

Cleidocranial Dysplasia (Yunis Varon syndrome) A case report

Author: Dr. Rahamathulla Khan M.D.S
Head of Department,
Department of Orthodontics
Dental Faculty, Al-Tahadi University, Sirte

Co-Author: Dr. Verdine Virginia Antony M.D.S
Lecturer, Department of Periodontics
Dental Faculty, Al-Tahadi University, Sirte

ABSTRACT

Cleidocranial dysplasia is a rare skeletal dysplasia characterized by short stature, distinctive facial features and narrow, sloping shoulders caused by defective or absent collarbones (clavicles). Major symptoms may include premature closing of the soft spot on the head (coronal), delayed closure of the space between the bones of the skull (fontanelles), narrow and abnormally shaped pelvic and pubic bones and deformations in the chest (thoracic region). Delayed eruption of teeth, pseudo anodontia, multiple impacted supernumerary teeth, a high arched palate, a wide pelvic joint, failure of the lower jaw joints to unite, and fingers that are irregular in length may also be present. Cleidocranial dysplasia is inherited as an autosomal dominant genetic trait.

This article presents a case report of a 17-year old male patient with the classical features of cleidocranial dysplasia.

Key words: Cleidocranial dysplasia, pseudo anodontia, multiple impacted supernumerary teeth, autosomal dominant.

INTRODUCTION:

:

Cleidocranial dysplasia is an autosomal dominant condition with generalised dysplasia of bone and teeth. This disorder was detected in the skull of a Neanderthal man.[\[1\]](#) The more obvious features of the defect in the clavicle and cranium prompted Marie and Sainton to utilise the term cleidocranial dysostosis for this condition.[\[2\]](#)

The disorder is characterised by frontal and parietal bossing, brachycephaly,

persistent open anterior fontanelle, and often with late closure of the other sutures. There may be macrocephaly. Associated with it is late or incomplete development of accessory sinuses and mastoid air cells, small sphenoid bones, calvarial thickening and wormian bones. Facial bones are small, mid-face hypoplasia with low nasal bridge, narrow high-arched palate, hypertelorism and conductive deafness may be seen. [3] The primary dentition appears late and is frequently incomplete. The secondary dentition is similarly delayed and often mal-aligned with some teeth malformed and hypoplastic. Supernumerary teeth are common, especially in the premolar area. There may be associated enamel hypoplasia. Proportionate short stature may also be exhibited. [4]

The clavicles are either absent or severely hypoplastic which leads to abnormally low positioning of the shoulders that can frequently be opposed anteriorly. Thorax is narrow with short oblique ribs and the most serious manifestation is the occasional occurrence of a severe scoliosis. Hand anomalies may include asymmetric length of fingers. Other skeletal abnormalities include delayed mineralization of pubic bone, wide symphysis pubis, narrow pelvis and spondylolysis. [2, 3] Though there is slight to moderate shortness of stature, intelligence is usually normal.

Hearing and dental problems may be anticipated. Oral manifestations include poor development of the premaxilla, relative mandibular prognathism, pseudo anodontia and multiple impacted supernumerary teeth. [5]

CASE REPORT

:

A 17-year-old male patient reported to the department of orthodontics with the chief complaint of crowded teeth in the upper as well as the lower jaws. The patient had a moderate build, clubbing, slurred speech, and shrugged shoulders. Intra oral examination revealed crowding of teeth in relation to the upper and lower arches and a high arched palate. The face showed increased inter-canthal distance. The profile view revealed frontal bossing, depressed nasal bridge and infra-orbital margins. The orthopantomogram revealed multiple unerupted teeth. Postero-anterior view chest radiograph revealed hypoplastic clavicles. Lateral cephalometric radiograph revealed hypoplastic maxilla, zygoma and infra-orbital rim.



Fig.1: Facial view showing increased intercanthal distance, depressed nasal bridge and infra-orbital margins.



Figs.2a, 2b & 2c: Intra-oral view showing crowding in relation to the maxillary and mandibular arches.

