Journal of Clinical Neuroscience 19 (2012) 969–974

Contents lists available at SciVerse ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Clinical Study

Brain MRI abnormalities in Brazilian patients with neuromyelitis optica

Cíntia Elias Pires [a,b,*], Christianne Martins Correa da Silva [a], Fernanda Cristina Rueda Lopes [a,b], Fabiola Rachid Malfetano [a], Valéria C.S.R. Pereira [a], Tadeu Kubo [b], Paulo R.V. Bahia [a], Soniza Vieira Alves-Leon [a], Emerson L. Gasparetto [a,b]

[a] Diagnostic Radiology and Neurology Department, University Federal of Rio de Janeiro, Rio de Janeiro, Brazil
[b] Clínica de Diagnóstico por Imagem (CDPI)/Diagnósticos da América (DASA), Avenue das Américas 4666, Sala 325, Barra da Tijuca, Rio de Janeiro 22649900, Brazil

ARTICLE INFO

Article history:
Received 18 July 2011
Accepted 4 October 2011

Keywords:
Brain abnormalities
Brazilian patients
Neuromyelitis optica
MRI

ABSTRACT

Brain abnormalities in neuromyelitis optica (NMO) have been reported previously, but the pathophysiological mechanism and clinical relevance of these abnormalities are poorly understood. We assessed the prevalence and patterns of brain MRI abnormalities in a Brazilian cohort of patients with NMO. Conventional brain MRI and medical records from 24 Brazilian patients with NMO were retrospectively evaluated. Brain MRI were classified into four subgroups: normal, non-specific lesions, multiple sclerosis (MS)-like lesions, and typical lesions. Brain lesions were detected in 19 patients (79.2%). Fourteen patients (58.3%) had non-specific lesions, three (12.5%) had MS-like lesions, and two (8.3%) had typical lesions. Differences between these subgroups with respect to the Expanded Disability Status Scale (EDSS) scores ($p = 0.86$) were not significant. This study demonstrates a high prevalence of brain abnormalities in Brazilian patients with NMO; however, we did not find a significant relationship between these abnormalities and EDSS scores.

1. Introduction

Neuromyelitis optica (NMO, also known as Devic's syndrome) is an uncommon but severe inflammatory/demyelinating disorder that affects the central nervous system (CNS), with a particular predilection for the optic nerves and spinal cord. NMO was previously considered an idiopathic disease, but as a result of new histopathological, immunological, and neuroimaging findings the condition has been characterised as the first CNS immune-mediated channelopathy. The target antigen in NMO is the aquaporin-4 (AQP4) protein, which is the predominant water channel in the CNS.[1]

There is currently no "gold standard" diagnostic test for NMO; rather, the diagnosis is based upon clinical, laboratory, and imaging findings. In 1999, Wingerchuk et al. proposed diagnostic criteria for NMO that emphasised the restriction of the disease to the optic nerves and the spinal cord.[2] However, studies later revealed that a significant number of patients who displayed a clinical course that was otherwise highly compatible with NMO had brain abnormalities that were detectable by MRI.[3–5] Therefore, in 2006, revised NMO diagnostic criteria were proposed that included an onset brain MRI non-diagnostic of multiple sclerosis (MS) as a supportive criterion for NMO.[6]

Many authors have reported varied frequencies of brain MRI abnormalities in patients with NMO. In Asia, brain MRI lesions have been observed in up to 46.0% of Japanese[7], and 84.8% of Chinese, patients with NMO.[8] Pittock et al. reported that brain lesions were detected in 60.0% of patients with NMO in the United States, and other research has shown that brain lesions were detected at a rate of 65.5% in a Cuban population.[3,9] In Europe, the following frequencies of brain MRI abnormalities in patients with NMO have been reported: 41.6% in patients in Great Britain, 17.3% in Italy, and 15.3% in France.[10–12] In a report discussing the clinical features and outcomes of 24 Brazilian patients with NMO, brain MRI abnormalities were observed at a frequency of 38.0%.[4]

Despite these reportedly high frequencies of brain lesions, brain involvement patterns in NMO are still poorly understood. Brain biopsies and pathological results from patients with NMO have shown a variety of brain lesions. Recently, distinctive lesions in sites of high AQP4 expression, such as the hypothalamus and periependymal regions, have been reported. These lesions could be characteristic of, if not specific to, NMO or its spectrum of disorders, and they have been reported in up to 10.0% of patients.[5,13,14] However, most brain lesions described in imaging studies to date have been non-specific and asymptomatic. Pittock et al. evaluated brain MRI from 60 patients with NMO and classified the lesions found into four subgroups: (i) normal/no lesion (40.0%); (ii) non-specific (42.0%); (iii) MS-like (10.0%); and (iv) typical (8%).[3] In this study, however, only 17.0% of the patients with brain lesions were

* Corresponding author. Tel.: +55 (21) 98563331.
E-mail address: cintiaepires@yahoo.com.br (C.E. Pires).

