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Periodontal management of gingival enlargement with Laser Therapy in Sturge-Weber Syndrome – A Case Report

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Abstract

Sturge-Weber Syndrome is a rare disorder that occurs with a frequency of approximately 1 per 50,000. The disease is characterized by an intracranial vascular anomaly, leptomeningeal angiomatosis, most often involving the occipital and posterior parietal lobes. Facial cutaneous vascular malformations, seizures, and glaucoma are among the most common symptoms and signs. The classic pathognomonic features of disease include angioma of the leptomeninges extending to cerebral cortex with ipsilateral angiomatous lesions, unilateral facial nevus after one division of trigeminal nerve and epileptic convulsions. The most characteristic oral manifestation is represented by gingival hemangiomatous lesion usually restricted to ipsilateral maxilla or mandible. The cutaneous angioma is called a Port Wine Stain. It is important for the clinician to be aware of the non- oral and oral manifestations in order to identify and manage appropriately to prevent development of complications. The present case report highlights a case of Sturge Weber syndrome associated with gingival enlargement and its management.

Keywords: Gingival Enlargement, Port Wine Stain, Sturge Weber Syndrome

Introduction

Sturge–Weber syndrome (SWS) or Sturge–Weber–Krabbe disease sometimes referred to as encephalotrigeminal angiomatosis, is a rare congenital neurological and skin disorder. It is one of the phakomatoses and is often associated with port-wine stains of the face, glaucoma, seizures, mental retardation, and ipsilateral leptomeningeal angioma (cerebral malformations and tumors). It is commonly referred to as “Sturge Weber syndrome” after Sturge and Weber who first described this affliction in 1879. Sturge Weber Syndrome can be classified into three different types. The Roach Scale is used for classification; Type 1 includes facial and leptomeningeal angiomas as well as the possibility of glaucoma or choroidal lesions. Normally only one side of the brain is affected. This type is the most common. Type

2 involvement includes a facial angioma (port wine stain) with a possibility of glaucoma developing. There is no any evidence of brain involvement. The symptoms can include glaucoma, cerebral blood flow abnormalities and headaches. Type 3 has leptomeningeal angioma involvement exclusively. The facial angioma is absent and glaucoma rarely occurs. This type is only diagnosed via brain scan.[1]

Pathophysiology

Sturge-Weber syndrome is an embryonal developmental anomaly resulting from errors in mesodermal and ectodermal development. Unlike other neurocutaneous disorders (phakomatoses), Sturge-Weber occurs sporadically (i.e., does not have a hereditary cause). It is caused by a somatic activating mutation occurring in the GNAQ gene. Radiological findings will show tram track calcifications on CT, bilaterally.[2]

Clinical Features: An estimated frequency of 1 per 50,000 live births have SWS.[3] Facial lesions of this syndrome include rosy purple nevus flammeus lesions that are sharply demarcated and usually flat; these occur on the ipsilateral side of the face in 90% of the patients. In some cases, they may extend onto the neck, chest, and back. The color varies from pink to purplish red and may decrease in intensity with increasing age.[4]

Seventy-five to 90% of children with SWS develop partial seizures by 3 years of age.[5] Of the longitudinal studies published, none demonstrate that early onset of seizures indicates a poor prognosis. In fact, retrospective studies do not support the widely held belief that seizure frequency early in life in patients who have SWS is a prognostic indicator.

However, some patients develop intractable epilepsy, permanent weakness, hemiatrophy, and visual field cuts, glaucoma, and mental retardation.[6,7] Other findings common to patients with SWS are a vascular headache (40-60%), developmental delay and mental retardation (50- 75%), glaucoma (30-70%), hemianopsia (40-45%), and hemiparesis (25-60%).[8] The hemiparesis and hemiatrophy are thought to arise from chronic cerebral hypoxia.

Oral manifestations include angiomatosis on the buccal mucosa and lips may present as a purplish red discoloration, which also may involve the soft palate, tongue, floor of the mouth, and gingiva. Gingival lesions may range from slight vascular enlargement to very large growths making closure of the mouth impossible.[9,10]

Diagnosis of SWS is made on the basis of the presence or absence of ophthalmologic or neurologic disease. The disease course, however, is variable and the patient must be continually monitored for complications.

Case Report

A 23 years old male patient presented to the department of periodontics with a chief complaint of swollen gums since two years. The patient did not complain of any pain and difficulty during mastication.The patient noticed the swelling which was gradually increasing and attained to the present size. The patient past medical history revealed that he was born full term and delivered normally and with no history of seizures but gradual blurring of vision was noticed in the right eye.

Extraoral examination revealed hemi-hypertrophy on the right side of the face with dusky red angimatous lesion extending from forehead to the lower border of mandible involving right eye. Face was asymmetrical and deviated towards left side and even deviation of nasal septum was seen towards the left, while examination of eye showed right reddish discoloration of bulbar conjunctiva, decreased vision and glaucoma. Increases in the size of upper and lower lips were noted (Figure 1).

