

## NEUROLOGICAL COMPLICATIONS OF COMMON VARIABLE IMMUNODEFICIENCY: THE IMMUNE SYSTEM IN CHAOS

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**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/m<sup>2</sup>/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.