Sturge–Weber syndrome

Natarajan Manivannan, Subramanium Gokulanathan, Ramakrishnan Swamy Ahathya[1], Gubernath[2], Rajkumar Daniel[1], Shanmugasundaram[1]

Departments of Oral and Maxillofacial Surgery, [1]Periodontology and Implantology, Vivekanandha Dental College for Women, Tiruchengode, [2]Periodontology and Implantology, Meenakshi Ammal Dental College, Chennai, Tamil Nadu, India

Address for correspondence:
Dr. Natarajan Manivannan
E-mail: ahathya@gmail.com

Received : 01-12-11
Review completed : 02-01-12
Accepted : 26-01-12

ABSTRACT

Encephalotrigeminal angiomatosis (Sturge–Weber syndrome) is a rather uncommon congenital condition characterized by the combination of venous angioma of the leptomeninges over the cerebral cortex with ipsilateral angiomatous lesions of the face, and sometimes the skull, jaws, and oral soft tissues. A case of portwine stain with intraoral gingival hemangioma is presented. There were no other systemic manifestations. Patient reported with a complaint of localized tumor-like swelling in gums. Based on the presence of sharply demarcated vascular lesion unilaterally on the face and with ipsilateral intraoral vascular hyperplasia in the lip and gingiva, a variant of encephalotrigeminal angiomatosis was diagnosed. Ultrasound Doppler flowmetry was used to determine the blood flow. Dental management included plaque control instructions, scaling, root planning, and excision of the lesion done under general anesthesia. Close follow-up and meticulous plaque control have kept the oral condition under fairly good control.

KEY WORDS: Hemangioma, portwine stain, Sturge–Weber syndrome, ultrasound doppler

Sturge–Weber syndrome is a rare congenital disorder characterized by the presence of leptomeningeal angiomas, portwine stains (sharply demarcated vascular lesions that occur unilaterally along the dermatomes supplied by first two divisions of trigeminal nerve), along with ocular disorders, mental retardation, and oral involvement. It was first described by Schirmer and later more specifically by Sturge in 1879. It is also known as Sturge–Weber disease, encephalotrigeminal angiomatosis, meningofacial angiomatosis, and Sturge–Weber–Dimitri syndrome.[1]

It is a congenital disorder occurring due to dysfunction of embryonal vascular system, resulting in hemangiomatosis. The classic feature of this disease is angioma of the leptomeninges. The other common clinical features are epilepsy (80%), dermal angiomas resulting in portwine stains (76%), abnormal findings in skull radiographs (63%), mental retardation (54%), ocular involvement (37%), and hemiplegia (37%).[1,2]

Access this article online

Quick Response Code:

Website: www.jpbsonline.org

DOI: 10.4103/0975-7406.100304

Portwine stains or nevus flammeus represent hamartomatous capillary malformations and are named so due to the deep purple hue that they leave on the skin or mucosa.[3] Intraorally, angiomatosis can involve lips, causing macrochelia, and also buccal mucosa, palate, and floor of the mouth. Gingival enlargement can vary from light vascular hyperplasia to monstrous overgrowth, making closure almost impossible. Pyogenic granulomas, unilateral hypertrophy of alveolus, ipsilateral premature eruption, or delayed eruption and malocclusion are the other abnormalities reported.[1,4] This syndrome is of rare occurrence and management becomes complicated due to the risk of hemorrhage.

This paper presents a case report of a patient with portwine stain and intraoral hemangioma.

Case Report

A 20-year-old female patient reported with a chief complaint of pain and bleeding gums while brushing in left upper posterior

How to cite this article: Manivannan N, Gokulanathan S, Ahathya RS, G, Daniel R, S. Sturge-Weber syndrome. J Pharm Bioall Sci 2012;4:349-52.

region and for replacement of a poorly constructed bridge in the upper anterior region. She was unable to follow proper oral hygiene because of recurrent bleeding from the swelling in the left upper posterior region.

