Multiple Supernumerary Teeth Associated with Bony Malformations

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Full blown cases of cleidocranial dysplasia (CCD) have been reported earlier, but a case with a rarity of 60 teeth associated with bony malformations, is seldom observed. Because of the oral findings this condition has been diagnosed at an early age, thus helping to achieve a better oral harmony. This article reports an atypical case with 16 supernumerary teeth associated with bony malformations.

Keywords: *Supernumerary teeth, Cleidocranial dysplasia*
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INTRODUCTION

Supernumerary teeth may be defined as any teeth or tooth substance in excess of the usual configuration of 20 primary and 32 permanent teeth.[1] They may occur in single, multiple, unilateral, or bilateral and in one or both jaws.[2] Different theories suggests that, these teeth are formed because of local, independent, conditioned hyperactivity of the dental lamina or dichotomy of tooth bud.[3] The etiology of supernumerary teeth still remains unclear but hereditary and environmental factors are considered important in the occurrence of supernumerary teeth. Over 20 syndromes and developmental conditions have been found to be associated with single, and multiple supernumerary teeth such as; Cleidocranial dysplasia, Gardner's syndrome, and cleft lip and palate.[4,5]

The purpose of this report is to document a case with 16 supernumerary teeth, which led to the diagnosis of associated bony malformations and to discuss the various treatment modalities that can be rendered for achieving better oral harmony.

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Case Report

An 11-year old female (Figure 1) reported to the Department of Pedodontics and Preventive Dentistry, Government Dental College and Hospital; Hyderabad; Andhra Pradesh; India; with a chief complaint of un-erupted upper front teeth. Family and health histories were non-contributory. On general examination, stature was normal but the forehead was prominent and there was reduced mid-face development. Chest was found to be narrow and shoulders sloping, but there was no hyper-mobility of the shoulders. Intraoral examination revealed the patient in mixed dentition with class I molar relationship. The teeth present were primary canines and molars in all the four quadrants, left maxillary first premolar, three permanent mandibular incisors and all four first permanent molars. Maxillary incisors were not erupted and mandibular left first permanent molar was carious.

Orthopantomograph (OPG) and full mouth periapical

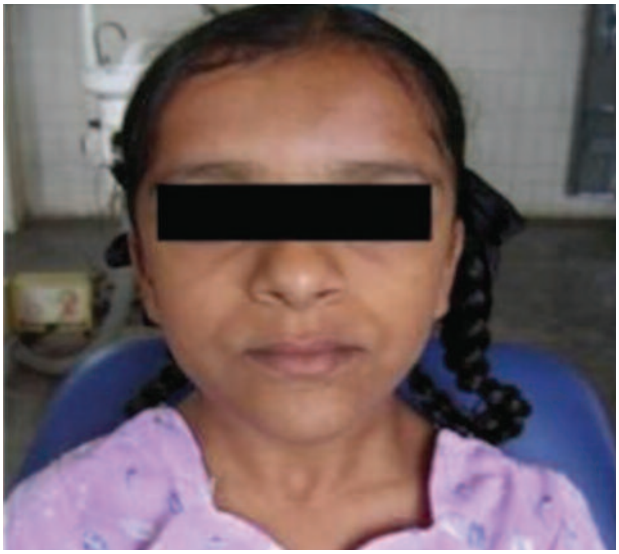

Figure 1. Anterior view of the patient.

views were taken, which revealed 16 supernumerary teeth; 10 in maxilla and 6 in mandible. There were a total of 60 teeth observed in the OPG; 16 un-erupted supernumeraries, 12 erupted primary, 8 erupted and 24 un-erupted permanent teeth. The dentition observed in panoramic view (Figure 2) was:

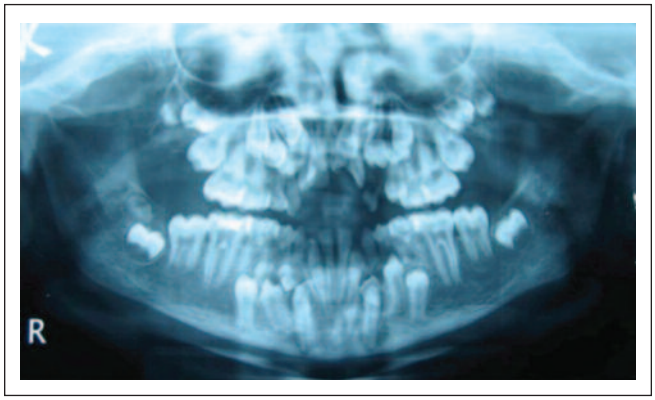

Figure 2. Orthopantomograph showing the presence of unerupted supernumerary teeth.

