

AMSER Rad Path Case of the Month:

41 year old with recurrent facial swelling

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Patient Presentation

HPI: 41 year old female with history of recurrent ameloblastoma presents with increased left facial swelling progressing steadily over several months with new onset intraoral lesion. The oral lesion is painful, but she denies dysphagia, weight change.

PMHx: Ameloblastoma of jaw, ESRD on dialysis, CHF, HTN, DMII

Surgical Hx: Mandibular resection and reconstruction (2016), Mandibular osteotomy (2017)

Social Hx: Non-smoker, no alcohol use, no illicit drug use

Physical exam: large mass left masseter space extending posteriorly, numbness left lower lip and left lateral chin, no neck or salivary gland masses

ACR Appropriateness Criteria

Variant 3: Parotid region mass(es). Initial imaging.		
Procedure	Appropriateness Category	Relative Radiation Level
CT neck with IV contrast	Usually Appropriate	☼ ☼ ☼
MRI neck without and with IV contrast	Usually Appropriate	○
US neck	Usually Appropriate	○
MRI neck with parotid sialography without and with IV contrast	May Be Appropriate	○
MRI neck with parotid sialography without IV contrast	May Be Appropriate	○
MRI neck without IV contrast	May Be Appropriate	○
CT neck without IV contrast	May Be Appropriate	☼ ☼ ☼
Fluoroscopy sialography parotid	May Be Appropriate (Disagreement)	Varies
CT neck with parotid sialography	Usually Not Appropriate	☼ ☼ ☼
CT neck without and with IV contrast	Usually Not Appropriate	☼ ☼ ☼
CTA neck with IV contrast	Usually Not Appropriate	☼ ☼ ☼
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☼ ☼ ☼ ☼
FDG-PET/MRI skull base to mid-thigh	Usually Not Appropriate	☼ ☼ ☼
MRA neck without and with IV contrast	Usually Not Appropriate	○
MRA neck without IV contrast	Usually Not Appropriate	○
Arteriography cervicocerebral	Usually Not Appropriate	☼ ☼ ☼

Radiology Images (not labeled)

