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Multiple Supernumerary Teeth Extraction and Surgical Exposure of Multiple Impacted Permanent Teeth with Orthodontic Traction inA Patient with CleidocranialDysplasia

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ABSTRACT: Cleidocranial dysplasia is a rare autosomal dominant inherited disease caused by mutations in the RUNX2 gene on chromosome 6p21 encoding transcription factor CBFA1. The most common clinical findings in these patients are the brachycephalic head, midface hypoplasia, hypoplastic or aplastic clavicles, a depressed nasal bridge with a broad base, open fontanelles, short stature, retention of primary teeth, delayed eruption of permanent teeth, supernumerary teeth and multiple impacted teeth. We are reporting a case of cleidocranial dysplasia in an 11-year-old patient with a retrognathic maxilla, underdeveloped premaxilla and right-side deviation of the nasal septum. Intraorally and radiographically, the patient was presented with seven supernumerary impacted teeth and six permanent impacted teeth within the maxilla; and eight supernumerary impacted teeth with eight permanent buccally impacted teeth visualised within the mandible. Based on the clinical and Cone Beam Computed Tomography findings, multiple extractions of supernumerary teeth and surgical exposure of multiple impacted permanent teeth was done under general anaesthesia followed by planning for orthodontic traction.

KEYWORDS: Cleidocranial dysplasia, Impacted permanent teeth, Orthodontic traction, Multiple supernumerary teeth.

I. INTRODUCTION

Cleidocranialdys plasia is ara recongenital disorder of bone formation with characteristic clinical findings and autosomal dominant inheritance.This rare hereditary skeletal disorder affects both the bones andteeth.[1]The term cleidocranialdys plasia is derived from the ancient Greek words cleido (c o l l a r bone), kranion (head), and dysplasia (abnormal formation).One of the earliest descriptions of this condition was given by Martin in 1765. Hence the disorder is also known as Scheuthauer-Marie-Saiton syndrome or cleidocranialdysostosis.The term 'Cleidocranial dysostosis' was used because it was thought to involve only the bones of intramembranous origin, but subsequent studies showed that the bones of endochondral ossifications were also affected, and therefore the term "cleidocranial dysplasia" currently implicates thegeneralised nature of thecondition rather than a specific diorama. [2]Cleidocranialdys plasia follows an autosomal dominant pattern of inheritance; however,in20-40%of reported cases, the disorder occurs sporadically.Herein, we present a case of a patient with features of cleidocranial dysplasia with multiple supernumerary teeth and multiple impacted permanent teeth and its management by extraction of multiple supernumerary teeth and surgical exposure of impacted permanent teeth and orthodontic traction.

II. CASE REPORT

An 11-year-old female with her parents were referred from a tertiary care centre to the Department of Oral and Maxillofacial Surgery, Kannur Dental College with a chief complaint of prolonged retention of deciduous teeth and multiple missing permanentteeth. Medical history of the

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patient revealed Epilepsy with generalised tonic-clonic seizures,visual hallucinations and behaviour arrest for the past three years. Reports of Electroencephalogram test (EEG) described rhythmic fast beta-activity over left hemisphere with an emphasis over the left parieto-occipital region. Her father showed similar external facial and general appearance with similar dentalconditions.

General physical examination demonstrated a thin build, short stature and slurred speech, the neck appeared long, and the shoulders were narrow with marked drooping. Extraoral examination revealed a prominent forehead with hypertelorism, mild exophthalmos, a depressed nasal bridge, hypoplastic maxilla and a groove along the metopic suture. Mid-facial hypoplasia with frontal, parietal and occipital bossing was also noted, giving the skull a large globular and brachycephalic shape. (Figure 1)

