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CASE STUDY

CLEIDOCRANIAL DYSPLASIA - A CASE REPORT

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ABSTRACT

Cleidocranial dysplasia is a congenital anomaly that is clinically characterized by complete defect or hypoplasia of the clavicle and abnormal ossification of the skull. In the maxillofacial region, it has various abnormalities such as reversed occlusion with hypoplastic midface, persistence of deciduous teeth with delayed eruption of permanent teeth, supernumerary teeth and malalignment. A case of cleidocranial dysplasia in a 17-year-old female visited our hospital. After the preoperative orthodontic treatment for 2 ½ years, Le Fort I osteotomy and Bilateral sagittal split osteotomy for the hypogrowth of the maxilla was performed to advance it anteriorly and set back the mandible. Her midface appearance and the occlusion were remarkably improved after the surgery.

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INTRODUCTION

Cleidocranial dysplasia (CCD) is an autosomal dominant disorder with a prevalence of 1 in 1,000,000 individuals. This condition is characterized by clavicular aplasia or deficient formation of the clavicles, delayed and imperfect ossification of the cranium, moderately short stature, and a variety of other skeletal abnormalities. The gene for cleidocranial dysplasia has been mapped on the short arm of chromosome 6p21, core binding factor a-1 (CBFA1). This disorder can be caused by a mutation in the transcription factor CBFA1 (RUNX2). The CBFA1 gene controls the differentiation of precursor cells into osteoblasts and is therefore essential for membranous as well as endochondral bone formation, which may be related to delayed ossification of the skull, teeth, pelvis, and clavicles. The most characteristic and pathognomonic feature of this disorder is hypoplasia or aplasia of the clavicles, which results in hypermobility of the shoulders allowing the patients to approximate the shoulders in the midline. The face appears small in relation to the cranium with hypoplastic maxillary, lachrymal, nasal, and zygomatic bones. The paranasal sinuses may be underdeveloped. The bones of the middle part of the face are less well developed than the cranial bones.

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The concave profile indicates a skeletal class III with a hypoplastic maxilla. Defects in the skull appear to be always symmetrical. The frontal, parietal, and occipital bones are prominent.

Case report

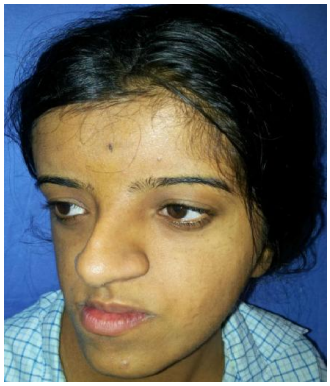
Clinical examination

A 17 year old girl with a history of CCD originally presented with a complaint of retained deciduous teeth and irregular alignment of teeth. Upon general examination she had short stature, aplasia clavicles, brachycephalic skull, concave profile with underdevelopment of the mid face and excessive mandibular growth. Intraoral examination revealed delayed exfoliation of primary teeth, delayed eruption of permanent teeth, impacted third molars, high arched palate and class III malocclusion. Radiographic examination too revealed a skeletal Class III malocclusion. (Fig. 1, 2 and 3)

Treatment planning

1. Extraction of all the supernumerary teeth and retained deciduous teeth.
2. Correction of malalignment of the permanent teeth.
3. Maxillary Lefort I osteotomy, BSSO and correction of skeletal maxillary retrusion, mandibular protrusion.

4. Post-surgical orthodontic treatment to establish the ideal occlusal relationship.
5. Retention to maintain proper occlusion after treatment.



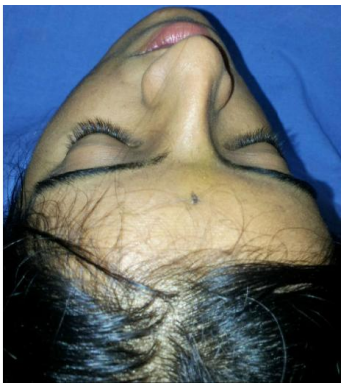
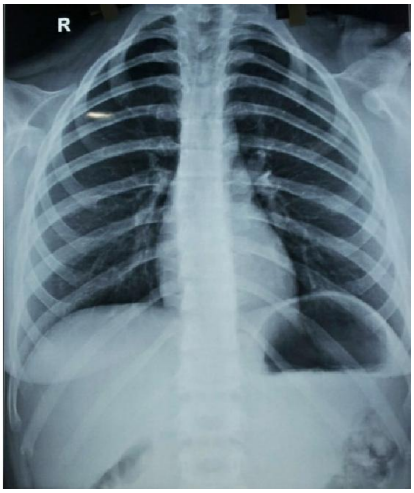
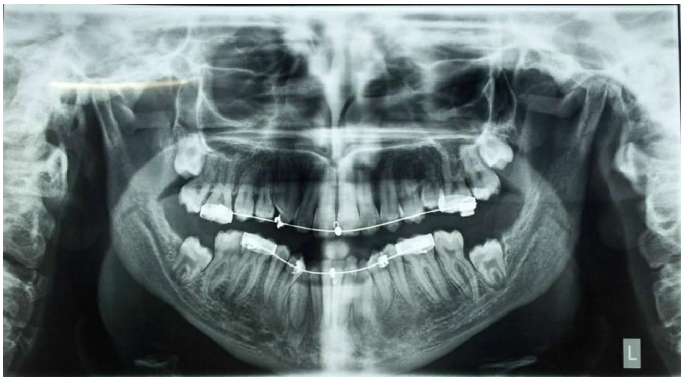


