www.contempclindent.org
DOI: 10.4103/0976-237X.72789

Sturge – Weber syndrome: A case report

Namrata C. Gill, Nandini Bhaskar[1]

Abstract

Sturge–Weber angiomatosis is a rare, nonhereditary developmental condition characterized by a hamartomatous vascular proliferation involving the tissues of brain and face. A report of a case with facial port wine stains, gingival overgrowth, and dilated ocular vessels is described.

Keywords: Angiomas, gingival overgrowth, port wine stain, Sturge–Weber syndrome

Introduction

Sturge–Weber syndrome (SWS) belongs to a group of disorders collectively known as the phakomatoses ("mother-spot" diseases). It consists of congenital hamartomatous malformations that may affect the eye, skin, and central nervous system (CNS) at different times, characterized by the combination of venous angiomas of leptomeninges, face, jaws and oral soft tissues.[1] SWS was first described by Schirmer in1860. More specific description was given by Sturge in 1879.[2]

SWS is believed to be caused by the persistence of vascular plexus around the cephalic portion of the neural tube. This plexus develops during the sixth week of I.U. development but normally undergoes regression during ninth week.[3]

Angiomas of leptomeninges are usually unilateral, located in parietal and occipital region. The presence of angioma results in alteration of vascular dynamics causing precipitation of calcium deposits in cerebral cortex underlying the angioma. Seizures, mental retardation, hemiplegia, or hemiparesis may develop secondary to this and their severity depends on the extent of lesion.[4]

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 be bilateral or totally absent or may extend to neck, limbs and other parts of the body.[4] Involvement of the area supplied by ophthalmic division is pathognomic. Ocular involvement can result in glaucoma, choroidal hemangioma, bupthalmos, or hemianopis.[5]

Intraorally angiomatosis may involve lips, buccal mucosa, palate, gingiva, and floor of mouth. This syndrome is of rare occurrence and management becomes complicated due to risk of hemorrhage.

Departments of Pedodontics and Preventive Dentistry and [1]Periodontics, Dr. HSJ Institute of Dental Sciences and Hospital, Sector – 25, Panjab University, Chandigarh, India

Correspondence: Dr. Namrata C. Gill,
House no. 1511, Phase 10, Sector 64, Mohali, Punjab, India.
E-mail: namratagill@gmail.com

Case Report

A 11-year-old female patient reported to the department of Pedodontics, with a chief complaint of bleeding gums and reddish discoloration on the left side of the face.

History revealed that the reddish discoloration (port wine stain) was present on the face since birth and was gradually darkening with age.

She was eldest of the three siblings born at full term by normal delivery. Family history was noncontributory. Patient was illiterate. There was no visible sign of mental retardation and patient had no history of convulsions.

Extra oral examination revealed that the port wine stain had unilateral distribution involving the left side of face. It extended from the middle of forehead and involved the eye, half of nose, cheek, philtrum, and left side of upper lip till the angle of mouth from where it extended to the left ear. The lower lip and lower jaw were not involved. A port wine stain was also seen unilaterally involving the left half of neck [Figure 1].

Examination of the eyes revealed that the blood vessels in the left eye were dilated [Figure 2]. Right eye appeared normal. Patient was advised ophthalmic consultation.

Intra oral examination showed a sharply delineated extension of port wine stain on the left half extending from the incisive papilla to the soft palate [Figure 3]. On the buccal aspect it involved the alveolar mucosa and attached gingiva.

Gingiva on the left side showed an overgrowth, while the right side appeared normal [Figures 4 and 5]. Patient had poor oral hygiene and bleeding on probing was present on the left side. Gingival overgrowth was bright red in color and showed blanching on applying pressure suggesting

angiomatous enlargement. Based on the clinical features, a diagnosis of Sturge-Weber syndrome was made. Physician's consent was taken prior to treatment.

Deciduous right maxillary second molar showed grade III mobility and was extracted under LA. A thorough plaque control regimen was started, which included oral prophylaxis at regular intervals, oral hygiene instructions, plaque index scoring, and motivation of patient at each visit.

After two months, patient reported no bleeding on tooth brushing and also no bleeding on probing was present. The size of gingival over growth did not decrease but the texture changed from edematous to firm. It was decided to follow-up the patient every month to evaluate plaque control.

