Revista Brasileira de Odontologia
Brazilian Journal of Dentistry
Associação Brasileira de Odontologia - Seção Rio de Janeiro
DOI: http://dx.doi.org/10.18363/rbo.v77.2020.e1884

The Contribution of Dentistry on The Sturge-Weber Syndrome Diagnosis and Management - Report of Two Cases

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• Conflicts of interest: none declared.

Abstract

Objective: The aim of this study was to report two cases of female patients diagnosed with Sturge-Weber syndrome (SWS), its main oral and maxillofacial clinicals manifestations and dental management. **Case report:** Case 1. A 30-year-old female patient, referred to the Oral Medicine Clinic of State University of Rio de Janeiro for evaluation of a diffuse reddish lesion over her face and mouth. During the clinical examination, port-wine stains (PWS) were detected on the right side of her face and intraoral discoloration were suggestive of SWS. As the patient referred no symptoms, she was referred to a neurology team to investigate alterations in the central nervous system (CNS). Case 2. A 19-year-old woman was referred for evaluation of a 5-month lasting gingival bleeding in the right mandibular and maxillary alveolar mucosa. Extraoral exam showed the presence of PWS on the right side of her face. Intraorally, the gingival hyperplasia caused malocclusion, plaque deposition and severe bleeding. The SWS diagnosis was established due to these findings. Patient was referred for periodontal treatment. **Conclusion:** Thus, it is essential that the dentist has knowledge of this condition clinical manifestations in order to do the correct diagnosis and purpose the correct treatment.

Keywords: Sturge-Weber Syndrome; Port-Wine Stain; Gingival Hyperplasia.

Introduction

Sturge-Weber syndrome (SWS) - also called as encephalotrigeminal angiomatosis - is a rare developmental non-hereditary congenital condition characterized by facial cutaneous vascular nevus (nevus flammeus or port-wine stain) in association with venous angiomas in leptomeninges over the cerebral-cortex, usually unilateral - which often follows the outline distribution of trigeminal nerve.[1,2]

SWS often shows morphological and histological alterations in the periodontal tissues - frequently gingival hyperplasia involving the maxilla, mouth floor, lips, buccal mucosa, palate or tongue is observed, ipsilateral to the port-wine stain on the face. Therefore, it is important for the clinician to know this syndrome oral manifestations and the complications that may occur - such as increased risk of hemorrhage during surgical procedures - in order to purpose the correct treatment amongst others medical specialties.[1,2,3]

Herein, we report two cases of female patients diagnosed with SWS presenting extra and intraoral manifestations.

Case Report

Case 1. A 30-year-old white female patient was referred from a dental clinic to the Oral medicine clinic, State University of Rio de Janeiro in order to evaluate reddish lesions on her face and mouth - that kept growing, but were present since younger ages. Family history was non-contributory, and the patient did not report any previous surgeries or neurological disturbs. On extraoral examination (Figure. 1.a, 1.b), a right-sided hemihypertrophy of the face with port-wine stains was observed, extending along the second division of the trigeminal nerve unilaterally. Her mouth was deviated toward the inferior-right side of her face. The upper lip and alar base were swollen, edematous, and incompetent. Also, there was increased malar prominence and facial asymmetry. Intraoral exam revealed the presence of a diffuse reddish to purple discoloration over the hard and soft palate on the right side, not exceeding the midline (Figure 1.c). Patient referred to no intraoral symptoms or periodontal alterations. The clinical findings confirmed the Sturge-Weber syndrome diagnosis. However, despite the oral manifestations, as the patient complained about no symptoms, she was oriented regarding the syndrome and referred to a neurology service in order to investigate possible alterations.

