Case Report

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A case report of a pregnant woman with Sturge-Weber syndrome

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Abstract

A 23-year-old pregnant woman presented with headache and paroxysmal spasm of the right limb. She was born with a port-wine birthmark around the left eye frame. Magnetic resonance imaging revealed evidence of atrophy, calcification and vascular malformation in the left cerebral hemisphere. She was diagnosed with Sturge-Weber syndrome. She gave birth to a child without a port-wine birthmark through a caesarean section and her headache eased without surgical intervention of the intracranial vessel hyperplasia. This case suggests that pregnant women with SWS can deliver safely.

Keywords

Sturge-Weber syndrome, vessel hyperplasia, port wine birthmark, pregnant woman

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Introduction

Sturge-Weber syndrome (SWS) is a rare but well-known disease that affects the development of certain blood vessels and causes abnormalities in the brain, skin and eyes. Its major features are a port-wine birthmark, leptomeningeal vascular malformations and glaucoma.[1] It has been found to be caused by a mutation in the G protein subunit alpha q (*GNAQ*) gene.[2] This gene provides instructions for making a protein to help control the development and function of blood vessels.[3] The mutation occurs in a cell during early development before birth and it is not inherited.[3] There is controversy as to whether the vascular malformation needs treatment.[4,5] This current case report describes a pregnant woman with SWS who gave birth to a child safely without requiring surgical intervention of

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her intracranial vascular malformation, which is a situation that has been rarely reported.[6]

Case Report

A 23-year-old Asian pregnant woman presented at the Department of Neurology, Affiliated Hospital of Southwest Medical University, Luzhou, Sichuan Province, China in June 2018 with a 20-year history of headache and paroxysmal spasm of the right limb, but in the past 1 day it had worsened with severe vomiting. The patient was born with a port-wine birthmark around the left eye frame (Figure 1a). Her symptoms involved episodes of temporary hemiparesis on the right side, seizures and migraine headaches beginning about age 2 without a history of head injury. All attacks began with cold infection. Headache was paroxysmal, lasting several hours and occurred 1–2 times every year. The headache was always on the left side, associated with the right hemiplegia and right limb seizure that lasted for minutes. The patient did

