Sturge-Weber syndrome with spontaneous intracerebral hemorrhage in childhood

Case report

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A girl aged 2 years 10 months suddenly went into a deep coma and demonstrated left hemiplegia. At birth, she had exhibited a left-sided facial port-wine stain typical of Sturge-Weber syndrome (SWS) and involving the V1 and V2 distributions of the trigeminal nerve. Computed tomography showed a right thalamic hemorrhage with acute hydrocephalus. Magnetic resonance imaging with Gd enhancement 8 months before the hemorrhage had shown a patent superior sagittal sinus (SSS) and deep venous system. Magnetic resonance imaging and MR angiography studies 2 months before the hemorrhage had revealed obstruction of the SSS and right internal cerebral vein (ICV). Given that a digital subtraction angiography study obtained after the hemorrhage did not show the SSS or right ICV, the authors assumed that impaired drainage was present in the deep venous system at that stage. The authors speculated that the patient’s venous drainage pattern underwent compensatory changes because of the occluded SSS and deep venous collectors, shifting outflow through other cortical venous channels to nonoccluded dural sinuses. Sudden congestion (nearly total to total obstruction) of the ICV may have caused the thalamic hemorrhage in this case, which is the first reported instance of pediatric SWS with intracerebral hemorrhage and no other vascular lesion. Findings suggested that the appearance of major venous sinus occlusion in a child with SWS could be a warning sign of hemorrhage.

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KEY WORDS • Sturge-Weber syndrome • intracerebral hemorrhage • sinus occlusion • venous congestion • congenital

Sturge-Weber syndrome (SWS) is a neurocutaneous syndrome characterized by the presence of facial port-wine stains and ipsilateral leptomeningeal angiomatosis affecting early fetal vascular development. It is thought that venous congestion due to cortical vein hypoplasia occurs and causes ischemic conditions related to decreased cerebral blood flow. The majority of patients experience progressive and paroxysmal neurological manifestations, including stroke-like episodes, seizures, hemiparesis, hemianopia, and cognitive delays or regression, which typically begin in infancy.

Spontaneous intracerebral bleeding has never been reported in childhood SWS. Here, we describe a patient with SWS who exhibited bleeding in the right thalamus with intraventricular hematoma penetration. Informed consent to publish this case report was obtained from the child’s parents.

Abbreviations used in this paper: DSA = digital subtraction angiography; ICV = internal cerebral vein; SSS = superior sagittal sinus; SWS = Sturge-Weber syndrome.

Case Report

At birth, a baby girl exhibited a left-sided facial port-wine stain typical of SWS and involving the V1 and V2 distributions of the trigeminal nerve. After the administration of oral phenobarbital (dosages of 90 mg/day), prescribed as an antiepileptic agent by her pediatrician, only two epileptic seizures occurred during the follow-up. In addition, she showed appropriate developmental milestones. At the age of 2 years 10 months, however, she had sudden impairment of consciousness to the level of a deep coma as well as left hemiplegia. Intraventricular hemorrhage and acute hydrocephalus were confirmed on plain CT studies performed at a local hospital, and emergency ventricular drainage was begun immediately (Fig. 1).

Just after transfer to our hospital was scheduled, the patient had rebleeding in the right thalamus. On admission to our hospital, no abnormalities were found in the blood
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She had undergone diagnostic imaging, such as MRI, on a regular basis before the hemorrhage. Magnetic resonance imaging with Gd enhancement obtained 8 months before the hemorrhage had shown an enhanced superior sagittal sinus (SSS) as well as enhanced internal cerebral veins (ICVs), Galenic vein, and deep venous system (Fig. 2). Magnetic resonance imaging and MR angiography 2 months before the hemorrhage had revealed obstruction of the SSS and dilation of the ICV (normal control 35.5 seconds; prothrombin time (PT) 14.0 seconds (normal control 13.0 seconds); and international normalized ratio (INR) 1.08 seconds. No other risk factors for stroke, including cardiac conditions, were present. Ventricular drainage was continued until the intraventricular hematoma cleared out. In addition, early rehabilitation was introduced at bedside, and motor power improvement was achieved within 8 weeks of admission. On Day 27 after the first hemorrhage, neuroendoscopy was performed in response to persisting hydrocephalus and showed an obstruction of the foramen of Monro, which was caused by the hematoma; we performed ventriculoperitoneal shunting after fenestration of the septum pellucidum. The child recovered steadily afterward and visited our clinic for follow-up after discharge. She regained normal motor function without any other deficit. She was observed for 2 years after bleeding. Although we did not register seizures on scalp electroencephalography, at the age of 4 years, the girl’s psychomotor development corresponded to that observed at 1 year and 8–10 months, indicating significantly delayed development. In addition, imaging findings revealed an atrophic tendency in the temporal lobe associated with aggressive behavior, making it likely that subclinical seizures were in the background clinical picture of this patient.

