Chapter 10

Jaw Tumors

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Introduction

Tooth development begins at 37 days of IU life with formation of a continuous horseshoe-band of thickened epithelium in the location of upper and lower jaws, the dental lamina. Each dental lamina develops 10 centers of proliferation from which tooth buds grow.

Tooth buds for the permanent teeth that have deciduous predecessors appear at 10 weeks, from deep continuations of the dental laminae, while the rest appear at different times. These laminae mature into enamel organs, one for each primary and permanent tooth. Cells of the enamel organ differentiate into ameloblasts and synthesize enamel.

The ameloblasts stimulate the dental papillary cells to differentiate into odontoblasts, which later synthesize dentin.

History

In the history of jaw tumors, one may elicit any of the following:

Pain, loose teeth, recent occlusal problems, delayed tooth eruption, swellings, dysesthesias or intraoral bleeding. There is often a history of a recent tooth extraction, with a resultant non-healing ulcer. Paresthesias, trismus, and malocclusion may indicate a malignant process if recent in onset. In the sub-Saharan African patient, this may just be the result of a neglected benign lesion. The onset and course of the growth rate of the mass is important; recent history of rapid growth after a long history suggests a malignant degeneration, for example.

General Head and Neck examination

Carefully examine the affected part of the jaw and overlying dentition, noting ulceration, degree of jaw involvement, and any nodal involvement. Palpation will differentiate soft tissue masses from bony lesions.

Investigation(s)
Radiologic examination of the mass is the investigation of choice in the evaluation of jaw related cyst and tumors. The type of examination is determined by availability of facilities, and the ability of the patient to pay for the test.

An Orthopantomogram (OPG)/Panorex will often confirm clinical suspicions and even allow the formulation of differential diagnoses – this however requires special equipment that may not be universally available. Skull radiographs may be more universally available, while chest radiographs are useful where malignancy is suspected.

Where available and affordable, CT Scans are excellent for delineation of jaw tumors: they help narrow down the differentials, and are useful for planning the extent surgical excision and reconstruction.

As a general rule, well-demarcated lesions outlined by sclerotic borders suggest benignity; aggressive tumors have a tendency to show up as ill-defined lytic lesions. With larger more aggressive lesions, CT scan may more clearly identify bony erosion and/or invasion into adjacent soft tissues.

Large tumors do not necessarily mean malignant lesions. They do, however make both the resection and reconstruction more difficult.

Where possible, tissue is obtained for histologic identification of the lesion; this may be done under local anesthesia, or may require a general anesthetic, depending on the ease of accessibility and the safety of the airway.

As far as possible, trans-oral biopsies are preferred. It is important for the biopsy scar site to fall within the anticipated tissue to be excised. Fine needle aspiration is excellent for ruling out vascular lesions prior to open biopsy and may be helpful in helping diagnose inflammatory or secondarily infected lesions.

The term, ‘jaw tumors,’ is an expansive and all-inclusive term. It includes a large number of different tumors, both benign and malignant. This chapter will only deal with tumors that patients present with, with relative frequency, in the context of sub-Saharan Africa.

The classification of jaw tumors is quite complex, but is significantly abbreviated here, in order to cover the commonly seen lesions:
<table>
<thead>
<tr>
<th>Benign Tumors</th>
<th>Jaw Classification</th>
<th>Tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Odontogenic Cysts</td>
<td>Dentigerous Cyst</td>
<td></td>
</tr>
<tr>
<td>Epithelial odontogenic tumor</td>
<td>Ameloblastoma</td>
<td></td>
</tr>
<tr>
<td>Other jaw lesions</td>
<td>Fibrous dysplasia</td>
<td></td>
</tr>
<tr>
<td>Malignant jaw tumors</td>
<td>Osteoclastic tumors</td>
<td>Squamous cell carcinoma</td>
</tr>
<tr>
<td>Osteoclastic/osteoblastic tumors</td>
<td>Osteosarcoma</td>
<td></td>
</tr>
</tbody>
</table>

**Dentigerous cysts**

Dentigerous cysts may constitute up to 24% of all developmental cysts. They are commonly located in the mandibular 3rd molars, maxillary canines, and maxillary 3rd molars.

They occur most commonly in the second to the fourth decades, and are largely symptomatic. Large cysts can however cause the displacement or resorption of adjacent teeth. They may also cause malocclusion, pain, and even interfere with respiration.

![Fig 1](image1.png)
Dentigerous Cyst (cystic area at end of red arrow)

![Fig 2](image2.png)
Ameloblastomas

Ameloblastomas are the most common odontogenic tumors. They are benign, but locally aggressive tumors, with the potential to grow to enormous sizes, resulting in severe bony/facial deformity.

Ameloblastomas may originate from any epithelium within the jaws that is involved with the formation of teeth. Although ameloblastomas may arise from the lining of a dentigerous cyst, they more often arise independently of impacted teeth.

