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Case Report

Successful corpus callosotomy for Doose syndrome

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Abstract

Doose syndrome (epilepsy with myoclonic-atonic seizures) is an epilepsy syndrome with an incidence of approximately 1–2% of childhood-onset epilepsies. Although this syndrome is associated with multiple types of generalized seizures, the diagnosis is based on the presence of myoclonic-atonic seizures. Eighteen percent of patients have refractory seizures and intellectual disabilities. There have, however, been a few reports on the efficacy of surgical treatment for Doose syndrome.

We describe a case of Doose syndrome in a 10-year-old boy. He developed generalized tonic–clonic seizures at 3 years 8 months of age and subsequently developed myoclonic-atonic, myoclonic, and tonic seizures. The frequent myoclonic seizures were refractory to multiple antiepileptic medications. His cognitive development was moderately delayed. Anterior four fifths corpus callosotomy was performed at 8 years 5 months of age. His seizures, especially myoclonic seizures, were markedly reduced. He was given vagus nerve stimulation therapy at 9 years and 1 month of age, which led to complete resolution of the myoclonic seizures. Corpus callosotomy can be a good treatment strategy in patients with Doose syndrome with medically refractory generalized seizures.

Keywords: Doose syndrome; Epilepsy with myoclonic-atonic seizures; Myoclonic seizures; Corpus callosotomy; Vagus nerve stimulation

1. Introduction

Doose syndrome is an epilepsy syndrome that was first reported in 1970 as "centrencephalic myoclonic-astatic petit mal" [1]. This syndrome was otherwise known as myoclonic-astatic epilepsy, and was redefined in 2010 by the International League Against Epilepsy as "epilepsy with myoclonic-atonic seizures" [2]. Doose syndrome is relatively common, with an incidence of approximately 1–2% of childhood-onset epilepsies. Although these patients can develop multiple types of generalized seizures, the diagnosis is based on the presence of myoclonic-atonic seizures. The seizures are generally refractory to antiepileptic medication. The seizure-free ratio in each antiepileptic medication ranges from 0% to 36%. Cognitive impairment affects 20–74% of the patients with Doose syndrome [3]. An electroencephalogram (EEG) demonstrates a 2–5 Hz synchronous spike and wave, and polyspike and wave complexes with abnormal parietal theta activities, although most of the posterior background rhythms and sleep architecture could be considered generally normal for age. This can help to distinguish children

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with Doose syndrome from those with LGS, in whom the EEG is much more abnormal with little or no normal background activity [3]. Brain magnetic resonance imaging (MRI) shows no abnormality.

Antiepileptic medications such as valproate, ethosuximide, and lamotrigine are beneficial for controlling the seizures [4–6]. For the refractory patients, a ketogenic diet has been reported to be efficacious [4,6,7]. Recently, a few reports described vagus nerve stimulation (VNS) therapy, which led to complete resolution of seizures in two patients [8,9]. However, there has been no detailed report of open cranial surgery for the syndrome.

Corpus callosotomy (CC) is a valuable palliative surgical option for cases of medically refractory epilepsy, especially in cases that include drop attacks [10,11]. CC is typically used in patients with diffuse or multifocal epileptic discharges resulting in generalized seizures [11]. There have been only a few reports of CC for children with refractory myoclonic seizures.

Here, we present a case of a child with Doose syndrome, whose myoclonic seizures were refractory to multiple antiepileptic medications. CC markedly decreased seizure frequency.

2. Case report

A 10-year-old boy developed febrile generalized tonic–clonic (GTC) seizures at the age of 3 years 8 months. One week after the febrile seizure, he developed daily myoclonic-atonic seizures. GTC seizures, myoclonic seizures, and tonic seizures also appeared after the onset. He was treated with valproate, lamotrigine, and clobazam, which controlled the myoclonic-atonic seizures. Other antiepileptic medications, including zonisamide, topiramate, carbamazepine, nitrazepam, and levetiracetam, were administered for the other types of seizures, and were partially effective in reducing the frequencies.