Case 2. A 19-year-old white female patient was referred for evaluation of a 5-month lasting gingival bleeding in the right mandibular and maxillary alveolar mucosa. Medical history was non-contributory, and patient denied any addictions regarding alcohol or tobacco abuse. Extraoral clinical examination showed right-sided hemihypertrophy

of the face with port-wine stains following the outline distribution of trigeminal nerve on the face crossing the midline. A significant facial asymmetry could be observed with mouth deviation toward the left side and both upper and lower lips presented swollen, edematous and incompetent (Figure 2a, 2b). Intraoral clinical examination revealed the presence of a diffuse gingival enlargement in both right maxilla and mandible along with reddish-areas throughout the right hard palate, buccal mucosa, tongue and mouth floor - that did not cross the midline. Blanching on pressure could be noted in the enlarged gingiva, suggesting angiomatous lesion. Both right maxillary and mandibular teeth were misplaced, and significant plaque and calculus deposition was presented due to lack of oral hygiene (Figure 2c). Due to the clinical findings the Sturge-Weber syndrome diagnosis was established. Patient was oriented regarding the syndrome and its complications and referred to the Periodontology clinic to start periodontal treatment - Plaque control and gingival excision - and to a neurology service to investigate alterations in the central nervous system.

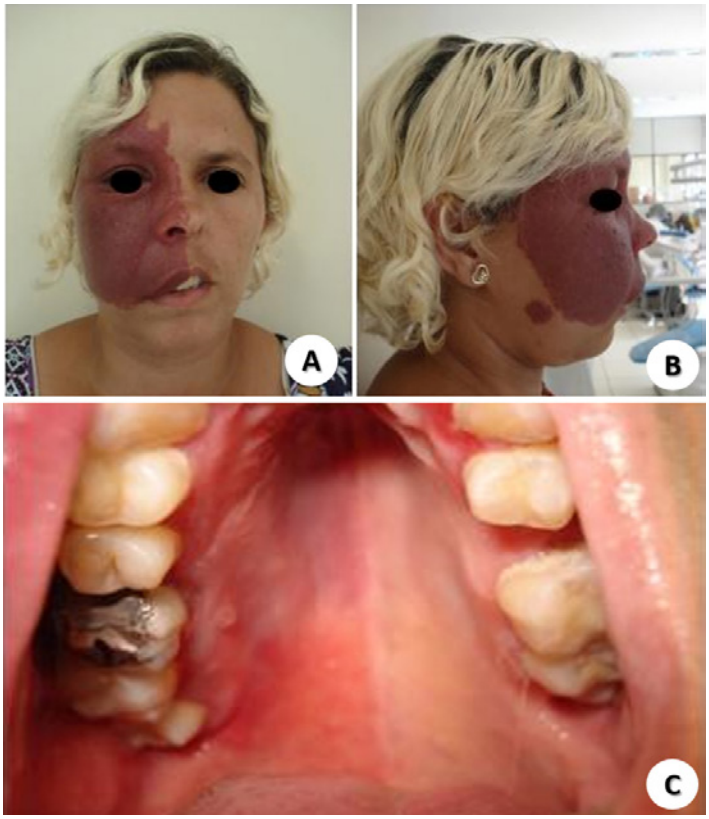

Figure 1. A - Frontal View - a right-sided hemihypertrophy of the face with port-wine stains following the trigeminal nerve second division is observed. B - Profile View. C - Intraoral features - Notice that the reddish discoloration on the right side do not exceed the midline and it is located ipsilateral to the PSW on the face.