Figure 1

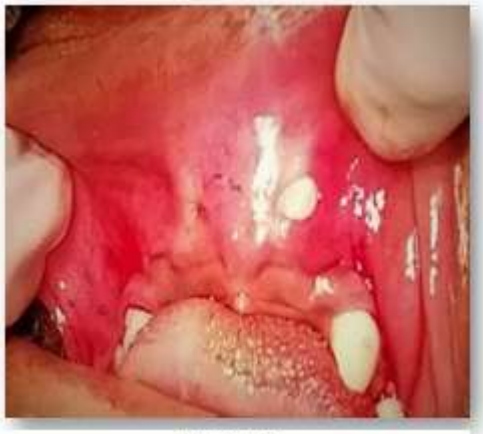

Figure 2

Intraoral examination revealed multiple over-retained primary teeth and multiple missing permanent teeth. Malocclusion and high narrow arched palate were also noted. (Figure 2) Her blood investigation reports were exceptional.

Radiographically, CBCT view of the maxilla (Figure 3) revealed seven supernumerary impacted teeth (shown in red circles) and six permanent impacted teeth (shown in grey rectangles). Displaced/rotated tooth buds were also noted in relation to 15 and 25 (shown in blue pentagons).CBCT view of the mandible (Figure 4) revealed eight supernumerary impacted teeth (shown in red circles) and eight permanent buccally impacted teeth (shown in grey squares), the supernumerary teeth were mimicking premolar. The panoramic view (Figure 5) revealed an increased alveolar bone density overlying the unerupted teeth. Chest x-ray posteroanterior view revealed small and bell-shaped thoracic cage with short, oblique ribs. Aplasia of clavicles (collar bones) was also noted, which leads to hypermobility of the shoulders including the ability to touch the shoulders together in front of the chest. (Figure 6)

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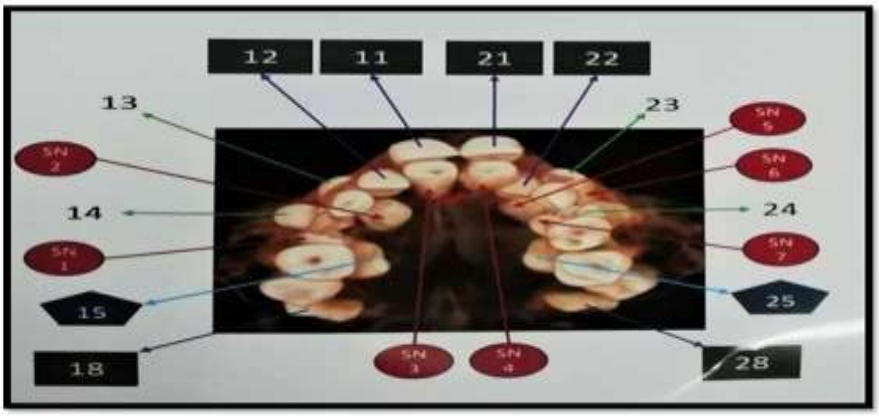

