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Plausible predicament in emergency endodontic rehabilitation of a child with Sturge-Weber syndrome: A case report

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Highlights

- Sturge–Weber syndrome (SWS) is a developmental disorder portrayed by a triad of unilateral capillary malformation, leptomeningeal hemangioma, and secondary glaucoma.
- This case report observes SWS in a child with acute periapical abscess requiring emergency intervention and observation of typical port wine lesion intraorally.
- The patient had a full mouth rehabilitation preceded by emergency endodontic intervention.

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Abstract

Sturge–Weber Syndrome (SWS) is a neuro-oculo-cutaneous vascular disorder that includes leptomeningeal hemangioma and port wine stains, usually ipsilaterally, with ocular manifestations and extended lesions over the oral cavity. It is an embryonic developmental disorder affecting both mesodermal and ectodermal germ layers, associated with somatic mutation of gene GNAQ. It has a sporadic occurrence of 1:50,000 and no gender predilection. This case report added to the knowledge and understanding of systematic rehabilitation of a child patient with SWS and its associated risk factors in an emergency scenario. A 3.5-year-old girl child with a known diagnosis of SWS presented abscess over the right lower mandibular deciduous teeth and distinct oral features of SWS, requiring emergency endodontic intervention, followed by full mouth rehabilitation. Thus, an in-depth knowledge of the clinical state that helps in the expert assessment of dental interventions, including emotional co-regulation in those children, is essential.

Keywords: Angiomatosis; Endodontics; Port Wine Stain; Syndrome

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INTRODUCTION

Sturge–Weber syndrome (SWS) is a sporadic inherent developmental vascular disorder portrayed by leptomeningeal hemangioma; a facial port-wine nevus ipsilaterally involving ophthalmic trigeminal dermatome (V1) distributed over the trigeminal nerve region with ocular and oral manifestations.[1,2] It is also termed encephalotrigeminal angiomatosis, and these hemangiomas cause neurological variations, such as epilepsy, mental impediment, and hemiplegia.[3] Developmental and neurologic morbidity includes seizures, strokes, cerebral pains, hemianopsia, mental disturbances, and formative irregularities.[4] SWS has a sporadic occurrence of -1:50,000, unlike other phakomatoses.[1] It was first described by Schimer (1860) and later named by William Allen Sturge (1879) and Frederick Parkes Weber (1929).[5,6] The cortex of the brain becomes atrophic and calcified due to the deranged development of primordial blood vessels resulting in highly vascularized leptomeninges.[7] SWS has been associated with a somatic mutation in the gene guanine nucleotide-binding protein, G alpha subunit q (*GNAQ*) c.548G4A on the long arm of chromosome 9, phosphatidylinositol 3-kinase (*PI3K*), and activation of mitogen-activated protein kinase (*MAPK*). These somatic mutations are associated with phenotypes, such as McCune Albright syndrome. Hemangiomatous lesion may be observed intraorally involving maxilla/mandibular gingiva, tongue, lip, and palatal region. In gingiva, the clinical presentation may vary from demarcated unilateral hyperplasia extending till the midline to severe hemangiomatous malformation, which results in profuse bleeding following minor trauma.[7] The differential diagnosis includes Blue rubber bleb nevus syndrome Klippel–Trenaunay–Weber syndrome, PHACES (posterior fossa abnormalities, hemangiomas, arterial anomalies, cardiac, eye, and sternal anomalies), Wyburg–Mason syndrome, Rendu–Osler–Weber syndrome, angio–osteodystrophy syndrome, Maffuci's syndrome, and Von Hippel Lindau disease.[7,9]

CASE REPORT

A 3.5-year-old girl with her mother reported to the Outpatient Department of Pedodontics and Preventive Dentistry of University College of Medical Sciences and GTB hospital, Delhi , India with the chief complaint of pain and swelling in the right lower back tooth region for 5 days.

Medical history revealed that she developed tonic-clonic seizures at the age of 2.5 years and since then has been on medication with oral sodium valproate. Currently, no similar episodes of seizure occurred after the commencement of the anticonvulsant drug. No signs of mental retardation were observed, and the patient had an Intelligent Quotient score of < 81. The patient was communicative but highly apprehensive and uncooperative. History adduced a neglected child as reported by her mother.