Intra-oral examination showed soft tender dusky red over growth of right side of the upper and lower gingiva and

also involving buccal mucosa, floor of the mouth, pharynx, palate and uvula. On palpation, pulsations were felt on upper and lower lips. A solitary well-defined ulcer of size 0.8×1cm roughly oval in shape seen on attached gingiva apical to 13 which was tender on palpation (Figure 2).

Investigations

Computed Tomography of the brain revealed no calcification, but enlargement of choroid plexus was seen on affected side of the brain. Ophthalmic examination revealed dilated and tortuous vessels, optic atrophy and presence of glaucoma in relation to right eye.

Differential Diagnosis

The finding of the various investigations and clinical examinations confirmed the diagnosis of SWS type II. However, Klippel-Trenaunay Weber Syndrome has a striking resemblance to SWS.

Periodontal Therapy

The patient was explained about the treatment plan and the written consent was taken and a medical consent was obtained from the general physician. Initially thorough scaling and root planing was done. Patient was educated about maintaining good oral hygiene and was instructed to use 0.2% chlorhexidine mouth rinse. The patient was recalled after one week and gingivectomy procedure with Diode Laser was planned to minimize bleeding and less postoperative pain.

Adjunctive Laser Treatments

After adequate Local anesthesia with 2% lidocaine with 1:80000 adrenaline (Lignox 2% A; Indoco Remedies Ltd, L-32, Goa) gingival overgrowth correction and gingivoplasty of maxillary right gingiva were done with Diode Laser provided by the manufacturer (DenLase; China Daheng Group, Inc. Beijing CHINA). The diode laser (DenLase; China Daheng Group, Inc. Beijing CHINA) was operated at a peak power of 5.0 W with a pulse length of 200µs and pulse interval of 200µs (average power 1.0 W), using a 400 µm fibre-optic tip and a wavelength of 980 nm (Figure 3 & 4). Additional Low Level Laser Therapy (LLLT) using a Biostimulation probe provided by the manufacturer (DenLase; China Daheng Group, Inc. Beijing CHINA) was done. The laser was fired at the orifice of the gingival margin at a distance of approximate 1-2 mm, using a setting of 1.5W as a continuous wave.

Postoperative Instructions

Patient was advised to take antibiotic analgesic regimen (Amoxicillin 500 mg, three times a day; for five days and Ibuprofen 250 mg + Paracetamol 250 mg, three times a day for three days). Patient was advised to rinse with 0.2% chlorhexidine twice daily for two weeks postoperatively. The healing was uneventful without any overt complications (Figure 5). The patient was advised to return for a checkup after 3 months and 6 months (Figure 6) thereafter to look for any sign of recurrence of the gingival enlargement up to 12 months.

Discussion

SWS consist of a group of disorders which may present with neurological, cutaneous, ocular and oral manifestations. The most common clinical characteristic is a port wine stain on the face which normally follows the course of the trigeminal nerve, especially the ophthalmic division.[11] It has usually unilateral distribution along one or more segments of the trigeminal nerve.[12] Occasionally bilateral involvement or additional Port Wine lesions are found elsewhere in the body. Leptomeningeal angiomatosis is another important clinical feature of this syndrome and can lead to contra-lateral hypertrophy, progressive cerebral calcification, epileptic convulsions, mental retardation and angiomas in the eye causing glaucoma.[13]

In the present case, the patient had a no history of epileptic convulsions and CT brain showed no intracranial calcification in the occipito-parietal region. In the extra oral examination, there was no ocular involvement with buphthalmos, but glaucoma was confirmed by the ophthalmologist. The fact that PWS was unilateral, present since birth, spanning V2 division of the trigeminal nerve, not have any CNS manifestations but leading to the intraoral clinical finding of increased bleeding on probing and gingival enlargement in the affected quadrant (right maxillary) lead to a diagnosis of SWS Type II in the reported case.

Conclusion

Management of patient with Sturge–Weber syndrome may be challenging due to the risk of hemorrhage. Extra care must be taken when performing surgical procedures in the affected areas of mouth. The key in the management of SWS is to prevent or reduce the intensity of complications as the underlying pathology cannot be treated. Early identification and early institution of treatment and regular follow up with psychological counseling is imperative for the patient to have a better quality of life.