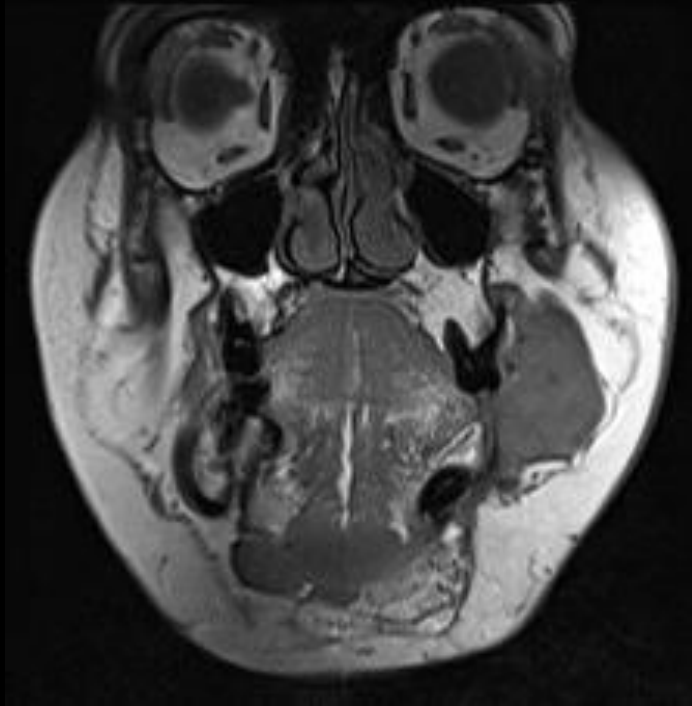


Axial CT with IV contrast

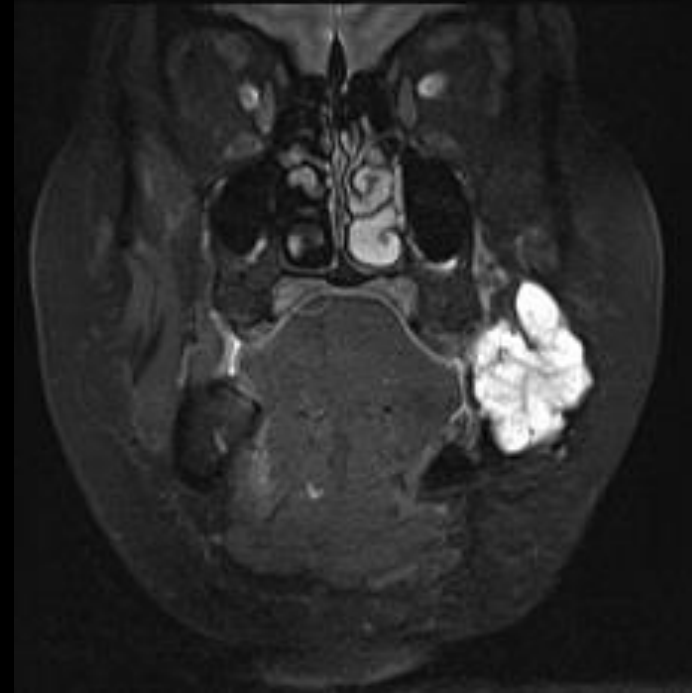


Coronal CT with IV contrast

Radiology Images (not labeled)

