Cherubism in 12 Year Young Female

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Abstract

Cherubism is a rare benign, autosomal-dominant inherited fibro-osseous lesion of jaw characterized by excessive bone degradation of the upper and lower jaws followed by the development of fibrous tissue masses. It is usually self-limiting; it starts in early childhood and involutes by puberty. The purpose of this clinical report is to describe a nonfamilial case of cherubism on a teenager female patient first treated by calcitonin nasal spray followed by surgical resection and recontouring after puberty.

Keywords: Benign fibro-osseous lesion, calcitonin, cherubism, self-limiting pathology

Introduction

Cherubism is a autosomal dominant inherited fibro-osseous lesion mainly causing painless bilateral expansion of the mandible, maxilla, or both.[1] Jones first described it as a benign, self-limiting bone disorder which is usually found among children of 5–10 years of age.[2] The word "cherub" refers to the spherical facial appearance of angels painted in the Renaissance era due to typical moon facies of patients having marked fullness of the jaws and cheeks with upward gazing of the eyes.[3] On the basis of the extent of involvement, Ramon and Engelberg graded such lesions as follows: Grade 1 (involvement of both mandibular ascending rami; Grade 2 (same as Grade 1 plus involvement of both maxillary tuberosities); Grade 3 (massive involvement of whole maxilla and mandible, except the condylar processes); and Grade 4 (same as Grade 3 with involvement of the floor of the orbits causing orbital compression).[4] Various multiquadrant osteolytic bone lesions which are included in differential diagnosis are brown tumor of hyperparathyroidism, giant cell lesions, Noonan/multiple giant cell lesion syndrome, fibrous dysplasia, aneurysmal bone cyst, and the hyperparathyroidism-jaw tumor syndrome. Here, in this article, we are presenting a case report of a 12-year-old female patient with swelling on both sides of the face for 5 years of age (Grade 3). It has been treated firstly by calcitonin nasal spray for 1 year followed by surgical resection and recontouring after puberty.

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Case Report

A 12-year-old female reported to the Department of Oral and Maxillofacial Surgery complaining of painless swelling over both sides of the face. History revealed that it had started as a small swelling at 5 years of age and progressively increased to the present size. None of her family members reported such type of swelling. On extraoral clinical examination, we noticed roughly oval shape, painless swelling involving all four quadrants of the jaws, that was hard in consistency with poorly defined margins [Figure 1]. Swelling on the left side was slightly larger than the right side of the face. On palpation, the temperature of the overlying skin was normal and no tenderness was elicited on palpation. The swelling extended below the lower border of the mandible. Intraorally, there was a marked expansion of buccal cortex with a shallow palate. There were a few remaining teeth that were rotated and malposed while the rest of the teeth had fallen out [Figure 2]. An orthopantomograph showed multilocular appearance of the maxilla and mandible (excluding condyle) with numerous unerupted and displaced teeth appears to be floating in

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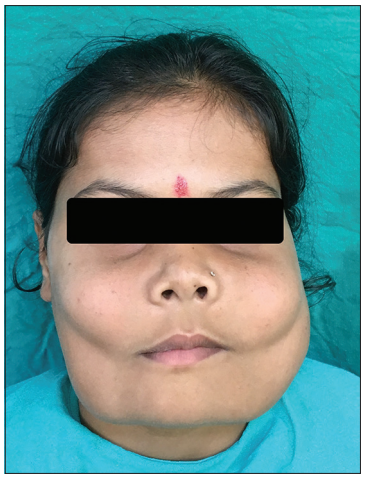

