Case Report

Sturge Weber Syndrome- A Rare Congenital Neuro-cutaneous Disease.

Wahida Sultana[a], Sufia Nasrin Rita[b], *S. M. Anwar Sadat[c],

a. Resident, Dept. of Oral & Maxillofacial Surgery, Dhaka Dental College Hospital, Dhaka
b. Associate Professor and Head, Dept of Orthodontics, Sapporo Dental College Hospital, Dhaka
c. Lecturer, Dept. of Oral & Maxillofacial Surgery, Dhaka Dental College, Dhaka

ARTICLE INFO

Article history:
Received : 23.08.15
Accepted : 30.09.15

Key words:
Sturge Weber syndrome, congenital, encephalotrigeminal angiomatosis, vascular lesions, oral and limb manifestations.

ABSTRACT

Sturge Weber Syndrome (SWS) or Encephalotrigeminal Angiomatosis is specifically a rare non hereditary congenital sporadic disorder of elusive etiology. It belongs to a group of disorders collectively known as the phakomatoses ("mother-spot" diseases). It has a vast continuum of cutaneous, neurologic, ophthalmic and oral manifestations. It consists of congenital hamartomatous malformations that may affect the eye, skin, and central nervous system (CNS) at different times, characterized by the combination of venous angiomas of leptomeninges, face, jaws and oral soft tissues. 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. We report a case of Sturge Weber Syndrome with its characteristic oral manifestations and review of relevant prevailing literature.

Introduction

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. Sturge-Weber syndrome is a sporadic neurocutaneous disease characterized by facial port-wine stain, ocular abnormalities (glaucoma and choroidal hemangioma) and leptomeningeal angioma [1,2].

SWS is rare disorder occurring with a frequency of 1:50,000 live births[3]. It is believed to be caused by the persistence of

*Address of Correspondence:
Dr. S. M. Anwar Sadat
BDS, BCS, MCPS, FCPS, MS (OMS)
Lecturer, Dept. of Oral & Maxillofacial Surgery
Dhaka Dental College, Dhaka, Bangladesh.
Contact No. +880 1711156023
E-mail: an_sadat@yahoo.com

vascular plexus around the cephalic portion of the neural tube. This plexus develops during the sixth week of I.U. development but normally undergoes regression during ninth week[4]. Both sexes are affected equally and no racial predilection is seen[5]. The classic feature of this disorder is the angioma of leptomeninges. Most common features are 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[2].

The cutaneous angiomas are called port wine stains, which usually occur unilaterally along dermatomes supplied by the ophthalmic and maxillary division of trigeminal nerve. It may

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A 12 years old Bangladeshi girl reported to us with a complaint of red pigmentation on her left face and oral cavity. According to her mother's statement, it appeared at birth and gradually increased with age and increased the intensity of color with age from light pink to deep purplish red. There were no complaints of burning sensation, pain, numbness or paraesthesia in the pigmented or surrounding areas except occasional mild bleeding from gum in different sites of oral cavity. There were no systemic symptoms but had a history of convulsion during fever 3-4 times during her childhood. During convulsion patient was treated by oral diazepam according to paediatrician's advice. Rapid sponging of the whole body was practiced during fever to preven rise of body temperature and thus prevented the convulsion. Patient's mother gave history of normal delivery in house without any complications; there was no history of any drug intake or trauma during pregnancy. No other family members have this entity.

epilepsy, Port-wine stain and dermal angiomas, abnormal findings in skull radiographs, mental retardation, ocular involvement and hemiplegia[6]. Oral manifestations of the disease may vary considerably and changes in morphology and histology of gingiva, periodontium and pulp

be bilateral or totally absent or may extend to neck, limbs and other parts of the body[7]. Intraorally angiomatosis may involve lips, buccal mucosa, palate, gingiva, and floor of mouth. This syndrome is of rare occurrence and management becomes complicated due to risk of hemorrhage. In this report, we present a case of SWS with its characteristic manifestations.

Examination of patient showed macular lesions on left side of the face along the distribution of ophthalmic, maxillary and mandibular division of the trigeminal nerve and also on bilateral hands and legs. The distribution of pigmentation varied in different sites of face; it did not cross the midline in upper lip skin, mucosa, hard and soft palate but crossed midline to some extent in nose, forehead, lower lip skin, mucosa and chin. The distribution of pigmentation is haphazard in extremities bilaterally. X-rays and other investigations revealed normal findings. The case was discussed in an expert board where it was diagnosed as Sturge Weber Syndrome. As there was no complications, it was decided to give follow up of the patient to see the progression and any complications.

Discussion

Sturge Weber Syndrome which is also known as Sturge Weber Disease, Leptomeningofacial Angiomatosis is characterized by facial port wine stains, ocular abnormalities and leptomeningial angiomas. The most

characteristic clinical appearance is that of nevus flameus or port-wine stains on the face along the distribution of Trigeminal nerve, of which ophthalmic division is the most commonly affected. Lesions vary in color and size ranging from pink, red and purple small to large macular lesions and do not cross the midline. However, few cases of bilateral involvement have also been reported[8].

