Cherubism – A Case Report

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Abstracts: Cherubism is an autosomal dominant fibro-osseous lesion of jaws involving more than one quadrant that stabilizes after growth period, usually leaving some facial deformity and malocclusion.Here we present a case of 7 years old male patient who came with complaint of bilateral enlargement of lower face since 4 years. A thorough physical and radiographic examination was done and a diagnosis of cherubism was made.

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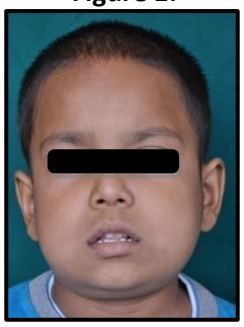
Introduction: Cherubism is a rare inherited fibro-osseous bone disease that affects the jaws. The first case study of this condition was published in 1933 by Jones, who coined the term cherubism to reflect the characteristic facial appearance of affected individuals.[1]

It was first described by Jones as "familial multi-locular cystic disease of the jaws," but the term "cherubism" was later coined to describe the rounded facial appearance resulting from jaw hypertrophy that was reminiscent of cherubs depicted throughout Renaissance art.[2]

Case Report: A 7 years old male patient reported to the department of Oral Medicine & Radiology department with the chief complaint of bilateral enlargement of lower face since 4 years.

Figure 1:



Patient was apparently alright 4 years back when his parents noticed enlargement of lower jaws which has increased gradually to the present size. Similar kind of abnormality was seen in his mother and uncle (maternal side) during their childhood but subsided on its own in adulthood.

Extra oral examination was done and on inspection a bilateral symmetrical enlargement of mandible was evident. On palpation swelling was firm in consistency with slight rise in local temperature over swelling. Examination of lymph node revealed bilateral enlargement of the submandibular lymph nodes which were firm, movable and non tender on palpation.

On intraoral examination of swelling, on inspection a diffuse enlargement was noted on posterior body of mandible bilaterally. On palpation the swelling was firm, fixed and non-tender on the posterior body and anterior ramus of mandible. Buccal and lingual cortical plate expansion was noticed. The overlying mucosa was intact with no pus discharge and ulceration.

Figure 2:

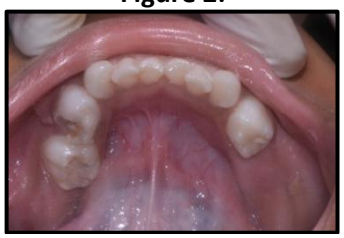
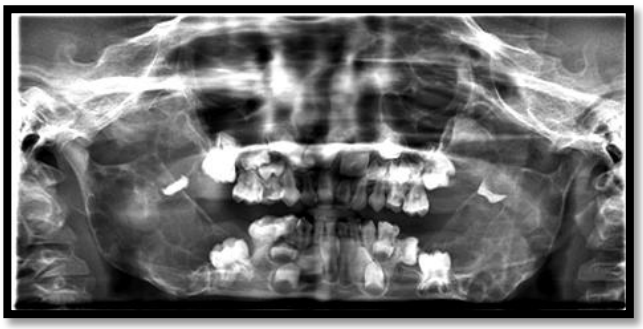


Figure 3:



Radiographic examination with O.P.G was done which showed bilateral multi-locular radiolucency

in posterior mandible extending from distal of deciduous 1st molar till the ramus of mandible sparing the condyles. There was increase in the width of ramus of mandible on both sides. There was loss of antigonial notch bilaterally.

Lab investigations were carried out and it showed increased level of Serum alkaline phosphatase (120 IU/L). Other findings like Serum calcium and Serum phosphorous were within the normal limits.

So based on clinical examination, family history, radiographic and lab examination a diagnosis of cherubism was made. Patient was recalled and reviewed after 1 year and similar features were revealed. So the patient was advised to have a regular follow up every year.

Discussion: Cherub means plump-cheeked little angels. The characteristic chubby facial appearance is because of progressive painless swelling of jaws and heavenward or angelic look is due to severe maxillary swelling causing pressure on floor of orbit leading to upward turn of pupils. Other features include regional lymphadenopathy.

Many cases are inherited as an autosomal dominant fashion. Although several cases without a family history have been reported, using two families with clinically, radiologically, and/or histologically proved cherubism, performed a genomewide linkage search and has localized the gene to chromosome 4p16.3.[3]

Cherubism is a rare osseous disorder of children and adolescents. Although the radiologic characteristics of cherubism are not pathognomonic, the diagnosis is strongly suggested by bilateral relatively symmetric jaw involvement that is limited to the maxilla and mandible. Imaging typically shows expansile remodeling of the involved bones, thinning of the cortexes, and multi-locular radiolucencies with a coarse trabecular pattern.

Oral manifestations includes enlarged Palate: expansion and widening of alveolar ridge causing flattening of palatal vault, premature exfoliation of deciduous teeth, hypodontia, diastema of permanent teeth, malocclusion, early exfoliation of deciduous teeth, impaction and/or displacement of teeth .

Radiographic examination shows **e**xpansive radiolucent, generally multiloculated lesions clearly delimited by cortical bone and distributed bilaterally in the posterior quadrants of the mandible and/or maxilla.[4]

Grading system was developed by Motanedi[5] :

- Grade I: lesions of mandible without signs of root resorption
- Grade II: mandible and maxilla involvement without root resorption
- Grade III: aggressive lesions of mandible with root resorption
- Grade IV: both jaws involved with root resorption
- Grade V: rare, massively growing, aggressive and deforming juvenile cases involving the maxilla and mandible and which may include the coronoid process and condyles

Treatment of cherubism should be based on known natural cause of disease and clinical behaviour of individual case. Surgery is rarely indicated and if necessary, to be undertaken after puberty. It is a self-limiting disease. There is general agreement that in extreme cases in which important functions are impaired, surgical intervention should be performed as early as possible. Radiation therapy has been abandoned as a treatment of cherubism .When possible, follow-up is always a valuable choice.[6]

Conclusion: Cherubism is seen commonly in younger population but can be seen upto the age of 20yrs. So the doctor should be aware of the pathology, its course and management. A consultation with the oral physician and radiologist can be very handy since the dentist might be the first professional sought for a diagnosis of this disease.

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