STURGE-WEBER SYNDROME IN ASSOCIATION WITH PYOGENIC GRANULOMA

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Abstract

Sturge-Weber syndrome is a rare nonhereditary developmental condition with neurological and skin disorder, characterized by presence of Port wine stain on the face along with ocular disorders, oral manifestations and leptomeningeal angiomas. Pyogenic granuloma or granuloma pyogenicum is a well known oral lesion characterized by localized granulation tissue overgrowth. Pyogenic granuloma of the oral cavity is known to involve the gingiva commonly. The present article reports a case of association of pyogenic granuloma in a patient suffering from Sturge-Weber syndrome.

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Introduction

Sturge-Weber syndrome or encephalotrigeminal angiomatosis (OMIM – 185300) is an uncommon nonhereditary developmental condition with neurological and skin disorder. The Port wine stain is a malformation of the dermis that involves venular, capillaries and possibly perivenular nerves [1].

It occurs in an estimated three per thousand births [2]. Most lesions appear on the face and approximately 5% of patients have associated ocular involvement, mental retardation and seizures due to involvement of vasculature of eye and central nervous system[3].

Oral changes occur in 40 % of cases of this syndrome and may include massive growth of the gingiva and asymmetric jaw growth [4].

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Case Report

A 12 year old boy was reported to Department of Pedodontics, SPPIDMS Dental College, Lucknow, India with chief complaint of gum enlargement and bleeding while brushing.

On extra oral examination bluish red discoloration suggestive of Port wine stain was noticed on left and right side of cheek, forehead, nose and left ear (Fig: 1).

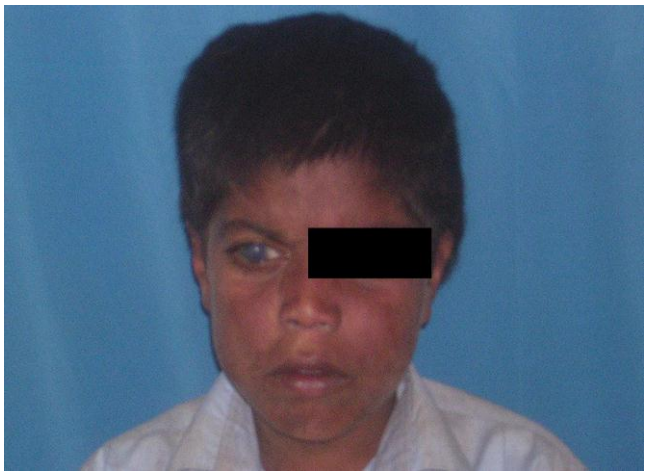

Figure 1. Photograph of patient showing Port wine stains on face and blindness of right eye.

The lesion was present since birth and shows gradual enlargement. None of the family members had similar abnormalities. Medical history revealed that patient is under medication for convulsion (Carbamezapine). There was history of swelling on right side of eye at the age of 3 months and which resulted into blindness at the age of 1 year (Fig:1). Detailed examination of the patient as well as imaging test done to know the involvement of meningeal angiomatosis showed no positive findings.

Intra oral examination revealed, port wine stain on upper and lower lip, gingival hyperplasia on right side of maxillary arch. Pedunculated soft tissue growth was noticed between upper central incisors region suggestive of Pyogenic granuloma (Fig: 2), which is present since 3-4 months as stated by patient. There was history of gingival bleeding during brushing. Blood investigation revealed normal bleeding and clotting time.

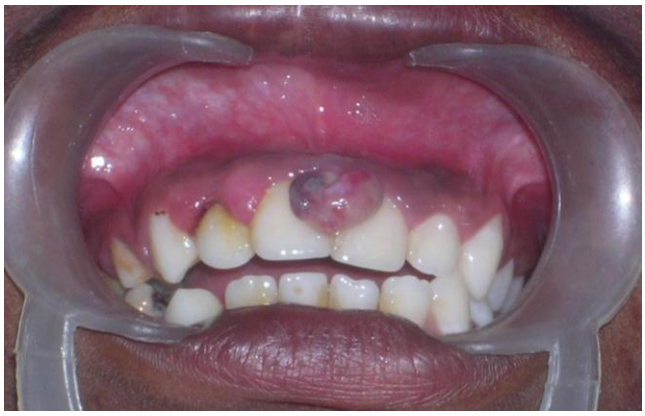

Figure 2. Photograph showing gingival hyperplasia of maxillary right region and Pyogenic granuloma between maxillary central incisors.

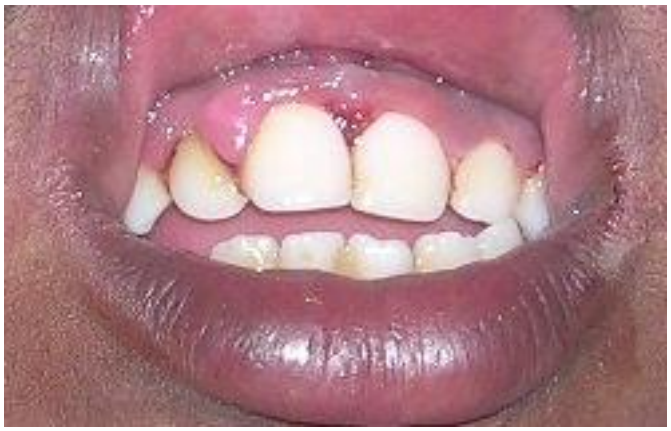

Figure 3. Maxillary incisor region after excision of pyogenic granuloma.