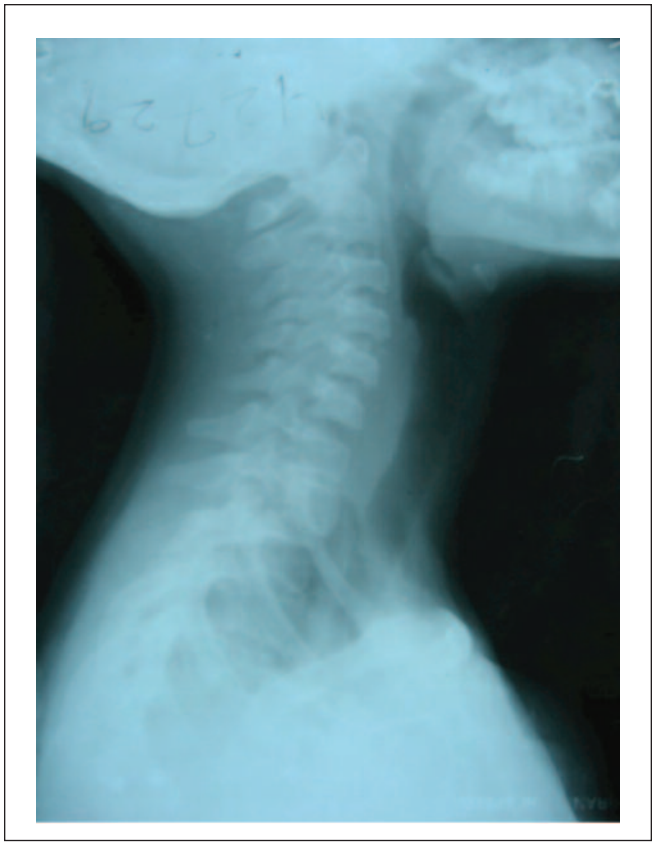

Figure 3. Radiograph of cervical spine.

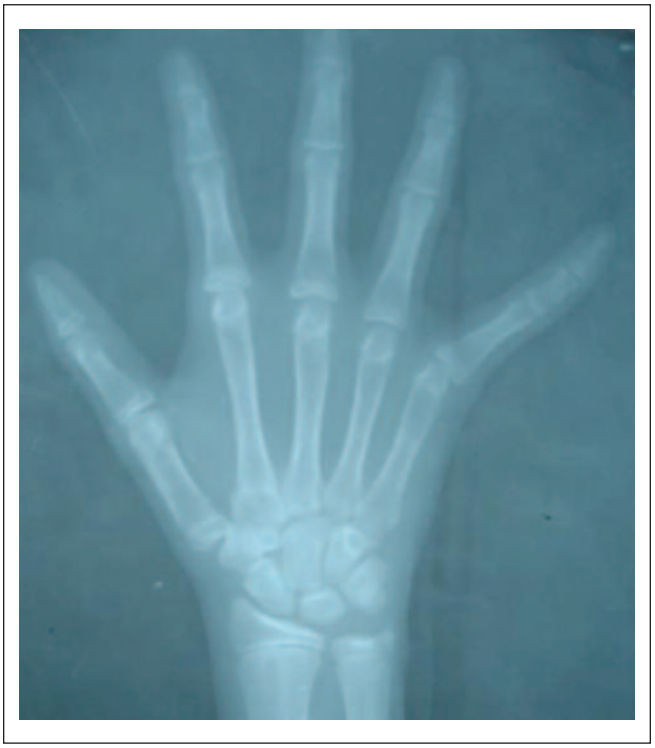

Figure 4. Hand wrist radiograph.

