CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY (AUSTIN-DYCK SYNDROME): STUDY OF 45 CASES (Abstract)*. **Thesis. Rio de Janeiro, 1993.**

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The clinical and laboratorial features of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) were analyzed in 45 patients. All of them fulfilled the present clinical criteria proposed for the identification of this polyradiculoneuropathy, particularly those recently presented by the American Academy of Neurology (AAN). None of the cases presented a concurrent disease, other than the one encompassed by the idiopathic modality of CIDP. The clinical, electrophysiological and pathological features of this disease were evaluated. The former included studies in semithin sections, teasing of fibers, histograms and electron microscopy. The sensitivity of the neurophysiological criteria of the AAN was considered in the identification of cases clinically defined as CIDP, in relation to the performed modifications. Histopathological findings of an axonal lesion were related with those of motor conduction of the peroneal nerve. Magnetic resonance images (MRI) of the head were obtained in order to search the possibility of simultaneous demyelinating lesions in the central nervous system (CNS).

Subjects were 26 males and 19 females, ranging in age from 2 to 72 (average of 38 years) at the onset of the symptoms. These symptoms had an average progression of 31 months, ranging from 2 months to 18 years. Twenty-eight cases (62.3%) had a progressive course and 17 (37.7%) a relapsing one. The average time of follow-up was 26 months. Weakness was constant in all cases and followed by sensory impairment, predominantly of touch and vibratory sensations, mainly in the extremities of the lower limbs. Six patients had an asymmetric weakness distribution with a multifocal pattern. Hypo or areflexia was present mainly in the lower limbs. Facial muscles paresis was found in 10 cases (22.2%) and papilloedema in only one. Enlargement of peripheral nerve trunks, tremor and fasciculations of muscles were found in 15, 14 and 2 patients, respectively. Respiratory and autonomic dysfunctions were not found.

Motor neuroconduction tests allowed to verify the demyelinating nature of the neuropathy in all of the cases, being F-wave alterations and the presence of conduction block the most frequent ones, pointing to the involvement of more proximal portions of peripheral nerves. The sensitivity of the electrodiagnostic criteria for establishing demyelination in peripheral nerves of AAN allowed the recognition of only 64.4% of cases clinically diagnosed as CIDP. Changes made in these criteria, although decreasing in specificity, increased the sensitivity in the recognition of the other 35.5% of CIDP cases. Six cases with clinical predominant multifocal motor presentation with conduction block were considered as CIDP variants.

A cerebrospinal fluid (CSF) examination showed increased total protein content in 80% of cases, with normal cell count.

In the histological examination of 44 sural nerve biopsies, the most frequent findings indicative of this neuropathy were active macrophage-associated demyelination found in 75% of nerves and the presence of endoneurial inflammatory infiltration in 40.9%. Reduction of myelin fibers density, particularly of large caliber with loss of bimodal pattern of the majority of histograms was frequent, although inespecific. There was a tendency of increasing the first peak of histograms in the spences of the relative increase of small caliber fibers, possibly due to the presence of fibers inclosed in sproutings and small demyelinated fibers. Remyelinating fibers were identified in 97% of biopsies, onion bulb formations in 54% and axonal lesions in 22%. In such cases which presented axonal lesions in biopsies a correlation with neurophysiological findings was found. Three patients were submitted to two biopsies, being the second made upon their clinical improvement stabilization. In these biopsies it was verified the presence of demyelinated fibers and inflammatory infiltrates, histologically demonstrating the persistence of the pathogenic process in CIDP.

In 83.3% of studied nerves, there was amyelinic fiber loss of mild to moderate intensity, which seemed to be a frequent finding in CIDP. However, no signs or symptoms of vegetative nervous system involvement were found in the patients.

Any of the cases showed signs or symptoms relative to CNS damage. The MRI's of the head, performed in 25 cases, have not shown T2 intense signals in the white matter, as occurs in central demyelination, in any of 17 patients aged under 50. This result shows that the association of CIDP with demyelination of the CNS seems to be merely a coincidence or does not happen in our country.

Treatment response could be evaluated in 39 patients. Clinical improvement was observed in all of the cases being prednisone the therapeutic agent of choice. Three patients were treated with high doses of intravenous immunoglobulin (IV Ig), but only one showed some improvement. However due to the intense headache and vomiting, without cellular reaction in the CSF, this patient was administered half of the original dose IV Ig. Pulsotherapy with metilprednisolone was not completely benefic. It helped 1 in 2 of the patients that received this treatment during the study period. Seven patients benefited from prednisone in association with azathioprine after no response with isolated prednisone (5 cases), pulsotherapy with metilprednisolone (1 case) and IV Ig (1 case). Ciclophosfamide has improved 1 case with multifocal clinical presentation.

The several aspects refered above are discussed particularly in relation to the three great casuistics with clinical, electrophysiological and histopathological studies found in the literature.

KEY WORDS: **chronic inflammatory demyelinating polyradiculoneuropathy, clinics, neuroconduction studies, sural nerve biopsies, CNS involvement, therapeutics.**

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