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Short communication

Transitional Syndrome: From West to Lennox-'Gastaut Syndromes"

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Abstract

Both West Syndrome (WS) or Infantile Spasms (IS) and Lennox-Gastaut Syndrome (LGS) have been considered catastrophic epilepsies and in some cases difficult to control, especially the latter.

Keywords

gastaut syndromes; infantile spasms; atypical absences; west syndrome

Introduction

There are theories that consider that they are a continuum of the same pathological process (1):

WS is a type of epilepsy that occurs between 3 and 18 months of age, with a peak between 6 to 9 months, there are spasms, which can be inflexion, extension or mixed, a characteristic EEG called hypsarrhythmia and psychomotor delay.

In contrast, the LGS starts after the first 3 years of age and sometimes until adulthood and it can demonstrate a variety of seizures such as: Tonic, Generalized Tonic-Clonic, Myoclonic, Atonic and Atypical Absences; these seizures are usually difficult to control and cause greater psychomotor retardation. In the EEG, spikes and slow-wave discharges of 1.5 to 2.5 Hz are shown, and during a Sleep EEG test, discharges of fast sharp waves will be seen.

There is multiple etiology for both syndromes including genetic mutations. For this reason, it has been classified as symptomatic if there is an obvious pathology, and it will be cryptogenic in case that the cause is not detected. There are cases where the patient has a normal psychomotor development and if the SW appears, it causes a psychomotor delay with the cause being unknown.

It has been determined that several cases of IS become GLS, however, there is no clear concept of what the reason is. It is suggested that the evolution in this syndrome may be due the etiology, the type of treatment, and even, some authors suggests it may be the same syndrome. Its worldwide incidence is reported to be 1 per 2,000 to 6,000 alive NB. Different reports say that the SW – GLS evolution is between 25% to 54%. SLGs that have a history of IS are up to 36%. (2) There are also some studies that do not report any case or that are minimal.

We've performed an observational study of 130 cases with SW between 2006-2016, confirmed by the presence of the clinical manifestations described above. EEG, brain CT and / or MRI were performed. During its evolution, 14 cases were detected (10.7%) that evolved to GLS, 11 of them were male. All patients were administered Vigabatrin at a dose of 45 to 200 mg / kg / day, associated with valproic acid at a dose of 20 to 50 mg / kg / day for 6 months to a year. Only in 2 cases methylprednisolone was added, because the spasms did not cease. None were treated with ACTH, nor a ketogenic diet. The change in symptomatology from SW to GLS on average occurred at 2.5 years old, with the manifestations of mixed seizures and peak and slow wave discharges of less than 3hz in the EEG. The cause in all the patients was symptomatic, being the antecedent of hypoxic-ischemic encephalopathy in 43%.

All patients underwent ophthalmologic assessment, with visual potentials, and no alterations were detected. The crises in the GLS were predominantly tonic in up to 63% of the cases, atonic in 6%, atypical absences in 5% and generalized tonic-clonic in 5%. In this treatment, several antiepileptic drugs were used such as valproic acid, lamotrigine, topiramate, clobazam, levetiracetam , oxcarbamazepine and prednisone.

We did not find great variation with other reports in its evolution and there was no correlation of the etiological diagnosis, nor the time of the manifestations of SW with the evolution to GLS, it is only to draw attention to the prescription of Vigabatrin. Several series have reported that in GLS, WS was present in around 30% to 60% of the cases (3). So, in our experience there has been a lower percentage. There are no reports in the literature that suggest the possibility of Vigabatrin reducing the risk of the evolution from SW to GLS, however, it has been suggested that the ketogenic diet may contribute to diminish this evolution, even though in any of our cases, no patient received such a diet. In Colombia there is a report in a review of 36 patients with IS, where only 1 case evolved to GLS, also treated with Vigabatrin and valproic acid, however there was no comments on the reason. There is a debate about whether IS and GLS are the same encephalopathy and that difference in the clinical manifestations depend on the level of cerebral maturation.

The mechanism of Vigabatrina is GABAergic, inhibiting the transaminase GABA enzyme, which metabolizes the GABA and increasing its synaptic level. Recent studies of epileptogenesis in IS, describe several processes of neuro-inflammation, alterations in the immune status, and, recruitment of the endocrine system (4), that suggest neuroimmunomodulation as the key to the epileptogenic mechanisms of IS.

There theories of antiepileptic mechanisms, such as the inhibition of inflammation, the modification of mitochondrial metabolism, the action of hormonal therapy and the ketogenic diet, that have been proposed as the processes that prevent the evolution of IS to GLS. Perhaps Vigabatrin should also be added as a preventive mechanism for this change. Although more information is necessary, this hypothesis should be discussed and demonstrated to support avoiding the transition to GLS.

Conflict of Interest

The Author declared there is no conflict of interest

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