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Case Report

CHERUBISM: CASE REPORT

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ABSTRACT

Cherubism is a rare hereditary disease having a non neoplastic origin. It occurs in childhood and regresses towards puberty. It has an autosomal dominant mode of inheritance and is characterized by bilateral painless enlargement of jaws. This bilateral enlargement gives a cherubic look to the patient. Apart from striking clinical features, it has a characteristic radiographic and histopathologic appearance. We report a classic case of cherubism in a 7 years old girl who presented with a chief complaint of bilateral enlargement of jaws. A brief of clinical, radiographic features along with review of literature of has been discussed.

INTRODUCTION

Cherubism is a rare, hereditary, non-neoplastic disorder occurring children and young adolescents and regresses after puberty. [1] It presents as a painless bilateral mandibular swelling which leads to fullness of the cheeks. [2] Radiographic features present as a multilocular radiolucency which involves maxilla and/or mandible. It has a characteristic histopathological picture showing the occurrence of giant cells in the fibrovascular stroma. Most cases show an associated family history, but cases with non familial history have also been reported. [2,3]

CASE REPORT

A 7-year-old girl presented to us with a chief complaint of painless swelling of the jaws since 2 years. There was no history of pain, dental or visual difficulty. On examination, enlarged jawsand hard mass in the cheek area were noted. [Fig 1] This was involving body to ramus area of bilateral mandible. The mass was non tender, with palpable bilateral submandibular lymph nodes. intraoral examination revealed a " V" shaped palate, multiple carious teeth and mal alignment of teeth due to enlargement of jaws.

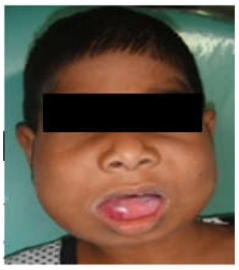

Fig 1 classic appearance of cherubs due to expansion of mandible bilaterally

Past medical and family history was non contributory. Routine blood picture was normal and the biochemical examination revealed alkaline phosphatase and serum calcium and phosphorus levels within normal limits.

Radiographic examination using Orthopantomograph revealed bilateral multilocular radiolucencies involving the maxilla and mandible extensively. The mandibular lesions showed extensive resorption of bone and thinning of cortical plates and the multilocular radiolucencies were extending bilaterally till the condylar portion. Apart from this, there were displaced and multiple unerupted teeth in maxilla and mandible. [Fig 2] Based on the clinical and radiographic features a diagnosis of cherubism was made. Biopsy was advised for jaw swelling and lymph nodes.

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Fig 2 Radiograph shows the extensive lesions & the characteristic multicystic appearance. The lesion extended across the midline & involved both rami upto the condyles

Microscopic examination revealed spindle cell stroma with numerous multinucleated osteoclastic giant cells distributed uniformly in the dense stroma. Few areas revealed dense fibrosis and focal areas showed the stroriform pattern of spindle cells. Areas of hemorrhage around these giant cells were also noted. [Fig 3] Lymph node examination did not render any significant changes. The overall histopathological features were suggestive of Cherubism.

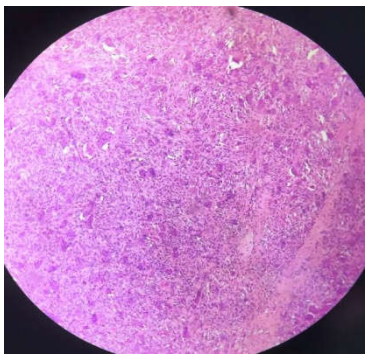

Fig 3 photomicrograph shows numerous multinucleated giant cells in a dense fibrocellular stroma

Patient was kept under follow up without any surgical intervention. No further complications were noted for a follow up of 3 years.

DISCUSSION

Cherubism is an autosomal dominant disorder with male predominance. [2] The present case shows a girl patient affected with the disorder which is quite uncommon. The disease was first described by Jones in 1933 as familial multilocular cystic disease of jaws. Later in 1948 the term 'cherubism' was coined by him which described the conventional distinctiveness of full round cheeks, akin to those of the cherubs commemorate of Renaissance art. [4] The condition has been described hereditary, but even sporadic cases have been reported in literature. The present case is not having any associated family history of the disease. The patient presents only with jaw enlargement without any physical or mental deformity. However, it has been associated with Noonan syndrome, Neurofibromatosis-I and Ramon syndrome. [5]

It has been recognized that the Chromosome 4p16.3, between 4p-telomere and D4S127 and SH3-binding protein2 is responsible for Cherubism. [6]

The children affected are clinically normal at the time of birth and lack evidence of the disease till 3 years of age. [3] Following this age, symmetrical growth of the jaw occurs and this continues till 12-15 years of age. Lesion begins to regress at puberty and jaw remodeling persists till the third decade of life by which the deformity generally becomes unapparent. [4] Signs and symptoms of diseases vary from almost unnoticeable to an apparantely enlarged and distorted maxillary and mandibular lesions. [6]

The jaw lesions are non tender and firm. Lesions start at mandibular angle and involve the ramus and body of mandible. [7] The maxillary lesions show V shaped palate and patient also shows eyes to heaven appearance due to orbital advancement. Dental abnormalities associated are malposed teeth, impacted and abnormally shaped teeth, loss of teeth, agenesis of permanent 2nd and 3rd molar due to intricated tooth germs. There is enlargement of submandibular lymph nodes. [8]

Radiographically, it presents as a well defined multilocular radiolucent lesion, with irregular intersecting bony septae, with thinning of cortical plates. The compressed trabeculae give rise to ground glass appearance. [9]

Based on radiographic features, Seward and Hankey contemplated a grading system on the basis of location of lesion in jaws as: [9]

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

Grade II: In addition to grade I lesions; Involvement of bilateral maxillary tuberosities and diffuse mandibular involvement.

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

Grade IV: Involvement of both jaws, including the condyles.
According to the grading system, the present case is placed under Grade IV.

Histopathology shows plentiful multinucleated giant cells with numerous vascular spaces distributed haphazardly in the dense fibrous connective tissue stroma. The stroma is dense collagenous type with deposits of hemosiderin pigment. Eosinophilic perivascular cuffing is noted and this is considered pathognomonic of cherubism. [10] IHC and histochemical analysis has revealed that multinucleated giant cells are osteoclasts and express vitronectin receptor and show positive reaction for tartrate-resistant acid phosphatase. [11]The histopathological differential diagnosis involves primary hyperparathyroidism and central giant cell granuloma. However, these can be easily ruled out on the basis of clinical examination and histochemical analysis. [12]

Surgery can be performed in cases with serious functional and cosmetic defects. Lesion undergoes regression so mostly the surgery is delayed until puberty. In cases with functional defect, curettage and calcitonin can be administered. Orthodontic treatment for malaligned teeth can be done after the disease has regressed. [4]

CONCLUSION

Cherubism is an autosomal dominant disorder of jaws occurring by the age of 5 years and regresses by puberty. It presents with some dental abnormalities and surgery is mostly avoided as the disease is self limiting.

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