Complex febrile seizures associated with Doose vs Dravet syndrome. Presentation of a clinical case

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Abstract

Introduction:

Febrile seizures have an incidence of 4-5 % of the pediatric population from 0 - 6 years. Incidence in males than in females 1.5 : 1. Simple 3 - 15 minutes and complex 15 - 30 minutes. Its physiopathology is unknown, associated factors such as increased circulation of toxins, myelination deficit in immature brain, immaturity of thermoregulation mechanisms, increased oxygen consumption, in any febrile process.

Objective:

To describe the etiology and evolution of the case treated in a second level at a private hospital in Los Cabos, Baja California Sur, Mexico.

Clinical case:

The patient is a Male, son of 30 – year - old healthy mother, 35 – year - old healthy father, second pregnancy, she was born with a weight of 3.2 kg, size 49 cm, respiratory had an Apgar 8 - 9, she was term, obtained by scheduled cesarean section, exclusively breastfed 6 months, complete immunizations and a normal psychomotor development. Parents deny history of febrile seizures or epilepsy.

When she is 7 – months - old, simple febrile seizures began as myoclonic spasms on left arm, 3 minutes last accompanied by loss of consciousness, first 24 hours evolving into complex 3 - 5 per day of 3 - 30 minutes last associated with body temperature 37.9 ° C - 38.3 ° C, her parents start to realize changes in evacuation characteristics. Recurrences of convulsive seizures, with same characteristic, more atonic in intervals of 30 - 60 days the following 5 months associated with febrile process of 38.3 ° C. When 13 – months - old, head movements are added without loss of consciousness, 1 - 3 per day called 'Head Drops'.

Physical exploration:

During my physical exploration I found the following: Weight of 9 Kg, Height of 71 cm, Body temperature from 37.9 ° C to 38.3 ° C, her heart Rate was at that time 124 per minute, Breathing Rate on 36 per minute, Oxygen saturation of 97 % and a Glasgow scale of 15 points.

GENERAL STOOL TEST. Entamoeba hystolitica

STOOL CULTURE. Salmonella enterica

LUMBAR PUNCTURE. Normal. Negative lcr culture.

ELECTROENCEPHALOGRAM. Wave tip image.

MAGNETIC RESONANCE IMAGING. Temporary Mesial Sclerosis

Febrile seizure plus is a term used to denote childhood onset febrile seizures, which (unlike typical febrile seizures) start earlier (less than 6 months with median of 1 year) than the classical febrile seizures. They are often multiple and continue beyond the age of 5 years usually remitting by mid-childhood (median of 11 years).

Individuals with febrile seizures polus may also have additional non febrile seizures. According to Berkovic, the critical feature in diagnosing the" febrile seizures plus phenotype is the continuity of generalized seizures from early to mid-childhood, not the presence or absence of fever. Some other authors have proposed a different name wich is the next one: " autosomal dominant epilepsy with febrile seizure plus maybe more appropriate than GEFS. (generalized epilepsy with febrile seizures plus. In a child presenting with recurrent convulsions precipitated by fever the diagnostic issue is whether they are typical febrile seizures or febrile seizures plus. This is because at onset, the febrile seizure plus phenotype resembles typical febrile seizures occurring between 6 months and 5 years. In most studies, the overall pictures are that generalized epilepsy with febrile seizures pluses usually benign and self-limited. Non-febrile occur in only approximately one-quarter of the patients and these are usually infrequent and often remit by mid-childhood (median 11 years). However, this overall good prognosis is now being reconsidered with the inclusion of Dravet syndrome. The age at the onset, from the first months of life to childhood varies considerable between individuals, in many cases we find individuals of the same family with the same patron. Both sexes are equally affected by this. The prevalence is still unknown now days, but may be high considering the increasing numbers of publications and the very broad spectrum of generalized epilepsy with febrile seizure plus, they show marked genetic and phenotypic heterogeneity. They are extreme intrafamilial and inter-familial clinical variations regarding seizure type seizure frequency, severity and prognosis. By definition, in all families some patients suffer from febrile seizures plus, which are often proceeded by classical febrile seizures. Febrile seizures may occur alone (approximately 75 % of patients) or combine with other type of seizures such as the following: Brief non febrile generalized convulsions. Other generalized seizures, such as a abscesses, myoclonic jerks and tonic seizures. Focal seizures of mainly frontal or temporal lobe origin. More severe forms of seizures such as those occurring in Dravet syndrome and epilepsy with myoclonic-astatic seizures typical febrile seizures and febrile seizures plus are the most common clinical phenotypes occurring in approximately 75 % of affect patients.

Results:

I started her medical treatment the next medication, initially diazepam 0.3 - 0.5mg for kilogram of weight per day, phenytoin 20 mg for kilogram of weight per day, 5 – 7 kg per day, metronidazole 30 mg for kilogram of weight per day, metamizole 10 mg for kilogram of weight per day, paracetamol 20 mg for kilogram of weight per day, and with valproic acid 25 – 40 mg for kilogram of weight per day, levetiracetam 30 mg per kg dose and clobazam 0.1 to

0.3 mg for kilogram of weight per day oral when we have the crisis already controlled. Psychomotor development has been normal. The etiology of the crises in this case is cryptogenic.

Discussion and conclusions:

Children under 12 – months - old with febrile convulsive seizures have a 50 % risk of recurrence with body temperature below 38 ° C at the time of the crisis and recurrence in the same febrile outbreak.

The seizures were refractory to antiepileptic drugs, behaving as a Syndrome of Epilepsy with myoclonic-atonic crisis; as described by Doose in 1970; children are previously normal, with a history of febrile seizures in 11 – 28 %, the evolution of this syndrome is variable, 50 – 89 % stops having crisis after 3 years and as another important fact for many families is that 58 % of this patients live with a normal IQ.



Biography

Dra. Laura Elena Castañeda Ramirez is a pediatrician with over 20 years of experience. She started her career as a general physician and then she starts the specialization in pediatrician. With many years working for the public sector in Mexico, Dra. Laura Castañeda is currently working as the boss of pediatrician in Hospiten Cabos San Lucas, México.

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Notes/Comments: