NON-SYNDROMIC UNILATERAL CONGENITAL MANDIBULAR HYPOPLASIA: A CASE REPORT.

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ABSTRACT

Mandibular hypoplasia is a frequently encountered craniofacial defect. It can be classified into three groups namely, the congenital, developmental and acquired. The focus of this case report is to highlight the non-syndromic sub-group of mandibular condyle hypoplasia which is a rare occurrence and to review the literature.

Keywords: Non-syndromic, Congenital, Hypoplastic mandible.

INTRODUCTION

Mandibular condyle hypoplasia is a rare prenatal condition, primarily involving the mandibular condylar growth centre,[1] resulting in hypoplasia of the condylar head and the ipsilateral hemi-mandible.[2] Condylar hypoplasia is caused by 'under-development' or defective formation of the mandibular condyle and may be unilateral or bilateral.[3] It may also either be congenital or acquired and affects both sexes equally.[4] Primary congenital condylar hypoplasia or aplasia may be part of a syndrome[5, 6] or non-syndromic.[7] Acquired or secondary condylar hypoplasia occurs if the condyle is injured during active growth, for example by trauma, infection or radiation, while the primary type is very rare and has no pre- or postnatal cause. The features of the congenital type occur slightly earlier than those of acquired hypoplasia, congenital heart disease and absence of the hyoid bone.[2] Early recognition of this abnormality and the severity of the problem are important in deciding the modality of management.[3]

A single case of mandibular hypoplasia may occur in 700 genetic syndrome craniofacial abnormalities in the United States,[8] while it constituted 0.8% consecutive new births in Kenya.[9]

Presented below is the case of an isolated congenital unilateral mandibular hypoplasia with tempero-mandibular joint ankylosis.

CASE REPORT

KO is a 5-year-old girl who presented to our hospital with pain and difficulty in opening the mouth of two years duration which was progressive. There was no intrauterine or post-delivery history of trauma, infection or radiation to the area of the jaws. The patient has been relatively healthy except for occasional malaria attacks. The parents discovered that as the baby was growing, there was a progressive asymmetry of the lower jaw, with the left side looking smaller. Physical examination showed asymmetry of the mouth and jaws with slight deviation of the former to the right side.

The teeth were intact bilaterally. No teeth fusion or under-development was seen. The palate and tongue were within normal limits. No cleft palate or lip was seen. Other organs were examined and were within normal limits. No webbing of the neck or fusion of fingers or toes was seen. Cardio-pulmonary examination revealed normal air entry into the lungs, and normal cardiac size and sounds.

Conventional radiographs showed a small right body of the mandible with ill-defination of the mandibular head and the tempero-mandibular joint (TMJ)._Fig.1. The left mandible and tempero-mandibular joint are within normal limits. Coronal slices of the CT showed a hypoplastic right mandible. There was also an expansile and flat ipsilateral mandibular head (see arrow). The mandibular fossa of the temporal bone was shallow. There was also associated narrowing of the tempero-mandibular joint space with sclerosis of the joint surfaces and partial fusion at the lateral edge. The ipsi-lateral mandibular shaft was also under-developed, which was also seen in a conventional radiograph of the jaws. No hyoid bone was identified on the ipsi-lateral side, but the contra-lateral hyoid bone was identified, and appeared within normal limits. No cardiac anomalies are seen.

Ultrasound of the abdomen showed normal abdominal organs. Also, a chest radiograph and echocardiography were within normal limits. Intravenous urography was done to asses any renal anomaly which turned out to be normal. A diagnosis of congenital hypoplasia of the right mandible with ankylosis of the temporo-mandibular joint was made.

The Dental Surgeons assessed the patient for a possible mandibular correction, but the patient's relations absconded at night without any given reason.

