# AN UNUSUAL CASE OF STURGE-WEBER SYNDROME

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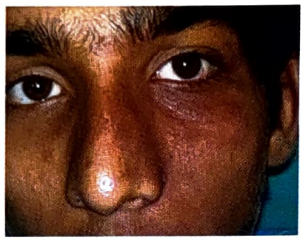
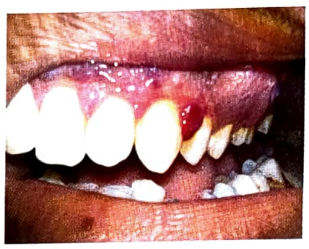
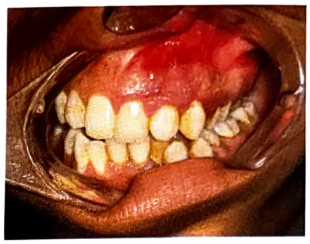
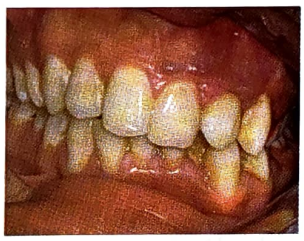
**Abstract**
Sturge-Weber Syndrome, also known as Encephalotrigeminal Angiomatosis, is a neurocutaneous disorder with angiomas involving the leptomeninges and skin of the face, typically in the ophthalmic (V1) and maxillary (V2) distributions of the trigeminal nerve. The cutaneous angioma is called a Port-wine Stain (PWS). This is a report of a 17 year old patient who came with a complaint of bleeding gums in left upper back region of the jaw since past 4 to 5 years. Thorough case history, clinical, radiographic, ophthalmic and neurological examinations point towards the diagnosis of Sturge-Weber Syndrome Type II. With this case we have tried to look for the implications that such findings may have on the treatment of gingival and periodontal diseases.

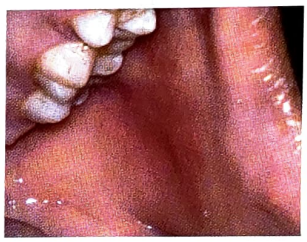
**Keywords:-** Sturge-Weber Syndrome, Port-wine Stain, angiomatosis

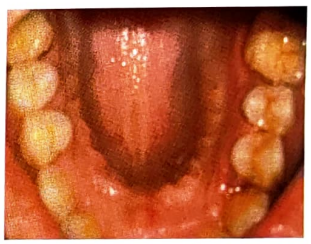

Extra Oral View

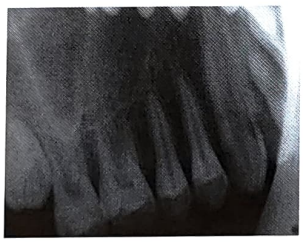

Intra Oral View-Pre operative

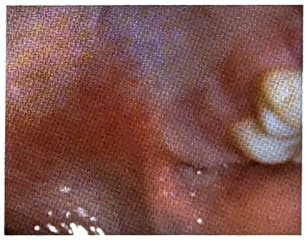

Intra Oral View - Affected side after scaling


Intra Oral View- Unaffected side


Intra Oral View - Buccal Mucosa


Palatal View- note expansion of bone on affected side


Intra Oral Radiographic View- Affected side


Intra Oral View of soft palate

## Introduction

The Naevus Flammeus (Port Wine Stain) is usually present at birth or develops on the face in early infancy in histologically normal skin. The lesion is flat and is not associated with any other underlying malformations and shows no tendency to regress.[1] Telangieclascia or dilatations of capillaries are often seen in irradiated skin, in elderly patients, following prolonged steroid therapy or in patients suffering from liver failure. The cutaneous angioma is called a Port-Wine Stain (PWS). PWS in the distribution of Trigeminal Nerve may be associated with the Sturge-Weber Syndrome (Encephalotrigeminal Angiomatosis). It is an extremely uncommon congenital disorder attributed to faulty development of certain mesodermal and ectodermal elements.[2] It is often associated Sturge-Weber Syndrome, also known as encephalotrigeminal angiomatosis, is a neurocutaneous disorder with angiomas involving the leptomeninges overt the cortex and by ipsilateral PWS on the skin of the face,[3] typically in the ophthalmic (V1) and maxillary (V2) distributions of the trigeminal nerve. The cutaneous angioma is called a Port-wine Stain (PWS).

