Case Report

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NONSYNDROMIC UNILATRAL CONGENITAL CONDYLAR HYPOPLASIA: A RARE CASE REPORT

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ABSTRACT:

Mandibular condyle hypoplasia 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 congenital unilateral condylar hypoplasia 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.

Major complains of patient being facial anomaly and deviation of chin .This case report deals with such a case which was effectively treated with camouflaging genioplasty of the mandible.

Key Words: Condylar hypoplasia, facial deformity, genioplasty.

INTRODUCTION:

Facial asymmetry is common in humans. Significant facial asymmetry causes both functional as well as esthetic problems. When patients complain of facial asymmetry, the underlying cause should be investigated. The etiology includes congenital disorders, acquired diseases, and traumatic and developmental deformities.

Temporomandibular joint (TMJ) is considered a ginglymous diarthrodial joint capable of both rotational and translator movements. It consists of the mandibular condyle, glenoid fossa and the articular eminence of the temporal bone. The congenital deformities and developmental abnormalities of the mandibular condyle can be classified as hypoplasia or aplasia, hyperplasia, and bifidity. Hypoplasia or aplasia of the mandibular condyle indicates underdevelopment or non-development associated mainly with various craniofacial abnormalities.

The anomaly can be acquired or congenital [1].

The TMJ first appears in the 7^{th}-8^{th} week of gestation, when two separate areas of mesenchymal blastemas appear near the eventual location of the mandibular condyle and glenoid fossa.[2]

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

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condyle is not visible at birth and seems to be gradually acquired during growth.[3]

CASE DETAIL:

A 20 year old female patient reported to us complaining of deviation of jaw and teeth to one side since childhood. She did not notice it during childhood and it became prominent during her adolescence. (FIG 1 a)

The patient denied neither any history of childhood trauma to the maxillofacial region nor any severe systemic illness. There was no familial trait to this problem also.

General Physical examination did not reveal any abnormalities .Her vital signs were within normal limits.

Extra oral examination revealed a hypoplastic and deviated chin to the left side. She had a convex profile with mandibular shortening on left side.

Intra oral examination revealed the patient had a wide arch and maxillary anteriors to be in a proclined state where as the mandibular teeth were deviated to the left side where there was open bite in the posterior teeth giving a bimaxillary protrusion appearance to the dentition.(FIG 1b)

An Orthopantomogram (OPG) was advised which revealed the anomaly of a hypoplastic left mandibular condyle head. This resulted in hypoplastic growth of mandibular left ramus body and deviation of gnathion to the left side. Glenoid fossa was underdeveloped in the left side.(FIG 1c & d)

The patient was given a treatment option of mandibular distraction osteogenesis which she declined due to financial reasons so a plan of camouflaging sliding genioplasty was advised which she accepted to correct somewhat of her complaints.

Under general anesthesia intraoral vestibular incision in the mandibular anteriors was made, periosteal dissection was done till the mandibular basal bone and the symphyseal region was exposed.(FIG 2)

Via rotary instruments the midline marking of the chin was made, then sliding genioplasty was performed via an horizontal osteotomy cut and fixing the osteotomized segment to the mandibular bone to its desired position via two 2.0 stainless steel two hole with gap miniplates .(FIG 3) The wound was closed layer by layer by 3.0 absorbable vicryl sutures and a compression bandage was placed for 5 days. The in hospital stay of the patient was uneventful. On 6th post-operative day she was discharged.

Post operatively her deviated chin was surgically camouflaged (FIG 4a,b,c)and she underwent post-surgical orthodontics treatment.

DISCUSSION:

The temporo mandibular joint (TMJ) is one of the joints of the human body which starts developing at 7th intrauterine week. The initial functions of mouth opening

movements start appearing by the twentieth week during foetal stage, but the development process continues until the twelfth year of life**.**

Developmental anomalies of the mandibular condyleare generally classified in terms of aplasia, hypoplasia,hyperplasia, and bifidity. Hypoplasia means the insufficient development of the mandibular condyle [4]

