Sturge-Weber Syndrome: A Case Report
Neha Khambete, Mukund Risbud, Avinash Kshar

Abstract
The Sturge-Weber syndrome (SWS) or Encephalotrigeminal Angiomatosis is specifically congenital, non-hereditary, rare condition of unknown etiology. The classic pathognomonic features of disease include angioma of the leptomeninges extending to cerebral cortex with ipsilateral angiomatous lesions, unilateral facial nevus after one division of trigeminal nerve and epileptic convulsions. The most characteristic oral manifestation is represented by gingival hemangiomatous lesion usually restricted to ipsilateral maxilla or mandible. A classic case of Sturge-Weber syndrome is reported here.

Key Words: Sturge-Weber Syndrome; Port-Wine Stain; Tram Line Calcifications.

Introduction
Sturge-Weber syndrome is a sporadic neurocutaneous disease characterized by facial port-wine stain, ocular abnormalities (glaucoma and choroidal hemangioma) and leptomeningeal angiomatosis. (1) Sturge Weber syndrome (SWS) was first described by Schirmer in 1860 and later more specifically by Sturge in 1879, who associated dermatological and ophthalmic changes of the disease to neurologic manifestations. Weber in 1929 complemented it with the documentation of radiologic alterations seen in these patients. (2)

It is rare disorder occurring with a frequency of 1:50,000 live births. (3) Both sexes are affected equally and no racial predilection is seen. (4) The classic feature of this disorder is the angioma of leptomeninges. Most common features are epilepsy, Port-wine stain and dermal angiomas, abnormal findings in skull radiographs, mental retardation, ocular involvement and hemiplegia. (5) Oral manifestations of the disease may vary considerably and changes in morphology and histology of gingiva, periodontium and pulp have been reported. However the most common feature is a gingival hemangiomatous lesion usually restricted to ipsilateral maxilla, mandible, floor of mouth, lips, cheeks, palate and tongue. (1) In this report, we present a case of SWS with its characteristic manifestations.

Case Report
An 11 years old male patient reported to Dept. of Oral Medicine & Radiology, Vasantdada Patil Dental College & Hospital, India with chief complaint of bleeding from gums since last 2 months. Past medical history revealed that patient had developed convulsive disorder at the age of 6 months for which he was under medication. His parents also gave history of delayed developmental milestones and learning disabilities. Patient was diagnosed with moderate mental subnormality with I.Q. of 52. Extra oral examination showed presence of port-wine stain on right side of face along the ophthalmic division of trigeminal nerve. Examination of eyes showed presence of dilated blood vessels in right eye (Fig 1).

Figure 1 Extra oral photograph showing port-wine stain along ophthalmic branch of trigeminal nerve, dilation of ocular blood vessels in the right eye

Intraoral examination showed erythematous area on attached gingiva in maxillary anterior region. The lesion showed positive diascopy (Fig 2).

Figure 2 A. Angiomatous lesion on gingiva in maxillary anterior region, B and C: Lesion showing positive diascopy.

Figure 3 A. Lateral cephalogram and B. cropped PA skull view showing “tram-line” calcifications.
Patient had poor oral hygiene and malocclusion. On radiographic examination, tram-line calcifications were seen on lateral cephalogram in the posterior parietal lobe and PA skull (Fig. 3). Blood investigations were normal. Based on history, clinical and radiological findings a diagnosis of Sturge-Weber Syndrome was made. A thorough plaque control regimen was started. It included oral prophylaxis, use of chlorhexidine mouth rinses, oral hygiene instructions. Gingival bleeding on chewing food and exposure to sunlight was stopped completely after giving plaque control therapy. It was decided to continue with same plaque control regimen and monthly follow-up was done. The patient and his parents were referred to counselor for psychological counseling.

Discussion

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 is used for classification, as follows(1):
Type I - Both facial and leptomeningeal angiomas; may have glaucoma
Type II - Facial angioma alone (no CNS involvement); may have glaucoma
Type III - Isolated leptomeningeal angioma; usually no glaucoma.

