Intraosseous Ameloblastoma – an Aggressive Lesion of the Jaw

Ameloblastoma Intraósseo – uma Lesão Agressiva da Mandíbula

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Abstract
The ameloblastomas are the most common odontogenic benign tumours, locally aggressive and with clinical impact. We report a case of an old woman with a mixed ameloblastoma. We also review the clinical, imagiologic and surgical state of art of ameloblastomas.

Key-words
Ameloblastoma; Odontogenic tumour; Computed tomography.

Resumo
Os ameloblastomas são os tumores odontogênicos benignos mais comuns, localmente agressivos e com impacto clínico. É relatado um caso de uma mulher idosa com um ameloblastoma misto. Foi feita ainda uma revisão do estado de arte clínico, imagiológico e cirúrgico dos ameloblastomas.

Palavras-chave
Ameloblastoma; Tumores odontogênicos; Tomografia computorizada.

Introduction
Ameloblastomas are benign but locally aggressive neoplasms originating from odontogenic epithelium and are the most common odontogenic tumours of clinical significance, accounting for 10% of all such tumours. They tend to present in the 2nd to 4th decades of life, without sex predominance and are slow-growing painless masses that present with local swelling.

Ameloblastomas can be classified as either intraosseous or extraosseous (peripheral). The intraosseous ameloblastomas arise in the jaw and are classified as unicystic, desmoplastic, and mixed cystic and solid. The radiographic appearance is variable and dependent on the histopathology of the lesion. Computed Tomography is useful to access the extent of the lesion, cortical perforation, and adjacent soft-tissue involvement. The mixed cystic and solid ameloblastoma form usually appears multiloculated with internal septations manifested by a “honeycomb” or “soap-bubble” appearance.

The treatment is surgical resection, and wide margin en-bloc resection is required for tumours that infiltrate through the cyst wall into adjacent bone. We report a case of a mixed cystic and solid ameloblastoma whose diagnostic was suspected in Computed Tomography and was confirmed with a local biopsy and treated with en-bloc surgery.

Case Report
A 72-year-old woman presented with a visible painless bulge over the anterior left mandible, which had been growing for three months, causing facial asymmetry. There was no past history of head and neck malignancy or surgery, including dental surgery. Trigeminal and facial nerve functions were intact, and no mucosal lesions were identified. A contrast-enhanced computed tomography (CT) of the facial bones was performed in order to clarify the diagnosis. It showed an expansive multilocular hypodense lesion - in the left horizontal branch of the mandible, with areas of cortical thinning and interruption (Fig. 1). The soft tissue window showed moderate soft tissue enhancement mixed with cystic areas, involving the left mandibular body including the oral platform (Fig. 2). The lesion was biopsied, and the diagnosis of ameloblastoma was confirmed. A complete surgical resection with curative intent was performed. It consisted of segmental mandibulectomy of the left condyle and left mandibular body (Fig. 3).

Discussion
A variety of benign and malignant lesions occur within the jaw, and they can develop from any tissue including dental elements, bone, nerves, or blood vessels. Mandibular lesions develop from both odontogenic and nonodontogenic origins and have varying degrees of destructive potential.
Odontogenic tumours represent 9% of all tumours of the oral cavity and are designated according to their origin: from the crown (epithelial origin) or apex (ectomesenchymal origin). Ameloblastoma is a benign ectodermal tumour of odontogenic origin and is the most common epithelial odontogenic tumour, that usually appears in the posterior mandible, typically in the third molar region. About 75% of ameloblastomas occur in the mandible, at the level of the bicuspsids and molars and in the angle of the mandible. About 25% of ameloblastomas occur in the maxilla. Follicular cysts or impacted teeth may be associated. The expansion of the mandible can occur as the tumour has slow growth. Ameloblastoma has no gender predilection and demonstrates a peak incidence between 20 and 40 years old. These tumours are slow-growing, painless and can reach a considerable size; swelling is the most common symptom. Small lesions can be asymptomatic. According to the World Health Organization (2005), the ameloblastomas can be categorized on the basis of characteristics such as the age at presentation, location in the body, imaging features, clinical behavior, and prognosis. They can be classified as either intraosseous or extraosseous (peripheral). The peripheral form appears as a sessile or pedunculated slow-growing mass that is confined to the gingiva or alveolar mucosa with no involvement of the underlying bone. The intraosseous form arises in the jaw and is subdivided in: unicystic, desmoplastic, and mixed cystic and solid. The solid/multicystic ameloblastoma is the most common type (85%), and constitutes the second most common odontogenic tumour (after odontoma). The mixed cystic and solid form usually appears in the posterior mandible, as occurs in our case. The mixed cystic and solid form is typically more aggressive and is more likely to recur than unicystic and desmoplastic forms. The radiographic appearance of ameloblastomas is variable and dependent on the histopathology of the lesion. Radiographic findings of the unicystic form include a unilocular, well-circumscribed and well-corticated lucent lesion often associated with the crown of an unerupted or impacted tooth. The peripheral ameloblastoma has a solid appearance on imaging. The desmoplastic variant can be...
distinguished from other variants by the presence of multiple coarse internal calcifications with significant surrounding cortical destruction. The mixed cystic and solid type of ameloblastoma appears as an expansile, radiolucent, uni- or multilocular mass, with internal septations that form a honeycomb or soap bubble. The hallmark of ameloblastoma is extensive tooth root absorption.

In order to plan surgery, helical CT with three-dimensional reformations and holograms should be done. These technique provide valuable insight for the surgeon when considering the primary abnormality and the surgical approach needed.

At CT the ameloblastoma appears with uni or multiple cystic areas of low attenuation and with enhancing solid areas. The unilocular lesions occur most often in the maxilla. The mixed cystic and solid form often has a honeycomb or soap bubble appearance, a classic finding, as occurs in our case. They can vary in size from a small to a large cyst that causes extensive destruction of the jaw. Thinning or frank dehisence of the adjacent cortical surfaces can occur, possibly associated with erosion of the root of the adjacent tooth. Variable resorption of the bone cortex can be present, with extension into adjacent soft tissues. Bony expansion may be found, but there is no periosteal bone formation. Loss of the lamina dura, erosion of the tooth apex and displacement of teeth are also common.

Magnetic Resonance Imaging (MRI) typically shows low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Contrast-enhanced imaging may show papillary projections, enhancing walls, mural nodules and septations.

Surgical resection is the treatment of choice and the prognosis after surgery is linked to the method of treatment, the age of the patient, and the cystic characteristics of the tumour. In order to avoid recurrence, early en-bloc surgical resection should be done. Radiation therapy may be considered if complete resection is not possible or if positive resection margins are not amenable to resection. There is no indication for chemotherapy treatment. Although ameloblastomas are defined as benign neoplasms, they are locally aggressive and infiltrative. If resection is incomplete, they may persist locally or rarely metastasize, although the foci of metastases demonstrate a cytologically benign tumour. Higher recurrence rates are seen in older patients and in those with tumours that are multilocular or exhibit soap bubble-like cysts. In those with multilocular or soap bubble pattern tumours long-term follow-up is needed, as recurrence can occur more than 10 years after initial treatment.

Jaw abnormalities - as ameloblastoma can present in a nonspecific manner, making imaging of utmost importance in elucidating the cause of the symptoms. The contrast-enhanced CT with soft tissue and bone algorithm is the first line and the most useful diagnostic imaging modality to delineate both focal enhancing mural nodules as well as tumor-bone relationships. CT identifies the full extent of the tumor to support surgical planning. The contrast-enhanced MRI can be used when is necessary to define extraosseous components and association with critical neurovascular structures.

References