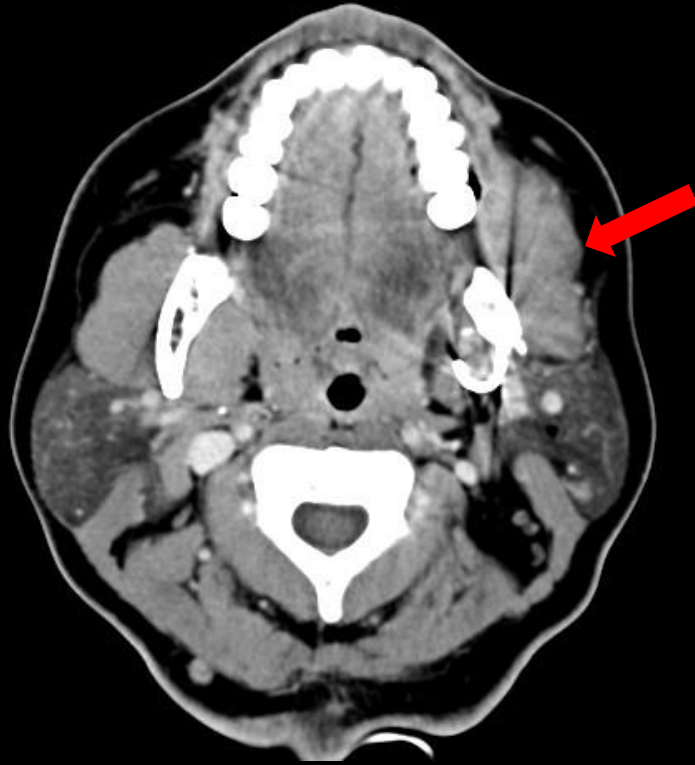


T1 Coronal MRI

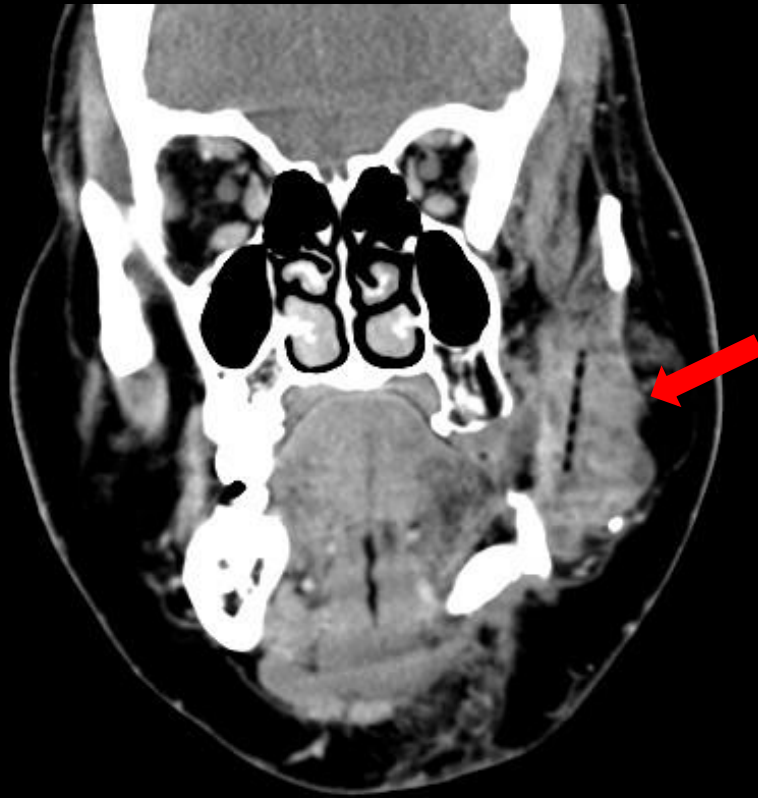


T2 Fat Sat Coronal MRI

Radiology Images (labeled)



