Cherubism

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DESCRIPTION

An 11-year-old boy came to the outpatient department for a routine dental check-up. On general physical examination, the boy was well built, healthy and well oriented with time and space. On extraoral examination, a diffuse swelling was noticed in mid-to-lower face region (figure 1A).

Intraoral examination was not remarkable. Orthopantomogram demonstrated bilateral multiple cyst like radiolucencies in the region of angle of mandible (figure 1B–D). Anterioinferior displacement of mandibular second molars was also seen (figure 1B–D).

On the basis of clinical and radiographic features, diagnosis of cherubism was made.

Cherubism is a non-neoplastic hereditary bone lesion characterised by a bilateral, painless swelling of the maxilla and mandible resulting in a fullness of the cheeks and retraction of the lower eyelids giving an upward turned appearance of the eyes comparable with a cherub angel.^{1 2} A molecular pathogenesis of cherubism has been proposed, with the detection of a mutation in the gene

encoding SH3-binding protein 2 (SH3BP2) and possible degradation of the Msx-1 gene which is involved in the regulation of mesenchymal interaction during craniofacial morphogenesis.¹ It is believed that the different clinical manifestations of cherubism are due to the changes secondary to mutations or incomplete penetrance. Ramon and Engelberg³ proposed a grading system for cherubism

- ▶ Grade 1—involvement of both mandibular ascending rami (as seen in our patient);
- ▶ Grade 2—grade 1 plus involvement of both maxillary tuberosities;
- ▶ Grade 3—massive involvement of whole maxilla and mandible, except the condylar processes;
- ▶ Grade 4—grade 3 with involvement of the floor of the orbits causing orbital compression.

The initial clinical signs of this disease usually begin at about 2 years of age, followed by accelerated growth from 8 to 9 years and spontaneous interruption after puberty. Being a self-limiting condition, treatment (liposuction and curettage) is mainly for the aesthetics.^{1 4}

Figure 1 An 11-year-old boy presenting with cherubism. (A) Clinical photograph of facial profile showing fullness of cheeks. (B) Orthopantomogram showing multiple radiolucencies bilaterally in the region of angle of mandible. (C) Cropped orthopantomogram of right side showing multiple radiolucencies involving the mandibular angle (white arrow). Note the antero-inferior displacement of mandibular second molar (black circle). (D) Cropped orthopantomogram of left side showing multiple small round-to-oval-shaped radiolucencies in the region of angle of mandible (white arrow). Developing second molar is also transposed anteriorly (shown by black circle).

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Learning points

- ▶ Cherubism classically presents as bilateral, multiple, multilocular, multicystic appearing expansile lesion affecting the jaw bones. Other features are expansion, thinning of cortices and displacement of teeth. The locules are almost always multiple with well-defined septa in between. The disappearance of internal loculation is a sign of resolution.¹
- ▶ Mutation in SH3BP2 and Msx-1 gene are likely to be responsible for cherubism.
- ▶ It may be associated with syndromes like neurofibromatosis type 1, Noonan-like/multiple giant cell lesion syndrome, Ramon syndrome and Jaffe-Campanacci syndrome.

Competing interests None.

Patient consent Obtained.

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