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Original Research Paper

Dental Science

CLEIDOCRANIAL DYSPLASIA- NON -SURGICAL TREATMENT OF RARE CASE

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ABSTRACT Cleidocranial dysplasia is an autosomal dominant condition caused by mutation of RUNX2, characterized by generalized dysplasia of the bones and teeth. Affected individuals have short stature, atypical facial features, and skeletal anomalies affecting mainly the skull and clavicle. The dental manifestations are mainly delayed exfoliation of the primary teeth and delayed eruption of the permanent teeth, with multiple impacted supernumeraries, and absence of cellular cementum. The frequency of this disorder is 1 per million individuals. Here we report a rare case of CCD in a 55-year-old male patient having most of the characteristic features of this syndrome with chief complaint of bleeding gums & swelling in gums since 2 years .

KEYWORDS : Cleiodocranial dysplasia. Supernumerary Teeth. Hypoplastic Clavicles.

INTRODUCTION

Cleidocranial dysplasia also termed as Marie & Saintain disease ,Mutational dysostosis was a congenital disorder of bone formation manifested with 1) Clavicular hypoplasia or Agenesis with narrow thorax 2) Delayed closure of frontanels ,3) Disorders of the jaws & dentition 4) skeletal abnormalities (ARNOLD HEAD)

This syndrome is familial & is transmitted as an autosomal dominant trait several chromosome abnormalities have been reported to be associated with this syndrome ,including rearrangement of long arm of chromosome 8 (8q22) & the long arm of chromosome 6 .Mutations in the core -binding factor alpha -1 (CBFA1) gene ,located on chromosome 6p21 have been shown to be the cause of cleidocranial dysplasia.

Clinical features-

General features-

CCD is characterized by abnormalities of the skull ,teeth , jaw & shoulder girdle as well as by occasional stunting of the long bones .In the skull the fontanels often remain open or at least exhibit delayed closing , & for this reason tend to be rather large. Based on the cephalic index ,the head is brachycephalic or wide & short ,with the transverse diameter of skull being increased . ARNOLD HEAD named after the descendants of a chinese who settled in south africa & changed his name to arnold . There is clavicular disturbance the patients have an unusual mobility of the shoulders & may be able to bring their shoulders forward until they meet in the midline .

Oral manifestations-

Patients with CCD exhibit a high ,narrow ,arched palate ,& some time with cleft palate. Maxilla is almost reported underdeveloped & smaller than mandible. Prolonged retention of deciduous teeth & subsequent delayed eruption of the seccedaneous teeth , sometime this delay in tooth eruption is permanent

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There is absence or paucity of cellular cementum on the roots of permanent teeth, & this may be related to failure of eruption so frequently seen. A surprising & unexplained feature was the absence of this cementum on the erupted teeth in both dentitions with no increased thickening of the primary acellular cementum . The manner of anchorage of periodontal fibers & maintenance of periodontal ligament. [1,2]

Radiographic Examination-

Reveals the widely patent anterior fontanel & sutures with wormian bones in cranium. The clavicles typically are reduced to single or double fragments on each side with middle part being deficient. Frequently the changes are symmetric .Hands & feet demonstrate various anomalies including shortening & broadening of carpal ,metacarpal ,tarsal ,metatarsal bones[3]

Treatment

Treatment of CCD includes dental procedures to address the affects of retention of deciduous dentition, presence of supernumerary teeth, noneruption of the permanent dentition along with related malocclusion and periodontal conditions accompanying them. Maintenance of periodontal health is of prime concern in order to allow for an absolute and complete oral rehabilitation of the patient. It was found that a diagnosis of the disorder should be made early so that formation of supernumerary teeth can be diagnosed and early intervention undertaken. Supernumerary teeth that pose a significant obstacle to tooth eruption begin their mineralization 4 years later than the corresponding permanent teeth.[4]

Case Report

A 55-year old male patient having Cleidocranial Dysplasia was referred to the Department of Periodontics with a chief complaint of bleeding gums and swelling at multiple sites in the gums for periodontal treatment. On performing an oral examination it was noticed that the patient had several missing and unerupted teeth. Over-retained deciduous molars and incisor were present. The color of the gingiva was reddish pink with evident pigmentation, all teeths are mobiles.