Paranasal sinus view revealed absence of maxillary sinus, zygoma and increased size of ethmoidal sinus. Considering all the clinical, oral, radiographic and skeletal anomalies he was diagnosed as a case of cleidocranial dysplasia with absence of clavicles.

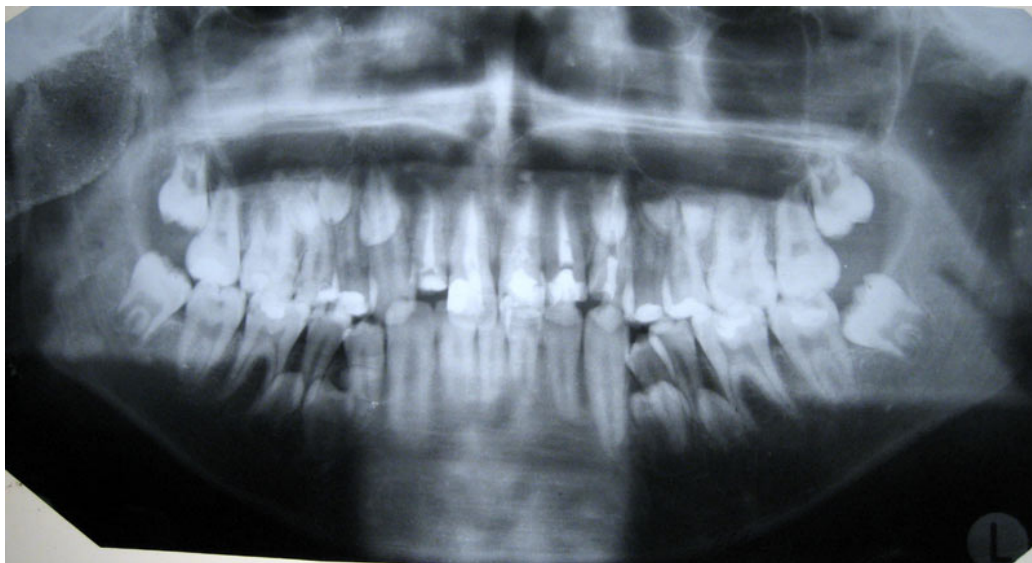


Fig. 3: Orthopantomogram showing multiple unerupted teeth.



Fig. 4: Postero-anterior view chest radiograph revealing hypoplastic clavicles.

DISCUSSION:

Abnormalities of the skull, teeth, jaws, and shoulder girdle as well as occasional stunting of the long bones characterize Cleidocranial dysplasia.[1,2,3]

A common feature of patients with cleidocranial dysplasia is delayed shedding of deciduous teeth and failure of eruption of permanent teeth, which result in pseudo anodontia or the presence of multiple supernumerary teeth[3,4,5] Patients with this disorder need extensive dental therapy in order to maintain efficient chewing.

Oral features seen in our case reports included crowding of teeth in relation to the upper and lower arches and a high arched palate,. Radiographic features are characterized by the presence of numerous unerupted supernumerary teeth both in the upper and lower jaws. [4, 5]

REFERENCES:

1. Shafer WG, Hine MK, Levy BM et al: A textbook of oral pathology, 4th edition, WB Saunders Co, 1983, P678-680.
2. Jensen BL, Kreiborg, S (1990) Development of the dentition in cleidocranial dysplasia. *Journal of Oral Pathology and Medicine* 19:89-93.
3. Gorlin RJ, Pindborg JP: *Syndromes of the head and neck*. McGraw Hill book Co, London, 1964, P. 138-145.
4. De Nguyen T, Turcotte JY (1994) Cleidocranial dysplasia: review of literature and presentation of a case. *Journal Canadian Dental Association* 60(12): 1073-8.
5. Richardson A, Deussen FF (1994) Facial and dental anomalies in cleidocranial dysplasia: a study of 17 cases. *International Journal of Pediatric Dentistry* 4(4): 225-31.