CHRONIC IDIOPATHIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY: CLINICAL, DIAGNOSTIC AND THERAPEUTICAL EVALUATION (Abstract)*. Thesis. São Paulo, 1994.

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The diagnosis of chronic idiopathic inflammatory demyelinating polyradiculoneuropathy is based on clinical, laboratory, electrophysiological and pathologic findings. To study these aspects and their correlations, and to analyse critically the therapeutics employed, verifying the presence of predictive of the clinical responses, twenty-one patients were prospectively followed for a ten-year and four-month period.

Seventeen patients developed a progressive course and four a relapsing course. Eighteen were male patients and three female. The average age was 41.1 years (range 6-63) with predominance in the fifth decade. The difference of age average for both clinical courses was statistically significant.

It was observed muscle weakness, sensory loss and areflexia in all patients. Trophic changes occurred in 52.4% and action tremor in 19.0%. Involvement of the cranial nerves was present in 33.3% of patients with facial weakness in five, unilateral optic disk atrophy in two, and ophthalmoparesis in one. Laboratory tests ruled out cases secondary to other diseases. All patients presented protein increase in the cerebrospinal fluid (CSF), with an increase of the gamma-globulin content in 35.4% which was not associated with serum increase of this globulin. Normal CSF protein concentration was observed during the development stage of the disease in four patients. Electrophysiological studies were compatible with demyelination process in 86.6% of the patients. Sural nerve biopsy showed demyelination and remyelination in all biopsies analysed, axonal changes in 90.9%, onion bulb formation in 18.2%, perivascular inflammatory cells in 54.5%, and increase in the endoneurial collagen in 100% of the patients. Neither CSF protein nor electrophysiological and pathologic findings showed correlation with clinical course.

Corticosteroids were used in all patients, being the isolated initial therapeutics in 85.7% of them; clinical improvement was observed in 61.1% and side effects in 22.2%. Sixteen patients were treated with nonsteroidal immunosuppressive agents (cytotoxic drugs): only 25% of the ones treated with azathioprine presented clinical improvement and 25% presented side effects; among patients treated with oral or "pulse" cyclophosphamide therapy, 63.6% presented improvement and 27.2% presented side effects. Eight patients were treated with plasma exchange and improvement was observed in 50%. Three patients were treated with intravenous immunoglobulin and presented no improvement. The beneficial effect of immunosuppressive therapeutics on final clinical response was statistically significant. The isolated therapeutics did not reveal improvement with statistical significance when the results were evaluated isolatedly. No predictive factors related with therapeutic responses were determined in this study.

KEY WORDS: chronic idiopathic inflammatory demyelinating polyradiculoneuropathy, clinics, diagnosis, therapeutics.

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