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Dental care challenges in Sturge-Weber syndrome: a case report

Desafios do tratamento odontológico na síndrome de Sturge-Weber: relato de caso

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Abstract
Introduction*: Sturge-Weber syndrome (SWS) is a rare condition characterized by facial capillary malformation, involves ocular, neurological, and cutaneous alterations. Associated with unilateral characteristic port-wine stains, gingival growth and purple-red coloration.* ***Aim:*** *his case aims to report dental treatment challenges in patients with SWS and importance of oral health maintenance in these individuals.* ***Case report:*** *a 20-year-old woman with an established diagnosis of SWS, presented bad breath and spontaneous gingival bleeding, with gingival growth and reddish-purple spots spread to labial and alveolar mucosa, tongue, and palate. Conditioning of the patient's oral environment by supra and subgingival scraping, dental unit extraction was performed. A conservative treatment plan was adopted for management adequacy of oral environment owing to possible complications inherent to the condition.* ***Conclusion:*** *it is important to emphasize the importance of dental surgeon's performance in relation to a multidisciplinary health team, as well as cooperation of patient, to obtain better results from the proposed therapy.*
Keywords*: Sturge-Weber Syndrome. Port-Wine Stai., Angiomatosis. Complications. Dental Care.*

Resumo
Introdução: a síndrome de Sturge-Weber (SSW) é uma condição rara caracterizada por malformação capilar facial, envolve alterações oculares, neurológicas e cutâneas. Associada a manchas unilaterais características do vinho do porto, crescimento gengival e coloração vermelho-púrpura. **Objetivo:** este caso tem como objetivo relatar desafios do tratamento odontológico em pacientes com SSW e a importância da manutenção da saúde bucal nesses indivíduos. **Relato de caso:** paciente do sexo feminino, 20 anos, com diagnóstico estabelecido de SSW, apresentou mau hálito e sangramento gengival espontâneo, com crescimento gengival e manchas roxas avermelhadas espalhadas pela mucosa labial e alveolar, língua e palato. Condicionamento do ambiente oral do paciente por raspagem supra e subgengival, foi realizada extração da unidade dental. Foi adotado um plano de tratamento conservador para adequação do manejo do ambiente bucal devido a possíveis complicações inerentes à condição. **Conclusão:** é importante enfatizar a importância do desempenho do cirurgião-dentista em relação a uma equipe multidisciplinar de saúde, bem como a cooperação do paciente, para obter melhores resultados com a terapia proposta.
Palavras-chave: Síndrome de Sturge-Weber. Mancha de Vinho do Porto. Angiomatose. Complicações. Cuidado Dental.

INTRODUCTION

Sturge-Weber Syndrome (SWS) is a rare congenital neurocutaneous disorder. It is characterized by the presence of facial port-wine stains, increased ocular pressure, and leptomeningeal angiomatosis[1]. The pathogenesis of SWS involves a somatic mutation in a nucleotide transition in the GNAQ gene on chromosome 9q21. The exact mechanism of how this activation results in macules with violet staining and in the establishment of SWS is not yet clear[2].

The clinical components of SWS can be divided into cutaneous, neurological, and ocular malformations. The most prominent malformation is a facial macula, usually unilateral and similar in color to port wine. The advancement of facial maculae to maxillary and mandibular areas is associated with soft tissue hypertrophy and excessive growth of the underlying bone regions. Glaucoma is the most common ophthalmic manifestation of SWS[3].

The clinical course of SWS is variable, but it is usually characterized by progressive neurological problems such as seizures, hemiparesis, headache, stroke, behavioral problems, mental retardation, and visual field defects[4].

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Radiographic features include intracranial calcifications known as "line" or "tram track"[5]. In fact, histological studies of cerebral angiomas in SWS revealed tortuous and abnormal vascular structures in thick leptomeninges[4].

SWS is classified into three types according to the Roach scale: type I, with vascular facial and endocranial malformations with the possibility of glaucoma (classic form); type II, involving the presence of facial angioma with the possibility of glaucoma, without evidence of intracranial involvement; and type III, often involving leptomeningeal angioma[4].

