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Case Report

The multidisciplinary management of the cherubism patient for function and aesthetics

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ABSTRACT

Cherubism is a nonmalignant, multicystic, fibro-osseous disease that is characterized by painless gradual enlargement of the jaws. Typical dental anomalies may manifest, such as displaced and missing teeth, which are associated with complex malocclusions that present a challenge for correction. This case report provides a description of the orthodontic management of an adolescent patient with cherubism. Adequate improvement of teeth malposition was achieved through the use of removable and fixed appliances; although, treatment duration time was lengthy due to delayed tooth eruption and level of difficulty. It is imperative to understand that dental practitioners may face an intricate task when a patient presents with this condition, but proper multidisciplinary treatment planning and execution can provide satisfactory final results.

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1. Introduction

Cherubism is a rare, inherited developmental fibro-osseous abnormality that affects the jaws by enlarging the maxilla and/or mandible [1,2]. First described by Jones in 1933, it is a benign disease that manifests itself usually at 2 to 3 years of age; although, the moment that it is recognized could vary depending on the acceleration of the disorder [3,4]. It is characterized by painless gradual swelling of the cheeks due to symmetrical skeletal enlargement, with frequently associated dental abnormalities; and usually, progresses until puberty, after which it shows partial or complete spontaneous involution.

The etiology was initially thought to be familial, but many sporadic cases have been reported. Also, the condition was considered a form of a localized fibrous dysplasia; nonetheless, recent studies have revealed that it is a genetically autosomal dominant gene at chromosome 4p16.3 with mutations in the gene *SH3BP2*. Males are affected twice as often as females, with penetrance reported to be 100% in men and 50% to 70% in women [5–11].

The diagnosis of cherubism is based on clinical, radiographic, and histological findings. The most common clinical manifestation is firm painless bilateral enlargement of the lower face. Swelling of the submandibular lymph nodes may occur, but no systemic abnormalities are involved. The visual appearance varies from a discernible posterior swelling of the mandible to a marked anterior–posterior expansion that may result in masticatory, speech, and swallowing difficulties. Maxillary expansion may produce the characteristic "*eyes raised to heaven*" look similar to the cherubic angels portrayed in renaissance religious paintings—a thin band of sclera is exposed between the iris and the lower eyelid. This disease is predominantly observed in the mandible; maxillary occurrence is variable, but always accompanied by mandibular involvement. Radiographic lesions appear as multiple, well-defined, multilocular, radiolucent areas in the maxilla and/or mandible. They begin in the posterior alveolar region and/or ramus and can spread anteriorly. The mandibular angle, ascending ramus, retromolar region, and posterior maxilla are mostly affected. The coronoid process can be involved, but the condyles are always spared [12–16].

These lesions are irregular in size and usually cause marked profound destruction of alveolar bone that affects tooth buds and incipient follicles. It is typical for displaced and erupted teeth to

This study was conducted while Dr. Picon was a resident at Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, New York.

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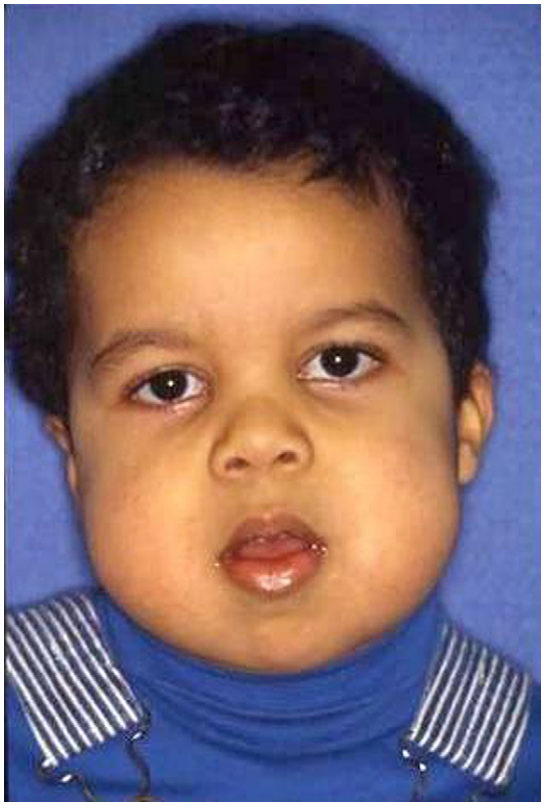

Fig. 1. Presentation at initial visit (3 years old).

