

Odontogenic Carcinoma Arising from Benign Odontogenic Cysts: Report of Two Cases, Management, and Review of the Literature

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ABSTRACT

Introduction: Odontogenic carcinomas arise either within a longstanding benign cyst that exhibits a rapid change in clinical behavior, or, de novo from a subclinical cyst. They are exceedingly rare.

Methods: The presentation, diagnosis, treatment, and prognosis for odontogenic carcinoma arising within odontogenic cysts was reviewed in the context of a case series. Two patients presented with rapidly enlarging masses of the maxilla (1), and, mandible(1). A review of English language case reports of such lesions was performed with PubMed and OvidWeb searches and is also presented as a context for the individual cases.

Results: Two cases met inclusion criteria for this report. The first carcinoma arose from an existing odontogenic keratocyst of the mandible and was pathologically consistent with odontogenic carcinoma. The second carcinoma was classified as an odontogenic ghost cell tumor (OGCT) of the maxilla arising from a mixed benign lesion with components of calcifying odontogenic cyst and compound odontoma. The management in both cases was composite resection with wide margins; Neck dissection was performed for the mandibular tumor, but withheld in the maxillary tumor. Radiation therapy was delivered to the maxilla and neck in an adjuvant fashion for the OGCT; this patient remains disease free at 29 months.

Conclusions: Odontogenic carcinoma from benign cysts are a rare occurrence best managed with surgery and reconstruction, followed by radiation therapy for concerning pathologic features or close margins, and, in certain pathologies thought to exhibit aggressive behavior; Applications of radiation therapy are extrapolated from experience with squamous carcinomas. Primary surgery for odontogenic carcinoma should include composite resection with or without neck dissection depending on location of the primary with immediate reconstruction.

INTRODUCTION

Few cases of odontogenic carcinoma have been reported, therefore, the presentation, natural history, and outcomes of such lesions are poorly understood. Tumors arise from pre-existing odontogenic cysts or benign tumors, and therefore undergo malignant transformation of the same cell-types that are features of the benign cyst of origin. Benign odontogenic cysts are broadly classified into developmental, inflammatory, or nonepithelial cysts. Tumors are generally grouped by the cell of origin; epithelial, mesenchymal, and unknown.

Odontogenic carcinomas arise in only 0.6-1% of all odontogenic tumors and include ameloblastic carcinoma, ghost-cell carcinoma, and odontogenic sarcoma, according to the WHO classification system.¹ Due to the rarity of such tumors, diagnosing them, particularly when arising from long-standing benign cysts, can be challenging. As a whole, these tumors have proven exceedingly rare with only a handful of reported cases and poor understanding of the continuum between benign and malignant. Occurrences of both pathologies encountered in this report are difficult to tabulate in the literature owing to their rarity and multiple nomenclatures. The two other benign cyst or tumor and malignant counterparts are included in the table below for completeness. As indicated, all reported incidences of these tumors still represent a small number of cases.

Benign Odontogenic Cyst/Tumor	Malignant Counterpart	Cases
Ameloblastoma	Ameloblastic Carcinoma	Rare – 37
Calcifying Epithelial Odontogenic Tumor (Pindborg Tumor)	Malignant CEOT	Very Rare – 3
Calcifying Odontogenic Cyst (Gorlin's cyst)	Odontogenic Ghost Cell Carcinoma	Rare – 31
Odontogenic Keratocyst	Primary Intraosseous Squamous Carcinoma	Rare – 40

