Case Report

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Gingival Overgrowth in a Patient with Sturge-Weber Syndrome

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Abstract

Sturge-Weber syndrome is a rare congenital neurological and skin disorder. The pathognomic features of disease include angioma of the leptomeninges, facial nevus, convulsions and gingival hyperplasia. In the present case, a 30-year-old male patient presented with a port wine stain on the right side of the face, dilated blood vessels of the right eye, epilepsy and intra-orally with gingival enlargement in both maxillary and mandibular arches.

Keywords: Sturge-Weber Syndrome; Port Wine Stain; Gingival Enlargement.

Introduction

The first detailed description of SWS was given by Schirmer in 1860. Sturge further described SWS-related skin, neurological and ocular manifestations in 1879 and Weber described radiological features seen in these patients in 1929 [1]. This syndrome occurs in 1:50 000 live births [2]. Both sexes are equally affected [2]. Cushing [3] noticed that port-wine stain tend to occur in the division of the trigeminal nerve. In this syndrome seizures aggravate the progression of brain injury. Morphological and histological changes of periodontium occur as oral manifestations of the syndrome [4]. This article presents a case of Sturge-Weber syndrome associated with severe gingival enlargement.

Case Report

A 30-year-old male patient was referred to the Department of Periodontics from the Department of Psychiatric at King George's Medical University, Lucknow, in march of 2017 with the chief complains of swelling of gums in both upper and lower jaws for the last 4 months along with bleeding from gums and difficulty in chewing.

The patient's past medical history revealed that apparently he was healthy until the age of 24 years when he had epileptic attacks for which an antiepileptic drug (phenytoin sodium) was prescribed. He was also diagnosed of SWS along with port wine stains that were present since birth. He began to use phenytoin sodium at an age of 24 years and was still continuing at the time he visited dental department. Patient did not give any relevant past dental and family history.

Extra oral examination (Figure 1) showed presence of a right-sided hemihypertrophy of the face with port wine stains along the ophthalmic and maxillary branch of trigeminal nerve. His mouth was deviated toward the left side of his face. The upper and lower lips were swollen, edematous, and competent. Examination of eyes (Figure 2) showed presence of dilated blood vessels in right eye and left eye appeared normal.

The intraoral examination (Figure 3 and 5) revealed sessile, lobulated gingival swelling involving all four quadrants of the oral cavity. The papillae were swollen and covering more than cervical third of the tooth. Local irritants were found around the involved teeth. On palpation, the growth was moderately firm in consistency and was non-tender. Bleeding on probing in all four quadrants of the oral cavity was

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apparent. Periodontal pockets (9 to 10 mm) were detected in the regions of teeth #11, #12, #21 and #22.

The OPG (Figure 4) revealed generalized crestal bone loss, severe bone loss at teeth #11, #12, #21 and #22. The complete hemogram showed normal haemoglobin levels with no other abnormality in the total or differential white blood cell count. Bleeding and clotting times were within normal limits. Hepatitis B test was positive.

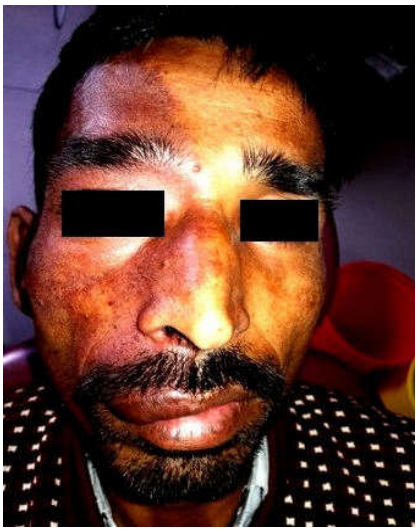

Fig. 1:

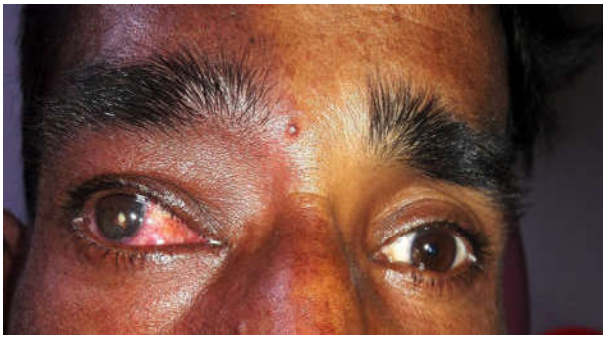

Fig. 2:

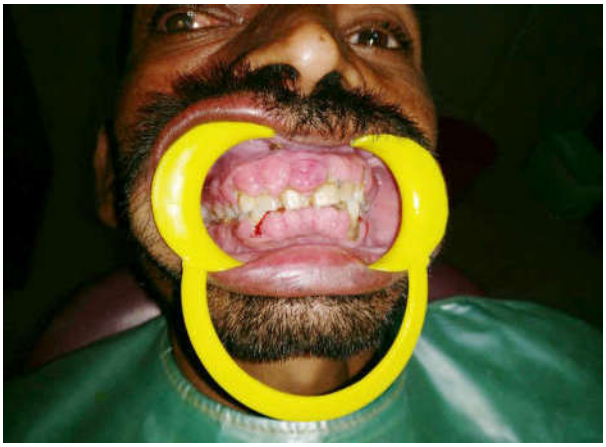

Fig. 3:

