**Journal of Advanced Medical and Dental Sciences Research**

@Society of Scientific Research and Studies ***NLM ID:*** *101716117*

Journal home page: www.jamdsr.com doi: 10.21276/jamdsr Indian Citation Index (ICI) Index Copernicus value = 91.86

(e) ISSN Online: 2321-9599; (p) ISSN Print: 2348-6805

# Case Report

## Gingival enlargement associated with type II struge-weber syndrome and its management- A case report

[1]Shubhangi Gupta Law, [2]Aditya Law, [3]Neha Joshi, [4]Sukanya Mohanty

[1]Senior Lecturer, Department of Periodontics, Kusum Devi Sunderlal Dugar Jain Dental College, Kolkata, West Bengal, India;
[2,3,4]Private Practitioners, India

***ABSTRACT:***
Gingival enlargement is one of the most common feature of gingival diseases. It can occur due to many underlying causes. The most common type of enlargement is the inflammatory type. There are also enlargements which are associated with underlying genetic disorder. This is a case of a 17-year old female, with gingival enlargement and an extraoral 'port-wine stain' on the ipsilateral side of the face since childhood. Since the gingival enlargement was erythematous, electrocautery was used for the excision after which it was sent for histopathological investigation. To treat any kind of gingival enlargement it is very important to know the underlying cause of it. Similarly, in this case we managed the enlargement according to the medical condition. Thus, the medical condition should be considered before proceeding for any treatment.
**Key Words:** Struge-weber syndrome, port-wine stain, Gingival enlargement.

Received: 10 April, 2022 Accepted: 13 May, 2022

**Corresponding author:** Shubhangi Gupta Law, Senior Lecturer, Department of Periodontics, Kusum Devi Sunderlal Dugar Jain Dental College, Kolkata, West Bengal, India

**This article may be cited as:** Law SG, Law A, Joshi N, Mohanty S. Gingival enlargement associated with type II struge-weber syndrome and its management- A case report. J Adv Med Dent Scie Res 2022;10(6):1-4.

### INTRODUCTION

Sturge–Weber syndrome (SWS) or Sturge–Weber–Krabbe disease, sometimes referred to as encephalotrigeminal angiomatosis, is a rare congenital neurological and skin disorder. It is one of the phakomatoses and is often associated with port-wine stains of the face, glaucoma, seizuresand ipsilateral leptomeningeal angioma (cerebral malformations and tumors)[1].It was first described by Schirmer and later more specifically by Sturge in 1879[2]. SWS is classified according to the Roach scale[3] as follows:

- Type I: Both, facial and leptomeningeal angiomas (LA); may have glaucoma.
- Type II: Facial angioma alone; may have glaucoma.
- Type III: Isolated LA; usually no glaucoma.

Port-wine 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[4].The colour varies from pink to purplish-red and may decrease in intensity with increasing age. Cushing[5] noted that the nevi tend to occur along the distribution of trigeminal nerve. However, Alexander and Norman[6] found the trigeminal relationship to be secondary. They assumed that the distribution of the facial nevus was determined, in part, by the position of the processes and fissures in the developing face.

Intraorally, angiomatosis can involve lips (causing macrochelia), and also buccal mucosa, palate, and floor of the mouth. Gingival enlargement can vary from a light vascular hyperplasia to a monstrous overgrowth, making closure of the jaws almost impossible. Pyogenic granulomas, unilateral hypertrophy of alveolus, ipsilateral premature eruption, or delayed eruption, and malocclusion are the other abnormalities reported that are associated with SWS[2,7]. Because of the vascular condition and its tendency to bleed easily, management of the lesions pose a challenge to the clinician.

This is a case of gingival enlargement in Type II SWS in which there were no neurological manifestations and the subsequent management of the lesion.

## CASE REPORT

A 17-yearold female patient, reported to the Department of Periodontics, Kothiwal Dental College and Research Centre, Moradabad, with a complaint of swelling in the upper right back tooth region of the jaw for the past 2 years. She had first noticed the swelling 2 years ago and also complained of bleeding from that side while brushing. She also reported that there was no change in the size of swelling in the past 2 years.