Serial radiographs were obtained to assess the skeletal growth and developmental pattern. Cervical spine (Figure 3), dorsolumbar spine and hand wrist (Figure 4) radiographs were noncontributory. Lateral and postero-anterior (PA) view of skull revealed mild opening of anterior fontanel. PA view of skull (Figure 5) showed delayed ossification of

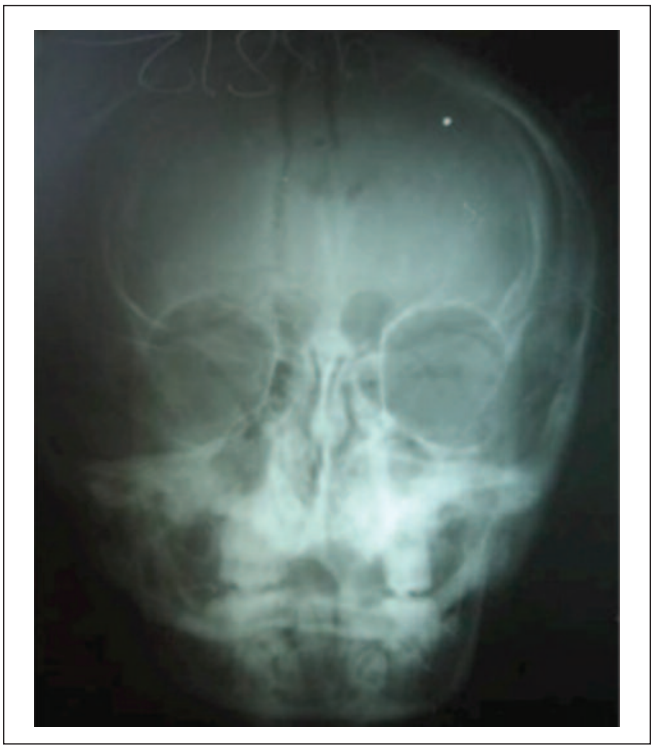

Figure 5. Postero-anterior view of the skull showing open anterior fontanel and delayed ossification of sutures.

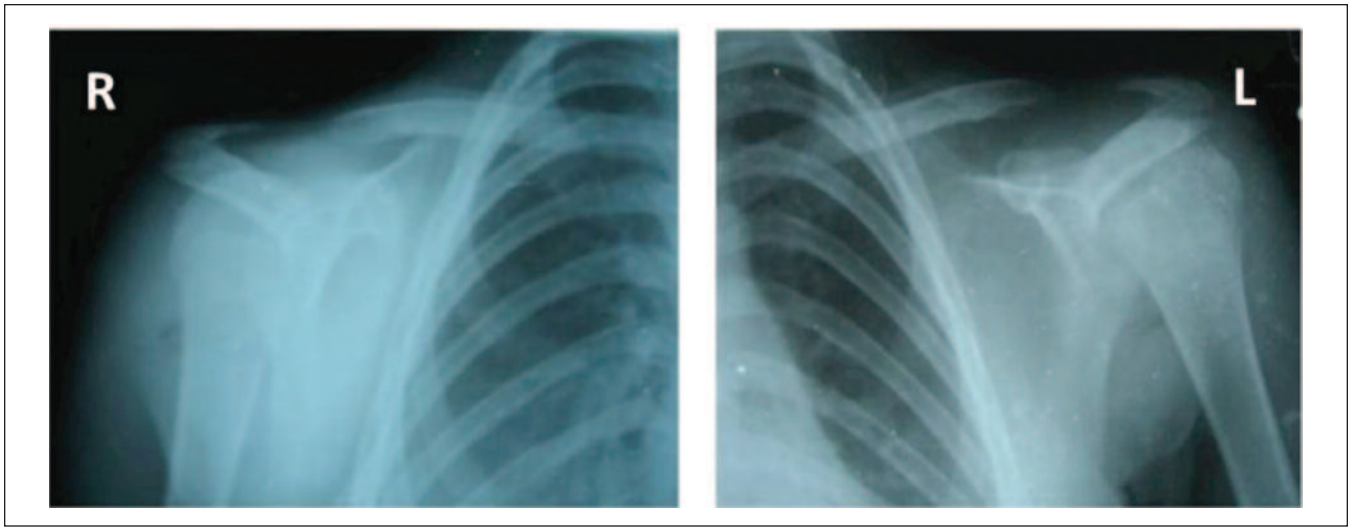

Figure 6. Right and Left clavicular views showing mild hypoplasia of right clavicle.

sutures and hypoplasia of maxillary sinuses and mild hypoplasia of both right and left clavicles in the chest radiographs (Figure 6) were evident. Hematological investigations revealed elevated level of alkaline phosphatase (*202U/L*).