Figure 3

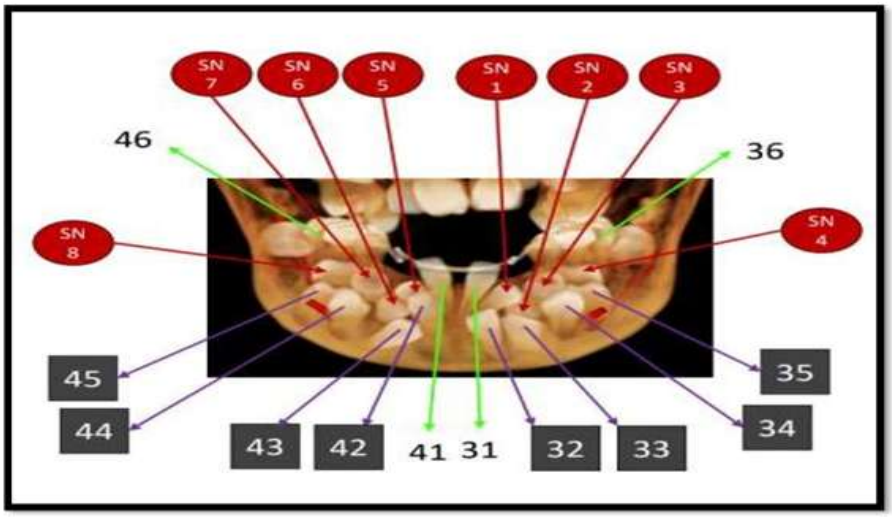

Figure 4

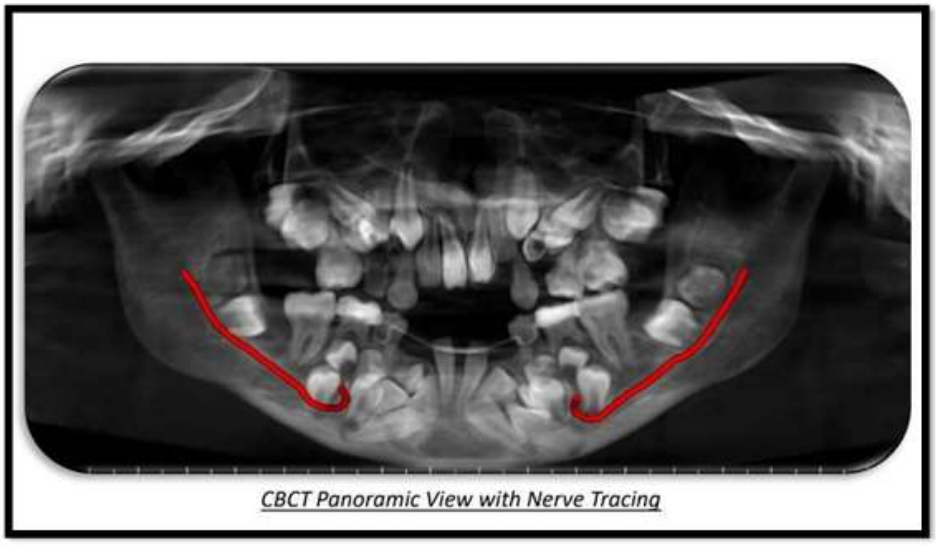

Figure 5

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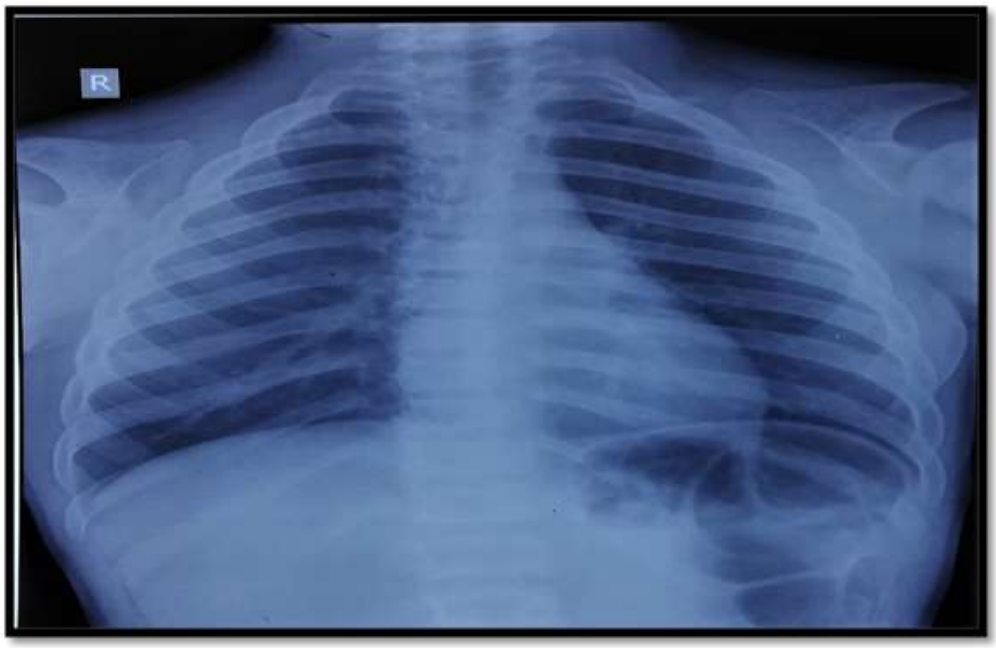