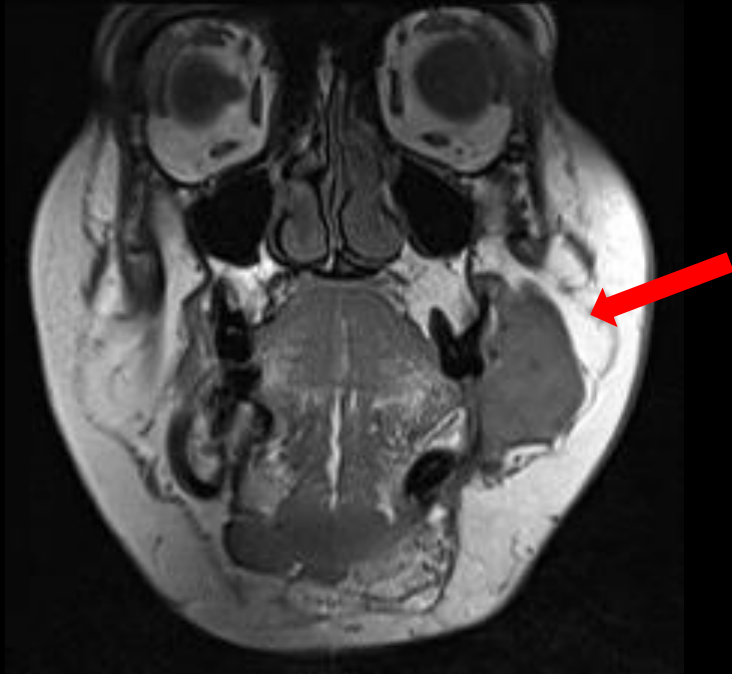
Axial CT with IV contrast



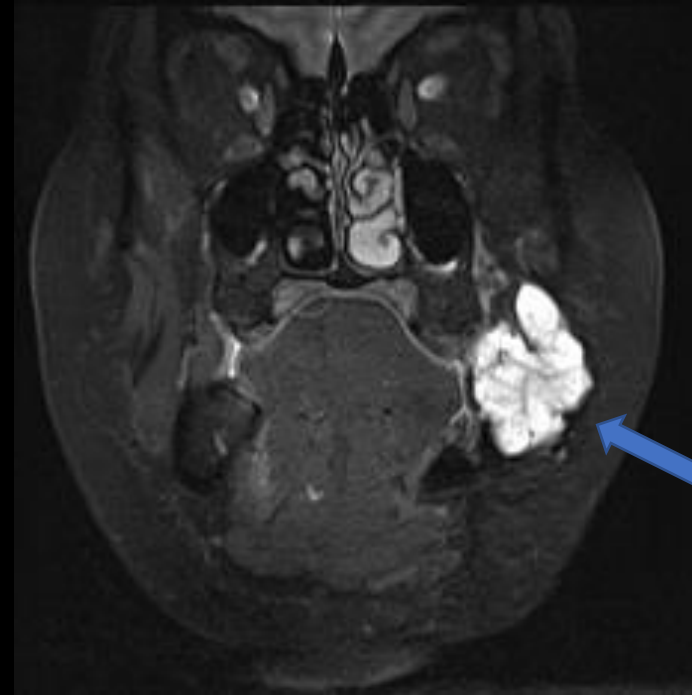
Coronal CT with IV contrast

Red arrow shows heterogenous mass measuring 5.7 x 2.6 x 4.6 cm, positioned within the superficial masticator space on the left in the expected location of the masseter muscle with extension along the mandibular body.

Radiology Images (labeled)



T1 Coronal MRI



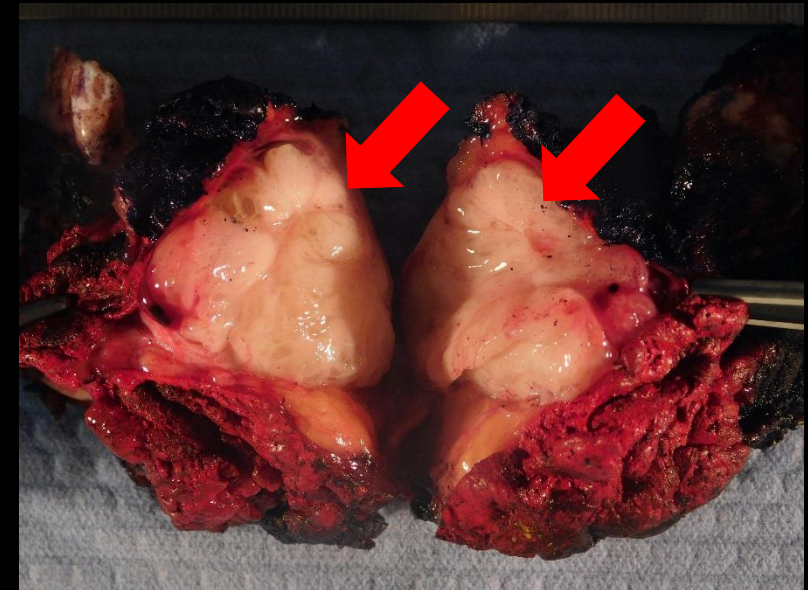
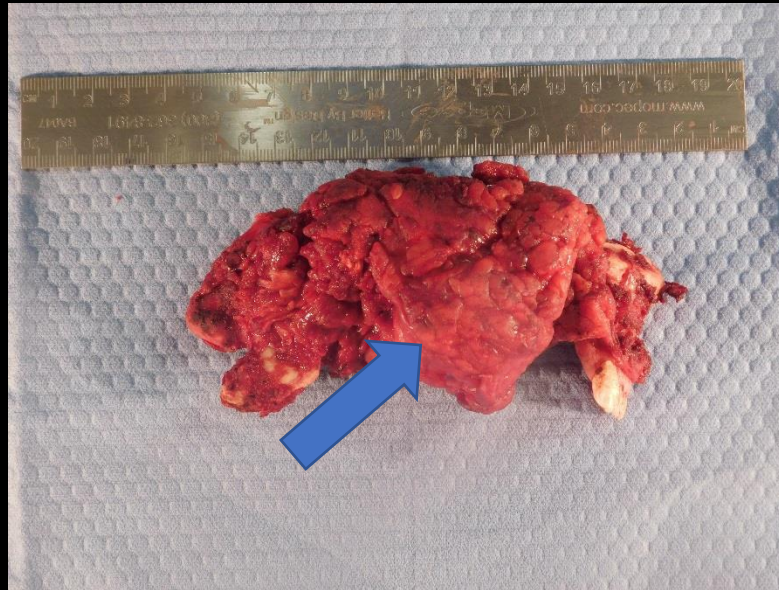
T2 Fat Sat Coronal MRI

Red arrow shows hypointense lesion on T1 weighted imaging. Blue arrow shows multilobulated heterogeneous hyperintense mass on T2 Fat Sat.

