Ameloblastoma of the mandible: a case report
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Section: Head & neck imaging
Area of Interest: Head and neck Musculoskeletal bone Oncology
Procedure: Diagnostic procedure
Procedure: Staging
Procedure: Contrast agent-intravenous
Procedure: Computer Applications-3D
Procedure: Computer Applications-Detection, diagnosis
Procedure: Computer Applications-General
Imaging Technique: CT
Imaging Technique: MR
Special Focus: Swallowing disorders Tissue characterisation Pathology Neoplasia Case Type: Clinical Cases
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Patient: 40 years, female

Clinical History:

A 40-year-old female patient came to our department for the recent onset of bleeding from the right inferior dental arch, gum pain and difficulty moving the tongue.
Imaging Findings:

An orthopantomography performed elsewhere showed a multilocular radiolucency with sclerotic border at the right jaw with loss of tooth 48.
CT examination (Fig. 1) confirmed the presence of a cystic mass in the right mandible, affecting the posterior part of the body (from 45) and the first part of the ramus.
The lesion caused cancellous bone osteolysis, deformation of the cortical bone with mass effect on the lateral (compressing the masseter and pterygoid muscles) and medial (compressing the tongue) side.
Tooth 48 was completely encompassed by the mass.
At MRI (Fig. 2) it showed to be polycystic with fluid and parenchymatous components and signs of recent bleeding.
It was encapsulated and had regular margins with compression of adjacent structures (including the major salivary glands).
A biopsy was performed and the diagnosis was ameloblastoma.
The patient was treated with "curettage".
After six months follow up CT (Fig. 3) and MRI (Fig. 4) were performed and showed persistence of disease with slightly decreased diameter (40x23mm; previously 40x33), regular margins, decreased displacement of adjacent organs, and heterogeneous contrast enhancement.

Discussion:

Ameloblastoma (AB) is the second most common odontogenic neoplasm [1], representing 1% of all oral odontogenic epithelial tumours and 11% of all odontogenic ones. The incidence of AB is 1 in a million, mainly affecting people between 30 and 60 years of age, with no gender difference [2]. Its aetiology is unknown. In 80% of cases AB affects the mandible (70% of those affecting molars or mandible rami, 20% premolars, 10% canines), in the remaining cases AB affect the maxillary bone [3].
AB has a slow growth and it is normally benign, however, cases of malignant AB are described in the literature, mainly with lung metastases [4, 5].
Symptoms are non-specific: firm swelling, facial asymmetry (or deformity if severe), pain (if AB spread to soft tissue), mobility or displacement of teeth, bleeding, ulcers and periodontal disease. When the tumour increases its dimension, it leads to cortical bone thinning resulting in an "egg shell cracking" (crepitus) sign. The slow growth allows reactive bone formation, leading to deformity of the jaw. In the end AB may perforate the bone spreading to soft tissues [6].
To diagnose AB, imaging findings are essential: Orthopantomography and CT may detect a unilocular/multilocular radiolucency of the jaw with soap-bubble sign or honeycomb sign.
Imaging findings are useful to study the cortical bone, its thinning, dislocation and resorption of teeth.
MRI is essential to study the compression or infiltration of adjacent structures [7].
Histologically AB may be divided in [8]:
- Solid/multicystic type, the most common form, divided in follicular and plexiform type. The follicular one may have prevalence of either spindle, acanthomatous, granular, or basal cells. The plexiform type has prevalence of basal cells in anastomosing strands. Stroma often has cyst-like degeneration.
- Extra-osseous/peripheral type, with the same histological patterns of the previous one but in a different location.
- Desmoplastic type, with a large amount of stroma, compressing the odontogenic epithelial components.
- Unicystic type.
Recent molecular studies associated AB with overexpression of RAB31, a member of RAS oncogene family, calretinin [9], polymorphism of P53 [10] and PTCH1.
Treatment planning should consider tumour size, location, histopathology and clinical/radiographic findings. Treatment is primarily surgical, including conservative surgery with curettage, tumour enucleation and cryosurgery. This option has a very high rate of recurrence (60-80%) probably because it may leave small islands of tumour within the bone [11]. Radical treatment includes marginal and segmental resections with safe margins to reduce recurrences.
In metastatic cases chemotherapy is the treatment of choice; encouraging results have been obtained with
carboplatin-paclitaxel or doxorubicin-cisplatin [12].

