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STURGE-WEBER SYNDROME : REPORT OF THREE CASES

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Abstract

Sturge - Weber Syndrome is a congenital disorder and characterized by facial hemangioma following one or more divisions of the trigeminal nerve, epilepsy, mental retardation, contralateral hemiplegia, occlular involvement, gingival involvement.

A 34 year old Korean man, a 25 year old Korean woman and a 48 year old Korean woman were found to have red - purple colored pigmentation on the hemifacial area and upper oral mucosal area.

INTRODUCTION

The Sturge-Weber syndrome was first described by Schirmer in 1860. Shirmer described a patient with a port-wine facial nevus associated homolateral glaucoma.

Schirmer, however, did not suggest a relationship between the neural and cutaneous lesions.

Sturge was the first to suggest a relationship between the facial and cranial lesions.

Weber and Dimitri contributed descriptions to complete the syndrome in 1922. Weber have demonstrated the intracranial calcifications in skull roentgenograms as a characteristic of the syndrome.

The sturge-Weber syndrome is a vascular dysplasia which is characterized clinically by the following classic symptoms.

Major symptoms are a nevus flammeus of the face along the distribution of one or more branches of the trigeminal nerve, roentgenographic evidence of intracerebral calcification on the same side as the facial nevus and convulsions, which are most frequently focal and on the contralateral side of the facial nevus.

Minor symptoms are atrophy or spasticity of the extremities on the contralateral side of the nevus, vascular changes in the eye, mental retardation, obesity, electroencephalographic abnormalities.

Oral manifestations are vascular hyperplasia of the oral tissues, telangiectasia, one-sided hypertrophy of the tongue, malerupted teeth, mouth breathing secondary to monstrous gingival enlargement, nevoid affection of the soft & hard palate, high palatal vault, ipsilateral immature eruption of the permanant teeth and macrodontia.

The most frequent site of oral involvement is the oral mucosa. Both the lips and the cheek may be the seat of vascular hyperplasia, the color is more bluish red than the surrounding normal mucosa. The palate and the mandibular gingiva are rarely affected.

The tongue may be affected, showing either telangiectasia or hemihypertrophy. The gingival mucosa exhibits characteristic changes, ranging from slight vascular hyperplasia to monstrous masses making closure of the mouth impossible.

The gingival enlargement is soft and purple and blanching in response to pressure.

The color usually demarcated at the midline of

the maxilla, often in contrast to the facial nevus which stops at the outer margin of the philtrum.

Thoma reported a case in which gingival tumors occurred during pregnancy; following electrosurgical excision, the gingiva remained normal until the next pregnancy when the lesions recurred. Sturge-Weber syndrome has been assigned many names, both eponymic and descriptive, such as Sturge-Weber-Dimitri syndrome, Krabbe's syndrome, Parkes-Sturge-Weber-Kalischer syndrome, encephalotrigeminal angiomatosis, nevoid amentitia, and encephalofacial neuroangiomatosis.

However, because of sturge's first case report and Weber's roentgenographic contribution, the eponym Sturge-Weber syndrome has been generally accepted by clinicians and pathologists.

Bergstrand differentiated three types of Sturge-Weber syndrome,

1. the more severe cases, with epilepsy and idiotism, in addition to the entire triad of symptoms,
2. the less severe cases which present in general only two symptoms, but in which intracranial angiomatosis, if present, is associated with epilepsy.
3. the third group contains those abortive cases in which the nevus and the ocular symptoms are absent and in which merely the cerebral process with induced epileptiform symptoms is the most characteristic feature.

REPORT OF CASES

Case I.

A 34-year-old male patient was admitted to our dept. for large ovoid mass on buccal side of right upper molar area.

Past medical history revealed only the usual childhood diseases and several cosmetic operations for removal of facial hemangioma. On physical examination, height, physical development and proportions were within normal range.

Mental retardation was not observed.

Extraoral examination showed facial hemangioma within the distribution of the first and second divisions of the trigeminal nerve (Fig. 1).

Intraoral examination revealed an extensive hemangiomatous mass, clinlcally resembling a large pyogenic granuloma superimposed on the area of hemangiomatous mucus membrane, which involved buccal side of maxillary right first premolar, second premolar, first molar, second molar (Fig. 2).