In Western series, these tumors have no gender predilection and occur in adults, with a mean age 40 years.

Radiographs may reveal well-circumscribed, expansile soap-bubble radiolucency (either unilocular or multilocular) with clearly demarcated borders. The lesions may be unilocular or multilocular: unilocular lesions are indistinguishable from
an odontogenic cyst, hence the need for a biopsy because of the differences in management of the two lesions. Clinicopathologic subtypes include the multilocular and multicystic/solid intraosseous, uni-cystic intraosseous and peripheral/extraosseous subtypes, constituting 86%, 13% and 1% respectively, of ameloblastomas. The solid (multicystic) subtype is the most aggressive of the three, requiring more aggressive treatment. Recurrence rates of 50 to 90% have been reported when curettage was the means of treatment.

♦ Clinical presentation
Ameloblastomas are slow growing, locally invasive odontogenic tumors of the jaws with a high rate of recurrence. Ameloblastomas frequently grow to enormous sizes, especially in the patient population in sub-Saharan Africa. The mandible is more frequently involved than the maxilla.

Ameloblastomas have a small risk of metastasis, usually to the lungs: ameloblastomas that behave in this manner have been called metastatic ameloblastomas. Histologically, these are indistinguishable from other ameloblastomas. Ameloblastic carcinomas have on the other hand, microscopic characteristics of malignant tumors.
Early symptoms are often absent, while late symptoms may include a painless swelling, loose teeth, malocclusion, fistula formation or nasal obstruction. Maxillary tumors may extend into the antrum, and thereafter grow into the nasal cavity, ethmoid sinuses, and skull base.

♦ Management

Anesthesia is a challenge in these cases. Often the surgery is relatively easy. The difficult part of the operation is securing a good airway for the case. In many locations this will not be possible and the patient must be referred to a large center. A tracheostomy may always be done when the intubation is difficult or impossible but postop care of the tracheostomy may be difficult. In the small masses an oral endotracheal tube may be inserted. Sometimes it may be possible to insert a nasotracheal tube. In ideal situations a fiberoptic bronchoscope is available and it is used to safely intubate the patient through the nose. It is to be hoped that after the mass is removed, the patient will not require intubation but with the large masses, it may not be wise to remove a tube for the first 12 hours. Suction must be available in these cases. One of the authors frequently uses one dose of an intravenous steroid as Solu Medrol before starting large head and neck cases with the expectation that this will reduce swelling. Several doses may also be given in the first 24 hours. There are other ingenious ways to intubate such a patient but the above methods are the safest. It is to be emphasized again that the surgical resection is the easiest part of the care of this patient. Anesthesia and postoperative care are most important.

Some use a GlideScope to help with intubation. This is a laryngoscope attached to a small monitor that may be watched while one attempts intubation. This allows for better visualization during intubation. A GlideScope is not cheap but would help less experienced anesthesia personnel with intubation. One may Google GlideScope for further information.
The incision for mandibular resections is a submandibular incision with a midline split of the lower lip. The incision can extend to the angles of the mandible. In ameloblastomas, much of the expanded mucosa may be saved and closure is relatively easy. Xylocaine ½ % with adrenalin is used to infiltrate the skin and mucosa before the incision to help reduce the bleeding. Care must be taken to place the incision below the mandibular branch of the facial nerve and elevate the skin below the platysma muscle. The facial vein can be identified, ligated and elevated with the skin flap to help protect the nerve.

The treatment of choice is resection of the involved mandible or maxilla, with surgical margins of 2.0 cm or 1.5 cm respectively, of the mandible or maxilla. Extension of tumor into surrounding soft tissues requires additional soft tissue wide resection. Postoperative follow-up is recommended, to monitor for recurrence and plan treatment accordingly.

Reconstructive challenges after resection of parts of the mandible or maxilla

♦ Small bone defects of 4 cm or less: may be reconstructed with a steel/titanium plate and bone from iliac crest, rib or best calvarium

♦ Defects larger than 4-6 cm: should ideally be reconstructed using vascularized bone – either as a pedicled flap as pectoralis flap with rib or as a free flap as fibula or scapula. While the former requires some training with a good understanding of the anatomy and vascular patterns around the ribs or scapula, the latter requires both specialized equipment, ideally an operating microscope and microvascular surgical training.

♦ In most of these cases with large tumors and defects, reconstruction using a plate of steel or titanium, will be an acceptable option. Some body, ramus, angle and condylar defects can be ignored, and although these will result in cosmetically unacceptable defects, the patients will be functional with the mandible loss and even with an “Andy Gump” deformity.

