Sturge-Weber Syndrome Associated with Maxillofacial Osteohypertrophy

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Abstract

This case report presents a rare case of an adult female patient of Sturge-Weber Syndrome with unilateral maxillofacial osteohypertrophy. Incidence and exact cause of association of Sturge-Weber Syndrome with osseous hypertrophy is not known and there are very few similar cases that have been reported in literature. Patient visited to the dental department with the chief complaint of malalignment of teeth for which orthodontic treatment plan was done.

Introduction

Soft tissue vascular malformations may rarely be associated with osteohypertrophies. There are only a few case reports showing association of Sturge-Weber syndrome (SWS) with osteohypertrophy of maxillofacial region [1-3]. SWS is a rare congenital neurocutaneous syndrome with a prevalence of 1:50,000 live births with equal sex distribution and no racial predilection [4]. This syndrome has variable phenotypic expression usually manifested as cutaneous capillary angioma, congenital glaucoma with ipsilateral leptomeningeal angiomatosis. Here we report an unusual case showing the association of SWS with maxillofacial osteohypertrophy in an adult patient.

Case Presentation

A 23 year old female patient reported with the complaint of malalignment of teeth. Her medical history revealed reddish discoloration on left side of face since birth. She also gave history of abnormal growth pattern of left maxillofacial region. The patient was mentally normal with no previous medication and non-contributory family history. On extraoral examination, port-wine stains were present unilaterally on left side of face extending from forehead to eyelids, cheek, nose, philtrum and upper lip (Figures 1a and 1b). On applying digital pressure, blanching of port wine stains was noticed. Facial asymmetry due to hypertrophy of left side of face was evident resulting in deviated plane of occlusion (Figure 1c). Intraoral examination showed reddish discoloration of gingiva in premolar-molar region (Figure 2). Dental malocclusion including deep overbite was present which might be due to regional osseous overgrowth. Clinical signs of port wine angioma were present in the eye of affected side.

Submentovertex radiograph revealed more anteriorly placed zygomatic prominence and zygomatic arch on the affected (left) side than normal side. The condyles and pterygomaxillary fissure were found to be bilaterally symmetrical in both sagittal and transverse direction. However, in the cranial base, foramen spinosum, on the left side, was placed more anteriorly than its counterpart (Figure 3).

Postero-Anterior cephalogram clearly showed increased vertical maxillary height on the left side which is in agreement with the transverse cant in the occlusal plane (Figure 1c). The ramus height on the affected side was marginally higher as compared to the opposite side as a compensation of the increased vertical maxillary height on the affected side which led to facial asymmetry.

Diagnosis of Sturge-Weber Syndrome associated with maxillofacial osteohypertrophy was made on the basis of clinical and radiographic examination.

Discussion

Osseous involvement in form of any hypertrophy, lysis, hypoplastic changes or other abnormality has been illustrated in relation to different vascular malformations but its incidence in association with SWS is not known [5,6]. Osteohypertrophy is benign osseous overgrowth that can...
be described as angiodysplasia that entail some vascular abnormality which is related to secondary changes including further vascular malformations and bony overgrowth [3]. There is scanty literature available and only a few number of cases [1-3,6] reported for SWS associated with maxillofacial hypertrophy as seen in this case. Facial bone overgrowth may lead to severe facial disfigurement with facial and dental asymmetry, occlusal canting and malocclusion. Gingival enlargement may also be present that can be due to hypertrophy, antiepileptic medications such as phenytoin sodium, or a combination of both [7].

A vascular malformation–induced osseous change can be a strong consideration by virtue of close association with the distribution of capillary angioma, the underlying mechanism may be varied and remains elusive [1]. Boyd et al. [5] has suggested possible mechanisms including mechanical, physiologic, and developmental processes that could explain for the alteration of skeletal growth pattern in various vascular malformations.

Treatment rendered depends on the severity of symptoms. Osseous overgrowth can be corrected by surgical intervention [6]. Laser therapy, dermaabrasion or tattooing can be done for port wine stain as these can cause psychological trauma to the patient. Further medication can be given on basis of any symptom such as seizure, headache, glaucoma or complaints of bleeding.

Conflict of Interest

None

References