POEMS syndrome, calciphylaxis and focal segmental glomerulosclerosis - VEGF as a possible link.

Polyneuropathy organomegaly endocrinopathy M-protein skin changes (POEMS) syndrome is a rare cause of polyneuropathy. Calciphylaxis, a severe disease leading to necrotic ulcers of the skin, is associated with POEMS syndrome and also with renal disease. This case report describes a patient with POEMS syndrome plus primary focal segmental glomerulosclerosis. A 27-year-old Caucasian woman with chronic renal insufficiency due to focal segmental glomerulosclerosis and calciphylaxis presented to our institution with polyneuropathy and encephalopathy. An extensive diagnostic workup revealed POEMS syndrome. Serum concentrations of vascular endothelial growth factor (VEGF) were highly elevated, consistent with POEMS syndrome. To our knowledge, this is the first report of a patient with POEMS syndrome and primary focal segmental glomerulosclerosis. The combination of POEMS syndrome, calciphylaxis and primary focal segmental glomerulosclerosis may be coincidental, suggesting the need for additional studies to confirm or exclude this association. VEGF may be an important pathogenetic link, suggesting that treatment with antiangiogenic agents may improve patient outcomes.