The main oral manifestations include changes in mucosa coloration, that is, the appearance of purple-reddish macules; gingival lesions that may include a significant vascular component; macroglossia; and hemihypertrophy of palatal and buccal mucosae[6,7]. Angiomatosis favors the development of pyogenic granuloma and malocclusion and increases the risk of bleeding during dental procedures[8,9].

Moderate-to-severe variable gingival growth may be present, which causes greater discomfort, impaired nutrition, difficulty in hygiene, and consequently favors the accumulation of a biofilm, to increasing the individual's susceptibility to oral infections and compromising systemic health. This gingival growth may be induced by anticonvulsive medications used in treating these individuals, which usually report epileptic seizures[10].

Considering the rarity of the disease and need for dental care procedures, this article aims to report a case of SWS patient with emphasis on challenges in dental treatment and the importance of oral health maintenance in SWS patients.

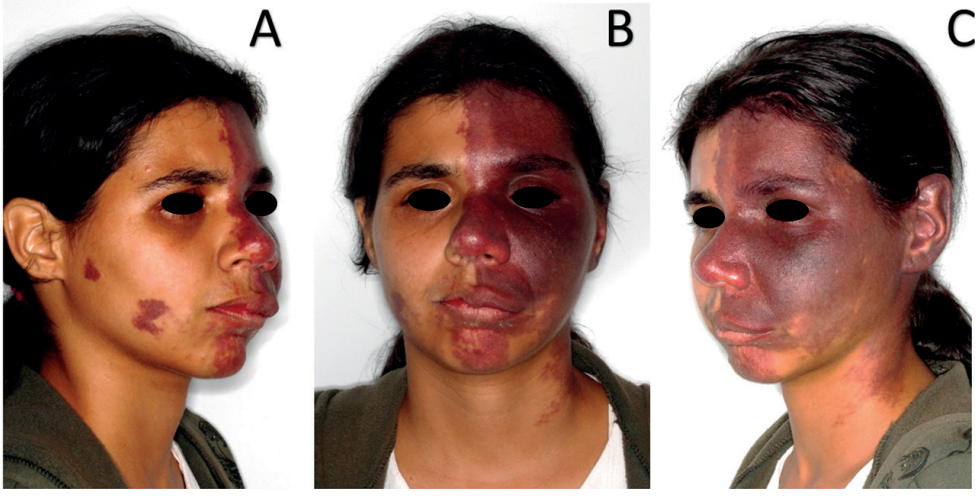
CASE REPORT

A 20-year-old woman attended the Special Patient outpatient clinic in a higher education institution. She was diagnosed with SWS since childhood with secondary epilepsy and right hemiparesthesia, and her last episode of crisis was more than 10 years prior. She reported regular use of Phenobarbital (Gardenal®, Sanofi-Aventis, Rio de Janeiro-Brazil) for 10 years. At the time of admission, she was using 200 mg of Carbamazepine (Tegretol®, Novartis, Taboão da Serra-São Paulo- Brazil) and 5 mg of Clobazam (Frisium®, Sanofi-Aventis, Rio de Janeiro-Brazil). The patient's medical report stated that she was vigil, oriented in time and space, and had preserved language and coordination, and tactile sensitivity. However, she presented with amaurosis and reduced visual acuity and temporal visual field on the right side.

The patient complained of bad breath and a fracture in one of the dental units. She also reported spontaneous gingival bleeding and mild pain in the fractured dental unit while chewing.

