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Case Report

A Rare Case of Encephalotrigeminal Angiomatosis: A Case Report

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Abstract

Aim: Sturge–Weber Syndrome, also known as encephalotrigeminal angiomatosis, is an uncommon, nonhereditary developmental anomaly.
Methodology: A 48-year-old woman presented for routine dental treatment. She had a history of seizures and had a port wine stain on the right side of her face, which followed the distribution of the trigeminal nerve. Skull radiographs revealed "tram-track" calcifications.
Conclusions: The early diagnosis of Sturge–Weber Syndrome requires a multidisciplinary approach. Oral health care professionals need adequate knowledge and understanding of the disease process to help diagnose and treat these patients.

Keywords: Encephalotrigeminal angiomatosis, port wine stain, tram-track calcifications

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Introduction

Sturge–Weber syndrome, also known as encephalotrigeminal angiomatosis, is an uncommon, nonhereditary developmental anomaly (1). Although the exact etiology is not known, it has been suggested that the main defect is a developmental injury that affects the precursors of tissue originating in the promesencephalic and mesencephalic neural crest, which leads to the various manifestations (2). It is classified with the phakomatoses (mother spot disease), and has no sex predilection (3).

The main feature is facial angiomatosis (nevus flammeus) with variable distribution along the dermatomes of the branches of the trigeminal nerve (5), most commonly the ophthalmic division (4). Other features include venous angiomas in the leptomeninges occurring on the cerebral cortex, the presence of ipsilateral gyriform calcification, which leads to seizures and contralateral hemiparesthesia, cognitive delay, and glaucoma (6). The most frequent neurological manifestation is epilepsy, which occurs in 75~90% of the cases. Ocular angiomas occur in approximately 30% of the cases, affecting the choroids and sclera. It is usually ipsilateral, like the cutaneous lesions (7). Generalized tonic–clonic motor convulsions are more prevalent in younger patients, peaking at 4–6 months of age (7).

Intra-oral manifestations include hypervascularity of the ipsilateral mucosa and

gingival hyperplasia or hemangiomatous proliferation that often resembles a pyogenic granuloma in its appearance (8). The gingival hyperplasia is attributed to the vascular component of the disease, the use of anti-epileptic drugs such as phenytoin, or both. Angiomatous lesions can also occur in the buccal mucosa (5). The oral lesions are usually unilateral and do not cross the midline (9). Rarely, alveolar bone is destroyed. Other rare oral manifestations include macroglossia and hypertrophy of the maxilla (7).

Case Report

A 48-year-old woman visited the Department of Oral Medicine and Radiology, Pushpagiri College of Dental Sciences, Thiruvalla, Kerala, South India, for routine dental treatment. She had a history of seizures, for which she was taking medication. The extra-oral examination showed a diffuse port wine stain on the right side of her face measuring 6 × 7cm (Figs. 1 and 2). She stated that it had been present from birth. No such skin lesions were found elsewhere on the body.

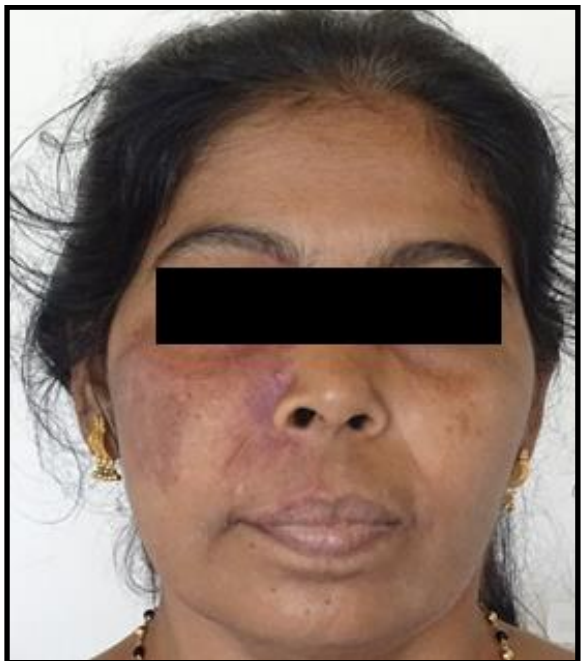

Figure 1. Extraoral view.

