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Cherubism: Report of a case

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Abstract
Cherubism is a dominantly inherited fibro-osseous lesion that occurs in the jaw bones. The clinical appearance of the patients varies greatly ranging from very few or no clinically recognizable changes to marked facial disfigurement. The mandible is primarily involved with the lesion starting at the angle of the mandible and the entire bone may be involved except for the condylar processes. The maxillary involvement is variable, less severe in most cases, and always associated with mandibular involvement. In this report, we describe a pediatric case of cherubism involving both jaws.

Keywords: Cherubism, mandible, maxilla, pediatric patient

I. Introduction

Cherubism is a rare benign fibro-osseous, hereditary bone disease that primarily affects the jawbones. The condition was first reported by Jones in 1933 who described it as the multilocular hereditary cystic disease of the jaw. [1] This Canadian radiologist used the term 'cherubism' because the symmetrical swelling of the cheeks and upturned eyes of the affected individuals resemble the angels painted during the Renaissance era. [2,3] The disease usually appears in childhood between two to five years of age as a painless and often bilateral swelling of the mandible or/ and maxilla. There is an increased growth from eight to nine years of age followed by stabilization and spontaneous regression of the lesion in most cases when the individual reaches early adulthood. The clinical appearance of the patients varies greatly ranging from very few or no clinically recognizable changes to marked facial disfigurement. Cherubism is now recognized as an autosomal dominant disorder (MIM 118400) with 100% penetrance in males and 50-70% penetrance in females. [4] The condition is associated with mutations in the SH3BP2 gene on chromosome 4p16.3.[5] Some investigators have reported that cherubism has been associated with Ramon syndrome, Noonan syndrome, neurofibromatosis, and Fragile X syndrome. [6-8] To date, nearly 300 cases of cherubism were reported in the literature and most of them were familial cases. In this article, we report a pediatric case of cherubism.

II. Case Report

A 4-year-old girl reported to the Department of Pediatric and Preventive Dentistry of our institution for non-erupting lower front teeth. She was the only child of her parents with no significant medical or family history. The girl presented with painless bilateral bony hard swelling of the lower jaw since two years. The cervical and submandibular lymph nodes were found nonpalpable. An intraoral examination of the patient revealed a primary dentition with dental anomaly. Clinically, the maxillary left lateral incisors, mandibular central incisors, and lateral incisors were absent. [Figure 1] There was no history of trauma, dental extraction, or pus discharge from the jaw. A cone-beam computed tomographic (CBCT) examination revealed a well-defined, expansile, multilocular radiolucent lesion on both sides of the mandible. Osteolytic lesions were seen in the coronoid processes, angle of the mandible, rami, and the body of the mandible. However, the condyles were not affected. A three-dimensional image of the CBCT displayed similar osteolytic lesions present in the tuberosity region and the lateral wall of the maxillae. [Figure 2] Radiographic examination also revealed multiple missing permanent tooth buds. The patient was referred to the Oral and Maxillofacial Surgery Department of the same institute for an incisional biopsy of the lesion. Sections stained with hematoxylin-eosin exhibited multiple multinucleated giant cells in the loosely arranged mesenchymal connective tissue stroma. Large number of fibroblasts and many small blood vessels were also seen.

[Figure 3] Biochemical investigation of the patient revealed that calcium, alkaline phosphatase, and parathyroid (PTH) were within normal limits. The clinical, radiographic, and histopathologic features and the results of the biochemical investigation of the patient are suggestive of cherubism. The condition was

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explained to the parents and follow-up visits were advised.

III. Discussion

Cherubism is a fibro-osseous lesion that exclusively affects the jawbones. The mandible is primarily involved with the lesion starting at the angle of the mandible and the entire bone may be involved except for the condylar processes.[3-6] However, some reports have described the involvement of the condyles.[2,9] The maxillary involvement is variable, less severe in most cases, and always associated with mandibular involvement [2] In this report, the mandible, as well as the maxilla, are bilaterally involved. Various dental anomalies observed in patients with cherubism include congenital absence of teeth, impaction, root resorption, and displacement of teeth.[2-6] In the present case, the impaction of primary incisors was noted.

