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Case Report

SURGICAL REMOVAL OF AN IMPACTED THIRD MOLAR IN A PATIENT WITH STURGE WEBER SYNDROME – A RARE CASE REPORT AND REVIEW OF LITERATURE

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A B S T R A C T

The association of the classic port-wine stain of the face with ipsilateral atrophy and calcification of the cerebral cortex is known as encephalotrigeminal angiomatosis or Sturge-Weber syndrome. Port-wine stains are a subgroup of capillary hemangiomas that usually involves the face and represent a cosmetically devastating disfigurement affecting many people.Other common findings of this syndrome are mental retardation, epileptic seizures, contralateral hemiparesis or hemiplegia, and congenital glaucoma. Intraorally, the lips, gingiva, tongue and buccal mucosa appear to be affected and exhibit vascular hyperplasia, hypertrophy and a red purplish color. Hard tissue involvement consists of bone overgrowth and tooth mal-eruption. Sturge Weber syndrome patients may or may not exhibit seizures, mental retardation based on the severity of the syndrome. Risk of massive haemorrhage is a constant threat while performing intraoral surgical procedures in such patients. The following case report highlights the case of a patient with Sturge Weber syndrome with a symptomatic impacted mandibular third molar which was surgically extracted under local anaesthesia while taking all necessary precautions. There was minimal bleeding intra and post-operatively and no post-operative complications were encountered.

Detailed clinical examination, investigations and adequate precautions should be taken during a surgical procedure to minimize any potential complications and manage these effectively.

INTRODUCTION

Sturge-Weber syndrome (SWS) is a sporadic neurocutaneous disorder characterized by a classic triad of facial capillary malformation, ipsilateral leptomeningeal angioma, and vascular eye abnormalities [1]. Also known as encephalotrigeminal angiomatosis, it is reported to affect 1 in 50,000 live births in the US [2]. The classic pathognomonic features of this disease are venous angiomas of the leptomeninges extending over the cerebral cortex, with ipsilateral angiomatous lesions; unilateral facial nevus (hemangioma) after one or more divisions of the trigeminal nerve; and epileptic convulsions (contralateral focus)[3].

SWS is a developmental anomaly of embryonic origin because of malformation in mesodermal and ectodermal development. It occurs due to a failure of regression of a vascular plexus around cephalic portion of neural tube which is destined to become facial skin. This vascular plexus normally forms at 6thweek of intrauterine life.

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In the normal course, it regresses by the 9thweek. Failure of its regression results in residual vascular tissue which forms angiomas of leptomeninges, face and the ipsilateral eye [4].The following case report describes the surgical extraction of a symptomatic impacted lower third molar in a patient with SWS.

CASE REPORT

A 26 year old male patient, reported to the Department of Oral and Maxillofacial Surgery with a chief complaint of pain in his lower right back tooth region since the past 10 days. The patient gave a history of being diagnosed with Sturge Weber Syndrome and a history of glaucoma in the right eye. There was no history of any episodes of epilepsy. On the date of examination of the patient, the patient was not consuming any medications and the pain was dull in nature and aggravated during mastication. The general examination revealed a normal mentation.

Clinical examination revealed the presence of a lightly pigmented port wine stain in relation to the right side of the face extending periorbitally around the right eye and extending inferiorly upto the ala tragal line. Blanching of the stain was positive. Findings of dilated episcleral and conjunctival vessels

were noted in the right eye. Vision was preserved. Examination by an ophthalmologist confirmed the presence of glaucoma in the right eye. Intraorally, well defined palatal, buccal and gingival erythema could be seen in relation to the 1st quadrant of the jaw. The palatal erythema did not extend beyond the midline and was restricted to the soft palate. Examination of the tooth in question revealed a deeply carious impacted 48 with associated chronic cheek bite. The tooth was slightly tender on percussion with no evidence of pus discharge.