Hypoplasia or Aplasia of the mandibular condyle is one of several manifestations of syndromes including hemifacial microsomia, Goldenhaar's syndrome, and Treacher Collin's syndrome,the literature also mentions Proteus syndrome, Morquio syndrome, and auriculocondylar syndrome, which can demonstrate condylar malformations and condyle agenesis. In each of these conditions, in addition to condylar aplasia, these are other facial or skeletal malformations [5-6]. Acquired (secondary) condylar aplasia or hypoplasia may occur due to mechanical trauma during active growth. Other causes may include inflammation in the TMJ area, rheumatoid arthritis and radiotherapy [7] .Parathyroid hormone-related protein deficiencies also affect bone formation and chondrocyte differentiation, which consequently affects the condyle formation. Recent reports have shown that various extracellular matrix proteins, such as transforming growth factor-b disorders affecting Meckel's cartilage for normal mandibular development.[8] Whereas some authors affirm that mandibular condyle deficiency can occur with no defined aetiology.[9]

Various treatment modalities have been proposed for the treatment of condylar hypoplasia, the timing of treatment and possibility of influencing mandibular growth has been the topic of numerous clinical and experimental studies. Early clinical procedures could prevent facial asymmetries as well as other functional disorders. The surgical treatment methods include restoring the mandibular dental base to its correct relationship with the maxillary dental base by osteotomy,distraction osteogenesis and bone-grafting operations in the mixed dentition stage so that the permanent dentition can take up a satisfactory occlusion.

Alternatively orthodontic treatment may be carried out in the mixed or permanent dentition, which would be followed by corrective surgery in the form of masking operations such as epithelial inlays or orthognathic surgeries including genioplasty [10].

Patients with non-syndromic mandibular hypoplasia are a rare subgroup; in which the condition does not seem to be progressive. Integration of surgical treatment with orthodontic treatment is required for a good prognosis and predictable outcome.

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TABLE:

TABLE 1 : Differential diagnosis of Syndromic /non syndromic- condylar aplasia [11]

CLINICAL PRESENTATION	SYNDROMIC MANDIBULAR HYPOPLASIA/ APLASIA	NONSYNDROMIC MANDIBULAR HYPOPLASIA/ APLASIA
HISTORY & EXAMINATION	• Generally diagnosed at birth • Soft-tissue defects(may be very mild to severe) • Masseter muscle hypoplasia • Ear defects, pre-auricular tags • Facial nerve deficit • Deviation of the chin on the affected side, associated with flatness on the affected cheek	• Usually not diagnosed at birth, history of trauma unusual • No soft-tissue defects • Well developed facial muscles • Normal external and middle ears • No nerve deficit • Deviation of the chin on the affected side,associated with fullness on the affected cheek
RADIOGRAPHIC	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

FIGURES:

