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Research Article

# STURGE-WEBER ANGIOMATOSIS: A CASE REPORT

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**A R T I C L E I N F O**

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**A B S T R A C T**

Sturge weber syndrome or encephalotrigeminal angiomatosis which is a rare sporadic, nonhereditary, neurocutaneous condition affecting other multiple system of body which includes CNS, ophthalmic, oral, endocrine, vascular, ENT. oral manifestation is seen in 40% cases. In this case 12/F patient presented with port wine stain involving left side of face involving maxillary division of trigeminal nerve, gingival over growth involving left maxilla and cone beam computed tomography suggestive hypertrophied lining of maxillary sinus associate with Osteohypertrophy of left maxilla. Surgical excision of lesion was done under GA & histopathology was suggestive of capillary haemangioma. In this paper a special emphasis is placed on other manifestation of Sturge weber syndrome and their management along with oral manifestation.



## INTRODUCTION

Sturge-weber syndrome is also known as Encephalotrigeminal Angiomatosis or Sturge-Weber Angiomatosis. Schirmer was first to describe the symptoms of this syndrome. The name Sturge weber is given by Professor Hilding Bergstrand in 1935 for appreciation of the work of William Allen Sturge who noticed the neurologic involvement in SWS In 1879 & Parkes Weber who described radiologic features in 1992[1].Frequency of occurrence of SWS is 1:20000 to 1:50000.with no sex predilection[1].SWS belongs phakomatoses group of syndrome which involves multiple system of body like CNS, Opthalmic,Vascular,endocrine,ENT [1,4,6].

### *Case Report*

12-year-old female patient was presented with complaint of painless swelling with respect to left palatal region enclosing gingiva of 25 to 28 region and frequent history of bleeding gums.Medical history patient revels no history of seizure, headache with no relevant past dental history or family history. General examination reveals normal growth, behaviour & speech.

Extraoral examination revealed presence of Port wine stain (Fig-1) involving left side of face predominantly along the maxillary division of trigeminal nerve. Intraoral examination showing large reddish growth with respect to buccal gingiva enclosing 26,27 and palatal gingiva with respect to 23 to 27 regions,28 was missing (Fig-2).

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Lesion blanched on palpation and having high bleeding tendency even on minor trauma with periodontal probe & presence osteo-Hypertrophy of left maxilla.

### *Investigation*

OPG, Lateral Cephalogram, CBCT & other hematologic investigation like complete blood count, bleeding time, clotting time Was done.

Lateral cephalogram is normal with no evidence of any calcification of leptomeninges. CBCT showing involvement of maxillary sinus with hypertrophied lining of maxillary sinus and Osteohypertrophy maxilla from 23 to 28. Showing large spacing of approximately 8 mm between 1st and 2nd molar and impacted 28. (Fig-3)

Blood investigation suggestive of anaemia with haemoglobin 7.2gm/dl.Patient was referred to department of haematology for management and patient again operated when haemoglobin concentration raised to 12gm/dl.

### *Management*

Patient was taken into OT, oral intubation was done and surgical excision of growth was done and extraction of 26,27,28 was done. greater palatine artery was found to be feeder vessel of lesion which was ligated and cauterized. Complete homeostasis was achieved. Blood loss during procedure was 800ml.1unit of blood transfusion was given postoperatively (Fig-4).

### *Histopathology*

As shown in (Fig-5) H & E stained section shows Parakeratinized stratified squamous epithelium showing

varying degree of proliferation. Underlying connective tissue shows lobulated growth separated by fibrillar collagenous tissue. Lobules contains numerous small capillaries and dilated blood vessels lined by flattened endothelial cells. Stromal tissue is moderately cellular containing spindle cell, fibroblast, chronic inflammatory cell infiltration and over all feature is suggestive of capillary haemangioma.