Pyogenic granuloma was surgically excised under local anesthesia and the bleeding was controlled using pressure pack (Fig: 3). In subsequent visits oral prophylaxis, restoration of carious teeth and oral hygiene maintenance instructions was carried out.

Discussion

Sturge-Weber syndrome was first described by Schirmer in 1960 and later more specifically by Sturge in 1879. It is also known as Sturge-Weber disease, encephalotrigeminal angiomatosis, meningofacial angiomatosis and Sturge –Weber Dimitri syndrome [5, 6].

Port wine stains represents hamartous capillary malformation and is named so due to deep red hue that they leave on the skin or mucosa [7].

Their presence can signify a developmental anomaly involving central neural axis. Sturge-Weber syndrome is one such hemartoneoplastic syndrome characterized by presence of Port wine stain. Port wine stains in child hood are classically faint, pink macules tend to darken progressively to red purple, may be isolated with well delineated border or may be very diffuse. It has usually unilateral distribution along with one or more segments of trigeminal nerve. Occasionally bilateral involvement or lesion on other parts of body may occur [8].

Most potent vascular lesion will blanch under pressure. Large lesions may be pulsatile if associated with large vessel. Thrombi in angiomas may eventually calcify and such lesion will feel hard on palpation. The calcified nodule or phlebolith may be radiographically evident. Meningeal angiomatosis is usually associated with a convulsive disorder and sometimes contralateral hemiplagia and mental retardation[8].

Characteristically angioma of the leptomeninges occurs as a unilateral lesion, overlying the posterior temporal, parietal and occipital area. Abnormalities of cerebro-vascular system such as AV malformations, arterial thrombosis, subdural hematoma and cerebral atrophy can occur secondary to the angioma. Ocular involvement can result in glaucoma, choroidal hemangioma, bupthalmas or hemionopsis [5].

In our patient none of these features were found except for port wine stain, blindness and convulsion.

Intra orally angiomas can involve lips, causing macrochelia, results in hemihypertrophy of buccal mucosa, palate and floor of mouth. Generalized enlargement might be associated with increased vascular supply [9].

Unilateral hypertrophy of alveolus, pyogenic granuloma, ipsilateral premature eruption or delayed eruption, malocclusion is the other abnormalities reported [5]. Differential diagnosis of Sturge-Weber syndrome includes, Klippel Trenauny-Weber syndrome, hereditary hemorrhagic telengectesia (Rendu-oslar- Weber disease), Mafucci syndrome, Von Hippel Lindau disease [5, 7].

The general management of the patient includes, control of epilepsy and ocular lesions. Port wine stains on the face can be a cosmetic problem and should be treated by high dose of hydrocortisone, dermabrasion, tattoing or lash lamp pulse tunable dye laser therapy. They can result in partial or complete clearing of the Port wine stain. Cryosurgery may be used to correct lip and other soft tissue deformities[10].

Patient was referred to Dermatologist for his cosmetic treatment of port wine stains. But patient did not show interest to take the treatment as he was poor and not affordable to the cost of the treatment.

Dental management of the patient should be stressed on behavior management and preventive measures. Poor oral hygiene can lead to secondary inflammatory gingival enlargement and high DMFT score. Therefore patient and parent education along with plaque control measures should be strictly followed to minimize or to prevent these problems. Endodontic treatment can be performed in these patients since angioma may not involve pulpal tissue. Over instrumentation should be avoided during periapical instrumentation of root canals. Pulpal bleeding can be controlled by cotton pellet and vasoconstrictors[11].

In the case presented here, pyogenic granuloma was surgically excised under local anesthesia. After excision the bleeding was controlled using pressure packs. Once the healing of excised region took place, thorough scaling of the teeth and restoration of carious teeth was carried out.

Gingival hyperplasia can be superimposed by phenytoin induced gingival hyperplasia. Thus proper history taking is very essential to rule out this diagnostic dilemma. In our case, as patient was taking Carbamezapine drug for his convulsions, gingival enlargement was considered as due to the vascular malformations and not because of the drug. Gingival enlargement can be corrected by use of CO_2 laser surgery. Laser surgical procedure can also produce immediate hemostasis, little damage to surrounding tissue and no pain compared to conventional surgical procedure [12].

Various methods used to manage risk of hemorrhage are use of haemostatic agent, provision for blood transfusion, use of post operative splints or injecting sclerosing solutions[10].

Conclusions

Management of patients with Sturge-Weber syndrome may be challenging due to risk of hemorrhage. Precautionary measures should be taken to control hemorrhage and complications during surgical procedures. Dental management should include behavior management and stress should be placed on preventive measures.

Declaration of Interest

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