Figure 6

III.DIFFERENTIAL DIAGNOSIS

- Gardner syndrome, characterised bymultiple unerupted supernumerary teeth, impacted permanent teeth, multiple intestinal polyps, skin lesions like epidermoid cysts and fibromas; and osteomas. The absence of multiple osteomas, intestinal polyps and skin lesions rules out the condition in thispatient.
- Craniofacial dysostosis, characterised by premature craniosynostosis, ocular hypertelorism, parrot-beaked nose, nystagmus, maxillary hypoplasia and mandibular prognathism. However, the absence of clavicle in the present case rules out the possibility of this condition.
- Apert syndrome, characterised by craniosynostosis, craniofacial abnormalities and symmetrical syndactyly of the hands and feet. Presence of symmetrical syndactyly differentiates it from cleidocranial dysplasia.
- Pyknodyostosis is a rare defect of osteoclast function in which though many of the clinical findings resemble cleidocranial dysplasia, the increased bone density on x-ray and the absence of supernumerary teeth should readily distinguish between the two.
- Crane- Heise syndrome, presence of genital hypoplasia and dysplastic ears in this condition differentiates it from cleidocranial dysplasia.
- Mandibuloacral dysplasia is mainly characterised by acroosteolysis with progressive loss of bone from the distal phalanges which is not seen in cleidocranial dysplasia.
- Hypohydrotic ectodermal dysplasia is characterised by anomalous dentition which is a similar clinical finding, but the presence of hypohidrosis, onychodysplasia and hypotrichosis in this condition differentiates it from cleidocranial dysplasia.
- Yunis-Varon syndrome the severity of the syndrome, together with limb malformations and a patchy, sometimes sclerotic bone structure differentiates it from cleidocranial dysplasia.
- Focal dermal hypoplasia hypoplastic or aplastic clavicles and anomalous dentition are common features between the two, but the presence of multiple features like skin manifestations and syndactyly differentiates the condition from cleidocranial dysplasia.
- CDAGS syndrome presence of genital and skin manifestations in the condition differentiates it from cleidocranial dysplasia.
- Hypophosphatasia recurrent unexpected fractures seen in this condition differentiates it from cleidocranial dysplasia.

IV. PROVISIONAL DIAGNOSIS

Based on clinical examination, radiographic and other necessary investigations, the patient was diagnosed as having cleidocranial dysplasia.

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V. TREATMENT

Surgical removal of seven impacted supernumerary teeth and two retained deciduous teeth along with the alveolar bone covering the impacted teeth which were causing hindrance in the path of eruption of permanent teeth was done in the maxilla and the mandible under general anaesthesia. Surgical exposure of permanent impacted teeth was done in both maxilla and mandible and bonded with Begg's brackets, and orthodontic traction was given. The surgical site was sutured with 4-0 vicryl suture material. (Figure 7)

VI. REVIEW OF LITERATURE

1. Galal Omami et al.[1] came to a diagnosis of cleidocranial dysplasia on a patient with multipleunerupted and supernumerary teeth based on various clinical and radiographic findings.
2. Stefan Mundlos[2]summarised the clinical and molecular genetics of cleidocranial dysplasia andshowed that a new master gene, CBFA1 was identified as the culprit gene underlying cleidocranial dysplasia.
3. S Mundlos et al.[3]showed that the mutations involving the transcription factor CBFA1 colonisedon chromosome 6p21 cause cleidocranial dysplasia.
4. Chin-Yun Pan et al.[4]showed different craniofacial features of cleidocranial dysplasia and the insights into its pathogenesis that may assist in the development of new treatment.
5. Ravi Prakash S Mohan et al.[5] illustrated the clinical features, radiological features anddental abnormalities in a rare case of cleidocranial dysplasia.
6. A Impellizzeri et al.[6] did a review and report of a family with cleidocranial dysplasia with the delayed eruption of permanent dentition and maxillary contraction and showed the importance of orthodontic techniques along with surgical approach and the use of cone-beam for better understanding of the cleidocranial dysplasia patient's dental situation and a better treatment.