DISCUSSION

Cleidocranial dysplasia (CCD), also known as *Marie* and *Sainton* disease, mutational dysostosis, craniocleidodysostosis and cleidocranial dysostosis, is a rare developmental defect of autosomal dominant inheritance.[6] This has been mapped to a microdeletion of chromosome band 6p21,[7] t(6;18) (p12;q24) translocation[8] and pericentric inversion of chromosome 6.[9] It is also thought to be caused by heterozygous mutations in *runt*-related gene 2 (*RUNX2)*/*polyoma virus enhancer binding protein 2*a*A (PEBP2*a*A)*/*core-binding factor A1 (CBFA1),* the pathogenesis being the impaired Smad signaling of transforming growth factor by bone morphogenetic protein pathways that target the activity of RUNX2 during bone formation, thus affecting all bones and cartilages.[10,11] It presents with skeletal defects of several bones, the most striking of which is partial or complete absence of clavicles and late closure of the fontanel resulting in frontal bossing.[12] Additional skeletal defects are spina bifida, failure of union of neural arches to the body of the vertebrae, union failure of the pubic bones, with patent sacro-iliac joints and wide spaces between the ossified ribs and vertebrae. Reported vertebral abnormalities are scoliosis, kyphosis, lordosis, and vertebral synostosis. Most individuals show delayed and deficient ossification of long bones, which accounts for a somatotypically short and broad body build.[13,14,15] The defects in the present case are confined to cranium and clavicle; incomplete closure of sutures and fontanel and hypoplasia of maxillary sinuses giving characteristic facial appearance.

The most presumptive clinical finding of CCD, i.e. hypermobility of the shoulders is not present in this case, as it depends on the amount of clavicular involvement.

It has also been reported that the serum alkaline phosphatase (ALP) levels [10,11,16] will be reduced in cases of CCD, but in the present case it is elevated.

Dental abnormalities of CCD include retained primary dentition, delayed eruption or retention of the permanent dentition, multiple supernumerary teeth, crown and root abnormalities, and a high palate. Other features include complete absence of cellular cementum, an increase in the amount of acellular cementum of the roots of the affected teeth, retention of the primary teeth and failure of eruption of the permanent successors.[17,18] In the present case multiple supernumerary teeth is the only feature. On surgically exposing the permanent maxillary central incisors, there was progressive eruption after one week follow up.

In the present case, defects in the clavicles, open anterior fontanel and delayed ossification of sutures and presence of 16 supernumerary teeth are in support of the diagnosis CCD. But elevated serum alkaline phosphatase levels and no other dental abnormality other than supernumerary teeth are opposing CCD. As parents were not willing for genetic tests, the diagnosis could not be confirmed.

Supernumerary teeth can cause disturbances in the developing dentition such as crowding, delayed eruption, diastema, rotations, and resorption of the adjacent teeth.[19] They can also result in cystic degeneration,[20,21] odontoma[22,23] or ameloblastoma[24] formation. Most problems associated with supernumeraries are because of their potential to interfere with normal occlusal development. In the present case supernumeraries caused delayed eruption/impaction of permanent teeth giving a clinical appearance of hypodontia.

There are different options of management of these conditions. The option of no treatment was common in the past,[25] according to which, observation and follow-up radiographs was the only management advised, if the un-erupted teeth are asymptomatic without any evidence of cyst formation, as the attempts at surgical removal might result in damage to vital anatomical structures such as inferior dental nerve or mental nerve. Periodic radiological assessment

should be done to enable early detection of any cystic degeneration associated with the un-erupted teeth.

Symptomatic multiple un-erupted supernumerary teeth should be surgically removed after appropriate education of the patient and parent about the risk of damage to adjacent teeth and vital anatomical structures.

Extractions followed by provision of dentures have also been suggested.[26] Some regard this approach as too invasive, especially considering the extensive bone loss experienced after removal.

Distraction osteogenesis with or without orthodontic treatment might play a role in the definitive treatment of multiple un-erupted supernumerary teeth.[27] The current "state-of-the-art" treatment involves a combination of orthodontics and maxillofacial surgery.

CONCLUSION

Early diagnosis of conditions such as this is difficult, as majority of the craniofacial abnormalities become obvious only during adolescence. In the present case mild skeletal defects confined to the clavicle and cranium have been diagnosed based on the dental findings. There is an elevated level of serum alkaline phosphatase, which is uncommon in case of CCD. Though diagnosis could not be confirmed with genetic tests, it can be an atypical case of CCD with incomplete genetic expression.

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