CASE REPORT

Giant Ameloblastoma: Radiologic Diagnosis and Treatment

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INTRODUCTION

The ameloblastoma is a benign ectodermal tumor of odontogenic origin and is considered to be the most common epithelial odontogenic neoplasm. It represents only 1% of all tumors or cysts of the jaw[1,2]. About *75%* of ameloblastomas occur in the mandible, especially in the regions of the bicuspids and molars and in the angle of the mandible.

The remaining *25%* of ameloblastomas occur in the maxilla. The tumor usually occurs in the 4th and 5th decades of life and shows no predilection for either sex. Ameloblastoma has been reported to metastasize to the lung, brain, and bone, but metastases remain rare[3,4]. A tendency to local aggressiveness is more common[1], and recurrence is frequent after inappropriate surgery[5].

Accurate preoperative delineation of the boundaries of the tumor is essential for achieving a complete resection. This delineation is best achieved with the complementary use of radiography, computed tomography (CT), magnetic resonance (MR) imaging, and angiography. Three-dimensional reformations and holographic imaging help portray the sectional data so that they can be integrated coherently.

Presentation and clinical course: A 45-year-old man presented with a 2-year history of a painless enlarging mass in his right mandible (Fig 1). Overlying skin appeared normal. There was no evidence of skin discoloration or ulceration. Patient was examined with axial CT and three-dimensional reformations. MR imaging was not used because of the large size of the tumor.

CT demonstrated a large mass centered at the right ramus of the mandible (Fig 3). The mass expanded and almost completely destroyed the right mandibular ramus and angle (Fig 6). The multilocular appearance of the mass was well appreciated on CT as well as multiple fluid-fluid levels could be identified (Fig 3,4). On panoramic coronal reconstruction the mass appears to have spared the ipsilateral temporomandibular joint and shows a soft tissue component in the body of the mandible giving heterogenously enhancement after I/V contrast (Fig 5). Superiorly the mass does not extend to the ipsilateral maxilla, however there is incidental evidence of bilateral maxillary sinusitis. Neither intracranial nor distant metastases were identified.

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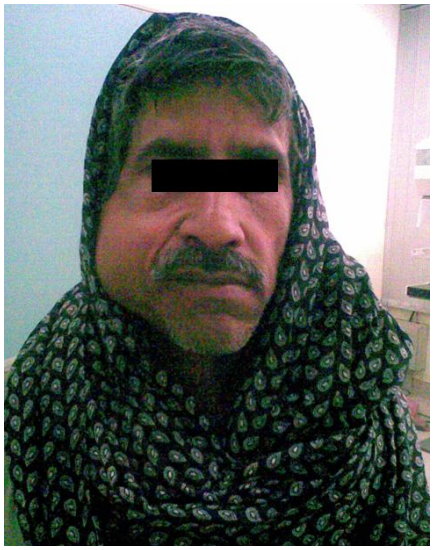

Figure 1: Giant soft tissue swelling on right side of the face, no ulceration or discoloration of overlying skin.

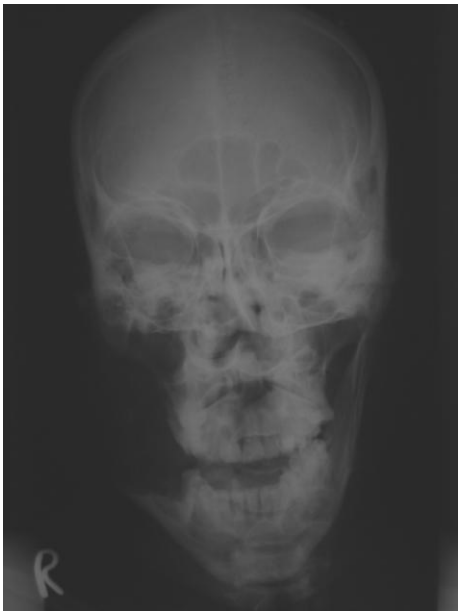

Figure 2: Frontal Radiograph of Face showing expansile lytic lesion involving right hemi mandible, causing thinning and destruction of the overlying cortex.

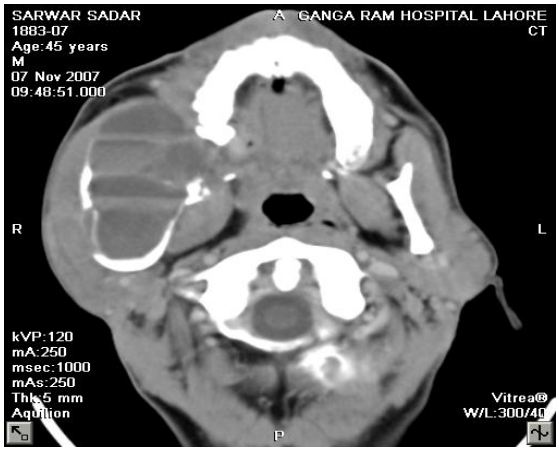

Figure 3: Axial postcontrast CT image showing multilocular cystic mass in the right ramus of mandible with fluid-fluid levels and extensive thinning/destruction of the overlying bony cortex.

