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AJNR Am J Neuroradiol 1993, 14 (1) 223-226 http://www.ajnr.org/content/14/1/223

This information is current as of May 5, 2025.

Ameloblastoma of the Maxilla: CT and MR Appearance

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Summary: This paper describes the CT and MR appearance of an ameloblastoma that involved the maxilla, infratemporal fossa, and adjacent structures. Although not pathognomonic, the multicystic appearance of an ameloblastoma may suggest the correct diagnosis.

Index terms: Paranasal sinuses, neoplasms; Maxilla, magnetic resonance; Maxilla, computed tomography; Maxilla, neoplasms

Ameloblastoma of the jaw is an odontogenic tumor comprising epithelial elements (1). Most ameloblastomas are benign. The plain film appearance of a multilocular "bubble-like" lucency (2) with no calcification in the matrix (3) has been described. However, the appearance of ameloblastoma on computed tomography (CT) and magnetic resonance (MR) imaging has received relatively little attention (4–6).

Case Report

A 70-year-old man presented with 1 year of progressive diplopia and proptosis, plus mild left otalgia. Twelve years earlier he had undergone partial maxillectomy for a left maxillary ameloblastoma. Physical examination revealed a firm submucosal mass involving the left half of the hard palate, the anterior tonsillar pillar, and the retromolar trigone. The left nasal cavity was obstructed by a firm submucosal mass, and the posterior choana was obstructed by a submucosal nasopharyngeal mass. There was mild prominence of the left cheek, and proptosis of the left eye.

A CT scan (GE 9800, Milwaukee, WI) was obtained after intravenous administration of iodinated nonionic contrast (Isovue (iopamidol), Squibb, Princton, NJ). Contiguous axial and coronal images 3 mm thick were obtained through the facial bones and skull base. An inhomogeneous mass with a multilocular, whorled appearance filled the left maxillary sinus (Figs. 1A–1C). Areas of low attenuation were separated by curvilinear areas of intermediate to high attenuation. There was one dense calcification (Fig. 1A). The mass eroded through the posterolateral wall of the sinus into the masticator space and pterygopalatine fossa (Fig. 1B), through the medial wall of the sinus into the nasal cavity (Figs. 1B and 1C), and through the roof of the sinus into

the floor of the orbit and ethmoid sinus (Fig. 1C). The maxillary tuberosity (Fig. 1A), posterior hard palate, the superior alveolar ridge, and the pterygoid plates were eroded (Fig. 1A). The pterygoid muscles on the left could not be identified (Figs. 1A and 1B). The left fossa of Rosenmuller was deformed by the mass, and the mastoid and middle ear were opacified (Fig. 1B). The mandible was intact.

An MR scan (GE Max, 0.5 T) was obtained (Figs. 1D-1G). Axial and coronal T1-weighted images 3 mm thick with a 1-mm intersection gap were acquired before and after the intravenous administration of gadopentetate dimeglumine (Magnevist, Berlex, Cedar Knolls, NJ). Axial T2-weighted images were also acquired. These were 5 mm thick, with a 2-mm intersection gap. The precontrast T1weighted images demonstrated a slightly inhomogenous tumor, largely isointense with muscle, with a few areas of lower signal intensity (Fig. 1D). There were two foci of high signal that could have represented hemorrhage or high protein (Fig. 1E). (On the corresponding CT images, no discrete areas of high density could be identified in these locations, but all CT images were obtained after contrast, which may have obscured subtle areas of high density.) The lesion enhanced intensely, but inhomogeneously, with a multilocular pattern created by many small rounded foci of low to intermediate signal intensity that did not enhance, separated by enhancing tumor tissue (Fig. 1F). On T2weighted images, the areas of low signal brightened considerably, while the septae between them remained of intermediate signal intensity (Fig. 1G). There was a discrete signal void (Figs. 1D, 1F and 1G) that corresponded to the calcification seen on CT.

Because of the patient's visual symptoms and the lack of other effective therapy, he underwent a left radical maxillectomy and a transfacial approach to the skull base. Grossly, all of the tumor was resected. The orbit was preserved. The surgical defect was reconstructed with temporalis muscle and skin graft.

Histologically, the tumor contained plexiform and follicular patterns, and many foci of cystic change (Fig. 1H). The large calcification seen on the CT and MR was composed of a poorly organized mass resembling cementum. Dentin and tooth enamel were not seen.

Received November 12, 1991; revision requested March 19, 1992; revision received April 7 and accepted July 8.

