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Successful Autologous Stem Cell Transplantation for POEMS Syndrome

Ali Ünal, Serhat Çelik, Zeynep Tuğba Güven, Mustafa Baydar, Muzaffer Keklik, Gülşah Akyol, Mustafa Çetin, Leylagül Kaynar

Erciyes University, Faculty of Medicine, Department of Hematology, Kayseri, Turkey

Introduction

(Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, Skin changes) is characterized by the presence of a monoclonal plasma cell disorder, peripheral neuropathy, and one or more of the following features: osteosclerotic myeloma, Castleman disease, increased levels of serum vascular endothelial growth factor, organomegaly, endocrinopathy, edema, typical skin changes, and papilledema. The underlying cause of the disorder is not well understood. POEMS syndrome is a chronic disorder, with a median survival time of 8-14 years. There is no standard treatment; management depends on the underlying plasma cell disorder and may include radiation therapy, chemotherapy(CT), and/or autologous hematopoietic stem cell transplantation(ASCT).

Case report

45-year-old woman has 6 years of neuropathy and 2 years of DM. She was admitted to our clinic with the complaint of weakness, abdominal swelling, 40 kg loss in the last year, and numbness in the hands. She has acrocyanosis and white nails. HSM and ascites were detected. There is polyneuropathy in Electromyography. Plasma cell neoplasia was reported as lambda monoclonal in her bone marrow biopsy. At the same time, hypothyroidism and adrenal insufficiency were detected. Because of she has Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal plasma proliferative disease and Skin changes, POEMS was diagnosed. Bortezomib, Cyclophosphamide, Dexamethasone(VCD) was started in May 2019 and 3 cycles were given. ASCT was performed in January 2020 due to the age and initial systemic involvement of the patient. She had engrafted successfully and discharged on the 15th day of the transplant.

Discussion

Treatment of POEMS syndrome is twofold. The first is directed at treating the underlying plasma cell disorder. The second is directed toward ameliorating the specific symptoms that are apparent in each individual. The use of radiotherapy or surgical removal of osteosclerotic lesions that are localized may temporarily lead to a remission of symptoms associated with POEMS syndrome. In many cases, including those with widespread osteosclerotic lesions or diffuse bone marrow involvement, therapy with certain CT, like VCD or melphalan may alleviate symptoms associated with POEMS syndrome. Many patients may be offered high-dose chemotherapy with ASCT.

Conclusion

POEMS is a very rare disease that is difficult to diagnose. As in our patient, ASCT is the most important treatment option in young patients with multiple lesions and bone marrow involvement.

Contact

Ali Ünal, hematoloji38@gmail.com