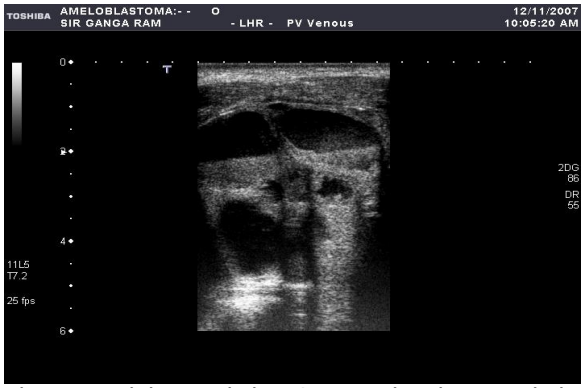

Figure 4: High Resolution Gray scale Ultrasound shows a large multiloculated cystic mass and confirms the finding of fluid-fluid levels.

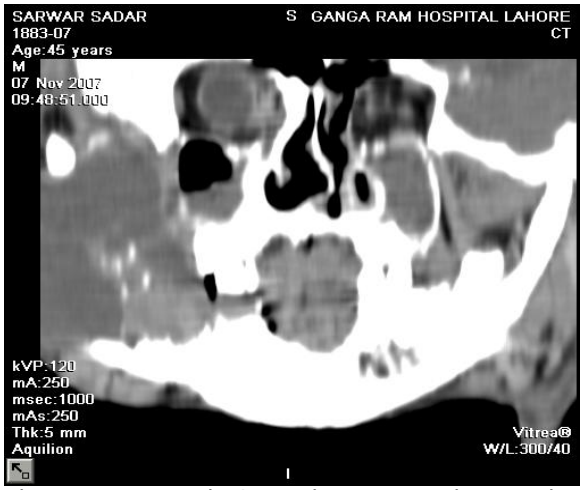

Figure 5: Panoramic Coronal Reconstruction: craniocaudal extent of the tumor is noted, showing that the ipsilateral condylar process is preserved and there is no extension of the mass in the maxillary sinus and orbital cavity. Soft tissue component shows enhancement after IV contrast. Incidental finding of bilateral maxillary sinusitis is seen.

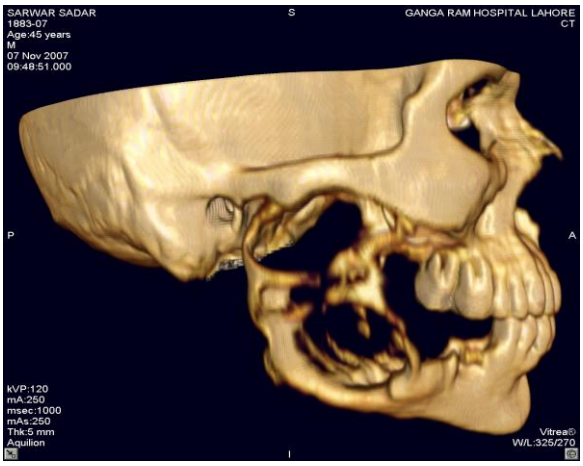

Figure 6: 3-D volume rendered image showing bony destruction of the ramus and angle of mandible.