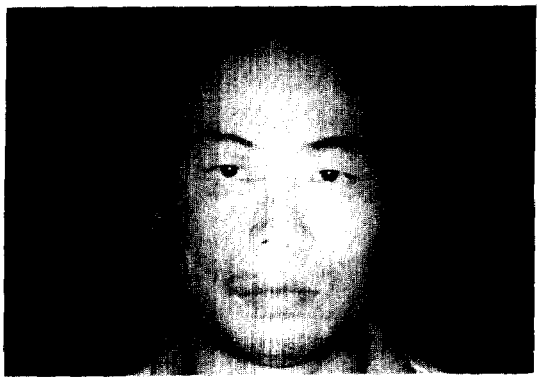

Fig. 1

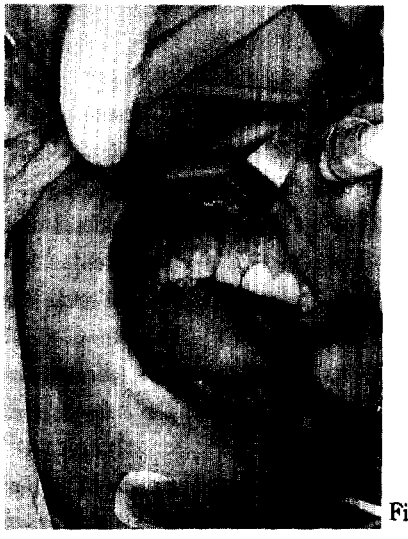

Fig. 2

He complained hypersensitivity to cold, intermittent dull pain and pain during mastication on affected side.

Mass was bluish-red, not tenderness to palpation,

thumb size and blanched on pressure.

Roentgenograms showed generalized alveolar bone resorption on maxillary right first premolar, second premolar, first molar, second molar, third molar. Maxillary right first premolar showed periapical radiolucent mass, first molar dental caries and periodontal ligament space widening, second molar periodontal ligament space widening.

The chest roentgenogram was normal.

Radiographically the bone of the maxilla appeared normal in structure, content, and configuration.

The hemoglobin was 15.3 grams.

All blood differential findings were within normal limits. The result of urine test was normal.

Under general anesthesia the entire mass was removed from the alveolar process and the teeth were removed on maxillary right first premolar, first molar, second molar, third molar. Bleeding was controlled by cauterization and pressure.

Mucosal graft from left cheek was performed (Fig. 3). Bleeding was well controlled throughout the procedure.

Fig. 3

The prefabricated acrylic splint was secured to the remaining teeth, and the resulting pressure provided hemostasis for the postoperative period.

Biopsy report

The specimen consists of an irregular fragment of soft tissue, measuring 3.5×1.5×1cm

Two sections are embedded, Blocked in A

Diagnosis is gingival cavernous hemangioma.

Case II.

A 26-year-old female patient was referred to the Dept of Dentistry, Guro Hospital, College of Medicine, Korea University, for oral surgery consultation because of papillary mass on nasolabial area and ovoid mass on palate, labial gingiva.

The trauma from mastication caused bleeding.

Clinical examination revealed marked facial hemangioma within the distribution of the first and second divisions of the trigeminal nerve (Fig. 4).

Fig. 4

Intraoral examination revealed bluish-red mass involved maxillary left incisor and lateral incisor and canine and first premolar teeth (Fig. 5, 6).

Entire masses were surgically removed from palate, labial gingiva and nasolabial area.

Biopsy report

The specimen is submitted in two parts.

No 1 specimen consists of an ellipse of skin, measuring 1×0.5cm in width and 0.5cm in depth.

No 2 specimen consists of an irregularly shaped fragment of pink grey soft tissue, measuring 1.5×1×0.5cm

Diagnosis is pyogenic granuloma.