Patient's history revealed that the lesion started 3 years back and slowly attained the present stage. She was free of any epileptic symptoms and there was no sign of mental retardation. The patient was communicative but apprehensive. The family history revealed no relevant findings.

Extraoral examination revealed the presence of portwine stains on the left-hand side of the face, extending along the second division of the trigeminal nerve unilaterally [Figure 1]. Facial asymmetry and macrochelia was noted with enlargement on the left side.

Intraoral examination revealed prominent reddish purple gingival enlargement posteriorly on the left side [Figures 2 and 3]. Blanching on pressure was noted in the enlarged gingiva, suggesting angiomatous enlargement. Upper anterior teeth had a broken fixed partial denture. Teeth on the lower arch were malpositioned. Oral hygiene was poor with extensive amounts of plaque and calculus.

Chest X-ray and PA view of the skull showed no abnormality. Blood picture was normal. Ultrasound Doppler flowmetric study [Figures 4 and 5] was done to determine the blood flow in the growth. Treatment plan consisted of oral prophylaxis, surgical excision of the lesion under general anesthesia (GA), and replacement of the worn out bridge in the upper anterior region.

The scope of the treatment and the goals were explained and informed consent was obtained from the patient. A thorough plaque control regimen was started from day 1 to minimize the inflammatory component of the gingival enlargement. It included oral prophylaxis, use of chlorhexidine mouth rinse, oral hygiene instructions, and motivating the patient at each visit.

Intralesional injection with sodium tetradecyl sulfate was given once a week for 2 weeks as a pretreatment to the excision of the growth.[5,6] Because of the risk of bleeding, surgical excision was done under GA following hospitalization. The excision of the growth was done using electrosurgery [Figure 6]. Periodontal pack was placed after the patient regained consciousness. Patient was given postoperative instructions and discharged after 24 hours.

Histopathologic examination of the excised tissue revealed excessive number of dilated blood vessels lined by endothelial



Figure 1: Extraoral view



Figure 2: Intraoral view of gingival hemangioma



Figure 3: Intraoral occlusal view (mirror image)

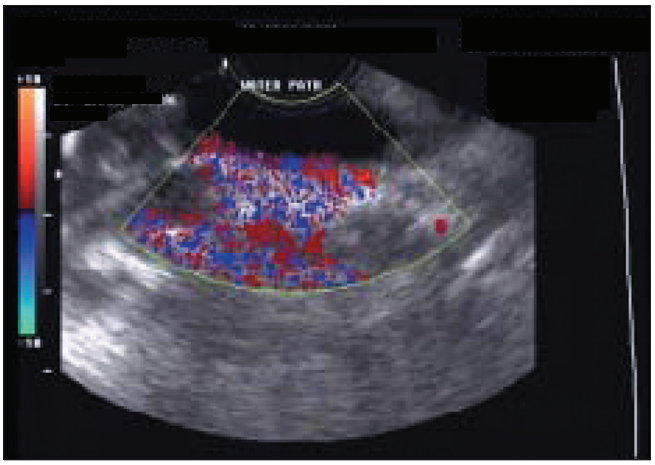


Figure 4: Color Doppler of the intraoral lesion

cells in the connective tissue matrix, confirming the lesion to be a hemangiomatous lesion [Figure 7].

Patient was reviewed on the 7th postoperative day. One-month follow-up showed satisfactory healing of the gingiva at the operated site [Figure 8]. A newly constructed fixed partial denture was given to replace the old one in the region of the upper anterior teeth. Oral hygiene instructions were reinforced and the patient was made to understand its importance. Sixth month follow-up showed complete regression of the lesion and her oral hygiene maintenance was satisfactory [Figures 9 and 10].

Discussion

Although Sturge–Weber syndrome appears with a number of manifestations, they are highly variable. In the case presented here, there were no systemic manifestations except for portwine stains on the face and intraoral hemangioma. Unless the vascular lesions include the region innervated by the ophthalmic branch of the trigeminal nerve, usually the patient does not have central nervous system involvement.

