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Familial Fibrous Dysplasia of the Jaws: Cherubism in Two Brothers

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Familial fibrous dysplasia (Cherubism) is a rare disease characterised by painless swelling of the jaws or cheeks, firm protuberant intraalveolar masses and missing or displaced teeth. This condition was first reported by Jones in 1933 in three Jewish siblings(1). It was termed 'Cherubism' because of the typical facial appearance in these children characterised by chubby cheeks with upward projection of the eyeballs, due to growth arising from the maxilla. The disease has a strong familial predilection and probably occurs due to local disturbance in the embryonogenesis.

This report illustrates cherubism diagnosed and treated surgically in two siblings.

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Case Report

Two brothers (PRP and DRP) (*Fig. 1*) aged 7 years and 4 years respectively presented with bilateral symmetrical swellings arising from the lower jaws. These swellings were painless, bony hard in consistency with no signs of inflammation. Dental examination revealed labially positioned mandibular canine and premolar teeth on both the sides which were out of occlusion, bulging into the buccal sulcus of the lower jaws on both sides from the canine to the 1st molar teeth. Maxillae and other bones of the body were unaffected.

Radiology of the lower jaws of both the cases revealed large well defined osteolytic areas within the body and part of the ramus of the mandible on both the sides. There was expansion of bone and thinning of the cortical margins more pronounced on the left side (PRP). A thin septae within the osteolytic lesions were also seen. The unerupted premolars were not well-developed and they were below the roots of the deciduous molars (DRP) (*Fig. 2*). A routine blood picture and urine examination were normal. Serum calcium, phosphorus and alkaline phosphatase levels were within normal limits.

The cases were planned to undergo an excision and curettage for histopathological confirmation and as a part of surgical management. The older child (PRP) was operated upon first on the right side of the lesion. After one week the younger

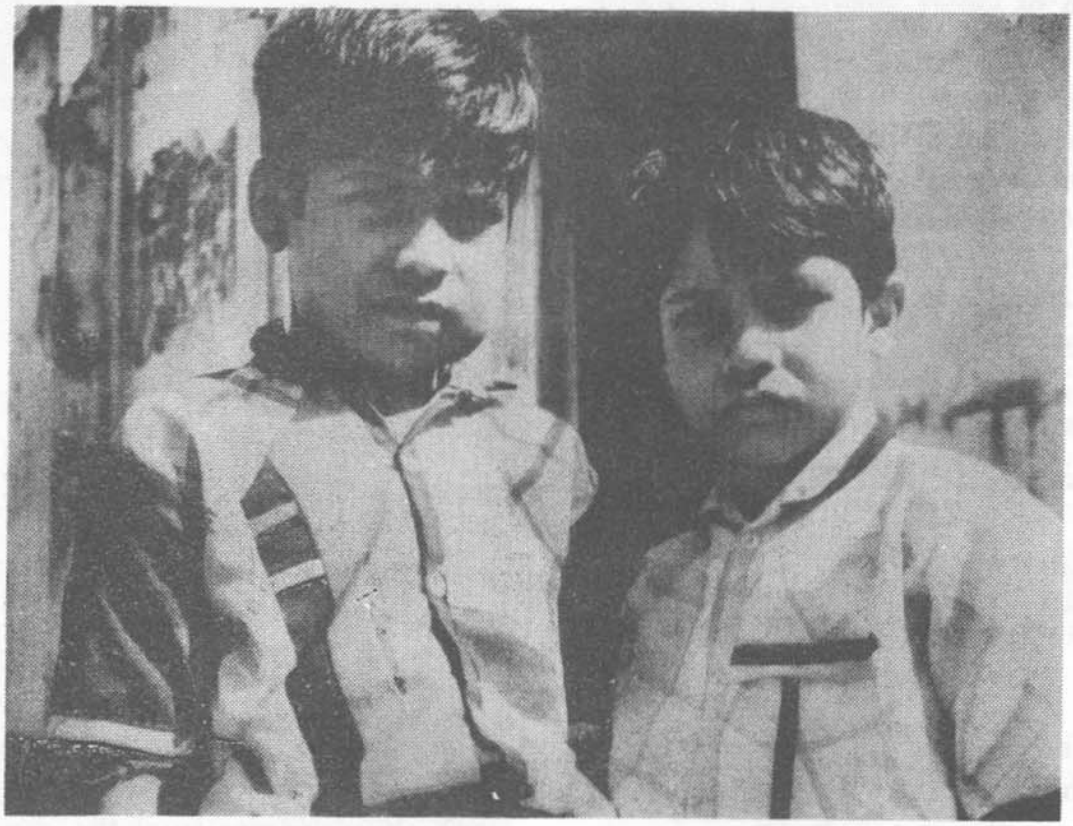

Fig. 1. Clinical photograph of two brothers with cherubism. (L) PRP; (R) DRP.

