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Case Report

Sturge–Weber syndrome – A case report

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ABSTRACT

Sturge–Weber syndrome (SWS), also called as encephalotrigeminal angiomatosis is an uncommon congenital neurological disorder & frequent among the neurocutaneous syndromes specifically with vascular predominance. This disorder is characterized by facial capillary malformation & other neurological condition. The oral manifestations are gingival hemangiomatosis restricting to either side in upper and lower jaw, sometimes bilateral. We report a case of SWS with oral, ocular and neurological features.

1. Introduction

Sturge–Weber syndrome (SWS), also called as encephalotrigeminal angiomatosis, is a rare non-hereditary condition characterized by a facial cutaneous vascular nevus (nevus flammeus or port-wine stain) in association with leptomeningeal angiomatosis.[1] Schirmer was first one to describe it nearly a century ago (1860) in association with facial angioma and buphthalmos. Later in 1879 while reporting a case of 6½ year old girl, William Allen Sturge acclaimed these features in accordance with neurological findings and termed it as Sturge–Weber syndrome.[2] There is a risk of 10–50 % for the involvement of brain if a child is born with Port-wine birthmark (PWB) on the forehead or the upper eyelid. When the upper and lower eyelids are involved in port-wine birthmark, the risk of developing glaucoma becomes as high as 50%.[3] The frequency of occurrence of SWS is between 1:20,000 and 1:50,000. Amongst these port-wine birthmark is the most common with the occurrence of 0.3% in live births.[2] In 1992, Roach categorized SWS variants into three types:

- Type I: individual has a facial PWS, leptomeningeal angioma, and may have glaucoma
- Type II: individual has a facial PWS, no leptomeningeal angioma, and may have glaucoma

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- Type III: individual has leptomeningeal angiomatosis, no facial PWS, and, rarely, glaucoma.[4]

The aim of this article is to report a case of Sturge–Weber syndrome and add to the existing literature.

2. Case report

An 11 year old boy reported to the Department of Pedodontics & Preventive Dentistry, I.T.S. Dental College, Hospital & Research Center, Greater Noida with the chief complaint of bleeding gums in upper and lower right posterior tooth region & dental caries affecting upper left posterior teeth. The patient's mother gave a history of pain due to multiple mobile teeth since 1–2 months.

Medical history of the patient revealed that there was no history of epileptic seizures, but the patient gave history of multiple attacks of migraine during day time. The natal history of the child was non-contributory as it was a normal full term delivery without any complications.

General examination revealed no developmental delay in motor & speech function without any evidence of cardiovascular & respiratory disease.

Extra oral examination revealed the presence of port-wine stain on right side of face along the upper eyelid & forehead. These stains were seen extending over the right side of neck, shoulder & also extending up to scalp but not crossing the midline (Fig. 1a). The ocular examination revealed presence of suprascleral hemangiomas, indicative of initial glaucoma (Fig. 1b).

Intraoral examination revealed inflamed & hypertrophied gingiva of the right upper and lower quadrants. 64 was carious & 53, 55, 84, 85 were mobile. An OPG, lateral ceph & PA view skull was advised to check for resorption of roots, osteohypertrophy & calcifications in frontal lobe. The vascular nevus extended up to the underlying oral mucosa and gingiva. Erythematous reddish pink patches were seen on the right side of the hard palate extending till the soft palate (Fig. 1c). He had poor oral hygiene with gross calculus in the posterior region (Fig. 1d).

Based on the present finding provisional diagnosis of Sturge–Weber syndrome was made & patient was advised to go for the ophthalmic examination & Magnetic resonance imaging.

MRI with contrast revealed non-visualization of right sigmoid sinus and proximal internal jugular vein with attenuated caliber of right transverse sinus. Prominent venous channels were noted in the left posterolateral aspect of craniovertebral junction and there was prominent right vein of Labbe (Fig. 2a). MRI showed very mild changes in leptomeninges not significant enough to report (Fig. 2b).

Ocular examination revealed presence of subscleral hemangiomas with right eye (Fig. 2c). After complete presurgical laboratory blood test, patient was advised a thorough plaque control regimen. It included oral prophylaxis, use of chlorhexidine mouth rinses and oral hygiene maintenance. Extraction of grade II mobile teeth 54, 64, 65, 84, 85 were

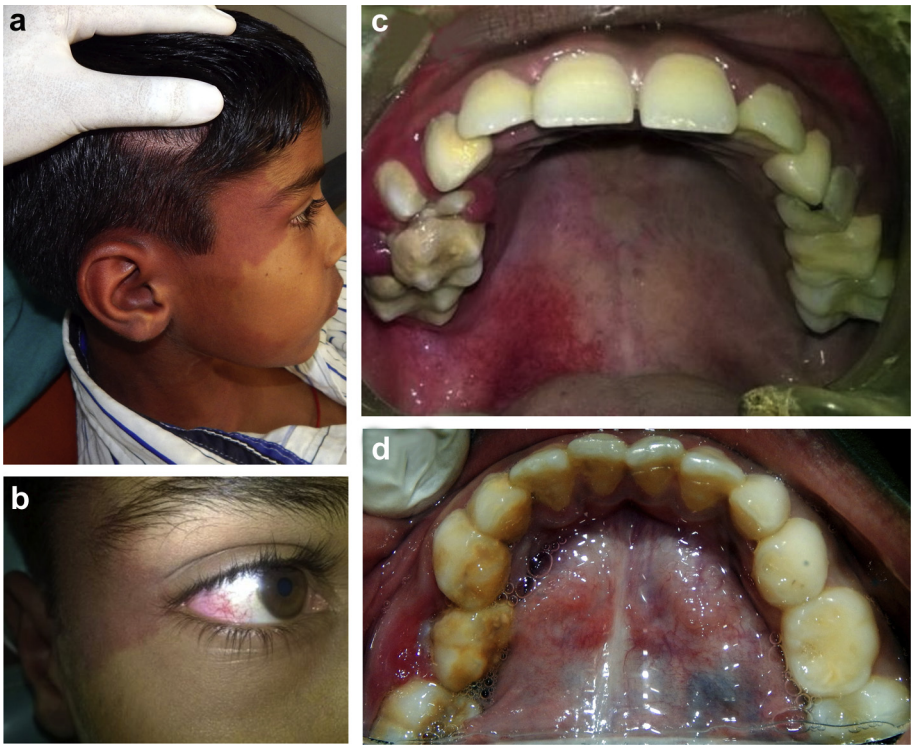

Fig. 1 – a: Port-wine stain on right side of face along the upper eyelid & forehead. These stains were seen extending over the right side of neck, shoulder & also extending up to scalp but not crossing the midline. b: Ocular examination revealing presence of suprascleral hemangiomas. c: Erythematous reddish pink patches were seen on the right side of the hard palate extending till the soft palate. d: Poor oral hygiene with gross calculus in the posterior region.