



## **A case report on Sturge Weber Syndrome Seizures with Stain**

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### **Abstract**

Sturge Weber Syndrome or encephalotrigeminal angiomas is non hereditary, congenital and rare disorder of unknown etiology. It is characterised by vascular malformation with capillary venous angiomas involving face, eye and leptomeninges resulting in neurological and orbital manifestations. A case of 46 years old female presented with history of tonic-clonic convulsions, evidence of Port wine stain on face since birth, characteristic CT findings diagnosed as a case of Sturge Weber Syndrome is reported here for its rarity.

**Keywords:** Sturge Weber Syndrome; Port wine stain; Seizures.

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### **Introduction**

Sturge Weber Syndrome (SWS) also called encephalotrigeminal angiomas is congenital non hereditary sporadic neurocutaneous disease. Shirmer in the year 1860 described this syndrome for the first time and more specific description was given by Sturge in 1879 who described eye and skin changes as well as neurological manifestations. Weber in the year 1929 described radiological changes occurring in these patients<sup>1</sup>. Sturge Weber Syndrome is characterised by facial port wine stain, ocular and neurological manifestations<sup>2,3</sup>. The typical feature of this disease is the angiomas involving the leptomeninges, skin of the face especially the ophthalmic division of the trigeminal nerve.

Neurological abnormalities include epilepsy, focal deficits, learning disorders, mental retardation including developmental delay, hemiplegia as well as orbital involvement resulting in bupthalmos and glaucoma<sup>4</sup>. Dental manifestations may occur in the form of gingival hemangiomas lesion involving mandible, maxilla, lip, cheeks, palate, tongue and floor of the mouth<sup>5</sup>. A rare case of SWS presenting as seizures is described here.

### **Case Report**

A 46 year old female presented with history of generalized tonic-clonic convulsions since 5 days.

She had history of seizures since childhood for which she had taken medication, on and off, from many doctors, quacks and places. The seizures started when she was 8-9 years old. No record was available with her regarding treatment or investigations. There was no history of headache, vomiting, fever and tuberculosis. Her mother denied history of delayed developmental stones and learning problems. Her vitals were normal. On examination, she was conscious and well oriented. Clinical examination revealed erythematous hyperpigmented plaque on the left face involving left side of upper lip with encroaching left side of nose, lips and chin

suggestive of superficial hemangioma.(Fig-1) The lesions were present since birth.

There were no signs of meningeal irritation. Examination of the oral cavity was unremarkable. Laboratory investigations including hemogram, liver and kidney functions, blood sugar, electrolytes HIV test were normal. Xray chest, Xray skull and ultrasound abdomen did not reveal any abnormality. Fundus and CSF examination were normal. EEG revealed focal and sharp waves confined parietal lobe. CT brain revealed hyperdense gyriform calcification in right fronto-parietal lobe (Fig-2).



Fig.1



Fig.2

CT brain depicting prominent subcortical white matter calcification involving left fronto-parietal lobe.

## Discussion

SWS is characterised by port wine stain over the face, ocular abnormalities (glaucoma and choroidal hemangioma) and leptomeningeal angiomas<sup>2</sup>. It is a rare disorder with incidence of 1:50,000 live births. Facial port wine stain is a congenital macular lesions which initially may be light pink in colour, thereby involving area of distribution of trigeminal nerve as documented in this case. Neurological manifestations may vary from minimal or neurological signs to uncontrolled epileptic crisis, hemiparesis, hemiatrophy, mental retardation, microcephaly and visual loss<sup>6</sup>. They commonly involve right side of face especially the ophthalmic distribution of trigeminal nerve and usually do not extend beyond midline<sup>7</sup>.

80% of the patients with seizures show involvement of the contralateral side of Port wine stain and the mechanism of the seizures is cortical irritation caused by the angioma resulting from hypoxia, ischemia and gliosis<sup>7</sup>. Neuroimaging studies are of great help in the diagnosis, assessment of severity and progression of CNS involvement in patients of Sturge Weber syndrome. Xray skull especially lateral view may show tram like calcification which is usually in the parietal and occipital regions. Calcification is rare before 2 years of the age. CT brain reveals contrast enhancement of angioma, abnormal brain ependymal and medullary veins as well as abnormal enlarged choroid plexus ipsilateral to the angioma<sup>8</sup>. Other modalities in the form of functional cerebral studies using PET and SPECT reveal abnormalities of metabolism and perfusion in SWS<sup>9</sup>. The treatment of SWS includes use of anticonvulsants, carbonic anhydrase inhibitors for the control of glaucoma, beta blockers for prevention of optic nerve atrophy and aspirin to prevent vascular disease and headache. As Port wine stain presents cosmetic problems, it is treated with pulsed tunable dye laser. Surgical extirpation of affected lobe or hemispherectomy is indicated in resistant cases with rapidly progressive seizures in order to improve outcome with an aim to prevent developmental delay.

## Conclusion

Early diagnosis and management is recommended in view of varied clinical manifestations of Sturge Weber Syndrome with unknown exact aetiopathogenesis. The combined team efforts of physician, radiologist and psychologist are required for the better management of the SWS.

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