**CHERUBISM: A CASE REPORT**

**ABSTRACT**

Cherubism is a congenital childhood disease of autosomal dominant inheritance. This disease is characterized by painless bilateral enlargement of the jaws, in which bone is replaced with fibrous tissue. This paper reports a case of cherubism in a 11 year old girl.

**Key words:** Cherubism; Multilocular Radiolucencies; Giant cell lesion; Calcitonin.

**Introduction**

Cherubism is a benign disease with a characteristic symmetrical involvement of the maxilla and mandible. It was first described by Jones in 1933 as a “familial multilocular disease of the jaws” in three siblings who appeared as though they were “looking towards heaven”. This inspired him to call the condition “cherubism”, to describe the round appearance of the cheeks, typical of cherubs, resulting from jaw hypertrophy.

Lannon et al mentioned the necessity to distinguish cherubism from central giant cell granuloma and giant cell tumour of the jaws, with which it holds a false synonymy. This paper reports a case of cherubism in a 11 year old girl.

**Case Report**

An 11 year old female child of non-consanguineous parents presented with painless progressive swelling of bilateral cheek with 5 years of duration. There was no history of similar disease or swelling in any other family members. Girl was healthy and had no relevant medical history. On clinical examination, bilateral swelling of the mandible was present but increased towards the left side than on right side (Figure 1). On palpation both side swellings were hard and non-tender. Submandibular and cervical lymph nodes were not palpable. Intraoral examination revealed missing 36 and erupting 44(Figure 2).

OPG findings (Figure 3) revealed a multilocular radiolucency involving both right and left side of the mandible extending to the angle and ascending rami along with left side coronoid involvement. Both condyles were unaffected and partially developed and the left mandibular first and second molars were displaced to the inferior border of mandible. Based on radiographic examination a provisional diagnosis of cherubism was given. Blood investigation was done for the evaluation of calcium, phosphorus and alkaline phosphatase levels. The report results were within normal limits. Histopathological evaluation (Figure 4) shows multinucleated giant cells, with ovoid to spindle shaped cells within a fine fibrillar collagenous stroma. Based on history, clinical features, radiological and histological findings a final diagnosis of cherubism was given with the grading system proposed by Arnott.

Patients with cherubism, generally males at a proportion of 2:1, present the same clinical characteristics: enlarged face due to swelling of the jaws which is bilateral in most cases, bone consistency of the lesion, intact mucosa, dental malocclusion, upward-looking eyes in the case of maxillary involvement, and absence of pain. In our case patient was female, with bilaterally mandibular involvement. Arnott proposed a grading system for cherubism, according to lesion location and the degree of expansion. Accordingly, grade 1 cases are limited to both ascending rami of the mandible; grade 2 cases involve the maxillary tuberosities and mandibular ascending rami (resulting in congenital absence of the third and occasionally the second molars); and grade 3 cases correspond to massive involvement of both jaws except the coronoid processes and condyles, resulting in considerable facial disfigurement. Ramon and Engelberg added grade 4 in application to cases where all of the classical features of the disorder exceed-
ing grade 3 are present.\textsuperscript{23} In the present case lesion was limited to ascending rami of the mandible excluding the condyle and coronoid.

Radiographically, cherubism is characterized by bilateral, multilocular, radiolucent areas within the jawbones. The lesions usually appear around the mandibular angle and spread to the ascending rami and body of the lower jaw. The maxillary processes may also be involved, and lesions can spread to other facial bones. The extent of the lesions varies from minor to massive involvement of both jaws.\textsuperscript{22} We took an OPG which showed bilateral, multilocular radiolucent areas extending the ascending rami, excluding the condyles of both sides, but both coronoid processes were involved. Radiographically the teeth appears to be floating in the radiolucencies giving “floating tooth appearance”.\textsuperscript{19} In our case at the age of 11 permanent left side of the molar was missing and radiographically showed displaced and tilted with second molar pushed towards the lower border of the mandible. Computed tomography is a useful tool for the assessment of the damage caused by the process either during the analysis of disease progression or during surgical planning.\textsuperscript{11,13} Advancements in virtual three-dimensional reconstruction of anatomic structures based on computed tomography or cone beam computed tomography data can provide for more predictable individual treatment planning.\textsuperscript{23} According to Mnari et al magnetic resonance imaging is useful for identifying orbital involvement.\textsuperscript{24}

Histology: The lesions of cherubism are not distinctive histologically and are difficult to differentiate from other giant cell-containing fibro-osseous disorders. As a result, the diagnosis depends on the clinical findings. Microscopy shows a highly vascular fibrous stroma with unevenly distributed osteoclastic-like multinucleated giant cells that tend to cluster near hemorrhagic foci and deposits of hemosiderin.\textsuperscript{25} With regard to biochemical parameters, serum calcium and phosphorus concentrations and TSH, FSH, LH, T4 and T3 hormone levels are usually within normal limits but alkaline phosphatase levels might be elevated.\textsuperscript{19} Differential diagnosis of cherubism consists of giant cell granuloma of the jaws, osteoclastoma, aneurysmal bone cyst, fibrous dysplasia and hyperparathyroidism.\textsuperscript{5,19}

Treatment: Once the diagnosis is established, therapeutic management should be evaluated. Treatment options include waiting for stabilization and spontaneous remission of the disease, tooth extraction in areas showing fibrous alterations, cosmetic osteoplasty of the affected jaws after regression of disease activity or, in the case of functional impairment, curettage of the lesions and treatment with calcitonin.\textsuperscript{19,26}

The policy of waiting for disease regression, followed by the evaluation of physiological bone remodelling, is the most recommended.\textsuperscript{9,10,12,19} Curettage has been suggested to be as a good approach since this intervention stimulates bone replacement.\textsuperscript{1} Radiation therapy has been abandoned as a treatment of cherubism because of the potential risk of osteo-radio-necrosis or even malignant transformation of the process resulting in osteosarcoma. When possible, follow-up is always a valuable choice.\textsuperscript{11,15,26,27} According to Novack and Faccio,\textsuperscript{19,28} hypothesis that cherubism is caused by enhanced cytokine tumour necrosis factor α (TNF-α) production by myeloid cells due to an activating mutation in Sh3bp2 not only represents a major advancement in the understanding of the disease but suggests new potential options for its treatment. Although cherubism was described more than 70 years ago, the rather sparse literature in this area has provided little insight on disease effective therapies. As a major pathogenic factor of the identification of TNF-α for the patients with this rare disease, is significant news. Anti-TNF therapies are already in clinical practice for the treatment of rheumatoid arthritis. If these drugs will prove the effectiveness in the treatment of cherubism it can be hoped that the interval from laboratory discovery to clinical use would be short.\textsuperscript{19,28}

**Conclusion**

In conclusion the triad of clinical, histologic and radiologic findings helps in the diagnosis of cherubism. However, laboratory investigations of serum calcium, phosphorus and alkaline phosphatase are necessary to differentiate this condition from other similar lesions.

**Authors Affiliations**

1. Nishat Sultana, MDS, Asst. Professor, Department of Oral Medicine and Radiology, 2. M.E. Sham. MD,MDS, Professor, Department of oral and Maxillofacial Surgery, Vydehi Institute of Dental Sciences, Bangalore, India.

**References**


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**Figure 1 Front View, Figure 2 Intra oral View, Figure 3 OPG, Figure 4 Histopathology**


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Address for Correspondence
Dr.Nishat Sultana, MDS, Asst. Professor, Department of Oral Medicine and Radiology, Vydheii Institute of Dental Sciences, Bangalore, Karnataka, India. Email: ehtaihsham@yahoo.com

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