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Case Report

Multiple unerupted and supernumerary teeth in a patient with cleidocranial dysplasia

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ABSTRACT

Cleidocranial dysplasia is an extremely rare familial disorder characterized by partial or complete absence of clavicles, characteristic craniofacial deformities, and the presence of numerous supernumerary and unerupted teeth. Here, the author reviews the striking radiographic findings of cleidocranial dysplasia in a 16-year-old adolescent boy who presented with delayed teeth eruption.

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Case presentation

A 16-year-old Caucasian adolescent boy presented to the dental clinic for consideration of orthodontic braces. Extraoral examination revealed a brachycephalic head, with pronounced frontal and parietal bossing, hypertelorism, depressed nasal bridge, and hypoplastic maxilla. On intraoral examination, there were multiple over-retained primary teeth and multiple missing permanent teeth. A panoramic radiograph showed multiple unerupted permanent and supernumerary teeth (Fig. 1). A lateral cephalogram showed calvarial thickening, with numerous wormian bones in the line of the lambdoid suture (Fig. 2). However, a previously taken posterior-anterior spine radiograph showed hypoplasia of the clavicles, absence of the 12th ribs, with thoracolumbar scoliosis (Fig. 3). The family history

identified other members with similar dental and skeletal conditions. On the basis of the clinical and radiographic findings, a diagnosis of cleidocranial dysplasia was made.

Discussion

Cleidocranial dysplasia is a rare autosomal dominant disorder affecting bones and teeth. Cleidocranial dysplasia is caused by a mutation in the *Runx2* gene involved in the differentiation of osteoblasts [1]. The prevalence of cleidocranial dysplasia is estimated at 1:1 million without sex or ethnic group predilection [2].

Affected individuals are usually short. The skull is brachycephalic, with pronounced frontal and parietal bossings, and

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Fig. 1 – Panoramic image shows multiple retained primary teeth and multiple unerupted permanent and supernumerary teeth. Note the dense alveolar bone overlying the unerupted teeth.

the maxilla and zygomas are underdeveloped. Thus, the face appears small in contrast to the cranium. The bridge of the nose is broad and depressed, with ocular hypertelorism.

There is delayed or failed closure of the anterior fontanel, open cranial sutures including a persistent metopic suture, and multiple wormian bones (small, irregular bones formed by secondary centers of ossification within the suture lines). The skull usually shows a segmental pattern of calvarial thickening in the supraorbital ridge of the frontal bone, the squamous part of the temporal bones, and the occipital bone above the inion. The paranasal sinuses and mastoids are often underdeveloped or absent [3].

There is aplasia or hypoplasia of 1 or both clavicles, frequently allowing the individual to approximate the shoulders in front of the chest. Deficiency of the clavicle is responsible for the long appearance of the neck and the narrow sloping shoulders.

The maxilla is characteristically hypoplastic, resulting in maxillary micrognathia. The growth of the mandible is usually normal. However, there may be delayed union of the mandibular symphysis [3].



Fig. 2 – Lateral skull image shows calvarial thickening with extensive wormian bone formation in the line of the lambdoid suture. Note the obtuse gonial angle of the mandible.



Fig. 3 – Posterior-anterior spine radiograph taken at age 15 shows defective clavicles with thoracolumbar scoliosis.

Characteristically, there is prolonged retention of the primary teeth, and multiple unerupted permanent and supernumerary teeth. The supernumerary teeth are most often in the mandibular premolar and maxillary incisor areas [4].

Although cleidocranial dysplasia primarily affects bones of membranous origin, other bones also may be affected, including the femur, pelvis, vertebral column, and bones of the hands and feet [3]. Mental development is usually normal.

Cleidocranial dysplasia may be diagnosed by the characteristic clinical and radiographic findings [4]. However, multiple unerupted supernumerary teeth have been reported in a number of conditions, including Gardner syndrome (familial polyposis coli and osteomata) and pyknodysostosis. Complete absence of both clavicles is seen in Yunis-Varon syndrome (skeletal defects, digital anomalies, and intellectual dysfunction).

The treatment plan comprised orthodontic treatment for the malocclusion with extraction of the retained and supernumerary teeth, and surgical exposure and orthodontic traction of the impacted permanent teeth. However, dental management in cleidocranial dysplasia primarily depends on the

patient's needs and age. The best results are obtained if the condition is diagnosed and treated at an early age [5]. Radiologists should be familiar with the constellation of findings associated with cleidocranial dysplasia so as to assist clinicians in the diagnosis of this disorder.

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