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Bilateral Condylar Hypoplasia – A Rare Case Report and Review of literature

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Abstract

Condylar hypoplasia is very rare condition, especially when this condition is not associated with any syndrome. Very few literatures are published on the non-syndromic bilateral condylar hypoplasia. Here, we report a rare case of congenital bilateral condylar hypoplasia in a 62 year old female patient.

Keyword: Condylar hypoplasia, Bilateral ,Congenital, Facial Asymmetry, Antegonal notch.

Introduction

TMJ abnormalities may be congenital or acquired. Growth disturbances in the development of mandibular condyle may occur in utero, late in the first trimester and may result in disorders such as aplasia or hypoplasia of the mandibular condyle[1]. Condylar hypoplasia is a bone disease characterized by the decreased development of one or both the mandibular condyles.Condylar hypoplasia may be congenital or acquired and may present as unilateral or as a bilateral entity. Congenital hypoplasia or

aplasia of the mandibular condyle indicates underdevelopment or non-development usually associated with various craniofacial abnormalities mainly originating in the first and second branchial arches which include Treacher-Collins, Goldenhar, Proteus, Morquio, and Auriculo-condylar syndrome[2]. Acquired condylar hypoplasia is usually initiated by any local or systemic pathological event. Developmental abnormalities at this location manifest as altered growth on the affected side of the condyle or lower third of the face, mandibular ramus, mandibular body, and alveolar process. Aberrations in the growth and development of the mandibular condyle can affect both the functional occlusion and the aesthetic appearance of the face and is usually asymptomatic. Early diagnosis by correlating clinical and imaging features is paramount in the management of these patients. An interesting rare case of congenital bilateral severe condylar hypoplasia in a 62 year old female is presented.

Case Report

A 62 year old female patient reported to the outpatient department of Oral medicine and Radiology with the chief complaint of missing teeth and an underdeveloped lower jaw which worsened over the years. It was first noticed during her childhood and it has gradually increased in severity.There was no complaint of pain or discomfort associated with the condition. Pre-natal, natal and post-natal history was uneventful. At the anamnesis there was no history of any trauma associated with the mandible or any systemic diseases. Patient's parents had a history of consanguineous marriage. There was no family history of the present problem. Due to unfavorable socioeconomic conditions, it was not possible to get the treatment done for the patient. General physical examination did not reveal any abnormalities. Her vital signs were within normal limits and the patient was in good general health.

Extraoral examination revealed gross facial asymmetry of the lower third of the face with severe retruded mandible giving a bird face appearance(Fig 1). Mouth opening was normal, 42 mm. with no deviation or deflection. On palpation, condyle were not palpableon both sides.

Fig 1:Extra-oral photograph of the patient showing facial asymmetry of the lower third of the face with retruded mandible Frontal view of face, 2 Lateral view of patient shows severe retruded mandible

Intra-oral examination revealed generalized plaque and calculus and maxillary and mandibular arches were partially edentulous with missing teeth-18,27,28,31,32,33,35,47,46 and root stumps in relation to 34, 36. There was proclination and crowding with upper anterior teeth with increased overjet. She had a deep palate. A deep antegonial notch was palpable on both sides. No other important clinical extraoral and intraoral finding were observed. As the clinical findings were consistent with those of TMJ ankylosis a working diagnosis of early ankyloses of the TMJ was made.

Panoramic radiographic (Fig.2) revealed a poorly developed condylar head on right and left side associated with a short condylar neck and an overall decrease in the height of the ramus of the mandible. The antegonial notch was more prominent on the left side as compared to the right side.Glenoid fossa was not developed on both sides.

CBCT(Fig.3&4)revealed a severely hypoplastic condyles on both sides. The condylar head on left side was smaller in size as compared to right side with flattened articular surface. There was no evidence of bony or soft tissue

ankylosis associated with the TMJ on any side. Based on the clinical and radiographic findings, a final diagnosis of nonsyndromic bilateral condylar hypoplasia was made.Patient was referred to oral surgery and prosthodontics for the treatment.

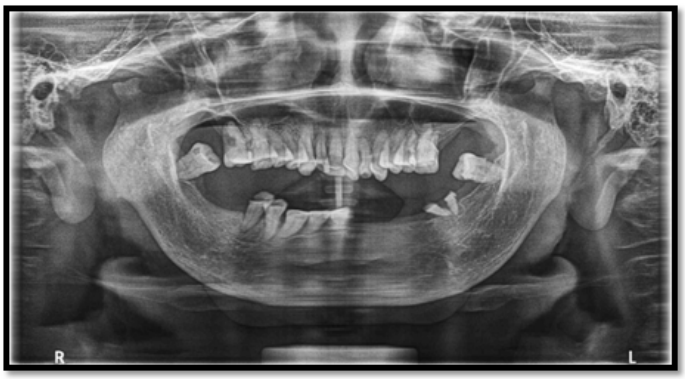

Fig. 2: Panoramic radiograph showing a poorly developed bilateral condylar head associated with a short condylar neck and an overall decrease in the height of the ramus of the mandible. The antegonial notch was more prominent on the left side as compared to the right side.

