Central Nervous system involvement in CIDP

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Background and Objective

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired heterogeneous autoimmune disorder affecting the peripheral nerves, causing weakness and sensory symptoms and signs.

The diagnosis of CIDP is based on a combination of clinical and electrophysiological criteria. The most recent guidelines were developed by the European Federation of Neurological Societies (EFNS)/Peripheral Nerve Society (PNS).

It is hypothesized that both cell-mediated and humoral mechanisms could act together in an aberrant immune response directed against myelin, thus inducing demyelination in peripheral nerves. However, no specific target has been still identified.

Since some structural proteins in myelin are shared between central and peripheral nervous systems, it could be hypothesized that both myelin types could share common antigens and induce a crossed immune response. However, a combined central and peripheral demyelination has been only occasionally reported in literature.

In this study we assess the prevalence of central nervous system (CNS) demyelination in CIDP patients with or without central signs, their clinical and prognostic features, disability, and therapy response

Methods

Fourteen consecutive patients, attending the neuromuscular service at the Department of Neurology in Portogruaro Hospital, were recruited, after having been diagnosed according to EFNS/PNS electrodiagnostic criteria.

All patients underwent brain magnetic resonance (MRI) without gadolinium enhancement using T1, T2, FLAIR and DWI sequences. Clinical examination and history, Overall Neuropathy Limitations Scale (ONLS) and Modified Rankin Scale (mRS) were also performed.

Results

Prevalence of central demyelination in CIDP patient was 38.5%. No subject fulfilled Barkhof's criteria on MRI and/or the McDonald criteria for MS. No central sign in the neurological examination was noted.

CIDP with CNS involvement showed earlier age at onset (57.60 ±10.33) than those without alterations (63.50 ± 9.04 p=0.32) and longer duration of disease (10.20 ± 6.94 in CIDP with CNS involvement vs. 5.03±5.04 in CIDP without CNS involvement p=0.20).

The CNS demyelination correlated with a higher disability, on ONLS (p=0.006).

No association between central demyelination and response to therapy was observed (r=0, 29).

Conclusions

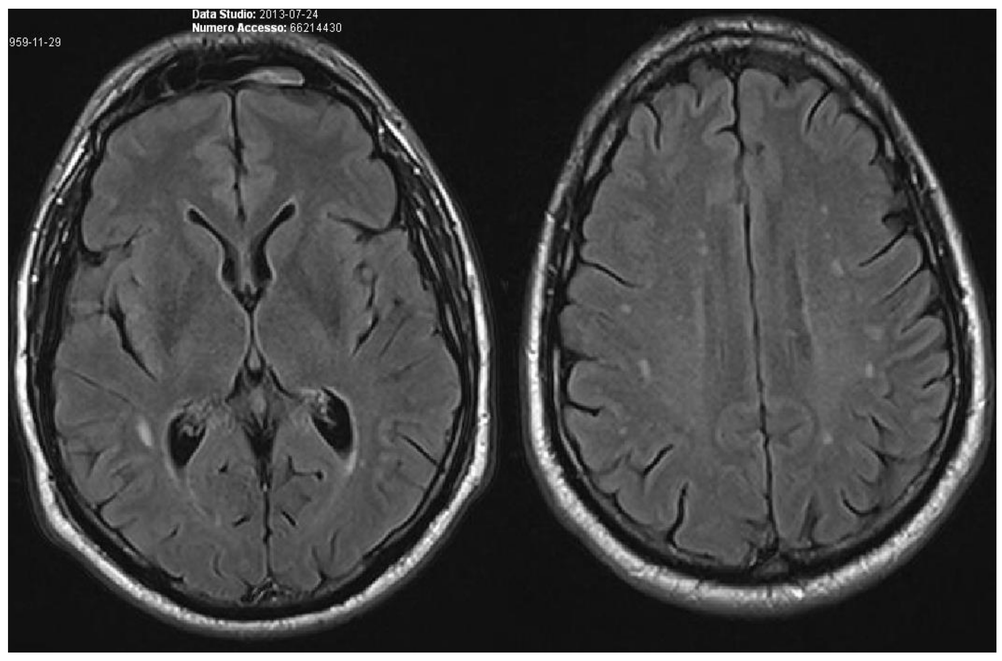
CNS demyelination in CIDP patients represents a frequent subclinical finding and the presence of central clinical signs is rare.

