



CASE REPORT

Neuromyelitis Optica Spectrum Disorders: a definite case with Autoimmune Thyroiditis

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Special Issue on Inflammatory Demyelinating Diseases of the Central Nervous System

Abstract

Introduction: The concept of Neuromyelitis Optica (NMO) spectrum disorders (NMOSD) encompasses patients with or without NMO-Immunoglobulin G antibodies, fulfilling several clinical and imagiological criteria defined in the international consensus diagnostic criteria of 2015. The coexistence of other autoimmune disorders is common and considered supportive of NMOSD" diagnosis.

Case Report: A 24 year-old female was admitted due to a sudden painful hypovision in the left eye. The etiological investigation was negative but brain MRI showed T2 hyperintensity of the left optic nerve with gadolinium enhancement, but no signal changes in the brain parenchyma. Eleven years before, she had been diagnosed with autoimmune thyroiditis based on hypothyroid goiter with biopsy-proven chronic lymphocytic inflammation and positive thyroid-related antibodies. Two years after optical neuritis, she developed sensory complaints below a thoracic defined level. MRI revealed a diffuse T2 hyperintense area from D2 to D5. She tested positive for antibody anti-NMO/Aquaporin-4. One month later she had a new episode of left optic neuritis. With two optic neuritis and one longitudinally extensive myelitis, with positive serum test for NMO antibody, she was diagnosed with NMOSD with hypothyroidism and started Azathioprine.

Discussion: In up to 20-30% patients with NMOSD there is an association with autoimmune thyroiditis. In the literature the reference to "thyroid disease" and seroprevalence of thyroid-related antibodies in NMOSD patients, ranges between 5.6% and 20.5%, but case reports/series with definite clinical, analytical and histological characterization of the thyroiditis are scarce. We present a case of a patient with clear data consistent with autoimmune thyroiditis who, more than a decade later, was diagnosed with NMO.

Keywords: Neuromyelitis Optica, Neuromyelitis Optica Spectrum Disorders, Autoimmune Thyroiditis.

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Citation: Braz et al. Neuromyelitis Optica Spectrum Disorders: a definite case with Autoimmune Thyroiditis. International Journal of Clinical Neurosciences and Mental Health 2016; 3(Suppl. 3):S04

DOI: [http://dx.doi.org/10.21035/ijcnmh.2016.3\(Suppl.3\).S04](http://dx.doi.org/10.21035/ijcnmh.2016.3(Suppl.3).S04)

Published: 20 Oct 2016

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

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Introduction

Neuromyelitis Optica Spectrum Disorders (NMOSD) defines an inflammatory autoimmune syndrome of the central nervous system, with distinctive clinical, neuroimaging and laboratory findings, that preferentially affect the optic nerves and spinal cord. Many NMOSD patients have detectable serum antibodies that target the water channel aquaporin-4 (AQP4-immunoglobulin G), which are highly specific and have pathogenic potential. Recently an international consensus defined clinical spectrum of NMOSD and unified diagnostic criteria [1].

In up to 20–30% of NMOSD patients, the disorder occurs in association with a variety of organ and non-organ-specific autoimmune diseases, which may become symptomatic before or after the NMOSD clinical syndrome. In addition, up to 40% of NMOSD patients have coexistent antibodies without an obvious autoimmune illness [2, 3]. Autoimmune thyroiditis is considered among the organ-specific autoimmune diseases that may co-exist with NMOSD [4].

Case Report

In 2010, a 24-year-old, Caucasian female was admitted in the Neurology department due to a sudden painful loss of vision in the left eye. Eleven years before, the patient had been studied for a hypothyroid goiter with low D4 hormone level (3.6 mcg/dL) and elevated thyroid-stimulating hormone (TSH) (139.0 mcU/mL). At that time, she undertook thyroid biopsy that showed a chronic inflammatory process with lymphocytic predominance; the study of anti-peroxidase and anti-thyroglobulin antibodies was positive at high titers (340.9 UI/mL and 180.3 UI/mL, respectively). She was then diagnosed with autoimmune thyroiditis and started on levothyroxine, lying thereafter asymptomatic, in an euthyroid state.

In 2010, the study done to assess left eye hypovision showed: visual evoked potentials suggesting left pre-chiasmatic and bilateral retro-chiasmatic commitment, normal somatosensory evoked potentials, absence of oligoclonal bands (OCB) in CSF and negative infectious studies. Brain MRI showed T2 hyperintensity of the left optic nerve and contrast enhancement in the same structure, but no signal changes in the brain parenchyma. The patient was diagnosed as having a left optic neuritis, and did intravenous (IV) corticosteroid for five days with partial clinical recovery.