Fig. 5

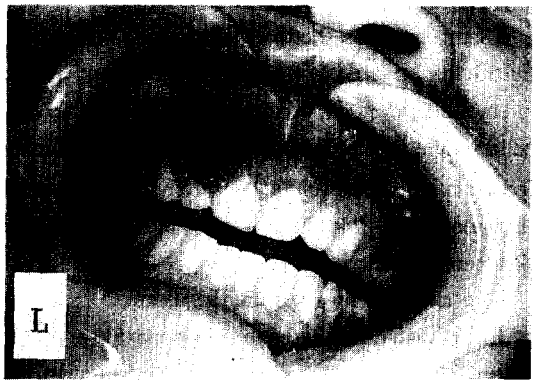

Fig. 6

Fig. 7

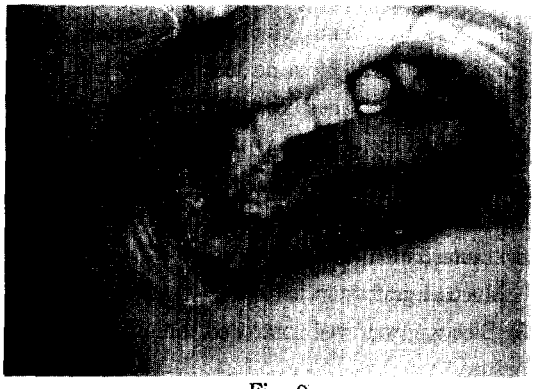

Fig. 8

Case III.

A 48-year-old female patient presented for intermittent sharp pain on maxillary right first molar.

Roentgenograms showed mesiobuccal root fracture on maxillary right first molar. Extraoral examination showed facial hemangioma within the distribution of the first and second and third divisions of the trigeminal nerve (Fig. 7).

Intraoral examination revealed bluish-red flat lesion which involved the entire right half of the maxilla (Fig. 8).

The teeth were removed on maxillary right first molar and third molar. Scaling was performed.

And flap operation was performed on maxillary right molar area.

DISCUSSION

The treatment of dental disease in patients who have oral manifestations of Sturge-Weber syndrome requires special care.

Treatment methods include surgical excision, injection of sclerosing solution, radiation therapy, carbon dioxide snow, electrodissection or insertion of steel wire electrodes.

Since the gingival lesion of Sturge-Weber syndrome was an angiomatous one associated with vascular dysplasia, the possibility of significant hemorrhage during and after surgery must be considered.

Hospitalization is recommended for any surgical treatment.

The preoperative fabrication of acrylic splints to apply pressure directly over the area is perhaps most rewarding.

The use of a local anesthetic with a vasoconstrictor, topical bovine thrombin, Surgicel, Gelfoam, Spungel, and electric cautery can make the operation simpler

by helping to contend with the bleeding.

Peterman and others, in an analysis of 35 cases, found the most common clinical features to be a convulsive disorder (89%), abnormal readiographic findings (63%), mental retardation (54%), ocular involvement (37%), and hemiplegia (31%).

The frequency of oral involvement reviewed by Gorlin and Pindborg in 111 cases, there was a 38% incidence of some oral manifestations. The facial neviod lesion is present at birth, does not enlarge with age, and is usually unilateral.

The area innervated by the ophthalmic division of the trigeminal nerve is the most frequently affected.

The lesion is nonraised, pink to purplish-red in color, well-bordered, and blanches readily.

The oral lesions, like the facial nevus, remain unilateral, ending abruptly at the midline.

Thoma has suggested that some alteration in the vascularity of the jaws occurs, citing abnormal ipsilateral dental eruption sequences and the occurence of macrodontia.

The causes of this disease are unknown, but a genealogic etiology with autosomal dominant and recessive forms with incomplete penetrance, has recently been described.

All the symptoms of this syndrome have rarely been reported in more than one member of the family, but abnormal skin manifestations are not unusual in families of patients with Sturge-Weber disease.

SUMMARY

The authors observed three cases of Sturge-Weber syndrome patients who had come to Department of Dentistry of Hospital in Medical College, Kerea University.

We performed the clinical, hematologic and radiologic examination, and surgical excision.

1. We observed nevus flammeus on facial area, hard palate and buccal mucosa
2. The oral gingival lesions described in the case report were related to the inherent facial and meningial vascular dysplasia of the disease syndrome.
3. The gingival enlargements, we thought, were due to inflammatory reaction.
4. The above clinical signs were found in accordance with the first and second branches of trigeminal nerve.
5. By the surgical excision of lesion and the mucosal graft, gingiva was formed to normal contour. We thought that this therapy was good to periodontal health.

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STURGE-WEBER 증후군의 문헌고찰과 증례보고

국문초록

Sturge-Weber syndrome은 드물게 볼 수 있는 선천성질환으로 일명 encephalotrigeminal angiomatosis, 또는 Sturge-Weber-Dimitri syndrome이라 하며 삼차신경의 분지를 따라 분포하는 안면 혈관종, 안면 혈관종과 같은 부위의 intracerebral calcification, 경련의 특징을 가진다.

이에 저자들은 Sturge-Weber syndrome 3예를 치험하였기에 문헌고찰과 함께 보고하는 바이다.