NMD VI

MDA

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PROGRAM

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☎ 467 0011

Telex: 203748 DARAYAH SJ

61. Remote Effects of Neoplasms-Mechanisms

61.1 SENSORY NEURONOPATHY AND NEURONAL ANTINUCLEAR ANTIBODY. I.Illa,J.L.Rodriguez,C.Juarez.Barcelona.Spain.

Subacute sensory neuronopathy is a clinically well defined paraneoplastic syndrome,usually associated with small cell carcinoma of the lung.Its etiology and pathogenesis is unknown.

Recently Graus et al.(Neurology 1985;35:538-543) described the presence of an antinuclear antibody "restricted" to neurons in two patients with Denny-Brown and oat cell carcinoma of the lung.

We present the immunological studies in a patient with a lung neoplasia and a sensory neuronopathy.

Our patient was a 63 years old man who had a subacute onset of difficulty in deambulation and numbness in the lower extremities.Neurological examination disclosed sensory deficit involving all modalities of sensation in a gloves and stocking distribution,global arreflexia and sensory ataxia.The electrophysiological study revealed absence of sensory action potentials.Motor nerve conduction velocities and amplitudes were normal.No denervation was observed.CSF was normal,except for increased protein content(183mg/dl.).Routine immunological studies were normal.Together with the neuronopathy an oat-cell carcinoma of the lung was diagnosed.The patient died one month later while on chemotherapy for the tumor without any improvement of the neuronopathy.

Indirect immunofluorescence studies with patient's serum disclosed staining of the nucleus of the dorsal root ganglia and cerebral cortex neurons.No staining was observed in the nucleus of the glial cells nor in the nuclei of other non-nervous tissues that were studied.The staining of the neuronal nucleus was homogeneous with exclusion of the nucleolus.The pattern and distribution of the reactivity of this antibody seen in the serum of our patient is similar to that described by Graus el al.(see above).The significance of this antibody in the pathogenesis of the sensory neuronopathy is not known.A suggestive hypothesis is that the neuronal antigen recognized by this antibody is also expressed by the tumor of the patient and the neuronopathy is caused by the antibody which cross-reacts with the tumor and the neuronal antigen.

62. Neuromuscular Disorders In Developing Countries

62.1 THE PATTERN OF CENTRAL NERVOUS DISEASE IN CHILDREN IN KING KHALID UNIVERSITY HOSPITAL IN RIYADH, SAUDI ARABIA. A. Al Frayh, N. Al Naquib. Riyadh, Saudi Arabia, Riyadh, Saudi Arabia.

Over a period of 15 months, 260 children were studied aged 3 months to 15 years in the inpatients and outpatients of King Khalid University Hospital, Riyadh. The objectives were to outline the types of childhood central nervous system disease, the aetiology and sex incidence. Diagnosis was based on clinical examination, laboratory, neuro-physiological and neuroradiological tests. The conclusions were the following:

Convulsive disorder 48.7%. Of these, there were (26.15%) of febrile convulsions, (20.38%) epilepsy and only (1.53%) due to metabolic causes.

The cerebral palsies presented as (20%), the types were the usual with a higher incidence of hypotonia.

Head injuries (11.5%(.

Congenital malformations 3.07%, hydrocephalus included as a separate group in an incidence of 3.84%. Other group of central nervous system disorder (13.84%) included degenerative disorders, microcephaly, tumors, rheumatic chorea, spinal cord injury, etc.

Aetiology was in 27.96% of cases prenatal in orgin, 12.68% perinatal in origin, 32.30% postnatal in origin, 3.88% mixed causes, 20.38% of unknown aetiology. Perinatal factors were highest among the cerebral palsy and convulsion groups.

Sex incidence was generally higher in the male group except in cerebral palsy. A comparison with figures in the literature from other countries is presented and suggestions for prevention.

62.2 SCIATIC NERVE LESIONS IN AKR MICE INOCULATED WITH Y AND CL STRAINS OF Tripanosoma cruzi DURING THE ACUTE PHASE OF INFECTION. A. Antunes-Barreira, J.S. Silva, M.A.Rossi. Ribeirão Preto, Brazil.

Eletroneuromyographic evidence of somatic muscle denervation in chagasic patients has been obtained over the last eight years. Quantitative and electroneuromyographic evaluation of these findings is compatible with the absence of clinical-neurological syndromes imputable to lesions of the lower motor neurons in these patients. This subclinical denervation may be due to neuronal loss at the level of the anterior gray horn of the spinal cord. Anatomical studies thus far published have not been sufficient to demonstrate the level and quality of the possible lesions in chagasic patients and animals inoculated with T. cruzi.

The objective of the present investigation was to study from a clinical-neurological and histopathological point of view the sciatic nerves of inbred AKR mice inoculated with T. cruzi during the acute phase of infection to determine the presence of flacid paresia or paralysis as well as the presence or absence of lesions.

During the observation period- 26 days for mice inoculated with the Y strain, and 32 those inoculated with the CL strain - no paresias or paralyses were detected. Epineural, and endoneural lesions were detected in the sciatic nerves of mice killed at 3-day intervals starting on the 5th day after inoculation. The lesions consisted mainly of the presence of pericapillary macrophage infiltrates (with or without amastigotes in the cytoplasm), as well as mononuclear infiltrates. Exceptionally amastigotes were seen in the cytoplasm of Schwann cells and in the endoneural connective tissue.

No differences in quality or extent of damage were noted between the animals injected with strain Y or strain CL. There were no demyelinating lesions, as is the case for CBA mice injected with the CL strain. The absence of demyelination and of Schwann cells invasion in the present model may contribute to a better understanding of the machanism of damage to Schwann cells and to the myelin sheat in comparative studies using more susceptible mouse strains.

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