Ameloblastic carcinoma in a 3-year-old: Case report and review of the literature

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Introduction
Epithelial malignancies of the odontogenic osseous structures are rare. Ameloblastic carcinoma is an aggressive tumor with a very poor prognosis. Less than 45 tumors of this type have been reported in the literature.1

Many times the tumors arise from prior ameloblastomas and should be suspected when previously resected ameloblastomas recur rapidly.2

The most common site is the posterior portion of the mandible. The most common site of distant metastasis is the lung, followed by bone, liver, and brain.2 Distant metastases can occur even without regional disease.6

Signs and Symptoms
The average age at presentation is 30-50 years-old, with ranges from 15-84 reported.2

Most patients present with a rapidly-enlarging mass in the mandible or maxilla and swelling of the facial soft tissues.2 Other symptoms can include pain, trismus, rhinorrhea, dysphagia, and dysphonia. The mass is often firm, well-circumscribed, and can ulcerate or soft tissue destruction.2

A 3-year-old boy presented to a local pediatric plastic surgeon with a 3 month history of rapid right-sided facial swelling. An incisional biopsy revealed ameloblastic carcinoma. He underwent a hemimandibulectomy and level I-III selective neck dissection. Reconstruction was performed with a condylar prosthesis and reconstruction bar covered with a radial forearm free tissue transfer.

Final pathology revealed negative bony margins with 0/7 lymph nodes involved. The primary tumor demonstrated lymphovascular invasion. Biopsies of the inferior alveolar nerve were negative.

His post-operative course was complicated by wound dehiscence and infection requiring debridement. He did well following debridement and intravenous antibiotics. A tracheotomy that was placed was removed 3 months after initial surgery. Unfortunately, 4 months after primary resection, he developed recurrent right-sided facial swelling. He was treated with platinum-based chemotherapy for four cycles, and radiotherapy to 68Gy.

Less than three months after completing chemoradiotherapy, the patient died from an infection associated with residual disease.

Computed tomography demonstrates destruction and expansion of normal bony with bony remodeling.6

Evaluation of the chest with CT or PET-CT is indicated due to the high rate of lung metastases.1 Also, metastases from the lung, breast, and GI tract must be ruled-out.1-4

Guidelines for treatment do not exist, but most ameloblastic carcinomas have been treated with surgical resection and neck dissection, followed by adjuvant chemotherapy and radiotherapy.

Diagnosis and Management
A high index of suspicion must exist when a patient presents with a rapidly-enlarging mandible or maxilla mass. Occasionally, salivary gland neoplasms can mimic ameloblastic carcinoma in presentation and histology.

Diagnosis begins with a thorough history and physical examination, with particular attention to the bony and soft tissues of the oral cavity and neck.

Radiographs and panorex films are helpful and often show radiolucent changes in the bone with root resorption.1

Computed tomography demonstrates destruction and expansion of normal bony with bony remodeling6

Radiotherapy is debatable, but can improve the likelihood of local control, especially with positive surgical margins.6 Chemotherapy often includes platinum-based agents, like cisplatin, with doxorubicin added if deemed clinically feasible.

Pathology

High-powered 40x H&E stained tissue showing follicles with palisading arrangement of cuboidal cells and disorderly arrangement1.

Tumor cells showing marked cellular atypia with necrosis (asterisk). No longer showing typical follicular arrangement.5

Adjuvant Therapy
Radiotherapy is debatable, but can improve the likelihood of local control, especially with positive surgical margins. Chemotherapy often includes platinum-based agents, like cisplatin, with doxorubicin added if deemed clinically feasible.

Conclusion
We present a case report of a 3-year-old boy with ameloblastic carcinoma and review of the literature. This patient was treated aggressively, but did not survive. Ameloblastic carcinoma is a rare and extremely aggressive odontogenic tumor of epithelial origin. It can arise from ameloblastomas, but often is a primary tumor. Treatment involves aggressive surgical resection with negative margins, free flap reconstruction and post-operative chemo-radiotherapy. Despite this extensive treatment, the prognosis is very poor, with average survival less than twelve months. Early diagnosis and management is essential for minimizing symptoms and maximizing quality of life.

References