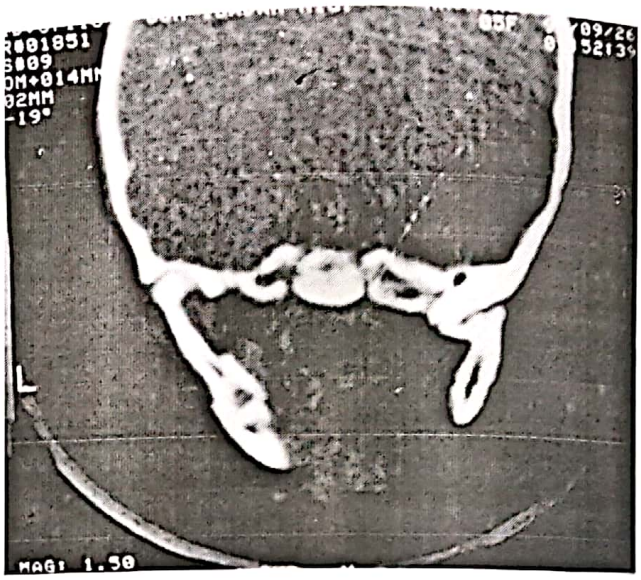

Fig. Coronal slice of the temporo-mandibular joint showing a flattened temporo-mandibular fossa and head of mandible. There is joint-space narrowing and medial ankylosis.

DISCUSSION

The term 'craniofacial' is that which relates to the bones of the skull and face. Congenital craniofacial anomalies are a group of defects caused by abnormal growth and/or development of the head and facial bones. They may be part of a syndrome or be isolated cases. These anomalies can affect any area of the craniofacial region. Multiple anomalies may be identified such as micro/megacephaly, microphthalmia, coloboma, hypertelorism, cleft palate or lip, micrognothia or microthia.[3] These patients may present as isolated cases, that is, without associated anomalies or developmental delays.[10, 11] Our patient showed normal facial features and abdominal organs except for the asymmetry of the jaw with the right side looking smaller.

In the US, available statistics show that 1 in every 700 normal live births showed features of a craniofacial syndrome.[8] Kenyan Hospital statistics was 0.8% of 7,355 consecutive births at the Kenyatta referral hospital.[9] However, one non-syndromic cranio-orofacial anomalies were seen in every 100 live births in India.[10] Mandibular hypoplasia constituted only 2.9%

of over 750 patients born with external congenital anomalies in Mulago Hospital, Uganda over a time frame.[12] There is no documented report of craniofacial anomalies in any centre in Nigeria. To the best of our knowledge, this is the first documented case of isolated hypoplastic mandible in this environment.

The most obvious facial bony deformity among craniofacial anomalies is the mandible, especially the ascending ramus, which can be absent or reduced in the vertical dimension.[13]The size of the condyle usually reflects the degree of hypoplasia of the ramus. Involvement of the temporomandibular joint (TMJ) can range from mild hypoplasia to only a pseudo articulation at the cranial base. Pruzansky proposed three types of the disease, depending on the severity of the disease. This classification was later modified by Kaban et al to include various subgroups.[12] This patient fell into type 1 condition, with only a moderate mandibular deficiency.

The etiology of these anomalies is complex, including multiple genetic and environmental factors.[7] A variety of congenital syndromes affecting the face occur due to defects involving the first and second brachial arches.[14] Embryologically, the mandible develops from the cartilage of the first pharyngeal arch, known as Meckel's cartilage. Developmental anomalies of structures derived from the upper half of the first brachial arch are common,[15] giving rise to deformities like mandibular hypoplasia, and cleft palate,[16] with occasional associated deformities of soft tissue structures in the neck. Such structures are from the lower brachial arch.[17]

The most common associated congenital abnormalities outside of the craniofacial area are congenital heart disease, absence of the hyoid bone, neck contracture and small stature. However, in the case under review, we could not identify other congenital abnormalities associated with it, hence highlighting its rarity. With the arrival of CT and MRI, diagnostic imaging of the TMJ had improved tremendously.[18] The CT examinations enable accurate diagnosis and surgical planning, and also provide quantitative information from skeletal and muscular parameters.[19]

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