Multiple sclerosis, Sjogren and/or another autoimmune disorder?

Adriana Rua¹, J. Alves², C. Pereira², G. Carvalheiras³, S. Cavaco⁴, and A. Martins da Silva¹

1Neurology, Centro Hospitalar do Porto - Hospital de Santo António, Portugal
2Neurorradiology, Centro Hospitalar do Porto - Hospital de Santo António, Portugal
3Internal Medicine - Clinical Immunology Unit, Centro Hospitalar do Porto - Hospital de Santo António, Portugal
4Neuropsychology Unit, Centro Hospitalar do Porto - Hospital de Santo António, Portugal

Correspondence: adrrua@gmail.com

A 28-year-old female, with prior history of peripheral facial palsy at age 12, developed signs of depression four months after giving birth to a healthy son. In the following years, her behaviour deteriorated (e.g., carelessness with chores, childish behaviour, hypersomnia, memory difficulties, psychomotor slowness, irritability, inappropriate laughter, unawareness of behavioural changes, and occasional incontinence). At 33, the patient was referred to neurology and the neurological examination only revealed fragmented saccadic movements and horizontal-torsional nystagmus. The neuropsychological assessment showed significant impairment in multiple cognitive domains (attention, psychomotor speed, visuo-spatial functions, memory, and executive functions). MRI scans disclosed multiple T2-weighted hyperintense lesions in supratentorial region (e.g., periventricular, subcortical, juxta-cortical areas, and corpus callosum), infratentorial territory (cerebellum and brainstem), and cervical medulla suggestive of inflammatory/demyelinating aetiology. There were also signs of subcortical atrophy. To exclude other causes of white matter disease, a comprehensive series of laboratory studies were carried out. CSF had elevated IgG index and oligoclonal bands. Visual evoked potentials revealed bilateral prolonged latencies. The immunological tests found ANA=1/640 and high anti-thyroid antibodies. These set of paraclinical findings were supportive of Multiple Sclerosis–cortical variant. A course of methylprednisolone did not have a significant effect. One year later, the patient developed Sicca syndrome and diarrhoea of unknown aetiology. Additional investigation revealed ANA=1/1280 and salivary gland scintigraphy and biopsy compatible with Sjogren syndrome. Neuropsychological and MRI findings did not show significant changes from prior examination. The prolonged neuropsychiatric symptoms associated with a puzzling set of clinical and paraclinical findings pose a diagnostic challenge.

Abstract

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