The role of MRI and CT in diagnosis and treatment planning of cherubism: a 13-year follow-up case report

ABSTRACT

Aim Cherubism is characterised by mesenchymal alterations during the development of the jaws secondary to perivascular fibrosis. According to the ALARA (As Low As Reasonably Achievable) principle, it is important to avoid conditions where the amount of radiation used is more than that needed for the procedure, because there is no benefit from unnecessary radiation. However, the use of MRI has been poorly studied in cherubism.

Materials and methods The patient underwent head and neck MRI and 3D CT for imaging assessment.

Results MRI is necessary to evaluate the extension of dysplastic tissue and the cystic part of the lesions. Bone window CT only allows evaluation of strong densitometric alterations of cherubism lesions. Moreover, on radiographic film it is not always possible to distinguish fibrous tissue from mucous pseudocystic tissue. By contrast, these differences are readily evident on MRI.

Conclusion MRI, in addition to other traditional radiographs and CT, could be useful in helping the clinician in the diagnosis and treatment of cherubism.

Keywords Cherubism; CT; MRI.

Introduction

Cherubism is a benign hereditary fibrous dysplasia of childhood limited to the maxilla and especially to the mandible. It usually manifests at 5 years of age [Faircloth et al., 1991; Peters, 1979], with painless swelling of the jaws. It is an inherited autosomal dominant disease; the penetrance is 100% in males and 50-70% in females [Quan et al., 1995], with a 2:1 male preponderance [Hitomi et al., 1996; Imai et al., 2003]. It is characterised by mesenchymal alterations during jaw development secondary to perivascular fibrosis.

Cherubism is usually characterised by rapid growth in childhood from 2 years of age until puberty. The lesions regress partially or fully in adulthood, showing sclerotic involution. The spontaneous regression distinguishes cherubism from most other craniofacial fibro-osseous lesions [Beaman et al., 2004]. Radiographs usually reveal multiloculated lucent lesions with well-defined edges and trabeculation. The mandible could display expansile lesions causing cortical thinning, even to the extent of erosion and perforation. The lower alveolar canal could also be displaced downward. Maxillary radiographs could show a lesion with soap bubble appearance involving antra. Non-erupted teeth seem to float in a cyst-like tissue [Cavezian et al., 1981]. A Computed Tomography (CT) analysis is necessary to evaluate the extension of the lesions, especially around the orbital areas, to prevent optic nerve compression [Colombo et al., 2001], and to evaluate the prognosis of the disease [Lupescu et al., 2001]. Cherubism is histologically similar to central giant-cell granuloma, with proliferating fibrous connective tissue containing numerous multinucleated giant-cells. Chronic lesions contain fibrous tissue more than giant-cells. Microvessels of the lesions are typically surrounded by collagen fibers and eosinophilic cells [Hammer, 1969; Zachariades et al., 1985; Kerley and Schow, 1981]. Differential diagnosis includes fibrous dysplasia, central giant-cell granuloma, proliferating fibrous connective tissue containing numerous multinucleated giant-cells. Chronic lesions contain fibrous tissue more than giant-cells. Microvessels of the lesions are typically surrounded by collagen fibers and eosinophilic cells [Hammer, 1969; Zachariades et al., 1985; Kerley and Schow, 1981]. Differential diagnosis includes fibrous dysplasia, central giant-cell granuloma, keratocysts, hyperparathyroidism, paediatric cortical hyperostosis, histiocytosis and ossifying fibroma [Jain and Sharma, 2006; Yamaguchi et al., 1999], and is very simple to perform through dental examination and conventional radiographic exams.

According to the ALARA (As Low As Reasonably Achievable) principle, it is important to avoid that the amount of radiation used is more than that needed for the procedure, because there is no benefit to the patient from unnecessary radiation. However, the use of Magnetic Resonance Imaging (MRI) has not been studied adequately in the diagnosis of cherubism [Beaman et al., 2004; Jain and Sharma, 2006]. We therefore wanted to explore the ability and limits of MRI in cherubism.

Case report

In 1996, a 7-year-old boy came to the Department of Oral Sciences of Sapienza University of Rome. The case history revealed a left hemimandibular, painless...
swelling since 3 years of age. The patient’s mother had cherubism; his 34-year-old father and 8-year-old sister were affected with microcythemia. The patient’s face showed lower third asymmetry, with painless, nontender, left hemimandibular enlargement. There was also a right enlargement, but it was smaller than that on the left side. The intraoral examination revealed swelling on the lower gingival fornix and under the tongue, near the retro-incisor area. A bilateral hard palate swelling simulating a median cleft was also observed. Many of the upper and lower teeth were absent. Perioral sensitivity and mandibular function were preserved.

