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POEMS Syndrome: Clinical Features and Outcomes in a U.K. Cohort

A retrospective analysis of cases shows that smooth meningeal thickening provides an additional diagnostic clue differentiating POEMS syndrome from chronic immune demyelinating polyneuropathy.

POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin lesions) is a rare but treatable paraneoplastic disorder of plasma cells associated with sensorimotor polyneuropathy, lambda gammopathy, and elevated cerebrospinal fluid protein and serum VEGF (vascular endothelial growth factor). Missed or delayed diagnosis is common. To investigate characteristics, outcomes, and risk factors, investigators retrospectively assessed data from 100 patients with POEMS syndrome at a U.K. institution who were followed for a median of 59 months.

Common features included hypogonadotrophic hypogonadism (72%), volume overload (70%), acrocyanosis (46%), lymphadenopathy (42%), weight loss, restrictive lung disease, and pulmonary hypertension. Mean onset-to-diagnosis time was 15 months. Patients diagnosed within 6 months had lower overall neuropathy limitation scores, although more than half of patients were previously misdiagnosed with chronic immune demyelinating polyneuropathy (CIDP). The neuropathy was length-dependent in 93% of patients, with conduction velocity slowing in 85% and secondary axonal loss in 72%. Conduction block was rare. POEMS patients were more likely to have meningeal thickening than were patients with CIDP (71% vs. 0%). Without serum immunofixation, the gammopathy would have been missed in 23 patients, and serum protein electrophoresis and immunofixation were both negative in 21%.

Wheelchair or bedbound status decreased from 35% to 11% after treatment. Risk factors for progression and death included hematologic nonresponse, vascular endothelial growth factor nonresponse, and non-transplant therapy. Five-year survival was 90%. The authors acknowledge potential patient selection bias in treatment-related outcomes.

COMMENT

These researchers nicely characterize the multisystem clinical features of POEMS in this European population and corroborate the benefit of transplant therapy in certain patients. This study highlights the insensitivity of the serum assays used in isolation to diagnose the plasma cell disorder, and it identifies meningeal thickening as another useful diagnostic clue. Larger sample sizes are needed to assess the prognostic value of cardiopulmonary features. ***— Leana Doherty, MD***

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