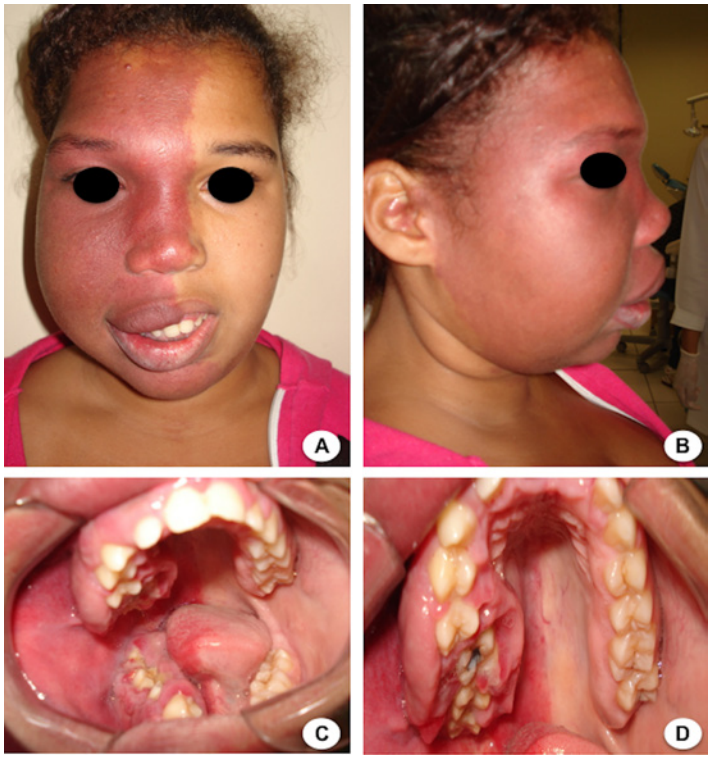

Figure 2. A - Frontal View - a right-sided hemihypertrophy of the face with port-wine stains. B - Profile View. C - Severe periodontal alterations may be observed on this patient presented with SWS in both right maxilla and mandible. D - Gingival hypertrophy on the right maxilla.

Discussion

Sturge-Weber syndrome is a very uncommon, non-familial, congenital condition of unknown etiology. SWS presents varied clinical features that may affect the central nervous system (CNS), eyes, skin (specially the face) and the oral cavity - which the dental clinician should be aware of when performing the routine stomatologic exam. Its diagnosis can be established by the presence of port-wine stain on the face followed by other signs, such as glaucoma, epilepsy, and mental retardation (Figure 3).[1,2,3] Table 1 highlights the extra and intraoral features reported on previously SWS cases as well as its dental management, thus highlighting the role of the dentist on its diagnosis and treatment.[1,2,4-17]

The Roach Scale is used as a classification of the SWS manifestations:[18] Type 1 presents with facial and leptomeningeal angiomas and glaucoma may occur; Type 2 involves facial angioma alone - no CNS alteration is observed - and may have glaucoma and finally, type III shows isolated leptomeningeal angiomas and usually glaucoma. Furthermore, when the CNS and facial angiomas are present, SWS is referred to as complete, whereas it is incomplete when only one of these areas is affected.[12] Both of our cases presented as the incomplete type II form of Sturge-Weber syndrome - with the presence of facial angiomas and no CNS alteration or glaucoma.

