

Sturge Weber Syndrome- A Case Report

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Abstract Sturge Weber Syndrome is a rare non hereditary congenital sporadic disorder of elusive etiology. It has a vast continuum of cutaneous, neurologic, ophthalmic and oral manifestations. Routine dental procedures are associated with increased jeopardy of hemorrhage making treatment in such patients an exigent task for oral health care practitioners. Untoward complications can be prevented by having profound knowledge of this rare disorder. We report a case of characteristic oral manifestations in Sturge Weber Syndrome along with a review of relevant prevailing literature.

Keywords: Sturge Weber syndrome, congenital, encephalotrigeminal angiomas, vascular lesions, oral manifestations

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1. Introduction

Sturge Weber Syndrome also known as encephalotrigeminal angiomas is a rare non hereditary congenital disorder having a prevalence of approximately 1: 50,000. [1] It was first described by Schirmer and later more precisely by Sturge in 1879. [2] It has a vast spectrum of cutaneous, neurologic and ophthalmic manifestations which may or may not be associated with one another. [3] Oral involvement has also been reported in a few cases with considerable variations. [4] Here we report a case of Sturge Weber Syndrome (SWS) in a female patient with characteristic oral manifestations.

2. Case Report

A 20 year old female patient reported to us with a complaint of deposits on her teeth and presented with pigmentation on right side of the face and oral cavity which was static in size since birth. However, she alleged an increase in intensity of color with age from light pink to deep purplish red. There were no complaints of bleeding, pain, burning sensation, numbness or parasthesia in the pigmented or surrounding areas. There were no systemic symptoms or seizure episodes. Patient's mother gave history of full term pregnancy followed by normal delivery; there was no history any drug intake or trauma during pregnancy. Patient's family and medical history were non contributory.

On general physical examination she was found to be moderately built and nourished with no abnormality in extremities and joints. On extraoral examination a macular area of deep purplish-red pigmentation was noted on right half of the face extending over the temple region,

supraorbital region, infra-orbital region, bridge of the nose, malar region and the supralabial region along with a gross enlargement of right half of the upper lip (Figure 1).



Figure 1. Extra oral Photograph of the patient shows macular area of deep purplish red pigmentation on right side of the face and enlargement of right half of the upper lip.

On palpation no bruit or pulsation was elicited. The areas partly blanched under pressure. There was also presence of increased conjunctival vascularity in the right eye. Complete evaluation by an ophthalmologist ruled out the presence of glaucoma.

On intraoral examination similar lesions were noted on right side of maxillary labial mucosa maxillary labial

vestibule, buccal mucosa, and palate. There was absence of visible pulsations, however, the lesions partly blanched under pressure (Figure 2, Figure 3 and Figure 4).

A lateral skull view and posteroanterior view was taken which did not delineate the presence of any tram line calcifications.

On the basis of history and clinical appearance a final diagnosis of SWS was given. Patient along with her family was educated and made sentient about the condition and the possible complications that could arise during the necessary dental procedures.

A thorough plaque regimen program was initiated. It included oral prophylaxis, use of chlorhexidine mouth rinse, oral hygiene instructions, recording plaque index and motivating the patient. Mild gingival bleeding was encountered during oral prophylaxis which was arrested by local application of pressure. However, after the first visit patient was lost to follow up.



Figure 2. Intraoral photograph of the patient shows macular area of purplish red pigmentation involving right maxillary labial mucosa and vestibule



Figure 3. Intraoral photograph of the patient shows macular area of purplish red pigmentation involving half of the right buccal mucosa.



Figure 4. Intraoral photograph of the patient shows unilateral involvement of the palate with purplish red macular pigmentation.



Figure 5. (a and b) - Posteroanterior view and lateral skull view did not show the presence of any pathology

3. Discussion

Sturge Weber Syndrome which is also known as Sturge Weber Disease, Leptomeningofacial Angiomatosis,

Sturge-Weber-Dimitri Syndrome is a rare sporadic neurocutaneous disorder and belongs to a group of disorders called Phakomatoses. [4] Its etiology remains elusive, however some authors believe that angiomas arise

due to persistence of vascular plexus around the cephalic portion of neural tube which normally regresses around 9th week of intrauterine life [3].

It has no racial or gender predilection. [4] It is characterized by facial port wine stains, ocular abnormalities and leptomeningeal angiomas. The most characteristic clinical appearance is that of nevus flammeus or port-wine stains on the face along the distribution of Trigeminal nerve, of which ophthalmic division is the most commonly affected. Lesions vary in color and size ranging from pink, red and purple small to large macular lesions which mostly affect the right side and do not cross the midline. However, few cases of bilateral involvement have also been reported [5].

Our patient showed macular lesions on right side of the face along the distribution of ophthalmic and maxillary division of the trigeminal nerve.