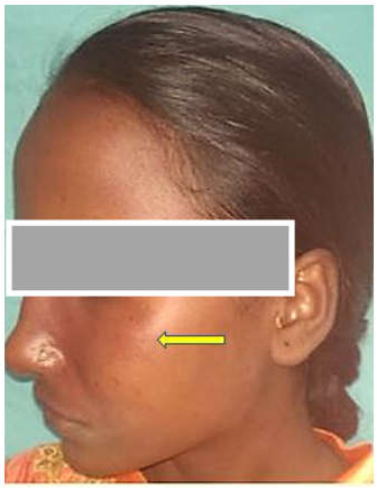

**Fig 1** Presence of Port wine stain on left side of face in distribution of maxillary nerve.

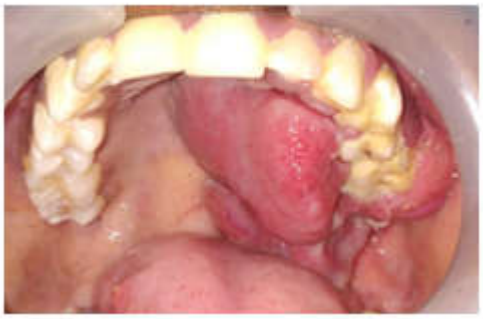

**Fig 2** Intra oral view of lesion involving left maxilla.

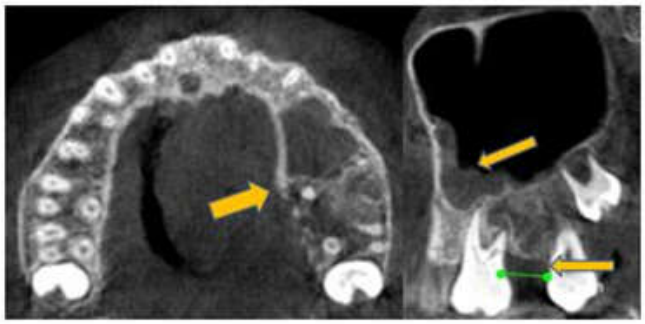

**Fig 3** Hypertrophied lining of maxillary sinus and Osteohypertrophy maxilla from 23 to 28.

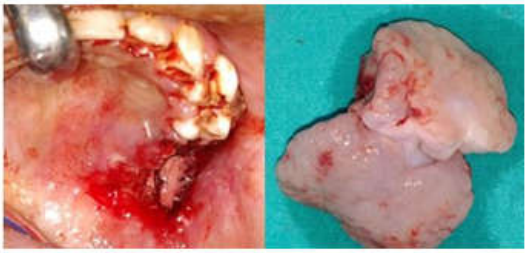

**Fig 4** Post-operative view of after excision of lesion.

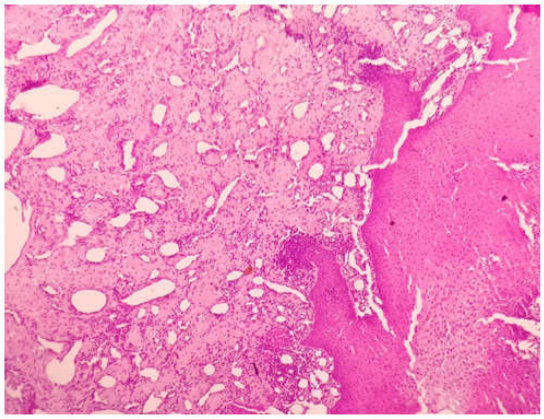

**Fig 5** Histopathology of the lesion

## DISCUSSION

SWS is rare disease from phakomatoses spectrum mainly affects neuro-oculo-cutaneous system of body [1]. Other disease includes in differential diagnosis is Von Hippel-Lindau disease (haemangioma of cerebellum, retina of eye, visceral cyst), Rendu-Osler-Weber syndrome (recurrent epistaxis, telangiectasia, family history, recurrent GI bleeding), Maffucci's syndrome (enchondromas, cavernous haemangiomas and lymphangiomas), angioosteodystrophy syndrome, Trenaunay-Weber syndrome (hypertrophy of bone and soft tissue associated with capillary malformation [4,2]

***Pathogenesis***

1. SWS is developmental anomaly affecting ectodermal and endodermal structure of face. Cephalic portion of neural tube will form structure of brain and face. vascular plexus around cephalic portion of neural tube normally form at 6week of intrauterine life and regression start at 9$^{th}$ week.Persistence vascular plexus leads to angioma of face, eye and leptomeninges [9,1,8].
2. Genetic basis for formation of SWS includes mutation of RASA1 gene, GNAQ gene both genes are responsible for normal function of vascular structure [1,5].

