Non-syndromic Unilateral Congenital Aural Atresia with Agenesis of Right Mandibular Condyle: A Clinical Report

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ABSTRACT

Ear and mandible share common embryologic origin. Any disturbance in these structures during developmental process may result in disorders such as aplasia or agenesis of ear along with Temporomandibular Joint (TMJ). This can result in facial asymmetry and malocclusion. The resulting malocclusion is best managed surgically at appropriate age to address the facial, skeletal and dental problems but till then a harmonious occlusion is important for sound function and esthetics. This case presents a comprehensive management of dental problems in a patient with unilateral congenital aural atresia with agenesis of TMJ till a definitive treatment is achieved.

A 5-year-old male patient diagnosed with congenital aural atresia with agenesis of right TMJ was referred to the Department of Pedodontics and Preventive Dentistry for dental management. After thorough examination and investigation, comprehensive dental management was done to provide ample function, occlusion and esthetics.

Keywords: *Agenesis, congenital aural atresia, temporomandibular joint.*

INTRODUCTION

The Temporomandibular Joint (TMJ) is composed of the temporal bone, mandible, articular disc, ligaments and associated muscles.[1] Ear and mandible share common embryologic origins. Thus, their embryologic development is linked processes. Condylar aplasia is a condition with under-development of condyle mostly associated with other syndromes and are considered extremely rare when not seen in conjunction with any other developmental anomalies.[2] Congenital aural atresia is the malformation of external auditory canal. Here with, a unique case of non-syndromic unilateral condylar aplasia with ipsilateral aural atresia is reported with an aim to summarize the clinical manifestations and treatment experience of this rare entity.

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CASE REPORT

A 5-year-old male patient diagnosed with congenital aural atresia with agenesis of right mandibular condyle was referred to the Department of Pedodontics and Preventive Dentistry for dental management.

Extraoral examination revealed facial asymmetry (Figure 1a and 1b) with right sided deviation of mouth on opening (Figure 2) that was considered adequate (32mm). The external ear was underdeveloped with small sized obliterated right pinna (Figure 1c).

On palpation, condyle could not be appreciated on right side. Intraorally, midline shift with Angle's class II molar relationship and deep bite towards the right side was and Angle's class III with open bite on left side was observed. Early Childhood Caries (ECC) involving 55, 54, 53, 52, 51, 61, 62, 63, 64, 65, 74, 75, 84 and 85 and exfoliative mobility on 71 and 81 were noted (Figure 3, 4 and 5)

Due to unavailability of 3D CT, temporal bone CT (Figure 6) was advised which revealed small sized, peanut shaped deformed right pinna (likely Grade III microtia) along with complete obliteration of the bony and cartilaginous right external auditory canal (likely aural atresia). Also, right middle ear cavity size was reduced with decreased volume presenting dysplastic and fused right ossicles (fusion of

