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Case Report
Cleidocranial Dysplasia

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Cleidocranial Dysplasia: A Case Report

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Cleidocranial dysplasia (CCD) is a rare autosomal dominant disorder. This congenital defect is characterized by hypoplastic or absent clavicles, delayed skeletal development .Along with hypoplasia of orofacial and skeletal structures the condition is also reported to have multiple supernumerary teeth, retained deciduous dentition, non-eruption of permanent tooth. We report a case of CCD in a 26 year-old female who had classical diagnostic feature.

Keywords: Cleidocranial dysplasia, multiple missing tooth, absence of clavicles

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Introduction

Cleidocranial dysplasia (CCD) is a rare congenital autosomal dominant condition, usually presents with skeletal defects of multiple bones, the most characteristic feature of which are partial or complete absence of clavicles, wormian bones ,presence of open skull sutures and delayed closure of fontanels, and. These patients usually complain of multiple missing teeth or delayed eruption of teeth. Ours was a typical case of CCD in a 26 year old female patient who had classical diagnostic feature of this syndrome.

Case Report

A 26 year old female patient reported our department with complains of missing multiple teeth (Fig 1). Her family history showed no similar history. She was moderately built and nourished. Extraoral examination showed the patient had a flat face, hypertelorism and sunken nasal bridge. There was evidence of clubbing of the digits (Fig 2), shoulders appeared narrow and showed marked drooping. On palpation, there was bilateral absence of clavicles and patient was able to approximate both shoulders in the midline on the chest. Intraoral examination revealed the presence of both deciduous and permanent tooth with several missing tooth in a narrow constricted maxillary and mandibular arch.

Orthopantomogram (Fig 3) showed mandible with coarse trabeculation, narrow ascending ramus and slender pointed coronoid processes and multiple impacted tooth .The posteroanterior view of chest radiograph (Fig 4) revealed complete bilateral absence of clavicle. Based on the typical clinical examination and radiographic feature, a diagnosis of cleidocranial dysplasia was made.