**Differential Diagnosis List:** Ameloblastoma of the right jaw relapsed after conservative surgery, Odontogenic keratocyst, Central giant cell granuloma, Calcifying epithelial odontogenic tumor (CEOT), Odontogenic myxoma, Calcifying odontogenic cyst, Ossifying fibroma

**Final Diagnosis:** Ameloblastoma of the right jaw relapsed after conservative surgery

**References:**

Figure 1

Description: Axial CT image: presence of an osteolytic mass of the right jaw

Origin: Department of Radiology, AOUP, Pisa, Italy
**Description:** Axial CT image: the mass compresses the adjacent soft tissue structures (masseter muscle, pterygoid muscle, but mostly the tongue) **Origin:** Department of Radiology, AOUP, Pisa, Italy
Description: Axial CT image: the mass includes tooth 48 and causes alterations of the right jaw, affecting mostly the posterior part of the mandible body. Origin: Department of Radiology, AOUP, Pisa, Italy.
Figure 2

a

Description: Axial T1-wt and T2-wt images: the mass is polycystic with fluid and parenchymatous components and signs of recent bleeding. Origin: Department of Radiology, AOUP, Pisa, Italy

b

Description: Axial T1-wt and T2-wt images: the mass compresses the adjacent organs (tongue, muscles and major salivary glands) Origin: Department of Radiology, AOUP, Pisa, Italy

c

Description: Coronal T2-wt images that shows the compression of the tongue and salivary glands. Origin: Department of Radiology, AOUP, Pisa, Italy
**Description:** Axial T1-wt during contrast administration: the second image is taken 5 minutes after the first. The mass has a heterogeneous contrast enhancement. **Origin:** Department of Radiology, AOUP, Pisa, Italy
**Figure 3**

Description: Axial CT image: persistence of disease (in its cranial portion), less compression of adjacent organs. 

Origin: Department of Radiology, AOUP, Pisa, Italy
Description: Axial CT images of the same layer, the first in bone window, the second in soft tissue window: persistence of the tumour, the extension is only slightly decreased in 6 months after curettage

Origin: Department of Radiology, AOUP, Pisa, Italy

Description: Axial CT image: the mass in its caudal portion still compresses the tongue

Origin: Department of Radiology, AOUP, Pisa, Italy
Description: Sagittal and coronal MPR images showing the extension of disease, the anterior lower part of the mass shows new bone apposition. **Origin:** Department of Radiology, AOUP, Pisa, Italy

Description: Sagittal MPR image showing the mandibular nerve canal, adjacent to the mass **Origin:** Department of Radiology, AOUP, Pisa, Italy
Description: Coronal MPR images, showing at two different levels the mandibular nerve canal, adjacent to but not infiltrated by the mass. Origin: Department of Radiology, AOUP, Pisa, Italy.

Description: 3D volume rendering image: the mass occupies the right posterior part of the mandibular body. Origin: Department of Radiology, AOUP, Pisa, Italy.
Description: 3D volume rendering image: reconstruction of the mandible viewed from above, useful to plan aggressive surgery

Origin: Department of Radiology, AOUP, Pisa, Italy
Figure 4

Description: Axial T1-wt and T2-wt images at the same level: persistence of disease

Origin: Department of Radiology, AOUP, Pisa, Italy
Description: Coronal T2-wt image: decreased compression of the adjacent structures

Origin:
Department of Radiology, AOUP, Pisa, Italy
**Description:** Sagittal T2-wt and T1-wt (during contrast administration): note the cystic areas hyperintense in T2-wt and hypointense in T1-wt with heterogeneous contrast enhancement

**Origin:** Department of Radiology, AOUP, Pisa, Italy