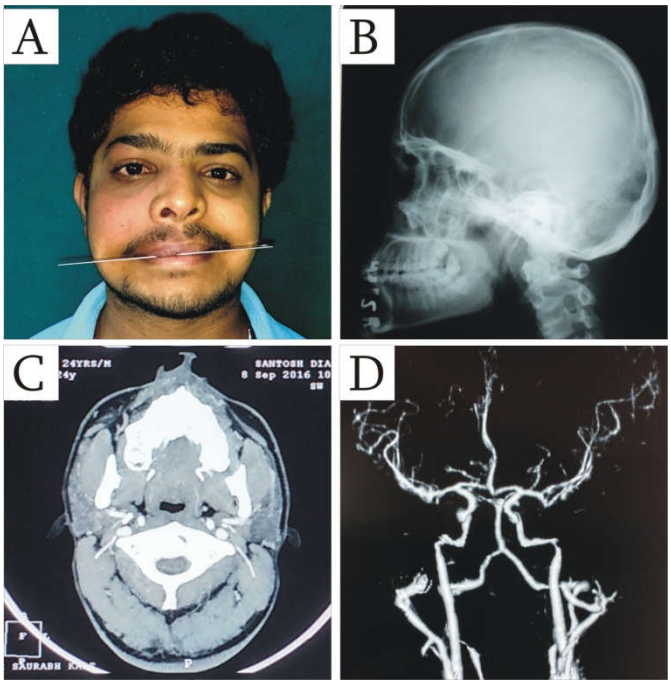

Figure 1 A –Clinical Frontal Photograph of the patient exhibiting the port wine stain in the right periorbital region and occlusal cant in relation to the maxilla. B – Lateral skull radiograph without any evidence of tramline calcifications. C – Axial view CT scan with contrast demonstrating no vascular malformation. D – 3D reconstruction of the head and neck vessels.

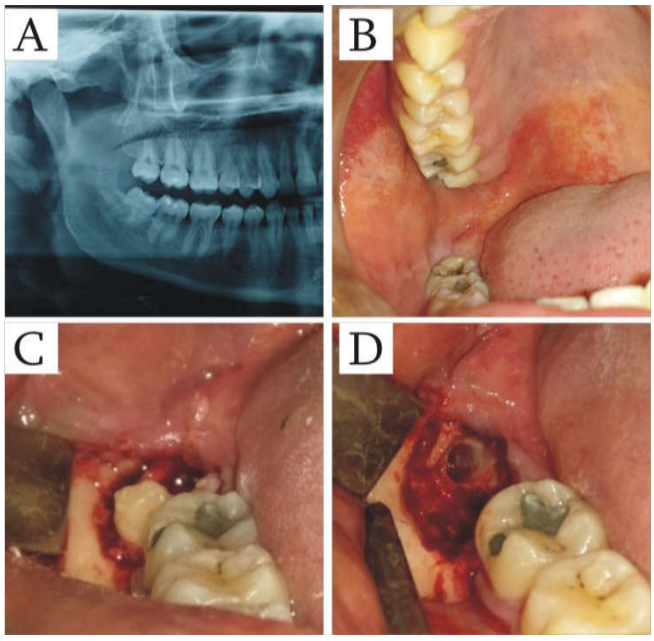

Figure 2 A – Pre-operative OPG showing the mesioangular impacted 48 with roots in close proximity to the inferior alveolar canal. B – Intraoral clinical photograph showing the increased vascularity in realtion to the soft tissues surrounding the 1st quadrant. C – Wards incision without a distal incision and flap elevation. Socket post extraction of 48.

A clinical diagnosis of apical periodontitis in relation to impacted 48 was given and surgical extraction was advised. OPG, CT angiogram and routine blood tests were advised. All the lab investigations were in the normal range. The OPG revealed the presence of a mesioangular 48 with close proximity of the roots to the inferior alveolar canal. While the distal root exhibited darkening, the mesial root exhibited a dilaceration in the distal direction. The radiographic examination of the skull did not show any characteristic tram line pattern. The CT angiogram of the head and neck performed with and without contrast failed to exhibit any significant abnormality. There was no arterio-venous malformation, aneurysms noted in the angiogram. The patient was deemed fit for a surgical procedure. However, evaluating the potential risks involved and the possible need for embolization or external carotid artery ligation in the event of massive uncontrolled haemorrhage, the patient was operated upon in a hospital setting under local anaesthesia. Local haemostatic agents were arranged prior to beginning the procedure. Informed consent was sought. To minimize tissue injury and the risk of bleeding, a wards incision without a distal releasing limb was given after administering an inferior alveolar nerve block and a buccal nerve block. A full thickness mucoperiosteal flap was raised. To minimize risk of injury to the inferior alveolar canal, the tooth was sectioned longitudinally using a bur under abundant irrigation. The distal half of the crown was removed first, followed by the removal of the mesial crown and root and lastly the distal root. There was no significant bleeding encountered. The socket was irrigated using butadiene and a gelatin sponge was placed to aid in haemostasis. The wound was closed using horizontal mattress sutures. The patient was reviewed on the 7th post-operative day for suture removal and no bleeding episodes were recorded during this period.

