

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, S. Batista¹

¹*Department of Neurology, Centro Hospitalar e Universitário de Coimbra, Portugal*

²*Department of Immunoallergology, Centro Hospitalar e Universitário de Coimbra, Portugal*

³*Department of Clinical Haematology, Hospital São Teotónio, Portugal*

⁴*Department of Anatomic Pathology, Hospital São Teotónio, Portugal*

amfloresnovo@gmail.com

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 dose 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 rehospitalized 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, cyclophosphamid 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.