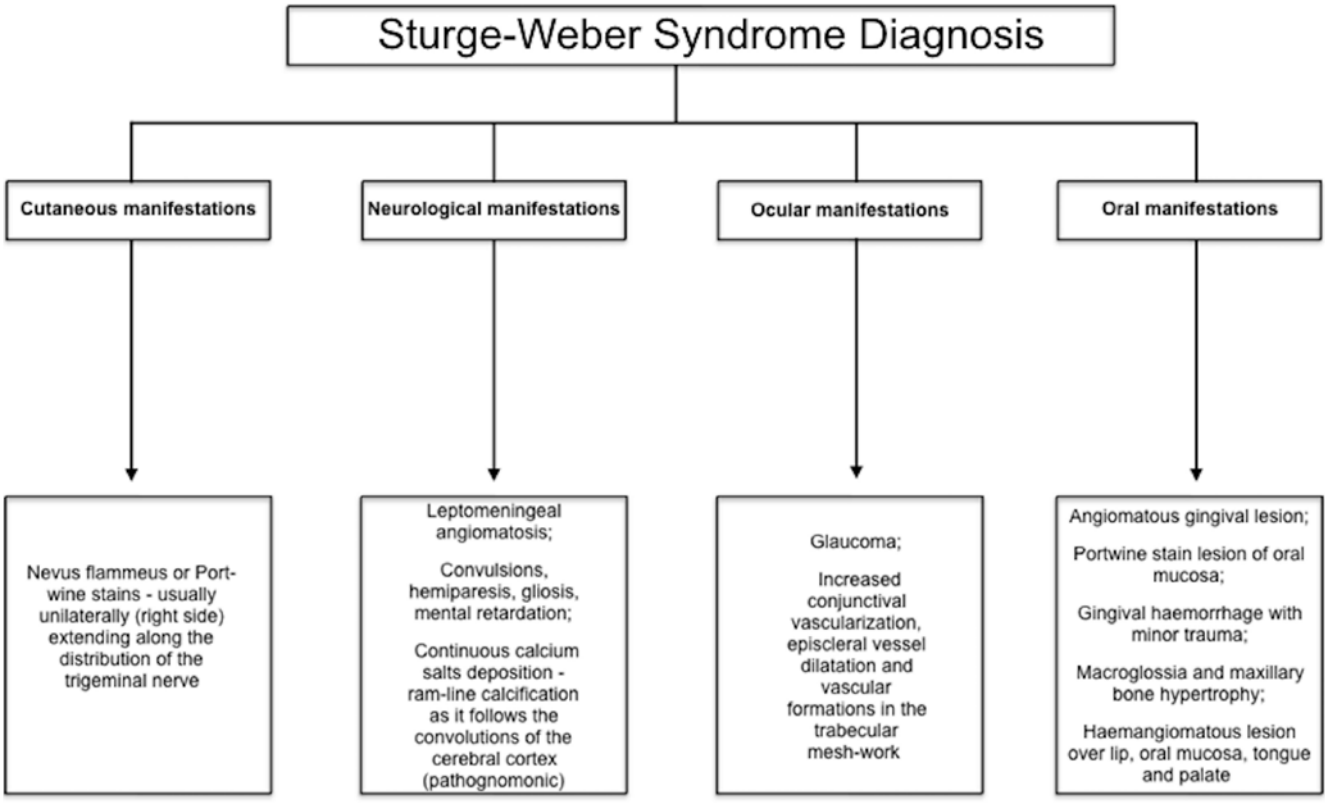

Figure 3. Sturge-Weber syndrome diagnostic features.

Table1. Sturge-Weber syndrome clinical findings and treatment

Author	Sex	Age (years)	Extraoral features	Intraoral features	Dental manegement
Gyarmati *et al*.[4]	Female	10	Bilaterally PWS; slight swelling of the upper lip;	1.5 cm gingival growth	NS
el-Mostehy *et al*.[5]	Female	14	PWS on the left face	Gingival swelling on the left face; mass of hypertrophied, deep red, easily bleeding angiomatous tissue on the left mandibular vestibular; periodontal pockets; teeth mobility	Gingival growth excision; debrided of tissue tags and calculus; root surfaces
Yukna *et al*.[6]	Male	25	PWS on the left face, extending from the forehead to the neck and from the midline to the ear	Dusky-red lesion on the left side involving the buccal mucosa, floor of the mouth, pharynx, palate, uvula, and the gingiva of both the maxilla and the mandible. Slight to moderate gingival hyperplasia, coupled with a marginal gingivitis; carious lesions associated with the left posterior teeth	Plaque-control instructions; sealing and root planning; extractions; pocket elimination by flap surgery
Perez *et al*. [7]	Female	6	Bilaterally PWS, bilateral congenital glaucoma	Red swelling involving the upper anterior gingiva in the region of the central incisors and the mucosa of the upper lip; carious lesions; malocclusion	Plaque control, carious teeth restore; forward to the Orthodontic clinic for evaluation and treatment.
Yamashiro *et al*.[8]	Male	35	Bilaterally PWS; Glaucoma	Gingival enlargement and periodontitis.	Extractions and gingivectomy.
De Benedittis *et al*.[9]	Male	25	Bilaterally PWS, hemiplegia, glaucoma, blind in the right eye,	Macroglossia secondary to the hemangioma; gross angiomatous mass	Gingivectomy using Nd:YAG laser
Bhansali *et al*.[1]	Male	15	Hemiplegia on the right side along with PWS; right-sided hemihypertrophy of the face; upper and lower lips swollen, edematous, and incompetent	Gingival enlargement involving all four quadrants of the oral cavity; periodontal pockets; purplish-red discoloration over the soft palate, the floor of the mouth bilaterally, and the buccal mucosa on the right side	Scaling and root planing; extraction; gingivectomy