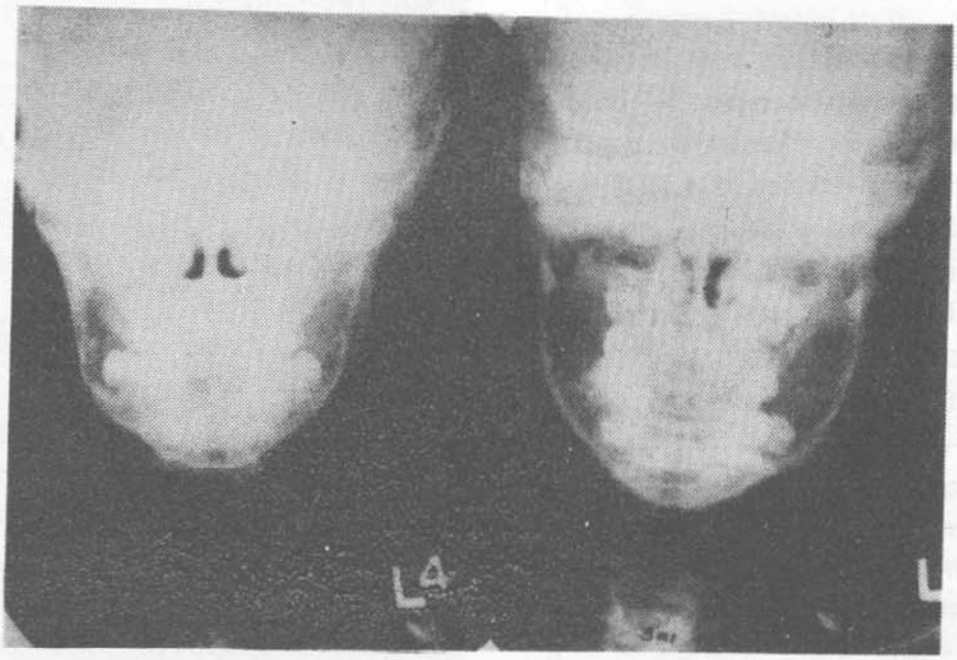

Fig. 2. Radiographic findings of cherubism. The posteroanterior view of mandible shows bilateral expansion of the mandible with extensive destruction, bony expansion and extensive disruption of dentition. (L) DRP; (R) PRP.

child (DRP) was operated on both sides simultaneously. Cosmetically he looked absolutely normal after three weeks. Tempted by this we curetted the left side of the older child (PRP) and postoperatively he developed huge swelling, severe inflammation and subcutaneous abscess formation which needed surgical drainage and antibiotic therapy. The older child (PRP) showed no response to surgical treatment.

The histopathological examination revealed fibrous tissue and collagen fibres replacing the normal bony architecture with presence of cellular elements in between.

Discussion

The clinical manifestations of cherubism varies from a slight jaw swelling causing a little cosmetic problem to a marked expansion of the bones with concomittant difficulties in mastication, speech, swallowing and respiration. The progression of the disease occurs in early years of illness with spontaneous regression reported at puberty and subsequently(2-5).

The exact etiology of this disease is not known, but it has very strong familial predilection(1,6) and involvement of upto five generations of one family have been reported(6). The cases encountered by us also occurred in two siblings suggesting a familial pattern. The suggested mode of inheritance in cherubism is autosomal dominance with varying expressibility of the genes(2,7).

The cases of cherubism have classical radiological features. However, confirmation of diagnosis is possible only by histopathological examination. The radiological features include well defined multilocular radiolucencies occurring due to the destruction of the bony tissue. Numerous displaced and uprooted teeth can be seen floating in the radiolucent spaces(3). The cases with fibrous tissue preponderance show ground glass appearance of the bone.

The histopathological examination reveals replacement of normal bony architecture by fibrous tissue and presence of collagen tissue and giant cells. The prominent eosinophilic perivascular cuffing around capillaries in these lesions is proven to be collagen and is the distinctive diagnostic feature of this condition(8). The index cases had typical radiological and pathological features.

The treatment modalities used for cases of cherubism vary considerably. The therapeutic options used for treating these cases include no active treatment, extraction of teeth in the involved area, surgical contouring of the expanded lesions and complete curettage(2,4,9-11). The controversy in the management exist due to varying clinical manifestations and a known spontaneous regression occurring in these children. Curettage showed good recovery in one out of the two cases treated by us. The other child showed worsening of clinical symptomatology. No definite cause could be attributed for this difference in the clinical response. The infection and abscess formation probably occurred due to an intraoral alveolar incision used by us.

In conclusion, cherubism is a benign tumour of the jaw presenting in early childhood with characteristic radiological and histopathological features. The clinical manifestations vary from mild enlargement of the jaw bone to severe disability. The management in these cases should be individualised and planned according to functional disability of the child. The

final outcome of surgical treatment is variable in these children.

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The Aicardi Syndrome

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Since, the description of Aicardi syndrome in 1965(1), several cases of this rare entity have been reported(2,3). The characteristics of this syndrome are, its occurrence in females, seizures (often flexion spasms), chorioretinal lacunae, agenesis of corpus callosum,mental subnormality and a variable association of vertebral or other osseous abnormalities. The electroencephalogram has multifocal epileptiform abnormalities with burst suppression, arising independently of two hemispheres or hypsarrhythmia(4). Wadia *et al.* perhaps reported the first and only case from India(5). We report a case of Aicardi syndrome who had asymmetry in EEG.

Case Report

An eleven month-old girl was referred to the neurological care of G.B. Pant Hospital for the control of her intractable seizures. She started, initially with myoclonic jerks (infantile spasms) since the age of 2 months. She was first in birth order and product of non consanguineous marriage. Both of her parents were healthy. She was born vaginally at 40 weeks of gestation.

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