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


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

**Major criteria**
- Castleman disease¹
- 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, thyroid² and pancreatic²)
- Skin changes (hyperpigmentation, hypertrichosis, glomeruloid hemangiomata, plethora, acrocyanosis, flushing, and white nails)
- Papilledema
- Thrombocytosis/polycythemia³

### Diagnosis of POEMS syndrome

<table>
<thead>
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<th>The two mandatory criteria</th>
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<tr>
<td>PLUS</td>
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<td>≥ 1 major AND ≥ 1 minor criterion</td>
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**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.