AJNR

A Cherubism with Aneurysmal Bone Cyst

S. Wang, H. Shi, Q. Yu and Y. He

AJNR Am J Neuroradiol published online 22 April 2009
http://www.ajnr.org/content/early/2009/04/22/ajnr.A1626.citation

Published April 22, 2009 as 10.3174/ajnr.A1626

A Cherubism with Aneurysmal Bone Cyst

We report the case of a young man who had cherubism, with secondary aneurysmal bone (ABC) cyst change, to illustrate the clinical characteristics and imaging findings.

A 26-year-old Chinese man was referred to our hospital in February 2006 for maxillofacial consultation. In June 2000, we curetted the enlarged right maxilla, and a pathologic diagnosis of fibrous dysplasia was made. A half-year later, the patient was re-admitted with swelling of the jaws, which had grown rapidly. In the past year before admission, the lesion had been stable. There was no family history of a similar condition.

CT scans showed expansile osseous remodeling with a multilocular appearance and a coarse trabecular pattern affecting the bilateral mandibular bodies and right maxilla (Fig 1). The cysts had fluid-fluid levels. MR imaging showed homogeneous isointensity, including a partial slight low-signal-intensity area on T1-weighted imaging and slight heterogeneous isointensity, including partial high signal intensity, on T2-weighted imaging. The patient underwent curettage. The histopathologic and radiologic findings were suggestive of cherubism with secondary ABC change.

Cherubism is an autosomal dominant inherited disease.[1] It was first described by Jones in 1933[2] as a "familial multilocular cystic disease of the jaws." Clinically, symmetric swelling and an indolent course are characteristic. The mandible is affected more extensively. Maxillary involvement is less extensive and does not occur in the absence of mandibular disease.

ABCs are benign expansile lesions that can be classified as primary (65%) or secondary (35%).[3] Secondary ABC is a pathologic entity that is superimposed on a pre-existing lesion. The most common is giant cell tumor, which accounts for 19%–39% of those cases in which the preceding lesion is found. Giant cell lesions of the jaw bone include giant cell reparative granuloma, brown tumor of hyperparathyroidism, true giant cell tumor, cherubism, and ABC. The typical radiographic appearance of a primary ABC is an eccentric, expanded, and sometimes destructive osteolytic lesion that may contain internal septa. CT and MR imaging often show multiple fluid levels contained within thin-walled vascular cystic spaces. The fluid levels represent hemorrhage. Most (80%) secondary ABCs show the radiographic characteristics of the pre-existing tumor.

Because cherubism may have multilocular radiolucencies with a coarse trabecular pattern, it may be difficult radiographically and pathologically to distinguish between ABC and the spontaneous bleeding into cystic areas of cherubism. In most cases, the radiographic and clinical appearance of the underlying abnormality predominates. To our knowledge, there have been no previous reports of ABC arising in cherubism.

Curettage is the treatment of choice. No clinical sign of recurrence in our patient was observed in the 36 months after the curettage was performed.

References

1. Barnes L, Eveson JW, Reichart P, eds, et al. *World Health Organization Classification of Tumours: Pathology and Genetics of Head and Neck Tumours.* Lyon, France: IARC Press; 2005:324
2. Jones WA. **Familial multilocular cystic disease of the jaws.** *Am J Cancer* 1933;17:946
3. Bonakdarpour A, Levy WM, Aegerter E. **Primary and secondary aneurismal bone cyst: a radiological study of 75 cases.** *Radiology* 1978;126:75–83

S. Wang
H. Shi
Q. Yu
Department of Radiology
Y. He
Department of Oral and Maxillofacial Surgery
Ninth People's Hospital
Medical School, Shanghai Jiao Tong University
Shanghai, P.R. China

DOI 10.3174/ajnr.A1626

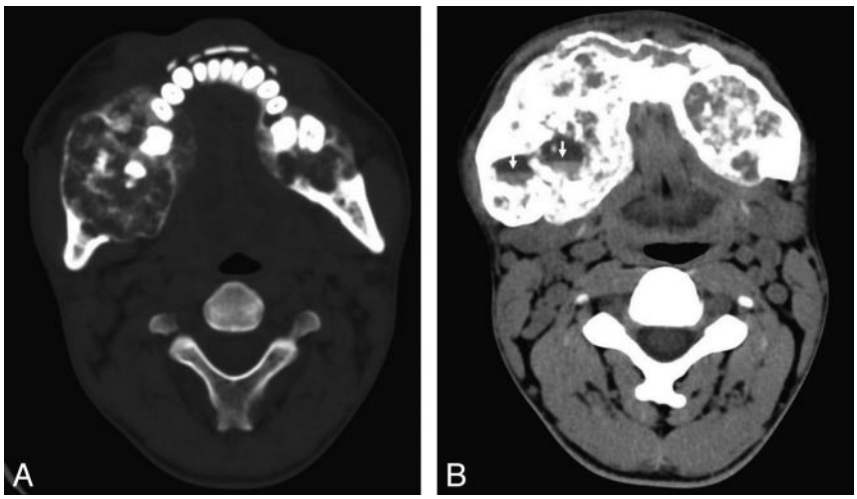

Fig 1. *A* and *B,* Axial CT scans show the extent of the bilateral mandible and right maxilla bone. Note osseous expansile remodeling and a multilocular contour with a coarse trabecular pattern. The cysts have fluid-fluid levels (*arrows*)