FAMILY HISTORY

Patient give family history that his children's die with in 20days after birth & having some tumour like abnormalities according to the doctors .Patients having no brothers, sisters.

Clinical features

EXTRAORAL FINDINGS

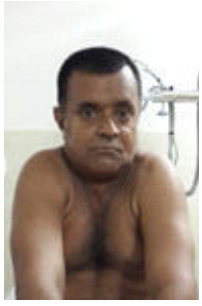
The extraoral findings were concurrent with the classical findings in a patient of CCD including: high prominent forehead, depressed nasal bridge, frontal bossing, hypoplasia of the maxilla along with false prognathism of the mandible resulting in a Dish Face appearance.The patient also exhibited features such as drooping shoulders with narrow and funnel shaped chest, excessive joint mobility and an ability to easily approximate his shoulders in front of the sternum.

INTRAORAL FINDINGS-

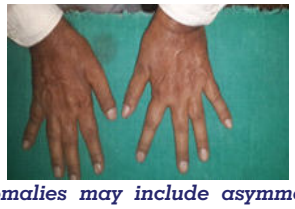
There is narrow highly arched palate, the retention of deciduous teeth, delayed eruption of permanent teeth, and the

presence of large numbers of impacted supernumerary teeth are all classical oral findings in cleidocranial dysplasia patients which were notice in this patient with other abnormal finding like .

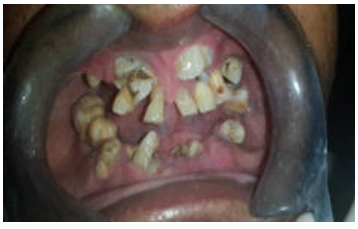
There was short roots with spike like apices often seen in unerupted teeth in this case due to inadequate bone resorption. Also occurrence of ankylosis in the deciduous teeth is indicative of tooth resorption which has ceased and the new bone that has been formed is very dense. Deciduous teeth and first permanent molars come into normal occlusion because there is little or no bone over them that may impede their eruption. This is a common feature of CCD patients which all are present in our patient.



a) patient easy made his shoulders in front of the sternum



b)- Hand anomalies may include asymmetric length of fingers

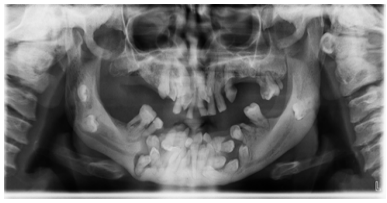


c) Pre-operative photograph of patient

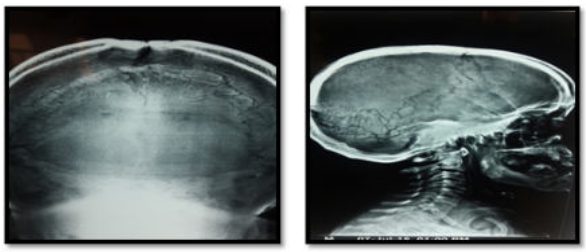
Radiograpic examination-

Radiological investigations included an orthopantomogram, lateral cephalogram, postero-anterior view of the skull, chest radiograph, anteroposterior and lateral views of the spine, antero-posterior views of the pelvis and both hips, shoulders x-ray both sides .

- The orthopantomogram revealed multiple impacted permanent and supernumerary teeth in the incisor and bicuspid regions of the maxilla and mandible (total number of teeth present, 46). The follicular spaces of some impacted teeth were enlarged in the mandibular incisor and right parasymphysis region, suggesting cystic transformation. The ascending ramus of the mandible appeared narrow, with nearly parallel borders and coarse trabeculation. The coronoid process appeared slender and pointed. The zygomatic arch was thin with a downward tilt, and there was increased density of the alveolar crest bone overlying the unerupted teeth

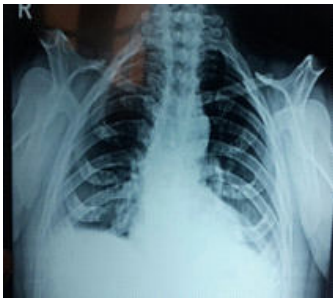


d) Orthopantomogram showing 46 teeth including multiple unerupted and supernumerary teeth along with other features.