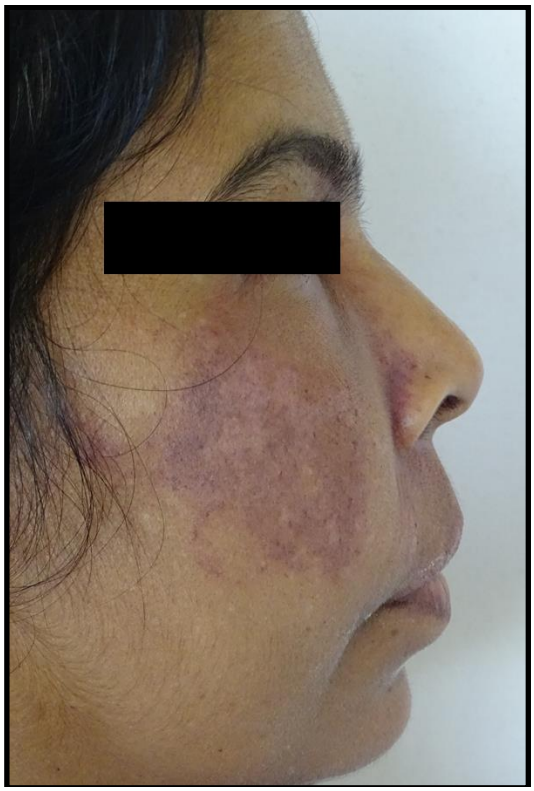

Figure 2. Appearance of diffuse port wine stain.

The intra-oral examination showed unilateral vascular involvement of the right buccal mucosa, right side of the maxillary and mandibular gingiva, and hard palate, extending to, but not crossing, the midline (Figs. 3 and 4). No gingival hyperplasia was observed. The left buccal mucosa, left maxillary and mandibular gingiva, and left side of the hard palate were normal. Diascopy was positive. A posteroanterior view of the skull showed tram-track calcifications (Fig. 5). Routine blood tests were within normal limits.

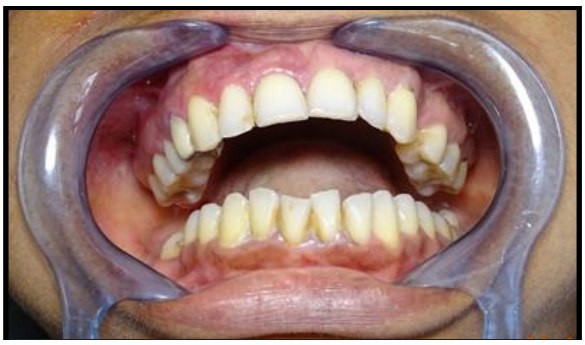

Figure 3. Intraoral view.

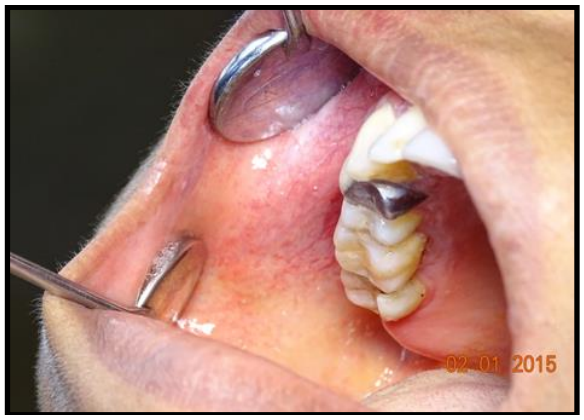

Figure 4. Intraoral view of right buccal mucosa.