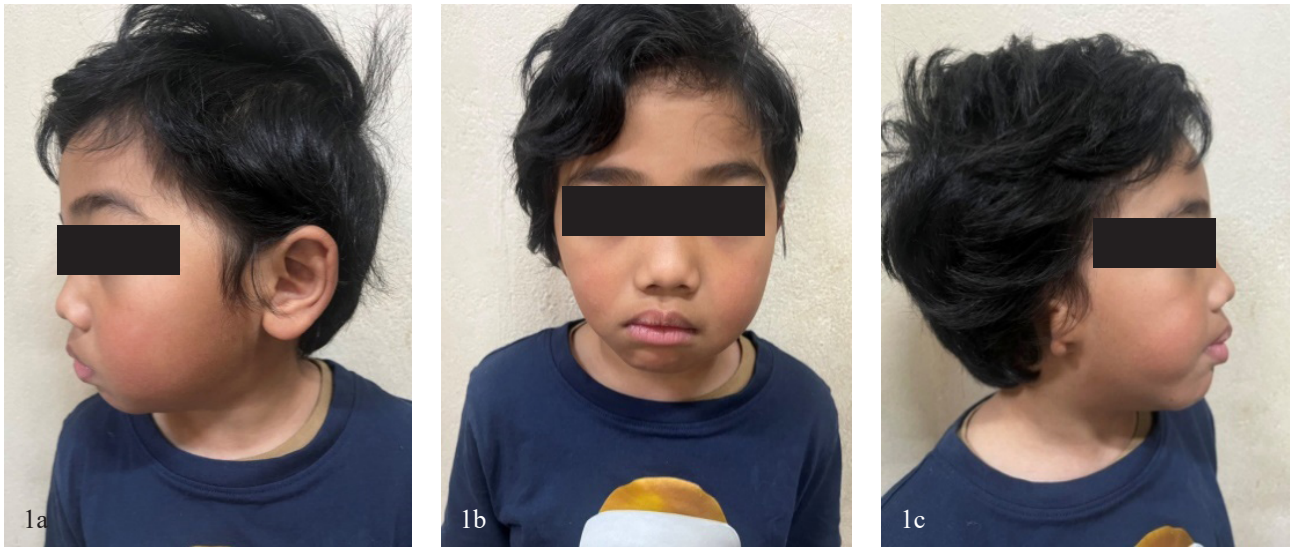

Figure 1. Preoperative extraoral photographs. a) Left profile b) Anterior profile c) Right profile.

