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Sturge–Weber syndrome in a 6-year-old girl

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Summary. Sturge–Weber syndrome is a congenital disorder characterized by vascular facial birthmarks and neurological abnormalities. Oral cavity involvement may occur, and the extent of the vascular abnormality may vary considerably. The present authors report the case of a 6-year-old girl with Sturge–Weber syndrome, focusing on the clinical and radiographic features. Her dental management involved a multidisciplinary team and included orthodontic treatment using removable appliances.

Introduction

Sturge–Weber syndrome (SWS), or 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 [1,2]. The origin of the disease has been related to a single vascular abnormality arising between weeks 4 and 8 of pregnancy [2]. Clinically affected patients most often present with unilateral cutaneous vascular nevi which follow the divisions of the trigeminal nerve. Neurological alterations, contralateral to the facial cutaneous lesions, are also common, with seizures occurring in 90% of cases. Varying degrees of intellectual disability may also occur [2,3]. Other manifestations of this syndrome include ocular involvement, where angioma of the choroid membrane may produce glaucoma, buphthalmos, loss of vision or contralateral hemiplegia [1,4]. Radiographically, tram-line gyriform calcifications of the ipsilaterally affected side of the brain may be visible [4].

Sturge–Weber syndrome may also show oral manifestations. These include unilateral vascular hyperplasia of the oral mucosa and/or gingival changes ranging from slight vascular hyperplasia to large masses, which may interfere with mouth closure. Affected patients may show ipsilateral hypertrophy of the tongue, macrodontia, and abnormalities in the vascularity of the mandible and/or maxilla. Abnormal ipsilateral dental eruption may result in considerable malocclusion [3–6]. Other radiographic findings include bone destruction, furcation involvement, reduced bone density and a loss of lamina dura over teeth present in affected areas [6].

Because of neurological changes, many patients with SWS find it difficult to maintain adequate oral hygiene, leading to an increased risk of periodontal disease and caries. In addition, the medication usually employed to manage their seizures may lead to gingival hyperplasia. Although the syndrome has been fully described, there is little in the literature about oral and dental care, including orthodontic treatment, for patients with SWS.

The aim of the present report is to describe the clinical and radiographs features in a patient with SWS, and the dental treatment that was provided.

Case report

A 6-year-old white female with SWS was referred to the Oral Diagnosis Clinic (Orocentro), School of Dentistry of Piracicaba, State University of Campinas, São Paulo, Brazil, for evaluation and dental treatment. She complained of bleeding in the upper incisor region. The preliminary examination revealed a bilateral port-wine nevus on her face, and the

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patient's parents reported that this had been present since birth. Her mother also reported that the child experienced seizures, which had started during the first year of life. She had taken carbamazepine (250 mg day^{-1}) since then and had had no further seizures. The patient's medical history also revealed that she suffered bilateral congenital glaucoma as a consequence of the disease, but there was no evidence of either contralateral hemiplegia or intellectual disability.

The diagnosis of SWS had been established when the patient was one year old by analysis of contrast-enhanced magnetic resonance imaging (MRI) and cerebral perfusion scintigraphy. Contrast-enhanced MRI had revealed leptomeningeal angiomatosis involving the right cerebral hemisphere. Cerebral perfusion scintigraphy had shown localized areas of hypoperfusion in the right parieto-occipital region, which was believed to correspond to areas of chronic hypoxia. Radiographically, there was no evidence of typical tram-line gyriform calcifications in the brain.