## MEDICAL HISTORY

Patient was systemically healthy. She had no history of seizures and was not on any medication.

## CLINICAL EXAMINATION

The patient did not show any neurological deficit, spoke in a clear and articulate manner, had moderate build,normal temperature, blood pressure and pulse.

Extraorally,aport-wine stain was seen on the right side of the face involving the malar region, upper eyelid and upper lip of that side (Figure I(a),(b)).

**Figure I(a): Port-wine stain on the right side of face and hemi-hypertrophy seen.**

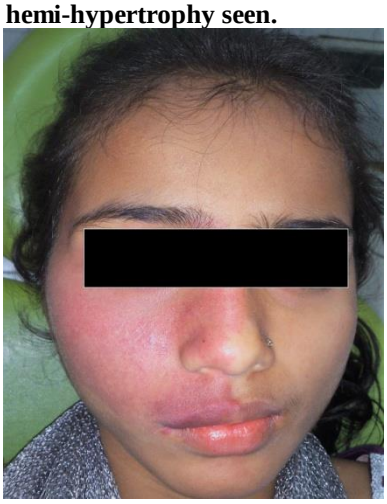
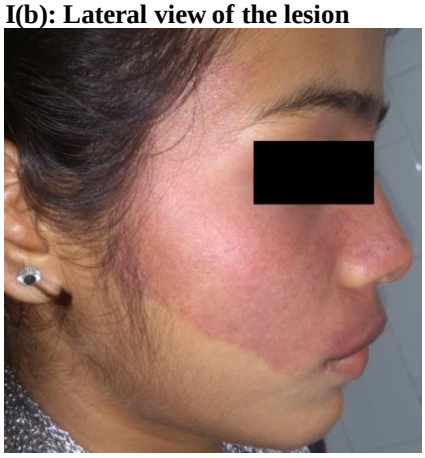
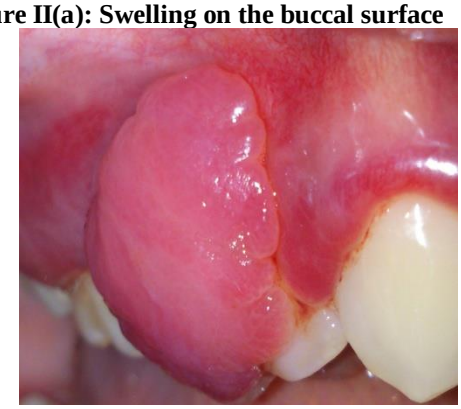


**Figure I(b): Lateral view of the lesion**



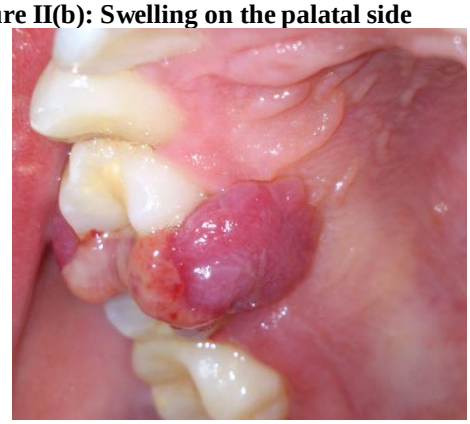
Patient mentioned that this stain was present since birth. Hemihypertrophy of that side was seen. The mouth was also deviated towards the right side. On intraoral examination, a well demarcated, pinkish red, erythematous, oval, pedunculated growth was seen extending from mesial side of #14 to the distal side of #15, including the interdental area, and covering the entire buccal surface of #15 (Figure II(a)).

**Figure II(a): Swelling on the buccal surface**



The swelling was also seen extended to the palatal side from mid palatal surface of 14 till the distal side of 15 (Figure II(b)).