DDX (based on imaging)

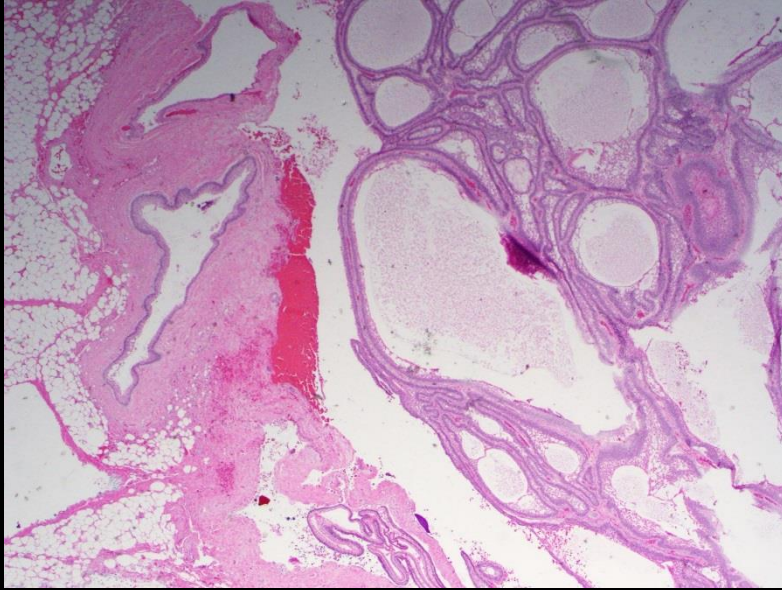
1. Ameloblastoma
2. Dentigerous cyst
3. Odontogenic keratocyst
4. Odontogenic myxoma
5. Aneurysmal bone cyst
6. Fibrous dysplasia
7. Giant cell reparative cysts/granulomas

Gross Path (labeled)

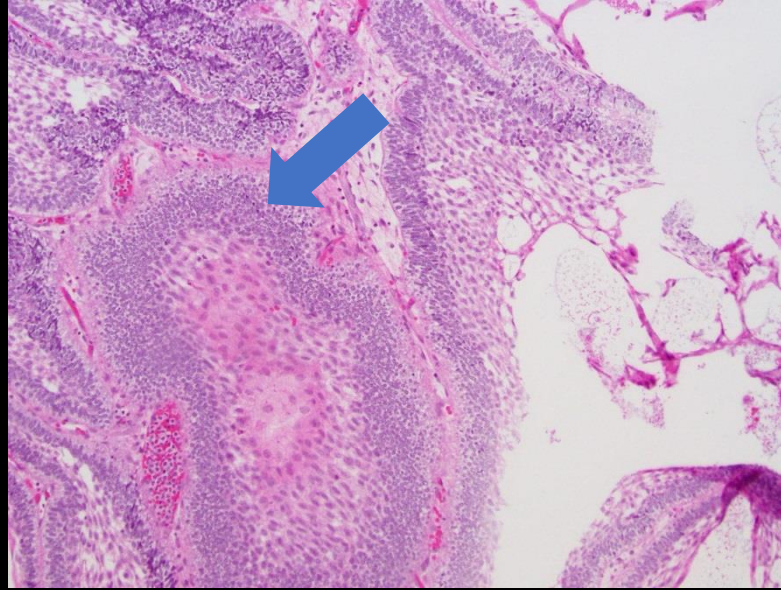


Gross images from above and laterally showing a 7.7 x 0.9 x 0.2 cm firm area along the mandible (Blue arrows). When sliced (Red arrows) the mass consists of lobulated yellow-tan soft tissue with focally cystic areas, measuring 3.6 cm in greatest linear extent.