VII. DISCUSSION

Cleidocranial dysplasia is an autosomal dominant generalised skeletal dysplasia affecting bones of intramembranous and endochondral ossification.Cleidocranial dysplasia results from a Runx2 gene mutation in the small arm of chromosome 6 at 6p21. Runx2 mutations, which functions as a heterodimer with core-binding factor ß (Cbfß), are found in most individuals with cleidocranialdysplasia.[3] Theprevalence of cleidocranial dysplasia isestimated at 1:1 million without sexor ethnic group predilection.[1,4] Clinical features includeopen fontanelle or delayed closure offontanelle, persistently open skullsutures, presence of Wormianbones, skull is usually large, broad and brachycephalic type (Arnoldhead), ocular hypertelorism and a mildexophthalmos, a depressed nasal bridge and midfacialhypoplasia, prominent frontal, parietal, and occipitalbones, a narrow high arched palate and infrequently cleftpalate, delayed tooth eruption and enamelhypoplasia, a long neck, narrow sloping shoulders and shorterstature hands with finger length asymmetry due to extraepiphysis in metacarpals II andV, conductivedeafness, scoliosis, hypoplasia of maxillae and a skeletal class IIItendency.[2,3]

Radiographically,multiple unerupted supernumerary teeth and multiple impacted permanent teeth, a parallel-sided ascending ramus ofmandible, a slender and pointed coronoid process, coarse trabeculation of the mandible, cystformation with supernumerary teeth and increased density of the alveolar crest bone over unerupted teeth, a thin zygomatic arch with a severe downwardtilt are usually seen. Skull radiographs show calvarial thickening especially over the occiput and Wormianbones, chest x-ray shows a narrow thorax, oblique ribs and absence or hypoplasia ofclavicles. [3]Otherradio graphic findings includes coliosis, vertebralanomalies, spinabifidaoccultaand a wide pubic symphyseal space with a 'chef's hat' appearance of the femoral head.[5]

Different approaches to the treatment of the dentition in cleidocranial dysplasia patients have been proposed in the past based on several premises for the patient's functional, aesthetic and psychological well-being.[6]

- The "Toronto-Melbourne approach"is a combined oral surgery and orthodontic approach based on timely serial extraction of deciduous teeth in the initial stages and surgical exposure of impacted permanent teeth later and it involves multiple surgical interventions under general anaesthesia.
- The "Belfast-Hamburg approach" is a single surgical approach that limits the number of surgeries that is the extractions of supernumerary and deciduous teeth and surgical exposure of permanent impacted teeth to a single episode under general anaesthesia.
- The"Jerusalem approach" is based on two surgical interventions depending on root development of the permanent dentition; in the

first phase, the treatment of anterior teeth is involved at the age of 10-12 years, and in the second phase, the posterior teeth are treated at around 13 years or older under general anaesthesia.

- The "Bronx approach" uses an interim overdenture prosthesis during the long course of treatment

The timing of diagnosis in cleidocranial dysplasia patients is essential in choosingan appropriate treatment plan and in attaining a successfulresult. From an oral surgeon's perspective, elimination ofclinical pathology followed by achieving a functional dentition and an overall aesthetically satisfying facial appearance should be the ultimate treatmentobjectives.

VIII. CONCLUSION

Cleidocranial dysplasia is a rare condition whichwith proper anticipatory guidance, early diagnosis and timely orthodontic- surgical intervention for dental and skeletal conditions; the patients with cleidocranial dysplasia usually lead healthy and productive lives, and it is also vital for theirpsychological well-being.

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