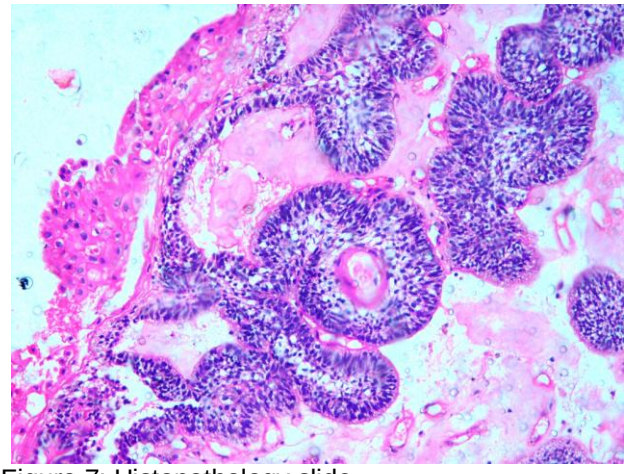

Figure 7: Histopathology slide

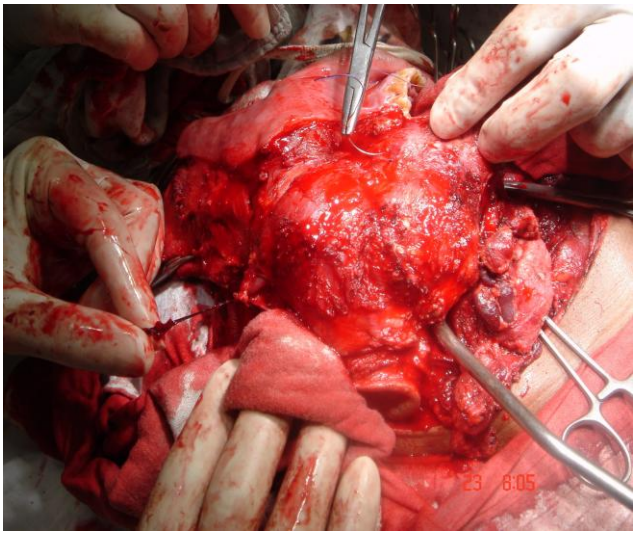

Figure 8: Per-operative photograph

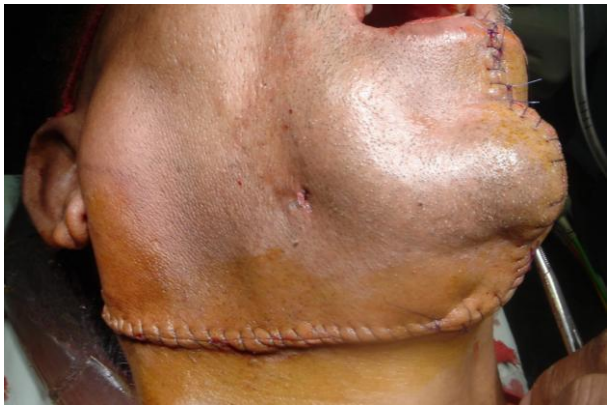
Figure 9

FNAC (Fine needle aspiration cytology) of the tumor was performed under ultrasound guidance however it turned out to be hemorrhagic aspirate and so excisional biopsy was planed. Excisional biopsy was done through per oral approach. On microscopy it showed features of a cellular lesion composed of nests and groups of stellate reticulum, like oval to spindly epithelial cells with slightly pleomorphic nuclei and palisaded basal layer. The intervening stroma reveals degenerated keratin squames and fibrosis. These findings are consistent with diagnosis of keratotic ameloblastoma (Fig 7).

Patient was admitted in surgical ward and underwent right hemi-mandibulectomy with suprahyoid en block lymph node dissection and plastic reconstruction (Fig 8, 9).

DISCUSSION

Cysts and tumors of the jaw are relatively common. Clinical, radiologic, and pathologic features allow the diagnosis to be made. Cysts and tumors (Table) can develop from tooth derivatives (odontogenic) or other tissues (nonodontogenic). Radicular cysts are by far the most common type of odontogenic cyst and are associated with caries. Dentigerous cysts are the next most common cyst, and they vary in size and may cause massive destruction of the jaw[6]. The cyst appears radiologically as a well-demarcated radiolucent lesion attached at an acute angle to the cervical area of an unerupted tooth[13]. The crown of the tooth is contained within the lesion, which was not seen in our case. Unlike ameloblastomas, dentigerous cysts do not exhibit an extracystic soft-tissue mass, but ameloblastomas, mucoepidermoid tumors, and carcinomas may develop on the wall of a dentigerous cyst[6]. Odontogenic keratocysts account for about 10% of cysts of the jaw[6]. Nonodontogenic cysts include the fissural cyst, solitary bone cyst, and static bone cavity. Cysts are usually small and asymptomatic; however, some may become multiloculated and are difficult to distinguish radiologically from an ameloblastoma. Benign odontogenic tumors include the ameloblastoma, calcifying epithelial odontogenic tumor (Pindborg tumor), odontoma, odontogenic myxoma, and cementoma. These tumors represent various histologic subtypes of odontogenic origin (epitheial, mesodermal, and mixed tissues)[6].

