POEMS syndrome is an acronym of polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes), it is rare paraneoplastic disorder, related to plasma cell neoplasm. The pathophysiology has not yet been clearly elucidated. The production of proinflammatory cytokines is thought to play an important role in the pathogenesis of the POEMS syndrome. Vascular endothelial growth factor level reflects disease activity, which is helpful for diagnosis and evaluation of treatment response. Conventional agents, such as corticosteroids and melphalan are effective and safe combination regimen. Autologous hematopoietic stem cell transplantation is the another option for high-risk, transplant eligible patients. Radiotherapy is effective in some patients with localized lesions. Anti-myeloma agents, lenalidomide, thalidomide, and bortezomib have shown good treatment outcomes for POEMS syndrome; however, large-scale studies with long term follow-up are required. Early identification and active treatment can improve the outcome of POEMS syndrome.