**Figure II(b): Swelling on the palatal side**

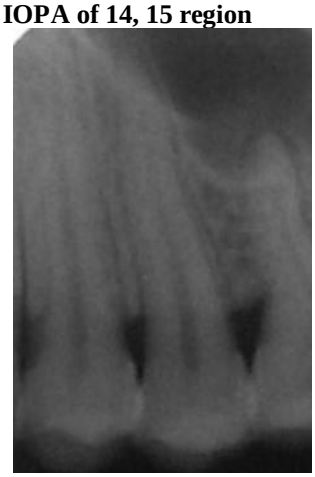


On palpation, the growth was soft in consistency and it bled readily on probing. It was also observed that gingiva on the ride side was more reddish in colour than the left side.

## INVESTIGATIONS

IOPA showed no evidence of bone loss in relation to #14,#15 (Figure III).

**Figure III: IOPA of 14, 15 region**



Routine blood investigationsshowed normal haemoglobin levels with normal red blood cell and white blood cell count.

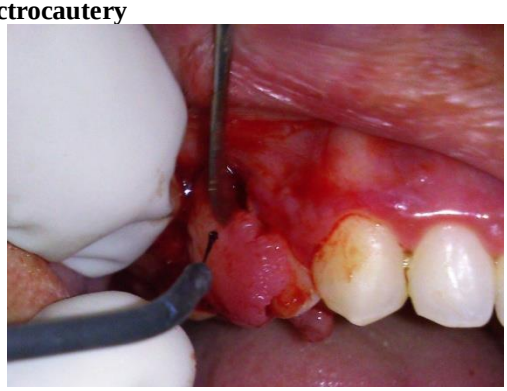
Based on the history and clinical findings, a

provisional diagnosis of gingival enlargement associated with Sturge-weber syndrome was made. Differential diagnosis was given as Klippel-Trinaunay Weber syndrome because it is also associated with port-wine stain,but it is not as intense in colour as it is in SWS.

**MANAGEMENT**

The complete treatment plan was explained to the patient and a written consent was obtained. Scaling and root planning was done to remove all the local irritating factors and the patient was educated regarding the oral hygiene maintenance. After phase I periodontal therapy, total surgical excision was planned. Since the swelling bled easily, electrocautery was used for the excision. A small ball-point electrode was used to excise the growth from the base, first from the buccal side and then from the palatal side (Figure IV).

**Figure IV: Excision of the growth using electrocautery**



Care was taken that the electrocautery tip did not touch the alveolar bone as the excess heat could cause necrosis of bone.

Post-operative instructions were given and the patient was kept on amoxicillin 500mg thrice daily for 5 days,andparacetamol 500mg was prescribed for 2 days. 10ml of 0.2% chlorhexidine mouthwash was advised twice daily for 10 days.

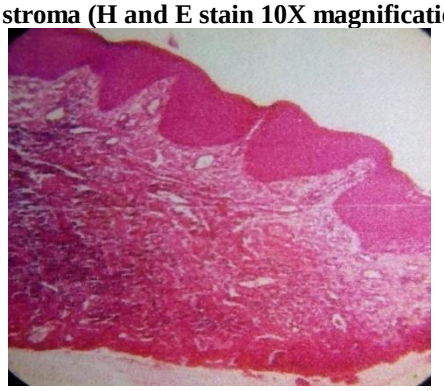
The excised tissuewas sent for histopathological analysis.Postoperative healing was satisfactory. There was no re-appearance of the enlargement when the patient was re-evaluated after 12 months (Figure V).

**Figure V: Post-operative after 12 months**

**HISTOPATHOLOGICAL EVALUATION**

H&E stained section revealed a hyperplastic epithelial lining with mitotically active cells. Connective tissue consisted of a proliferation of multiple blood vessels of varying calibre, surrounded by a dense stroma resembling granulation tissue. Overall histopathologic features were suggestive of telangiectatic granuloma (Figure VI).

**Figure VI: Histopathologic figure showing proliferation of blood vessels and dense connective tissue stroma (H and E stain 10X magnification)**



**DISCUSSION**

SWS has many manifestations that are highly variable. In this particular case, there were no neurological manifestations, only the port- wine stain and gingival enlargement was present. Unless the vascular lesion involves the region innervated by the ophthalmic nerve, the patient does not usually have central nervous system involvement[7].