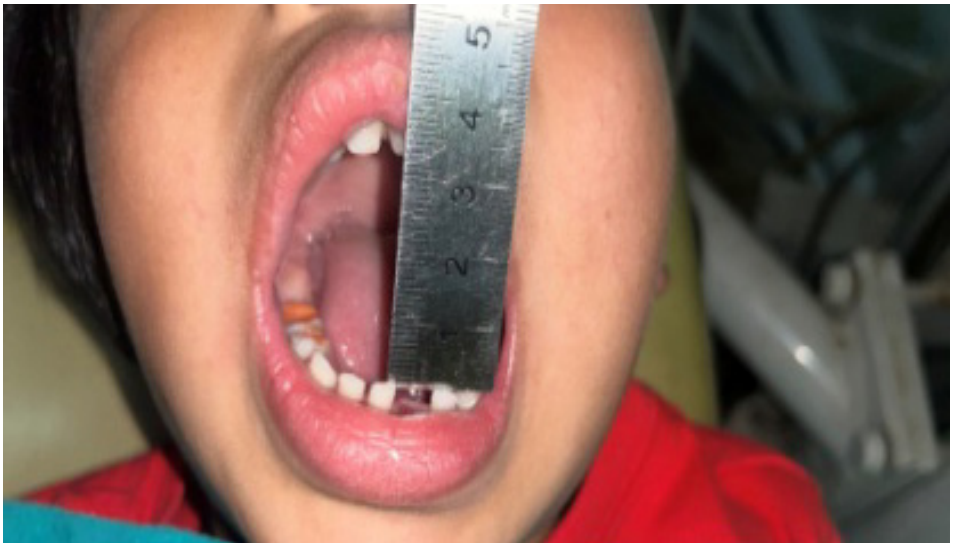

Figure 2. Adequate mouth opening with right side deviation

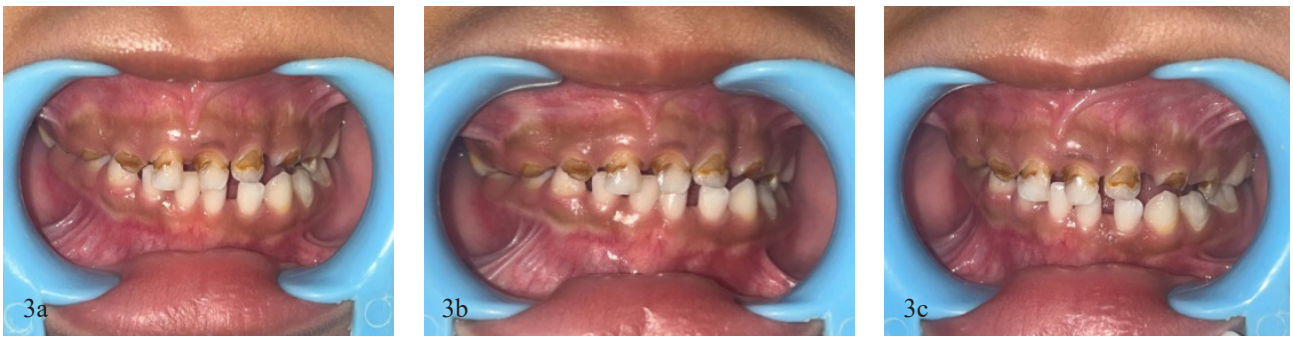

Figure 3. a) Frontal view b) Right view c) Left view. Preoperative intraoral findings

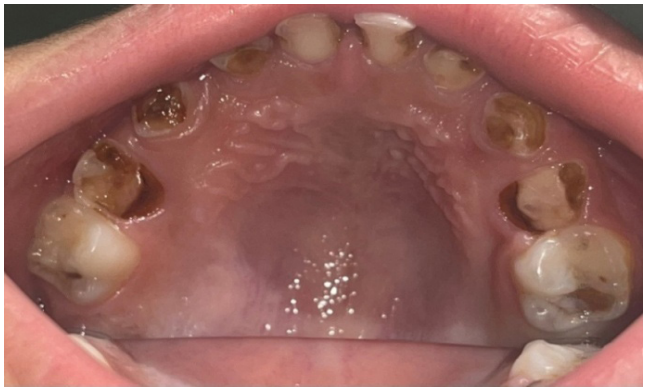

Figure 4. Preoperative maxillary arch showing ECC.

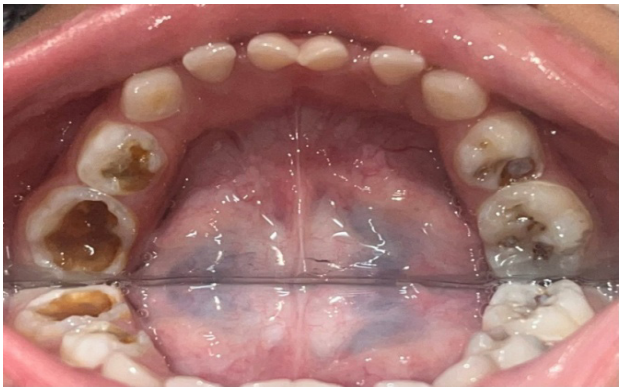

Figure 5. Preoperative mandibular arch showing spared anteriors teeth by caries.

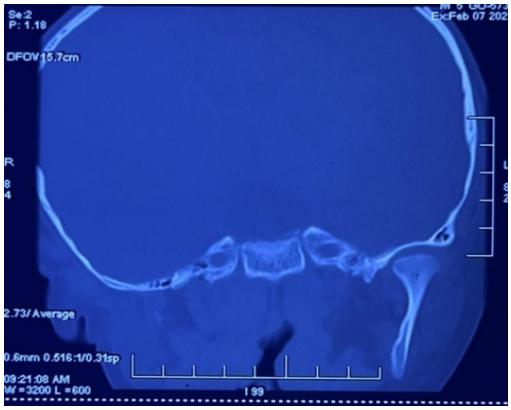

Figure 6. CT showing absence of right TMJ.

