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Original article

Clinical features of epilepsy with pervasive developmental disorder

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Abstract

Purpose: To clarify the clinical features of patients with epilepsy and pervasive developmental disorder (PDD). *Methods*: We examined 12 outpatients with epilepsy as well as PDD at Seiai Rehabilitation Hospital. *Results*: The patients comprised 7 males and 5 females. The initial neurological symptoms appeared between 5 months and 4 years of age. The interval between the initial neurological symptoms/developmental delay and seizure onset ranged from several months to twenty years. The seizures started at 10–19 years of age in 8 out of the 12 cases. The types of seizures were astatic-drop in 2 cases, tonic-to-astatic in one, atypical absence (decreased consciousness) and generalized tonic clonic seizures (GTCS) in 3 cases, GTCS in 4 cases, or myoclonic and psychomotor in 2 cases. The mental development distributed from normal to extremely severe retardation. Paroxysmal abnormalities on eegs were focal at the frontal area in 7 cases (58%) and other findings in 5 cases. Presumptive risk factors were prenatal in 6 cases (family history for PDD in 1 case, for epilepsy in 1, twin pregnancy in 2 cases, and other in 2 cases), perinatal in 2 patients, postnatal in 1, and unknown in 3 cases. *Conclusions*: The seizures occurred most frequently after the onset of neurological symptoms or developmental delay. The frontal lobe dysfunction was associated with seizure onset in 58% of the cases based on the EEG findings. The risk factors were prenatal in 50% of the cases.

Keywords: Epilepsy; Autism; Pervasive developmental disorder; Electroencephalogram; Risk factor

1. Introduction

Pervasive developmental disorder (PDD) is a serious and nationwide social problem in the fields of education, administration, medicine, welfare, and justice in Japan since the introduction of a new law regarding special aid education in 2002. In this context, some of the classical "epileptic characters" that have been described for a long time might be also be symptoms of PDD and other developmental disorders [1]. Thus, an understanding of developmental disorders is indispensable for discussion on non-paroxysmal symptoms of epilepsy. It is also important to think about PDD from the viewpoint of epilepsy and vice versa.

2. Purpose

The present study reviewed and discussed the clinical features of epilepsy with PDD.

3. Materials and methods

This study analyzed 12 outpatients with epilepsy and PDD. The diagnosis of PDD was based on past and present medical history, physical and psychological examination including the Child Behavior Checklist and other psychological tests, and according to the DSM-IV. The electroencephalogram and MRI results were reviewed by one of the authors (T.K.).

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4. Results

The patient age at the first presentation to our hospital ranged from 3 to 51 years and could be grouped as follows: 3–9 years, 4 cases; teenager, 4 cases, 20s; 2 cases; 40s and 50s, 1 cases of each. There were 7 males and 5 females. The ages at onset of the initial neurological symptoms/developmental delay were 5 months for wry-neck in one case, 6 months for delayed head control in two cases, 2–4 years for language delay in 6 cases, and 2–4 years of indifference to people, hyperactivity, and uncontrolled crying in 1 case of each. The age of seizure onset ranged from 1 to 23 years of age: from 1 to 9 years in 4 cases, teens in 7 cases, and over 20 in one case (Fig. 1). Seizures therefore seemed to first appear predominantly in the teenage years. The interval from initial neurological symptoms/developmental delay to seizure onset ranged from several months to twenty years, and the signs of developmental delay preceded seizure onset in all but one case, in which generalized tonic clonic seizures (GTCS) started at the age of 1 year and 8 months while language delay was noticed at the age of 2 years. The types of seizures were astatic-drop in 2 cases, tonic-to-astatic in 1 case, atypical absence (decreased consciousness) and GTCS in 3 cases, GTCS only in 4 cases, and myoclonic and psychomotor in 2 cases. Seizure frequency until the initial hospital presentation was once in 2 cases, less than once a year in 1 case, several times a year in 5 cases, monthly in 3 cases, weekly in 1 case, and daily in 1 case. Thus, the frequency of seizures among this study cohort ranged from low to high.