Panoramic radiography, lateral and frontal view of the skull (Fig. 1), and a 3D CT were performed. The 3D CT revealed important dysplastic lesions involving the mandible (condyles spared), the maxillary alveolar process and antra. The bone tissue appeared thinned, and cortical integrity was interrupted in many areas, especially in the mandible. The infratemporal and oral floor spaces were reduced by the swelling of the mandible. There was a prominence of the posterior part of the hard palate and of the orbital floor that appeared convex upward, with orbital apex constriction, especially of the left orbit, and with a mild bilateral exophthalmus. No cerebral parenchyma alterations were found.

The patient was classified as a “Seward and Hankey” 3rd degree case [Seward and Hankey, 1957].

Orthodontic treatment started in 1997, using a removable lower appliance to gain a comfortable occlusion and to protect erupted teeth.

The biopsy analysis of 1999 revealed “fibrous connective loose tissue fragments containing numerous multinucleated giant cells in open order. Some fragment was adjoining with trabeculae of lamellar or hypocortical bone”. This report confirmed the diagnosis of cherubism.

Orthodontic treatment was resumed in July 2001 to recover and preserve the teeth, and in October 2004 a temporary removable prosthesis was applied to improve dental occlusion.

In March 2007, at the Department of Radiology of Sapienza University of Rome, the patient underwent head and neck MRI and 3D CT. A superconducting magnet of 1.5 Tesla (Siemens, Erlangen, Germany) with head and neck coil was used for the MR exam. The CT exam was performed with a Somatom Sensation Cardiac 64-slice scanner (Siemens, Erlangen, Germany). These examinations (Fig. 2-5) confirmed the presence of dysplastic lesions (4x5 cm), especially involving mental symphysis, and non-embedded teeth. Bone window CT images showed the thin and soft tissue aspect of the dysplastic tissue. The MRI revealed a heterogeneous sign of the lesions with areas of isointensity with skeletal muscle on T1-weighted (T1-W) images, and a heterogeneous aspect with very hypointense areas within a hyperintense tissue on T2-W images. A cystic lesion was clearly delineated near the symphysis area over an embedded tooth, although it was not revealed in CT images. The last 3D CT analysis indicated that bone alterations were steady and the patient had no particular aesthetic requests. A prosthetic rehabilitation will be considered in the future.

**Discussion**

Cherubism is usually progressive until puberty, after which it shows partial or complete spontaneous regression. Bone remodelling goes on until 30 years of age, when clinical signs could be minimal.
Active treatment of the disease is generally not necessary. Surgical treatment should be performed only in case of functional or psychological manifestations. Surgical resection, with or without bone graft, should
be preferred, even more than once. Dukart et al. [1974] believe that surgical treatment during disease development not only provides good and immediate results, but also stops expansion of the lesion, thereby helping bone regeneration. However, other authors [Seward and Hankey, 1957; Riefkohl et al., 1985] have described cases in which surgery, if performed during the acute stage, caused disease aggravation. Bone grafts are indicated in severe lesions with the risk of pathologic jaw fracture.

There are no cranial basis alterations in cherubism; thus, cranial nerves are safe, even if the mandibular nerve could be displaced. 3D CT is the best analysis to evaluate the dimensions of the lesion, the adjoining anatomic structures, and dental malformations. The extension of the lesion using MRI has been evaluated by two authors [Beaman et al., 2004; Jain and Sharma, 2006]: dysplastic bone had T1-W and T2-fat suppression (FS) images isointense with skeletal muscle signals. In the case of cherubism, MRI is necessary to evaluate dysplastic tissue extension and the cystic part of the lesions. In fact, bone window CT allows the evaluation of only strong densitometric alterations of cherubism lesions. Moreover, it is not always possible to distinguish fibrous tissue from the mucous pseudocystic tissue on radiographic film, in contrast to MRI, where these differences are clearly evident. On CT images, it is difficult to distinguish the mandibular nerve within a scantily mineralised bone tissue; by contrast, on T2-W MRI the nerve appears clear because of its characteristic hypointense signal. It is very important to be able to visualise the exact path of the inferior alveolar nerve in surgical treatment planning.

Conclusion

A 13-year follow-up case is reported. The importance of radiographic analysis in evaluating lesion development is emphasised. It is suggested that especially MRI, in addition to other traditional radiographs and CT, could be useful in helping the clinician in the diagnosis and treatment of cherubism.

References