Although radiographic features are not characteristic, bilateral multilocular radiolucent areas in the mandible and/ or maxillae are common. [4,6,7] The patient in this report showed multiple radiolucent areas in the upper as well as the lower jaw. Computed tomography (CT) is preferred over conventional radiography to determine the extent of a lesion. Furthermore, three-dimensional reconstructions and multiplanar views provide a more accurate image of the disease. Both radiographs and CT scans display multilocular radiolucency, expansive remodeling of the jaws, coarse trabeculae, thinning of cortical bones, and absence of periosteal reaction. [2]

Ramon and Engelberg introduced a grading system for cherubism according to the extension of the lesion as follows:[10]
Grade 1: Involvement of ascending ramus of mandible bilaterally
Grade 2: Involvement of ascending ramus and maxillary tuberosity bilaterally
Grade 3: Involvement of entire maxilla and mandible except condylar processes
Grade 4: involvement of whole maxilla and mandible, except the condylar processes with the involvement of the floor of the orbits causing orbital compression
According to Ramon and Engelberg grading system the present case belonged to grade 2 cherubism because mandibular ramus and maxillary tuberosity were involved.
According to the severity of the lesion, Seward and Hankey classified cherubism as follows:[11]

Grade I: Involvement of bilateral mandibular molar region and ascending rami, mandible body or mentis.
Grade II: Involvement of bilateral maxillary tuberosities as well as the lesion of grade I, diffused the whole mandible.
Grade III: Massive involvement of the entire maxilla and mandible except for the condyles.
Grade IV: Involvement of both jaws with condyles.
According to this grading system, the present case belonged to grade II.

Cherubism should be differentiated from fibrous dysplasia, hyperparathyroidism of the jaws, and central giant cell granuloma because of the resemblance of radiologic and histopathological features.[2-4,6] These diseases show a similar radiographical soap bubble appearance with scalloped margins. Despite similarities, cherubism can be distinguished by its limitation to the jawbones, bilateral distribution, early onset, no significant change in biochemical analysis, and regression of the lesion in adulthood.[2,3] Significant alterations in blood calcium, phosphorus, and parathyroid hormone level are features of hyperparathyroidism.[3] Noonan syndrome is characterized by short stature, congenital cardiac disease, facial dysmorphism, and bleeding problems.[3] Central giant cell granuloma is a nonhereditary reactive lesion, unilateral in distribution, the usual age of onset is the second and third decades of life, and it does not regress spontaneously.[2,3] Therefore, fibrous dysplasia, brown tumor of jawbones, and central giant cell granuloma were unlikely diagnoses in the case reported here.
Treatment of cherubism is largely individualized. In most of cases, the condition regressed spontaneously when the patient reaches adulthood. A review of the literature shows that surgical intervention, medical management in the form of calcitonin therapy, and a simply wait-and -see approach were adopted with varying degrees of success. [2,7,8] In our patient we adopt wait and see approach considering the age of the patient.

IV. Conclusion

Cherubism is a benign fibro-osseous lesion that manifests in early childhood. In our patient wait and see approach was taken considering her young age and the absence of serious complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parent has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Legends

Figure 1. Intraoral photograph of the patient
Figure 2. CBCT showing multilocular radiolucent lesion in the mandible as well as maxilla.
Figure3. Hematoxylin-eosin staining of the sections of the lesion showing multinucleated giant cells in loosely arranged connective tissue stroma.