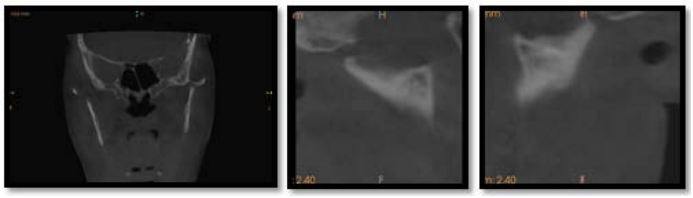

Fig. 3: A coronal section of CBCT showing a significant reduction in the mediolateral width of the condyle on right and left side.

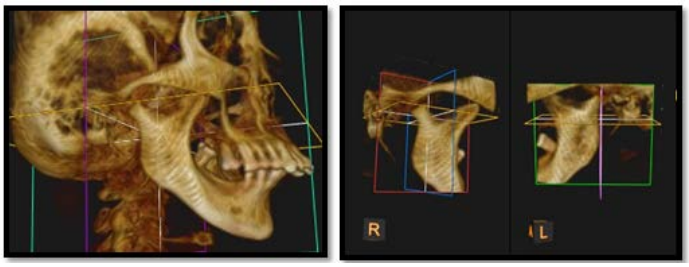

Fig.4: CBCT showing three dimensional reconstruction in the sagittal plane showing flattening of right and left side of condylar head.

Discussion

Developmental disturbances involving the TMJ may result in anomalies in the size and shape of the condyle. Condylar hypoplasia, hyperplasia, and bifid condyle are commonly observed developmental disturbances of TMJ. Condylar hypoplasia is characterized by unilateral or bilateral underdevelopment of the condyle and maybe either congenital or acquired. Congenital type presents at birth and is often associated with some systemic condition originating in the first and second branchial arches, such as Mandibulofacial dysostosis (Treacher Collins syndrome), Hemifacialmicrosomia (first and second branchial arch syndrome), Oculoauriculovertebral syndrome (Goldenhar syndrome), Oculomandibulodyscephaly (Hallermann-Streiff syndrome), Hurler's syndrome, Proteus syndrome, Morquio syndrome and Auriculocondylar syndrome[1]. Acquired or secondary condylar hypoplasia may be caused if the condyle is injured during active growth, because of which development may be arrested. The common factors include chronic trauma, infection of the mandibule or middle ear, irradiation of head and neck region, and systemic conditions like rheumatoid arthritis, mucopolysaccharoidosis. There have been previous reports of aplasia or congenital condylar hypoplasia without any syndrome, family history or any history of trauma and can occur without any distinct etiology. Aplasia or hypoplasia of the mandibular condyle without any associated facial malformations is an extremely rare condition. Cases of nonsyndromic mandibular condyle aplasia have been earlier described by Krogstad [3], Prowler and Glossman [4], Akihiko et al. [5], Santos et al. [6], Bowden Jr. and Kohn [7], Canger and Celenk [8] and so forth. Our case also presented with bilateral condylar hypoplasia without any other features suggestive of any syndrome.

The deformity involves fullness of the face, deviation of the chin toward the affected side, and flatness of the face on the unaffected side, the side to which the ramus is short causes muscles to appear fuller; the muscles on the unaffected side are stretched so that side appears flatter in unilateral condylar hypoplasia. Mandibular deviation causes malocclusion. In bilateral condylar hypoplasia there is mandibular retrognathia resulting in asymmetry of the face. The severity of the deformity depends on the degree of hypoplasia or agenesis of the tissues involved and the more severe the deformity, the greater the probability that it will worsen with growth.

Diagnosis is based on a history of progressive facial asymmetry during the growth period, clinical examination, and radiographic evidence of condylar deformity and antegonial notching. Several treatment methods have been recommended for treating condylar hypoplasia and possibilities for influencing the mandibular growth and therefore enhancing the facial appearance. These patientsoften manifest with many underlying skeletal and dental problems and a multidisciplinary approach (oral surgeon, general surgeon, plastic surgeon, and orthodontist) for of this condition is recommended[9,10,11].Surgery is often required, but the timing and regimen of this choice is still an issue to be resolved. After the patient has stopped growing, skeletal deformities can be corrected only by double jaw surgery and/ or genioplasty or unilateral mandibular augmentation. Since our patient was aged and her main concern was replacement of the missing teeth she was not advised any surgical correction.

Conclusion

We report a rare case of bilateral condylar hypoplasia, not related to any clear pathological disorder. This case of unknown etiology was thoroughly examined; based on clinical and radiographic findings, we suggest that this case is of congenital origin. Non-syndromic condylar hypoplasia and aplasia are exceedingly rare conditions and very few case reports are published till date. In this context, our case is an important addition to the literature. Early detection and prompt treatment are imperative to restore esthetics and thus provide psychologic benefit to these patients.

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