DISCUSSION

Sturge Weber syndrome was first described by Schirmer in 1860 in association with facial angioma and buphthalmos. Later in 1879, William Allen Sturge while reporting a young girl, acclaimed these features in accordance with neurological findings and termed it as Sturge Weber syndrome [5]. Bergstrand, in 1936, differentiated this syndrome into three types based on its severity [6]. In 1992, Roach categorized SWS variants into three types: Type I: individual with a facial port wine stain (PWS), leptomeningeal angioma, and who may have glaucoma, Type II: individual having a facial PWS, no leptomeningeal angioma, and who may have glaucoma and Type III: individual having leptomeningeal angiomatosis, no facial PWS, and rarely, glaucoma [7]. Due to the presence of the port wine stain and glaucoma in the right eye but absence of episodes of seizures or any significant findings on skull radiographs or CT angiogram, our patient was categorized as Type 2 according to Roach scale.

The most common clinical features (with approximate frequencies) are convulsive disorder (80%), dermal angiomas (76%), abnormal radiographic findings (63%), mental retardation (54%), ocular involvement (37%), and hemiplegia (31%) [3]. Control of epilepsy is the major goal in treatment of SWS patients. Medical treatment with various anticonvulsants has been attempted with varying results. Cases refractory to medical therapy may need surgical intervention [8]. Our patient did not give any history of seizures in the past.

Cutaneous capillary malformations in SWS are often referred to as port wine stains (PWS) or nevus flammeus (a well-demarcated red macular stain present since birth). Facial PWS may involve only the forehead and upper eyelid following the path of the ophthalmic branch of the trigeminal nerve (20%) ormay show involvement of the maxillary or maxillary and mandibular branches. Our findings are consistent with this [2]. Our patient showed a port wine stain along the distribution of the right ophthalmic and maxillary nerve division. Owing to its potential psychological impact, some clinicians advocate its removal/ lightening. Several methods to achieve this include dermabrasion, tattooing and lash lamp pulse tunable dye laser therapy. Cryosurgery may be considered for treatment of lip and other soft tissue lesions [9]. The hue of the stain was not as deep as often documented and the patient hence did not seek lightening or its removal.

Glaucoma and choroidal hemangioma are the most common ocular manifestations. Both the conditions are usually ipsilateral to the facial PWS. Increase in the episcleral venous pressure and developmental anomalies in the anterior chamber angle have been considered to be the main causal factors for the glaucoma associated with SWS. Other ocular abnormalities that have been reported in SWS include dilatation and tortuosity of conjunctival and episcleral vessels, buphthalmos, iris heterochromia, optic disc coloboma and cataract. Control of glaucoma may be achieved with medical treatment alone (beta-blockers and carbonic anhydrase inhibitors) or via surgery [10].

Intraorally, the most common feature is the hemangiomatous lesion, usually restricted to the ipsilateral maxilla, mandible, floor of the mouth, lips, cheeks, palate and tongue. The affected sites may exhibit vascular hyperplasia, hypertrophy and a red purplish color. Hard tissue involvement consists of bone overgrowth and tooth mal-eruption. Such lesions tend to bleed profusely when traumatized. Macroglossia and hypertrophy of the maxillary bone, found in some patients, might result in malocclusion and facial asymmetry.Our patient showed a significant occlusal cant on the affected side. However no malocclusion was noted.

Dental management of these patients is often complicated by the history of seizures, making dental management further difficult. The concurrent use of anticonvulsant therapy with resultant drug induced gingival hyperplasia further compromises the oral hygiene. A number of cases reported in literature comment on the periodontal and surgical management of such patients, ranging from simple extractions to gingivectomy. A report by Deeb M *et al* described a case of SWS which underwent anterior segmental osteotomy without any intra/ post-operative complications [11]. Another report by Caiazzo *et al* reported on performing full mouth extractions in a patient of SWS using pre-operative embolization of the internal maxillary artery, greater palatine and the superior alveolar artery with minimal blood loss of 200 ml [3].

Management of patients of SWS with symptomatic impacted third molars is scarce in the literature. Our patient exhibited clinical signs of erythema extending on the right side of the palate and the maxillary right buccal vestibule. To avoid complications related to the extraction of the impacted 48, a wards incision without a distal extension was given to minimize the risk of haemorrhage due to accidental nicking of the dilated vessels.

The use of lasers has often been advocated in the management of such patients to minimize bleeding. Advantages associated with lasers are haemostasis, sterility, minimal postoperative swelling and reduction in postoperative pain [2].

CONCLUSION

Treatment of patients with SWS poses a challenge to dental health professionals and they must always be mindful of the possible risks associated with each clinical procedure. Suitable hemostatic agents and emergency equipment must always be readily available when treating these patients. Special attention must be given to behavioral management of these patients especially those with a history of epilepsy or mental retardation and kids. Routine procedures must be performed as atraumatically as possible while surgical procedures, if necessary, must preferably be performed only in a hospital setting.

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