

Introduction:

Optic neuritis (ON) is a common opening symptom of the various demyelinating diseases of the central nervous system (DD-CNS).

Objective:

To describe the frequency and characteristics of typical and atypical forms of ON at disease presentation in a cohort of patients with DD-CNS.

Methods:

We selected patients with DD whose initial symptom was ON. We classified ON as typical and atypical ON if it had or not the characteristics as shown by the Optic Neuritis Treatment Trial studies. We divided atypical ON into groups according to the presence of specific autoantibodies, the association with vaccines, previous infections or acute disseminated encephalomyelitis, or the presence of relapses.

Results:

We analyzed 271 patients who presented ON at disease onset (101 of them typical and 170 atypical ON). Typical ON comprised idiopathic isolated ON in 13 (overall percentage of 4.8%), and ON associated with multiple sclerosis in 88 (overall percentage of 32.5%). Final diagnosis of patients with atypical ON included neuromyelitis optica spectrum disorders (NMOSD) in 73/170 (26.9%), with an overall percentage of 42.9%; recurring idiopathic ON in 26/170 (15.3%); MOG-related ON in 2/170 (1.2%); ADEM-related ON in 7/170 (4.1%); systemic autoimmune-related ON in 7/170 (4.1%); chronic relapsing inflammatory optic neuropathy in 2/170 (1.2%); parainfectious ON in 7/170 (4.1%); vaccine-related ON in 14/170 (8.2%). Thirty-two (18.8%) patients had atypical ON of unknown etiology. In an accompanying work we show that atypical ON could be distinguished from typical ON by their distinct phenotype and visual outcome.

Conclusions:

Optic neuritis at disease presentation occurs in different frequencies in the various DD of the CNS. Multiple sclerosis related ON and NMOSD-ON are the most prevalent forms. Their careful characterization may help to establish the differential diagnosis between them.

Table. Forms of optic neuritis at onset of demyelinating diseases. N = 271

Forms of optic neuritis		N (%)
Typical		101 (37.3)
	Idiopathic isolated ON	13/101 (12.9)
	MS-related ON	88/101 (87.1)
Atypical		170 (62.7)
	NMOSD-related ON	73/170 (26.9)
	Recurring idiopathic ON	26/170 (15.3)
	Vaccine-related ON	14/170 (8.2)
	ADEM-related ON	7/170 (4.1)
	Systemic autoimmune-related ON	7/170 (4.1)
	Parainfectious ON	7/170 (4.1)
	MOG-related ON	2/170 (1.2)
	CRION-related ON	2/170 (1.2)
	ON of unknown etiology	32/170 (18.8)

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