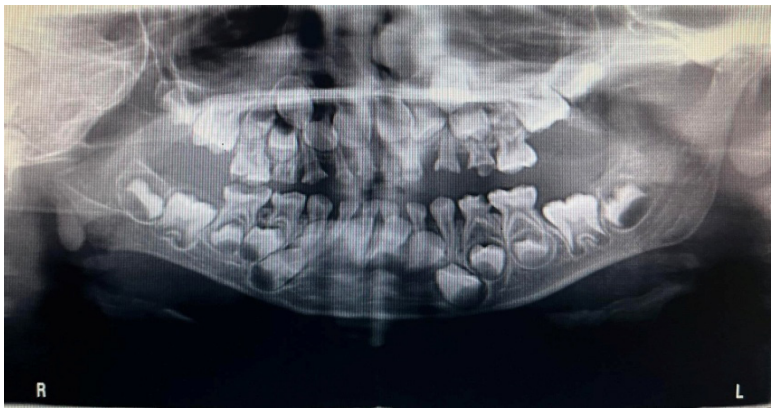

Figure 7. Preoperative OPG showing absence of right condyle.

malleoincudal and incudostapedial joints) along with the missing right tympanic membrane. Right mandibular fossa and right mandibular condyle were missing with intact coronoid process, while the TMJ on the left side appeared intact.

Further panoramic radiographic findings (Figure7) also revealed absence of the mandibular fossa and condylar head on the right side with an intact coronoid process and a prominent antegonial notch.

After radiographic confirmation, patient was referred to the pediatric OPD to rule out any associated disease/ conditions. Medical and clinical reports revealed no any significant findings. Based on the clinical and radiographic evaluation, a differential diagnosis of non-syndromic case of unilateral congenital aural atresia with agenesis of right TMJ condyle was made.

Comprehensive management with GIC to 55, 65, 74, 84 and 85 and Composite restorations with respect to 51, 61,62 and 63 was done respectively (Figure 8). Deep caries management was done on 75 with calcium hydroxide base (Dycal) and GIC restoration followed by stainless steel crown placement on 54 and 64. Pulpectomy followed by composite buildup was done with respect to 52 and 53 (Figure 9,10,11).

Treatment strategies for TMJ abnormalities in such patients remain unclear, however, periodic follow-up and radiographs including 3DCT face every 3 months was planned followed by surgical reconstruction in the future. At 3 months follow up, patient turned up with fall of 54 stainless steel crown which he didn't want to go for replacement and OPG was taken (Figure 12). Patient was kept under wait, watch and recall basis.

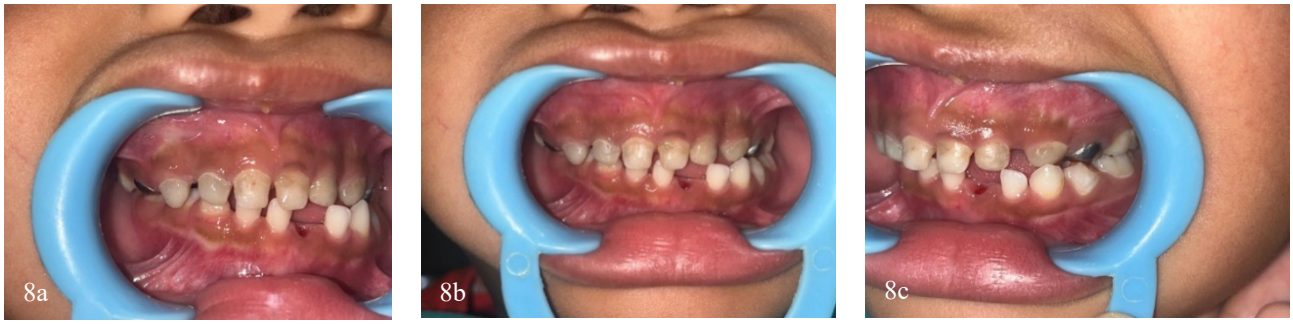

Figure 8. Postoperative intraoral findings. a) Right view b) Frontal view c) Left view.