Figure 1: Extraoral preoperative frontal view

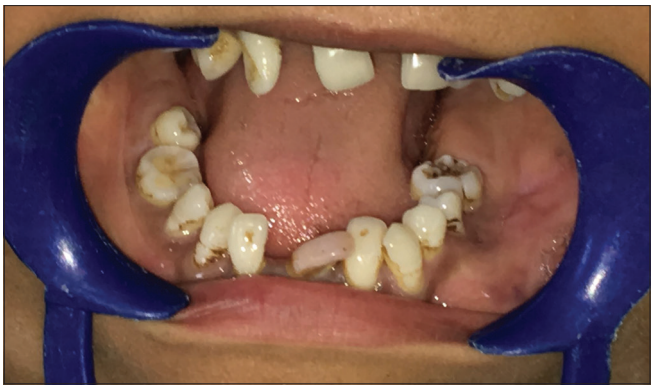

Figure 2: Intraoral preoperative view

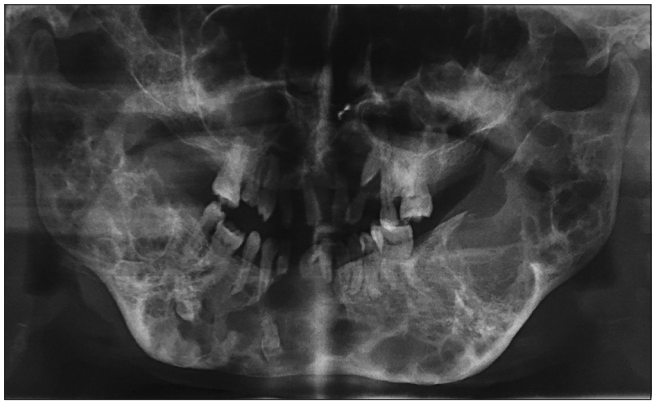

Figure 3: Preoperative orthopantomograph view

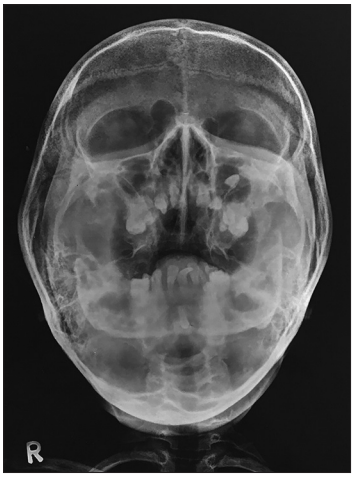

Figure 4: Paranasal sinus view

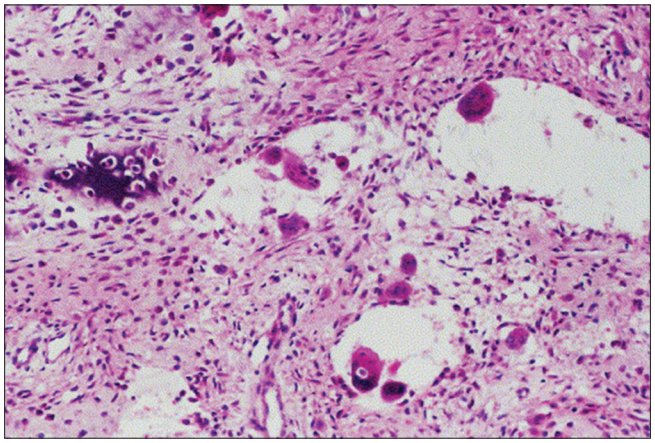

Figure 5: Histopathological view

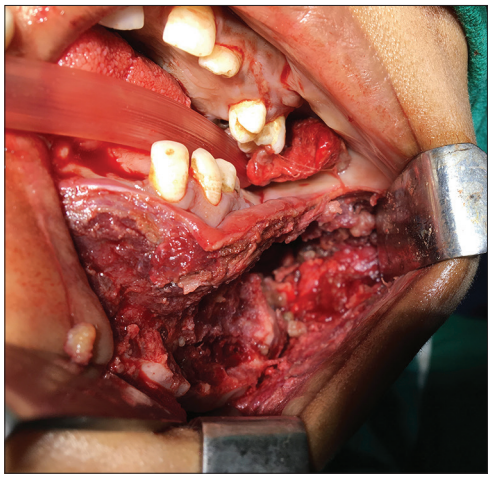

Figure 6: Intraoperative view- left side of mandible

numerous cyst-like spaces [Figures 3 and 4]. On the basis of clinical and radiographic findings, a provisional diagnosis of cherubism of jaw was given. The laboratory values of serum calcium, phosphorus, and alkaline phosphatase were found to be within the normal range. Incisional biopsy was taken from one site in the left mandible which on histopathological examination revealed a highly cellular mature fibrous connective tissue with numerous endothelial cell proliferations along with multinucleated giant cells. There was evidence of prevascular eosinophilic cuffing of collagen around small capillaries [Figure 5]. Thus, the final diagnosis of cherubism was established. Surgical resection and recontouring of lesion was planned after puberty [Figures 6-8]. Until then, medicinal therapy was prescribed in the form of salmon calcitonin nasal spray (Calcispray,

