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Clinical and Imagiological Findings of Central Giant Cell Lesion and Cherubism

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Cone beam computed tomography (CBCT) is the best examination for bone lesions of the maxilla, allowing the dentist to evaluate precisely the behavior and components of the lesion and their relationship to the surrounding structures. Central giant cell lesion and cherubism are histologically very similar lesions. Therefore clinical and radiological examinations are fundamentally important for the diagnosis. The aim of this paper is to report two cases diagnosed as central giant cell lesions and cherubism using CBCT. This imaging modality was very important for the diagnosis of the lesions presented in the current study. It also allowed observing precisely the limits of the lesions, the components, the behavior and the exact relationship to adjacent structures.

Key Words: CBCT, cherubism, central giant cell lesion.

Introduction

Computed tomography (CT) is currently the imaging modality most suitable to study bone lesions of the maxillofacial complex, permitting the dentist to evaluate precisely the behavior and components of the lesion. Cone beam computed tomography (CBCT) is widely used in the diagnosis and monitoring of pathologies of the bone and teeth. The interpretation of CBCT images must be carefully performed, always considering lesion location, size and development and relation to the adjacent structures as well as enlargement or disruption of the cortical bone (1-4). With the introduction of CBCT, bone lesions have been more effectively assessed in comparison with conventional radiography, which does not provide details of the lesion and also produces superimposition of images (5-11). Nevertheless, the use of CBCT is still in discussion, since the radiation dose is higher than for the conventional radiographs.

This paper describes the clinical and radiographic findings of central giant cell lesion (CGCL) and cherubism and emphasizes the importance of CBCT images for the diagnosis, follow-up and treatment of these conditions by the presentation of two case reports.

Case Report

Case 1 (Central Giant Cell Lesion)

A 7-year-old male patient presented with extensive swelling on the left side of the mandible, extending to the right side, which had been growing for 6 months. Intraorally this swelling caused bulging of the cortical vestibular bone, and dislocation of the mandibular incisors. The growth caused facial asymmetry and had a hard consistency. A panoramic radiograph revealed a multilocular, radiolucent image, causing a significant bone enlargement. However, owing to its excessive size, it could not be seen completely in the radiograph (Fig. 1).

The patient was then sent to a private radiology clinic for CBCT examination, where an extensive multilocular, hypodense area was observed, causing severe enlargement of the cortical vestibular bone with disruption at some points. Dislocation of the tooth germs was observed (Fig. 2). Proliferation of predominantly spindle-shaped mesenchymal cells, permeated with multinucleated giant

Figure 1. Panoramic radiograph, showing the lesion to be radiopaque at the base of the mandible and extending to the area of the chin, causing dislocation of the tooth germs. The components of the lesion cannot be seen clearly, and the lesion itself does not appear completely.

cells arranged diffusely throughout the lesion was also observed. There were areas of hemorrhage and hemosiderin deposition and presence of reactive bone. Correlating the clinical, radiographic and histopathological findings, a diagnosis of CGCL was established.

The patient underwent treatment involving periodic injections of corticosteroids, with the intent of avoiding mutilation of the mandible. The lesion showed slight regression after several applications. However, the treatment was interrupted because the patient no longer returned to the clinical facility.

Case 2 (Cherubism)

A 30-year-old male patient presented with facial asymmetry, absence and displacement of several teeth. A panoramic radiograph revealed several missing teeth, impacted teeth in the mandible, malposition of teeth and bone loss in the maxilla and mandible (Fig. 3). Patient was sent to a private radiology clinic for CBCT examination for a better evaluation of the lesion. The CBCT scans showed osteolytic lesions extending from the base to the mandibular ramus, causing bone loss and displacement of teeth to the base of the mandible (Fig. 4).

Bringing together the patients medical history, clinical and radiographic findings, it was suggested the diagnosis of cherubism. Review of the patient's family history revealed that his 8-year-old daughter had a similar condition, also presenting progressive bilateral growth, upturned eyes, with no intraoral swelling, which had appeared approximately 10 months before. Physical examination of the girl showed facial deformity characterized by bilateral, asymmetric growth of the cheeks and the infraorbital region. There was no loss of vision or alteration of ocular mobility. Panoramic radiograph showed bilateral osteolytic lesions in the mandible, causing dislocation of the tooth germs (Fig. 5). She was sent to a private radiology clinic for

Figure 2. CBCT axial (A), coronal (B) and sagittal (C) scans showing an extensive multilocular, hypodense area, causing severe enlargement of the cortical vestibular bone with disruption at some points and dislocation of the tooth germs. D: Three-dimensional reconstruction using volume-rendering technique to illustrate the lesion.

