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Case Study

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# RARE CASE OF STURGE WEBER SYNDROME DIAGNOSED ON NCCT HEAD

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**ABSTRACT**

Sturge-Weber syndrome is a syndrome of unknown etiology that is characterized by port wine vascular nevus flammeus in the trigeminal nerve distribution, leptomeningeal venous angiomatosis and they presents with seizures, dementia and hemiplegia. In patients with Sturge-Weber syndrome, X-ray findings in the skull usually show a tram-track pattern of calcification that is caused by calcification in opposing gyri on either side of an intervening dilated sulcus. On CT, curvilinear calcifications in a gyral pattern are often seen, primarily in the occipital and posterior parietal lobes ipsilateral to the facial angioma.

**KEYWORDS:** Portwine stain, gyral calcifications, cortical atrophy.

## INTRODUCTION

Sturge-Weber syndrome (SWS) is a rare neurocutaneous syndrome. It is usually sporadic in nature, which occurs due to occlusion or stasis of the persistent fetal vasculature resulting in reduced blood supply to brain and cerebral anoxia.[1] It is characterised by facial port-wine stain, leptomeningeal angiomatosis, congenital glaucoma, intractable epilepsy and progressive mental retardation.[1,2] These classical brain parenchymal changes can be demonstrated by imaging modalities such as CT and MRI. Since patients usually present with seizures, medical management is by the use of anticonvulsants with or without resection of affected lobes.

### Clinical details

35 years old female presented with multiple episodes of partial seizures since childhood which were refractory to the treatment. There was presence of port-wine stain on the right side of her forehead (Fig 1). For further investigation, NCCT head was done which showed extensive serpentine gyral calcifications in the cortex and subcortical white matter of right parietal lobe with atrophy and thinning of cortex and surrounding significant volume loss.

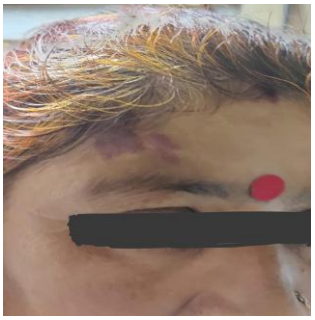
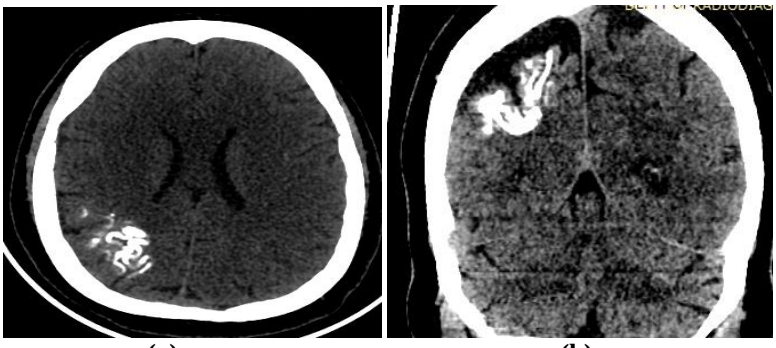


**Figure 1: Shows port-wine stain on the right side of forehead.**



**(a)** **(b)**

**Figure 2: Axial (a) and coronal (b) NCCT head: Shows extensive gyral calcifications involving right parietal lobe with significant volume loss in the affected lobe.**

## DISCUSSION

Sturge-Weber syndrome (SWS) is a rare congenital neurocutaneous syndrome. It was first described in 1879 by Sturge in a patient with facial angioma, glaucoma and seizures.[3] The current definition of SWS includes a cutaneous angioma of the face reaching territory V1, neurological malformations (most often, ipsilateral leptomeningeal angioma, which can be responsible for convulsions, mental retardation or neurological deficit) and ophthalmological anomalies (angioma choroid, congenital glaucoma) of inconsistent presence.[4,5] Our patient showed cutaneous angioma with presence of cerebral involvement in the form of gyriform hemisoherical calcifications. On cerebral computed tomography, the cerebral atrophy associated with gyriform hemispherical calcifications were typical enough to confirm the diagnosis despite the impossibility of visualizing the pial angioma through these calcifications. Choroidal angioma, on the other hand, is well visualized by cerebral CT with injection of contrast product.

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