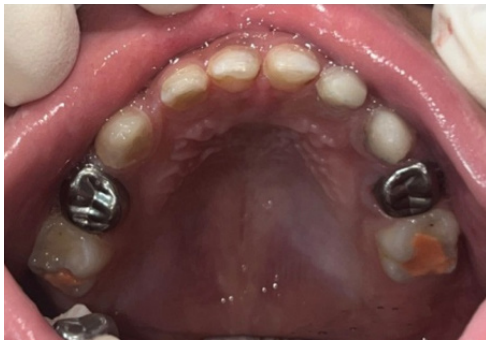

Figure 9. Postoperative maxillary arch with SSC on 54, 64 and type VII GIC restoration on 55 and 65

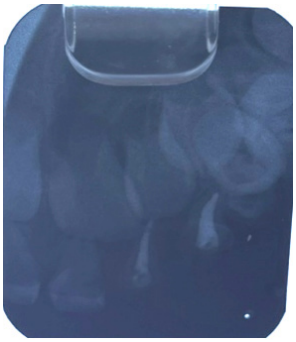

Figure 10. Postoperative IOPAR with Metapex obturation on 52 and 53

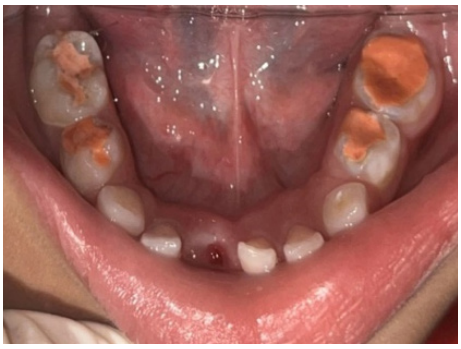

Figure 11. Postoperative mandibular arch with extraction of 81 and Type VII GIC restoration on 74, 75, 84 and 85

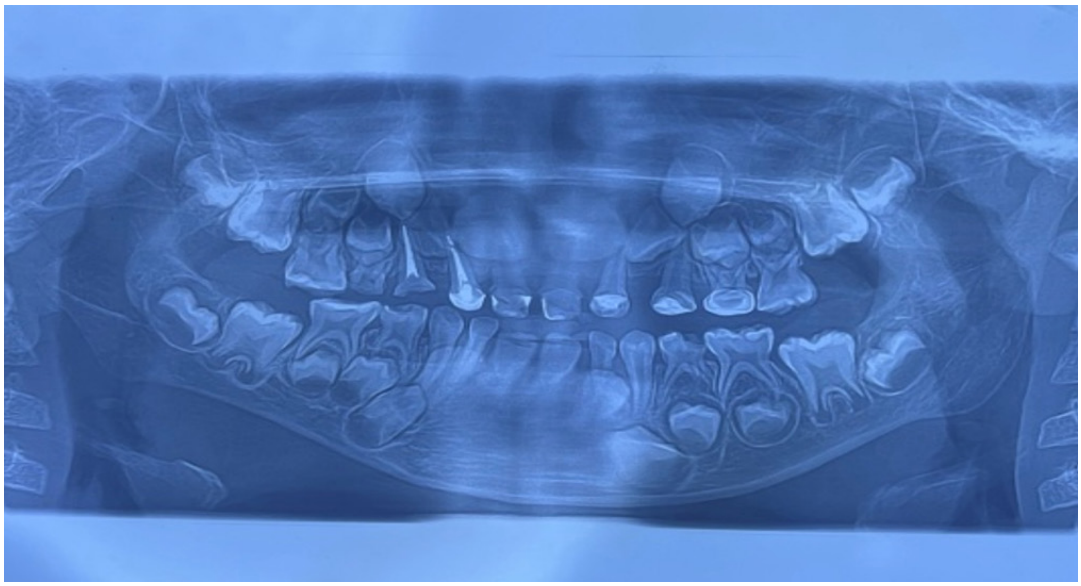

Figure 12. OPG at 3 months followup.