Conceição *et al*.[2]	Male	29	Lower lip hemangioma	Hemangioma in retromolar trigone and in the lower surface of the tongue seen as purplish red spots without symptomatology	None
Pagin *et al*.[10]	Male	43	PWS on the right side of the face	Stain in the entire hard and soft palates, and the alveolar ridge and buccal mucosa on the right side. Poor oral hygiene, calcified masses in both supragingival and subgingival sites, with swelling and generalized inflammation throughout the gingiva	Technical hygiene instruction; successive sessions to remove dental calculus by ultrasound.
Manivannan *et al*.[11]	Female	20	PWS on the left side of the face; facial asymmetry and macrochelia	Prominent reddish purple gingival enlargement posteriorly on the left side; teeth on the lower arch were malpositioned; poor oral hygiene, extensive amounts of plaque and calculus.	Oral prophylaxis, surgical excision of the lesion under general anesthesia and replacement of the worn out bridge in the upper anterior region.
Babaji *et al*.[12]	Female	8	PWS on the right side of the face	Maxilla on the right side revealed reddish discoloration of gingiva extending from labial frenum to the first molar region with osseous enlargement and drifting of teeth; gingival hyperplasia	Plaque control, oral prophylaxis at regular interval, oral hygiene instructions. Mobile deciduous were extracted
Kalakonda *et al*.[13]	Male	23	PWS on the left side of the face	Severe gingival overgrowth; loss of knife edge contour and bulbous papillae	Gingivectomy with diode laser (Picasa 810 nm 1.8 W continuous mode)
Kalakonda *et al*.[13]	Male	32	PWS on the left side of the face; facial asymmetry; swelling of the left side of the upper lip	Gingival overgrowth	Nonsurgical periodontal therapy under strict aseptic conditions.
Kalakonda *et al*.[13]	Female	12	PWS on the right side of the face; reddish discoloration of the sclera of the right eye	Discrete, sessile, reddish-pink gingival overgrowth; bleeding	Gingivectomy;
Tripathi *et al*.[14]	Female	15	PWS on the right side of the face	Unilateral hyperplastic lesions on the right side of the maxilla	Oral plaque control; prophylaxis
Pontes *et al*.[15]	Female	23	PWS on the right side of the face; swelling of the superior lip	Large overgrowth in the right maxilla extending from teeth 11 to 18 and a minor growth in the right mandible encompassed the area between teeth 42 and 48	Surgical excision with an electric scalpel; flap surgery, gingivectomy, osteotomy, osteoplasty, and extractions
Shaikh *et al*.[16]	Male	11	PWS on the right side of the face; Initial glaucoma	Inflamed and hypertrophied gingiva of the right upper and lower quadrants; carious lesions; calculus; erythematous reddish pink patches on the mucosa	Plaque control; extraction
Neerupakam *et al*. [17]	Female	18	PWS on the right side of the face; glaucoma	Hyperplasia of right side gingiva, including interdental, marginal and attached gingiva; palatal ecchymosis	Curettage of right maxillary region; gingivectomy using the dioode 980 nm laser
Present case 1	Female	30	Right-sided hemihypertrophy of the face with PWS; facial asymmetry	Diffuse reddish to purple discoloration over the hard and soft palate on the right side, not exceeding the midline	None
Present case 2	Female	19	Right-sided hemihypertrophy of the face with PWS; facial asymmetry	Diffuse gingival enlargement in both right maxilla and mandible along with reddish-areas	Referred to the Periodontology clinic for gingival excision and plaque control

PWS - Port-Wine Stain; NS- Not stated

The SWS cases reported in table 1[1,2,4-17] showed a slight preference for women (55%), most of them diagnosed in the second decade of life with a mean age of 20,65 - when perceptive signs of the syndrome manifests such as the port-wine stains which 50% occurred unilaterally on the right side and 25% on the left side; 25% bilaterally and only 5% no PWS were present. The case 1 reported in our study has an interesting fact due to the late diagnosis - age 30 - even though the patient presented with a right-sided PWS since younger ages. Both of our cases presented with unilaterally PWS on the right side - which is most commonly reported.