The patient was referred to our hospital at the age of 8 years 3 months. Brain MRI showed no suspicious epileptogenic lesions. The patient's Tanaka-Binet intellectual test score was 45. Scalp video EEG monitoring for 24 h showed dominantly bilateral parietal 6–7 Hz theta activity that was not attenuated by eye opening, and diffuse 1.5–2 Hz high-amplitude spike and wave burst interictally (Fig. 1). Bilateral myoclonic seizures with frontal dominant high-amplitude spike or polyspike and waves were captured approximately one

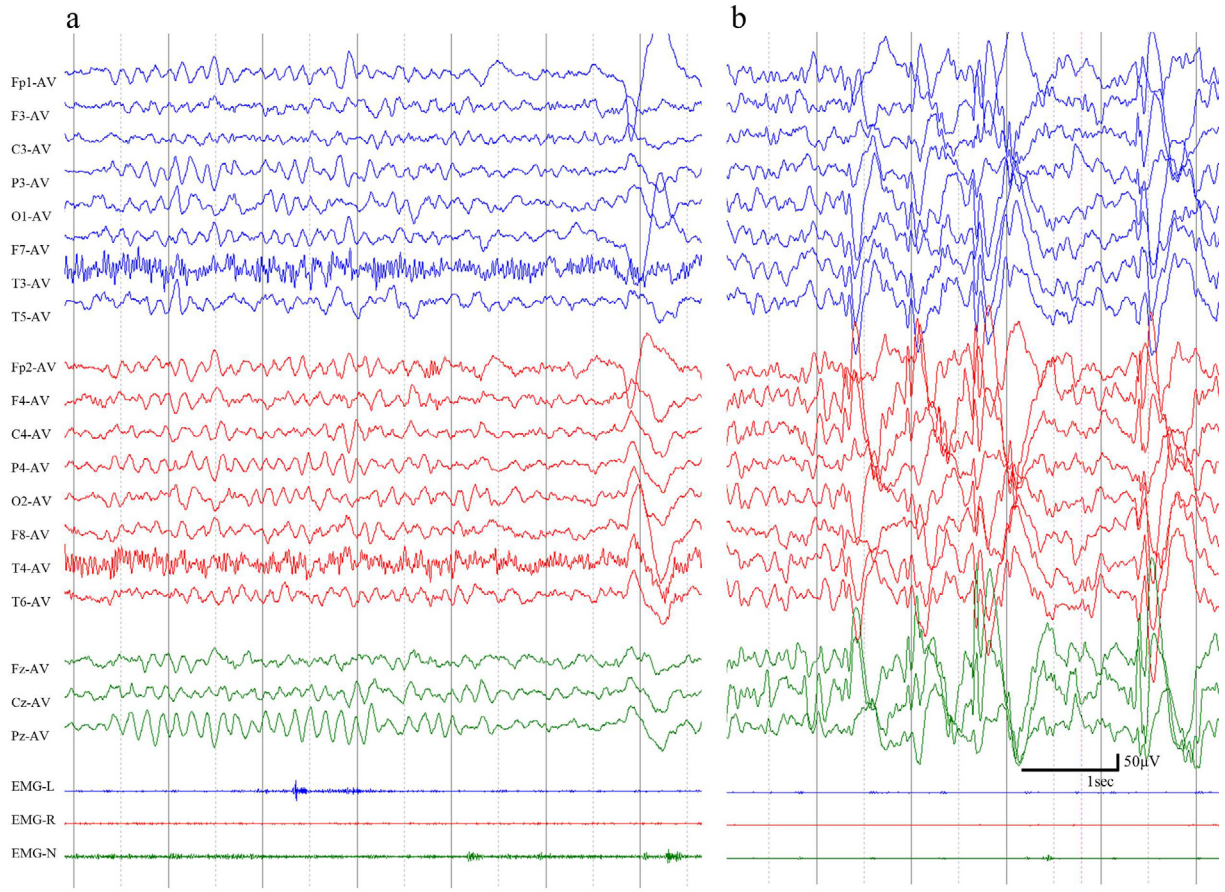

Fig. 1. The interictal EEG showed (a) bilateral parietal dominant theta activity that was not attenuated by eye opening, (b) diffuse 1.5–2 Hz high-amplitude spike and wave burst.