Two years later (2012), she was admitted for paresthesia and dysesthesia with sensory level by T4 and again was treated with IV corticosteroid for five days without significant recovery. Serum biochemistry, virological and immunological studies revealed no changes. The CSF study presented slightly perproteinorrhachia (56mg/dL), 1 cell-lymphocyte and normal glucose level. The OCB and infectious study on CSF were again negative. Spinal MR revealed a diffuse hyperintense area on T2 images from D2 to D5 (Figure 1a).

About 3 weeks later she was again evaluated for severe dysesthesia and hypoesthesia with sensory level for D4 and a new distal motor deficit (G4+/5) in her right lower limb. She was treated with IV corticosteroids for 8 days and recovered from the motor deficit but kept sensory changes and tonic spasms in the right leg (managed with gabapentin and clonazepam). She tested positive for AQP4-IgG in serum. Spinal MRI revealed a decrease in the extension of the spinal cord lesion identified one month before. One month later the patient had a new episode of left optic neuritis from which she recovered fully after five days of IV corticosteroid therapy.

Given the two left optic neuritis and the longitudinally extensive myelitis, with positive AQP4-IgG, she was diagnosed with Neuromyelitis Optica (actually NMOSD with AQP4-IgG). The patient started Azathioprine with increasing doses until 125mg daily. One year later (2013), a spinal MRI was done and showed complete absence of lesions in the spinal cord (Figure 1b). The patient is currently asymptomatic and without neurologic sequelae.

Discussion

Autoimmune thyroiditis is considered among the organ-specific autoimmune diseases that may co-exist with NMOSD [4] and reported as one of the most common autoimmune disease associated with it [5]. However, the literature is ambiguous and imprecise in what concerns to thyroid disease associated with NMOSD. In a large Japanese cohort of NMOSD patients, “history of thyroid disease” (including chronic thyroiditis, Graves’ disease or benign thyroid tumors-without further clinical or analytical definition) was present in 13.6% of patients [6]; in a Chinese study the prevalence of co-existing “thyroid disease”

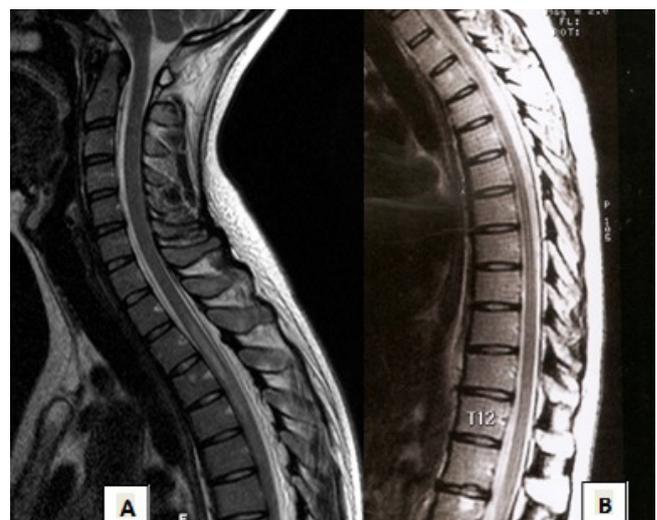


Figure 1. Spinal cord MRI: Axial T2-weighted sequences showing: (a) extended hyperintense signal in the thoracic spinal cord from D2-D5 level (taken during the myelitis episode) and (b) showing no sign of spinal lesion (taken 1 year after the treatment initiation).

among NMOSD patients, based on seroprevalence of thyroid-related antibodies, ranged between 5.6% and 20.5% [7]; in an American cohort of NMOSD patients, the coexistence of autoimmune thyroid disease was 16.7% [8].

To our knowledge, since the identification of AQP4-IgG, only one case-report of NMOSD associated autoimmune thyroiditis (Hashimoto thyroiditis) exists; however the clinical, analytical and histological characterization of the thyroiditis are not provided [9]. The available data suggests that the co-existence of autoimmune thyroiditis and NMOSD might be greater than what is indicated by the number of case-reports with well-defined data. We present the case of a patient with clinical, histological and analytic data consistent with autoimmune thyroiditis, who more than a decade later was diagnosed with Neuromyelitis Optica.

Abbreviations

AQP4: Aquaporin-4; IgG: Immunoglobulin G; IV: Intravenous; NMOSD: Neuromyelitis Optica Spectrum Disorders; OCB: Oligoclonal bands; TSH: Thyroid-stimulating hormone

Competing interests

The authors declare no conflict of interest.

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