Non Syndromic 'Aplasia' of Mandibular Condyle - A Case Report

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Abstract

Mandibular condyle aplasia is an anomaly which usually manifests in association with various syndromes. When not seen in conjunction with any other developmental anomalies, it is an extremely rare condition. Only a few cases of Non syndromic condylar aplasia have been reported in literature till date. Proper diagnosis along with differentiation from the syndromic cases is important as treatment plan and prognosis for each varies. Here we report a case of Non syndromic hypoplasia of mandibular condyle which was asymptomatic till the age of 61years.

Keywords: Hypoplasia Mandibular Condyle, Temperomandibular Joint.

Introduction

The temperomandibular joint (TMJ) is the most complex elegantly designed joint in the human body. It is a ginglymoarthrodial joint, a term that is derived from ginglymus, meaning a hinge joint, allowing motion only backward and forward in one plane, and arthrodia, meaning a joint which permits a gliding motion of the surfaces.[1] When compared to other diarthrodial joints, during prenatal life the TMJ lags morphologically behind other synovial joints in both the timing of its appearance and its progress, so that at birth the joint is still largely underdeveloped.

The TMJ first appears in the 8th week of gestation, when two separate areas of mesenchymal blastemas appear near the eventual location of the mandibular condyle and glenoid fossa.[2,3] Bone and cartilage are first seen in the mandibular condyle at approximately the 10th gestational week. First condylar blastema developed from which the mandibular condyle cartilage, the aponeurosis of the lateral pterygoid muscle, and the disc and capsule component composing the lower portion of the joint are derived. Next is the temporal blastema, which eventually forms the articular surface of the temporal component and the structures of the upper portion of the joint.

The mandibular condyle and temporal blastemas begin their growth at relatively distant sites; they then move towards each other as the joint develops by the 12th week. At birth, the articular surfaces of both the mandibular condyle and temporal bones are covered with fibrous connective tissue. Later, this tissue is slowly converted to fibrocartilage as the fossa deepens and the mandibular condyle develops under functional influences.[4,5] 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. As compared to hypoplasia, hyperplasia of the mandibular condyle is not visible at birth and seems to be gradually acquired during growth.[5]

Case Report

A male patient, age of 61 years visited to department of Oral Medicine & Radiology, M. R. Ambedkar Dental College & Hospital, Bengaluru, with chief complaint for replacement of missing upper and lower teeth. The patient gave no significant medical history. Patient gave history of extraction of all his teeth in a private clinic 2 years back. On general physical examination, patient was well oriented, cooperative and no other abnormalities found. On extra oral examination, Mouth opening was 4.2 cm. On inspection presence of asymmetry in the right side of face, fullness of face on the left side was noted and deviation of chin towards right side on mouth opening on palpation of TMJ, there was reduced movements on the right side, hypermobility of TMJ on left side, and prominent antegonial notch on the right side was detected. On intraoral examination presence of occulsal slant (Fig. 1) towards right on occluding with upper and lower edentulous ridge. Considering the patients history and clinical features, a provisional diagnosis of completely edentulous maxillary and mandibular arches and Condylar hypoplasia on the right side was made. Hemifacial microsomia was considered as differential diagnosis. Orthopantomogram (Fig. 2) was taken and it revealed completely edentulous maxillary and mandibular arches, absence of condylar head and rudimentary neck of the condyle was present, reduced height of ramus, deepening of ante-gonial notch was noted. Lateral TMJ view (Fig. 3) was taken, it also revealed the absence of right condyle and hypermobility of left condyle. With thorough clinical evaluation and radiological investigations, final diagnosis of non syndromic condyle hypoplasia was made.