Physical examinations at the initial visit showed marked obesity in 2 cases, marked obesity and atopic dermatitis in 1 case, atopic dermatitis in 2 cases, short stature and irritable colon in 1 case each, and no abnormalities in 5 cases. The mental developmental status at initial visit ranged from normal or mild retardation (1 case of each), moderate retardation (6 cases), and severe and extremely severe retardation (2 cases of each). PDD symptomatology in the present study comprised narrow interest, adherence to specific routine habits, short temper, or panic in 83% of the cases, speech delay and short temper in 75% of patients, attention deficit in 67%, and language regression in 33% of the 12 patients (Fig. 2).

The initial EEG findings were normal in 2 cases, borderline in 1, slightly abnormal in 6, moderately abnormal in 2, and severely abnormal in one (Table 1). The EEGs were recorded for more than 30 min in all but one case. Patients were recorded awake and in the natural sleep state mostly, but awake and in a drug-induced sleep in 2 cases, and only while awake in 1 case. The severity of EEG findings was classified by the degree of background abnormalities and by the frequency and distribution of seizure discharges. Paroxysmal abnormality localizations were frontal in 7 cases (58%), multifocal in 2 cases (17%), and hemispheric in 1 case (8%), with one of the cases of multifocal spikes showing a shift to frontal spikes at follow-up. Therefore, 8 out of 12 cases (67%) showed frontal spikes. The MRI revealed diffuse cerebral atrophy in 2 cases, with left parietal white matter lesions, cerebellar atrophy, or hypoplasia of the right cerebellar hemisphere repored in 1 case each. There were no abnormalities on MRI in 5 cases, while the remaining 2 cases were not examined.

The risk factor was prenatal in 6 cases: family histories for epilepsy, for PDD, advanced maternal age, or hypoplasia of right cerebellar hemisphere, with 1 case of each, and twin pregnancies in 2 cases. In the case of advanced maternal age, the age of mother was 38, and the history was a primi para, gestational age of 40 weeks, birth weight of 2.484 kg, and no asphyxia at birth, but MRI showed hypoxic-ischemic change at the follow-up. The perinatal risk factor was attributed to threatened abortion in 2 cases. The identified postnatal cause in one case was arsenic poisoning form milk production, which was

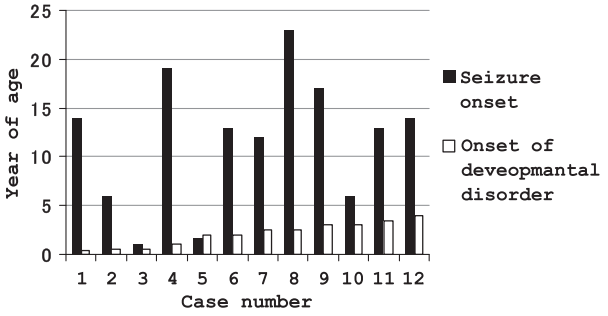

Fig. 1. Age at initial onset of neurological symptoms/developmental delay and seizures.

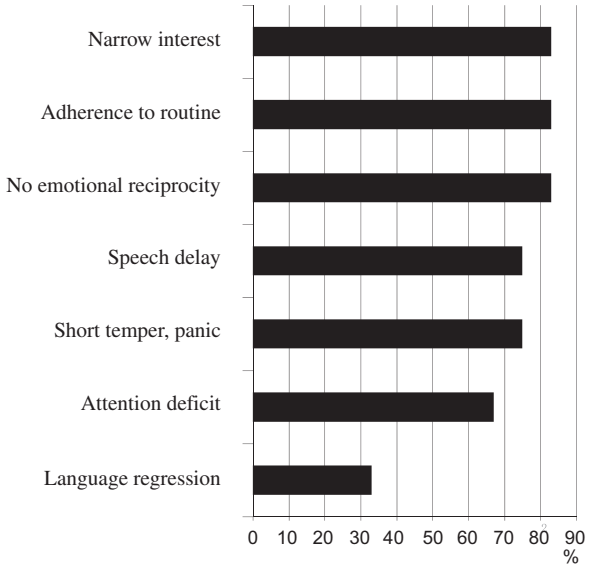

Fig. 2. Frequency of pervasive developmental disorder symptoms in 12 cases of epilepsy.