CBCT examination in order to have a better evaluation of the lesion. The CBCT scans showed osteolytic lesions, located in the maxilla and mandible, causing bilateral bone enlargement. Pathological progression was observed in the maxilla with a bone enlargement that caused bulging of the orbital floors and partial obliteration of the maxillary, ethmoid and sphenoid sinuses (Fig. 6). The patient's occlusion was compromised by the presence of impacted teeth, agenesis alteration of the axis of eruption of some teeth and problems with the positioning of other teeth. The patient did not undergo any treatment, and is being followed by CBCT once a year.

Discussion

CBCT is widely used in the diagnosis of bone lesions because it provides a better observation of lesion location and its relation with the surrounding structures by the analysis of multiplanar (MPR) and three-dimensional reconstructed images (3D-CBCT) (5). Despite the advantages of this exam, accurate knowledge of its use and limitations is required. In both cases reported in this paper, there is a clear indication for the use of CBCT exam because both lesions are histologically very similar, usually revealing numerous multinucleated giant cells and collagenous stroma, which contains a large number of spindle-shaped fibroblasts (10). Therefore, as much as the clinical examination, radiography is critical for the diagnosis.

CGCL is a benign bone lesion, located in the maxilla and more commonly in the mandible of female patients and usually causes swelling. Facial growth is the primary factor that causes patients to seek help from the health services (6). Conventional radiography is generally the first imaging modality performed in clinical practice. However, it offers limited information about cortical integrity, limits and size of the lesion, requiring an exam that offers more details such as CT (1,7). Cortical disruptions and soft tissue involvement can be observed in CT images, and it is useful for defining the limits of the lesion, its components and in determining its dimensions for diagnosis and follow up purposes (7). Nicolai et al. (8) reported a case of growth of osteolytic lesions on the vestibular cortical bone causing its enlargement. Earlier, the lesion extended to the lateral wall of the nasal cavity, reaching the pre-molars and the canine roots, without causing mobility. Next, the lesion extended to the anterior wall of the maxillary sinus and the superior part expanded to the inferior margin of the orbit (8). A retrospective study with 22 patients (12) showed that painless swelling was the most common clinical feature and the CT images clearly showed the presence of trabeculae within the lesions. Non-aggressive lesions were more frequent.

Jadu et al. (13) reported a case of a rare CGCL in the condyle. The authors described the importance of CT in lesion mapping and treatment planning. In another case report (14), a CGCL was associated with a fibro-osseous lesion and the authors emphasized that the CT images were important to reveal the localization, nature and extent of the lesion.

Similar to the findings in literature, the CGCL reported in the present paper was located in the mandible with large multilocular patterns causing enlargement of the buccal cortical bone and crossing the midline. The lesion behavior did not appear in the panoramic radiograph, and CBCT was very useful for the assessment of lesion dimensions,

Figure 3. Panoramic radiograph showing several missing teeth, impacted teeth in the mandible, malposition of teeth and bone loss in the maxilla and mandible.

location and behavior. Nevertheless, CBCT cannot acquire images of soft tissues. This limitation does not permit the creation of a vascular protocol using intravenous contrast, which would increase the accuracy to determine the limits of the lesion.

Cherubism is a benign giant cell lesion, characterized by symmetrical involvement of the maxilla and the mandible. Some studies report cherubism to be a family pathology, but both hereditary and non-hereditary cases have been described (9). Some reports claim that the lesion is transmitted by a dominant autosomal gene, which was recently located on chromosome 4p16. (15,16). It is more common in males (9), the panoramic radiography characteristics are usually well-defined, radiolucent, multilocular, located bilaterally in the mandible and/or maxilla, in addition to showing dislocation of permanent tooth germs (9,16). CT is very useful in complex cases of cherubism, considering the limitations of conventional