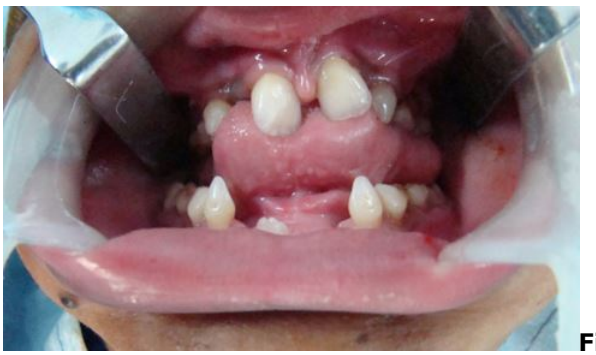

Fig 1: Showing the Intra oral Picture

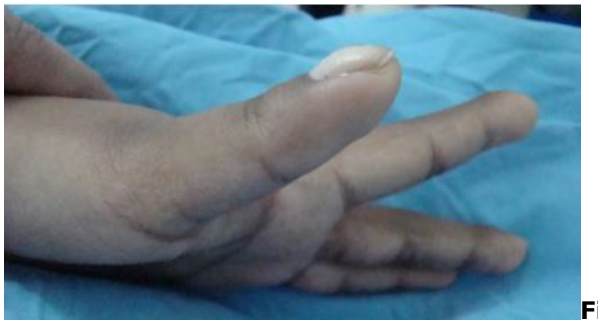

Fig 2: Clubbing of the digits

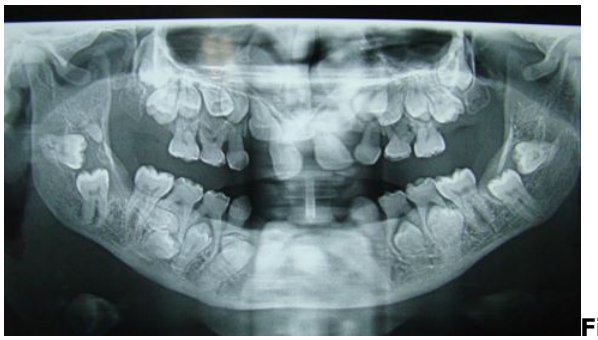

Fig 3: OPG showing multiple impacted tooth and retained deciduous tooth

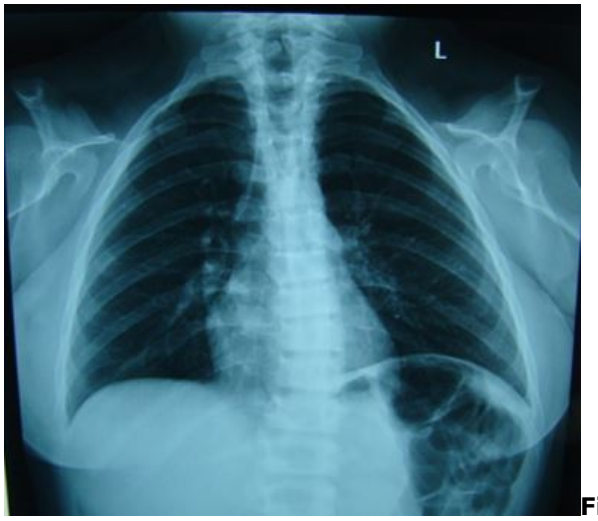

Fig 4: Chest X-ray (PA view) showing bilateral absence of clavicle

Discussion

Cleiodocranial dysplasia (CCD), also known as Marie and Sainton's disease; Scheuthauer - Marie – Sainton syndrome; Mutational Dysostosis and cleidocranialdysostosis [1].

CCD is generally accepted as an autosomal disorder, affecting both sexes with equal frequency. In 1765 CCD was first described by Martin [2] and in 1897 Marie and Sainton termed

It as 'cleidocranial dysostosis' [3]. The aetiology of cleidocranial dysplasia is unknown but Genes has been mapped for this kind of disorder ([4, 5]).

CCD is usually seen involved with the bones that get ossified early in foetal life; the clavicle is the first bone to show ossification. The disease characteristically causes a retardation or partial aplasia of bones that ossify intramembraneously ([4, 6]). It is rather an uncommon disorder with a prevalence of 0.5 per 100,000 live births [7]. The condition is characterised by abnormalities of the clavicles, skull and jaws as well as by occasional stunting of long bones. The patient is usually of short stature, but the neck appears relatively long and the most characteristic and differentiating skeletal feature is that of partial or completely absence of one or both clavicles and drooping of shoulders can be seen as a result of this. Similar features were seen in our case.

The intraorally the condition is characterised by the presence of multiple supernumerary teeth and the delayed eruption of the succedaneous dentition [8]. The middle third of face is usually hypoplastic both in the antero-posterior as well as in vertical dimensions. Hard palate is high and narrow. The causes of unerupted teeth in CCD is said to be [9] (i) a disturbance of bone resorption and delay in formation of tooth and its maturation (ii) delay and defectiveness in formation of cellular cementum or (iii) interposed fibrous tissue acting as a barrier to eruption resulting in a non union between the dental follicle and the mucosa.

The diagnosis is confirmed by radiographs [10]. Chest radiograph in these patients shows hypoplastic or aplastic clavicle. Radiographically, the mandible and maxilla contained many unerupted teeth that are greatly deviated from the normal position [10] similar features were reported in our case.Crane- Heise syndrome, mandibuloacral dysplasia, pycnodysostosis, yunis varon sydrome, CDAGS syndrome and hypophosphtasia etc. are usually considered in the differential diagnosis of CCD [11].

There has been no specific treatment for the patient with CCD; the correction of the dentofacial deformities both aesthetically and functionally has been mentioned in literature. Most of the times the age of the patient determines the treatment plan; in the early age group, the supernumerary teeth are Surgically exposed and orthodontic traction is applied with the help of brackets until the teeth are brought into normal occlusion [12].Orthognathic surgery has been suggested by Trimble et al, [13] for the correction of middle - third of the face. Removable prosthesis has been advocated by Winter after the extraction of all the primary and permanent teeth [13] but these patients most of the time ends up with alveolar hypoplasia which compromises the retention of prosthesis. Suggested removal of only the erupted teeth and use of a removable prosthesis inorder to minimize the alveolar bone loss was proposed by Pusey & Durie [14].It is not always easy to determine the most appropriate aesthetic and functional treatment for dentofacial deformities in patient with CCD ,so the ideal treatment for these patients still remains a myth.

Conclusion

A multidisciplinary approach to management of dental abnormalities on a long-term basis is necessary in the treatment of CCD [15]. Early diagnosis of CCD ensures an aesthetic facial appearance, functional occlusion and better quality of life for the patient.

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