0967-5868/$ - see front matter © 2011 Elsevier Ltd. All rights reserved.
doi:10.1016/j.jocn.2011.10.009

Table 1
MRI sequencing protocols used in Brazilian patients with brain MRI abnormalities in neuromyelitis optica

Parameter	T1-weighted MRI with magnetisation transfer	Three-dimensional FLAIR	T2-weighted MRI	DWI
Imaging planes	Axial	Sagittal	Axial	Axial
Repetition time (ms)	505	5000	3700	4000
Echo time (ms)	9	418	102	82
Field of view (mm)	187 × 250	226 × 260	198 × 220	377 × 615
Section thickness (mm)	5	1	3	3
Intersection gap (mm)	2	None	3.3	4.2
Matrix	256 × 144	256 × 218	320 × 288	160 × 160

DWI = diffusion-weighted image, FLAIR = fluid-attenuated inversion recovery.

Table 2
Imaging features used to classify brain lesions detected by conventional MRI

Non-specific	Non-enhancing deep white matter lesions that were not ovoid, did not abut, or were not perpendicular to, the ventricles, or were too few to satisfy the Barkhof et al. criteria for multiple sclerosis (MS)[15]
MS-like	Multiple deep white matter ovoid lesions, with peri-ventricular predominance, often oriented perpendicular to the ventricles
Typical	Large (> 3 cm) confluent hemispheric lesions, periependymal and diencephalic lesions (involving the thalamus and hypothalamus)

Table 3
Demographic and clinical features of 24 Brazilian patients with neuromyelitis optica

Variable	Patients with NMO n = 24, mean ± SD, (%)
Gender	
Male	5 (21)
Female	19 (79)
Ethnicity	
African	16 (67)
Caucasian	8 (33)
Age (years)	43 ± 11.7
Age at onset	31.66 ± 9.66
Disease course	10.54 ± 6.97
No. of attacks	6.12 ± 3.79
EDSS score	5.12 ± 2.78
NMO–IgG	
Negative	7 (53)
Postive	6 (47)
Not tested	11

EDSS = Expanded Disability Status Scale, IgG = immunoglobulin G, NMO = neuromyelitis optica, SD = standard deviation.

Table 4
Brain MRI findings of 24 Brazilian patients with neuromyelitis optica

Brain MRI patterns	Patients, n = 24, (%)
Normal	5 (20.83)
Abnormal (total)	19 (79.17)
Non-specific	14 (58.33)
Other abnormalities	5 (20.83)
Multiple sclerosis–like	3 (12.5)
Typical	2 (8.33)

symptomatic. In agreement with these findings, Li et al. found no association between Expanded Disability Status Scale (EDSS) scores and brain abnormalities in Chinese patients with NMO.[8] Therefore, it remains unclear whether the presence of brain abnormalities is related to clinical status or prognosis in patients with NMO.

The goal of this study was to assess the prevalence and patterns of brain MRI abnormalities in a Brazilian cohort of patients with NMO.

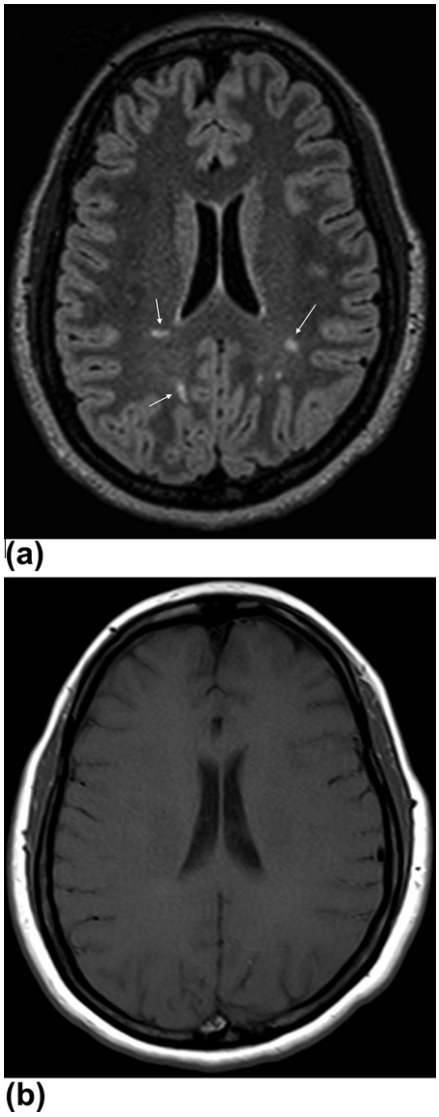

Fig. 1. A 33-year-old female with a two-year history of neuromyelitis optica. (a) Axial fluid-attenuated inversion recovery and (b) axial T1-weighted post-gadolinium MRI demonstrating subcortical and periventricular small hyperintense non-enhancing lesions (white arrows, non-specific pattern).