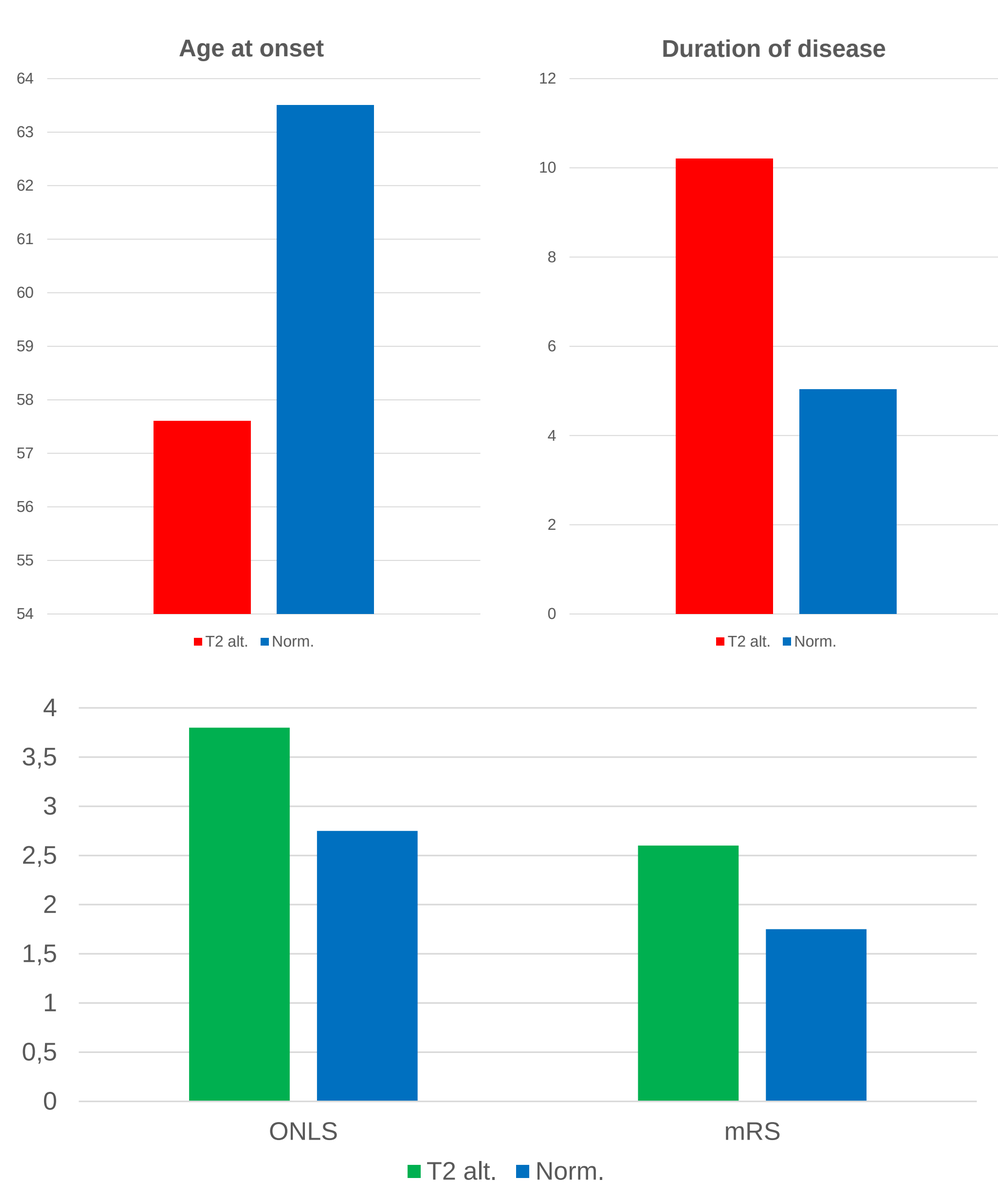
The CNS involvement is more frequent among patients with early onset and longer duration of disease and may be, likewise, an early feature or a late complication of the disease.

Central demyelination is correlated with higher disability and may be considered a negative prognostic index.

Cerebral MRI could be a useful tool to better understand the clinical course in CIDP.

	MRI +	MRI -	p
AGE	67.80 ± 9.63	68.50 ± 8.03	0.89
AGE AT ONSET	57.60 ±10.33	63.50 ± 9.04	0.32
DURATION OF DISEASE	10.20 ± 6.94	5.03 ± 5.04	0.20
ONLS	3.80 ± 0.84	2.75 ± 0.88	**0.06**
MRS	2.60 ± 0.89	1.75 ± 0.88	0.13





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