SWS is referred to as complete when both CNS and facial angiomas are present and incomplete when only one area is affected without the other. According to Roach Scale,[4] it may be classified as:

**Type I** - Both facial and leptomeningeal angiomas; may have glaucoma

**Type II** - Facial angioma alone (no CNS involvement); may have glaucoma

**Type III** - Isolated leptomeningeal angioma; usually no glaucoma

This case report is a Type II SWS which presents with congenital macular lesions that can be progressive; they may be a light pink color initially and then progress to a dark red or purple nodular lesion. These lesions may be isolated to the skin in case of Type II or may be associated with lesions in the choroidal vessels of the eye or the leptomeningeal vessels of the brain. Patients with Type I or Type III may present with seizures, headaches, ocular manifestations, glaucoma and blindness.

## Case Report

A 17 year old patient reported to the Dept. of Oral Medicine, SGT Dental College with a complaint of bleeding gums in left upper back region of the jaw since past 4 to 5 years. The patient was then referred to the Dept. of Periodontics for the needful. Henceforth the patient was examined and a thorough history recorded.

## Clinical Features

Extra-orally, the patient presented with a clearly demarcated Port-wine Stain (facial capillary hemangioma) on the left side of the face which has been present since birth as revealed by the accomplices of the patient. This stain extended from the bridge of the nose, spanning the lateral surface upto the lower eyelid and the outer canthus of the eye, and inferiorly to the angle of the mouth. On the left side a part of the upper lip was also seen to be involved.

Intra-oral examination revealed reddish pink gingiva only in the 2nd quadrant in an otherwise healthy mouth. The size was seen to be enlarged due to inflammation involving the marginal and papillary gingiva and bleeding even on slight probing was present in the same quadrant. The rest of the quadrants showed signs of mild inflammation. Intra-orally the Port-wine Stain could be seen extending anteriorly from the attached gingiva of the maxillary 2nd molar to the distal of the canine and inferiorly to the angle of the mouth indicative of cutaneous angiomatosis. The buccal and labial mucosa of the same area was also seen to be involved, with the labial stain seen to coalesce with the extra-oral PWS.

## Investigations

Radiographic investigations included an OPG and an occlusal view. Both did not reveal any significant changes in the periodontium. Further radiographic evaluation was ruled out in view of the radiation exposure and lack of any other apparent clinical signs and symptoms. The patient was also referred to an ophthalmologist and investigated for any ocular disturbances but no abnormalities were detected.

## Treatment

Symptomatic treatment was rendered to the patient which included scaling and root planing. A chlorhexidine mouthrinse was also prescribed.

## Discussion

Bioxeda *et al*[5] found facial PWS distributed predominantly over the distribution of V2 branch of the trigeminal nerve in 88% of a total of 121 patients with PWS affecting skin. The lesions were unilateral in 86% of the patients. Extra-facial PWS was more common when the V3 division was involved. Their conclusion was that only those with V1 involvement are at risk of epilepsy and glaucoma.

Tallman *et al*[6] conducted the largest study on 310 PWS patients. 85% were found to have unilateral involvement. Only 8% of the patients with trigeminal involvement had CNS or eye involvement. 9% of the patients had lower eyelid involvement alone. The authors recommended screening for glaucoma and CNS involvement when PWS involved the eyelids.

In the present case, a careful history to eliminate ocular manifestations (glaucoma, blindness) and CNS involvement (headaches, seizures) was recorded and consultations with a neurologist and an ophthalmologist were done to rule out the same.

The facts that PWS was unilateral, present since birth, spanning V2 division of the trigeminal nerve, not having any ocular/CNS manifestations but leading to intra-oral clinical finding of increased bleeding on probing and gingivitis in the affected quadrant (left maxillary) points to a diagnosis of Sturge-Weber Syndrome Type II in the reported case.

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