Port-Wine stains which are present since birth can be a cosmetic problem and should be treated by a plastic surgeon. This stain is most commonly unilateral and seen along the trigeminal nerve distribution[7]. Bioxeda et al. found facial PWS distributed predominantly over the distribution of V2 branch of the trigeminal nerve in 88% of a total of 121 patients with PWS affecting skin. The lesions were unilateral in 86% of the patients. Extra-facial PWS was more common when the V3 division was involved[8].

The dental management of such patients require conservative management initially, followed by surgery. Due to the presence of local factors, secondary inflammatory gingival enlargement is mostly encountered. Accordingly, the patient was trained in oral hygiene methods and then phase I periodontal therapy was carried out. Hancock et al.[9] achieved regression of gingival enlargement by vigorous plaque control measures alone.

After proper oral hygiene measures, the inflammatory component of the swelling regressed and the enlargement had to be removed surgically. Whenever a surgical procedure is planned, achieving hemostasis can be a significant problem[2]. The various methods used to manage the risk of par-operative hemorrhage are[10]:

Provision for blood transfusion

Use of hemostatic agents–topical bovine thrombin

Use of postoperative splints
Injecting sclerosing solutions
Percutaneous transcatheter vascular embolization using gelfoam or polyvinyl alcohol.

**Electrosurgery was preferred in this case since electrosurgical procedures have several advantageslike[11]:**

- Clean tissue separation, with little or no bleeding
- Clear view of the surgical site
- Time and operator fatigue are reduced

In this case since the enlargement was present from a very long time, so histologically there were features of a granuloma along with enormous blood capillaries. There was no history of neurological disorders or of ocular disorders, but there was a unilateral port wine stain and telangiectatic granuloma on the same side. Hence, this case was diagnosed as SWS type II.

## CONCLUSION

SWS is a rare genetic disorder and is associated with intraoral haemangiomas which should be carefully managed by a clinician. The surgical management of this vascular lesion in SWS should be performed with utmost care and planning. To prevent any further complication and recurrence the patient should be under regular follow-up.

## CONFLICT OF INTEREST

None

## REFERENCES


1. Dorland WA. 30th ed. Philadelphia, Pa: Saunders; 2003. Dorland's Illustrated Medical Dictionary.
2. Gorlin RJ, Pindborg JJ, editors. Syndromes of head and neck. New York: McGraw-Hill; 1964. pp. 406–9.
3. Roach ES. Neurocutaneous syndromes. PediatrClin North Am1992;39:591-620.
4. Fishman SJ, Muliken JB. Hemangiomas and vascular malformations of infancy and childhood. PediatrClin North Am. 1993;40:1177–200.
5. Cushing H. Cases of spontaneous intracranial haemorrhage associated with trigeminal nevi. JAMA. 1906;47:178–83.
6. Alexander GL, Norman RM. Sturge-Weber syndrome. In: Vinken PJ, Bruyn GW, editors. Handbook of clinical Neurology. Vol. 14. Amsterdam: Holland Publishing Company; 1972. pp. 223–40.
7. Manivannan.N et al. Struge Weber syndrome. J Pharma Bioallied Sci2012 Aug; 4(suppl 2):S349-352.
8. Bioxeda P, de Misa RF, Arrazola JM. Facial angioma and the Sturge-Weber syndrome: A study of 121 cases. Med Clin (Barc) 1993;101:1-4.
9. Hancock RH, Swan RH. Nifedipine induced gingival overgrowth. Report of a case treated by controlling plaque. J ClinPeriodontol. 1992;19:12–4.
10. Suprabha BS, Baliga M. Total oral rehabilitation in a patient with portwine stains. J Indian SocPedodPrev Dent. 2005;23:99–102.
11. Gnanasekhar, Al-Duwairi Electrosurgery in dentistry. Quintessence Int. 1998;29:649–54.