

Encephalotrigeminal Hemangiomas: Two Case Reports

Authors

Dr. Muthu Kumar.S¹, Dr. Mohsin Khan², Dr. Mubeen³. Dr. Vijayalakshmi. K.R⁴

^{1,3,4} Government Dental College and Research Institute Bangalore – India

²Maruthi Dental College

Email: *muthukumardoc@gmail.com, mubeendr@yahoo.com, drvijayalakshmikr@yahoo.co.in*

Corresponding Author

Dr. Muthu Kumar. S

Government Dental College and Research Institute Bangalore – India

Email: *muthukumardoc@gmail.com*

ABSTRACT:-

Encephalotrigeminal Hemangiomas also known as sturge- weber syndrome is specifically congenital, non-hereditary, rare condition occurring with a frequency of 1:50,000 live births, with equal predilection for either sex. This syndrome consists of a constellation of clinical features like cutaneous angiomas (Port-wine stain), glaucoma and variable neurological manifestation including seizures with or without mental retardation. Two male patients aged 17 and 29 year old reported to our department at different time interval with a complaint of swelling in the upper lip since 4 and 2 years respectively. Similar features of reddish discoloration noted in right side of face since childhood was reported by both the patients; there was no history of trauma or pain. However there was positive history of seizure in one of the patient. This paper reports two cases of this syndrome in detail with classic oral manifestations, radiological features and management.

Conclusion:- *Dentists play key role in identifying oral manifestations and aid in early recognition of this syndrome and prevent cranial complications thereby improving the prognosis.*

Keywords:- *Encephalotrigeminal Hemangiomas, Sturge- Weber syndrome (SWS), Port wine stains, angioma.*

INTRODUCTION

Encephalotrigeminal Hemangiomas is also known as Sturge - Weber syndrome & mother spot disease. It is a rare neurocutaneous syndrome that occurs with a frequency of approximately 1 per 50,000 and is characterized by constellation of clinical features like cutaneous angiomas (Nevus flammeus), glaucoma and neurological manifestation including epilepsy, mental retardation and focal neurological deficits. Oral changes occur in 40% cases of this syndrome which manifests as unilateral hemangiomas lesions of gingiva, lips, buccal, mucosa, palate, tongue, floor of the mouth resulting in malocclusion and facial asymmetry^{1,2,3,6}.

In depth knowledge of this syndrome will help a clinician in identifying early oral manifestation which is essential for the diagnosis and management, thereby preventing future cranial complications. The present paper reports two cases of sturge - weber syndrome with orofacial manifestations, role of imaging, diagnosis and its multispecialty approach in the management of this syndrome.

CASE REPORT

CASE 1:- A 29 year old male patient reported with a chief complaint of swelling in right upper lip to our department. Patient noticed swelling 2 years ago which was gradually progressing in size to the present size, medical history revealed that patient was under anticonvulsants for seizures since 10 years. On general physical examination, patient was moderately built and nourished with good orientation towards time, place and person.

No apparent abnormality was noticed on head & neck region.

Examination of face revealed a flat, uniform, diffuse dark reddish discoloration on right side of face suggestive of port wine stain, extended to the midline and up to angle of the mouth (Figure 1). However there was no surface changes evident over the skin.

Examination of eyes revealed dilated blood vessels w.r.to right lateral aspect of sclera in the right eye. However the movement of eyeball was normal, eyelashes & eyelid appeared normal. There was localized swelling on right side of upper lip extending to philtrum giving a fullness of upper right side of the lip. The swelling was uniformly soft in consistency, non-tender, compressible and pulsatile on palpation.