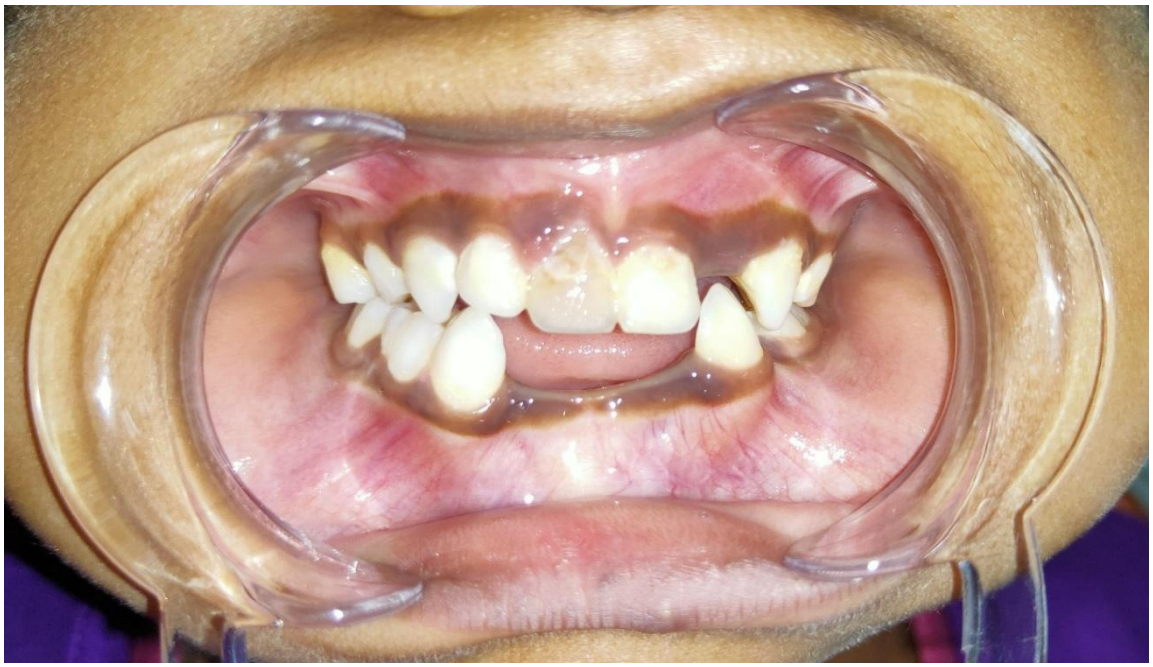

Figure 1

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