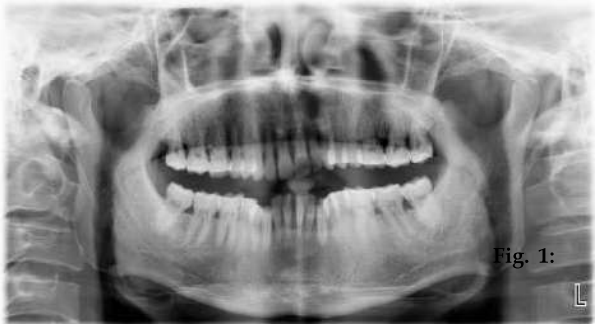

Fig. 1:

Fig. 4:

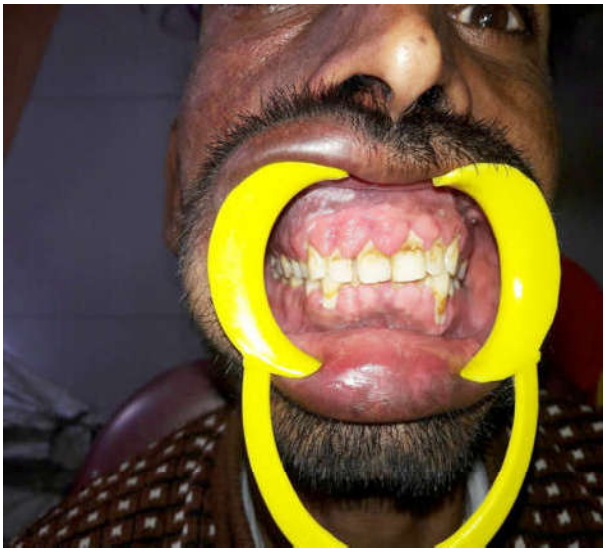

Fig. 5:

Differential Diagnosis

Klippel-Trenaunay-Weber syndrome, neurofibromatosis, Bannyan-Riley-Ruvalcaba syndrome, Maffuci's syndrome, Rendu-Oslar-Weber syndrome, Von Hippel Lindau disease and Coats disease [5].

Treatment

Treatment line begins with the phase 1 therapy which consists of patient motivation and education about concern of proper oral plaque control regimen. Oral prophylaxis (scaling and root planing) was carried out and the patient was instructed on oral hygiene.

Outcome and Follow-up

On recall visit after 1 month, when patient was examined clinically, bleeding on probing was reduced and patient was motivated for proper hygiene maintenance and further treatments. Patient was under treatment for hepatitis B.

Discussion

SWS is caused by a somatic mutation in a nucleotide transition in the gene GNAQ on chromosome 9q21 [6].

SWS is classified according to Roach's scale as follows [4]:

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.

Port-wine stains are present on face since birth and may range from small red macules to large red patches which blanch on pressure [7]. They occur more commonly on right side and do not extend over midline [7] as presented in our case. Most patients with brain involvement will begin seizure activity during infancy, but new neurological symptoms have been shown to develop in adulthood in certain cases [8]. In 1923, Dimitri demonstrated a classic radiographic features of the typical intracranial calcifications which were called as a ''ribbon-like'' or ''tram line'' pattern [9].

Most common oral manifestation is hemangiomatous lesion of gingiva. The gingival hyperplasia in these patients could be related to angiomatous proliferation in the connective tissue of the involved gingival tissues, secondary to anticonvulsant therapy further complicated by poor oral hygiene [4] as presented in our case.

Gingival overgrowth can be surgically removed with lasers, electrosurgery, or cryosurgery. Laser is use for the lightening of port wine stains. For the management of glaucoma, the first line of treatment modality is medication, when medication alone is failed to lessen elevated eye pressure then should go for surgery to preserve optimal eye function [10]. Only symptomatic treatment can be given for the facial disfigurement and neurologic problems. Anticonvulsants are treatment modalities for epilepsy. One of the most important parts of the treatment of such patients includes psychological counselling for the patient as well as the parents.

- Drugs which are causing gingival enlargement should be replaced with a suitable alternative.
- Multidisciplinary approach is crucial for proper management of the patient's need.
- Gingival hyperplasia in these patients could be further complicated by poor oral hygiene.
- Psychological counselling for the patient as well as the caregivers is important in such syndrome.

References

1. Neto FXP, Junior MAV, Ximenes LS, et al. Clinical features of Sturge-Weber syndrome. Intl Arch Otorhinolaryngol 2008;12:565–70.
2. Welty LD. Sturge-Weber syndrome: a case study. Neonatal Netw 2006;25:89–98.
3. Cushing H. Cases of spontaneous intracranial hemorrhage associated with trigeminal nevi. JAMA 1906;47:178-183.
4. Royle HE, Lapp R, Ferrara ED. The Sturge-Weber syndrome. Oral Surgery, Oral Medicine, Oral Pathology 1966;22(4):490-7.
5. Taly AB, Nagaraja D, Das S, et al: Sturge-Weber-Dimitri disease without facial nevus. Neurology 1987;37: 1063–1064.
6. Shirley MD, Tang H, Gallione CJ, et al. Sturge-Weber syndrome and port-wine stains caused by somatic mutation in GNAQ. N Engl J Med. 2013;368(21): 1971–9.
7. Di Rocco C, Tamburrini G. Sturge–Weber syndrome. Child's Nervous System 2006;22(8):909-21.
8. Raches D, Hiscock M, Chapieski L. Behavioral and academic problems in children with Sturge-Weber syndrome: differences between children with and without seizures. Epilepsy Behav. 2012;25(3):457–63.
9. Alexander GL, Norman RM. Sturge-Weber syndrome.In: Vinken PJ, Bruyn GW, eds. Handbook of Clinical Neurology, Vol. 14. Amsterdam: Holland Publishing Company; 1972:223-24.
10. Basler L, Sowka J. Sturge-Weber syndrome and glaucoma. Optometry. 2011;82(5):306–9.