

Sturge–Weber Syndrome: A Case Report and Review of Literature

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Abstract

Sturge–Weber syndrome (SWS) is a rare congenital developmental disorder manifesting with a facial port-wine birthmark, and a vascular malformation of the brain. It affects the skin in the distribution of the ophthalmic branch of the trigeminal nerve, abnormal capillary venous vessels in the leptomeninges of the brain and choroid, glaucoma, seizures, stroke, and intellectual disability. We reported a case of a 37-year-old male who had an unusual overgrowth of the left maxillary region, associated soft tissue hypertrophy directly corresponding to the distribution of the cutaneous port-wine stain, choroidal hemangioma, and hemianopsia of the left eye, hemiparesis of the right side of the body. Radiographic evaluation revealed tram-line like intracranial calcification, suggestive of SWS.

Key words: Choroidal hemangioma, Hemianopsia, Hemiparesis, Port-wine stains, Sturge–Weber syndrome, Tram-line like intracranial calcification

INTRODUCTION

Sturge–Weber syndrome (SWS) or encephalotrigeminal angiomatosis is a congenital, non-hereditary, condition of unknown etiology. The disease shows facial port-wine stain, ocular abnormalities (glaucoma and choroidal hemangioma), and leptomeningeal angioma.¹ It belongs to a group of the disorder known as the phakomatoses (“mother-spot” diseases). SWS was first described by Schirmer in 1860 and later more specifically by Sturge in 1879. He associated dermatological and ophthalmic changes of the disease to neurologic symptoms. Weber, in 1929, stated the radiologic alterations seen in patients of SWS.²

It is rare disorder occurring with no racial predilection³ equally affecting males and females with 1:50,000 live births.⁴ The classic features of SWS are angioma of

the leptomeninges epilepsy, Port-wine stain, ocular involvement, dermal angiomas, mental retardation, hemiplegia, and abnormalities in skull radiographs.⁵ Oral manifestations of the disease varies considerably. The morphological and histological changes in gingiva, pulp, and periodontium have been reported. However, the most common feature is a gingival hem-angiomatous lesion usually restricted to ipsilateral maxilla, mandible, lips, tongue, cheeks, palate, and floor of mouth.¹

SWS are caused by the persistence of vascular plexus around the cephalic portion of the neural tube. The development of plexus starts during the sixth week of intra-uterine life and usually undergoes regression during the 9th week.⁵ Angiomas of the leptomeninges are located in the parietal and occipital region and are usually unilateral. The presence of angioma alters the vascular dynamics causing precipitation of calcium deposits in the cerebral cortex. Seizures, mental retardation, hemiparesis, or hemiplegia develops secondary to this, and the severity depends on the extent of lesion.⁶

The cutaneous angiomas are called port-wine stains, which usually occur unilaterally along dermatomes supplied by the ophthalmic and maxillary division of trigeminal nerve. It may

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be bilateral or absent or may extend to the neck, limbs, and other parts of the body.⁶ Involvement of the area supplied by ophthalmic division is pathognomonic of the disease. Ocular involvement results in glaucoma, choroidal hemangioma, buphthalmos, or hemianopsia (decreased vision).⁷

Intraorally angiomatosis may involve lips, gingival, buccal mucosa, palate, and floor of the mouth. The oral manifestations of SWS are port-wine stain lesion of oral mucosa and hyper vascular changes. Most common manifestation is the angiomatous lesion of gingiva that varies from slight vascular hyperplasia to massive hem-angiomatous proliferation. There is an increase in the vascular component and gingival hemorrhage at minimal trauma. The oral manifestations are unilateral and finish abruptly in the midline. Macroglossia and maxillary bone hypertrophy are seen in some patients leading to malocclusion and facial asymmetry. This syndrome is of rare occurrence and management becomes complicated due to risk of hemorrhage.¹

In this report, we present a case of SWS with its characteristic manifestations.

CASE REPORT

A 37-year-old male patient reported to the Department of Oral Medicine & Radiology, Swargiya Dadasaheb Kalmegh Smruti Dental College and Hospital, Nagpur, Maharashtra, with a chief complaint of bleeding gums and pain in teeth and gums of upper and lower jaw. His past medical history revealed seizures, since 1 year of age, and lasted until the age of 15 years. He also gave the history of hemiparesis of the right side of the body at the age of 1 year and reddish discoloration on the left half of the face and neck since birth that became darker with the advancing age.