Micro Path (labeled)



Low power H&E stain showing multilocular cystic spaces



High power H&E stain displaying characteristic palisading of basaloid columnar epithelium (blue arrow)



High power H&E stain showing stellate reticulum (red arrows) encompassed by palisading columnar epithelium

Final Dx:

Recurrent Ameloblastoma of the mandible
Multicystic Conventional

Case Discussion: Characteristics

- Ameloblastoma arises from ameloblasts.
 - Components of the odontogenic epithelium responsible for producing enamel and crown formation.
- A locally aggressive benign tumor that arises from the mandible or maxilla.
- There is equal gender distribution with a mean age of 34 years at first diagnosis.
- Ameloblastoma is the 2nd most common odontogenic tumor
- WHO classifies ameloblastoma into four categories
 - Solid/multicystic, extraosseous/peripheral, desmoplastic, unicystic

Case Discussion: Presentation and Treatment

- Ameloblastoma presents slowly and progressively as hard lesion near the angle of the mandible
 - Can cause dysphagia and neuropathies
 - Often painful as mass grows
- It is a locally aggressive neoplasm with a high rate of recurrence.
- The mainstay of treatment is surgical resection of the tumor and local bone
 - More conservative treatment can be accomplished with BRAF inhibitors
 - However, non-surgical treatment has a 3.15 x higher rate of recurrence

Case Discussion: Radiographic Findings

- CT findings
 - Multicystic ameloblastoma
 - Classic “soap bubble” lesion
 - Well demarcated border with no matrix calcification
 - Larger lesions can erode the nearby bone
 - Unicystic ameloblastoma
 - Well demarcated unilocular lesions
 - Unable to distinguish from other unilocular lesions
- MRI Findings
 - Have a mixed solid and cystic pattern
 - Thick irregular wall
 - Hyperenhancing allowing differentiation from lucent lesions of the mandible

Case Discussion: Histologic Characteristics

- Gross Characteristics
 - Solid and cystic
 - Can be both intraosseous and extraosseous
- Microscopic Characteristics
 - Odontogenic epithelial islands with possible macrocystic degeneration
 - Peripheral palisading columnar cells with frequent reverse polarization of the nuclei and subnuclear vacuolization
 - Cellular stellate reticulum surrounded by the palisading columnar cells
 - The subtypes of ameloblastoma are determined histologically but have no formal impact in prognosis or treatment
 - Follicular, Acanthomatous, Plexiform, Granular cell, and Basaloid
 - These are histologically diagnostic tumors, however, immunohistochemical stains for cytokeratins CK14 and CK13 can be used to highlight the dual population of cells.

References:

1. OA Effiom, OM Ogundana, AO Akinshipo, SO Akintoye. Ameloblastoma: current etiopathological concepts and management. *Oral Diseases* (2018) 24, 307– 316 doi:10.1111/odi.12646
1. Hendra FN, Van Cann EM, Helder MN, Ruslin M, de Visscher JG, Forouzanfar T, de Vet HCW. Global incidence and profile of ameloblastoma: A systematic review and meta-analysis. *Oral Dis.* 2020; 26: 12-21. doi:10.1111/odi.13031
2. Troiano G, Dioguardi M, Cocco A, Laino L, Cervino G, Cicciu M, Ciavarella D, Lo Muzio L. Conservative vs Radical Approach for the Treatment of Solid/Multicystic Ameloblastoma: A Systematic Review and Meta-analysis of the Last Decade. *Oral Health Prev Dent.* 2017; 15: 421-426. doi:10.3290/j.ohpd.a38732
3. Almeida Rde A, Andrade ES, Barbalho JC, Vajgel A, Vasconcelos BC. Recurrence rate following treatment for primary multicystic ameloblastoma: systematic review and meta-analysis. *Int J Oral Maxillofac Surg.* 2016 Mar 45(3): 359-67. doi:10.1016/j.ijom.2015.12.016.
4. ACR appropriateness criteria. <https://acsearch.acr.org/docs/69504/Narrative/>
5. Magliocca K, Martinez A. Mandible and Maxilla Benign Tumors/Tumor like Conditions Ameloblastoma. PathologyOutlines.com website. <http://www.pathologyoutlines.com/topic/mandiblemaxillaameloblastoma.html>.