♦ Where the entire hemimandible including the condyle may need to be removed and one does not have microsurgery capabilities, one may reconstruct the defect with a condylar plate which extends up into the TMJ. If such a plate is unavailable, a titanium plate can be used to reconstruct the ramus, angle and body and a large non-absorbable suture can used to suspend the plate from the zygomatic arch. At least 3 screws must be placed into the mandible on the opposite side. The disadvantages of plate reconstructive include hardware failure and exposure/extrusion
When one does not have microvascular capability, plate reconstruction can first be done as described above. Then a few months later (3-6) tightly packed iliac crest cancellous bone grafts can be place along the entire length of the plate. This is done via an external approach, not entering the oral cavity and will permit bone reconstitution within the defect over time, with minimum risks of bone graft infection.

Hardware failure or exposure necessitates revision; vascularized bone would be preferable, but the realities of practice in some environments would only permit the use of a flap to cover the exposed hardware/bone. Most likely the extrusion of a metal plate will occur in the anterior midline. Flaps used are the deltopectoral, pectoralis major, or supraclavicular perforator flap. See the Flap chapter.

**Fibrous dysplasia**

Fibrous dysplasia (FD) is a benign tumor in which normal bone is replaced by fibrous connective tissue resulting from a defect in osteoblast differentiation and maturation. Fibrous dysplasia may affect a single bone (monostotic type) or multiple bones (polyostotic). Fibrous dysplasia may also occur as part of the McCune-Albright syndrome, consisting of polyostotic fibrous dysplasia, café-au-lait spots and an endocrinopathy (hyperthyroidism or precocious puberty in females).

Monostotic FD is the most common type; 70 – 80% is found in the long bones or ribs. The head and neck region is involved in 25% of cases. Polyostotic FD is less common, found in 15% of all FD; 50% of the cases involve the head and neck region.
Clinical presentation

Fibrous dysplasia in the jaws presents as a painless mass, most commonly in the maxilla. The lesion usually does not cross the midline and is usually limited to the bone involved, hence causing asymmetry. The antrum is often obliterated, and the orbital floor may be involved, leading to displacement of the eye.

Radiologically, FD may be radiolucent, or reveal a mixture of radiolucency and radio-opacity, with a ‘ground-glass’ appearance. The border of the tumor is ill-defined, merging with normal bone; when well defined, it is difficult even on microscopy, to differentiate it from ossifying fibroma.

FD usually presents in childhood or early adolescence, during the period of greatest skeletal growth. FD may have exacerbations, may occur during pregnancy and with oral contraceptive use, and usually stops growing with cessation of growth of the individual in late adolescence, although there are exceptions to this.

FD has no sex predilection.

Massive cranial bony involvement, may lead to a lion-like facies, ‘leontiasis ossea’ or bilateral maxillary involvement called ‘cherubism.’ It may also occur in the skull.
Fig 19
Fibrous dysplasia in maxilla: required near total maxillectomy and bone grafts

Fig 20
Fig 21
Fig 22
Fibrous dysplasia in skull and orbital roof: If symptomatic, it will require the expertise of a neurosurgeon and plastic surgeon working together

Treatment

Treatment is determined by the following variables:

- The age of the patient,
- The rate of tumor growth,
- The location and extent of the lesion,
- Degree of cosmetic deformity, and
- The resulting functional impairment.

There are three approaches to the treatment of FD: observation, conservative surgery - sculpturing/contouring (surgical debulking) the lesion to as normal a shape and size as possible with preservation of alveolar ridge and teeth and radical surgical excision and reconstruction. Surgical intervention is aimed at
correcting or preventing functional deficits, and the restoration of as near normal facial cosmesis as possible. Radical excision is most appropriate for extensive mandibular involvement where tumor debulking is impractical, and reconstruction a viable option.

Surgical debulking involves significant blood loss. Bone wax is a very handy hemostatic agent, as the bleeding is frequently unresponsive to cauterization. The administration of calcitonin preoperatively has been reported to reduce operative bleeding, as well as aid in bone remodeling. Pamidronate has also been used to treat pain and pathological fractures from lesions in long bones.

Where possible, treatment should be deferred for as long as possible, especially until skeletal maturity. Children with FD require long term follow-up. When the patient is near the end of physical growth, debulking with preservation of teeth may be carried out. Younger patients with large masses may also undergo surgical debulking with preservation of teeth but close followup is necessary and second stages will likely be necessary.

There is a small (less than 1%) risk of malignant transformation into osteogenic sarcoma, and therefore, accelerated growth or aggressive lesions will require early surgical intervention with en bloc resection and reconstruction. Radiation therapy has been reported to cause malignant transformation of FD, and is therefore contraindicated.

♦ Technique

Many cases of FD are in the maxilla and require a hemimaxillectomy. The maxilla is approached through a Weber-Ferguson incision which extends through the upper lip, around the nose and just below the lower eyelid. If possible the inferior orbital rim is left with a small portion of the maxilla as well as zygoma. In very large tumors, the entire rim must be taken. Care is taken to preserve the suspensory ligaments of the orbit. If necessary the rim can be reconstructed with calvarium.