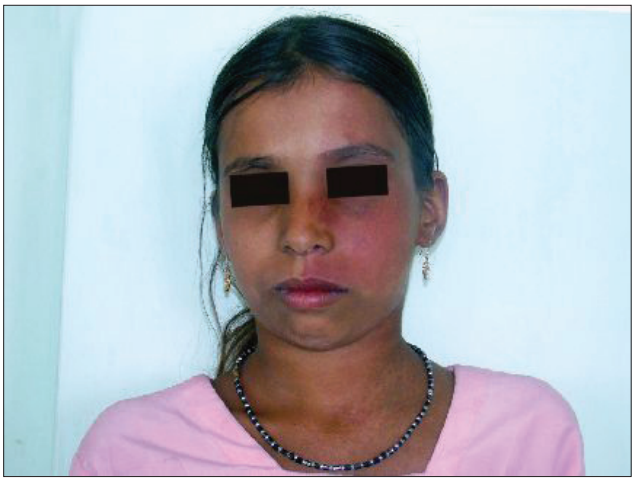

Figure 1: Portwine stains on left side of face and neck

Discussion

Port wine stains represent hamartomatous capillary malformations and are named so due to the deep red hue that they leave on the skin or mucosa.[6] Such lesions characteristically bleed profusely when traumatized.

Not all patients with facial port wine stains have Sturge – Weber angiomatosis. Only patients with involvement along the distribution of the ophthalmic branch of trigeminal nerve are at risk for the development of full condition.[3]