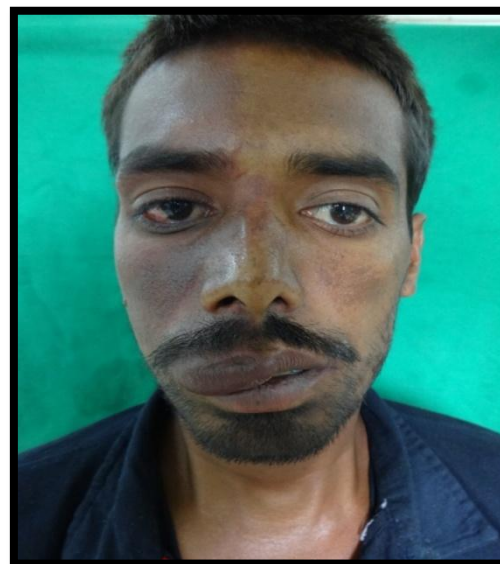


Figure 1:- Extra oral photograph showing port wine stain involving right side of face with dilated blood vessels in right eye and diffuse swelling in right side of upper lip.

On intra oral examination there was a diffuse gingival enlargement which appeared fiery red in

color involving marginal & attached gingiva w.r.to teeth numbers 11 to 18 and extending upto the vestibule (Figure 2) .However enlargement was more prominent in canine and premolar region with local deposits, physiological blackish pigmentation on tongue and on right side of palate there was diffuse ecchymosis extending to midpalatal region. Diascopy test was carried and was positive.



Figure 2:- Intra oral photograph showing gingival enlargement w.r.to right side of upper teeth.

Based on the history and clinical presentation a provisional diagnosis of Sturge - Weber syndrome was considered with differential diagnosis of Klippel-Trenaunay syndrome, Parkes weber syndrome, Rendu ossler weber syndrome.

Routine blood examination, OPG and lateral skull view were noncontributory towards the diagnosis. Ultrasound imaging showed venous stasis and chronic hypoperfusion of right side of face .Further MRI of the brain with contrast revealed enlarged right choroidal plexus measuring approximately 15x13mm showing significant post contrast enhancement suggestive of Angioma (Figure3) .

Based on the clinical feature, ultra sound & MRI findings a final diagnosis of Sturge -Weber

syndrome was established. Further patient was managed by Neurophysician with anticonvulsant drugs and surgical correction of lip and gingiva were carried out by vascular surgeon. Patient is under regular follow up.



Figure 3:- Contrast Enhanced MRI of brain indicating Angioma of right choroidal plexus

CASE 2

Another patient with identical features reported to our department with swelling in right side of upper lip since 4 years which was progressively increased to the present size. However the medical history was not significant.

Head and neck examination revealed no gross abnormality except for the uniform dark reddish discoloration in the right upper cervical 3rd of neck .Examination of face revealed port wine stain in right side of face not crossing the midline (Figure 4).

Examination of eyes revealed dilated blood vessels in sclera of right eye with normal movement of eyeballs; eyelashes & eyelid appeared normal. There was localized swelling on right side of upper lip extending to philtrum giving fullness to upper right side of the lip . The

swelling was uniformly soft in consistency, non-tender, compressible and pulsatile on palpation.



Figure 4:- Front view of patient showing Port wine stain involving right side of face with dilated blood vessels in right eye and diffuse swelling in right side of upper lip.

Routine examination of oral cavity revealed a uniform, diffuse gingival enlargement appeared fiery red in color involving attached gingiva w.r.to teeth numbers 11 to 18 and extending up to the vestibule & diffuse ecchymosis on right side of palate was noted extending to midpalate (Figure 5) & discoloration of 11 tooth was evident. Diascopy test was positive.



Figure 5:- Mirror image of the diffuse ecchymosis on right side of palate was extending to mid palate.

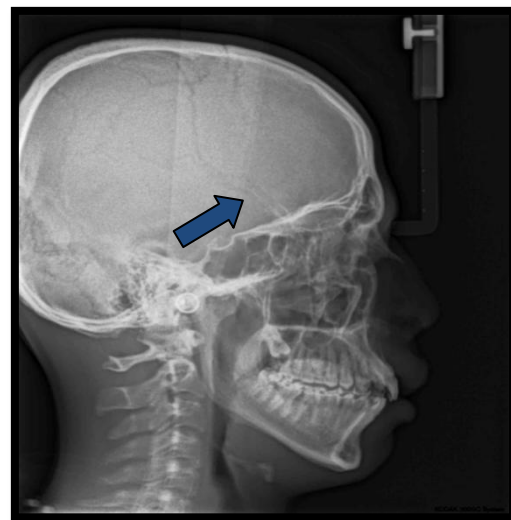


Figure 6:- Lateral Skull view showing “tram line calcification”.

