Neurological complications of common variable immunodeficiency: the immune system in chaos

Ana Novo1, P. Carvalho1, A. Nogueira1, J. Pita2, C. Constanço3, F. Vieira4, L. Sousa1, E. Faria2, and S. Batista1

1Department of Neurology, Centro Hospitalar e Universitário de Coimbra, Portugal
2Department of Immunoallergology, Centro Hospitalar e Universitário de Coimbra, Portugal
3Department of Clinical Haematology, Hospital São Teotónio, Portugal
4Department of Anatomic Pathology, Hospital São Teotónio, Portugal

Correspondence: amfloresnovo@gmail.com

Special Issue on Controversies in Neurology. From the 10th World Congress on Controversies in Neurology (CONy), Lisbon, Portugal. 17–20 March 2016.

Abstract

Introduction: Autoimmune disorders and granulomatous disease are well established complications of Common Variable Immunodeficiency (CVID). However, central nervous system (CNS) involvement is rare in these disorders.

Case 1: 42-year-old woman with recurrent occipital headaches since the age of 32 (1997). Brain-MRI showed hyperintense periventricular T2-lesions. Autoimmunity, serologic and CSF tests, SACE and visual evoked potential were normal. Protein electrophoresis revealed hypogammaglobulinemia G and A. In 2006, the patient presented left hemiparesis and hemihypoesthesia with spontaneous remission after 3-4 days, and recurrent mucocutaneous infections. Brain-MRI: multiple T2 hyperintense lesions in periventricular and bilateral frontal subcortical white matter. Cerebral vasculitis associated with CVID was diagnosed. Patient started a 30gr/month intravenous immunoglobulin (IVIG) therapy and achieved remission of infections and neurological symptoms.

Case 2: 21-year-old man with a previous severe dental abscess, megaloblastic anemia and hypogammaglobulinemia (G and A). Bone marrow biopsy showed non-caseous granulomas. Infectious and neoplastic causes were excluded. In 2014, the patient was hospitalized due to new onset headache. Brain-MRI: multiple large and enhancing brain lesions. Granulomatous disease associated with CVID with CNS involvement was diagnosed. A 35gr/month IVIG therapy was started. In 2015, patient was rehospitized due to flaccid paraparesis and urinary retention. Neuraxis-MRI: new multiple brain lesions and active cervicothoracic lesions. Improvement was observed in response to corticotherapy. Due to persistence of lesional activity, cyclophosphamide 400mg/m2/month was initiated.

Conclusion: These cases illustrate the difficulty in diagnosis and treatment of CVID with neurological complications, since they entail the approach of a paradoxically hypoactive and hyperactive immune state.

Citation: Novo et al. Neurological complications of common variable immunodeficiency: the immune system in chaos. International Journal of Clinical Neurosciences and Mental Health 2016; 3(Suppl. 1):P137

Published: 16 March 2016

Open Access Publication Available at http://ijcnmh.arc-publishing.org

© 2016 Novo et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.