Cleidocranial Dysplasia - surgically uncovering and bonding multiple impacted permanent teeth

Case Report

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ABSTRACT

Skeletal, craniofacial, and Oro-dental abnormalities are hallmarks of Cleidocranial Dysplasia (CCD), an autosomal dominant bone disorder brought on by a mutation in the CBFA1 gene. People have open fontanelles, hypoplastic/aplastic clavicles, small height, many impacted teeth, delayed eruption of permanent teeth, supernumerary teeth, and retained primary teeth. When this ailment is diagnosed early, the treatment plan that gives these people the best quality of life can be implemented. Notably, CCD patients have been shown to have mutations in the Runx2 gene. Thus, a better understanding of dental development in CCD may result from additional clarification of the molecular process underlying the creation of extra teeth linked to Runx2 mutations. In this case study, a 17-year-old female patient with Cleidocranial Dysplasia had her unerupted maxillary anterior permanent teeth surgically exposed for aesthetic and orthodontic purposes. Clinical follow-up, the patient's continued therapy, and the family's understanding and drive are all crucial in these situations.

Key Words : Cleidocranial Dysplasia, multiple impacted teeth, supernumerary teeth, hypoplastic/aplastic clavicles.

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INTRODUCTION

In 1897, Marie and Sainton coined the term "cleidocranial dysplasia" (CCD) to describe a condition characterized by delayed ossification of the skull, aplastic or hypoplastic clavicles, and an enlarged transverse diameter of the cranium. Males and females are equally likely to have CCD, which has an incidence of one per million [1]. Mutations in RUNX2, a transcription factor-encoding gene unique to osteoblasts located in the 6p21 chromosome, induce skeletal dysplasia in this autosomal dominantly hereditary condition [2]. The condition is typically an inherited autosomal dominant feature. Nonetheless, the disease manifests intermittently in 20–40% of documented instances.

Clinical Manifestations:

Individuals who are affected usually have small stature, a long neck, and shoulders that slope noticeably [3]. The shoulders can be close to the chest due to a thin thorax [4]. There have also been reports of conduction hearing impairment [6] and delayed closure of the pubic symphysis, coxa Vara, or coxa valga [5]. These patients typically have normal mental development. brachycephalic head with an increased transverse diameter of the cranium,

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A brachycephalic head with an increased transverse diameter of the cranium, delayed ossification of the skull, prominent frontal and parietal bone, occipital bossing, Wormian bone development, ocular hypertelorism, and a broad-based nose are some examples of facial and cranial manifestations [3, 4]. Due to mandibular hyperplasia and mid-face hypoplasia, patients typically exhibit a skeletal Class III malocclusion. Alveolar bone hypoplasia results in a reduction in vertical face growth [7]. Multiple impacted permanent and supernumerary teeth, severe malocclusion, crossbite, and delayed primary tooth exfoliation are examples of oral characteristics [8]. Spontaneous eruption is typically delayed, but the permanent first and second molars are rarely impacted [4]. Even within families, the clinical spectrum varies greatly, ranging from mild cases with merely oral anomalies to severe ones with pronounced skeletal deformities.

Radiographic manifestations:

Short or missing clavicles, delayed ossification of the skull bones, and delayed ossification of the pelvic bones are the characteristic radiological findings of CCD. CCD patients' chest radiographs reveal that their clavicles may be smaller than usual (hypoplasia) or totally absent (aplasia). Usually unilateral or bilateral, the clavicles are hypoplastic or discontinuous; in 10% of cases, they are absent altogether. Acromial end hypoplasia or a sternal end missing with acromial end present are examples of hypoplastic clavicles. Multiple unerupted permanent and supernumerary teeth, as well as prolonged retention and delayed shedding of the primary teeth, are characteristics of CCD. Some times these unerupted teeth are associated with dentigerous cysts. Root resorption and main tooth exfoliation may be delayed, although primary tooth growth is rarely impacted.[9] Nowadays, CBCT imaging is frequently utilized for three-dimensional dentition, which improves anatomical localization of impacted and supernumerary teeth and eliminates uncertainty. The exact placement of a supernumerary tooth in respect to significant structures including the mandibular canal and nearby root apices, the labial cortex of the nasal ridge, the nasal floor cortex, and the nasopalatine duct are among the other relevant details that CBCT provides. CBCT is helpful for both diagnosis and treatment planning in CCD because it clearly shows the location and structure of impacted teeth.[10]

CASE REPORT:

A17-year-old female patient reported with chief complaint of failure of eruption of the upper permanent teeth. She suffered from disabilities in eating due to the distribution and eruption of anomalous teeth, dental aesthetics, and facial appearance. A family history did not reveal similar signs or a hereditary disorder on either side of the family. There was no parental consanguinity. Generalexamination: Diagnosis of CCD was based on the bilateral hypoplasia of the clavicles, the presence of an enlarged cranium, frontal bossing, depressed suborbital region, defective nasal bones, failure of eruption, and multiple supernumerary teeth (fig.1&2). This patient had a moderately short stature and a history of slow growth. Cognitive development was entirely within normal limits. The craniofacial findings included delayed closure of cranial fontanels and suture and brachycephalic. The patient had a skeletal class III malocclusion and a balanced facial pattern. Intraoral examination revealed a mixed dentition, with poor oral hygiene and an angle class III molar relationship.