Extraoral examination of the patient revealed a bilateral facial vascular nevus (port wine stain), which extend from her forehead to the medial third of her face, including the upper lip, following the ophthalmic and maxillary course of the trigeminal nerve (Figs 1 & 2). Intraoral examination revealed red swelling involving the upper anterior gingiva in the region of the central incisors (Fig. 3) and the mucosa of the upper lip (Fig. 4). The child had adequate oral hygiene, with caries of the occlusal surfaces of the lower first and second primary molars, and the lower left first permanent molar. There was marked proclination of the permanent upper central incisors, which prevented lip closure. There was no obvious difference in the tooth eruption pattern between the left and right sides of the mouth. A panoramic radiograph showed the absence of the left upper and lower second premolars, but there was no evidence of any loss of alveolar bone (Fig. 5). Over a series of nine appointments, the patient and her mother were given advice on plaque control through regular and effective tooth brushing, and the carious teeth were restored.

The girl was referred to the Orthodontic Clinic of the School of Dentistry for evaluation and treatment of the malocclusion. Orthodontic assessment showed that the patient was in the mixed dentition phase, with a distal relationship of the primary second

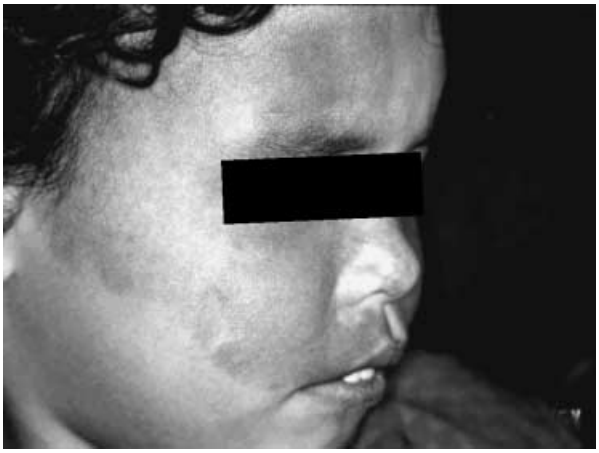

Fig. 1. Cutaneous vascular nevus of the right side of the face that follows the ophthalmic and maxillary divisions of the trigeminal nerve.

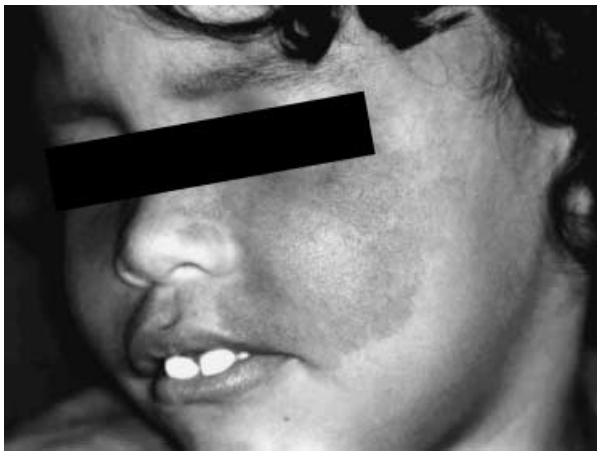

Fig. 2. Cutaneous vascular nevus of the left side of the face that, like that involving the right side (see Fig. 1), follows the ophthalmic and maxillary divisions of the trigeminal nerve, showing bilateral involvement of the facial cutaneous vascular nevus.

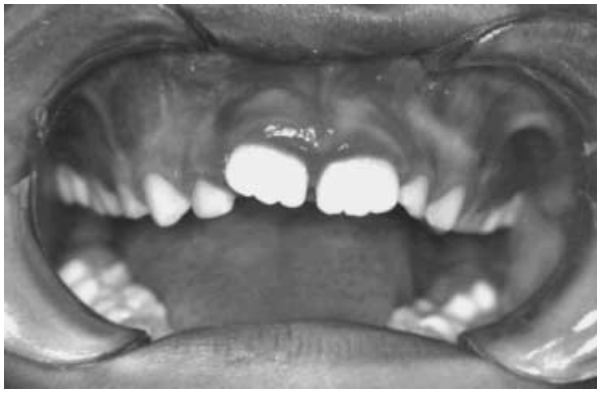

Fig. 3. Marked proclination of the permanent upper central incisors and red swelling on the anterior upper gingiva in the region.

