PS05

Sindrom POEMS - rijedak, ali često zaboravljen uzrok neuropatije

Ante Stojanović[1]; Domagoj Sajfert[1]; Maro Brbora[1]; Josip Batinić[1,2]

1 Medicinski fakultet, Sveučilište u Zagrebu, Zagreb, Hrvatska
2 Klinički bolnički centar Zagreb, Klinika za unutarnje bolesti, Zavod za hematologiju, Zagreb, Hrvatska

ID Ante Stojanović 0009-0009-9310-4426; Domagoj Sajfert 0009-0000-6367-1264; Maro Brbora 0009-0004-3482-1293; Josip Batinić 0000-0001-5595-9911 https://doi.org/10.26800/LV-145-supl7-PS05

KLJUČNE RIJEČI: POEMS sindrom; polineuropatije; autologna transplantacija

UVOD: POEMS sindrom (eng. Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal plasma cell proliferative disorder, Skin changes) je rijetki sindrom karakteriziran prisutnošću monoklonalne plazmastanične bolesti i perifernom neuropatijom, uz ostale sistemske simptome.
PRIKAZ SLUČAJA: u pedesetdevetogodišnjeg pacijenta je u travnju 2022. godine započela neurološka obrada zbog vertiginoznih smetnji i progresivne mišićne slabosti donjih ekstremiteta koji su se javili nakon preboljene COVID19 infekcije. U studenom 2022. dolazi do pogoršanja u vidu razvoja tetrapareze. Elektromiografskom obradom dijagnosticirana je teška senzomotorna polineuropatija. Laboratorijskom obradom utvrđen je monoklonski protein u serumu, zbog čega je učinjena hematološka obrada radi potvrđivanja dijagnoze monoklonske gamapatije. Temeljem navedenog, utvrđeno je da pacijent zadovoljava kriterije za sindrom POEMS. Provedeno je liječenje uvodnom terapijom po VRd protokolu (bortezomib, lenalidomid, deksametazon) uz primjenu intravenskih gamaglobulina zbog sekundarne hipogamaglobulinemije. Klinički se prati postupno kliničko poboljšanje (samostalno sjedi, hoda uz pomoć druge osobe). Liječenje je nastavljeno transplantacijom autolognih krvotvornih matičnih stanica što je učinjeno u srpnju 2023. godine; sam postupak transplantacije protekao je bez komplikacija. Nakon transplantacije prati se daljnje poboljšanje pacijentovog stanja i postupni napredak u pokretljivosti i kvaliteti života. Na kontrolu u listopadu 2023. godine pacijent dolazi samostalno pokretan.
ZAKLJUČAK: Sindrom POEMS rijedak je entitet iz grupe monoklonskih gamapatija, a može dovesti do teških oštećenja i invaliditeta. Dijagnostika je teška i najčešće dugotrajna. Pravovremenom i ciljanom terapijom može se postići reverzibilnost oštećenja i povratak kvalitete života.
Ključne riječi: POEMS sindrom; polineuropatije; autologna transplantacija.

POEMS syndrome – a rare but often forgotten cause of neuropathy

INTRODUCTION: POEMS syndrome (eng. Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal plasma cell proliferative disorder, Skin changes) is a rare syndrome characterized by the presence of monoclonal plasma cell diseases and peripheral neuropathy, along with other systemic symptoms.
CASE REPORT: A fifty-nine-year-old patient underwent neurological treatment in April 2022 due to vertigo disorders and progressive muscle weakness of the lower extremities that occurred after COVID-19 infection. In November 2022, deterioration in the form of the development of tetraparesis was observed. Electromyography showed severe sensorimotor polyneuropathy. Laboratory analysis revealed a monoclonal protein in the serum. Hematological work-up was done which confirmed the diagnosis of monoclonal gammopathy. Based on the above the patient met the criteria for POEMS syndrome. Treatment commenced with the VRd protocol (bortezomib, lenalidomide, dexamethasone) along with intravenous gammaglobulins due to secondary hypogammaglobulinemia. Gradually, clinical improvement was observed (sitting independently, and walking with assistance). Treatment continued with autologous hematopoietic stem cell transplantation in July 2023, which proceeded without complications. After the transplantation, the patient's condition and gradual progress in mobility and quality of life are observed. In the October 2023 follow-up, the patient arrived independently mobile.
CONCLUSION: POEMS syndrome is a rare entity from the group of monoclonal gammopathy, and it can lead to severe organ damage and disability. Diagnostic is difficult and usually long-lasting. Timely and targeted therapy can achieve reversibility of damage and return of quality of life.
KEYWORDS: POEMS Syndrome; polyneuropathies; autologous transplantation