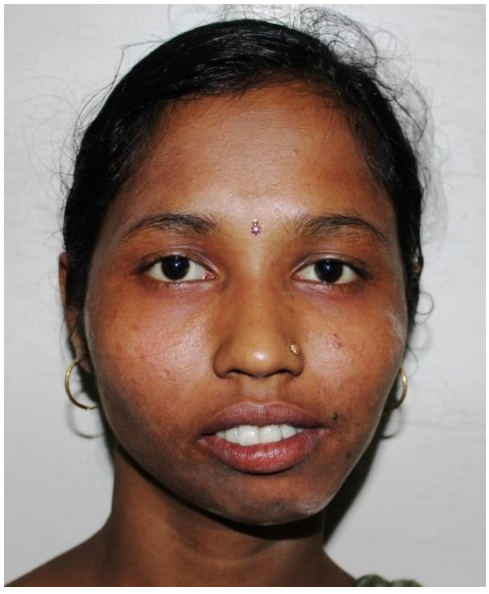

FIG 1A

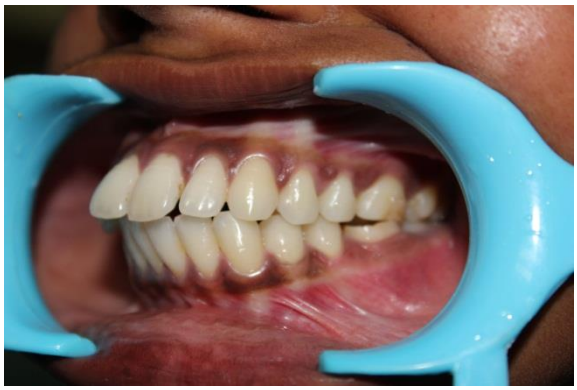

FIG 1B

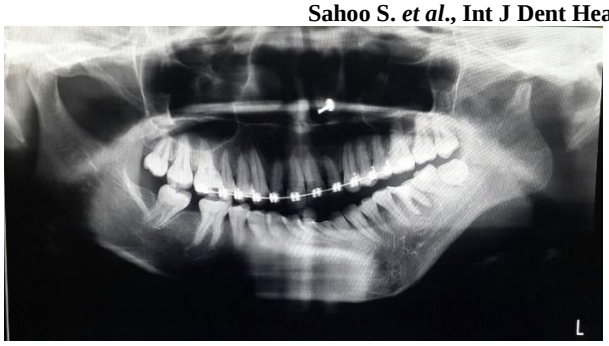

FIG 1C

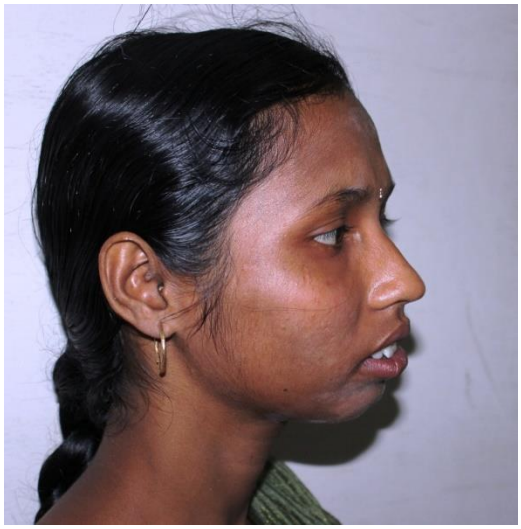

FIG 1D

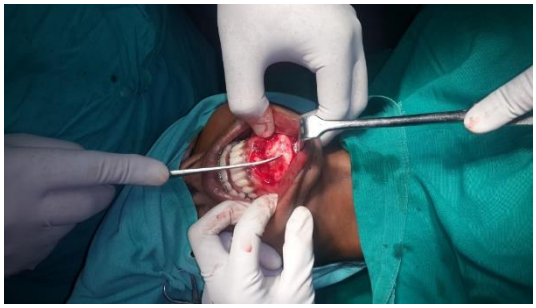

FIG 2

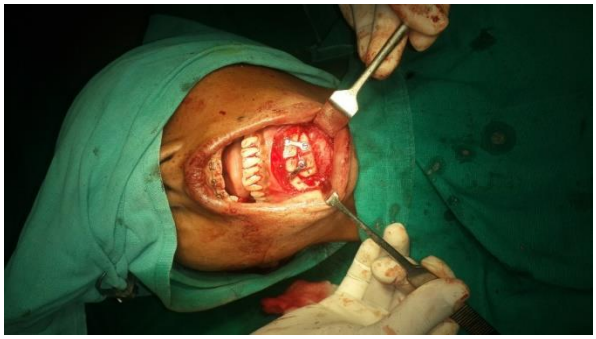

FIG 3

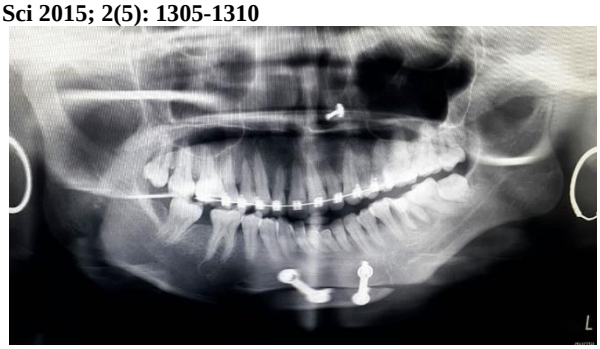

FIG 4A

FIG 4B

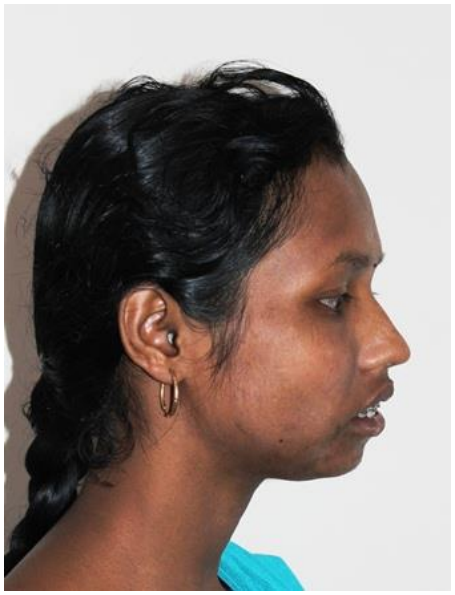

FIG 4C