



Management of Gingival Enlargement in Sturge-Weber Syndrome -A Case Report

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ABSTRACT

The Sturge-Weber syndrome is a rare uncommon neurocutaneous disorders with angiomas involving the leptomeninges (Leptomeningeal Angiomas) and skin of the face, typically in the ophthalmic (V_1) and maxillary (V_2) distributions of the trigeminal nerve. The cutaneous angioma is called a Port-Wine Stain. It is commonly referred to as "Sturge-Weber syndrome after Sturge and Weber who first described this affliction in 1879.

I. INTRODUCTION

Sturge-Weber syndrome (SWS) is a rare congenital disorder characterized by leptomeningeal hemangiomas; a facial Port-Wine nevus distributed over the trigeminal nerve area, usually unilateral. This syndrome is also called encephalotrigeminal angiomatosis. These hemangiomas cause neurological abnormalities, including epilepsy, mental retardation, and hemiplegia. Resection of gingival tissue and professional oral care are required because of enlargement of the soft tissue as a result of hemangiomas. SWS is caused by residual embryonal blood vessels and their secondary effects on surrounding brain tissues. Angiomas of SWS result due to failure of regression of a vascular plexus around cephalic portion of neural tube which is destined to become facial skin. This vascular plexus normally forms at 6th week of intrauterine life and regresses by 9th week. Failure of its regression results in residual vascular tissue which forms angiomas of leptomeninges, face and ipsilateral eye. These blood vessels show abnormal blood flow pattern as vasomotor phenomenon, venous occlusion, thrombosis and "vascular steal phenomenon" resulting ischemia, gliosis, atrophy and calcification of underlying cortical tissues. Although the leptomeningeal angioma in SWS is typically a static lesion, it has been demonstrated by some to be of progressive nature. The main

ocular manifestations buphthalmos and glaucoma occur due to secondary increase intra ocular tension due to increased secretion of aqueous humor by choroidal haemangioma.

II. CASE REPORT

An eighteen year old female patient named Kavitha reported to the Department of Periodontics, Tamilnadu Government Dental College, Chennai complaining swollen gums in maxillary and mandibular anterior gingiva for past 6 months. Patient has noted the increase in size of gums. Patient appears to have a thin physique, normal mental intelligence with normal appetite. There was no history of seizures and she was born full term and delivered normally.

Extra oral examination

Port wine stain or nevus flammeus on the right side of face and an ocular manifestation of cataract on right eye seen as clouding of lens. Patient right eye vision is blurry and cloudy. Face appears to be deviated to the right due to hemangioma.



Intraoral examination



Gingival enlargement of maxillary and mandibular anterior teeth along with pseudopockets. Subgingival deposits and plaque was noticed on both maxillary and mandibular anteriors..

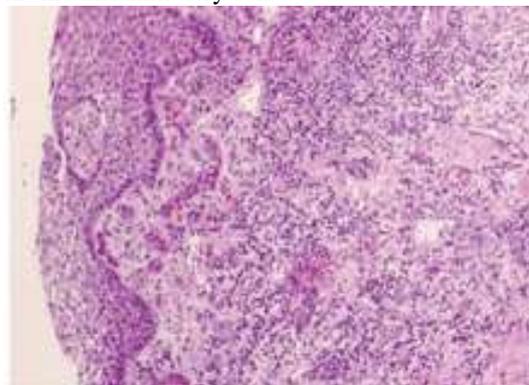


Reddish discoloration of alveolar mucosa in relation to teeth 11 to 15 and discoloration seems to extend upto attached gingiva, marginal gingiva and interdental papilla of teeth 11, 12, 13, 14, 15. Reddish discoloration also appears on mandibular labial mucosa in relation to 32, 31, 41, 42.

Blood investigations were normal. Normal RBC count, normal bleeding time, normal clotting time, International normalized ratio was 0.96. Orthopantomogram, CT

Histopathological report (Histopathology was done at Department of Oral Pathology and Microbiology, Tamilnadu Government Dental college, Chennai)

Histopathology report shows stratified squamous epithelium of variable thickness and the underlying connective tissue characterized by dense collagen fibres, blood vessels and scattered chronic inflammatory cells.



Management of Gingival enlargement

Non surgical periodontal therapy was done under aseptic conditions to remove plaque and subgingival deposits and patient was advised to brush twice daily and use 0.2% chlorhexidine mouthwash twice daily and the patient was recalled after 3 weeks.

Surgical procedure was explained to patient and a written consent was obtained from the patient. Medical consent was obtained from general physician. Excisional biopsy was taken from gingival enlargement in relation to 11 and 12. Biopsy was done under LA and excised tissue was kept in 10% formalin and sent for histopathological examination. No post biopsy complications occurred.

After 1 week External bevel gingivectomy was done on gingiva in relation to 15 to 25 under local anesthesia and antibiotic coverage of amoxicillin 3 times daily one day prior to procedure. Intra operative bleeding was controlled with pressure packs. Patient was advised to continue antibiotic analgesic regimen for 5 days after procedure. After 1 week external bevel gingivectomy was done on gingival enlargement in relation to 32 to 43. Antibiotic analgesic regimen continued for 3 more days. Patient was advised to use chlorhexine mouthwash after 2 weeks. The healing was satisfactory.



gingivectomy done on maxillary arch



POST OPERATIVE PHOTOGRAPH AFTER 1 week(mandibular anteriors)



gingivectomy done on mandibular arch after one week



POST OPERATIVE PHOTOGRAPH AFTER 1 week(maxillary anteriors)

III. DISCUSSION

Sturge Weber syndrome has extremely varied clinical features like portwine stain on face, cataract, glaucoma and mental retardation. In this case report patient has clinical manifestations of portwine nevi on right side of face, cataract on right eye. Patient is not mentally retarded. Pre operatively blanching of enlarged gingiva was observed. While treating the patient profuse bleeding was encountered intraoperatively. Pressure pack was given to manage the profuse bleeding. No postoperative complications were encountered.



IV. CONCLUSION

Nd Yag laser has affinity to hemoglobin. Nd Yag laser gingivectomy will achieve more hemostasis when combined with scalpel gingivectomy.

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