Discussion

Causes of Intracerebral Hemorrhage

Patients with SWS seem to have dilated transmedullary veins because they originally have insufficient emissary veins over the brain’s surface. To our knowledge, all patients with intracranial hemorrhage associated with SWS in previous reports had some type of vascular le-

**Fig. 1.** Axial plain CT obtained at the onset of right thalamic hemorrhage with acute hydrocephalus. Ventricular drainage was performed for intracranial pressure reduction.

**Fig. 2.** Sagittal T1-weighted MR image with Gd enhancement obtained 8 months before the bleeding, showing a patent SSS (white arrows) and ICV (asterisks).
sion, including angioma,7 arteriovenous malformation,1,11 or aneurysm.13 Therefore, spontaneous intracerebral hemorrhage in a patient with SWS is extremely rare. Because patients with SWS have a tendency to exhibit low cerebral blood flow, arterial bleeding is hardly expected without arterial pathology. In our patient, MRI 8 months before the hemorrhage showed patency of the SSS as well as the ICV. However, MRI and MR angiography studies 2 months before the hemorrhage did not show the SSS and deep venous system. We assume that the patient’s venous drainage pattern underwent compensatory changes because of the occluded SSS and deep venous collectors, shifting outflow through other cortical venous channels to nonoccluded dural sinuses. As DSA performed after the hemorrhage did not show a completely patent SSS, ICV, or basal vein of Rosenthal, we can speculate that the venous drainage from deep brain structures might have gone through collateral venous channels to the straight sinus. Sudden congestion (nearly total to total obstruction) of the ICV might have caused the thalamic hemorrhage in this case. Conversely, recanalization of the ICV might have contributed to her dramatic neurological recovery.

Diagnostic imaging, such as MRI and DSA, revealed that SSS and ICV obstruction developed over time in this patient as the result of events that occurred during the observation period. Venous congestion could be considered a cause of hemorrhage in cases of SWS, as cortical venous hypoplasia and dilated transmedullary veins are characteristic findings in SWS. Additional compromise of the outflow was probably related to the first bleeding. Progressive stenosis of the venous sinuses in a neonate has been reported by Curé et al.; apparently, such a process is a possible evolution in SWS. Based on these findings, major venous sinus occlusion may be a warning sign of hemorrhage in patients with SWS.

Treatment Interventions for SWS

Childhood sinus thrombosis is a rare disease, with an annual incidence rate of 0.4–0.7 per 100,000 individuals.6,10 Patients with sinus thrombosis often have local symptoms, such as venous infarction or cortical, subcortical, or basal ganglia hemorrhage.6 Consequently, it has been reported that sinus thrombosis causes impaired deep venous return and increased venous pressure, which may lead to intraventricular hemorrhage originating in the basal ganglia or thalamus.14,15 We have to consider the benefit of antiplatelet or anticoagulation therapy in such patients. However, preventive antiplatelet therapy for sinus thrombosis has not been definitely accepted. Therefore, the administration of an antiplatelet drug 2 months before the hemorrhage in our case was a difficult decision. In a report associated with aspirin and SWS, low doses of aspirin might have been beneficial for stroke-like episodes, such as venous obstruction, in patients with SWS.2,8 However, aspirin is used to maintain cerebral blood flow, not to prevent sinus occlusion. Although there is no definite evidence of the prevention of sinus thrombosis by the administration of any antiplatelet medications, these drugs may be the only way to prevent intracerebral hemorrhage in cases with a major venous sinus occlusion. In acute conditions, such as the sudden deterioration of epilepsy or severe migraine, we must confirm the patency of the major venous sinuses and use heparin at an adequate dosage when sinus occlusion is detected.

In conclusion, we encountered an extremely rare case of SWS with intracranial hemorrhage. The evidence obtained from cerebral angiography showed that increased venous pressure was probably involved in causing the hemorrhage because of impaired intracranial perfusion due to SSS and ICV obstruction. Patients in such cases should be closely monitored given the possibility of rebleeding.
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Disclosure

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