Diagnostic criteria for POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy, Skin changes)

(Dispenzieri A. et al., Am. J. Hematol. 89:214-23, 2014. Accessible at http://onlinelibrary.wiley.com/doi/10.1002/ajh.23644/full)

Mandatory criteria

- Polyneuropathy (typically demyelinating)
- Monoclonal plasma cell-proliferative disorder (almost always λ)

Major criteria

- Castleman disease[1]
- Sclerotic bone lesions
- Vascular endothelial growth factor (VEGF) elevation

Minor criteria

- Organomegaly (splenomegaly, hepatomegaly, or lymphadenopathy)
- Extravascular volume overload (edema, pleural effusion, or ascites)
- Endocrinopathy (adrenal, pituitary, gonadal, parathyroid, thyroid[2] and pancreatic[2])
- Skin changes (hyperpigmentation, hypertrichosis, glomeruloid hemangiomata, plethora, acrocyanosis, flushing, and white nails)
- Papilledema
- Thrombocytosis/polycythemia[3]

Diagnosis of POEMS syndrome

The two mandatory criteria
PLUS
≥ 1 major AND ≥ 1 minor criterion

Notes:
[1] Castleman disease is a variant of POEMS syndrome that occurs without evidence of a clonal plasma cell disorder. This entity should be considered separately.
[2] This diagnosis alone is not sufficient to meet this minor criterion, due to the high prevalence of diabetes mellitus and thyroid abnormalities.
[3] Approximately 50% of patients will have bone marrow changes that distinguish it from a typical MGUS (Monoclonal Gammopathy of Undetermined Significance) or myeloma bone marrow. Anemia and/or thrombocytopenia are distinctively unusual in this syndrome unless Castleman disease is present.