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Case report

Neonatal status epilepticus controlled with levetiracetam at Sturge Weber syndrome

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Abstract

Sturge–Weber syndrome is a rare, sporadic, congenital neurocutaneous syndrome characterized by facial cutaneous vascular malformation, leptomeningeal angioma and eye abnormalities. Seizures develop during the first year of life, may become refractory to multiple anticonvulsants and status epilepticus may develop. A rare subtype of Sturge–Weber syndrome with bilateral facial vascular malformation, unilateral cerebral involvement and neonatal status epilepticus is reported here. Neonatal status epilepticus was successfully controlled with intravenous levetiracetam infusion.

Keywords: Neonatal seizure; Status epilepticus; Levetiracetam; Sturge Weber syndrome

1. Introduction

Sturge Weber syndrome (SWS) is a sporadic neurocutaneous disease characterized by cutaneous, neurologic, or ocular manifestations [1,2]. Facial cutaneous vascular malformations (port wine stain), seizures and glaucoma are among the most common symptoms [2]. It is quite rare with an incidence of 1/50,000 and bilateral facial cutaneous vascular malformation can be seen in only 15% of the patients [3,4]. Cerebral lesions are typically ipsilateral to the facial cutaneous vascular malformation. Radiologically, a leptomeningeal vascular malformation which is commonly located in the parietooccipital region, cerebral atrophy and calcifications may be seen [4].

Seizures are the most common neurologic manifestations and occur either on the contralateral side of the facial cutaneous vascular malformation or are generalized [2,5]. Status epilepticus may also be observed [6].

Many reports on status epilepticus have excluded children under 1 month of age, and those that do include neonates differ in their criteria for diagnosing neonatal status epilepticus (NSE) [7,8]. Clear treatment guidelines for NSE do not exist [9]. Levetiracetam is an effective and well tolerated antiepileptic drug for adjunctive treatment of partial-onset seizure in children. Several pediatrics studies have reported that the safety and efficacy of levetiracetam in neonates [10–12].

A rare subtype of SWS with bilateral vascular malformation and unilateral cerebral involvement, having NSE which was controlled successfully with levetiracetam infusion.

2. Case Report

The 3000 g female infant was born to a mother of 26 years of age, via elective cesarean section at 40 week

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of gestation, following an uncomplicated pregnancy. The patient was admitted to the neonatal intensive care unit because of clonic seizures on left arm at 20 days of age. She had not been diagnosed with neurocutaneous disorder based on her facial lesion that is stated to have been present at birth. Her weight was 3770 g (percentile was bigger than 90th), length was 53 cm (percentile was bigger than 90th), head circumference was 37 cm, (percentile was bigger than 90th), heart rate was 120/min, respiratory rate was 40/min, arterial blood pressure was 61/39 mm Hg and arterial oxygen saturation was 99%. Physical examinations were unremarkable other than for facial cutaneous vascular malformation covering almost all of her face (Fig. 1) and her routine laboratory tests were within normal range.

The infant's left focal clonic seizures were initially treated with phenobarbital with a loading dose of 20 mg/kg and a maintenance dose of 5 mg/kg/day. The seizures were uncontrolled and phenytoin of same dose was then combined. Phenobarbital and phenytoin max dose were 20 mg/kg/day. First phenobarbital plasma level at 7th day of treatment was 2072 mg/L (reference values 15–40 mg/L), second phenobarbital plasma level at 14th day of treatment was 42 mg/L (reference values 15–40 mg/L) and phenytoin plasma level was 9 mg/L (reference values 8–20 mg/L). Her seizures were refractory to this combination despite serum levels of phenobarbital and phenytoin reached to maximum tolerable dosages. She had a focal seizure which generalized and lasted 40 min at her 10th day of hospitalization and NSE was defined with continuous clinical seizure activity lasting longer than 30 min. EEG revealed epileptic activity with isolated sharp spike-wave discharges at parietal right hemisphere and at the frontotemporal areas of left hemisphere (Fig. 2a).

Intravenous levetiracetam infusion of 20 mg/kg loading dose followed by 2 times daily 10 mg/kg/dose was added and NSE was controlled clinically after the onset after intravenous levetiracetam infusion (20 mg/kg/day). A few isolated suspicious convulsive movements were observed in the following hours which were not considered as convulsions by the Pediatric Neurology team. Her seizures were controlled electrographic after phenobarbital, phenytoin and levetiracetam treatments (Fig. 2b).

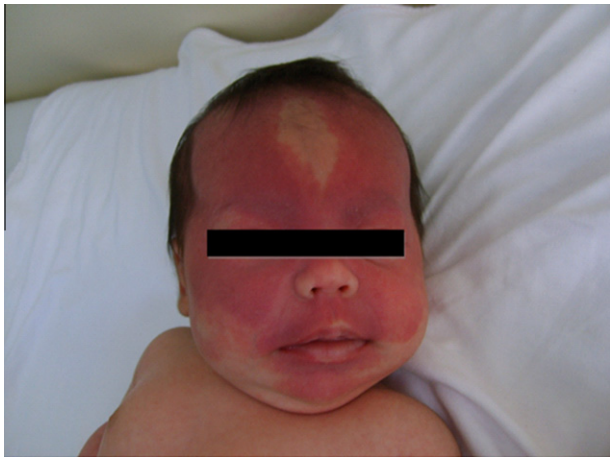

Fig. 1. Extensive facial cutaneous vascular malformation (facial port wine stain).

Cranial ultrasound showed findings correlating with lentikulostriate vasculopathy and cranial CT showed focal hypoplasia in right lateral hemisphere and occipital areas Cranial MRI and MR angiography revealed focal ischemia in the right occipital region, probably due to intrauterine tissue loss. Diffusion-weighted MRI images did not reveal evidence of acute ischemia. Intracranial segments of both vertebral artery, basilar artery and posterior cerebral artery were normal and intracranial aneurysms and stenosis were not detected (Fig. 3).

Glaucoma and other eye abnormalities were not observed and intraocular pressure and corneal diameters were normal. The patient was discharged on the 15th day of admission for outpatient follow up with oral levetiracetam and phenobarbital treatment.

3. Discussion

Sturge Weber syndrome is a sporadic neurocutaneous disease characterized by facial port wine stain, ocular abnormalities (glaucoma and choroidal hemangioma) and leptomeningeal angioma [1].

Facial cutaneous vascular malformation in SWS are well demarcated red macular stains present at birth and turn into the characteristic dark red color similar to the Portuguese liquor. The facial cutaneous vascular malformation typically involves the forehead and upper eyelid, in a distribution that resembles the area innervated by first branch (ophthalmic branch) of the trigeminal nerve. With increasing age the stain darkens in color and becomes raised and thickened [1]. However, unlike ordinary cases, our patient had a vascular malformation in dark red color covering the entire face with sharp boundaries separated from the intact skin.

Seizures occur in 23–83% of the patients with SWS [4,13]. In the majority of patients, they develop before 2 year of age most frequently around 6 months, but neonatal onset is quite rare. Focal tonic clonic motor seizures on the contralateral side of the facial cutaneous vascular malformation predominate but secondary generalization may develop [2,5]. Early onset of seizures may correlate with poorer prognosis and difficult epilepsy control [4]. Refractory seizures may require hemispherectomy or lobectomy [14] .

In an already compromised vascular system, such as a vascular steal from the angioma, seizures are more likely to cause injury, even when short. Episodes of status epilepticus are therefore, especially dangerous in SWS [6].