POEMS症候群

～いかに考え，治療するか～

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●VEGF
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●サリドマイド
●レナリドミド

Abstract

POEMS syndrome is a rare plasma cell dyscrasia presenting with polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal plasma cells (almost always λ type), and skin changes. The pathogenesis of POEMS syndrome is not well understood ; however, overproduction of vascular endothelial growth factor (VEGF), probably secreted by plasma cells or abnormally released by activated platelets, has been considered responsible. Increased VEGF production appears to cause most disease features by inducing angiogenesis and microvascular hyperpermeability. Diagnoses are often delayed because of the rarity of the disease and can be mistaken for other neurologic disorders. Treatment was largely unsuccessful until the past decade. More recently, considerable progress has been made in interpreting the pathogenesis, clinical features, and diagnosis, and new treatment options have become available, including novel agents such as thalidomide, lenalidomide, and bortezomib, and high-dose melphalan followed by autologous stem cell transplantation (ASCT). ASCT has been reported to be a successful treatment strategy in patients < 65 years old. With stabilization of the serum VEGF level with induction therapy, the transplant-related mortality has been low and the periengraftment complications become manageable. However, a certain number of patients experience disease relapse after ASCT in the long-term follow up. Therefore, all patients should be followed up carefully, and effective salvage strategies are required.

POEMS syndrome：
pathophysiology, diagnosis, and treatments.
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