According to the above criteria, our case is complete Type I SWS case. Characteristically leptomeningeal angiomas occur as unilateral lesions affecting the pia arachnoid membrane over the posterior temporal, parietal and occipital areas.(2) It is typically a static lesion but review of literature also reveals some progressive lesions.(2) It commonly shows abnormal blood flow pattern as venous occlusion, thrombosis, vasomotor phenomenon and vascular steal phenomenon resulting in cortical ischemia. This in turn gives rise to epileptic convulsive crisis, transient hemiparesis, glossitis and progressive deposition of calcium salts. These calcifications produce a characteristic double contoured “tram-line” appearance following the convolutions of cerebral cortex. Brushfield and Wyatt stated that these tram-line calcifications are pathognomonic of SWS.(6) These calcifications appear after the patient reaches 2 years of age and remain stationary after second decade of life. These calcifications are gyriform and curvilinear and most commonly seen in parietal and occipital lobes as seen in our case. These are seen best in the lateral skull view with affected side closer to the film. CT scan shows calcifications in the areas of atrophy. (7)

The most evident clinical manifestation is presence of nevus flammeus or Port-wine stains on the face within the distribution of Trigeminal nerve especially the ophthalmic division. They are present since birth and may range from small red macules to large red patches which blanch on pressure.(4) A large variation has been reported in their pattern of occurrence. They occur more commonly on right side and do not extend over midline.(4) They can be bilateral or completely absent or may extend to neck, limb and other parts of body.(6) However only carriers of port-wine stains along the ophthalmic branch develop the syndrome in its classical form as presented in our case.(5)

The oral manifestations include Port-wine stain lesion of oral mucosa along with the hypervascular changes. Most common manifestation is angiomatous lesion of gingiva which can vary from slight vascular hyperplasia to massive hemangiomaticus proliferation. It is characterized by increase in the vascular component and gingival hemorrhage at minimal traumatisms. The oral manifestations are generally unilateral and finish abruptly in the midline. Macroglossia and maxillary bone hypertrophy found in some patients can cause malocclusion and facial asymmetry. The gingival hyperplasia in these patients could be secondary to anticonvulsant therapy further complicated by poor oral hygiene secondary to mental retardation.(1, 3, 8) Table 1 illustrates the classic manifestations of SWS and manifestations present in our case.

<table>
<thead>
<tr>
<th>N.o.</th>
<th>Clinical Manifestation</th>
<th>Incidence (%)</th>
<th>Present Case</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Epilepsy</td>
<td>80</td>
<td>+</td>
</tr>
<tr>
<td>2.</td>
<td>Port-Wine Stain</td>
<td>76</td>
<td>+</td>
</tr>
<tr>
<td>3.</td>
<td>Abnormal Radiographic</td>
<td>63</td>
<td>+</td>
</tr>
<tr>
<td></td>
<td>Findings</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4.</td>
<td>Mental Retardation</td>
<td>54</td>
<td>+</td>
</tr>
<tr>
<td>5.</td>
<td>Oral Manifestations</td>
<td>38</td>
<td>+</td>
</tr>
<tr>
<td>6.</td>
<td>Hemiparesis</td>
<td>37</td>
<td>-</td>
</tr>
<tr>
<td>7.</td>
<td>Ocular Manifestations</td>
<td>37</td>
<td>+</td>
</tr>
</tbody>
</table>

Table 1 Clinical manifestations of SWS and manifestations seen in our case.

To conclude, the large spectrum of clinical manifestations of Sturge-Weber syndrome shows its multifactorial nature and difficulty in diagnosis. As the exact etiopathogenesis is not known, its prevention is difficult and its early diagnosis is critical, since it allows the control of future complications, mainly those relating to the Central Nervous
System, considering the inexistence of specific treatments for such pathology.

Authors Affiliations: 1. Dr. Neha Khambete B.D.S, Post graduate Student, 2. Dr. Mukund Risbud M.D.S, Professor and H.O.D, 3. Dr. Avinash Kshar M.D.S, Associate Professor, Department of Oral Medicine & Radiology, Vasantdada Patil Dental College & Hospital, Maharashtra, India.

References

Address for Correspondence
Dr. Mukund Risbud M.D.S, Professor and H.O.D, Department of Oral Medicine and Radiology, Vasantdada Patil Dental College and Hospital, A/P Kavalapur, Tal: Miraj, Dist: Sangli, Maharashtra, India.
Email: mukundrisbud@gmail.com

Source of Support: Nil, Conflict of Interest: None Declared