

THESES

LANDAU-KLEFFNER SYNDROME: CLINICAL AND ELECTROENCEPHALOGRAPHIC ASPECTS (ABSTRACT)*. Dissertation. São Paulo, 1995.

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Landau & Kleffner, in 1957, described 6 children who presented aphasia, after apparently normal speech acquisition and had epileptic seizures, readily controlled or even spontaneously remitted. All these children had abnormal electroencephalograms (EEG), consisting of bilateral discharges of spike-wave complexes, mainly in the temporal regions, without evident structural brain lesion. At that time, the authors suggested that such abnormalities could cause an "ablation" of primary cortical language area. The purpose of this literature review is to carry out a metanalysis of 180 cases of Landau-Kleffner syndrome (LKS) reported from 1957 to 1995, with emphasis on the clinical and electroencephalographical aspects. It was observed acquired aphasia of gradual onset, occurring in childhood, mainly between the ages of 4 and 6 years, predominating in males. The speech problem was an isolated one, and most of these patients had normal non-verbal abilities. Epileptic seizures of different types occurred in about 80% of the cases, between the age of 4 and 6 years, predominating tonic-clonic-generalized seizures, atypical absences and partial seizures. Behavior disturbances were very common, consisting of hyperactivity, attention deficit and also bizarre behavior resembling psychotic disturbances. The EEG was always abnormal during the aphasia, consisting of generalized or focal discharges, more accentuated in the temporal regions, mainly in the left hemisphere. Less frequently there were associated discharges in the parietal, central, occipital and frontal regions. Electrical Status Epilepticus of Slow Sleep was a common finding in LKS, but it is not specific of this entity, and probably represents an electroencephalographical pattern of some types of epilepsy in childhood. Most cases did not have a precise etiology and it is discussed if the epilepsy is the cause or the consequence of the acquired neurological disease. Neuroradiological abnormal findings were rarely seen. Functional studies (positron and single photon emission computed tomography) revealed irregular activity in the temporal regions, especially in the sylvian area. The treatment with antiepileptic drugs does not seem to change the evolution of the disease. Corticosteroids have been said to be beneficial by some authors. Surgical treatment, by cortical resections and multiple subpial transection have been used recently, with improvement of symptoms. The language evolution was regular, and many patients were socially inserted, even though in jobs that require low academic achievement. Therefore we classified the LKS among the idiopathic epilepsies related to localization, that in this syndrome corresponds to the speech area around the sylvian fissure, that could be damaged at a critical age to the development and maturation of this ability.

KEY WORDS: Landau-Kleffner syndrome, electroencephalography, aphasia, epilepsy.

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