MANAGEMENT OF AGGRESSIVE FORM OF CHERUBISM

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ABSTRACT:
Cherubism is a rare non-neoplastic hereditary disease related to genetic mutations characterized by symmetrically swollen cheeks, particularly over the angles of the mandible, and an upward turning of the eyes. The affected mandible and maxilla begin to swell in early childhood, and are gradually increased until the age of puberty. Apparently, surgical intervention is unnecessary unless significant functional, esthetic, or emotional disturbances develop. The purpose of the present paper is to report a case of cherubism in which all the classic features of the ailment were seen to a degree exceeding even that of the grade 3 classification.

KEYWORDS: Cherubism, hereditary multilocular cystic lesion, non neoplastic lesion, surgical procedure

INTRODUCTION
Cherubism is an inherited, fibro-osseous condition characterized by firm, painless swellings of the jaws, and was first described by Jones in 1933. Cherubism is usually diagnosed in the first decade of life, and has never been observed at birth. Clinically, cherubism is characterized by fullness of the cheeks and jaw bones that results in a round face with retraction of the lower eyelids and exposure of the sclera below the irises; the raised-to-heaven look produced is suggestive of a cherub and gave rise to the name of the condition. The radiographic appearance is characterized by bilateral, multilocular radiolucent lesions, which often begin near the angle of the mandible and spread to the ramus and body of the mandible. Expanding maxillary lesions can result in obliteration of maxillary sinuses. 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 also depends on the clinical findings. The presence of eosinophilic, collagenous material around small capillaries is of value in the diagnosis of cherubism.

Case report
A 20 year old male patient of non-consanguineous parents reported to our department with a complaint of bilateral painless swellings of both the jaws. The boy was apparently asymptomatic till 8 years of age. There after he noticed progressive swelling in his jaws which continued to attain the present size. The size of the swelling has been such since 1 year and has not regressed or increased. There is a family history of similar condition. Apparently the boy’s paternal grandfather and aunt were affected by similar swellings in their jaws. Extra-orally the swellings appeared ovoid in shape, with well defined borders. The swelling was more prominent in the parasymphysis and body of the mandible and also malar regions. Eyes were upturned with sclera visible below the iris (giving eye to heaven appearance) with a difference in the ocular levels right eye being apparently at slightly higher level.

Intraoral examination revealed multiple missing teeth in maxilla and mandible. Obliteration of buccal sulcus of mandible was noticed in the molar regions. In the maxilla multiple swellings were seen, which was hard, sessile, non tender and covered with pink mucosa. The orthopantamograph revealed multiple impacted teeth in both the jaws, with multiloculated osteolytic lesions involving the entire body as well as the rami of the Mandible sparing the condyles. A 3-D CT showed multiloculated cystic lesion affecting the body and rami of the mandible and also the maxilla. Both the orbital floors were seen to be raised. The lesion caused marked expansion of the bones, with a multifocal cortical breakthrough.

Histopathology
Biopsy of the lesion was taken under LA, from both the jaws in the alveolar region. A fleshy, red, profusely bleeding tissue was removed with a surgical curette and hemostasis was achieved followed by primary closure. Microscopy reveals highly cellular stroma consisting of plenty of multinucleated giant cells. The cellular stroma consists of spindle cell fibroblasts with vesicular nuclei.
arranged in fascicles, whorled pattern and also irregularly in few areas. The giant cells were unequally distributed and were of different sizes with varied number of nuclei resembling osteoclasts. There were few blood vessels and areas of extravasated blood and minimal amount of collagen was seen. No evidence of atypia (Fig.3).

Surgical procedure

Because of the grotesque appearance and the parents insistence on esthetic improvement compelled the surgeon to attempt cosmetic surgical recontouring of the jaws under GA. The entire mandible and maxilla was degloved intra orally, and exposed the lesional tissue consisted of multiple locules with reddish hue and semi hard consistency. Buccal surfaces of maxilla and mandible were decorticated and curetted as much as possible to provide an optimal cosmetic result. Impacted and loose teeth were removed.

Discussion:

The pathogenesis of cherubism remain controversial. No cause and effect relationship with trauma, infection, or hemorrhage has ever been verified. Anderson suggested that a genetically induced biochemical abnormality stimulates the giant cell lesions characteristic of cherubism.

Seward and Hankey have proposed a grading system based on the radiographic location of the lesions in the jaws. It is as follows:

Grade 1: Involvement of bilateral mandibular molar regions and ascending rami, mandible body or mentis.

Grade 2: Involvement of bilateral maxillary tuberosities (in addition to grade 1 lesions) and diffuse mandibular involvement.

Grade 3: Massive involvement of the entire maxilla and mandible, except the condyles.

Grade 4: Involvement of both jaws, including the condyles. According to Ramon and Engelberg grade 4 lesions not only push the orbital floor upward but also penetrate it.

Our case could possibly be classified as above grade 3 cherubism as there is huge deformity of the mandible, and especially the bilateral posterolaterosuperior extension of maxillary lesions with slight elevation of orbital floor. The lesions of cherubism are not distinctive histologically and are difficult to differentiate from other giant cells containing fibro-osseous disorders. As a result, the diagnosis also depends on the clinical findings. In our patient differential diagnosis included fibrous dysplasia, central giant cell granuloma, hyperparathyroidism. Giant cell granuloma can be excluded on clinical grounds because it is not bilateral condition, is not inherited and does not regress in adulthood. Blood chemistry studies helped to rule out hyperparathyroidism. Bone changes in hyperparathyroidism rarely cause isolated jaw lesions, but do cause abnormal serum calcium and phosphorus levels. In our case laboratory findings were within normal limits, and there were no associated systemic manifestations.

Treatment of cherubism has not been standardized, making it difficult to recommend management. The possibility of spontaneous involution is recognized, but the frequency is not known because most patients have been treated surgically prior to adulthood. As stated by Laskin “the treatment of cherubism should be based on the known natural course of the disease and the clinical behavior of the individual case.” Surgical treatment appears to be unnecessary for grade 1 and 2 cases in the absence of secondary disturbances. Curettage appears to be necessary in more aggressive cases (grade3), to reduce maxillofacial deformity after puberty. Calcitonin therapy seemed to be effective and resulted in remission of the lesion. Radiation therapy is ineffective and contraindicated in view of the risk of osteoradionecrosis, interference with dentofacial growth and development, and the affect on future surgical procedure.
In this case the patient was an adolescent affected psychologically by his grotesque appearance which was also a concern for his parents. Therefore a conservative surgical curettage and recontouring of the jaws was planned under GA as an attempt to address his aesthetic concern. One year of post operative follow up showed a sustained esthetic improvement achieved by the surgery of the grade 3 lesion (Fig.4). There was a radical improvement in the patient’s psychological and behavioral problems.

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References

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