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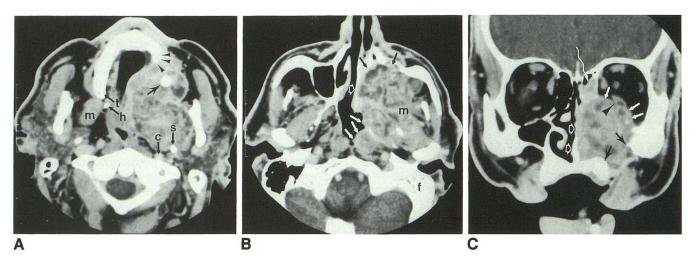


Fig. 1. A-C, Contrast-enhanced CT scan.

A, Axial image through the superior alveolus. An inhomogeneous mass with one focus of dense calcification (*unlabeled arrow*) erodes the left maxillary tuberosity, superior alveolar ridge (*arrowheads*), and hamulus, and extends posteriorly as far as the internal carotid artery (c) and styloid process (s). The mass has many foci of low density interspersed with more dense septations. The pterygoid muscles cannot be identified. Normal right maxillary tuberosity (t), hamulus (t), and medial pterygoid muscle (t).

B, Axial image through the maxillary sinus. The left maxillary sinus is completely filled by the inhomogeneous mass, which has partially destroyed the sinus walls (*small black arrows*). The mass extends medially up to the nasal septum (*open arrow*), and through the pterygopalatine fossa into the maxticator space (*m*). The pterygoid processes are destroyed. The left nasopharynx is deformed (*small white arrows*). There is fluid in the left mastoid air cells (*f*).

C, Coronal image through the maxillary sinus and orbit. The mass in the maxillary sinus has extended through the sinus roof (*small arrows*) into the orbit. The mass abuts the inferior rectus muscle (*black arrowhead*). Medially, the superior extent of the mass is the ethmoid sinus (*wavy arrow*), and, below this, the nasal septum (*open arrows*). Inferiorly, the maxilla is eroded (*black arrows*).

Discussion

Ameloblastoma is an odontogenic tumor comprising epithelial elements (1). The tumor may arise from the enamel organ (odontogenic apparatus) (1), follicle, periodontal membrane (2), lining of an odontogenic (dentigerous) cyst (1, 2), or marrow of the jaw (2). The name "adamantinoma" is sometimes used interchangeably with ameloblastoma. However, ameloblastomas never differentiate to form enamel, and may not even contain ameloblasts (7). Adamantinoma, suggesting the presence of hard material (3), is therefore a misnomer (1).

Ameloblastomas represent 1% of all jaw tumors (1, 8). Most present in the third and fourth decades of life as a painless swelling. Most series have found an equal sex distribution or slight male predominance (1). Eighty percent of ameloblastomas arise in the mandible, most in the molar region and ramus. The remaining 20% arise in the maxilla. The maxillary tuberosity is the most common location for ameloblastomas of the upper jaw (1). The tumor described in this report appears to have arisen from the maxillary tuberosity.

Ameloblastomas are nonencapsulated and locally invasive. There are two histologic patterns, and both patterns are usually present in the same lesion, as in the case presented here. The more common follicular pattern consists of islands of epithelial elements in a stroma of mature fibrous connective tissue (1). The plexiform pattern contains cords of epithelial elements in the same mature fibrous stroma. Areas of cystic transformation are often seen.

The histology does not predict the biologic behavior. Recurrence after excision is seen in 33% of reported cases (8). Metastasis, indicative of malignancy, is rare (8), and tends to occur after many resections and recurrences (1). The metastases are histologically benign (3). The presence of metastases, rather than their histology, determines the tumor's classification as malignant.

An ameloblastoma may cause resorption of the roots of an adjacent tooth, which is most often a molar. The maxilla of the patient described here was edentulous.

Most of what little has been written about the CT or MR appearance of ameloblastomas deals with mandibular lesions. Hertzanu et al (4) described the CT appearance of mandibular ameloblastoma as a multiseptated soft-tissue mass. Heffez et al (5) compared CT with MR in assessing

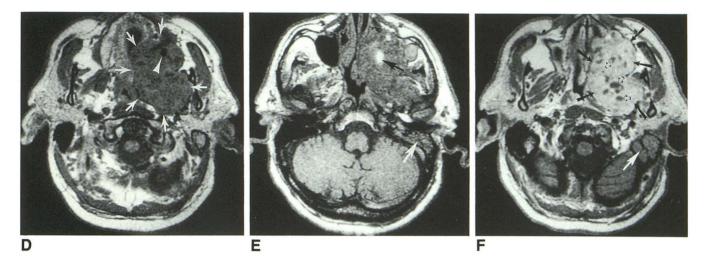


Fig. 1. — Continued. D-F, Axial MR images.