Extra oral examination revealed the presence of port wine stains on the right half of the face, head, and neck in the front and back region and faded patches on the left hand. The patient has a larger head compared to a normal child. Ocular anomalies were ruled out by the Ophthalmology Department, and the absence of significant findings was confirmed. Asymmetrical facies with macrochelia were noted according to the enlargement on the right side (Fig. 1A–D).

Intraoral examination observed reddish discoloration of the right buccal mucosa, hard palate, the floor of the mouth, and dorsum and ventral surfaces of the tongue and gingiva. (Fig 2A, B). The mother confirmed the presence of the reddish discoloration since birth and, with time, resolved the discoloration of the hands, abdomen, and leg. The tongue was enlarged on the right side. Patients had deeply carious lower primary second molars on both sides. Gingival

abscess and swelling were associated with the right mandibular deciduous second molar. The oral hygiene was moderate with slight increments in plaque.

A computed tomography (CT) scan revealed cortical hyperdensities in the right occipito-temporo-parietal region with moderate enlargement of the lateral and third ventricles. Bilateral cerebral sulci were prominent. (Fig. 3A) Ultrasonography (USG) of the skull suggested communicating hydrocephalus.

Radiographs of limbs and chest X-ray did not show any abnormality. The blood picture was normal. Magnetic resonance imaging (MRI) revealed diffuse prominence of the cerebral sulci spaces in the bilateral fronto-temporal-parietal regions, but no blooming was evident on gradient recalled echo (GRE) (Fig. 3B). Intraoral periapical radiograph (IOPA) showed periapical pathology of the lower right deciduous second molar (Fig. 4A). The above clinical and radiographic features extended the diagnosis to Sturge–Weber Syndrome.

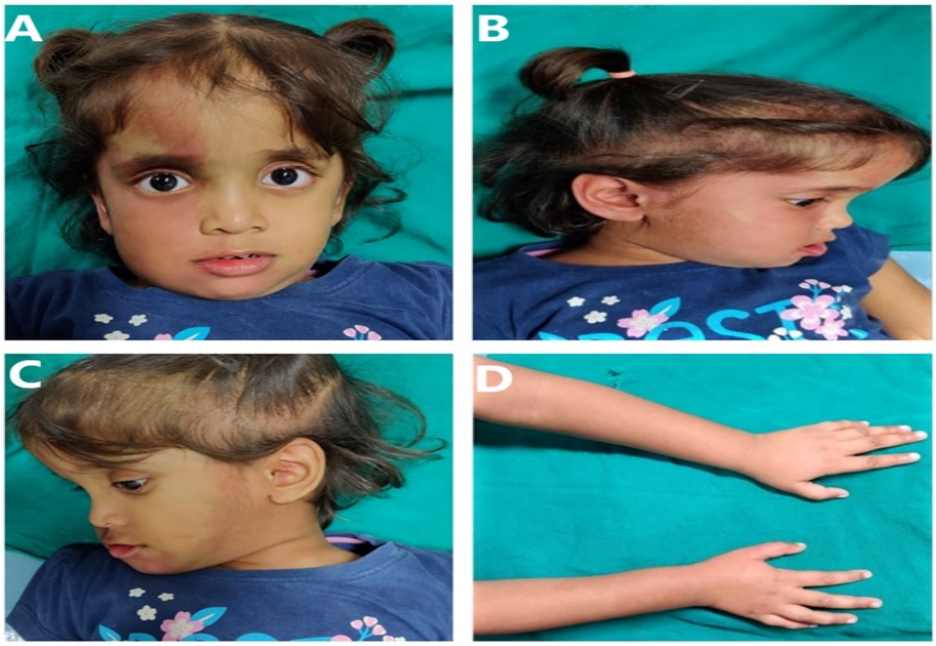

Figure 1. Extra-oral clinical manifestations of child with Sturge-Weber Syndrome. A: port wine stains on right half of the face and head. Asymmetrical facies and macrochelia of the lower lip seen; B: right lateral view of the child with observance of port wine stains; C: port wine stains on the right upper extremities