Differential Diagnosis of a Lytic Bone Lesion in the Jaw

- Cysts
  - Odontogenic
    - Radicular
    - Dentigerous
    - Odontogenic keratocyst
    - Basal cell nevus syndrome
  - Nonodontogenic
    - Fissural
    - Solitary, simple, or hemorrhagic bone cyst
    - Static bone cavity
- Benign tumors
  - Odontogenic
    - Ameloblastoma
    - Calcifying epithelial odontogenic tumor (Pindborg tumor)
    - Odontoma
    - Odontogenic myxoma
    - Cementoma
  - Nonodontogenic
    - Giant cell granuloma
    - Langerhans cell histiocytosis
    - Fibrous dysplasia
    - Hemangioma
    - Neurogenic tumors
- Malignant tumors
  - Secondary
    - Local invasion
      - Carcinoma from oral cavity
    - Metastatic
      - Lung
      - Breast
      - Kidney
      - Thyroid
  - Primary
    - Odontogenic
      - Central epidermoid carcinoma
    - Nonodontogenic
      - Mucoepidermoid carcinoma
      - Sarcoma (osteogenic, fibrosarcoma, and Ewing)
  - Other
    - Lymphoma, leukemia, and multiple myeloma
- Infection

Table: Differential Diagnosis of a lytic bone lesion in the jaw

Benign nonodontogenic tumors such as giant cell granuloma and Langerhans cell histiocytosis are not unique to the jaw[6] and may be found in other parts of the body. Other lytic benign bone lesions include fibrous dysplasia, hemangioma, and neurogenic tumor. Fibrous dysplasia shows typical ground glass appearance[14] which was not present in

our case. Many of these benign lesions appear radiolucent and have unilocular or multiocular cystic characteristics similar to those of ameloblastoma and can be difficult to differentiate from it when they occur in the jaw. Aneurysmal bone cyst also presents as an expansile multilocular mass with fluid-fluid levels however it is more common in children than adults[15], while in our case the age of the patient was around 45years thus making diagnosis of aneurysmal bone cyst less likely. Osteomyelitis, or infection of both the bone and marrow of the mandible, is rare in healthy individuals due to early administration of antibiotics[16]. Patient presents with history painful swelling and fever. Its radiographic appearance ranges from acute or chronic suppurative to sclerosing osteomyelitis. Acute suppurative osteomyelitis usually demonstrates no imaging findings in the early stages, whereas chronic lesions demonstrate a variety of bone reactions, including radiolucent and radiopaque areas.

Malignant tumors of the jaw include primary and secondary tumors. Carcinomas of the oral cavity that invade the jaw are the most common. Metastases from other sites, most commonly the lung, breast, thyroid, and kidney, are also frequent[6]. Malignant neoplasms that originate in the jaw are rare and include nonodontogenic tumors such as mucoepidermoid carcinoma and various sarcomas and an odontogenic tumor referred to as central epidermoid carcinoma. Other malignancies in the differential diagnosis include leukemia, lymphoma, and multiple myeloma. Malignant tumors can usually be differentiated radiologically from the less aggressive appearing benign tumors, but final diagnosis requires biopsy[6]. Our case of giant ameloblastoma is one of the larger tumors of its kind. The patient had the radiologic and laboratory findings typical of other cases of giant ameloblastoma[7-9]. Ameloblastomas rarely metastasize, but the local recurrence rate is high[1]. Early en bloc surgical resection is the treatment of choice to avoid recurrence. Radiation therapy can be used in addition to or, in certain cases, in place of surgery[10]. There is no effective role for chemotherapy[10]. Prognosis after surgical resection is linked to the method of surgical treatment, the age of the patient, and the cystic characteristics of the tumor. Higher recurrence rates are seen in older patients and in those with tumors that are multilocular or exhibit soap bubble-type cysts. Some studies report that the follicular histologic subtype has a substantially higher recurrence rate than does the plexiform subtype, but the majority of surgeons and pathologists believe that there is no correlation between the histologic pattern of the tumor and its prognosis[11]. Ucno et al[11] reported that the average interval between initial treatment and recurrence was 2.7 years and that 78.8% of cases recurred within *5* years.

Effective resection of an ameloblastoma depends on an appropriate three-dimensional understanding of the extent of the tumor[12]. Traditionally, this is derived from radiography, axial CT, multiplanar MR imaging, and angiography. When planning for surgery, especially of the head and neck, we have successfully used helical CT with three-dimensional reformations and holograms. These techniques provide valuable insight for the surgeon when considering the primary abnormality and the surgical approach needed. Use of helical CT with three-dimensional reformations and holograms was especially beneficial in the preoperative approach to this case of giant ameloblastoma because of its extensive bone involvement.

CONCLUSIONS

Thorough preoperative evaluation and an appropriate surgical approach are crucial to achieving a good prognostic outcome when treating giant ameloblastoma. Although benign, these rare tumors produce high morbidity with their extensive local destruction and their high rate of local recurrence following resection[1]. With the successful outcome that can be achieved by combining traditional imaging modalities and more recent advances in three-dimensional imaging, the radiologist plays a pivotal role in the diagnosis and management of these tumors.

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