Case Report

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Hemifacial Microsomia- A Clinical Dilemma, Case Confirmed by Radiodiagnosis.

Abstract:
Aplasia of condyle along with soft tissue manifestation seen as a part of a syndrome. We report a similar case along with condylar aplasia on the right side The patient reported to the department with a chief complaint of forwardly placed upper front teeth. Clinical examination, radiographs, and 3D CT images revealed complete absence of condyle on the right side along with soft tissue features which directed us to a final diagnosis of hemifacial microsomia.

Key-words: Hemifacial Microsomia, Hypoplasia of Condyle, Aplasia of Condyle.

Introduction:

Hemifacial microsomia was first described by German physician Carl Ferdinand Von Arlt in 1881.[1]

Gorlin and colleagues invented the word hemifacial microsomia. Other synonyms are "otomandibular dysostosis"-first branchial arch syndrome" " second branchial arch syndrome" oculoauriculovertebral sequence" Goldenhar syndrome" "lateral facial dysplasia" and "craniofacial microsomia".[2] Hemifacial microsomia manifests in highly variable phenotype. It is the second most common craniofacial disorder after cleft lip and cleft palate. Any structure derived from the first and second pharyngeal arches can be affected.[2] Hypolasia of the mandibular condyle is an extremely rare condition as it is generally present as apart of developmental disorder ranging from minimal to complete absence. The present case demonstrates this condition very well where, although absence of the condyle is clear, other manifestations were also seen.[3]

Case Report:

A 21-year-old Asian woman with facial asymmetry required radiographic examinations for later treatment. Her chief complaint was forwardly placed upper front teeth and wants orthodontic treatment. The patient was concerned about her facial appearance and thought that her face is deviated or turned to right side because of her forwardly placed upper front teeth. There was no reported history of this condition in the family. The patient's parents were normal. and her accompanying sibling was also normal.

On extra oral examination facial asymmetry and a severe deviation of the midline to the right side was seen.(fig1)

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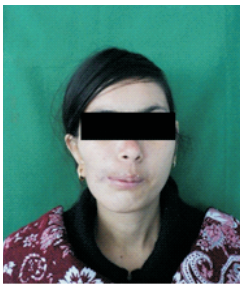

Figure 1: Clinical examination revealed facial asymmetry and a severe deviation of the midline to the right side.

Although only the left condyle was clinically palpable, the condylar movements were not affected by the condition. The lower part of the ear lobule was slightly raised on the right side which was the affected side. TMJ examination of the patient was done and it was observed that on palpation right side of the condyle was not palpable. After clinical examination, a provisional diagnosis of right side condylarhy poplasia was given with a differential of aplasia of condyle. A radiographic examination was indicated for further elucidation of the case.

Radiographic examination:

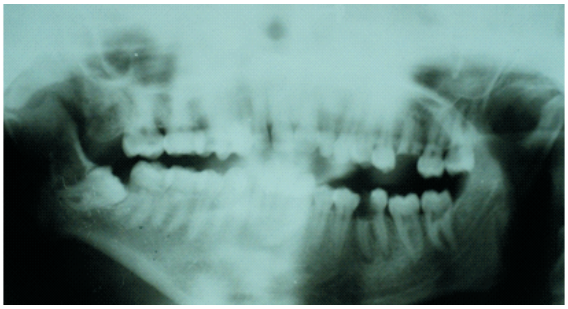

Figure 2: Panoramic radiography showing missing condyle on the right side along with Ill-defined coronoid process and height of the ramus also appears reduced, flattened condyle can also be seen.

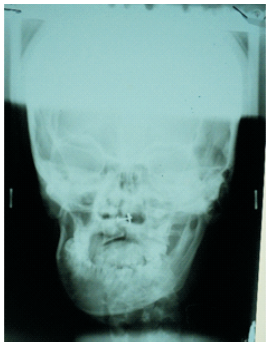

Figure 3: PA skull showing marked deviation towards affected right side along with deviation of nasal septum ,deviated maxilla and mandible .Lack of development of ramus ,coronoid and condylar process on right side.