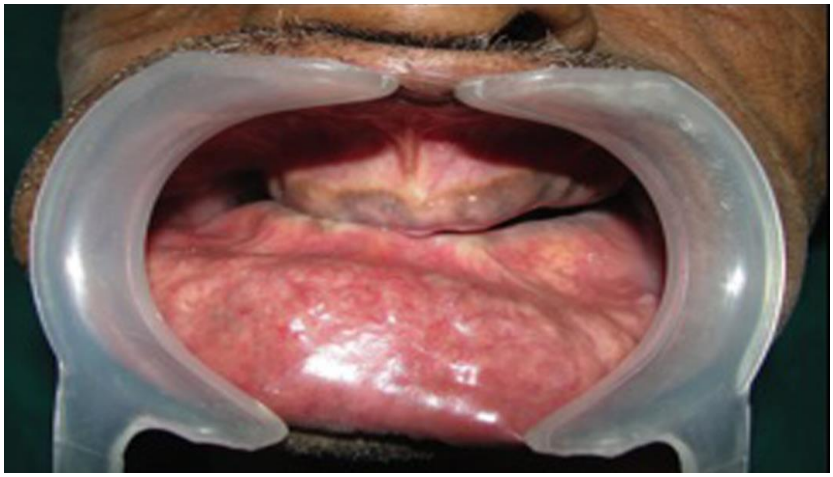

Fig. 1: Intraoral view

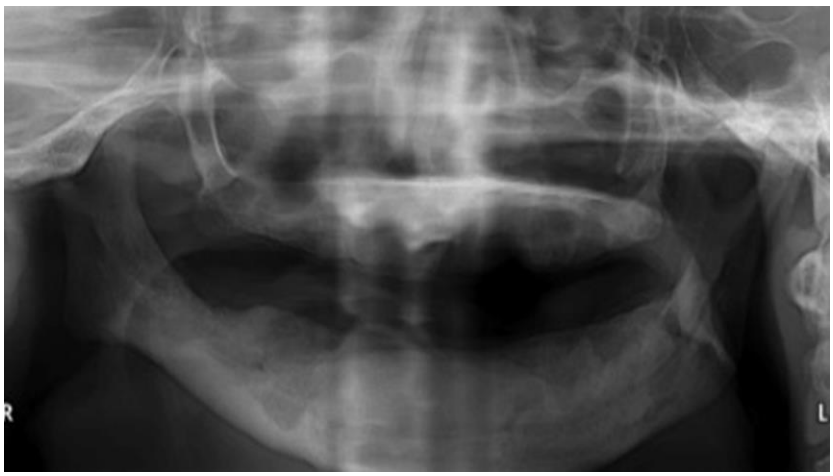

Fig. 2: OPG

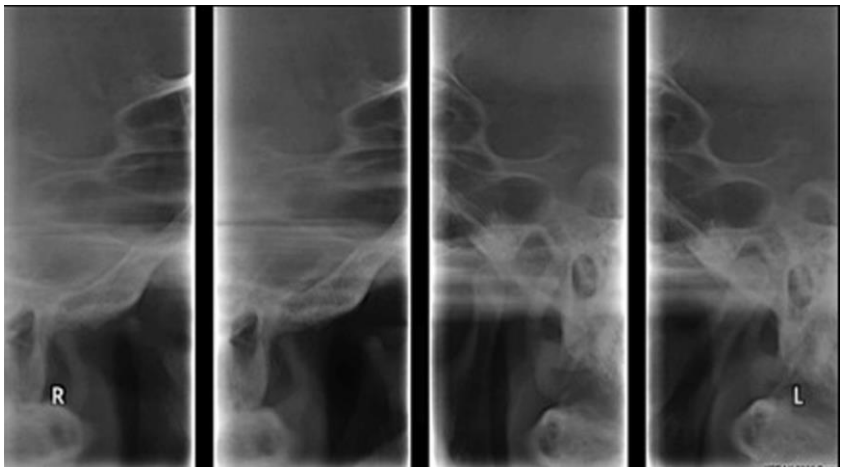

Fig. 3: TMJ

Discussion

Condyle hypoplasia is failure of the condyle to attain normal size because of congenital and developmental abnormalities or acquired diseases that affect condylar growth. The condyle is small, but condylar morphology usually is normal. The condition may be inherited or may appear spontaneously. Some cases have been attributed to early injury or injury to the articular cartilage by birth trauma or intraarticular inflammatory lesions.[6]

The cause of bone growth disorders is not completely understood. Trauma in many instances is a

contributing factor and, especially in a young joint, can lead to hypoplasia of that condyle, resulting in an asymmetric shift or growth pattern. Condylar hypoplasia usually is a component of a mandibular growth deficiency and therefore often is associated with an underdeveloped ramus and (occasionally) mandibular body. Congenital abnormalities may be unilateral or bilateral and usually are manifestations of a more generalized condition; they also may be associated with congenital defects of the ear and zygomatic arch.

Developmental abnormalities that manifest during growth usually are unilateral. Acquired abnormalities are the result of damage during the growth period from sources such as therapeutic radiation or infection that diminish or prevent further condylar growth and development.

Patients with condylar hypoplasia have mandibular asymmetry and may have symptoms of TMJ dysfunction. The chin commonly is deviated to the affected side, and the mandible deviates to the affected side during mandibular opening. Degenerative joint disease is a common long term sequela.[6] Table 1 shows distinguishing features between syndromic and non-syndromic condylar aplasia.

Congenital (primary) condylar hypoplasia is characterized by unilateral or bilateral underdevelopment of the mandibular condyle and usually occurs as a part of some systemic condition originating in the first and second branchial arches, such as Mandibulofacial dysostosis (Treacher Collins syndrome), Hemifacial microsomia (first and second branchial arch syndrome), Oculoauriculovertebral syndrome (Goldenhar syndrome), Oculomandibulodyscephaly (Hallermann- Streiff syndrome), Hurler's syndrome, Proteus syndrome, Morquio syndrome and Auriculocondylar syndrome.[7-10] Table 2 illustates the skeletal and general manifestations of various syndromes.

