

CASE REPORT

CLEIDOCRANIAL DYSPLASIA: A CASE REPORT

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ABSTRACT:

The Cleidocranial dysplasia is a dominant autosomal inheritance disease which is actually rare in occurrence. This has no race or gender predilection. Dental surgeons are usually the first professional to come across this disease, as patient first visit dentist with the chief complaint of missing teeth which is one of the most common clinical feature associated with this. In this series also the patient reported to us with the chief complaint of missing teeth. In this article we are presenting one case of patient with Cleidocranial dysplasia.

Key words : Cleidocranial dysplasia, Developmental anomaly

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This article may be cited as: Sharma R, Awasthy D, Chauhan R, Singh D. Cleidocranial Dysplasia: A Case Report. J Adv Med Dent Sci Res 2016;4(4):125-128.

Access this article online	
Quick Response Code 	Website: www.jamdsr.com
	DOI: 10.21276/jamdsr.2016.4.4.27

INTRODUCTION

Cleidocranial dysplasia is a rare dominant autosomal disease. It is a developmental anomaly which affects skeleton and teeth¹. The most common seen skeletal anomaly are hypoplastic clavicle, late closure of the fontanelles, presence of open skull sutures and multiple wormian bones². Cleidocranial dysplasia is also known as Marie and Sinton Disease, Scheuthauer Marie-Sainton Syndrome and Mutational dysostosis³. In these patients clavicle is either small or in not present so patients is able to move shoulders medially without any discomfort. It is also seen that there is late closure of fontanelles resulting in frontal bossing. For dentist it is of more clinical significance due to involvement of facial bones, altered eruption patterns

of teeth, presence of multiple impacted supernumerary teeth and retained deciduous teeth⁴. This disorder primarily affects bones showing intra-membranous ossification, i.e. calvarial bones and clavicles^{1,5}. Maxilla and paranasal sinuses are also underdeveloped. Sometimes some other bones are also affected which includes long bones, the vertebral column, the pelvis and the bones of hands and feet⁶. It is mostly seen that Adults with Cleidocranial dysplasia have mixed dentition in their oral cavities as there is prolonged retention of deciduous dentition and delayed eruption of permanent teeth in them. Frequently they show a large number of unerupted supernumerary teeth⁷

CASE REPORT

An eighteen year boy reported to the department of oral medicine and radiology with the chief complaint of missing teeth in upper anterior. His family history was non contributory but his medical history was contributory. On general examination the patient did not had normal height and weight. His IQ was normal; Brachycephaly, frontal bossing, depressed mid-face profile and hypertelorism were noticed. Patient was able to move his shoulder towards mid line, suggestive of hypoplastic clavicle which is one of the suggestive features of Clediocranial dysplasia. Oral examination showed prolonged retention of deciduous teeth and failure of permanent mandibular and maxillary teeth to erupt. The patient was

subjected to critical radiographic examination of chest, skull and major joints. (Figure 1-patient oral cavity)

Cephalometric and panoramic radiographs with supplementary periapical and occlusal views were done. The X-rays revealed the following positive findings:

Skull: Open sutures, large wormian bones, calvarial thickening and characteristically sunken sagittal suture giving the skull a flat appearance.(Figure 2-skull radiograph)

Chest: Right clavicle was absent while left clavicle was hypoplastic.

Hands : Metacarpal pseudoepiphyses



Figure 1: Intra-oral photograph



Figure 2: Skull radiographs



Figure 3: OPG

DENTAL FINDINGS

Cephalogram showed hypoplastic nasal bone and maxilla while the mandible was prognathic having characteristic obtuse mandibular angle. OPG and intraoral (occlusal and periapical) radiographs revealed abnormally retained. Primary teeth and so many impacted permanent and supernumerary teeth (excluding second and third molars). (Figure 3)

DISCUSSION

The most significant clinical findings of Cleidocranial dysplasia are Frontal bossing and excessive mobility of shoulder girdle^{1,4}. The clinical findings of CCD, although present at birth, are often either missed or diagnosed at a much later date. The radiographic evaluation of patients is the most important and reliable means to confirm the diagnosis. CCD is characterized by abnormalities of the skull, teeth, jaws, and shoulders girdles as well as by occasional stunting of the long bones⁸.

One of the outstanding oral finding is prolonged retention of the deciduous teeth and subsequent delay in eruption of the succedaneous teeth³. Scientists have postulated various views regarding the etiology of non-eruption, such as lack of cellular cementum², defectiveness in post cementum formation⁶, presence of thick connective tissue between oral epithelium and dental follicle⁹, delayed tooth formation and maturation¹⁰.

Characteristic dental findings are numerous supernumerary teeth, particularly in mandibular

premolar and maxillary anterior regions. The removal of primary or supernumerary teeth does not usually promote eruption of unerupted permanent teeth.³ The permanent molars generally erupt at proper time.^{1,4}

A multidisciplinary approach to treatment of these patients utilising a pedodontist, an orthodontist and an oral surgeon is recommended. The retained primary teeth should be restored if they become carious since extraction does not necessarily induce eruption of the permanent teeth³. The current mode of therapy for the dental anomalies is:

- Planned removal of nonresorbing primary teeth
- Surgical removal of supernumerary teeth
- Surgical exposure of permanent teeth
- Orthodontic alignment
- Growth modulation of maxilla and mandible with Delaire's face mask and chin cup, as per the requirement.
- When growth is complete consideration of orthognathic surgery in severe skeletal Class III malocclusion cases.

CONCLUSION

Cleidocranial dysplasia is generally missed or diagnosed at a much later time. This is generally diagnosed incidentally. Family history, excessive mobility of shoulders and radiographic pathognomonic findings of the chest, skull and jaws are useful in confirming the diagnosis.

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Source of support: Nil

Conflict of interest: None declared

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