Cementoblastoma

Cementoblastoma is another uncommon benign jaw tumor that usually occurs in people under the age of 25 and often involving the molars or premolars. It forms an irregular or round mass attached to the root of a tooth. On x-ray it is a well-defined radiopaque mass. Usually it is small but when consultation is delayed, it may grow quite large. It is very similar in appearance to ossifying
fibroma and osteoblastoma. Resection is the only treatment and reconstruction is similar to the reconstruction for ameloblastoma.

**Osteogenic sarcoma**

Is a malignant tumor of bone, and is the most common primary malignancy of bone (apart from myeloma). Only 5–10% of these tumors occur in head and neck, primarily in the jaws. They are usually painless swelling in jaws, although pain and paresthesia may result from nerve involvement or compression. Osteosarcoma of the jaws may present at any age, but peaks in the fourth decade.

Some reports suggest a slight male predilection, with mandible predominance. Jaw osteosarcomas are less likely to metastasize, but unfortunately, prognosis has not improved with the use of chemotherapy as with osteosarcomas of long bones. The main cause of death in jaw osteosarcomas is local recurrence.

Radiologically, tumors may exhibit the classic ‘sunburst’ appearance, best seen on CT images. Lesions may otherwise exhibit ill-defined bone edges, with radiolucency or a combination of radiolucent and radiopaque pattern.

**Burkitt’s lymphoma**

This is not a surgical disease, but is included in this text for completeness’ sake. This tumor may present as a jaw tumor, and lack of familiarity with the pathology may lead to unnecessary surgical intervention. It may present as any of the three variants; the endemic, sporadic and immunodeficiency (HIV) associated types. The endemic and immunodeficiency-associated variants are relevant to the surgeon working in sub-Saharan Africa.
A fine needle aspirate is sufficient to provide a diagnosis, while chemotherapy is often rapidly effective in controlling and decreasing the size of the tumor. Cure may be possible if the tumor is caught early.

**Rhabdomyosarcoma**

Rhabdomyosarcoma is the most common soft tissue sarcoma in children under 15 years of age except in some parts of Africa where Burkitt’s lymphoma is more common. Biopsy must be taken to make an accurate diagnosis. Whereas ameloblastoma is can be both a benign and malignant odontogenic tumor, Burkitt’s and rhabdomyosarcoma are nonodontogenic malignant tumors. Burkitt’s can be treated by high doses of chemotherapy, so it is important to get an accurate tissue diagnosis.
Squamous cell carcinoma

Squamous cell carcinoma is the most common malignancy of the oral cavity. It has a higher prevalence in certain regions because of cultural practices e.g. betel nut chewing, inverse smoking, etc. It afflicts more males than females, and is most frequently found in the tongue and floor of mouth, but can arise in any parts of the oral cavity. It may metastasize to regional lymph nodes, leading to a poor prognosis.

A number are recognized after a tooth extraction because of a non-healing ulcer.

In radiographs, bone radiolucency, with a ‘moth-eaten’ appearance adjacent to the soft tissue mass, is indicative of bone involvement. Most squamous carcinomas found in jaws have invaded from lesions of the oral cavity, but primary intraosseous carcinoma may arise within the jaw presumably from residues of odontogenic epithelium. Squamous cell carcinoma has also been reported occurring as a Marjolin’s ulcer in an orocutaneous ulcer in a large mandibular ameloblastoma.

Surgical resection with adequate margins may be sufficient treatment for early lesions. Late lesions amenable to surgical therapy will need wide excision and neck node dissection, with or without radiotherapy. Surgical excision is the most durable form of treatment available for most patients in sub-Saharan Africa, as radiotherapy may not be as easily accessible. For lesions that are not amenable to surgery, radiotherapy if accessible in a timely fashion, may downgrade a tumor, and make surgery feasible. Various flaps may be used to reconstruct defects including deltopectoral and pectoralis myo/osteocutaneous flaps.

Microvascular Reconstruction:

In the rare case where microvascular reconstruction is possible this is an example of what can be done with a fibula bone graft.
A titanium plate is used and the bone graft is anchored to the plate before the anastomosis if performed between the peroneal blood vessels and the major vessels in the neck.

Another microvascular case: (courtesy of Dr. David Chang)

Fibula reconstruction (postoperative, same case as above)
This is tedious surgery and requires microvascular instruments, loupes, a microscope and someone skilled in microvascular surgery. In addition, since these cases are long, good anesthesia and a postop ICU is necessary.

Surgery of the jaws requires histopathology in most cases. Ameloblastoma may be obvious with a cystic mandibular lesion in a young adult. Otherwise it is best to have a pathological report before beginning extensive surgery. On the other hand, some may be in remote areas where you will have to proceed based on your experience.