Fibrous Dysplasia - Cherubism
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History
2 year old with sinusitis and broad nasal bridge.

Diagnosis
Fibrous Dysplasia-Cherubism

Discussion
Cherubism is a subtype of fibrous dysplasia and the result of a point mutation of the SH-3 binding protein on the short arm of chromosome 4 (4p16.3). Radiographically cherubism is characterized as expansile remodeling of the maxilla and/or mandible with mild sclerosis, disorganized trabeculation and absence of periostitis. Differential diagnosis includes craniofacial fibrous dysplasia, brown tumor of hyperparathyroidism, Jaffe-Campanacci syndrome, and familial gigantiform cementoma. Although cherubism and craniofacial fibrous dysplasia of individual lesions show radiologic similarities, they may be distinguished clinically and histologically. Features more specific for the diagnosis of cherubism include bilateral mandibular involvement, limitation to the maxilla and mandible, and involution at the time of puberty. In contrast, patients with fibrous dysplasia typically do not present with swollen cheeks, upward turning of the eyes, or dental derangement. Histologically, patients with cherubism typically have a prominent number of multinucleated giant cells, which are rarely seen in fibrous dysplasia. Brown tumor and Jaffe-Campanacci syndrome are readily distinguished on clinical grounds and are easily eliminated from the differential diagnosis. Familial gigantiform cementoma is usually located primarily in the maxilla and focal rather than diffuse.

Findings
CT-Axial and coronal noncontrast images show symmetric expansion of the maxillae and mentum of the mandible with "cotton-wool" matrix.

Reference