

# Intracerebral hemorrhage in Sturge Weber Syndrome: A case report

*By Rizaldy Taslim Pinzon*

## Intracerebral hemorrhage in Sturge Weber Syndrome: A case report

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### Abstract

Encephalotrigeminal angiomatosis also called Sturge Weber Syndrome (SWS) is a neurocutaneous abnormality with angioma affecting the leptomeninges and the face skin, commonly in the Eyes and maxillary distribution of trigeminal nerve. The characteristic of Sturge Weber Syndrome is the dilated the face skin veins, and referred to nevus flammeus and port wine stain. We report a case of Sturge Weber Syndrome in a 41 years old male with uncontrolled hypertension and smoking, left-sided weakness, and left-sided facial port wine stain. Neurological examination was suggestive of SWS and further radiological examination confirmed the diagnosis.

**Keywords:** Intracerebral hemorrhage, encephalotrigeminal angiomatosis, Sturge Weber Syndrome

### INTRODUCTION

Sturge Weber Syndrome (SWS) is a rare neurovascular congenital abnormality which impacts the skin, eyes, and brains. SWS is related to the mutation of gene GNAQ. The incidence of the syndrome between 1/50.000 until 1/230.000 live births [1,9]. Typically, the patient with SWS develops facial capillary malformations, also recognized as port wine birthmarks or nevus flammeus, Leptomeningeal vessel defects commonly present in the trigeminal nerve region. SWS is the third most prevalent a neurocutaneous syndrome subsequent to those of neurofibromatosis and tuberc sclerosis. The neurological manifestations of SWS can involve tonic, myoclonic, or atonic seizures, mental disability, a learning problem, and Attention deficit hyperactivity disorder.

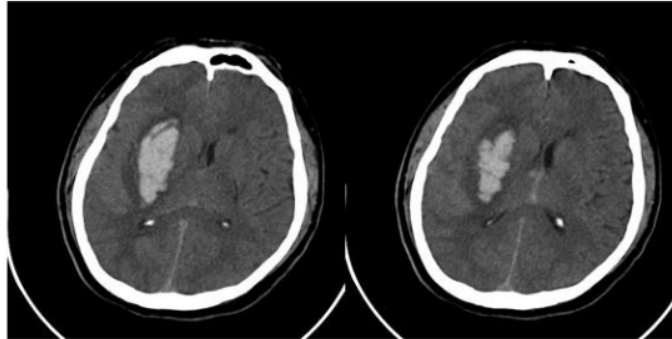
The patient will has impaired blood supply to the brain and is at increased risk of developing venous strokes and stroke like episodes [1,2,7,8]. Capillary malformation and leptomenigeal angiomasitosis can result from a failure in the primitive vein system in initial stages of developmental [10]. Whereas, intracranial hemorrhages in patients suffering from SWS rarely occur. We are reporting a rare case of SWS presenting with intracerebral hemorrhage with concomitant risk factors of uncontrolled hypertension.

### CASE REPORT

A 41 year old male suddenly hemiplegia in the left side was admitted to our emergency department after 4 hours onset. Its neurological status was scored according to GCS 13 (E3V4M6) on the left side weakness. Upon delivery, he developed a trademark SWS port wine stain on the left side of his face affecting the eye, submaxillary, and mandibular division of the trigeminal nerve.

He has no clinical history of any ophthalmic or neurological signs including seizure, glaucoma, and stroke like episodes, aside from visual disturbances in his left eye. He was not treated with anticonvulsants or antiaggregant agents, and had none past heart problem history or any cancerous tumors. He had history of uncontrolled blood pressure and smoking. During admission, his blood pressure was 220/110 mmHg and with pulse rate 72 pulses a minute in a normally rhythmic sinus rhythm, elevated body temperatures was 37.1° C.

His neurologic condition was scored as GCS 13 (E3V4M6) with left side hemiparesis, and focal seizure in the left extremities, and the National Institute of Health Stroke Scale scored is 16. Laboratories findings showed: hemoglobin 13.8 g/dl, the hematocrit 41.2 %, blood urea nitrogen 32.5 mg/dl, and creatinine 1.20 mg/dl. Thrombocytes activation factor and coagulation rate are both normal. Computed tomography showed right thalamic hemorrhage with peri lesion edema. The patient was managed conservatively with tranexamic acid, furosemide, nicardipine, amlodipine, candesartan, and anti-seizure medication. His consciousness disturbance improved slowly to GCS 15 (E4V5M6). Left side hemiparesis It slowly eased, and began to walk with support of the side rail. After 10 days the patients was discharged. In 6 month evaluation, the modified Rankin scale was 2. He continue the anti-hypertension and anti-seizure medication.



**Figure 1. The brain CT showed right thalamic hemorrhage**



**Figure 2. The illustration of port-wine birthmark in our patient**

## **DISCUSSION**

We reported rare cases of intracerebral hemorrhage in SWS. SWS is a manifestation of congenital neurocutaneous syndromes which presents as facial angiomas (also called wine stains) in the higher facial areas and frequent intracranial leptomeningeal angiomas [3].

Most of the patients with SWAS had signs and symptoms: headache, recent focal seizures and hemiparesis. Patients with SWS commonly having temporary episodes of hemiparesis, or stroke like episodes, which may be most puzzling serious neurologic symptoms in patients affected by Sturge Weber syndrome (SWS). Although this episode is similar to an ischemic stroke, the clinical presentation is quite variability,

and MRI of the brain rarely shows persistent infarcts that usually occur in cerebrovascular originating from arteries [4,5].

SWS patients with intracranial hemorrhage were very rare. Angiomatous defects, venous angiomas over plexus of the choroid, and the sinus thrombosis are recognized as prevalent in SWS patients. Though the correlation among SWS and intracranial hemorrhage is unclear in other situations [4,6]. In our patient the uncontrolled blood pressure can be the important trigger and confounding factors.

## **CONCLUSION**

To summarize, we report a very uncommon intracranial hemorrhage cases among SWS patients. Uncontrolled blood pressure may be the important confounding factor. Outcome of SWS patients is suffering from intracranial hemorrhage is not favorable.

## **Ethics**

<sup>6</sup> The authors certify that they have obtained verbal patient consent. The patient identity cannot be identified.

## **Conflict of Interest**

We certify that none of the writers of this work have any conflicts of interest. <sup>1</sup> The conduct, preparation, collecting, analysis, interpretation, and writing of the report were all done without any financial assistance.

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