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DENTAL TREATMENT OF A STURGE-WEBER PATIENT

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ABSTRACT

AIM

The aim of this study is to present the dental treatment under general anaesthesia of a patient with Sturge-Weber Syndrome (SWS) at the Dental Unit for Patients with Special Needs of an urban Hospital. Sturge-Weber syndrome (SWS) is a rare congenital neurocutaneous disorder. It is characterized by the presence of facial port wine stains, neurological abnormalities and mental retardation, ocular disorders, oral involvement and leptomeningeal angiomas.

METHODS AND RESULTS

The patient, diagnosed with SWS at birth, exhibited no cooperation with the dentist due to severe mental retardation. He also suffered from epilepsy and was receiving special antiepileptic medication. For the above mentioned reasons the dental team decided to proceed with the dental treatment under general anesthesia in the operating theatre. The patient presented swollen and inflamed gums, factors that made the dental treatment difficult to perform. After the dental team assessment, fillings, removal of tartar and extractions were carried out.

CONCLUSION

Good oral hygiene and regular dental follow-ups are critical factors for the prevention of oral and dental diseases. The degree of mental retardation of these patients and therefore

their cooperation with the dentist, determines the form of the dental treatment approach.

DENTAL TREATMENT OF A PATIENT WITH STURGE-WEBER SYNDROME: A CASE PRESENTATION

Key words: Sturge-Weber syndrome, dental treatment under general anesthesia

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Introduction

Sturge-Weber syndrome (SWS) is a rare non-inherited congenital neuro-oculo-cutaneous disorder characterized by an intracranial vascular anomaly and leptomeningeal angiomatosis (1) It belongs to the group of phakomatoses and is recognized by the characteristic congenital "port-wine stain"(PWS) located in the frontal area around the eyes. It is caused by a somatic activating mutation in GNAQ[1].

Seizures are the most common neurological manifestation and typically present in the first months of life. Glaucoma may be present at birth or develop later.

Other clinical features of affected individuals are:

• Muscle weakness or paralysis on one side of the body (hemiparesis)

[1] GNAQ gene provides instructions for making a protein called guanine nucleotide-binding protein G(q) subunit alpha (Gαq)

• Epileptic seizures, often resistant to antiepileptic therapy.

• Developmental disorders.

• Glaucoma.

The SWS patient can be recognized by the distinctive usually unilateral port-wine stain, with a distribution through the 1st and 2nd branches of the trigeminal nerve. About one-third of patients have bilateral appearance. The color of the skin lesion at birth varies from light pink to deep purple and is due to capillary overabundance. There is also a malformation of the blood vessels in the brain's meninges, which is lateral to the skin lesion, that causes tissue to clog and loss of nerve cells in the brain cortex. Abnormalities of the facial and brain arteries cause disorders in the blood flow and ischemia of the affected areas (1) (2). The syndrome is usually accompanied by migraine headaches or mere headaches and seizures that begin in infancy and usually worsen over the years. Epileptic seizures affect the opposite side of the skin lesion and present with varying degrees of severity and often do not respond to medication. Muscle weakness, hemiplegia, hemiparesis on the opposite side of the skin lesion can also occur (3).

Many patients have developmental and mental retardation (4). About 50% of people with the syndrome develop glaucoma at birth or later. Glaucoma can damage the optic nerve. Glaucoma

usually affects the eye on the same side as the PWS. The same eye may became extremely enlarged (bupthalmos) (2).

Diagnosis of Sturge-Weber syndrome is usually easy with the evaluation of skin symptoms and the presence of neurological and / or ophthalmic damage (5).

Radiologic findings of the brainstems that are visible in plain radiography as 'tram tracks' are characteristic. Brain atrophy is visible on axial and on magnetic resonance imaging. Brain angiography usually confirms the slowdown of cerebral blood flow. Functional brain imaging with PET or SPECT shows reduced metabolic activity and blood flow to the area under the vasculature (6).

The SWS occurs in 1 / 20,000 to 1 / 50,000 live births in males and females respectively (7).

The syndrome is thought to be due to a developmental abnormality of the facial and brain arteries that cause disturbances in the blood flow to the brain and ischemia of the affected areas. In particular, it results from errors in the embryonic development of the mesoderm and epidermis (8).

