

# Sturge-Weber Syndrome – A Case Report

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## Abstract

Encephalo-trigeminal angiomas otherwise known as Sturge Weber syndrome is characterised by port-wine stain. Here we report a case with typical characteristics of the same and description about the patient's clinical presentations

**Key words:** Encephalotrigeminal angiomas, Sturge Weber syndrome, Port wine stain, Neurocutaneous disorder

## INTRODUCTION

Encephalotrigeminal angiomas commonly named as Sturge-Weber syndrome (SWS) is an uncommon neurocutaneous syndrome. It is characterized by unilateral facial nevus flammeus (or port-wine stain) with associated ipsilateral leptomeningeal angiomas. In the year 1860, first described by Schirmer and but more specifically in 1879 by Sturge. It is also known as Sturge-Weber disease, leptomeningofacial angiomas, and Sturge-Weber-Dimitri syndrome.<sup>[1,2]</sup> Though rare, this is the most frequent disease among the neurocutaneous syndromes.<sup>[2]</sup> It often follows the outline distribution of the trigeminal nerve.<sup>[3]</sup> Occurs in equal frequency in both the sexes. Intraorally, angiomas can involve lips causing macrocheilia, leading to hemihypertrophy of the buccal mucosa, palate, and the floor of the mouth. Gingival involvement varies from light vascular hyperplasia to severe overgrowth making it difficult for mouth closure or almost impossible.<sup>[4]</sup>

In 1992, Roach categorized SWS variants into three types:

- Type I: Individual has a facial port-wine stain, leptomeningeal angioma and may have glaucoma
- Type II: Individual has a facial port-wine stain, no leptomeningeal angioma and may have glaucoma
- Type III: Individual has leptomeningeal angiomas, no facial port-wine stain, and, rarely, glaucoma.<sup>[5]</sup>

Our aim with this case report is to present a case with classical signs of the disease being survived with this condition for 45 years.

## CASE REPORT

A 45-year-old female reported to the department with complaints of swollen and bleeding gums and loose tooth on the upper left side back tooth region. Her medical history revealed getting seizures on and off most frequently but was always manageable due to proper medication. She was on long-term multiple antiepileptic drugs (sodium valproate, clonazepam, and topiramate) for intractable seizures. Hematological and biochemical profile was within the normal range. The patient was apprehensive. She has no knowledge about the birth type or any uneventful birth events. No family histories relevant to her condition were elicited.

On extraoral examination, the patient had a unilateral port-wine stain centered around the right forehead, the right eye, over the nose, and the right side of upper lip. A reddish lesion similar to hemangioma was observed over the right side of the face. The entire lesion was centered toward one side without crossing midline.

Intraoral examination of the patient showed few missing teeth which patient reveals of being exfoliated on its and not being extracted by any dentist. No other obvious hard tissue abnormalities. Oral hygiene of the patient was poor with extensive calculus.

Gingiva in the upper right side region appeared inflamed, reddened with a tendency toward bleeding on probing.

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Figure 1: Extra oral view



Figure 2: Intra oral view

However, there was an absence of blanching on the application of pressure. The tongue also appeared stained. The palate showed marked port-wine stain over the entire right side without crossing midline. The buccal mucosa, floor of the mouth, and rest of the gingival appeared normal.

The ocular examination revealed the presence of suprascleral hemangiomas, indicative of glaucoma but the patient has not revealed any ocular checkup history for the same.

Radiographs did not show any relevant findings.

Based on the clinical features and vitropression execution, provisional diagnosis of SWS was made and the patient was advised to go for the ophthalmic examination and further medical evaluation for medical counseling in these specialties as a preventive measure.

## DISCUSSION

In our case, even though the patient has port-wine stain from birth onward, it remained asymptomatic till date. However, it is essential to be alert on having future complications as well. The lack of awareness about the disease among the dentist may lead to serious complication. This syndrome is associated with a port-wine stain in the face (naevus flammeus), ocular involvement leptomenigeal angiomas, ipsilateral gyriform calcification, convulsive crisis, hemiparesis, and hemiplegia.<sup>[3,6-8]</sup> Sturge-Weber oral manifestations occur in 38% of the patients, who may have hemangiomas lesion in lips, mucosa, gingiva, tongue, and palate.<sup>[9]</sup>

In this case report, the patient was identified with lesions resembling hemangioma, on the right side of the face extending from forehead covering cheeks, upper lip and clearly demarcated till the midline with intraoral findings involving gingiva and tongue.

## CONCLUSION

SWS is a neurocutaneous disorder. It includes the following triad port-wine stain involving the trigeminal nerve, ipsilateral leptomenigeal angiomatosis, and ipsilateral vascular malformation of the choroidal vasculature of the eye (Florine, 2011).

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