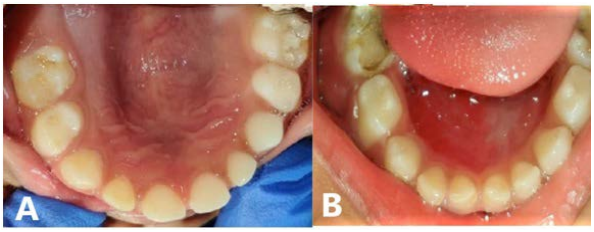

Figure 2 .Intraoral images of child with Sturge-Weber Syndrome; A: maxillary occlusal view with lesions over the posterior surface of the hard palate; B: mandibular occlusal view revealing unilateral port wine stain in right half of the floor of mouth. 85 with periapical abscess of deep carious lesion and dentinal caries of 75

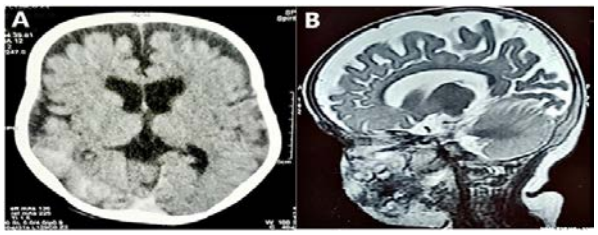

Figure 3. A: CECT scan of brain reveals cortical hyper densities in right occipito-temporo-parietal region and moderate enlargement of third and lateral ventricles; B: MRI reveals diffuse prominences of cerebral sulci. Spaces in bilateral fronto-temporo-parietal region. No blooming seen on GRE

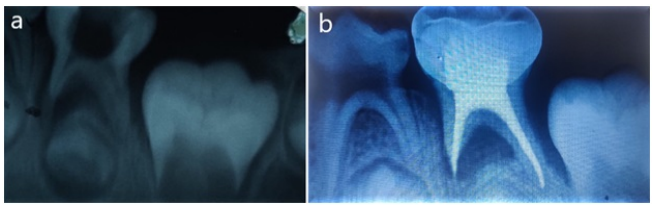

Figure 4. A: pre-operative intraoral periapical radiograph of 85 tooth; B: intraoral periapical radiograph postoperatively depicting the rehabilitated 85 tooth with stainless steel crown restoration

Pediatric department consent was obtained prior to the treatment. Pre-procedural counseling sessions were carried out to alleviate the fear and anxiety in children with a supportive psychotherapy schedule. The cause of swelling was confirmed using the standard diagnostic protocol and clinical investigations. Emergency access opening was performed with an endo access bur (Dentsply, Mallifer, Switzerland) under the administration of intraligamentary and intrapulpal anesthesia (Lidocaine 2%) under a rubber dam, followed by irrigation with 1% sodium hypochlorite (20 mL) (Parcan, Septodont Healthcare India Pvt Ltd, Raigad, India) and saline (20 mL) for the bacterial decontamination of canals. After open dressing, the child was medicated and recalled after 5 days. The patient was asymptomatic with resolved swelling at the next appointment. After disinfection with irrigants, biomechanical preparation was carried out using Kedo-S-Square (Reeganz Dental care Pvt Ltd,Chennai, India) rotary endodontic file system. After disinfection, the canals were dried and obturated using Metapex (Meta Biomed Co. Ltd, Cheongju, Korea). The next appointment was scheduled for full coronal restoration with a stainless steel crown (3M™ ESPE™ Stainless steel crowns, 3M, St. Paul, MN, USA) (Fig. 4b). Class I carious 55 and 65 were excavated partially (one-step partial excavation) and subsequently placed with final restoration using Glass Ionomer Cement(Ketac molar, 3M Corp., Minnesota, USA). A plaque control regimen was started from day one to minimize gingival enlargement, which included oral prophylaxis that was carried out every week for the first month, and 0.2% w/v chlorhexidine (Hexidine, ICPA Health Products Ltd, Mumbai, India) mouth wash was advised. Oral hygiene maintenance was reinforced during every visit, and the patient was decided monthly.