Three major forms of the syndrome (9) have been identified:

• Type 1, where both meningeal and skin vasculature coexist, along with ocular manifestations. This is the most common type of syndrome

• Type 2, where there is only facial skin irritation without the involvement of the brain, whereas ocular manifestations may exist.

• Type 3, where leptomeningeal angioma involvement is exclusively observed. The facial angioma is absent and glaucoma rarely occurs. This type is only diagnosed via brain scan.

The following disorders have a clinical presentation similar to that of Sturge-Weber syndrome (SWS) and must be included in the differential diagnosis:

- Klippel-Trenaunay-Weber syndrome

Kippel-Trenaunay-Weber Syndrome (features of this syndrome include PWS at the extremities)

- Rendu-Osler-Weber (vascular malformations observed throughout the body)
- Maffuci (asymmetric hemangiomas appear as pressure-disappearing nodules)
- Beck-Wiedman (presented with PWS-like capillary malfunction and upper eyelid) (10) (11) (12).

The treatment of the syndrome is considered difficult due to its complexity. Symptoms are usually treated symptomatically. Standard treatment for Sturge-Weber syndrome includes laser treatment for the Port-wine stain, anticonvulsants for the seizures, and medical or surgical treatment for the glaucoma. As regards glaucoma, prognosis depends on the extent of leptomeningeal involvement and the severity of the glaucoma. It should be noted that there is no specific treatment for the

syndrome except for seizures. In the most severe cases of seizures and at a young age, a functional hemispherectomy is suggested where some parts of the subdural hemisphere are removed and the neural pathways that connect it to the other hemisphere are interrupted to reduce the number and severity of the hemisphere (13). Neurological and ocular disorders can be treated with a combination of pharmacological and surgical treatment, ie, low dose aspirin, antiepileptic drugs and surgical treatment. Chronic aspirin use has been suggested as a means of limiting recurrence of cerebral thrombosis. Study to inhibit the mechanism of mutation induction will be a new future treatment for the syndrome (3).

The clinical picture varies in patients with the syndrome. In its typical form, the facial skin vasculature and its mucosal manifestations in the oral cavity are evident.

The classic oral manifestations involve haemangiomatous gingival lesion limited on the same side of upper or lower jaw.

Poor oral hygiene combined with the anti-epileptic treatment that many of these individuals pursue causes gum inflammation and hyperplasia. The dentist needs to be aware that hemorrhage may occur laterally.

Presentation of the clinical case

A 26-year-old male with Sturge-Weber Syndrome presented at the Dental Unit of an urban Hospital, accompanied by his brother and aunt.

We were told that the syndrome was diagnosed shortly after the patient's birth, mainly due to the characteristic hemangioma in the right half of the face extending from the frontal area, around the eye across the almost nearly lower jaw (Fig. 1). Most probably our patient belongs to type 1 of the syndrome. Upon receiving his medical history, we were informed by his relatives that the patient had severe mental retardation (I.Q. <30), psychomotor retardation, and epileptic seizures. Epileptic seizures began in infancy. The patient is receiving valproic acid and risperidone and his seizures and behavior are well controlled.

The patient had never visited a dentist before and when relatives were asked what he uses for his daily oral hygiene they said they did not know because the patient was staying at an institution.

Due to the complete lack of cooperation with the dentist probably because of his severe mental retardation and seizures, the dental and medical team decided to treat the patient under general anesthesia (GA) in the operating theatre.

Instructions from the anesthesiologist included the administration of oral antiepileptic drugs in the morning of the surgery with a minimal amount of water.

Following routine preoperative screening and anesthesia evaluation, GA was administered with special precautions particularly to prevent seizures. Anesthetic agents that could cause Central Nervous System stimulation (i.e. convulsions) were excluded.

As in all patients receiving general anesthesia, oxygenation, acid-base balance, depth of anesthesia, analgesia and muscle relaxation were maintained at the desired levels. The overall duration of the dental treatment was 2 hours and 5 minutes. In more detail, two simple amalgam fillings (teeth 16 and 48), two composite resins (teeth 46 and 47), five extractions (teeth 14,15,17,28,36) gingivitis treatment and topical application of fluoride were done. A gentle and smooth recovery followed the surgical procedure. Due to the adoption of the appropriate measures, no complications occurred preoperatively, intraoperatively or postoperatively (14).