Sturge-Weber syndrome is clinically important to the dentist because of the periodontal alterations often present and their higher risk to develop excessive bleeding. The most frequent oral manifestations include gingival hyperplasia[2] - usually the maxillary - and diffuse reddish to purple discolorations throughout the oral anatomic sites usually ipsilateral to the PWS on the face.[1,2,4-17] Also, macroglossia may be presented secondary to an angioma located in the tongue.[9] Furthermore, a periodontal sounding examination must take place because patients with SWS - due to the gingival growth - may develop periodontal pockets[1,5,10] and periodontitis.[8] Carious lesions and calculus or even teeth mobility and malocclusion secondary to poor oral hygiene are also clinical features observed within patients with this condition,[5,6,7,10,11,12] thus enhancing the importance to rigorously follow-up these patients with oral prophylactic measures. Both right maxillary and mandibular teeth were misplaced and significant plaque and calculus deposition secondary to the gingival growth were observed in case 2 of this study.

Sturge-Weber syndrome treatment and its prognosis will depend on the severity of its clinical manifestations - depending on the type.[18] CNS alterations are treated by a medical team and it includes non-invasive methods such as anticonvulsants for seizures, medications for controlling the intraocular pressure in glaucoma and symptomatic treatment for manifestations like headache to even surgeries - including glaucoma surgery, lobectomy or hemispherectomy - and PWS can be treated by laser therapy.[19,20] However, the dentist must integrate the multidisciplinary team that will aid a patient with SWS as periodontal alterations are often observed. Large hyperplastic gingival growth can be surgically excised with an scalpel[5,6,13] electric scalpel[15] (less risk of bleeding) or even by high intensity laser therapy.[13,17] Mild to moderate periodontal alterations could be treated by a non-surgical periodontal treatment like oral plaque control; prophylaxis, calculus removal and sealing and root planing.[5,6,10,11,12,13,14,16] Also, the restoration of the carious lesions and orthodontic treatment for malocclusions must be indicated on some cases.[7] Even though both of our cases did not present any CNS alteration at the moment of diagnosis, they were referred to a medical team to investigate possible alterations. Case 1 did not present any significant periodontal alterations, so no treatment was recommended at that time. However, case 2 was referred to initiate a periodontal treatment at the Periodontology clinic of State University of Rio de Janeiro.

Periodontitis is a condition of the periodontal tissues that results in attachment loss and destruction of alveolar bone - which involves a complex variety of immune system cells and inflammatory cytokines.[21] An increasing number of reports have shown associations between the presence of HLA antigens and the periodontal disease - the upregulation of HLA-DR antigens on epithelial cells at all sites in the oral cavity in patients presenting periodontitis is observed.[22] Therefore, we wonder whether this association could be studied on the periodontal manifestations often observed in the SWS and if it plays an important role in its development and severity.

Conclusion

Sturge-Weber syndrome (SWS) is a rare developmental non-hereditary congenital condition characterized by facial cutaneous vascular nevus in association with venous angiomas in leptomeninges that often manifest periodontal alterations, usually the gingival hyperplasia. The syndrome manifestations may vary and are better treated by a multidisciplinary team - in which the dentist must integrate in order to provide oral health and quality-of-life to these patients.

Acknowledgment

We would like to thank Professor Carlos Marcelo da Silva Figueredo - Department of Integrated Clinical Procedures - Periodontics of State University of Rio de Janeiro - for the reception and treatment of the patients presented in this study after diagnosis.

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Submitted: 08/03/2020 / Accepted for publication: 10/08/2020

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