

Introduction:

Demyelinating diseases of the central nervous system (DD-CNS) may share common features, including their opening symptom. However, the frequency of these symptoms at disease presentation may differ for each condition.

Objective:

To describe the frequency of the various inaugural manifestations of CNS demyelination in a large cohort of patients.

Methods:

We selected patients with DD-CNS seen at CIEM MS Research Center from January 2010 to December 2019. Optic neuritis (ON) was classified as typical or atypical if they had or not the characteristics shown by the Optic Neuritis Treatment Trial. McDonald 2017 criteria and Wingerchuk 2015 criteria were used for diagnosis of multiple sclerosis and neuromyelitis optica spectrum disorder (NMOSD) respectively.

Results:

Out of 955 patients with DD-CNS, myelitis occurred at disease opening in 480 (50.3%) patients and ON in 311 (32.6%). Typical ON was found in 101 (37.3%) (monophasic isolated idiopathic ON in 13 (4.8%), and multiple sclerosis related ON in 88 (32.4%) patients. Atypical ON occurred in 170 (62.7%), comprising recurring idiopathic optic neuritis (RION) in 26 (9.6%) patients, chronic relapsing inflammatory optic neuritis (CRION) in 2 (0.74%), optic neuritis of the neuromyelitis spectrum disorder (NMOSD-ON) in 73 (26.9%); post-immunization and para-infectious ON in 21 (7.8%); ON in ADEM (ADEM-ON) in 7 (2.6%); and ON in association with systemic autoimmune diseases in 7 (2.6%) patients. In 32 (11.8%) patients the ON etiopathogenesis could not be determined. Other symptoms at disease onset included brainstem syndrome in 281 (29.4%); area postrema syndrome in 45 (4.7%); cerebral symptoms in 22 (2.3%); and diencephalic syndrome in 3 (0.3%) patients.

Conclusions:

Myelitis and the different forms of ON are the most common inaugural symptoms of DD-CNS.

References:

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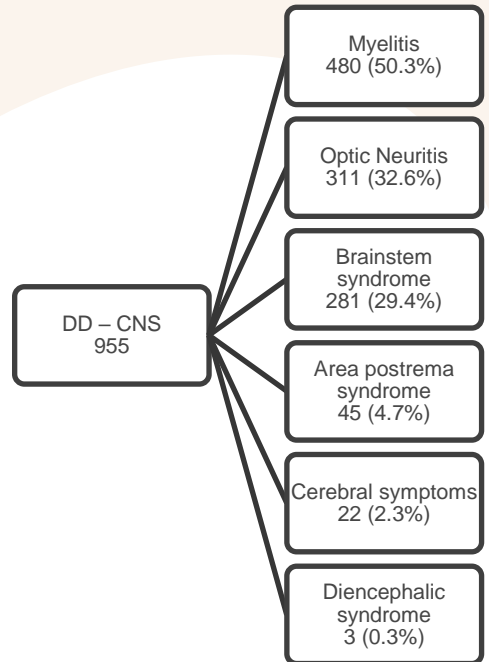


Figure. Opening symptoms of DD – CNS for study patients.

Table. Types of ON at onset of DD – CNS.

Types of optic neuritis		N
Typical 101 (37.3%)	Idiopathic isolated ON	13 (4.8%)
	MS-related ON	88 (32.4%)
Atypical 170 (62.7%)	NMOSD-related ON	73 (26.9%)
	Recurring idiopathic ON	26 (9.6%)
	Other etiologies of ON	71 (26.2%)