- D, Unenhanced T1-weighted 480/25/4 (TR/TE excitations) image through the alveolus. The mass (*arrows*) is isointense with muscle, with foci of lower signal. One focus of signal void (*arrowhead*) corresponds to the dense calcification seen on CT (Fig. 1A).
- E, Unenhanced T1-weighted image through the maxillary sinus. There is a focus of high signal (*black arrow*) within the tumor which could represent methemoglobin on this unenhanced scan. A left mastoid effusion is present (*white arrow*).
- F, Gadolinium-enhanced T1-weighted (480/25/4) image through the floor of the maxillary sinus. The tumor (*solid black arrows*) enhances intensely but inhomogeneously. Nonenhancing low-signal foci (*open arrows*) persist throughout the mass. The material in the mastoid (*white arrow*) does not enhance, indicating that this is fluid and not tumor.

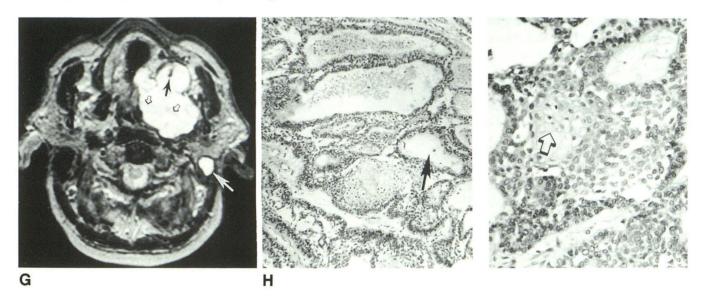


Fig. 1. — Continued. G, Axial MR T2-weighted (2500/80/1) image through the alveolus. The tumor is hyperintense, though septations of lower signal intensity are still seen (open arrows). The signal void of the calcification (solid black arrow) seen on the CT and T1-weighted images is again apparent. The mastoid effusion is hyperintense (white arrow).

H, Pathology specimen. Both follicular and plexiform growth patterns are present, with foci of cystic transformation (*solid arrow*). At the center of the epithelial islands, there is a loose network of cells resembling stellate reticulum and showing squamous metaplasia (*open arrow*). Hematoxilyneosin ×40 at left; hematoxilyneosin ×350 at right.

conspicuity, artifact degradation, and edge definition in three cases of ameloblastoma, one of which had inhomogeneous signal representing solid and cystic areas, confirmed histologically. Schultz et al (6) described a multiseptated, expansile ameloblastoma of the mandibular ramus in a patient with basal cell nevus (Gorlin) syndrome. Although the appearance of this tumor on CT or MR is not pathognomonic, there are features that may suggest the diagnosis, or help exclude other more common sinus tumors. The importance in differentiating ameloblastoma from the more common malignant neoplasms of the sinonasal region lies in the different prognosis each

tumor carries, and therefore the different therapeutic implications. Squamous cell carcinomas of the paranasal sinuses are generally homogeneous, without the many cystic foci seen here. The degree of contrast enhancement of this ameloblastoma, as well as the marked hyperintensity on T2-weighted sequences, would be unusual for a squamous cell tumor (3). Minor salivary gland malignancies such as adenoid cystic carcinoma and the more rare colonic-type adenocarcinoma may be inhomogeneous. Adenoid cystic carcinoma may demonstrate perineural spread, which was not seen here. Most colonic-type adenocarcinomas arise in the ethmoids (3). The calcification in this tumor would also be unusual for squamous cell and adenoid cystic carcinomas.

In summary, ameloblastoma is an invasive tumor of the jaws that most frequently arises from the mandible. The less common maxillary ameloblastoma, as in the case presented here, usually originates around the maxillary tuberosity. This location, and the extensive local invasion, may suggest a more common malignancy, such as squamous cell or adenoid cystic carcinoma of the paranasal sinuses or oral mucosa.

With the development of sophisticated techniques facilitating resection of skull base tumors, even invasive maxillary ameloblastomas may be amenable to surgery. As with other tumors of the

skull base, CT and MR contribute to preoperative planning by delineating the anatomic extent of the tumor. The CT and MR appearance of a multicystic, enhancing mass with aggressive local invasion but no evidence of perineural spread is unusual for the more common malignancies of the sinonasal cavity and skull base. Therefore, this appearance could raise the diagnostic possibility of an ameloblastoma.

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