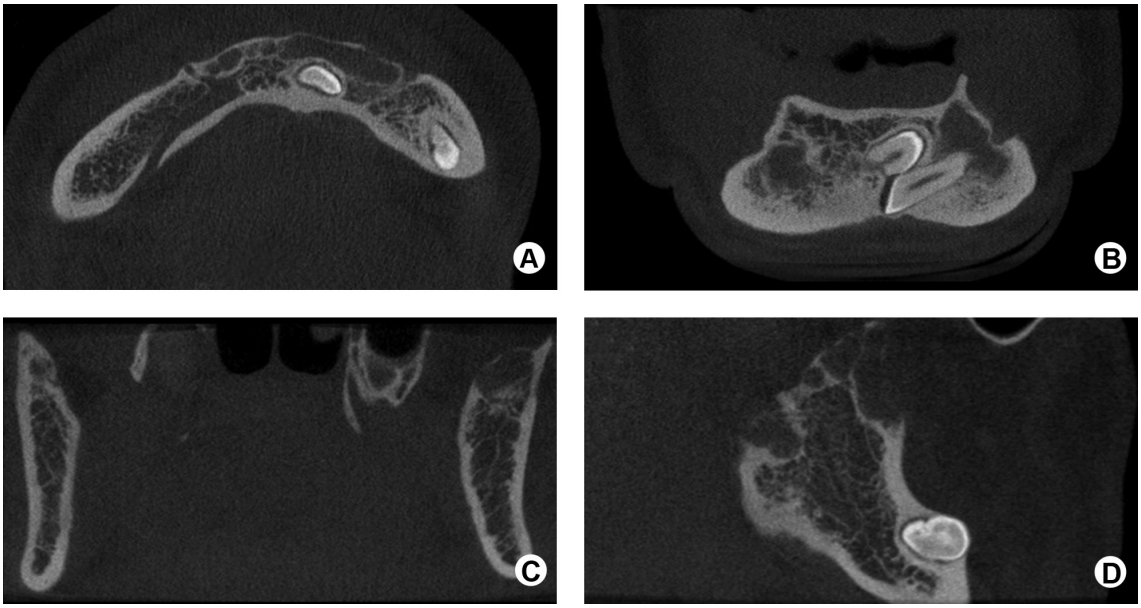

Figure 4. CBCT axial (A), coronal (B and C) and sagittal (D) scans showing osteolytic lesions extending from the base to the mandibular ramus, causing bone loss and displacement of teeth to the base of the mandible.

Figure 5. Panoramic radiograph showing bilateral osteolytic lesions causing dislocation of tooth germs in the mandible. In the image, it can be seen clearly that the lesion also affects the maxilla.

radiography. It makes it possible to verify in greater detail the limits of the lesion and dislocation of tooth germs (11).

In a case reported by Jain et al. (9), CT showed the presence of multilocular, cystic lesions, affecting the body and the sides of the mandible. Similar lesions were also observed in the maxilla (the left side was more affected than the right), ßinvolving the maxillary sinuses associated with sinusitis and causing severe bone enlargement.

In another case report, Colombo et al. (16) reported that CT was effective to provide a bilateral, multicystic image of the orbital floor, and the lesion extending to the edge of the orbit. In our case report, genetic influence was observed in the patient's father, who also showed signs of the lesion. Osteolytic lesions in the maxilla and the mandible were found in the CBCT images, causing bilateral bone enlargement. The progression of the lesion observed in the maxilla also caused bulging of the orbital floors and partial obliteration of the maxillary, ethmoid and sphenoid sinuses.

In a case report described by Wagel et al. (17), the authors found in the panoramic radiograph only expansive multilocular cystic lesions distributed in the mandible and maxilla with loss and medial displacement of the primary teeth. The CBCT revealed a destruction of the maxilla and mandible with soap bubble-like bone remodeling, sparing of the mandibular condyles and thinning of the adjacent cortical rims. The partially overgrown maxilla expanded toward the orbits, with a greater extent on the right side. The anterior wall of the maxilla was also expanded by intraosseus changes. The maxillary sinuses were surrounded by modified tissue, yet with aerial space. The body of the maxilla presented bilateral overgrowth (17).

Chavali et al. (18) reported a case of cherubism in which the panoramic radiograph revealed a well-defined multilocular radiolucency involving the posterior body of the mandible and ascending rami with displaced teeth. Maxillary lesions could not be identified. CT scan showed a surface shaded display image revealing expansible cystic