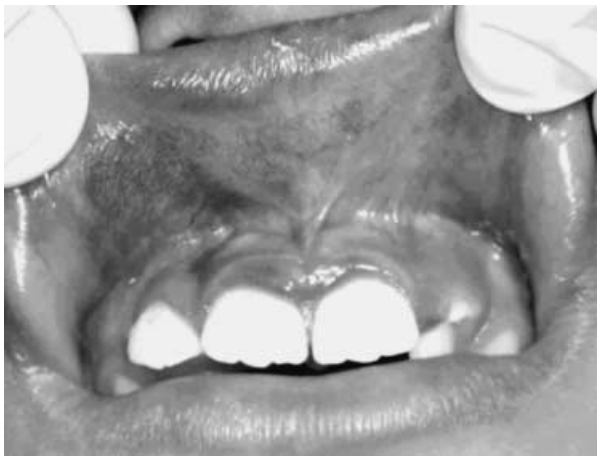

Fig. 4. Red spots with increased superficial vascularization on the mucosa of the upper lip.

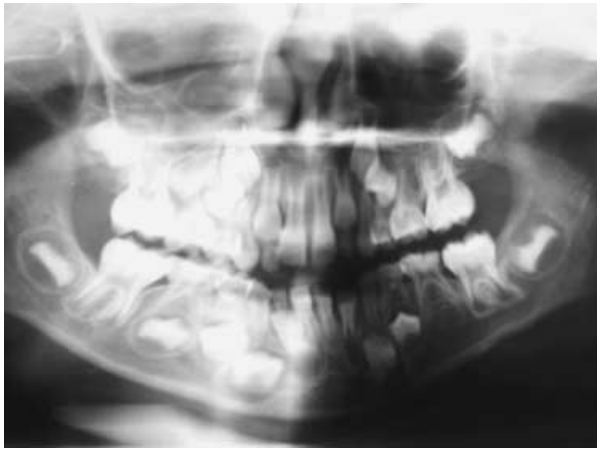

Fig. 5. Panoramic radiograph showing agenesis of the left upper and lower second premolars.

molars and anterior open bite, associated with atypical tongue position and incompetent lip pattern. Radiographic cephalometric examination revealed proclination of the lower incisors (IMPA: 105°) and proclination of the upper incisors with an overjet of 10 mm. Other measurements included SNA (85°). Clockwise rotation of the mandible was estimated to reduce SNB (75°) and ANB was 10°. Treatment was begun using a removable orthodontic appliance consisting of a lingual re-educator with tongue crib (Fig. 6). Subsequently, an appliance with a palatal bar and a lingual re-educator was used to improve the positioning of the tongue and to re-establish a normal swallowing pattern. Following this phase, a functional orthopaedic appliance is presently being used to redirect mandibular growth in an anti-clockwise

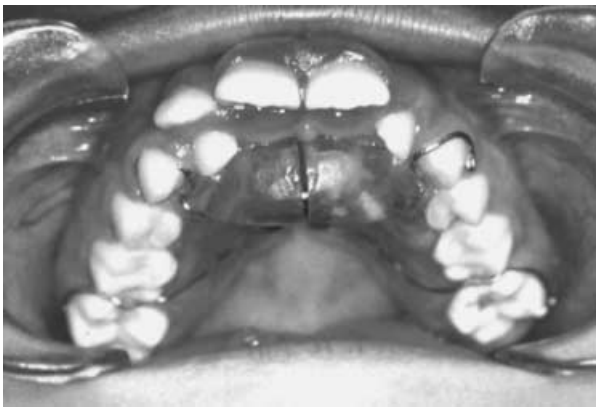

Fig. 6. Removable orthodontic appliance shown in position.

