Neurology. 2007 Oct 23;69(17):1680-7.

Multifocal motor neuropathy: the diagnostic spectrum and response to treatment.

Slee M, Selvan A, Donaghy M.

Department of Clinical Neurology, University of Oxford, West Wing, John Radcliffe Hospital, Oxford OX3 9DU, UK.

Abstract

OBJECTIVE: To define the clinical spectrum in a large cohort of patients with multifocal motor neuropathy (MMN) and the effectiveness of IVIg treatment. We also test two neurophysiologic criteria for conduction block (CB) for relevance to treatment responsiveness.

METHODS: Retrospective case cohort analysis of 47 patients with MMN followed for up to 12 years.

RESULTS: A total of 32 (70%) had an upper-limb onset with most showing clinical features of conduction block: weakened but non-wasted muscles (67%) and differential weakness across muscles supplied by a common terminal motor nerve (54%). Differential weakness of finger extension was a characteristic early sign. Application of consensus criteria for definite CB would have denied a trial of treatment to 6 patients with a typical phenotype compared with new criteria. No association was found between CB and presence of anti-GM1 ganglioside antibody. A total of 24 (51%) patients were treated with IVIg, which was associated with a marked initial improvement in self-reported disability in most patients. The magnitude of initial disability improvement was not sustained in all patients over time. However, the majority of treated patients reported significantly less disability at last follow-up than prior to treatment. Patients converted to a domiciliary IVIg program maintained function at least as well as hospital treated patients.

CONCLUSION: The importance of the clinical phenotype of multifocal motor neuropathy (MMN) is emphasized. Neither conduction block (CB) nor antibody status is a reliable predictor of treatment responsiveness. Over-reliance upon consensus CB criteria can deny IVIg to patients with MMN who are treatment responsive.

PMID: 17954783 [PubMed - indexed for MEDLINE]