Cherubism: Presentation of a case

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Cherubism is a rare, inherited fibro-osseous bone disease that affects the jaws. The first case study of this condition was published in 1933 by Jones,[1,2] who coined the term *cherubism* to reflect the characteristic facial appearance of affected individuals. Bilateral enlargement of the mandible produces a full, round lower face. The skin on the cheeks stretches, pulling the lower eyelids down and exposing a thin line of sclera, resulting in eyes that appear raised to heaven, similar to those of cherubs portrayed in renaissance religious paintings.

Cherubism is caused by an autosomal dominant gene and is usually manifest in males; this was confirmed by Anderson et al.[3]

A case of cherubism was reported by Gosh et al.[4] at the College of Dental Surgery, Manipal, India, in 1984, and an another case was reported by Lakhar et al.[5] in 1990 at the Kasturba Medical College, Manipal. In the latter case, the patient had three siblings; two showed no signs of Cherubism while the youngest brother was affected.

Case presentation

A male, 20 years old, reported to the orthodontic department of the College of Dental Surgery, Manipal, in July 1996, complaining of irregular teeth. A marked facial distortion was noted during the examination. The patient reported that the condition had been present for a long time and was getting worse. There was no familial history of the condition.

Extra oral examination showed bilateral swelling of the mandible, upturned eyes (Figure 1), and facial asymmetry. The facial profile was convex. Intraorally, the soft tissues were normal; teeth #18, 28, 48, 45, 44, 38 35, 34 (ISO/FDI) were missing and spaces were present in the lower arch.

Maximum incisal opening was 39 mm. The molar relationship on the right side was in crossbite, and the left side was Class II (Figure 2).

Abstract

Cherubism is a rare, fibro-osseous bone disease that affects the jaws. Bilateral enlargement of the mandible produces a full, round lower face. The skin over the cheeks stretches and pulls the lower eyelids down, exposing a thin line of sclera and eyes that are raised, seemingly heavenward. The patient in this report was diagnosed but not treated.

Key Words
Cherubism

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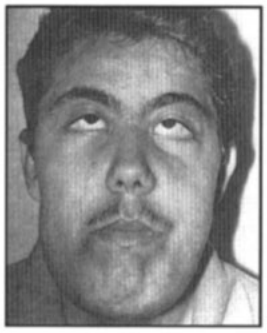

Figure 1

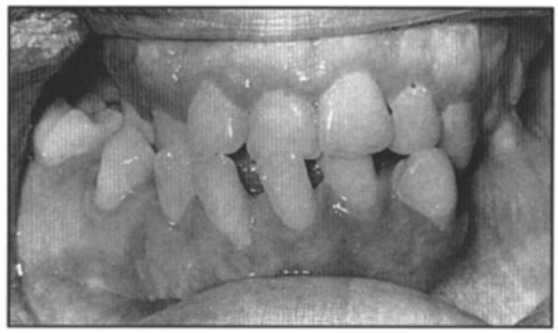

Figure 2

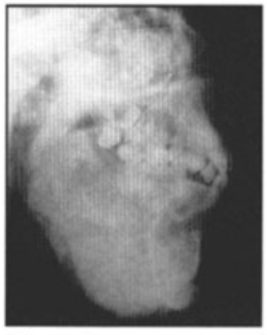

Figure 3

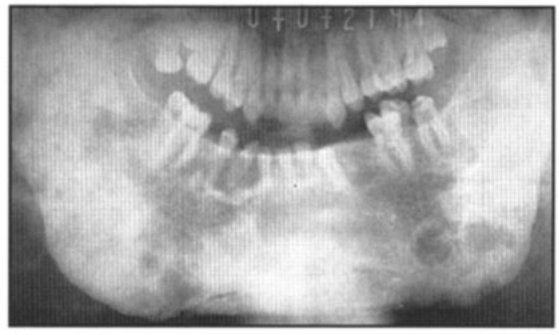

Figure 4

Figure 1
Classic appearance of cherubism

Figure 2
Irregular dentition

Figure 3
Lateral cephalogram with enlarged mandible and cystic lesions

Figure 4
Panoramic radiograph with multilocular radiolucencies and spacing of teeth

Radiographs

Multilocular radiolucencies involving the ramus and posterior part of the body of the mandible were visible on the right lateral cephalogram. The vertical depth of the mandible was larger than normal.

The lateral cephalogram showed that the mandible was enlarged with cystic lesions (Figure 3). The panoramic radiograph (Figure 4) revealed bilateral multilocular radiolucencies involving the mandible with spacing of teeth in lower arch. Teeth #48, 45, 44, 34, 35, and 38 were missing.

A diagnosis of cherubism was made. Because the patient was a visitor in our area and planned to leave soon, we were not able to provide any treatment. He had undergone surgery in the U.K. and was referred for orthodontic treatment there, but treatment had not been started.

General view of the disease

There has been a controversy whether this condition is an anomaly of dental development or whether the dental abnormalities are secondary to changes in the bone. According to the report by Goaz and White,[6] Cherubism is regarded as a bony dysplasia of uncertain nature.

The lesions develop during early childhood, between the ages of 2 and 6 years; bilateral swelling of the lower face continues gradually until about the age of 20. Radiographically, cherubism is characterized by multicular radiolucencies, which are usually well defined. There is an expansion of the buccal and lingual cortical plates. The destructive central jaw lesions have an understandably profound effect on the developing dentition, resulting in displacement of tooth buds.

Reports have so far been unavailable on the orthodontic management of cherubism. Goaz and White[6] are of the opinion that cherubism should not be treated surgically because it will regress on its own. Morley et al.[7] restated that orthodontic management is challenging, but no reports on the technical aspects are available.

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