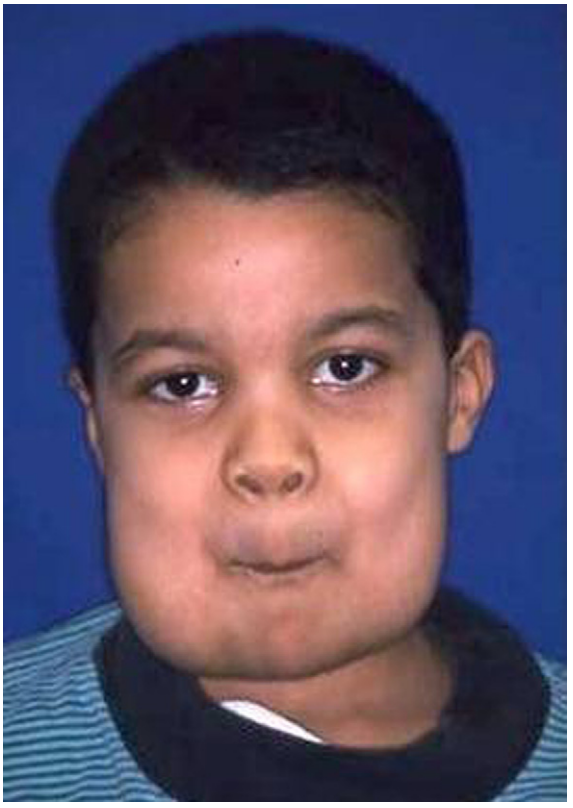

Fig. 2. A severe expansive area at the right angle of the mandible (5 years old).

appear to be floating in radiolucent spaces. Depending on the extension and involvement of the lesions observed in the radiographic images, a classification grading system ranging from 1 to 5, is used to define severity of the condition. Histologically, the lesion is composed of mature fibroblasts in a surprisingly pale edematous background that contains little collagen. Multinucleated giant cells are few and occur in clusters. A distinctive feature is the presence of an acellular, *eosinophilic cuff,* deposit that appears to ring small vascular channels. This feature, although considered pathognomonic, is not a sensitive marker because it is absent in many patients [17,18].

Active treatment of the disease is generally not necessary, and is largely dependent on the degree of bone involvement and the rate of progression. Reported treatments have varied considerably and can include extraction of teeth in the areas involved, surgical contouring of expanded lesions, and complete surgical curettage. Surgical resection may be performed for cosmetic reasons or if function can be improved, but treatment can be delayed because the cystic lesion usually becomes static and fills in with granular bone at the end of the skeletal maturational growth period. Long-term clinical studies have revealed that childhood lesions proceed to partial or complete resolution in adulthood. Finally, radiation therapy is totally contraindicated due to possible retardation of jaw growth and the increased incidence of osteoradionecrosis or malignancy [19–22].

2. Diagnosis and etiology

The patient was a 2-year-old Hispanic male, with a history of cherubism involving both maxilla and mandible, who was referred for evaluation and treatment to the Orthodontic Division. Past medical history indicated a full-term birth delivery without complications. No previous history of cherubism and/or any type of bony dysplasia were reported to have occurred in any family member. Adequate growth and development was observed until he was 18 months old, which was when his mother perceived abnormal bulging of the face and oral gingival tissue.

The patient was eventually referred to the Oral and Maxillofacial Surgery Division at the Albert Einstein College of Medicine for initial consultation at 3 years old (Fig. 1). Upon exam and evaluation, the attending physician determined that no active treatment was to be rendered at the time indicating that, “cherubism frequently burns itself out and regresses during puberty.” The recommendation was to follow up patient with an annual exam.

Bilateral enlargement of both maxilla and mandible, the latter being more pronounced, was observed throughout time. Laboratory blood workup revealed no changes in calcium, parathyroid hormone, and/or phosphatase levels which ruled out the possibility of hyperparathyroidism. The patient was finally diagnosed with cherubism at age 5 and his mother requested a surgical procedure to reduce size of the mandible because the patient had “become more self-conscious of his facial deformity” (Fig. 2).

The panoramic radiograph, taken at 6 years old revealed multilocular bilateral lesions primarily of the mandible with displacement of teeth out of their normal position. Intraoral surgical curettage and debulking were indicated and executed at the mandibular right side without any surgical complications, and a surgical dressing pack of a steroid-anesthetic-vasoconstrictor solution was placed over the cortices around the entire lesion. The extraction of the impacted right permanent mandibular canine was also performed. Surgical intervention was postponed in the left mandibular area due to the moderate amount of blood loss from the contralateral side but the steroid-anesthetic-vasoconstrictor solution was injected inside the cystic cavities on the left side (Figs. 3A and 3B). Biopsy of the tissue

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