Portwine stains on the face can be a cosmetic problem and if needed should be treated by a plastic surgeon. A high dose of hydrocortisone given orally in infancy results in regression of well-localized small lesions.[7] Other methods of treatment include dermabrasion, tattooing, and lash lamp pulse tunable dye laser therapy. They can result in partial or complete clearing of the portwine stains. Cryosurgery may be used to correct lip and other soft tissue deformities.[8,9] Although all the treatment modalities were explained to the patient, she was not interested in correcting the extraoral lesion.

The dental rehabilitation of patients with Sturge–Weber disease is a complex process requiring initial conservative management and later surgery.[10] Poor oral hygiene leading to secondary inflammatory gingival enlargement is often encountered. Patient education and implementation of preventive procedures should be done.[11] A thorough plaque control regimen can go a long way in avoiding gingivectomy which is risky in these patients. Hancock *et al*.[12] achieved regression of gingival enlargement by vigorous plaque control measures alone.

Drug-induced gingival enlargement is reported to be often superimposed on the existing hemangioma when the patient is under epileptic medication, as epilepsy is one of the common clinical features in these patients.[1,2] A distinction should be made between hemangioma and drug-induced gingival

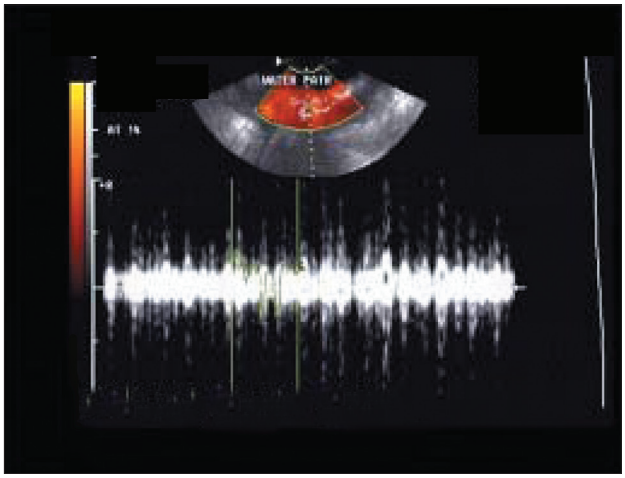


Figure 5: Spectral Doppler flowmetry of the intraoral lesion



Figure 6: Electrosurgical excision under GA

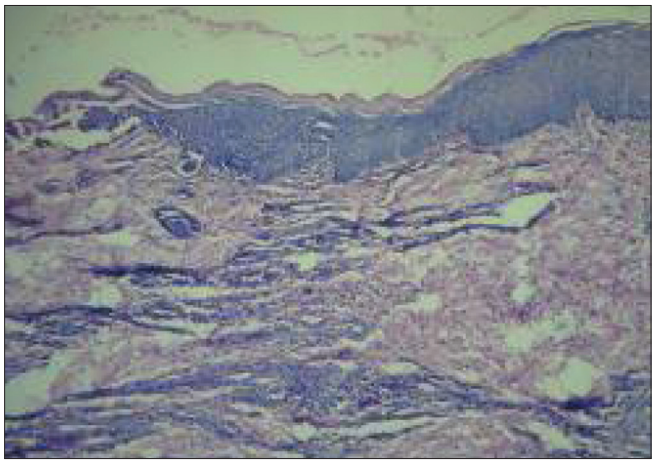


Figure 7: Histopathologic view showing numerous dilated capillaries



Figure 8: One month postoperative view showing satisfactory healing



Figure 9: Sixth month postoperative view showing remission of the lesion

enlargement with the help of angiography.[13] Although phenobarbitone can also cause gingival hyperplasia,[14] in the present case, it should be noted that the patient had no history of epilepsy and the blanching effect seen on examination confirmed the angiomatous nature of gingival enlargement.