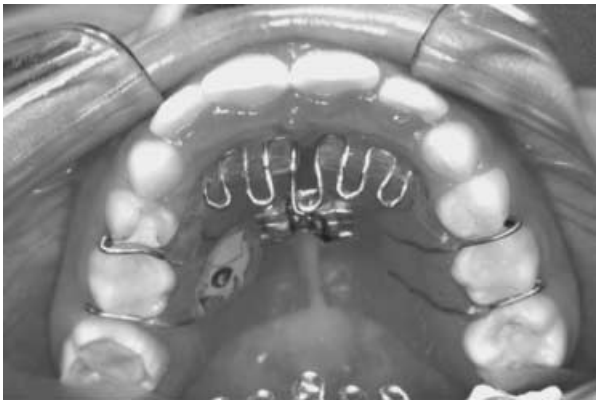

Fig. 7. Clinical view showing improvement of the upper teeth.

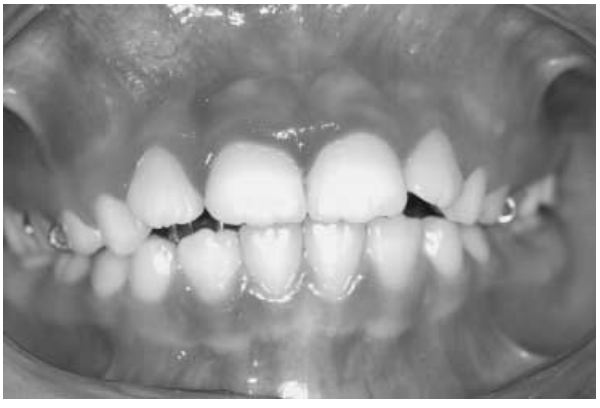

Fig. 8. Frontal view showing better positioning of the teeth.

direction. Currently, the patient is on follow-up. She has shown improvement of the malocclusion, mainly in terms of a decrease in the inclination of the lower incisors (IMPA = 99°), and a great improvement in swallowing, which is approaching normal after 1.7 years of treatment (Figs 7 & 8). The patient

Table 1. Main previous and current cephalometric data.

Cephalometric measurement	Normal (°)	Present case (°)	
		Initial (September 2002)	Current (April 2004)
IMPA (proclination of the lower incisors)	88	105	99
SNA (relationship between maxilla and skull base)	82	85	83
SNB (relationship between mandible and skull base)	80	75	76
ANB (difference between SNA and SNB)	2	10	9

continues to use her functional appliance. Table 1 shows her past and current cephalometric data.

Discussion

In general, SWS presents with a unilateral facial vascular nevus (port wine stain), which is present from birth, following one or more divisions of the trigeminal nerve, and which is associated with ipsilateral venous angioma of the leptomeninges [6,7]. Vascular abnormalities may be bilateral in up to 30% of cases [2]. In the present case, there was a bilateral facial port wine stain following two divisions of the trigeminal nerve, but leptomeningeal angiomatosis was present only on the right side of the brain and was limited to the right parieto-occipital region.

The development of glaucoma, as seen in this case, is thought to be the result of an abnormal plexus of episcleral vessels that results in increased intraocular pressure [8]. Other ocular involvement, such as buphthalmos (enlargement of coating of the eye) and loss of vision, may also occur [1]. Sujansky & Conradi [9] reviewed findings in 52 patients with SWS and reported that 60% presented with glaucoma. The prevalence was higher when the facial areas supplied by both the ophthalmic and maxillary divisions were bilaterally involved. The present report supports this finding because this patient presented with a bilateral facial port wine stain following the ophthalmic and maxillary divisions, and had bilateral congenital glaucoma.

Radiographically, patients typically present tramline gyriform calcifications on the brain, which may be associated with intellectual disability. In the present case, there was no sign of either calcifications or intellectual disability. Radiological advances have greatly improved the ability to characterize the involvement of central nervous system in SWS [1]. Contrast-enhanced MRI may demonstrate cortical atrophy, choroid plexus enlargement, prominence of the deep venous system, and the angiomas of the leptomeninges and eyes [1]. In this case, the method revealed leptomeningeal angiomatosis involving the right cerebral hemisphere. In addition, further information was gained using cerebral perfusion scintigraphy, which demonstrated cerebral perfusion abnormalities in the right parieto-occipital region.