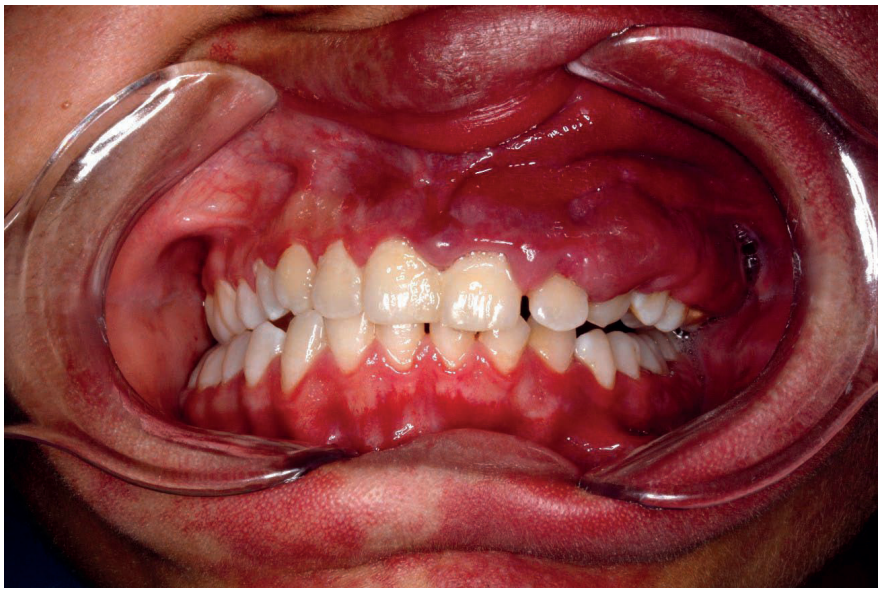
The extraoral examination revealed a port-wine stain in the left hemiface, causing a deviation from midline, and extending from the scalp line to the cervical region. It involved the auricular pavilion, nasal apex, and areas in the lower third of the face contralateral (Figure 1). The intraoral examination revealed extensive port wine lesion affecting labial, alveolar mucosa, gingiva, tongue, hard and soft palate in the left region, respecting the median line (Figures 2 and 3). The patient presented gingival hyperplasia in the second, third, and fourth sextant of the upper and lower arches. The patient had poor oral hygiene resulting in supra- and subgingival calculus accumulation and gingival inflammation, especially at the sites of vascular alterations. The patient also presented dental unit 2.7 with extensive carious lesion in dental crown (Figure 3). After a careful periodontal assessment, the patient was diagnosed with localized mild chronic periodontitis.

Figure 1 – *Side view (A and C) and frontal (B) photographs showing extensive port-wine stains in the left hemiface, extending from the scalp line to the cervical region, and contralateral facial areas.*