Based on the history and clinical presentation a provisional diagnosis of venous haemangioma involving right side of face was considered with differential diagnosis of Sturge-Weber syndrome, Klippel-Trenaunay syndrome, Parkes weber syndrome.

Routine blood investigations were under normal limits & there was no significant findings in the OPG, lateral skull view revealed "tram line" calcifications (Figure 6). The patient was further advised ultra sound and MRI investigations.

DISCUSSION

Encephalotrigeminal Hemangiomatosis is a rare neurocutaneous syndrome that occurs with equal frequency in both sexes, in our cases both the patients were male⁶.

ETIOLOGY

The exact etiology remains unknown, however it has been postulated that angiomas of this syndrome 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 formation of angiomas of leptomeninges and face.

The facial cutaneous capillary venous angiomas (Port-wine stains) are usually the first component of the syndrome to be observed since birth and are confined almost exclusively to the skin that are supplied by the trigeminal nerve, which may range from small red macules to large red patches^{4,6}. They occur more commonly on right side and do not extend over midline. Another feature noted with this syndrome is seizure. Similar such feature was noted in our both cases except for seizure in our 2nd reported case.

It was coincidence that identical findings was evident in right side of eye, lip, gingiva and palate in both the cases which are classical features of Sturge -Weber syndrome.

Based on ES Roach classification of Sturge weber syndrome (SWS) as shown in table 1(2,4). Our 1st reported case was grouped under Type 1 SWS & 2nd reported case was grouped under Type 2 SWS. Differential diagnosis what we considered were

Klippel Trenaunay syndrome, Parkes -Weber syndrome & Rendu ossler weber syndrome which was ruled out due to the absence of other features as shown in table 2⁴.

The radiographic findings shows "tram track, tram line" calcification in lateral skull radiograph(5,7). The MRI with contrast showed a high diagnostic accuracy in our case which has given the details of angioma of the right choroidal plexus. Management of this syndrome requires multidisciplinary team approach. Though there are different modalities which include medical, embolo therapy, sclerotherapy, laser, cryotherapy and surgical therapy, treatment should be individualized depending on tumor location, accessibility, depth of invasion, patient age and cosmetic consideration.

Oral physicians play a key role in early recognition of classic features in the maxillofacial region and the radiographic changes in skull of patients with Struge-Weber syndrome. Early diagnosis is crucial considering the fact that future complications especially those related to CNS can cause further morbidity to these patients.

Table 1 :-showing classification of sturge weber syndrome Based on ES Roach classification .

Type I (classic) SWS	Type II SWS	Type III SWS (forme fruste)
Most common form with Facial and leptomeningeal angiomas, Seizures, and ocular involvement.	Facial angioma and ocular involvement, but with no evidence of intracranial disease.	This type involves only leptomeningeal angioma, with no facial nevus and usually no ocular manifestation of glaucoma

Table 2 :- Showing the differential diagnosis of Sturge - Weber syndrome.

Klippel-Trenaunay syndrome	Parkes -Weber syndrome	Rendu-ossler weber syndrome
1. Overgrowth of limbs 2. varicose veins Intra orally 3. Deep palatal vault, early teeth eruption and anterior open bite. 4. The involvement of head and neck region is only about 5%.	1. Fistulas 2. Skeletal or soft tissue hypertrophy affecting multiple limbs, trunk, head & most commonly affects the lower extremities. 3. The incidence rate is only 0.3% worldwide.	1. Multiple small aneurysmal telangiectases of the skin and mucosa which is commonly associated with bleeding. 2. Progresses as age advances.

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