The oral findings related to SWS have varied from one report to another. Gorlin & Pindborg [10] reviewed 111 cases with SWS and found that 38% had some oral manifestations of the syndrome. The most frequently affected sites were the lips and cheeks, which showed a flat red lesion that blanches on pressure. Other sites may also be involved, such as the gingiva, palate and the floor of the mouth [6]. The case reported here presented a red swelling involving the upper maxillary gingiva with frequent spontaneous bleeding and red spots on the upper lip mucosa. When oral surgical procedures are needed in the areas affected by vascular lesions, such as periodontal surgery or tooth extraction, special attention must be given to the risk of severe intra- and postoperative haemorrhage. Special care to prevent and treat complications may include hospitalization, and the use of local anaesthesia with vasoconstrictors, dressings and splints [4,6]. The risks involved help to reinforce the importance of good dental health for affected patients.

Analysing panoramic and intraoral radiographs, some authors have reported that alveolar, periodontal and pulpal structures may be involved, and that alterations in vascularity within the jaws may lead to altered ipsilateral dental eruption sequences, with early eruption of the permanent teeth in affected areas causing malocclusion [4–6]. In the present case, a panoramic radiograph failed to show bone alterations or suggest alterations to dental eruption, except for demonstrating the absence of the left upper and lower second premolars. The present authors believe that this finding may be related to changes in bone vascularity by SWS not shown more directly on radiographs. It is well known

that malocclusion is present in most cases of SWS; however, to the authors' knowledge, this is the first report of documented orthodontic treatment in a patient with this syndrome. Treatment has been successful and has not been complicated by the superficial vascular lesions. However, treatment may be more difficult in the presence of larger and more deep-seated vascular abnormalities.

In conclusion, patients with SWS may present with oral lesions. The dentist has an important role in advising and maintaining good oral hygiene and periodic oral assessment, thereby providing better oral health and an improved quality of life.

Résumé. Le syndrome de Sturge-Weber est une maladie congénitale caractérisée par un hémangione facial congénital et des anomalies neurologiques. La cavité buccale peut être concernée et l'extension de l'anomalie vasculaire peut varier considérablement. Nous rapportons le cas d'une jeune fille de 6 ans présentant un syndrome de Sturge-Weber, en insistant sur les caractéristiques cliniques et radiographiques. La prise en charge dentaire a impliqué une équipe multidisciplinaire et inclus un traitement orthodontique à l'aide systèmes amovibles.

Zusammenfassung. Sturge-Weber Syndrom ist eine angeborene Erkrankung charakterisiert durch einen angeborenen vaskuläre Nävus und neurologische Störungen. Die Einbeziehung der Mundhöhle und der Grad vaskulärer Veränderungen kann erheblich schwanken. Wie berichten einen Fall eines sechsjährigen Mädchens mit Sturge-Weber Syndrom mit dem Schwerpunkt auf klinische und radiographische Befunde. Die Zahnbehandlung beeinhaltete eine multidisziplinäre Herangehensweise einschließlich kieferorthopädischer Behandlung mit herausnehmbaren Apparaturen.

Resumen. El síndrome de Sturge Weber es una alteración congénita caracterizada por una marca facial vascular desde el nacimiento y anomalías neurológicas. Puede estar implicada la cavidad bucal y la extensión de la anomalía vascular puede variar considerablemente. Informamos de una niña de 6 años con síndrome de Sturge-Weber, basado en hallazgos clínicos y radiográficos. El tratamiento odontológico implicó a un equipo multidisciplinario e incluyó tratamiento ortodóncico usando dispositivos removibles.

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