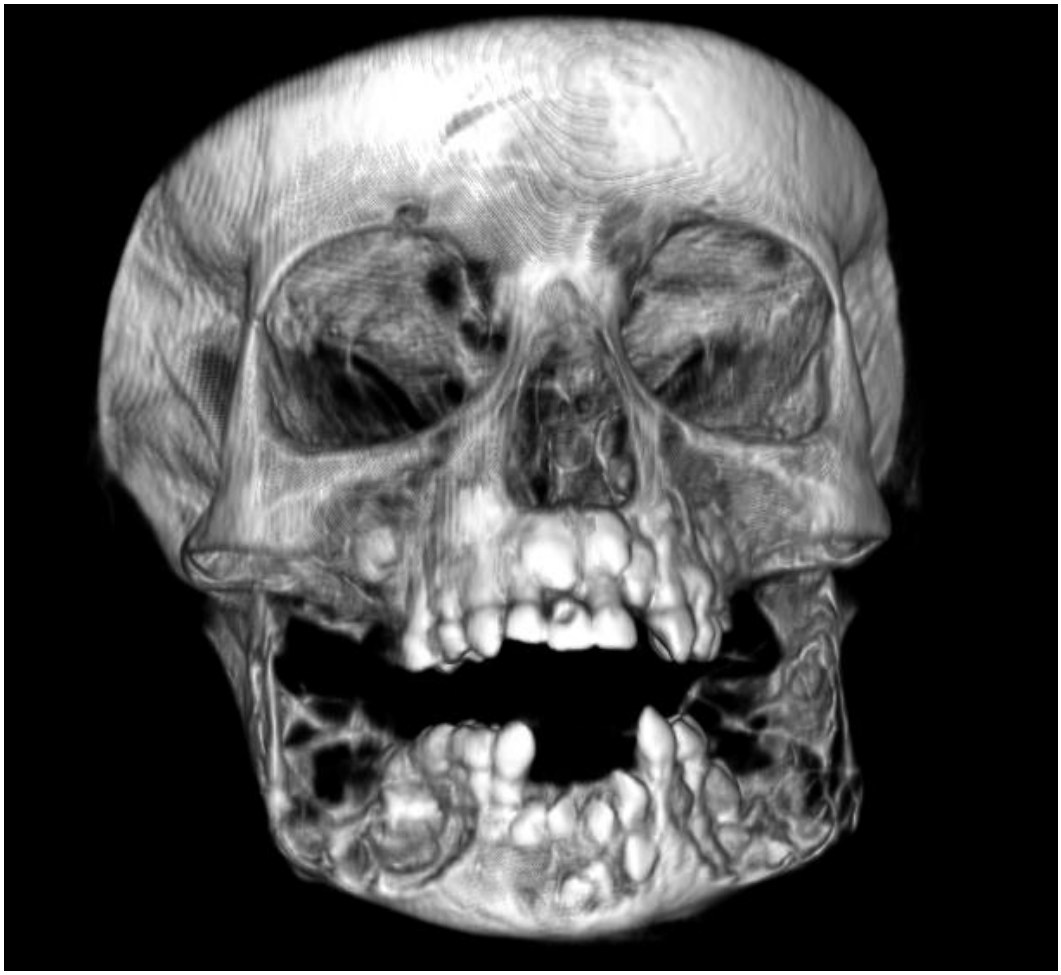

Figure 2

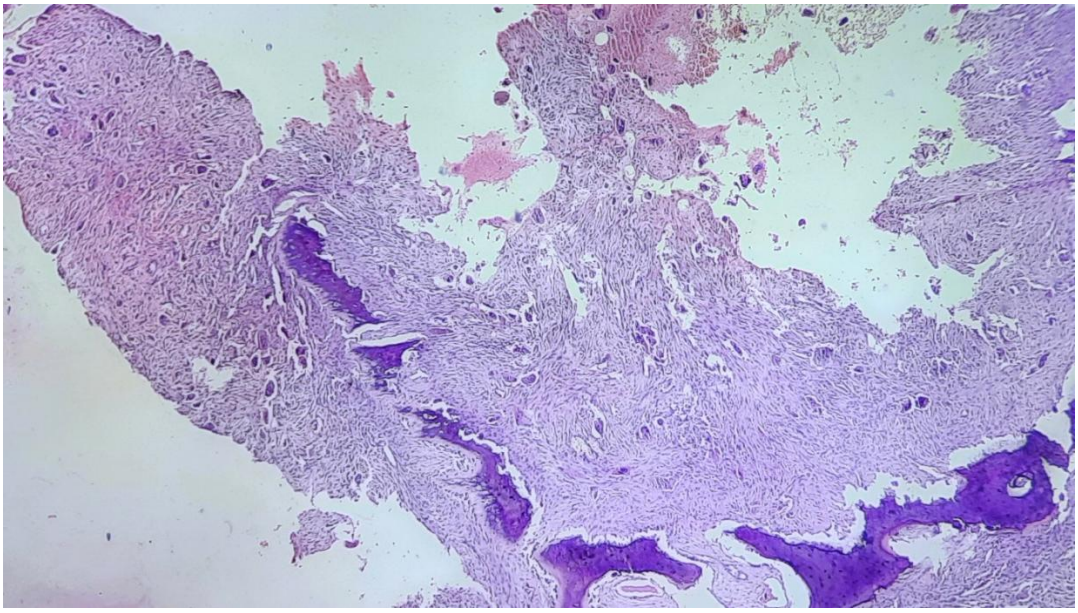

Figure 3