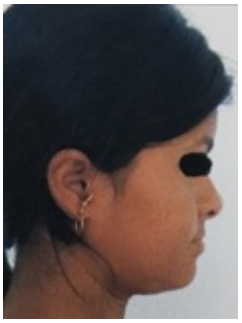

Fig.1 (profile)

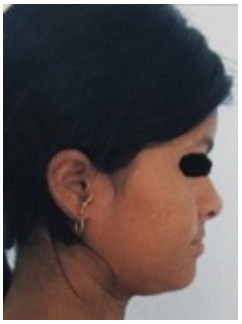

Fig.2 (hypoplastic clavicles)

RADIOLOGICAL EXAMINATION:

Radiological examination was done to analyse the skeletal morphology of the skull and the face, and to observe the dental development. The patient cranial sutures and frontal bossing were observed in the lateral cephalometric radiograph. The clavicles of both sides were hypoplastic and cone shaped thorax with narrow upper thoracic diameter was also identified in the chest radiographs (fig.3)

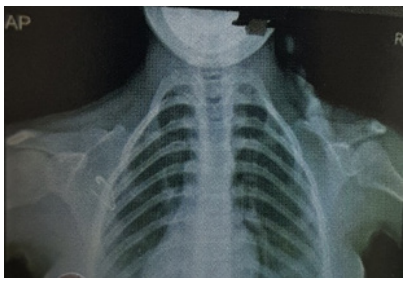

Fig.3 (chest X-ray)

The panoramic radiograph and CBCT scan demonstrated maxillary and mandibular impacted teeth, and presence of one impacted supernumerary tooth in the left maxillary anterior region. There were 16 over-retained deciduous teeth (as 51,52,61,62 deciduous teeth were extracted previously in the private dental clinic) and only 8 permanent molars (first and second molars except third molars) were present in the oral cavity. Other permanent teeth were unerupted though this patient was 17 years old. (fig.4,5&6)

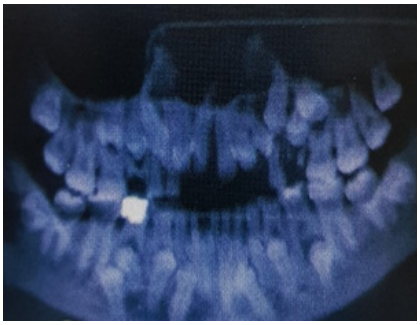

Fig.4 (OPG)

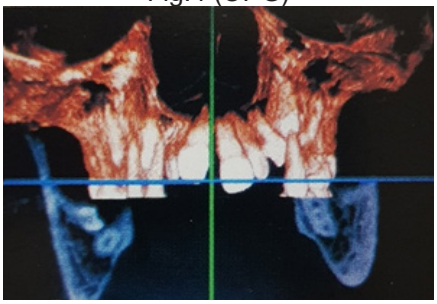

Fig.5 (CBCT scan)

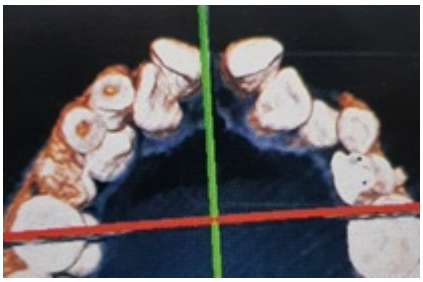

Fig.6 (CBCT scan)

Clinical examination:

This was the sporadic case in which the patient has all the typical features of CCD including hypoplastic clavicle, craniofacial and dental abnormalities. Concave profile due to maxillary hypoplasia was noted. The maxillary arch was constricted and both anterior and posterior cross bite were present. Severe class III malocclusion was obvious from intraoral photograph. (fig.7).