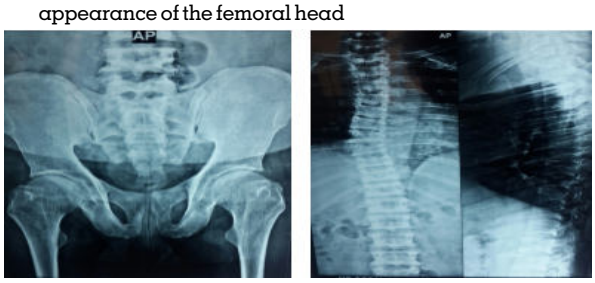
e)PNS view and lateral cephalogram showing open sutures, large fontanelles and multiple wormian bones.

- Radiograpic examination in the PA view of the skull revealed widely open fontanels and sutures . Lateral view of the skull showed enlarged skull bones along with multiple wormian bones.
- X-Ray of the chest showed asymmetrically deficient clavicles along with narrowing of the chest cage. & abnormal spine shape



f-chest radiograph showing a narrow thorax, oblique ribs and hypoplastic clavicle.

- X-ray of the spine showed normal vertebral bodies and posterior elements. Pelvic radiography showed widening of the pubic symphyseal space along with a "chef's hat" appearance of the femoral head



g) scoliosis,vertebral abnormalities , widening of the pubic symphyseal space

- After completing all the necessary investigations, the patient was confirmed as having cleidocranial dysplasia. He is currently being treated by a team comprising periodontics , oral surgeon along with psychological support. In our department of periodontics we done scaling & rootplaning & refer to department to oral surgery for further treatment . Patient is on regular follow-up & care

Discussion

CCD is an autosomal dominant condition characterized by generalized dysplasia of the bones and teeth. The more obvious features of the defect in the clavicle and cranium prompted Marie and Sainton to coin the term cleidocranial dysostosis for this condition. However, the more generalized dysplasia of bones and teeth has led to the abandonment of "dysostosis" in favour of "dysplasia". The frequency of this

disorder is one per million individuals. The CCD gene is located on either the long or short arm of chromosome 6p21

The radiological appearance of CCD is almost sufficient for diagnosis. Various features that are evident on panoramic radiographs are multiple unerupted abnormal teeth, a narrow ascending ramus, a slender and pointed coronoid process, a thin zygomatic arch with a severe downward tilt, small or absent maxillary sinuses, coarse trabeculation of the mandible, cyst formation with supernumerary teeth mainly in the anterior region, and increased density of the alveolar crestal bone over unerupted teeth . Skull radiographs show brachycephaly, a persistently openanterior fontanelle, multiple wormian bones, open skull sutures, small sphenoid bones, and calvarial thickeninge specially over the occipital and wormian bones. Chest radiography shows a narrow thorax, oblique ribs and partial part of the clavicle present.[1,5,6]

The planning of treatment for patients with CCD is complicated by a number of factors, and largely depends on both the chronological and dental ages of the patient. The timing of diagnosis is not only important for choosing an appropriate treatment plan but also for obtaining successful treatment results. A team approach to management of dental abnormalities on a long-term basis is necessary. The overall goal is to provide an esthetic facial appearance and functional occlusion . mainly consider in child patients ,in case of adults go for extraction & fabrication of prosthesis [7].

CONCLUSION

As this condition is very rare in medical & dental field one amongs millions so not many periodontitis routinely have the opportunity to treat patients with this rare genetic disorder and as a result often remain unsure of the periodontal treatment needs in such patients. The present case report is being presented to address this very gap between knowledge and the practical clinical applications. It is hoped that by looking at and treating such cases from a periodontal perspective will help the fellow periodontitis to gain their rightful place in the multidisciplinary treatment approach

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