Discussion

Diagnosis of Sturge-Weber syndrome is based on the characteristic clinical picture and usually easy to assess with skin symptoms and typical neurological and radiological findings (3).

Intra-oral manifestations of SWS include small or large vascular lesions in the gums. Intra-oral angiomas occur more frequently in the oral mucosa. The severity of the lesions varies from a small vascular hyperplasia to an oversized

vascular lesion. Port-wine stains (PWS) are found in the oral mucosa accompanied by vascular changes. Gum bleeding after a minor injury is a characteristic finding of the syndrome. Occasionally, hypertrophy of the upper jaw is observed, resulting in facial asymmetry. Vascular damage to the lips, the mucosa of the oral cavity, tongue and palate is seen in about 38% of cases. The manifestations in the oral cavity are bilateral and stop in the midline (15) (16).

Inflammation of the palate is less common. Inflammation of the tongue may also be accompanied by semi-hypertrophy. The gum lesions can range from a slight vascular hyperplasia to monstrous hyperplasia that interferes with the occlusion of the teeth. The vascular hyperplasia of the gum (blanches on pressure) should be differentiated from the fibrous hyperplasia. Multiple purulent granulomas have been reported and concomitant hypertrophy or alveolar cleft palate. Anomalies at the time of teeth eruption and the shape of the teeth (most often large teeth) have also been observed, resulting in abnormalities in the occlusion of the affected side (5).

In our case, the patient's gums were inflamed and with hyperplasia. It was observed that on the right side, corresponding to the presence of the vasculature, significantly more bleeding during scaling and extraction of the teeth than the left side was observed.

Our clinical experience of increased tooth decay prevalence in people with mental retardation is consistent with similar studies where it is found that tooth decay and periodontal diseases' rate in children and adults with intellectual disabilities is statistically significantly higher than in the mentally healthy age group (17). Consuming a lot of carbohydrates in patients' diet combined with poor oral hygiene can further increase tooth erosion and caries attack (18). This patient lives since his early childhood in an institution where the level of his oral hygiene cannot be monitored. This is likely to be irregular to non-existent. It is also difficult to control our patient's eating habits. The patient's antiepileptic treatment in combination with poor oral hygiene caused worsening gum inflammation and hyperplasia. It was observed that on the right side, corresponding to the presence of the vasculature, significantly more bleeding during the extraction of the teeth than the left side was present. Although this syndrome has been extensively studied, there are not many studies regarding the oral care of people with SWS. Periodic oral examination and maintaining good and meticulous oral hygiene are necessary to prevent any complications due to lesions in the mucosa of the oral cavity. In addition, severe gingival hyperplasia causes difficulty in feeding and inability to perform proper oral hygiene, making these patients susceptible to oral cavity infections and thus deteriorating general health. The dental

treatment of the patient with SWS is a difficult task for the dentist due to the increased risk of bleeding (19) (20).

Conclusions

The oral condition of patients with Sturge-Weber's syndrome worsens if oral hygiene is poor and especially if anti-epileptic treatment is taken, often causing inflammation and hyperplasia of the gums. Good oral hygiene and frequent re-examinations help to prevent dental problems, which is especially important for overweight patients whose treatment with GA presents additional risks and requires special measures.

The degree of mental retardation, and therefore the co-operation of these individuals, determines how they will be treated either at the outpatient dental clinic or under GA in the operating room. However, special precautions must be taken in the case of treatment under GA. The dentist should thoroughly check for bleeding to avoid postoperative adverse events that are impossible to handle when the patient does not cooperate. The key to effective treatment lies in alleviating the intensity of the complications. Early diagnosis and treatment are the best options for improving the quality of life of the SWS patient. However, it is very difficult for people with mental disabilities to properly apply oral hygiene. Additionally when a person resides in an institution away from his relatives, it is extremely difficult to assess

his/her oral hygiene and eating habits, two crucial factors for the prevention of tooth decay and periodontal disease(21) (22).

To our knowledge, this is the first presentation of a case of SWS patient dental treatment under GA to be published in our country.

The challenge for the dentist is to be aware of the Syndrome’s oral manifestations and any other general factors that can impact on the dental management. The dentist cooperation with other medical specialties is crucial for the safe provision of dental treatment. The key in the management of SWS is to prevent or reduce the intensity of complications as the underlying pathology cannot be treated. The early dental intervention is of utmost importance for the improvement of quality of life of SWS patients.

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