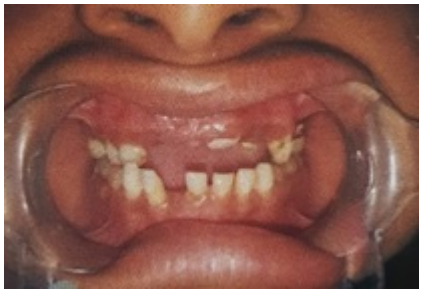

Fig.7 (pre operative intraoral)

MANAGEMENT:

Under all aseptic precautions and under local anaesthesia, extraction of deciduous teeth in right maxillary anterior region was done a full thickness mucoperiosteal flap was raised. The crowns of maxillary central incisors, lateral incisors and right maxillary canine was exposed after removing of overlying bone covering the unerupted teeth. The morphology of the 4 incisors (central and lateral) and right maxillary canine crowns was normal. Orthodontic brackets were bonded to the labial surface of the impacted teeth. (fig.8).

Fig.8 (surgical exposure and orthodontic brackets placed)

after this elastomeric power chains were placed on the brackets. Using closed eruption technique, the gingival flap was repositioned and sutured back in such a way that the bracketed crowns were not exposed to the oral cavity leaving the elastomeric power chains protruding through the mucosa. A prefabricated removable traction apparatus with traction hooks was attached to the mandible and used to guide the impacted maxillary anterior teeth into the correct position. (fig.9).

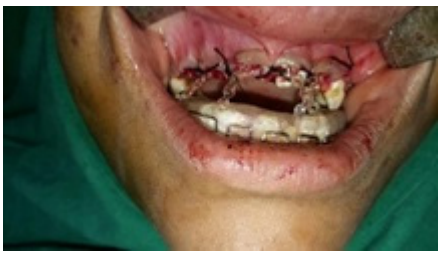

Fig.9 (closure)

Oral hygiene instructions were advised and patient was recalled after 7 days for follow-up and after suture removal wound healing was satisfactory. 5 months post operative photograph showing maxillary centrals, laterals, and right maxillary canine intraorally. (fig.10).

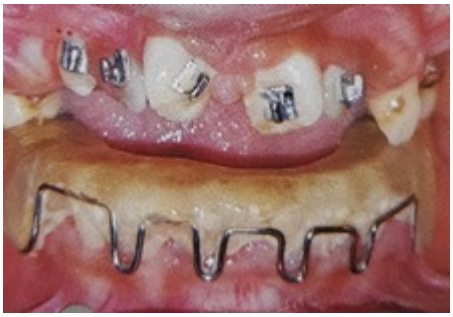

Fig.10 (post operative 5 months follow-up)

DISCUSSION:

A dental team has numerous treatment challenges when dealing with a patient who has cleidocranial dysostosis. Using traction mechanics to guide the unerupted permanent teeth into the arch was the most difficult aspect of this patient's treatment.The preferred treatment for this patient was surgical exposure of the permanent teeth that were not yet erupting, accompanied by orthodontic directed eruption. This prevented the need for a prosthesis that would need to be maintained or replaced multiple times during her lifetime, allowing her to preserve her natural teeth. Both functionally and aesthetically, the patient and family were satisfied. Other treatment options, including as surgical relocation and detachable prosthesis, have drawbacks like a poor prognosis and results that may worsen over time.[11] In the past, various methods for treating the dentition in CCD have been put forth. One could consider the approach proposed by Becker et al.[12,13] to be the most promising. The suggested approach is based on a number of tenets, including (1)the necessity of removing all barriers to the eruption of the permanent teeth that have not yet erupted and applying traction forces at the biologically appropriate time, (2) the necessity of applying extraneous force to induce tooth eruption and the corresponding vertical alveolar development, and (3) the importance of focusing initial efforts on bringing anterior teeth into the mouth as soon as possible for the patient's psychological welfare. For CCD,the intervention's timing is crucial. According to Hitchin and Fairley, the failure of eruption in CCD was caused by the overlying alveolar bone's failure to resorb. When exposed, the impacted teeth would have a typical eruption pattern [14]. Later, Farrar and Van suggested serially exposing the impacted teeth as part of an early surgical treatment [15]. The Toronto-Melbourne technique was demonstrated in Frame's case, when a 9-year-old boy's impacted teeth spontaneously erupted after the eruption path was cleared [16]. The early surgical exposure intervention reduced the difficulty of future orthodontic treatment and promoted the natural eruption of teeth.

CONCLUSION:

CCD is a complicated congenital condition that causes uneven dentition and skeletal abnormalities. Patient preferences, age, financial situation, permanent dentition eruption state, and periodontal and endodontic health all have a significant impact on the treatment strategy. For patients with CCD, therapeutic timing is crucial. The spontaneous eruption of impacted teeth can be facilitated by early removal of the primary and supernumerary teeth as well as the bone covering the affected teeth. Although it takes a long time and requires multiple procedures, combined surgical-orthodontic treatment for adults can result in a nearly full dentition and stable occlusal contact.

If at all possible, the combination of orthognathic surgery and orthodontic therapy can improve the facial profile and lessen the time and difficulties of orthodontic treatment.

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