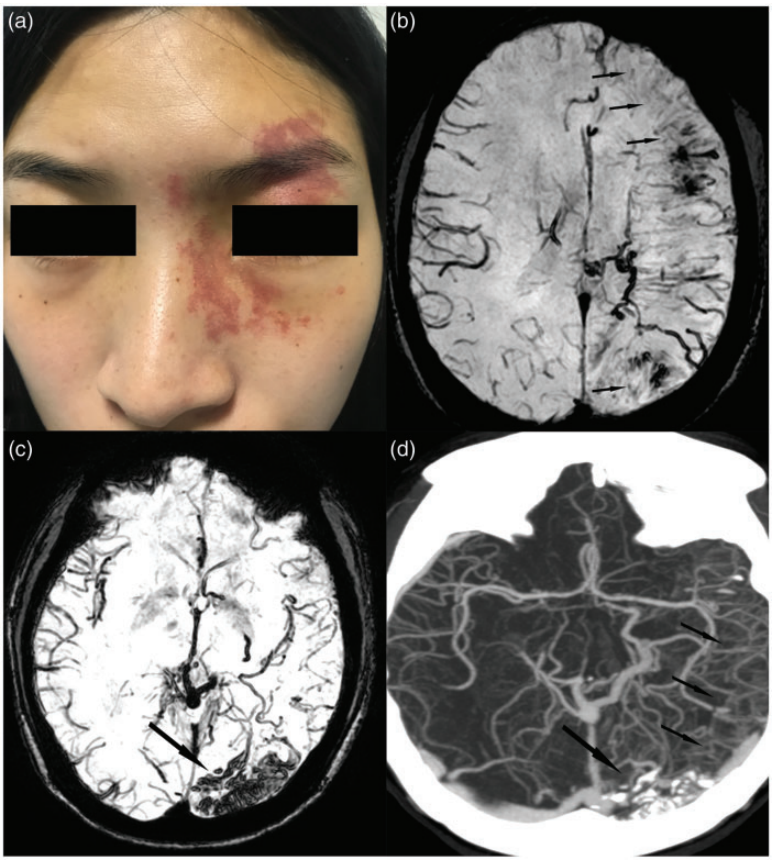

Figure 1. Representative photographs of a 23-year-old pregnant woman with a 20-year history of headache and paroxysmal spasm of the right limb. A port-wine birthmark around the left eye frame (a). Susceptibility weighted magnetic resonance imaging showed the left cerebral hemisphere was atrophied (b, black arrows) and left occipital cortical vascular malformation and calcification (c, black arrow). Computed tomography angiography showed vascular malformation (short arrows) and calcification (long arrow) (d).

not receive surgery to ligature the malformed blood vessels. The patient received 2.4 g benzylpenicillin sodium intravenously every 12 h for 3 days to treat a bacterial upper respiratory infection.

The results of physical and neurological examinations revealed right hemiplegia (4/5). Her cognitive function was normal. The vision and intraocular pressure were normal and the eyeballs did not appear enlarged or bulging. Routine blood biochemistry analyses were as follows: white blood cells, $13.20 \times 10^9 \times$cells/l; neutrophils, 11.73×10^9 cells/l; high-sensitivity C-reactive protein, >5 mg/l. Colour ultrasonography found that she was 17 weeks pregnant with a single live birth in the womb. Transcranial Doppler found normal cerebral blood flow. Dynamic electroencephalography showed an increase of the slow wave in the bilateral frontal lobe and a β-fast wave activity in the left frontal lobe. Magnetic resonance imaging of brain showed hypointensity in the left occipital and temporal region on susceptibility weighted imaging; and it showed evidence of atrophy, calcification and vascular malformation in the left cerebral hemisphere. She was diagnosed with Sturge-Weber syndrome. She refused surgery after careful consideration and was discharged. Her headache relapsed after 21 weeks. She gave birth to a child without a birthmark through a caesarean section at 38 weeks. Computed tomography angiography of the brain showed vascular malformation, atrophy and calcification in the left occipital and temporal region after the caesarean section. She refused surgical intervention of the intracranial vessels hyperplasia and her headache eased.

All procedures involving this patient were undertaken in accordance with the approval of the Ethics Committee of the Affiliated Hospital of Southwest Medical University, Luzhou, Sichuan Province, China (no. IRB-SOP-V3-27) and with the ethical standards of the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Discussion

The manifestations of SWS are the highly complex and diverse. They include seizures, glaucoma, migraine headaches, fluctuating hemiparesis, developmental delay, stroke-like episodes, cognitive impairments and mood fluctuations.[7] Treatment is based on the person's signs and symptoms and may include anticonvulsant medicines, eye drops or surgery to treat glaucoma, laser therapy for port-wine stains, physical therapy and possible brain surgery to prevent seizures.[3] Trabeculotomy *ab externo* that was used to treat glaucoma was safe and lead to good outcomes.[8] Surgical excision of part of the brain tissue achieved good seizure control and improved psychomotor development.[9] However, there are no data indicating whether surgery is needed for intracranial vessel hyperplasia. Some researchers believe that cortical dilated blood vessels are commonly seen in adults with SWS who are fairly neurologically intact and stable and are believed to be primary lesion.[4] Abnormally proliferating vascular malformations may cause cerebral haemorrhage and seizures, so need surgical intervention.[5] For pregnant women, the volume of blood in the third trimester increases dramatically, which increases the risk of cerebrovascular rupture.[10] This current case suggests that the surgical correction of the intracranial vessel hyperplasia may not be necessary in pregnant woman with SWS.

Declaration of conflicting interest

The authors declare that there are no conflicts of interest.

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