DISCUSSION

The complete form of the Sturge–Weber syndrome consists of a triad that includes the unilateral manifestation of congenital capillary malformation in the trigeminal nerve distribution, vascular malformations, such as ipsilateral leptomeningeal involvement with intracranial calcification, and neurologic signs, and vascular malformations of choroid plexus, often with secondary glaucoma.[10] Since only sporadic cases have been reported, inheritance is unclear; thus, authors have hypothesized the complex interaction between genetic alterations and MAPK and PI3K activation.[8] Tan et al.[11] reported that the clinicopathological vasculature dilatation might be due to the coexistence of vascular identities, namely Eph receptor B1 (ephb1) and ephrinb2 in Port wine stain endothelial progenitor cells that contribute to pathoanatomical vasculature. SWS is a rare but most frequent neurocutaneous syndrome and embryonic developmental disorder affecting both the mesodermal and ectodermal germ layers.[3,12] No racial predilection has been observed, and both sexes are equally affected. The central nervous system (CNS) involvement with facial angiomas is labeled as complete SWS. SWS could be categorized clinically according to the Roach [13] scale that would be enumerated as follows: a) Type I: Both facial and leptomeningeal angiomas; may have glaucoma; b) Type II: Facial angioma alone (no CNS involvement); may have glaucoma; c) Type III: Isolated leptomeningeal angiomas; usually no glaucoma.

Reportedly, there is a risk of 10–35% brain involvement when the patient has facial Port wine stain, and if both eyelids are involved, the risk of

glaucoma would be 50%. On the other hand, ipsilateral choroidal hemangioma is seen in 20% of affected individuals.[9]

In the present case report, the 3.5 -year-old child had unilateral facial Port wine-stains since birth. The child was not mentally retarded but had pronounced CNS involvement and a history of seizures, which might be considered a case of incomplete SWS with ipsilateral involvement; however, Roach classification may deem it type I without glaucoma.

The current case had a history of mild episodic attacks of seizures at the age of 2.5 years, for which he was on antiepileptic drugs as prescribed by the physician. The neurological manifestations may depend on the site of leptomeningeal angiomas with a common location in the frontal, parietal, and occipital regions. These incorporate focal deficit seizures in the form of transient hemiparesis and hemianopsia that concord with the findings of Maria et al. Cortical ischemia may develop around the angioma, clinically manifesting as progressive calcification, gliosis, and atrophy that results in neurological deterioration. This deterioration could be prevented by seizure control, aspirin medication, and early surgical intervention.[14]

The vascular plexus developed at the sixth week of embryonal life Is ordained for facial skin development around the cephalic region of the neural tube underlying the brain tissue. These advancements normally regress at the ninth week of gestation. The failure of this natural apoptosis results in the formation of the angiomata of the leptomeninges, face, and ipsilateral eye.[15]

Oral pathoanatomic vascular lesions account for approximately 40% of syndromic patients. Intraorally, angiomatous red-purple discoloration is expanded over regions, such as buccal mucosa, palate, tongue, the floor of mouth, gingiva, and lips. The observations on gingiva may vary from mild vascular hyperplasia to severe hemangiomatous proliferation accounting for massive growth. Intraoperative and postoperative oral hemorrhages pose a risk for routine dental and surgical oral procedures. Yamashiro and Furara[16] reported the postponement of the gingivectomy procedure due to excessive hemorrhage.

The dental treatment of patients with Sturge–Weber ailment is a complicated procedure requiring conservative management post-surgical intervention. Behavioral issues might be experienced because of past presentation to the Emergency Clinic or because of mental deficit, which utilizes behavioral management techniques.

As seen in our case of excessive swelling, emergency endodontic treatment does not convey the hazards of uncontrolled hemorrhage as angioma rarely includes the pulpal tissue. Draining can be constrained by cotton pellets and vasoconstrictors; also, the use of tranexamic acid might be propagated in an emergency if bleeding id not arrested. Nonetheless, safeguard is essential during root canal treatment and rehabilitation for undue injury of periapical tissue by over instrumentation and excessive subgingival preparation. This would aid in full coronal restoration placement for strategic precaution from hemorrhage, as angiomas have been reported in the periapical area.

CONCLUSIONS

Rendering dental treatment for a patient with Sturge–Weber ailment is confounding for the specialist due to the prevalence of oral vascular lesions and their associated menace factor. The risk of hemorrhage and epilepsy episodes with behavioral disorder worsens the scenario intra- and postoperatively. Thus, the dental specialist needs elite attention on early comprehensive treatment, beginning with behavior management and focusing on preventive measures similar to any patient having developmental disorders following the most desirable treatment.

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Declarations

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