In addition to the OPG and PA skull view. The 3D CT images(Figure 4a and 4b) obtained in this clinical case were enough to clearly diagnose the total absence of the right condyle. Other examinations could give similar information, such as 3D-CT, MRI and CBCT. The 3D CT examination enables accurate surgical planning and even helps to obtain a model by stereo lithography from CT data. It also provides quantitative information from skeletal and muscular parameters.[3]

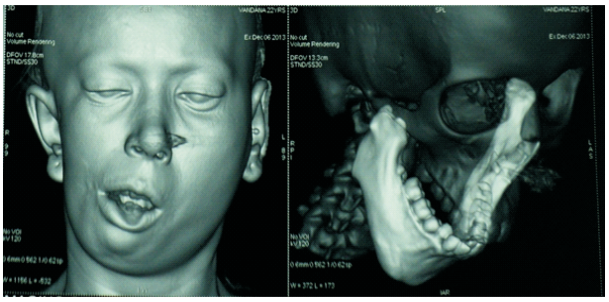

Fig 4a: Right side mandible showing hypoplastic coronoid with short and widened ramus suggestive of condylar aplasia and absence of 3rd molar and articulation

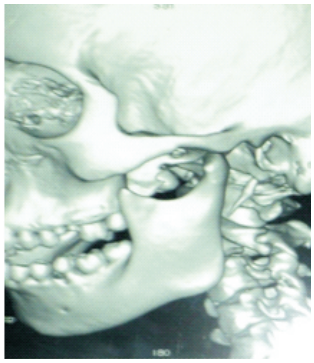

Figure 4b Left side mandible showing anteriorly displaced condyle and the condyle is articulating with the zygomatic arch.

Both 3DCT reconstruction images Fig4a)and 4b) confirm the radiodiagnosis of aplasia of condyle on right side

Discussion:

TMJ developmental disorders may result in irregularities in the size and shape of the condyle. Hyperplasia, hypoplasia, agenesis, and formation of a bifid condyle may be evident on radiographic examination of the joint. Local factors, such as trauma or infection, can initiate condylar growth disturbances.[4] Hemifacial microsomia, Goldenhar, Treacher Collins, Hallerman–Streif, Morquio's and Proteussyndromes are present concomitantly with condylar aplasia. As a rule, in each of these conditions some soft tissue manifestations accompany the condylar agenesis and/or condylar malformations.[5].

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Acquired (secondary) condylar hypoplasia takes place if the condyle is injured during active growth, because of which development may be arrested. The most common causes are mechanical injury, such as trauma (before the age of 2), infection of the joint itself or the middle ear, childhood rheumatoid arthritis, radiotherapy, and parathyroid hormone-related protein deficiency which affect bone formation and chondrocyte differentiation.[6] In the present case, there were no anomalies either of the ear or the eyelids. Agenesis of the mandibular condyle without infection of the soft tissues has been reported earlier (Prowler and Glassman, 1954)[7]. However, in that report the condition obviously started after the age of six and was probably of an acquired nature (Poswillo and Robinson, 1992)[8].

On the other hand the clinical features of HFM include flattening of the face on the affected side due to hypoplastic maxillary and malar bones and aplasia of the mandibular ramus and condyle[9] which can be seen in our patient. The eye may thus appear to be on the lower level than that of the affected side. Malformation of the external ear may vary from complete aplasia to a crumpled, distorted pinna. The chin and the facial midline are off centered and deviated to the affected side. Often, one corner of the mouth is situated higher than the other, giving rise to an oblique lip line which was also seen in our patient.(fig1) There can be hypoplasia of muscles such as the masseter, temporalis, pterygoids and muscles of facial expression on the involved side. Sensorineural hearing loss and facial nerve dysfunction are common in HFM. Auditory problems are present in 30-50% of the patients. In oral manifestations, there is macrostomia, aplasia of the mandibular ramus and condyle. Agenesis of the third molar (seen in our patient on the right side(fig 4a)and second premolar may be present on the affected side as well as there can be the presence of supernumerary teeth, enamel malformations, delay in tooth development and hypoplastic teeth.[9]. The first classification used in HFM patients was done by Samuel Pruzansky in 1969 using x-rays of the jaws of patients with this condition but this classification includes only the description of the mandible, therefore, when used in HFM patients, it leaves out many aspects of the pathology.[10] Thus two most frequently used classifications are the skeletal–auricular–soft tissue (SAT) and the orbital asymmetry–mandibular hypoplasia–ear malformation–nerve dysfunction–soft tissue deficiency (OMENS) classifications.[11] The OMENS classification is the most comprehensive one and, therefore, it is one of the most commonly used systems.

Conclusion:

In our case, besides aplasia of the mandibular condyle which is unilateral, a systemic factor is excluded; the history also indicates no systemic diseases .There were few soft tissue and dental features similar to hypodontia manifestations pushing the diagnosis towards hemifacial microsomia which according to OMENS would be O0M1E1N1S1.

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