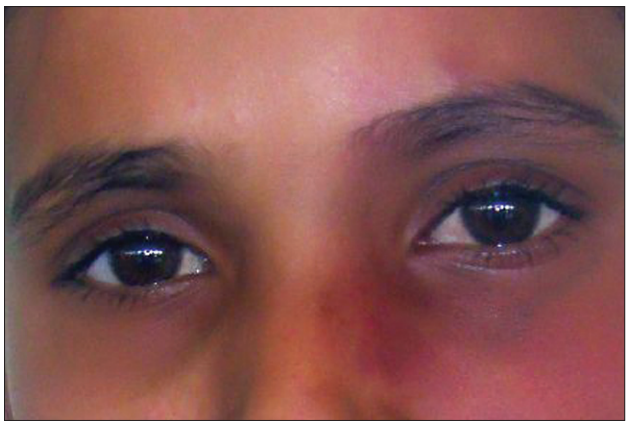

Figure 2: Dilated blood vessels of left eye

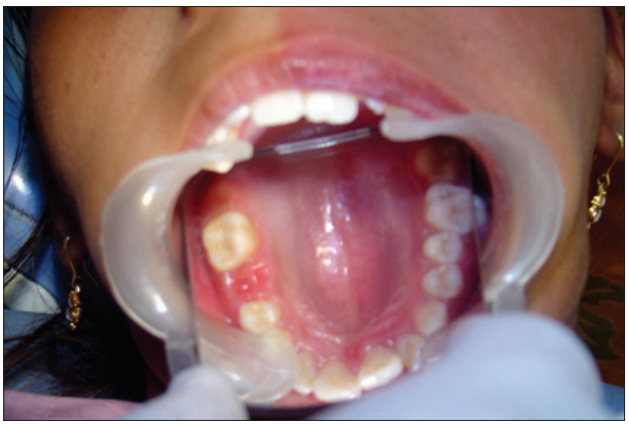

Figure 3: Unilateral (left) port wine stains on the palate

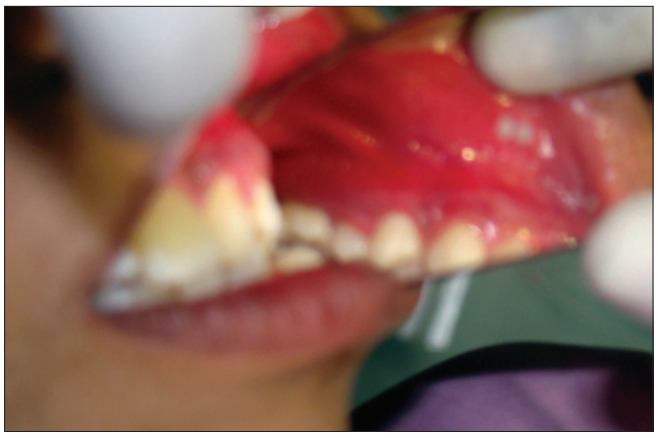

Figure 4: Gingival overgrowth on the left side

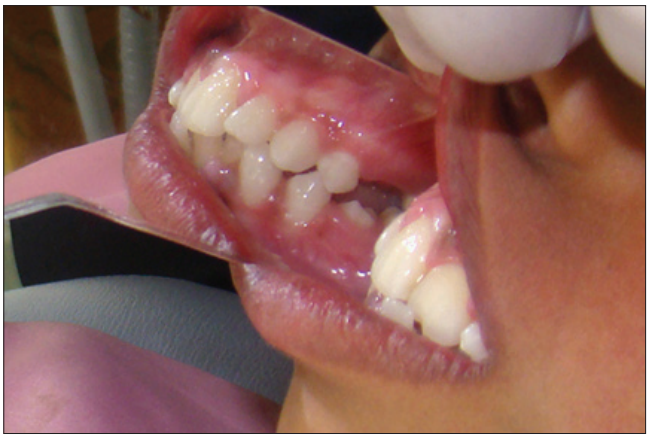

Figure 5: Normal gingiva on the right side

SWS is referred to as complete when both CNS and facial angiomas are present and incomplete when only one area is affected without the other. The Roach scale[7] is used for classification -

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

Differential diagnosis includes Rendu-Oslar-Weber syndrome, angio-osteodystrophy syndrome, Maffuci's syndrome, Von Hippel Lindau disease, and Klippel Trenaumy-Weber syndrome.[4,8]

Treatment and prognosis depends upon the nature and severity of clinical features. Presence of port wine stain can cause deep psychological trauma to patient and development of personality is affected in almost all patients.[8] Port wine stains can be improved by dermabrasion, tattooing, and flash lamp pulsed dye lasers.[3,8]

Intraoral involvement is common, resulting in hypervascular changes to the ipsilateral mucosa.[3] The gingiva in the present study showed hemangeomatous proliferation that felt soft on palpation and blanched under pressure. Such gingival overgrowth might be attributable to increased vascular component.[9]

In the present case, gingival overgrowth was managed by scrupulous oral hygiene maintenance. But even despite stringent plaque control, in some cases, gingivectomy may be required.[2,4] In such cases, achieving hemostasis can be a significant problem. Use of NdYag LASER for surgery provides the advantage of immediate hemostasis, minimal damage to surrounding tissues, and no post operative pain. Combination of gingivectomy and LASERs can be used if the overgrowth is very large. To prevent reoccurrence maintenance of good oral hygiene is very important even after gingivectomy.

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.

References

1. Rajendran R. Benign and malignant tumors of oral cavity. In: Rajendran R, Sivapathasundharam B, editors. Shafer's Textbook of Oral Pathology. 6th ed. Noida: Elsevier; 2009. p. 144-5.
2. Caiazzo A, Mehra P, Papagearge MB. The use of preoperative percutaneous transcatheter vascular occlusive therapy in the management of Sturge –Weber syndrome – Report of a case. J Oral Maxillofac Surg 1998;56:775-8.
3. Neville BW, Damm DD, Allen CM, Bouquot JE, editors. Oral and Maxillofacial Pathology. 3rd ed. St. Louis: Elsevier; 2009.
4. Suprabha B, Baliga S. Total oral rehabilitation in a patient with port wine stains. J Indian Soc Pedod Prev Dent 2005;23:99-102.
5. Gorlin RJ, Pindborg JJ. Syndromes of head and neck. New York: McGraw- Hill; 1964. p. 406-9.
6. Fishman SJ, Muliken JB. Hemangiomas and vascular malformations of infancy and childhood. Pediatr Clin North Am 1993;40:1177-200.
7. Roach ES. Neurocutaneous syndromes. Pediatr Clin North Am 1992;39:591-620.
8. Mukhopadhyay S. Sturge –Weber syndrome: A Case report. J Indian Soc Pedod Prev Dent 2008;26:29-31.
9. Yukna RA, Cassingham RJ, Carr RF. Periodontal manifestations and treatment in a case of Sturge –Weber syndrome. Oral Surg Oral Med Oral Pathol 1979;47:408-15.

Source of Support: Nil, **Conflict of Interest:** None declared.