DISCUSSION

Congenital deformities of the temporomandibular joint (TMJ) can cause disturbances in the growth of mandible. They can occur as single condition such as agenesis, dysplasia, hypoplasia/hyperplasia of mandibular condyle or can develop as a component of different syndromes like hemifacial microsomia, Treacher Collins syndrome or the Pierre Robin sequence.[3]

Aplasia of the mandibular condyle with no any association with other anomalies is an extremely rare condition.[4] The present case demonstrates similar condition with agenesis of mandibular right condyle with aural atresia without association with the other syndromes.

First and second pharyngeal arches give rise to a wide variety of facial structures including skeletal, muscular and neural elements through a complex signaling.[5]

The first pharyngeal arch develops into maxilla, mandible, zygoma, part of the temporal bone, malleus, incus, parts of the auricle and muscles of mastication, whereas the second arch gives rise to stapes, body and lesser horns of hyoid and muscles of facial expression. The external auditory meatus and tympanic membrane is evolved from the first pharyngeal cleft whereas the first pharyngeal pouch develops into the eustachian tube and middle ear cavity. Similarities in the embryonic origin between mandible and the external and middle ear can be affected by several other factors causing abnormal auricular morphology and mandibular deformity.[6]

In this case, no any genetic correlation or other syndromic association was found in the detailed patient medical and family history. There was no any history of prenatal or birth trauma that correlates with the differential diagnosis of congenital aplasia of the mandible with auricular deformity. Hence, the clinical evaluation, orthopantomogram and CT images obtained in this case were enough to clearly diagnose the total absence of the right condyle and atresia of the right ear with contrast to the normal opposite.

It is extremely important to perform a detailed family history and clinical examination in such condition as this will allow the appropriate counseling to the parents and patient regarding their comprehensive dental management including reconstructive surgery and orthodontic correction.[4] Treatment of such conditions is mainly corrective through surgical interventions.[7]

However, a differential diagnosis of hemifacial microsomia can be made based on the clinical manifestations. It is the second most common congenital craniofacial birth defect after cleft lip and palate. This disorder is mostly unilateral. The organs affected are ear with conduction defects, mandible, orbit, zygomatic arch and maxilla. This leads to unevenness of the mandible and TMJ with dental consequences such as malocclusion, delayed eruption and noticeable jaw deviation.[8]

It was also found that various dental disorders like untreated dental caries cause a profound impact on self-esteem, confidence, aesthetics, function and psychosocial behavior thus, affecting their quality of life. Therefore, in this case, a comprehensive dental treatment was performed to help the child to get the restored function and esthetics that inadverently improve the overall quality of life.

There is still controversy on the best approach and time to treat a mandibular condyle aplasia. Today, many agree that early manipulation and correction of the mandible is crucial for symmetric growth and development of the facial skeleton during childhood.[7] Whereas, others state that the best time to initiate therapy is shortly before the pubertal growth spurt.[9]

Finally in this case, after complete dental treatment the patient was kept under regular followup and periodic monitoring of OPG to help evaluate whether the lesion was progressive or arrested. Surgical repair of the joint and ear with costocondral graft and orthodontic rehabilitation of the occlusion could be planned in future.

CONCLUSION

Based on the clinical and radiographic evaluation of the findings, this condition was considered as a rare case of total condylar aplasia on the right side with right aural atresia with no association with any pathological disorder/syndrome. A differential diagnosis of hemifacial microsomia could also be made but further diagnosis is required to confirm the case. Such cases are rarely

reported and very few studies are pubished till date. Hence, this case might be an important addition to dental literature. Childhood is a crucial stage of life in which current impairment and future illness can be prevented by understanding the probable risk factors. Comprehensive dental treatment on this child helped to gain the confidence, restore the function and esthetics thus improving his quality of life.

Conflict of Interest: None

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