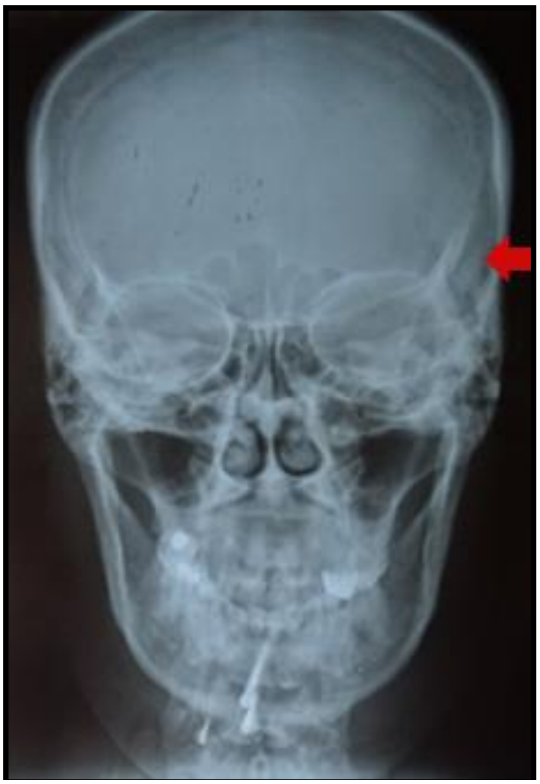

Figure 5. Posteroanterior view of the skull

Discussion

Sturge–Weber syndrome is usually not life threatening (10). The port wine stains are a congenital malformation of the dermis involving the capillaries, venules, and perivascular nerves (11). The majority of such lesions are on the face. Ocular manifestations, cognitive delay, and seizures may be present due to the vascular involvement of the eye and central nervous system (11). Sturge–Weber syndrome is progressive and is associated with neurological decline, but the patient's quality of life can be improved with regular monitoring and symptomatic treatment. The prognosis is variable, and the overall life expectancy is thought to be normal (4).

Oral manifestations occur in around 40% of cases and include gingival overgrowth and asymmetrical jaw growth (11).

A thorough history and physical, neuropsychological, and radiological examinations are required. Intra-oral radiographs may reveal vertical and horizontal bone loss with loss of the lamina dura (7). Skull radiographs may reveal gyriform calcifications resembling a tram track (5). Cerebral disturbances such as intracranial calcifications and cerebral atrophy can be visualized with computed tomography. Magnetic resonance imaging (MRI) and angiography are good for detecting the presence of cerebral vascular malformations. MRI is useful for detecting leptomeningeal angiomatosis and cerebral parenchymal and venous abnormalities. Ocular manifestations may be observed on a thorough retinal examination and with ocular ultrasonography (7).

The differential diagnosis includes Klippel–Trenaunay–Weber syndrome, Bannayan–Riley–Ruvalcaba syndrome, Osler–Weber–Rendu syndrome, Cobb syndrome, and Divry–Van Bogaert syndrome (2).

The treatment is usually symptomatic, involving control of seizures with anticonvulsants. Surgery is indicated for refractory cases of seizures and glaucoma (2). Other treatments include sclerotherapy, surgical resection, laser photocoagulation, or a combination of these (1). Invasive dental procedures in the affected area may be associated with an increased bleeding tendency. The management of these patients can challenge the dentist. Necessary precautions should be taken in case any complication arises, and a suitable armamentarium for hemostasis must be readily available (2).

Conclusions

The early diagnosis of Sturge–Weber syndrome requires a multidisciplinary approach. Oral health care professionals need adequate knowledge and understanding of the disease process to help diagnose and treat these patients. Care must be taken during the dental management of these patients to prevent hemorrhagic complications.

Acknowledgments

The authors deny any conflicts of interest related to this study.

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