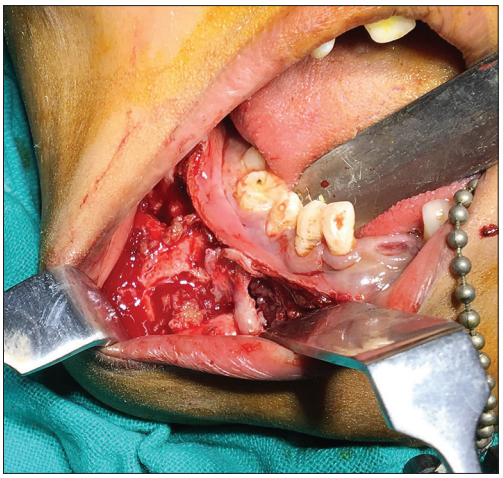

Figure 7: Intraoperative view- right side of mandible

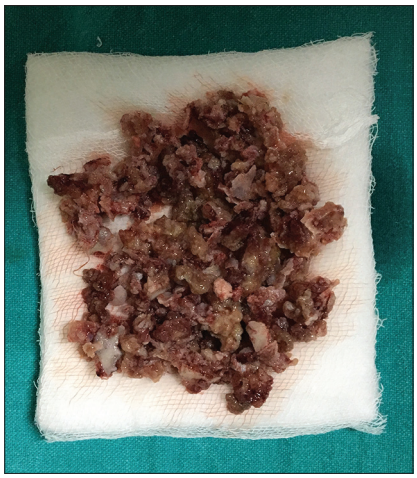

Figure 8: Biopsy specimen

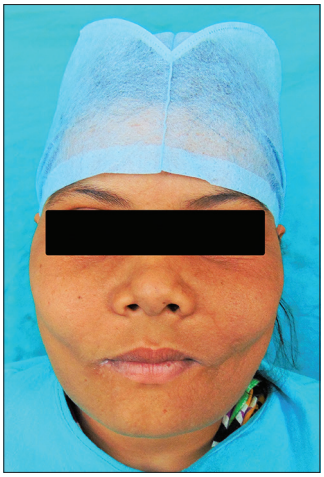

Figure 9: Postoperative extraoral frontal view

Zydus Cadila), alternating nostrils daily, in the dosage of 200 UI. After the surgical procedure, the patient submitted to radiographic examinations annually during 4 years' recall evaluation [Figure 9].

Discussion

According to the WHO histological typing, cherubism belongs to a group of nonneoplastic bone lesions that affect mainly the jaws.[5] Anderson and McCledon reviewed 65 cases from 21 families and suggested autosomal dominant pattern of inheritance for such lesions.[6] In 2016, Ueki. described the pathogenesis of cherubism as heterozygous mutations in 12 families in the gene for the signaling adapter SH3-domain binding protein 2 (SH3BP2) on chromosome 4p16.3. SH3BP2 is a major regulator of bone resorption, and its mutation leads to increased bone resorption of jaws.[7] Sporadic nonfamilial cases have also been reported like our case report whose pathogenesis suggested was of *de novo* type. Clinical manifestation varies from a barely perceptible posterior swelling of a single jaw usually observed at about 2 years of age followed by marked anterior and posterior expansion of both the jaws on both sides of the face from 8 to 10 years and spontaneous cessation of growth after puberty.[8] Rapidly growing lesions requires early surgical interventions to avoid optic nerve compression, respiratory incompetency, mastication, swallowing, and phonation.[1,3]

However, surgery should be avoided during the growth phase as it causes stimulation of fibroblasts, osteoclasts which results in a relapse of the lesion quickly and at a greater rate.[4,6] Hyckel *et al*. advocate radical surgery when the lesion occurs in all four quadrants. Thus, depending on the extent and rate of growth, mainly surgical and medicinal therapy is instituted. Regarding surgical resection, the strategy of wait and watch is the most commonly used until regression of lesion. Before puberty, medical treatment by systemic calcitonin, tacrolimus had reported success due to inhibition of bone resorption.[9,10]

Conclusion

Cherubism although rare has a significant impact on affected children and their families by causing facial deformity and functional problems. As most of the cases are self-limiting, so, surgical treatment should be reserved for aggressive growing lesions and esthetic desire of the patient.

Right to privacy and informed consent

The authors declare that no patient data appear in this article and written informed consent was taken from the patient.

Declaration of patient consent

The authors certify that consent had been taken from patient for her images and other clinical information reported in this article. The patient understood that identity and name was concealed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Kozakiewicz M, Perczynska-Partyka W, Kobos J. Cherubism – Clinical picture and treatment. Oral Dis 2001;7:123-30.
2. Carvalho Silva E, Carvalho Silva GC, Vieira TC. Cherubism: Clinicoradiographic features, treatment, and long-term follow-up of 8 cases. J Oral Maxillofac Surg 2007;65:517-22.
3. Atalar MH, Albayrak E, Erdinc P, Bulut S. Cherubism as a rare cause of bilateral expansion of the mandible: Radiological manifestations. J Hong Kong Coll Radiol 2008;11:76-80.
4. Ramon Y, Engelberg IS. An unusually extensive case of cherubism. J Oral Maxillofac Surg 1986;44:325-8.
5. Pindborg JJ, Kramer IR. In: WHO Histological Typing of Odontogenic Tumours, Jaw Cysts and Allied Lesions. 1st ed. Geneva: Springer-Verlag; 1971. p. 18-9.
6. Anderson DE, McCledon JL. Cherubism-hereditary fibrous dysplasia of the jaws, Part-I. genetic considerations. Oral Surg Oral Med Oral Pathol 1962;15:5-15.
7. Ueki Y. Molecular and cellular pathogenesis of cherubism. Clin Calcium 2016;26:918-26.
8. Beaman FD, Bancroft LW, Peterson JJ, Kransdorf MJ, Murphey MD, Menke DM, *et al.* Imaging characteristics of cherubism. AJR Am J Roentgenol 2004;182:1051-4.
9. Kadlub N, Vazquez MP, Galmiche L, L'Herminé AC, Dainese L, Ulinski T, *et al.* The calcineurin inhibitor tacrolimus as a new therapy in severe cherubism. J Bone Miner Res 2015;30:878-85.
10. Fernandes Gomes M, Ferraz de Brito Penna Forte L, Hiraoka CM, Augusto Claro F, Costa Armond M. Clinical and surgical management of an aggressive cherubism treated with autogenous bone graft and calcitonin. ISRN Dent 2011;2011:340960.