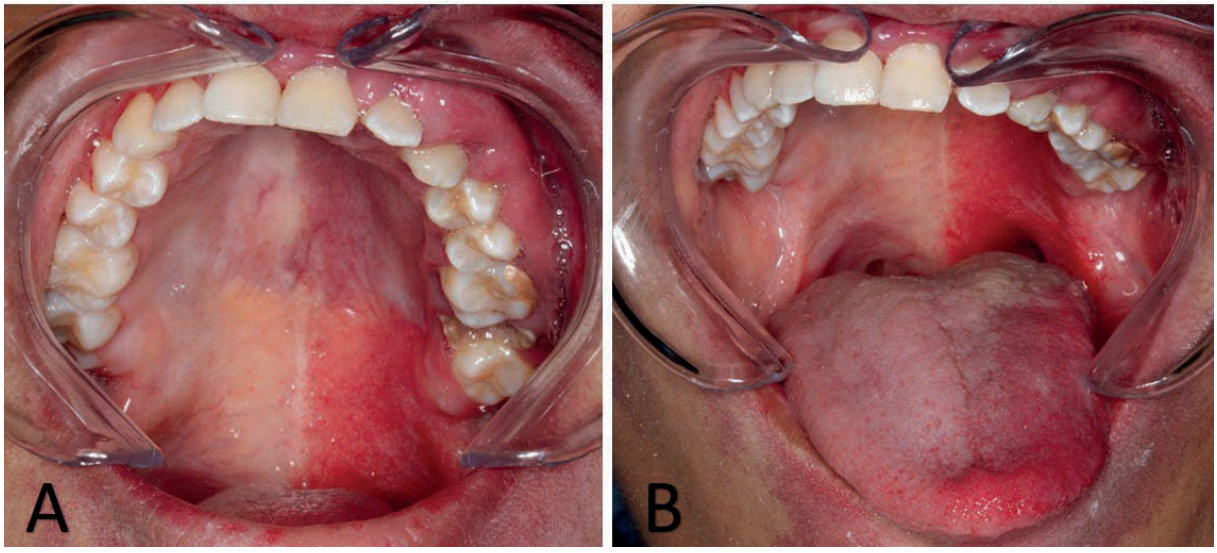
Source: Own authorship

Figure 2 – *Intraoral photograph showing an extensive violaceous lesion affecting the labial, alveolar mucosa and left maxillary gingiva.*



Source: Own authorship

Figure 3 – *Intraoral photographs showing (A) gingival hyperplasia observed in the second and third sextants, and in dental unit 2.7 with extensive coronary destruction and violaceous macula on the back of the tongue, hard and soft palate in the left region (B), respecting the median line.*



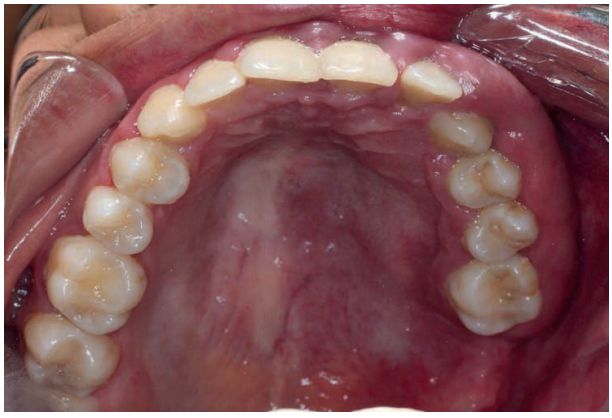
Source: Own authorship

The proposed treatment plan included scaling and root planing of all sextants, prophylaxis, topical application of fluoride, extraction of units 2.7 and 2.8, and oral hygiene instruction and motivation. In association with mechanical periodontal therapy, use of 0.2% chlorhexidine gel (Perioxidin®, Lacer, Spain) associated with mechanical control of biofilm with interdental brush. The patient accepted the protocol for oral care proposed by dental team and authorized publication of photographic and medical records by signing the Term of Free and Informed Consent (TFIC).

Before tooth extraction of unit 2.7, a nuclear magnetic resonance imaging examination was performed to reduce the risk of possible excessive bleeding during surgical procedure. The trans-surgical procedure was performed in a surgical center without any complications. Extraction of unit 2.8 was indicated to prevent the need for further surgical intervention at the site of vascular enlargement because of the increased risk of trans- and postoperative hemorrhage. After the surgical procedure, scaling and root planning (SRP) of the third and fourth sextants were performed to remove possible retentive factors of the bacterial biofilm.

In the postoperative period, the patient progressed favorably, and attended the outpatient control visits. After 5 months of oral environment conditioning, the patient had good oral hygiene with no pain complaints and no lesions suggestive of dental caries, and a stabilized periodontal condition (Figure 4).

Figure 4 – *Intraoral photograph after conditioning of the buccal medium. Note the partially edentulous upper arch due to the extraction of units 2.7 and 2.8.*



Source: Own authorship

DISCUSSION

The periodontal changes due to SWS can result in poor oral hygiene and chewing difficulties, predisposing an individual with SWS to local and systemic infections[10]. In fact, in the present case, we verified that the patient had difficulty in biofilm control because of pain and bleeding during the assessment of areas most affected by the syndrome.

Previously Tripathi and co-workers[11] reported on a 15-year-old girl diagnosed with SWS who presented with pain and gum bleeding during chewing. After one year, patient had a convulsive condition, and the extraoral examination showed port-wine stain in the right hemiface and dilated vessels in the right eye. The authors reported that the patient had a right unilateral hyperplastic lesion in the maxilla, which bled after application of local pressure. The treatment consisted establishment of an oral biofilm control regimen, oral prophylaxis associated with scaling and root planing, and oral hygiene instructions on using a chlorhexidine mouthwash.