FIGURES



Figure 1. Physical exam appearance

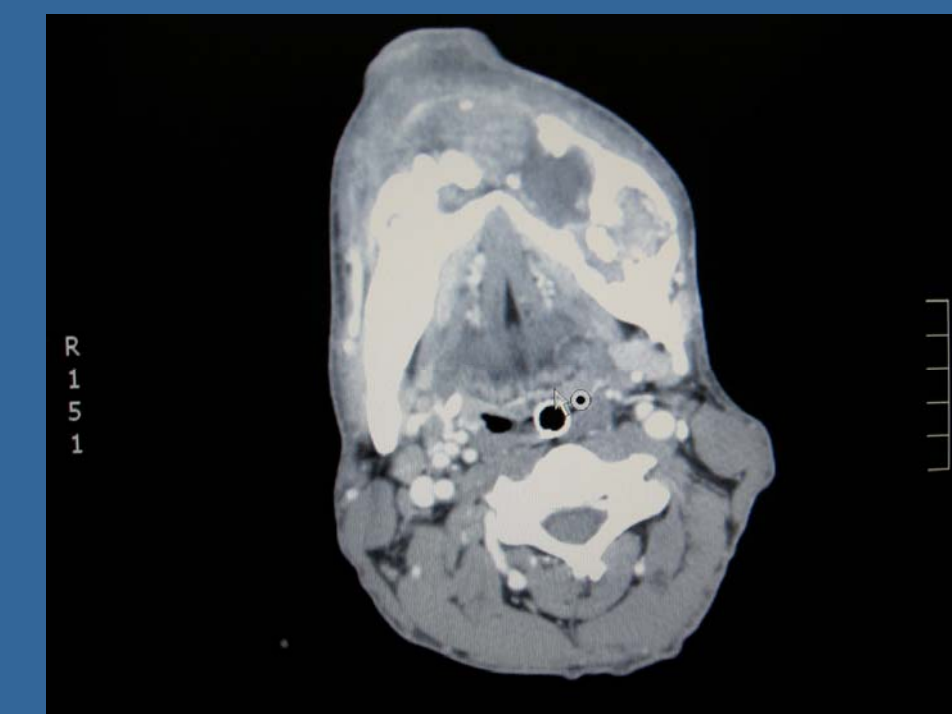


Figure 2. Radiographic appearance

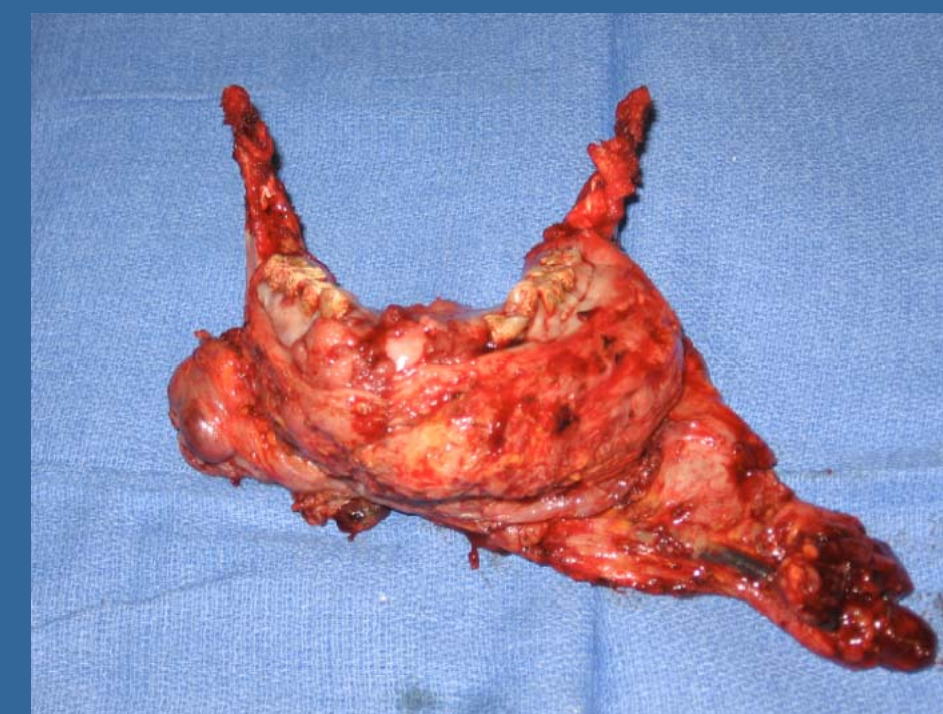


Figure 3. Resected tumor



Figure 4. Harvested fibula

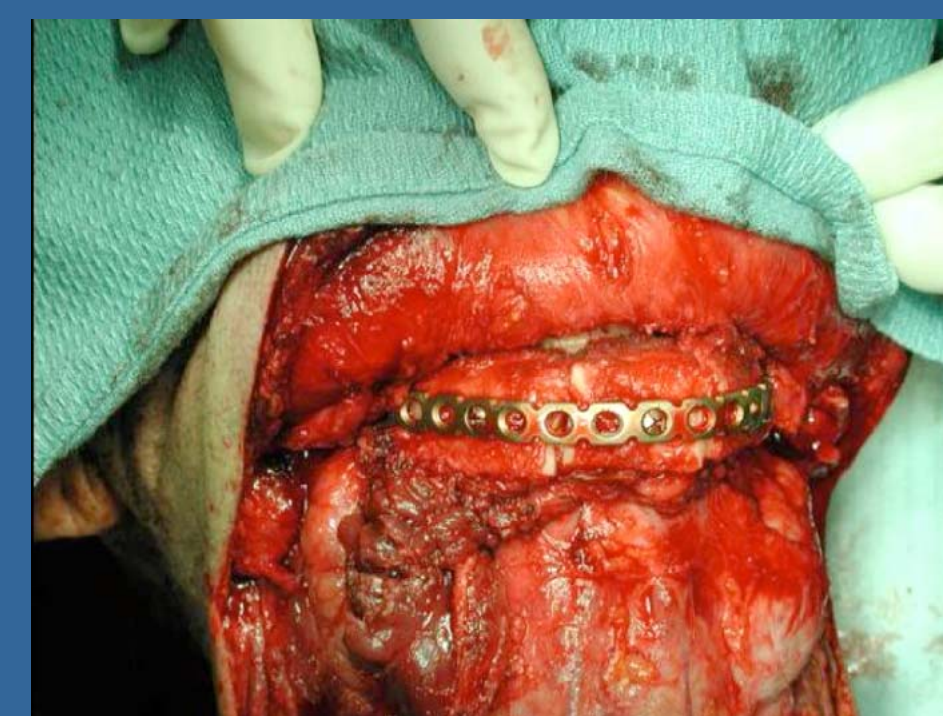


Figure 5. Reconstruction with fibula



Figure 6. Final appearance with fibula

METHODS AND MATERIALS

The presentation, diagnosis, treatment, and prognosis for odontogenic carcinoma arising within odontogenic cysts was reviewed in the context of a case series. Two patients presented with rapidly enlarging masses of the maxilla (1), and, mandible(1). A review of English language case reports of such lesions was performed with PubMed and OvidWeb searches and is also presented as a context for the individual cases.

RESULTS (Cases)

Two cases met inclusion criteria for this report. Both patients were worked up with imaging studies (CT with contrast in both cases, MRI for maxillary tumor) and tissue biopsy. In both cases, a definitive diagnosis was not determined until the time of ablative surgery. Biopsies revealed malignant cells but the time of tumor was unclear initially.

Management in both cases was composite resection with wide margins; Bilateral neck dissection was performed for the mandibular tumor and withheld in the maxillary tumor. Fibula free flap reconstruction was performed in both cases.

The first carcinoma arose from an existing odontogenic keratocyst (OKC) of the mandible and was pathologically consistent with odontogenic squamous carcinoma. The second carcinoma was classified as an odontogenic ghost cell tumor (OGCT) of the maxilla arising from a mixed benign lesion with components of calcifying odontogenic cyst and compound odontoma. Radiation therapy was delivered to the maxilla and neck in an adjuvant fashion for the OGCT; this patient remains disease free at 29 months. Radiation was withheld in the mandibular tumor due to low grade features on pathology and absence of metastatic lymphadenopathy; This patient remains disease free at 10 months.

DISCUSSION

Odontogenic carcinoma arising from preexisting benign cysts is a rare occurrence best managed with surgery, immediate reconstruction, and followed by radiation therapy for concerning pathologic features or close margins, or, in certain pathologies thought to exhibit aggressive behavior. Because of the rarity of these lesions, applications of radiation therapy are extrapolated from experience with more common head and neck carcinomas. Primary surgery for odontogenic carcinoma should include composite resection with or without neck dissection depending on location of the primary with immediate reconstruction with composite tissue (usually free flap) that best rehabilitates the patient's defect. In most cases, this will include an osseous or osteocutaneous free flap such as the fibula, radial forearm, or scapular free flap.

The workup of malignant mandibular lesions begins with determining the pathology and exclusion of metastatic origins or invasion from adjacent tissue sources such as the sinuses or muscles of mastication.

CONCLUSIONS

Odontogenic carcinoma from benign cysts are a rare occurrence best managed with surgery and reconstruction, followed by radiation therapy for concerning pathologic features or close margins, and, in certain pathologies thought to exhibit aggressive behavior; Applications of radiation therapy are extrapolated from experience with squamous carcinomas. Primary surgery for odontogenic carcinoma should include composite resection with or without neck dissection depending on location of the primary with immediate reconstruction.

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