In spite of strict oral hygiene measures, angiomatous gingival enlargement poses a threat to gingival health and may at some point require gingivectomy.[9] Whenever a surgical procedure is planned, achieving hemostasis can be a significant problem making hospitalization mandatory.[2] The various methods used to manage the risk of perioperative hemorrhage are:[15]

1. Patient's blood typed and cross matched
2. Provision for blood transfusion
3. Use of hemostatic agents–topical bovine thrombin
4. Use of postoperative splints
5. Injecting sclerosing solutions
6. Percutaneous transcatheter vascular embolization using gelfoam or polyvinyl alcohol.

Electrosurgery was preferred in this case since electrosurgical procedures have several advantages which include the following:[16]

1. Clean tissue separation, with little or no bleeding
2. Clear view of the surgical site
3. Planing of soft tissue is possible
4. Access to difficult-to-reach areas is increased
5. Time and operator fatigue are reduced
6. The technique is pressureless and precise.

Conclusion

Despite their benign origins and behavior, hemangiomas in the region of oral cavity are always of clinical importance to the dental profession and require appropriate clinical management. Dental practitioners and oral surgeons need to be aware of these lesions because they may pose serious bleeding risks. Suitable armamentarium should be at disposal to manage any complications that may arise.



Figure 10: Sixth month postoperative view showing remission of the lesion

References

1. Gorlin RJ, Pindborg JJ, editors. Syndromes of head and neck. New York: McGraw-Hill; 1964. p. 406-9.
2. Caiazzo A, Mehra P, Papagearge MB. The use of preoperative percutaneous transatheter vascular occlusive therapy in the management of sturge weber syndrome-Report of a case. J Oral Maxillofac Surg 1998;56:775-8.
3. Fishman SJ, Muliken JB. Hemangiomas and vascular malformations of infancy and childhood. Pediatr Clin North Am 1993;40:1177-200.
4. Ahluwalia TPS, Lata J, Kanwa P. Sturge-Weber Syndrome with intra oral manifestations: A case report. Indian J Dent Res 1998;9:140-5.
5. Cek DI. Sclerosing agent therapy for venous malformations of the lip and perioral region. Eur J Plast Surg 1992;15:276-8.
6. Woods JE. Extended use of sodium tetradecyl sulfate in treatment of hemangiomas and other related conditions. Plast Reconstr Surg 1987;79:542-9.
7. Saddan NS, Wolach B. Treatment of hemangiomas of infants with high doses of prednisone. J Pediatr 1996;128:141-6.
8. Stewart RE, Barber TK, Trontman KC, editors. Pediatric dentistry, scientific foundations and clinical practice. St. Louis: CV Mosby; 1982. p. 502-3.
9. Laskin MD, editors. Oral and maxillofacial surgery. Vol. 2. St. Louis: CV Mosby Co.; 1985. p. 528-9.
10. Perkins TM, Duncan WK, Hill WJ, Krolls SO. The sturge weber syndrome. A case involving a 13 year old black male. Ann Dent 1992;51:40-3.
11. Yukna RA, Cassingham RJ, Carr RF. Periodontal manifestations and treatment in case of sturge weber syndrome. Oral Surg Oral Med Oral Pathol 1979;47:408-15.
12. Hancock RH, Swan RH. Nifedipine induced gingival overgrowth. Report of a case treated by controlling plaque. J Clin Periodontol 1992;19:12-4.
13. Wilson S, Venzwel JM, Miller R. Angiography, gingival hyperplasia and sturge weber syndrome: Report of a case. J Dent Child 1986;53:283-6.
14. Gregarion AP, Schneider PE, Shaw PR. Phenobarbitone induced gingival over growth report of two cases and complications in management. J Dent Child 1996;63:408-13.
15. Suprabha BS, Baliga M. Total oral rehabilitation in a patient with portwine stains. J Indian Soc Pedod Prev Dent 2005;23:99-102.
16. Gnanasekhar, Al-Duwairi. Electrosurgery in dentistry. Quintessence Int 1998;29:649-54.

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