COMPARISON OF CLINICAL FEATURES IN A LARGE COHORT OF TYPICAL AND ATYPICAL CIDP

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Introduzione

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is a rare disease where an aberrant immune response against the components of the peripheral nervous system. Clinical manifestations are heterogeneous and various phenotypes have been described, so the correct classification may be difficult. The aim of the study was to characterised a large cohort of CIDP patients in the province of Padua, Treviso and Venice.

Metodi

This was a retrospective observational epidemiological study on CIDP in the provinces of Padua, Treviso and Venice. The study population was recruited from the Rare Diseases Registry of Veneto region and local registries in almost all neurological units in the three provinces. Typical and atypical variants were analyzed. All patients fulfilled the EFNS/PNS CIDP diagnostic criteria. Clinical features, biochemical data, cerebrospinal fluid analysis, comorbidities and response to common therapies were recorded and compared between the two groups.

Risultati

141 patients fullfilling the EFNS/PNS CIDP diagnostic criteria were identified. 86 (61,0%) had typical CIDP while 55 (39,0%) had atypical CIDP (18,4% purely sensory, 7,8% MADSAM, 7,2% DADS, 3,5% purely motor and 2,1% focal). Mean age was similar in the two groups (65 yrs vs 65,9 yrs in typical and atypical forms), M:F ratio was 1,7 in the typical and 2,4 in atypical forms (p=0,32). From the clinical point of view neck, facial, respiratory and ocular muscles were involved only in typical form in 18.6%, 4,7%; 4.7% and 9.3% respectively but none in atypical patiens. CSF protein was 98,4 mg/dL and 73,3 mg/dL in typical and atypical forms (p=0,20). Monoclocal gammopathy was found in 12 out of 86 patients with typical form and in 13 out of 55 patients with atypical CIDP (p=0,17); diabetes was found in 17 out of 68 patients with typical form and 9 out of 44 of atypical form (p=0,57). No difference was found in response to IVIg therapy.

Conclusioni

CIDP is a heterogeneous disease and diagnosis can be difficult in atypical cases. From our retrospective analysis we found an involvement of neck, facial, ocular and respiratory muscles in typical forms. The involvement of these muscles is reported in the literature with variable percentage but interestingly we didn't found it in atypical CIDP. No differences were found in other features such as CSF protein content or associated diseases (monoclonal gammopathy or diabetes). IVIg seems equally effective in both typical and atypical forms.