Our patient showed macular lesions on left side of the face along the distribution of ophthalmic, maxillary and mandibular division of the trigeminal nerve and also on the corresponding hand and leg. Patients affected with this syndrome have also been reported to have mental retardation, delayed development, seizures, headache and hemiparesis. Ocular involvement commonly manifests as glaucoma and buphthalmos[9].

Sturge Weber Syndrome is a constellation of numerous signs and symptoms and an affected person may not display all the features. 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[10] is used for classification -

Type I- Both facial and leptomeningeal angiomas; may have glaucoma

Type II – Facial angiomas alone; may have glaucoma

Type III – Isolated leptomeningeal angiomas; usually no glaucoma

According to the above criteria, our case is Type II SWS case.

The oral manifestations include ipsilateral port-wine stains of oral mucosa along with the hypervascular changes which finish abruptly in the midline of the palate. Angiomatous lesion of gingiva may be present which can vary from slight vascular hyperplasia to colossal hemangiomatous proliferation. Gingival hyperplasia can also be attributed to anticonvulsant medication and secondary to poor oral hygiene in mentally retarded patients. Macroglossia and maxillary bone hypertrophy have also been reported in a few cases[8].

Tram track calcification caused by calcification in apposing gyri, ipsilateral calvarial thickening and enlargement of the paranasal sinuses and mastoid may be visible in skull films. Higher imaging modalities like CT for calcification and MRI for brain assessment can also be used. MRI is the current gold standard for diagnosis of this disease especially in infants[9].

Differential diagnosis includes Rendu-Oslar-Weber syndrome, angio-osteodystrophy syndrome, Maffuci's syndrome, Von Hippel Lindau disease, and Klippel Trenaumy-Weber syndrome[7,11]. Diagnosis is made only on the basis of clinical and imaging features. Our patient exhibited characteristic clinical manifestations of Sturge Weber Syndrome[12].

Treatment and prognosis of SWS depends on the extent of involvement. Port wine stain can cause severe psychological trauma to the patients and hamper their personality development. Port wine stains can be improved by dermabrasion, tattooing and flash lamp pulse dyed lasers. Various treatment modalities like sclerotherapy, cryotherapy, laser and surgical excision have been tried with varying degrees of success to surmount intraoral lesions[13]. 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[2,3,14].

Dental management should be mostly stressed on preventive measures. Patients should be

educated, motivated and complied to follow a strict oral hygiene regimen to prevent dental caries and secondary gingival inflammatory enlargement. These patients can undergo endodontic procedure but over instrumentation should be avoided. Gingival hyperplasia has been reported to be successfully managed with carbon dioxide laser surgery with minimal hemorrhage[15]. To prevent reoccurrence maintenance of good oral hygiene is very important even after gingivectomy.

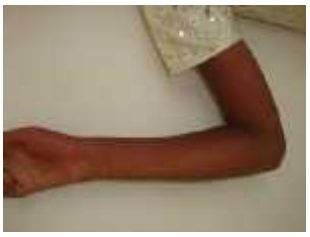
It is a challenging task to carry out dental procedures in a SWS patient due to risk of severe intra- and postoperative haemorrhage. Special precautions to prevent and treat complications may include hospitalization, application of local anaesthetics with vasoconstrictors, dressings, splints[13].

Conclusion

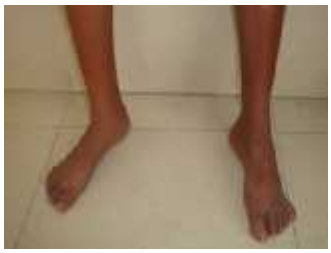
The large spectrum of clinical manifestations of Sturge-Weber syndrome shows its multifactorial nature and difficulty in diagnosis. As the exact etiopathogenesis is not known, its prevention is difficult and its early diagnosis is critical, since it allows the control of future complications, mainly those relating to the Central Nervous System, considering the inexistence of specific treatments for such pathology. Patients affected with SWS may or may not exhibit intraoral manifestations. It is imperative for oral health care practitoners to have profound knowledge of this rare congenital disorder and exhibit superfluous vigilance during routine dental procedures to prevent untoward life threatening complications.

Images:


Photograph 1: Extra oral Photograph of the patient shows macular area of deep purplish red pigmentation on left side of the face and enlargement of the lower lip. **Photograph 2:** Intra orally red pigmentation involves left half of hard and soft palatal mucosa with sharp mid line demarcation.




Photograph 3: Red pigmentation in right arm and half of right palm.
Photograph 4: Red pigmentation in left palm and fingers



Photograph 5: Red pigmentation involves bilateral leg skin

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