Patient's extra oral examination revealed facial asymmetry, overgrowth of the left maxillary regions, associated soft tissue hypertrophy directly corresponding to the distribution of the cutaneous port-wine stain involving V1 and V2 Distribution, and deviation of the nose toward the right side (Figure 1a and b). Pinkish red staining or so-called port-wine stains of left facial skin extending from the midline, involving forehead, nose, left half of upper lip and cheek (Figure 1a-c). The lower jaw, lower lip, and left ear were not involved. Similar type of stains seen on the left half of the neck, extending to left shoulder and left arm. Examination of the eye revealed hemianopsia and dilated blood vessels of the left eye. The right eye appeared to be healthy and patient was referred to the ophthalmologist for consultation. An angiomatous enlargement of the left upper lip was present.

Intraoral examination revealed erythematous, swollen attached gingiva involving the left maxillary arch soft in consistency, and restricted to the midline (Figure 2a and b). The Diascopy test was positive, suggestive of the angiomatous enlargement. Similar growth saw involving the left side of the palate extending to the midline (Figure 2c), areas of decapitation seen on the tongue (Figure 2d). Patient's oral hygiene was poor and showed chronic generalized periodontitis and generalized spacing resulting in malocclusion (Figure 2a and b).

Radiographic examination shows tram-line like calcification in the parietal lobe, on lateral skull projection (Figures 3 and 4) and posterior anterior skull radiograph (Figure 5). Based on the history, clinical findings and radiographic evaluation a diagnosis of SWS was made.



Figure 1: Port-wine stains in the distribution of trigeminal nerve



Figure 2: Intraoral photographs



Figure 3: Orthopantomogram



Figure 4: True lateral skull radiograph intracranial tram track calcification



Figure 5: Posterior anterior skull view osteohypertrophy of left maxillary bone

Blood investigations were found to be normal. Physician's consent taken before the extraction of Grade III mobile permanent mandibular right second molar and plaque control regimen started at regular intervals, proper instructions regarding maintaining oral health care, and use of chlorhexidine mouthwash were advised. Patient's evaluation after 1 month shows no evidence of gingival bleeding. Moreover, gingivectomy was planned for gingival enlargement. The patient is on follow-up. For the purpose of social acceptance, patients counseling was done to undergo treatment such as maxillectomy and laser treatment for facial stains.

DISCUSSION

SWS referred to as complete when both central nervous system and facial angiomas are present and incomplete when only one area is affected without the other. The Roach scale⁸ for SWS is as follows:

Type I: Both facial and leptomeningeal angiomas; may have glaucoma

Type II: Facial angiomas alone; may have glaucoma

Type III: Isolated leptomeningeal angiomas; usually no glaucoma.

According to the Roach scale, our case is a Type I SWS. Leptomeningeal angiomas are unilateral lesions affecting the pia-arachnoid membrane over the posterior temporal, parietal, and occipital areas. It is typically a static lesion, but a review of literature also reveals some progressive lesions.² It commonly shows abnormal blood flow pattern as venous occlusion, thrombosis, vasomotor phenomenon, and vascular steal phenomenon resulting in cortical ischemia. This results in an epileptic convulsive crisis, transient hemiparesis, gliosis, and progressive deposition of calcium salts. These calcifications produce a characteristic double contoured "tram-line" appearance following the convolutions of the cerebral cortex. Brush field and Wyatt stated that these tram-line calcifications are pathognomonic of SWS.⁹ These calcifications appear after the age of 2 years and remain stationary after the second decade of life. These calcifications are pyriform and curvilinear and most commonly seen in parietal and occipital lobes as seen in our case. These are best seen in the lateral skull view. If Magnetic resonance imaging (MRI) does not confirm the diagnosis in a suspected case of SWS, then computed tomography should be done because it is more likely to show calcification that can be elusive in MR imaging.¹⁰

Differential diagnosis includes Rendu-Osler-Weber syndrome, Maffucci's syndrome, angina-osteodystrophy syndrome, Von Hippel-Lindau disease, and Klippel Trenaumy-Weber syndrome.^{6,7,10-12}

According to Inan and Marcus, the port-wine nevi are localized on the face, especially on the right side and are detected in 87-90% of the cases. The lesion extension over the middle line is seen in 50% of the patients, and bilateral involvement can be detected in about 33% of the cases.¹³ In our case, the patient showed nevus flammeus only on the left side of the face without extension over the middle line.

CONCLUSION

The treatment of the Surge–Weber's syndrome is variable and depends on the presentation or intensity of its possible clinical features. The wide spectrum of clinical manifestations of SWS leads to multidisciplinary approaches for its management, such as Neurophysician, Ophthalmologist, Cosmetologist, Physiotherapist,

Radiologist, and Dentist. As a dentist, one must be aware of this condition and their possible complications arising during dental procedures and precautionary measures to avoid these complications.

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