

Unlocking the Mysteries: Stroke-like Episodes in Sturge-Weber Syndrome

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Sturge–Weber syndrome (SWS) is a rare neurocutaneous syndrome characterized by facial port-wine birthmarks and associated eye and meningeal angiomas. Its neurological manifestations include intellectual disability, seizures, and stroke-like episodes (SLEs), often misdiagnosed as stroke. We report a case of SWS presenting with left-sided weakness. Cranial computerized tomography revealed SWS-related venous anomalies, while digital cerebral angiogram confirmed proliferative angiopathy of the distal middle cerebral artery (MCA) with proximal MCA spasm, resolved by intra-arterial verapamil. Despite initial symptom resolution, electroencephalography (EEG) revealed electroclinical seizures. The patient was treated with anti-seizure medications (ASMs) and aspirin, achieving full recovery. SLEs in SWS can result from vascular malformations or postictal phenomena, with ischemia-related events often complicated by vasospasm and altered hemodynamics. Our case highlights the importance of multimodal imaging and video EEG in distinguishing SLE etiologies in SWS, which can guide timely intervention with ASMs and aspirin to reduce recurrence and improve prognosis.

Keywords: Sturge-Weber syndrome; Deficit, focal neurologic; Angioma

Sturge–Weber syndrome (SWS), or encephalotrigeminal angiomas, is a rare neurocutaneous disorder involving vascular malformations of the leptomeninges and facial skin, typically within the ophthalmic and maxillary trigeminal distributions.¹ Its neurological features include epilepsy, migraines, intellectual disabilities, and stroke-like episodes (SLEs), often misdiagnosed as strokes. SLEs arise from complex mechanisms, including seizures, impaired autoregulation, and vascular anomalies. Chronic ischemia contributes to cortical atrophy and cognitive decline.²

This case report discusses a patient with SWS presenting with an acute SLEs due to intracranial vasospasms and subsequent electroclinical seizures, emphasizing timely diagnosis and management with anti-seizure medications and aspirin.

Case Report

A right-handed man, age 35 years, with congenital purple discoloration of the skin on his face, trunk, upper limbs, and

feet presented to the emergency department with an unwitnessed onset of left-sided weakness. He was last seen appearing well 6 hours prior. On examination, his blood pressure was 154/90 mmHg, heart rate was 88–92 beats per minute, oxygen saturation was 99% on room air, random blood sugar was 120 mg/dl, and electrocardiogram showed sinus rhythm. He was drowsy, with a Glasgow Coma Scale of 12. Neurological examination revealed a National Institutes of Health Stroke Scale score of 9 (left facial weakness +2, left arm no movement +4, left leg drift not hitting the bed +1, left-sided sensory loss +2).

Non-contrast computerized tomography (CT) of the head showed right cerebral hemisphere atrophy with gyriform cortical–subcortical calcifications and choroid plexus engorgement, consistent with SWS. A dilated right periventricular venous structure extending to the straight sinus was visualized without evidence of acute hemorrhage or ischemic infarction. A CT angiography (CTA) revealed the