Corroborating with these authors, Pagin et al.[8] also reported on a patient with SWS and maxillary gingival hyperplasia presenting supra- and subgingival calculus in the affected region. One can assume that due to oral vascular alterations, toothbrush movement is restricted. This triggers greater accumulation of plaque and supra- and subgingival calculus. The authors[8] observed regression of gingival inflammation only with SRP and use of mouthwashes containing 0.12% chlorhexidine. In the present case, oral hygiene instructions and use of chlorhexidine mouthwash for plaque control were instituted. However, SRP was initially limited to the sextants which did not have marked gingival hyperplasia and inflammation, since the patient presented with severe bleeding with least local trauma. The complete SRP was only possible during trans-surgical procedure.

Another study[10] described the case of a 15-year-old boy who complained of edema in the left anterior region of the jaw that started at age 6 years but had no pain or bleeding. Extraoral examination revealed hemihypertrophy on the right side of the face and the presence of a port-wine stains, lip edema, increased malar prominence with frontal bossa, scarring of the frontal region, and widening of the frontal septum. Intraoral examination revealed gingival enlargement, presence of biofilm, supragingival calculus, and purple-reddish coloration on the soft palate, buccal floor, and direct side of the buccal mucosa. The therapeutic management[10] included SRP associated with oral hygiene instructions. The authors[10] performed extraction of a dental unit, with an unfavorable prognosis. Corroborating with these authors, in the present case, It was performed extraction of unit 2.7 with a restrictive and unfavorable prognosis, and chose to extract unit 2.8 during the trans-surgical procedure, as it was in a region of limited access difficulting its hygiene by the patient, representing a possible risk to the patient's systemic health.

Previously Gill and Bhaskar[12] suggested that gingival volume increase can be treated by laser gingivectomy, as there is an added advantage of homeostasis and minimal damage to oral tissues. However, this therapeutic option was not choosen for this patient. It was because it is essential to first establish patient's oral hygiene control, before proceeding with surgical techniques. Removal of local infectious foci precedes aesthetic surgeries.

Shaikh et al.[13] as the same way as in the presented study, adopted a conservative therapeutic approach in a 11-year-old child diagnosed with SWS. The patient complained of painful symptoms in dental units with mobility for about 2 months, and presented with gingival bleeding in the region of upper and lower right teeth, and dental caries in the left upper posterior teeth. At examination, vascular nevi were extended to the oral mucosa and underlying gingiva. The patient was instructed to use chlorhexidine mouthwash for controlling biofilm formation[14] and was submitted to extraction of the indicated dental units under local anesthesia. Likewise, in the present case, the treatment plan included similar prophylactic and preventive actions for biofilm control; using a mouthwash with 0.12% chlorhexidine solution two times a day. After two weeks, mouthwashes were replaced by the application of 0.5% chlorhexidine gel using interdental brushes and SRP. Due to the degree of inflammation, gingival hypertrophy, and hemorrhagic conditions, a more conservative approach was chosen to manipulate the second quadrant. Tooth extraction and scaling of supra- and subgingival of the affected sextants were performed after angiology, thus avoiding any possible trans- and

post-surgical complications.

SWS is included in the group of phakomatoses that are characterized by hamartomas involving the brain, skin and eyes[15,16]. It is important to provide these patients multi-professional care to treat various complications inherent to the syndrome. In the present case, the patient attended dental care, since she already had follow-up with ophthalmology and neurology teams.

CONCLUSION

Oral vascular manifestations of SWS result in important periodontal changes. Therefore, patients diagnosed with SWS should perform regular follow-ups with their dental surgeon to monitor and control the onset of inflammatory and infectious processes. This will help patients become aware of their condition and commit to maintaining good oral hygiene techniques. Together this will lead to better therapeutic results and prevent conditions that require more invasive therapies.

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