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Figure 1: Port Wine Stain on the Right Side of the Face

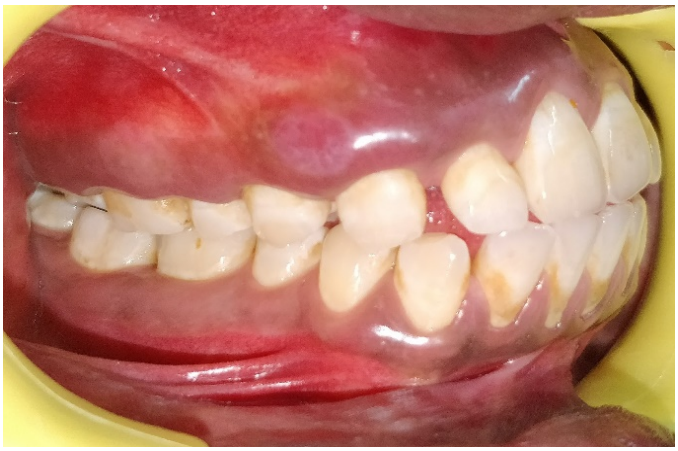

Figure 2: Right lateral view showing gingival enlargement

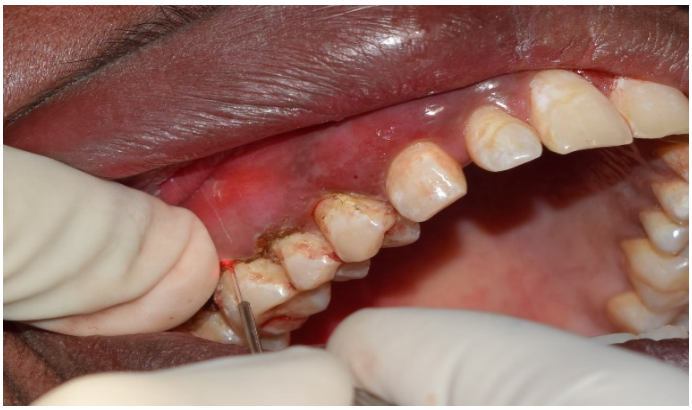

Figure 3: Laser gingivectomy

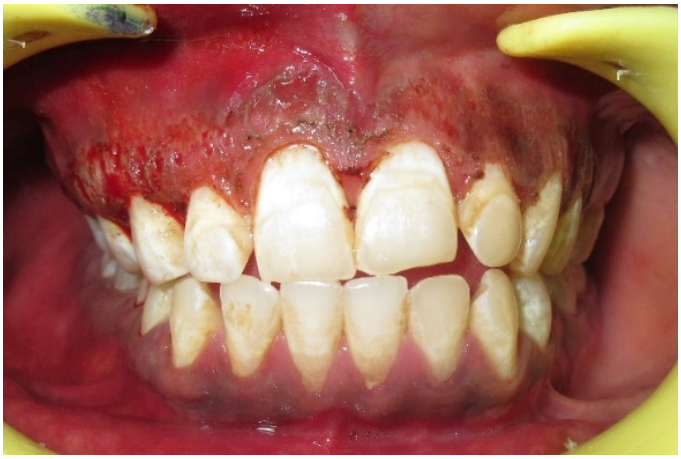

Figure 4: Post-Operative (Immediate)

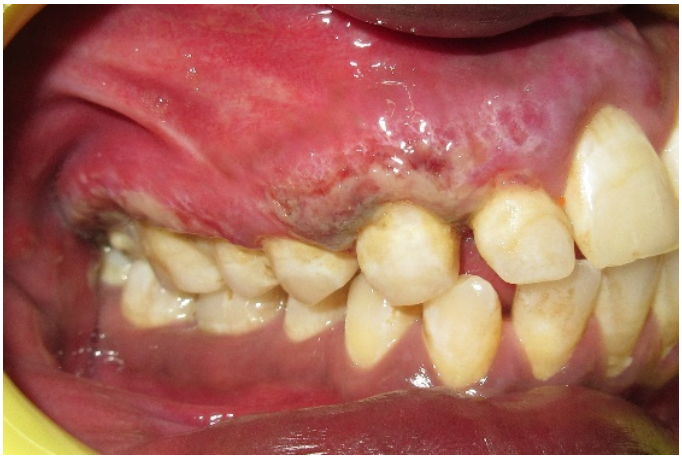

Figure 5: Post-Operative (One week)

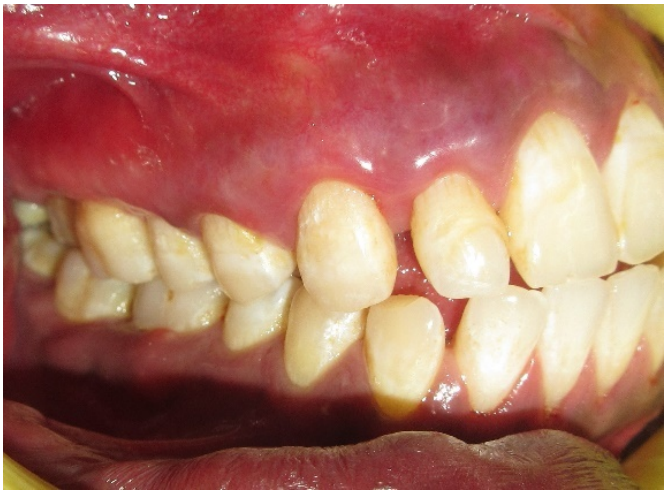

Figure 6: Post-Operative (Six Months)