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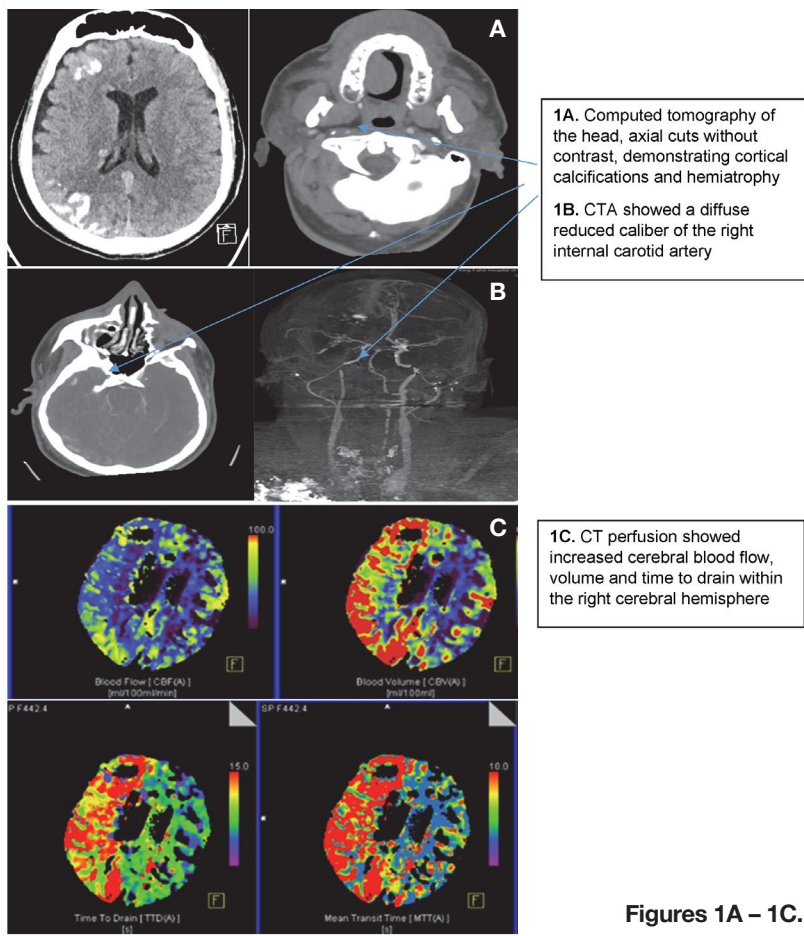
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caliber of the right internal carotid artery (ICA) at the petrous and proximal cavernous segments was reduced, with a small-caliber right middle cerebral artery (MCA) and hypoplastic right A1 segment. A small left MCA was also noted. Perfusing imaging showed increased cerebral blood flow, volume, and delayed venous drainage in the right hemisphere, likely due to angiomatosis (Figures 1A–C).

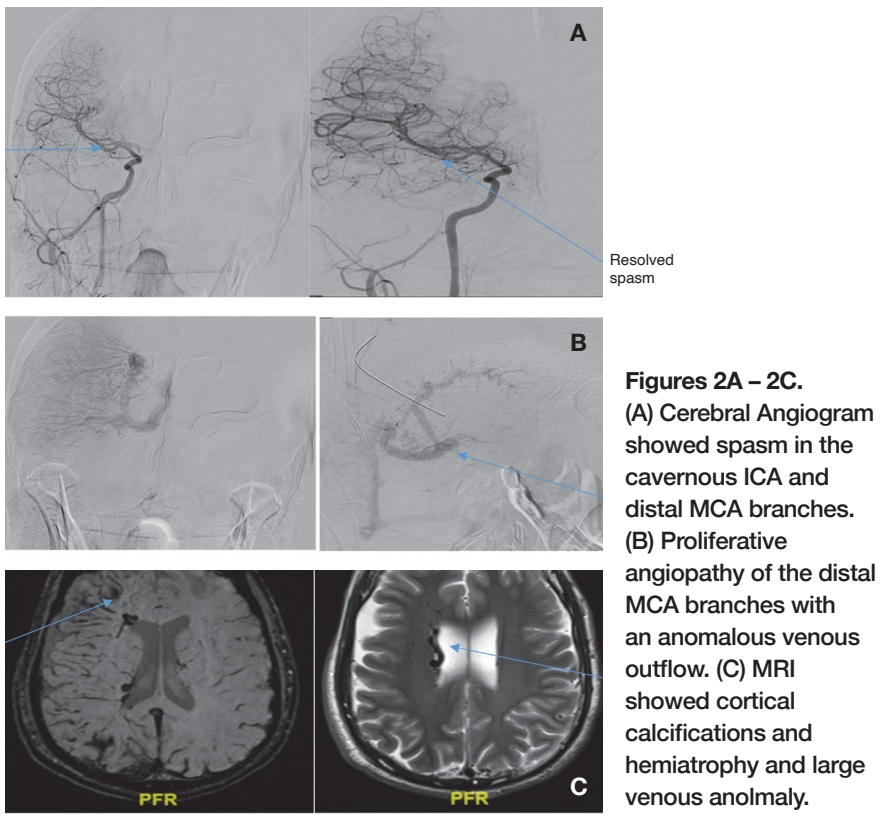
A digital cerebral angiogram identified severe vasospasm in the cavernous ICA and distal MCA branches without vessel occlusion, and there was proliferative angiomatosis of the distal MCA branches with an anomalous venous outflow (Figures 2A and B). The spasm resolved after the intra-arterial verapamil. Magnetic resonance imaging (MRI) of the brain showed cortical calcifications and hemiatrophy and large venous anomaly (Figure 2C). The patient was admitted to the intensive care unit for observation. Continuous electroencephalography (EEG) revealed recurrent focal ictal and interictal epileptiform discharges, primarily from the right temporal and central regions (maximum negativity over T4 and T6), with secondary spread. Associated clinical signs included subtle periodic limb movements, jerks, and lip-smacking, consistent with electro-clinical seizures (Figures 3A-B). The patient was treated with anti-seizure medications (ASMs), aspirin, and vasodilators. He regained full consciousness and was discharged stable with a good functional status (Figure 3C).