Fig. 1. Pre-operative photographs





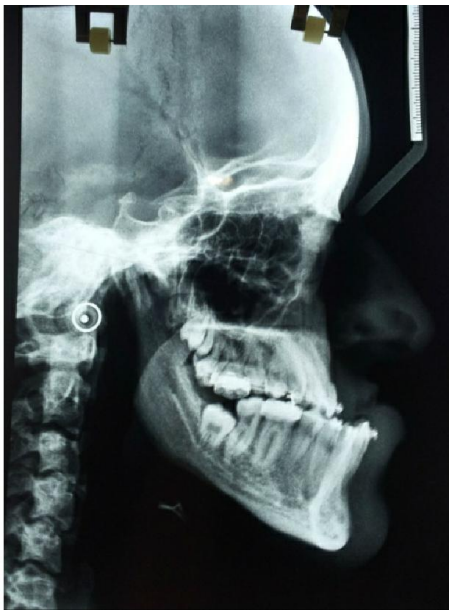
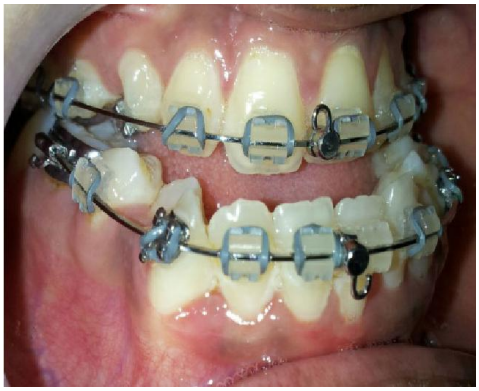
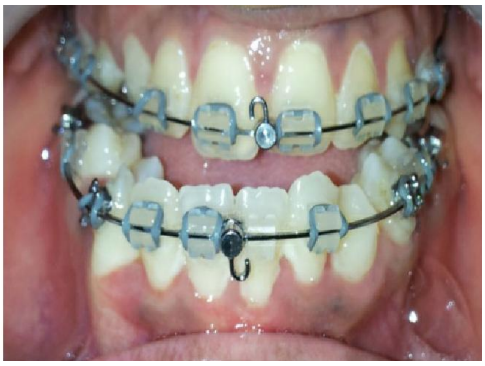


Fig. 2. Chest X-ray, pre-operative OPG and lateral Cephalogram





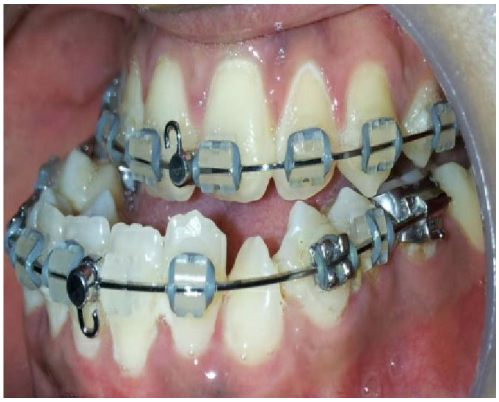


Fig. 3. Pre-operative intraoral photographs

Case management

The patient underwent extraction of retained deciduous teeth and orthodontic treatment for 2 ½ years. Eventually, good arch alignment of the permanent teeth was achieved, although a class 3 interarch relationship and an anterior cross bite remained due to maxillary hypoplasia. Maxillary Lefort 1 osteotomy advancement and BSSO set back was planned.

Bilateral Sagittal split Osteotomy

Incision was placed on the superior end of the ascending ramus, extending anteriorly through the external oblique ridge upto the 3rd molar. Subperiosteal flap was raised, superiorly, laterally and medially over the ascending ramus. Initially extraction of lower third molars was done. Osteotomy cuts were done medial, buccal, distal followed by cut on the inferior border of mandible. Both distal and proximal segments were mobilized. Splints were placed with elastics and internal fixation was done with miniplates. (Fig. 4)