Treacher Collins syndrome (TCS) is a rare congenital disorder of craniofacial development that arises as the result of mutations in the TCOF1 gene. TCS is inherited in an autosomal dominant fashion with variable penetrance and phenotypic expression. Anomalous development in TCS is characterized by a combination of findings isolated to the head and neck. Facial bone hypoplasia, involving the mandible and zygomatic complex in >75% of patients, is an extremely common feature of TCS. The maxilla may also be hypoplastic but sometimes can be seen as over projection. Other characteristic abnormalities include downward slanting of the palpebral fissures, the nose may be broad or protruding, absent external auditory canal (EAC), middle ear malformations, and pinna deformities.

Craniofacial radiologic abnormalities include hypoplastic or aplastic zygomatic arches, choanal shortening, micrognathia and maxillary narrowing, or over projection. Cleft palate is a common co-occurrence and may be severe. Craniofacial defects in TCS are often bilateral and relatively symmetric. Limb anomalies do not occur in TCS, which helps differentiate it from other syndromes that manifest with similar facial features.[11]

Hemifacial Microsomia (HFM) involves first and second branchial arch derivatives with highly variable phenotypes. It is also known as first and second branchial arch syndrome, otomandibular-facial dysmorphogenesis and lateral facial dysplasia. HFM is primarily a syndrome of the first branchial arch, which involves underdevelopment of the temporomandibular joint, mandibular ramus, muscles of mastication and the ear. Abnormal development of the auricular hillocks leads to microtia or atresia of the pinna and it is proportional in severity to the abnormal external auditory canal development.[12]

Various treatment approaches have been proposed for treating condylar aplasia, and possibilities for influencing mandibular growth have been the topic of numerous clinical and experimental studies. A costochondral rib graft can be used to help establish an active growth centre. Surgery is often required, but the timing and regimen of this choice is still an issue to be resolved. It is advisable to postpone surgical treatment to a later time. Orthognathic treatment or a combination of such treatment and a graft can be successful. In addition, reported that effective results were obtained through the application of a form of orthodontic activator which aimed to swing the mandible to the unaffected side and promote formation of a mandibular condyle, albeit irregular in shape.[14] In our case a modified complete denture was fabricated.

Table 1: Differential diagnosis of Syndromic /non syndromic- condylar aplasia.

	Syndromic-condylar aplasia	Non-syndromic condylar aplasia
History	Generally diagnosed at birth	Usually not diagnosed at birth, history of trauma unusual
Clinical presentation (as identified)	Soft-tissue defects(may be very mild to severe)	No soft-tissue defects
	Masseter muscle hypoplasia	Well-developed facial muscles
	Ear defects, pre-auricular tags	Normal external and middle ears
	Facial nerve deficit	No nerve deficit
	Deviation of the chin on the affected side, associated with flatness on the affected cheek	Deviation of the chin on the affected side, associated with fullness on the affected cheek
	Mild deviation to the affected side during opening	Significant deviation to the affected side during opening
Radiographic (as identified)	Hypoplasia of the ramus and condyle and coronoid processes up to absence of the condyle and temporal fossa	Hypoplasia of the ramus and condyle and coronoid processes, the temporal fossa is always present
Karyotyping (as suspected)	OMIM (Online Mendelian Inheritance in Man) 164210: Hemifacial microsomia, OMIM 602483: Auriculo-Condylar Syndrome identified.7	Normal genetic makeup

Table 2: Syndromes and their skeletal and general manifestations[13]

Syndromes	Skeletal traits	Other traits
Hemifacial microsomia	Underdevelopment of the temporomandibular joint, mandibular ramus	Underdevelopment of ear and mastication muscles, hearing loss, cardiac anomalies
Goldenhar	Most severe form of microfacial microsomia	Eye tumours, fused spine
Treacher Collins	Mandibular hypoplasia, hypoplasia/aplasia of the zygomatic arch, coronoid process and condyle	Underdeveloped/malformed ears, ocular anomalies, hearing loss
Hallerman–Streiff	Dyscephalia and bird face, hypoplastic mandible/ ramus, missing condyle	Cataract, microophtalmia, skin atrophy, dental anomalies
Morquio's	Abnormal development of bones, including the spine, a prominent lower face	Respiratory, cardiac, ocular and hepatic abnormalities, enamel hypoplasias
Proteus	Atypical growth of the bones, cranial hyperostosis	Thickening of skin, vascular anomalies, verrucous epidermal naevi

Conclusion

Patients with non syndromic unilateral condylar hypoplasia is very rare. Only a few cases have been published in the literature. In our case, it was an incidental finding after the thorough history taking, clinical evaluation and radiological investigations. This highlights the importance of diagnosis for the better outcome of the treatment.

Conflict of Interest: None

Source of Support: Nil

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