Discussion

This case highlights an acute presentation of SWS with ischemic symptoms in the right MCA territory. Patients with SWS often present with SLEs, which are challenging to differentiate from stroke due to their overlapping acute neurological deficits. These episodes can last more than 24 hours and arise from vascular or postictal origins. Initial investigations of our patients revealed severe vasospasms in the cavernous ICA and distal MCA branches, proliferative angiomatosis, and anomalous venous outflow. Intra-arterial verapamil resolved



Figures 1A – 1C.



Figures 2A – 2C. (A) Cerebral Angiogram showed spasm in the cavernous ICA and distal MCA branches. (B) Proliferative angiomatosis of the distal MCA branches with an anomalous venous outflow. (C) MRI showed cortical calcifications and hemiatrophy and large venous anomaly.

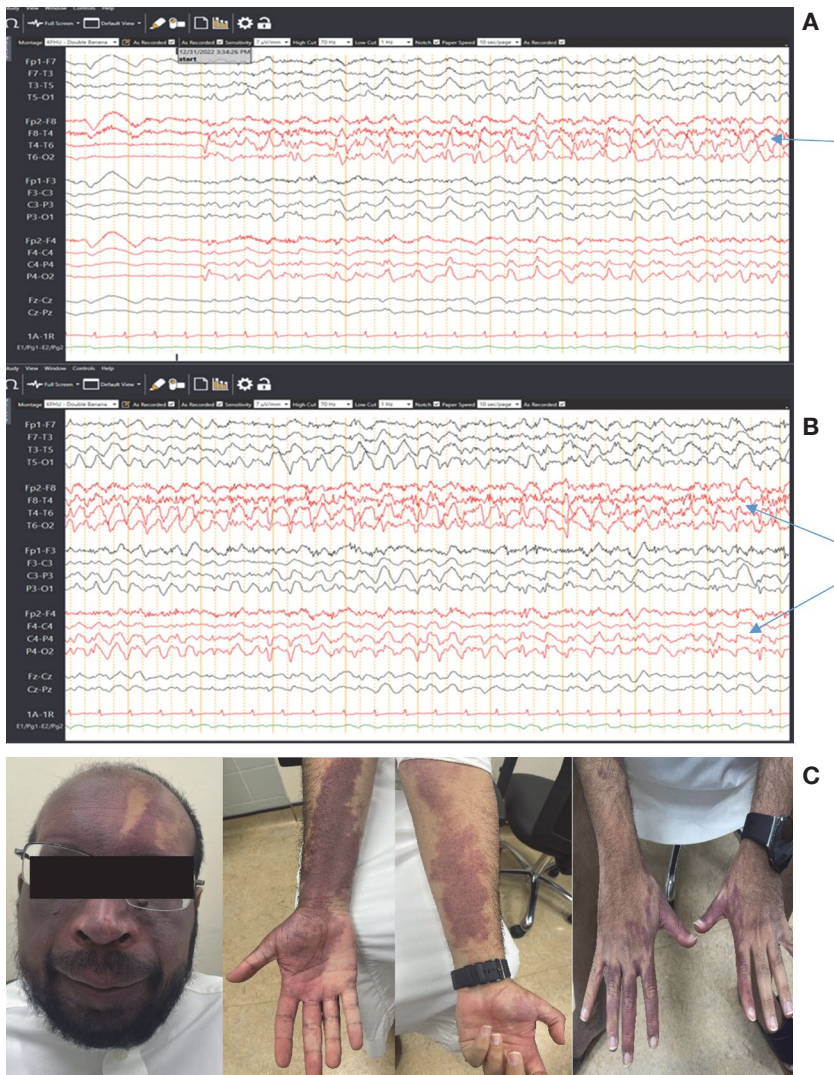


Figure 3. (A, B) Focal spike and slow wave ictal and inter-ictal epileptiform discharges. (C) Facial and limb discoloration due to cutaneous angiomas.

