 (IgA3) protein but no quantifiable monoclonal protein. A vascular endothelial growth factor (VEGF) level of 3,010 pg/ml (range 31–86 pg/ml) was elevated. The hepatitis panel was negative. The computed tomography scan of the abdomen revealed hepatosplenic megaly and ascites. Bone marrow aspiration and biopsy revealed 3% plasma cells without monoclonal protein. Serum protein electrophoresis revealed an immunoglobulin-A3 (IgA3) protein but no quantifiable monoclonal protein.

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