Mr. Y, a 60-year-old Caucasian male, presented to the clinic with complaints of fatigue, headache, and dry eyes but no visual 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 a physical examination and had increased in severity and frequency.

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 not tender 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 cytology and cultures were negative as was the computed tomography scan of the head.

Mr. Y was diagnosed with polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes syndrome (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, polynuropathy) syndrome. Important traits not included in the acronym include elevated levels of VEGF, sclerotic bone lesions, Castleman disease (a rare disorder characterised by benign lymph node tumors), papilledema, peripheral edema, ascites, effusions, thrombocytosis, polycythemia, fatigue, and clubbing (Dispensieri, 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 cytokines, such as vascular endothelial growth factor (VEGF), has been implicated.