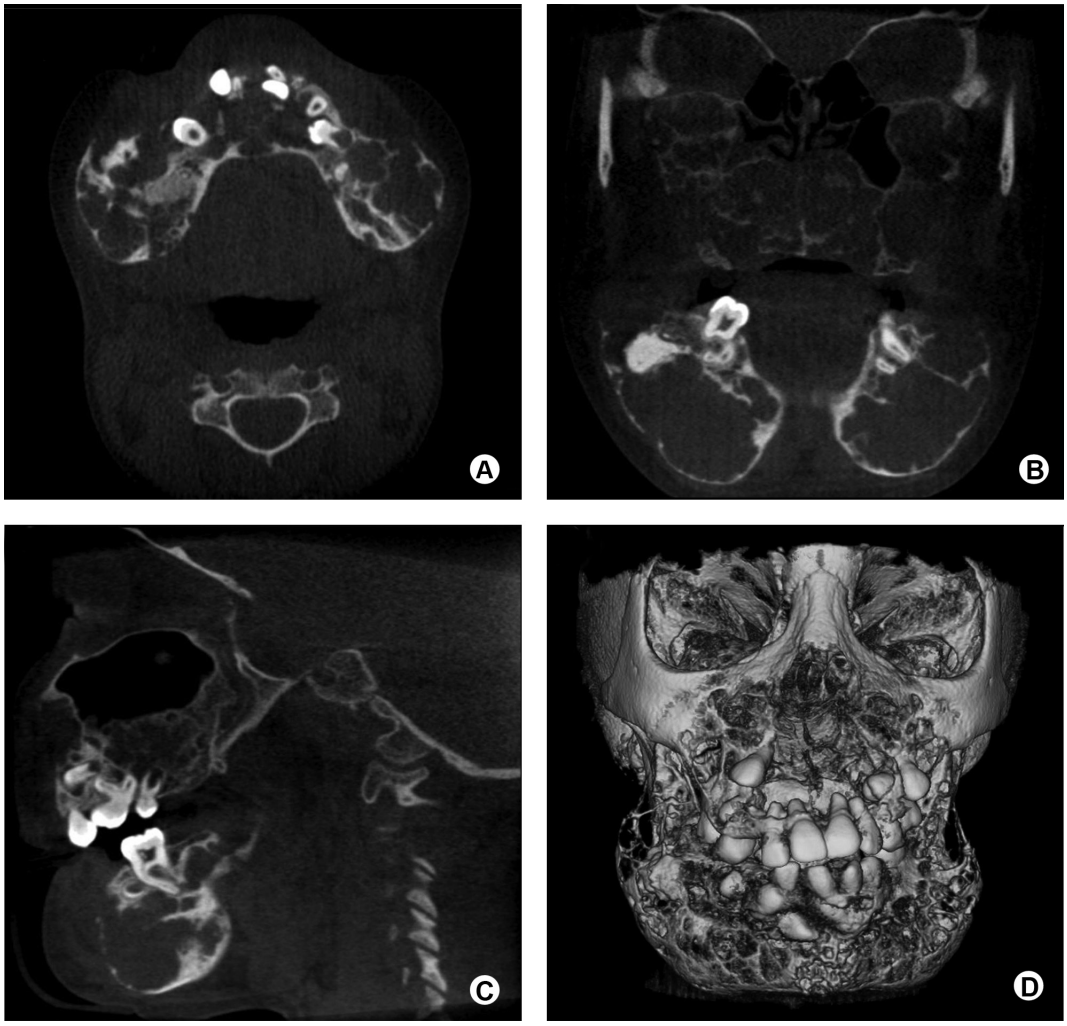

Figure 9. CBCT axial (A), coronal (B) and sagittal (C) scans showing osteolytic lesions located in the maxilla and mandible, causing bilateral bone enlargement. D: Three-dimensional reconstruction, using volume-rendering technique to illustrate the lesion.

lesions with bone remodeling, a mildly sclerotic matrix and internal trabeculations involving both mandible and maxilla (18).

In a previously reported case of cherubism (10), panoramic radiographic examination showed extensive involvement of the mandible. Bilateral, multilocular, radiolucent lesions were found extending from the ascending ramus to the body of the mandible excluding the condyle region. The cortical borders were thin and destroyed in some areas. The panoramic radiograph was insufficient for studying the involvement of maxilla. CT showed bilateral multicystic bone lesions in the mandible and maxilla, expansion and erosion in both cortices of the mandible, bilateral involvement of the maxillary sinus by soft tissue, and invasion of the temporal bone of the left side by the lesion, causing changes of pneumatization of the mastoid cells (10).

The cases of cherubism reported in this paper are similar to those reported by Colombo et al. (16) and Jain et al. (6). In all cases, CBCT added important information, such as involvement of the maxilla, maxillary sinus and orbital floor in the lesion.

In conclusion, CBCT images were very important to observe precisely the limits of the lesions, their components, behavior, the exact relation with surrounding structures, as well as provide more relevant information for the follow-up and treatment. Despite being helpful in the diagnosis, panoramic radiography underestimated the size and limits of the lesions and did not demonstrate clearly their components. All these data are important to establish a better prognosis and treatment planning. These findings confirm the usefulness of CBCT for the diagnosis of bone pathologies.

Resumo

A tomografia computadorizada de feixe cônico (TCFC) é o melhor exame para lesões ósseas da maxila, permitindo que o dentista possa avaliar com mais confiabilidade o comportamento, os componentes da lesão, e sua relação com estruturas adjacentes. A Lesão central de células gigantes e o querubismo são patologias muito semelhantes histologicamente, portanto, exames clínicos e radiológicos são de fundamental importância para o diagnóstico. O objetivo deste trabalho é relatar dois casos diagnosticados usando TCFC, um de lesões centrais de células gigantes e um de querubismo. Esta modalidade de imagem foi muito importante para o diagnóstico das patologias apresentadas neste estudo. Também permitiu observar com mais confiabilidade os limites das lesões, os componentes, o comportamento e a relação exata com as estruturas adjacentes.

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