Patients affected with this syndrome have also been reported to have mental retardation, delayed development, seizures, headache and hemiparesis. Ocular involvement commonly manifests as glaucoma and buphthalmos [3].

Sturge Weber Syndrome is a constellation of numerous signs and symptoms and an affected person may not display all the features. Roach developed a scale for classification which is as follows [6].

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.

According to the above criteria, our case is Type II SWS case.

The oral manifestations include ipsilateral port-wine stains of oral mucosa along with the hypervascular changes. Angiomatous lesion of gingiva may be present which can vary from slight vascular hyperplasia to colossal hemangiomas proliferation. Gingival hyperplasia can also be attributed to anticonvulsant medication and secondary to poor oral hygiene in mentally retarded patients. Macroglossia and maxillary bone hypertrophy have also been reported in a few cases [5].

Tram track calcification caused by calcification in apposing gyri, ipsilateral calvarial thickening and enlargement of the paranasal sinuses and mastoid may be visible in skull films. Higher imaging modalities like CT for calcification and MRI for brain assessment can also be used. MRI is the current gold standard for diagnosis of this disease especially in infants [3].

Absence of tram track calcification in skull films, seizures and mental retardation in our patient confirmed the absence of cerebral involvement.

Differential diagnosis include Rendu Osler Weber syndrome, Von Hippel Lindau Disease and Maffucci Syndrome. Diagnosis is made only on the basis of clinical and imaging features. Our patient exhibited characteristic clinical manifestations of Sturge Weber Syndrome [7].

Treatment and prognosis of SWS depends on the extent of involvement. Port wine stain can cause severe psychological trauma to the patients and hamper their

personality development. Port wine stains can be improved by dermabrasion, tattooing and flash lamp pulse dyed lasers. Various treatment modalities like sclerotherapy, cryotherapy, laser and surgical excision have been tried with varying degrees of success to surmount intraoral lesions [8].

Dental management should be mostly stressed on preventive measures. Patients should be educated, motivated and complied to follow a strict oral hygiene regimen to prevent dental caries and secondary gingival inflammatory enlargement. These patients can undergo endodontic procedure but over instrumentation should be avoided. Gingival hyperplasia has been reported to be successfully managed with carbon dioxide laser surgery with minimal hemorrhage [10].

It is a challenging task to carry out dental procedures in a SWS patient due to risk of severe intra- and postoperative haemorrhage. Special precautions to prevent and treat complications may include hospitalization, application of local anaesthetics with vasoconstrictors, dressings, splints [8].

4. Conclusion

Vast spectrum of clinical manifestations of SWS makes diagnosis a challenging task. Patients affected with SWS may or may not exhibit intraoral manifestations. It is imperative for oral health care practitioners to have profound knowledge of this rare congenital disorder and exhibit superfluous vigilance during routine dental procedures to prevent untoward life threatening complications.

References

- [1] Welty LD. Sturge-Weber Syndrome: A Case Study. Neonatal Network:© The Journal of Neonatal Nursing 2006;25(2):89-98.
- [2] Neto FXP, Junior MAV, Ximenes LS, de Souza Jacob CC, Junior AGR, Palheta ACP. Clinical Features of Sturge-Weber Syndrome. Intl Arch Otorhinolaryngol 2008;12(4):565-70.
- [3] Wahab Arif, Wahab Shagufta, Khan Rizwan Ahmad, Goyal Ruchi, Dabas Nisha, Sturge Weber Syndrome: A Review Bombay Hospital Journal, 2008; 50(1):55-58.
- [4] Gill NC, Bhaskar N Sturge Weber syndrome- a case report. Contemporary Clinical Dentistry 2010;1(3):183-185.
- [5] Khambete N, Risbud M, Kshar A. Sturge-Weber Syndrome: A Case Report. International Journal of Dental Clinics 2011; 3(1):79-81.
- [6] Royle HE, Lapp R, Ferrara ED. The Sturge-Weber syndrome. Oral Surgery, Oral Medicine, Oral Pathology 1966; 22(4):490-7.
- [7] Godge P, Sharma S, Yadav M, Patil P, Kulkarni S. Sturge Weber syndrome: A case report. Rev Odonto Cienc 2011; 26(4):366-369.
- [8] D. E. C. Perez, J. S. Pereira Neto, E. Graner, M. A. Lopes. Sturge-Weber syndrome in a 6-year-old girl. International Journal of Paediatric Dentistry 2005; 15:131-135.
- [9] Crinzi RA, Palm NV, Mostofi RM, Indresano AT. Management of a dental infection in a patient with Sturge-Weber disease J Am Dent Assoc 1980; 101: 798-800.
- [10] Darbar UR, Hopper C, Spoight PM. Combined treatment approach to gingival over growth due to drug therapy. J Clin Periodontol 1982; 23: 940 1996; 23(10):941-944.