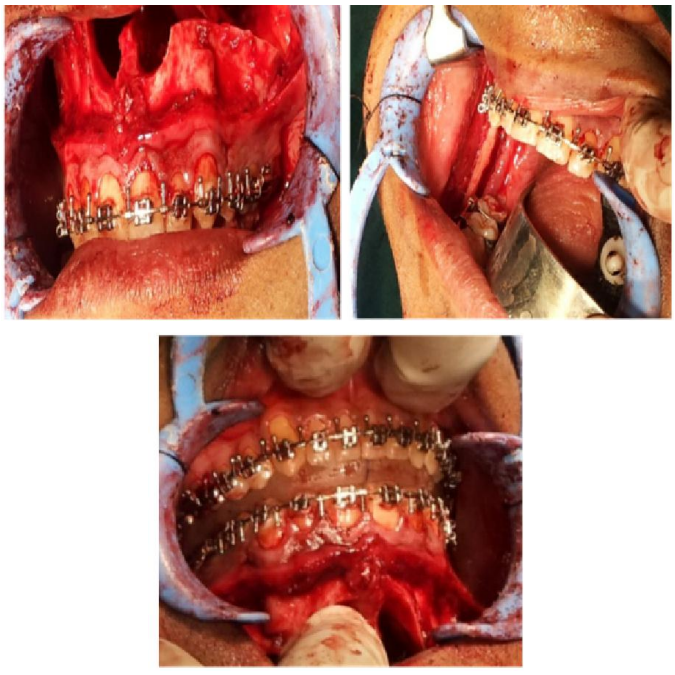


Fig. 4. Intra-operative lefort 1 osteotomy and bsso

Lefort 1 Osteotomy

Maxilla degloving incision was given, subperiosteal flap was reflected, osteotomy cuts were initiated from zygomatic buttress, upto piriform aperture and nasal septum followed by dysjunction. Maxilla was mobilized completely. Splint was placed with elastics by advancing the maxilla. L-form miniplates were fixed at zygomatic buttress bilaterally. Closure was done with 3-0 vicryl sutures.

Post-operative complications

Postoperatively patient had hematoma and swelling in the infraorbital region on the left side. Haemetmeis and epistaxsis were seen 5hours after surgery. It was managed by releasing the intraoral sutures in the distal region of the maxilla for draining the hematoma and nasal pack was inserted. Hydrocortisone 200mg IV, fortwin 30mg IM, otrivin nasal drops were administered. (Fig. 5) & (Fig. 6)

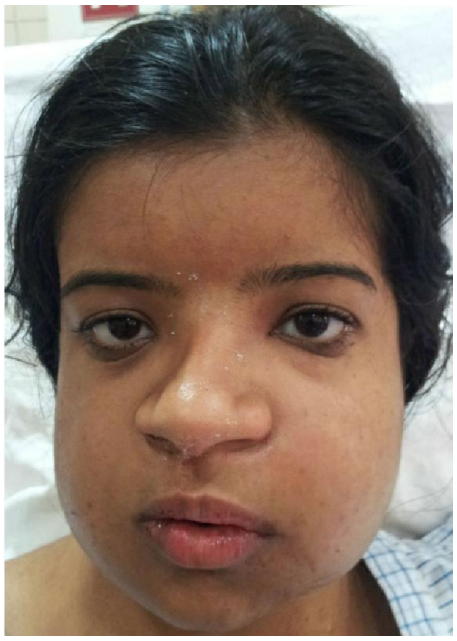


Fig. 5. Immediate post-operative photograph

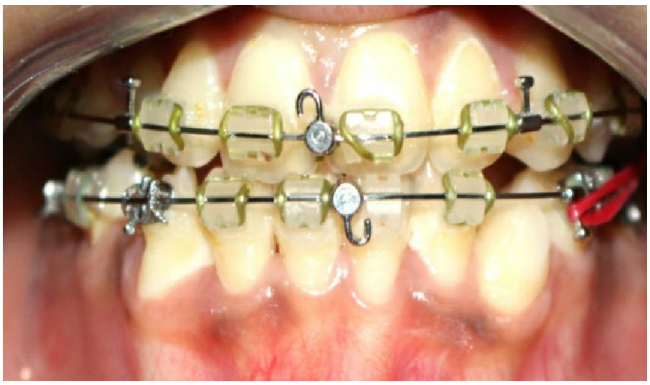


Fig. 6. 6 Months post-operative photographs

DISCUSSION

Management of the orofacial manifestations of CCD is lengthy and challenging, and requires careful planning and execution by an interdisciplinary team. The length of treatment ranges widely and is generally not completed until growth has seized. In this case, a 17 year old girl presented CCD. Initial treatment was extraction of all the retained primary and supernumerary teeth which was followed by orthodontic treatment to establish an intact and aligned dental arch.

Following this, the underlying skeletal deformity was corrected and an improved occlusal relationship was attained through a maxillary advancement lefort 1 osteotomy and BSSO followed by post-operative orthodontic treatment. This combined orthodontic–surgical approach yielded satisfactory results, as the natural dentition could be spared and good occlusal function and esthetics achieved. After the 3 years of interdisciplinary complex treatment, the patient had an acceptable facial appearance and an appropriate oral function.

Conclusion

A very different case of CCD is presented. The combined ortho-surgery treatments successfully met the objectives set out for the case. When establishing an appropriate treatment plan for a patient with CCD, the expected duration of treatment, the age of the patient and the patient's attitude toward treatment are important considerations.

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