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Editorial

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Pulsed intravenous corticosteroids in chronic inflammatory demyelinating polyneuropathy: why not?

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Chronic inflammatory demyelinating polyneuropathy (CIDP) is a remitting/relapsing and/or chronic autoimmune disorder, characterized by symmetrical, sensorimotor neuropathic involvement and a slowly progressive onset. There are many clinical variants, suggesting that this disorder may not be an unique entity but rather a spectrum[1]. CIDP diagnostic criteria combine clinical and electrophysiological features. Supportive data include increased cerebrospinal fluid (CSF) protein levels[1].

Many, but not all, patients may be successfully treated with therapies aimed at arresting immunological mechanisms, such as corticosteroids[2] and intravenous immunoglobulins (IVIg)[3]. A systematic review concluded that there was no clear short-term difference with IVIg when compared with intravenous methylprednisolone and likely no improvement when compared with either oral prednisolone or plasma exchange[4]. More randomised trials are strongly needed[4].

Recently, a multicentre retrospective study compared safety and efficacy of daily prednisolone, pulsed dexamethasone, and pulsed intravenous methylprednisolone. Interestingly, corticosteroids led to improvement in 60% of subjects and to clinical remission in 61% of responders[2]. There were no significant differences in terms of safety and efficacy[2]. A therapeutic protocol with corticosteroids, with IVIg as an adjunctive treatment in case corticosteroid treatment was insufficient, could lead to improvement in 90% of CIDP patients[2].

Some patients may not respond to IVIg. For instance, we follow a 37-year-old male patient diagnosed with CIDP at age 7, based on a sensory ataxic phenotype. The diagnosis was supported by typical

electrophysiological features, CSF analysis and sural nerve biopsy. Molecular studies for hereditary neuropathies were unremarkable. At age 33 he had a severe relapse leading to a subacute flaccid, areflexic tetraparesis unresponsive to IVIg. He was then successfully treated with plasma exchange and intravenous corticosteroids. Subsequently he became corticosteroid-dependent needing chronic treatment with oral prednisone (25 mg every other day). Unfortunately, he developed bilateral cataract, right hip osteonecrosis and cushingoid appearance. Therefore, we switched the treatment to pulsed intravenous methylprednisolone (1 g daily for three consecutive days every two months). After one year of this schedule, the neuropathy is excellently controlled (apart from mild distal leg weakness) and corticosteroid toxicity is minimized, with improvement of hip osteonecrosis and regression of the cushingoid features.

In conclusion, corticosteroids are cheaper, easier to use, and much more widely available than IVIg. They have been suggested to lead to long-term remission more often than IVIg[2]. Furthermore, even if there are no significant differences in response and remission rate between these two regimens, pulsed intravenous corticosteroids have lower rates of serious adverse effects than long-term daily use[2]. Therefore, in our opinion pulsed intravenous methylprednisolone should be considered in CIDP patients, especially in non-responders to IVIg. In fact, it may represent the therapy of choice in these patients.

DECLARATIONS

Authors' contributions

Made substantial contributions to conception and design of the study and performed data analysis and interpretation: Orsucci D

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Not applicable.

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Conflicts of interest

The author declared that there are no conflicts of interest.

Ethical approval and consent to participate

Not applicable.

Consent for publication

Not applicable.

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