Abstract

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PML-IRIS in a rare myeloma subtype, the non-secretory multiple myeloma (NSMM)

E. Marrone[1], R. Della Pepa[2], G. Maniscalco[3], F. Gallucci, U. Malgeri[1], C.Romano[1], F. Zeccolini[4], P. Morella[1]

[1]Internal Medicine Unit 3, A. Cardarelli Hospital, Naples
[2]Department of Hematology, Federico II University Hospital, Naples, Italy
[3]Multiple Sclerosis Regional Center, "A. Cardarelli" Hospital, Naples, Italy
[4]Neuroradiology Unit, "A. Cardarelli" Hospital, Naples, Italy

Background

Progressive multifocal leukoencephalopathy (PML) is a rare opportunistic infection caused by John Cunningham virus (JCV) in the context of immunosuppression such as HIV, malignancy, and certain immunomodulatory medication. PML has been reported only rarely in multiple myeloma (MM) patients. We describe a rare case of PML with immune reconstitution inflammatory syndrome (IRIS) in a patient with non-secretory multiple myeloma (NSMM).

Case report

A 65-year-old woman with a medical story of NSMM for about 10 years, treated with multiple lines of chemotherapy and undergoing two autologous stem cell transplants, now refractory, was admitted to the hospital presenting confusion, short-term memory impairment and behavioral changes. CT head showed bilateral white matter changes raising the possibility of central nervous system (CNS) infection or intraparenchymal CNS myeloma infiltration. MRI brain revealed multiple areas of hyper intensity on T2-weighted sequences which did not enhance but many of which showed diffusion restriction suggesting for PML-IRIS. A lumbar puncture was undertaken. Cerebrospinal fluid was positive for the JCV confirming the diagnosis of PML. Because patient presented PML-IRIS, steroid therapy was prescribed with a modest benefit. Three months later the patient died of a massive cerebral haemorrhage.

Conclusions

PML in MM is a rare consequence of the disease and its immunosuppressive profile, but it is underdiagnosed. Thus, an appropriate clinical approach compatible with early investigation and diagnosis and treatment of PML should be developed.