the vasospasms, but the patient subsequently developed electro-clinical seizures, which were managed with ASMs, aspirin, and vasodilators. The sequence of events raises questions about whether seizures triggered ischemia through dysautoregulation or whether ischemia provoked the seizures. Differentiating SLEs from ongoing seizure activity, postictal Todd's paresis, or hemiplegic migraine requires a detailed history, video EEG monitoring, and advanced imaging, such as dedicated MRI sequences.³⁻⁶ Seizures and migraines are frequently associated with venous congestion, supported by findings like focal hyperemia on single-photon emission CT and contrast retention on CTA.⁷

SWS vascular abnormalities more commonly involve venous malformations and hypoplastic cortical vessels. MCA

vasospasm is rarely reported in SWS. Potential mechanisms of MCA vasospasm in these cases include seizure-related sympathetic overactivity,⁸ endothelial dysfunction from chronic venous congestion,⁹ and cortical spreading depolarization during prolonged seizures or migraine aura.¹⁰

Previous literature on acute vascular narrowing in SWS is scarce. Aylett et al.⁸ observed altered cerebral hemodynamics during seizures, including transient arterial constriction, but did not document angiographic vasospasm. Iizuka et al.⁷ reported vasogenic leakage and perfusion changes during prolonged migraine aura in SWS, suggesting dynamic vascular tone changes. However, to our knowledge, there are no prior detailed angiographic images demonstrating MCA vasospasm in SWS with clear pre- and post-treatment comparison, making this case report a potentially novel contribution.

During prolonged seizures, impaired autoregulation of cerebral blood flow may exacerbate ischemia, resulting in neurological deficits and brain injury.⁸ This highlights the need for early and aggressive seizure management to mitigate long-term damage.¹¹ Similarly, ischemia-related events are multifaceted, often involving vascular malformations, hypoplastic cortical vessels, and impaired venous drainage, which may lead to severe cerebral vasospasm, hemorrhage, or infarction. Altered blood flow dynamics can also elevate intracranial pressure, contributing to headaches and hemiplegia.

Perfusion studies suggest impaired venous drainage from the affected regions reduces arterial perfusion, culminating in cortical atrophy, neuronal loss, and astrogliosis.¹² Baseline and SLE-specific imaging comparisons using multimodal techniques, including CTA and MRI, could shed further light on the pathophysiology of these events. Understanding these mechanisms is essential to advancing preventative and therapeutic strategies.

Preventive measures such as daily low-dose aspirin (3–5 mg/kg/day) show promise in reducing the frequency of SLEs and seizures.¹³ Small studies, including that by Udani et al.,⁹ have reported decreased seizure burden in patients with SWS receiving aspirin, suggesting its potential as an affordable and effective therapy. Further research is needed to validate these

findings and explore aspirin's broader role in managing SWS-related vascular and neurological complications.⁹

Conclusion

In conclusion, SLEs in SWS may be vascular or postictal in origin. Ischemia-related episodes are sometimes tricky, as these can be either vascular malformations related, which is consistent of hypo-plastic cortical vessels associated with impaired venous drainage can lead to hemorrhage, infarction, and even severe cerebral vasospasm. This case underscores the importance of tailored, multidisciplinary management of SWS to address its diverse and complex manifestations. Monitoring with video EEG is crucial in distinguishing postictal hemiparesis from paroxysmal vascular events to ensure timely and appropriate treatment, and interventions combining seizure control, vascular spasm management, and stroke prevention are critical to optimizing outcomes. Such strategies may also have significant implications for improving care in resource-limited settings, where access to advanced diagnostics is often constrained. Given the rarity of this complication, collaborative registry-based studies may help clarify its incidence, risk factors, and optimal treatment strategies.

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