**Clinical feature & management of SWS**
**Port wine stain (PWS)**

SWS generally associated with port wine stain Which is unilateral in nature along the distribution of trigeminal nerve [1,3]. Maxillary division is most commonly affected by PWS [1,7] .In our case there was involvement of both V1, V2(predominantly) division.

Port wine stain negatively affect the psychology of patient. Management of port wine stain includes laser therapy, dermabrasion, tattooing [4,8]. Early treatment of PWS is advisable for better result [5]

***Oral Manifestation***

***Intraoral changes occur in 40% cases if SWS***

***Soft tissue Oral manifestation includes [4]***

1. Unilateral or bilateral Erythematous mucosa on side of port wine stain.
2. Haemangioma of gingiva of involved side
3. Pyogenic granuloma

***Hard Tissue Manifestation Includes [4]***

1. Osteohypertrophy involved side [4].in this case CBCT shows the Osteohypertrophy of left maxilla. fig [3]
2. periodontitis management includes meticulous maintenance of oral hygiene, surgical excision of lesion under Local or general anaesthesia using scalpel or Nd: Yag laser based on the size of lesion. Laser is preferable as it cause less trauma to surrounding structure and easy to achieve haemostasis [4,8]

***Ocular Manifestation***

Involvement of V1 division associated more prone to neurologic and ophthalmic problems (78%), half of the cases SWS shows ocular involvement which includes

1. Episcleral vasodilation in 50% of cases [6]
2. Glaucoma (30-70%) -most commonly open angle glaucoma occurs trimodal distribution,40% cases are affected by congenital glaucoma in 1st year of life ,28% cases in childhood,20% cases in 3rd decade of life [1,6]
3. Choroidal haemangioma occurs in (20%–70%) [6]
4. Retinal abnormalities -detachment, haemorrhage [1,6,9]
5. Buphthalmos [9]

Ocular manifestation can be prevented by regular check up and medical or surgical management based on age of patient [6].Choroidal haemangioma can be treated by photodynamic therapy, intravenous bevacizumab which prevents the growth of blood vessels [9].

***CNS***

1. SWS associated with unilateral intracranial angiomatosis [1]
2. "tramline" or gyriform calcifications in temporal and occipital lobe can be seen in CT, MRI, Lateral cephalogram [4]
3. Epileptic convulsion, migraine, stroke like episode [1]
4. Mental retardation

Mainstay for management of convulsion is anticonvulsant drug but refractory to anticonvulsant drug may require surgery like lesionectomy, callosotomy and hemispherectomy [1].In cases where surgery is contraindicated and associated modified Atkins's diet reduces episode of convulsion [1]. Aspirin in low dose (3-5mg/kg/day) showing reduction in frequency of convulsion, headache, stroke like episode [1,5].

***Endocrine***

Endocrine problems most commonly associated with SWS is deficiency of growth hormones (18fold increase in incidence) and hypothyroidism (500-10000 times prevalent) [1]. Patient of SWS needs regular medical follow up and management.

***ENT***

Higher incidence of ENT problems is associated with SWS which mainly includes nasal bleeding upper respiratory tract infection, sinus infection and problems, sleep apnea secondary to tissue hypertrophy [10]. Through management of ear and sinus infection also positively affect the CNS symptoms of patient [10]. Based on clinical feature of patient roach scale classified the SWS into 3 type as [1,8,9]

| Type | facial angiomas | leptomeningeal angiomas | glaucoma |
|---|---|---|---|
| 1 | ✓ | ✓ | ✓ |
| 2 | ✓ | Not associated | May or may not |
| 3 | Not associated | ✓ | May or may not |

This case is type 2 SWS associated with only facial angioma.

## CONCLUSION

SWS is very rare disease with enigmatic clinical feature affecting various system of body. Patient of SWS must me managed with multidisciplinary team approach and also requires psychological counselling.Management of intraoral lesion may be difficult due to haemangiomatous nature of lesion and can leads to excessive bleeding. precautionary measures must be taken to avoid inadvertent compilations before surgical excision.

**Conflict of Interest:** None

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