

Pediatric Odontogenic Tumors



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KEYWORDS

• Odontogenic tumor • Ameloblastoma • Odontoma • Jaws • Mandible • Maxilla

KEY POINTS

- Pediatric odontogenic tumors are mostly benign lesions, and are categorized based on their cells and tissues of origin.
- They typically occur in the posterior mandible region and are often associated with an impacted tooth.
- Most odontogenic tumors are amenable to simple enucleation and curettage; however, more aggressive or recurrent lesions may require radical surgery with appropriate reconstruction.
- Fear of disfigurement or psychosocial reasoning often influences surgeons to perform conservative treatment on children; however, serious consequences can occur, which include recurrence or malignant transformation of the tumor.

INTRODUCTION

Odontogenic tumors are rare tumors that affect patients in the maxillofacial region. These tumors can be varied in their presenting symptoms, overall growth rate and magnitude, and degree of tissue destruction. Many odontogenic tumors present with minimal symptoms, and are discovered only incidentally on radiographic examination.

If the lesion is symptomatic, patients typically present with rapid growth and expansion of their jaws. Cosmetic appearance may be altered with facial asymmetry, and there may be displacement and loosening of teeth in the affected area. Paresthesia of the inferior alveolar nerve is an unusual symptom because of the benign nature of these odontogenic tumors. If the inferior alveolar nerve is affected, this could indicate a long-standing lesion, or a malignant process.

Most of these tumors are benign and easily amenable to extirpative surgery, such as enucleation and curettage. More aggressive benign

lesions, as well as malignant tumors, require radical ablative surgery with appropriate immediate or delayed reconstruction. Although benign, if not treated appropriately, more aggressive odontogenic tumors will recur in anatomic locations of the head and neck that may be unresectable, thereby becoming life threatening because of airway compromise or cranial base involvement.¹

On a histologic level, odontogenic tumors are divided into 3 categories: epithelial, mesenchymal, and mixed epithelial and mesenchymal odontogenic tumors. Each category is defined based on the tissue from which the tumors arise.

Overall, oral and maxillofacial tumors in the pediatric population are rare. Studies are retrospective in nature, and usually combine multiple surgeon and institutional experiences. Overwhelmingly, these tumors are benign in most case series.² Soft tissue tumors, such as hemangiomas, papillomas, and lymphangiomas, make up close to 70% of all the head and neck tumors in the pediatric population.³ Pediatric odontogenic

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tumors consist of only one-third of the tumors seen in the maxillofacial region. The most common lesions are odontoma, ameloblastoma, and the keratocystic odontogenic tumor (KCOT).

Most literature reviews agree that the preponderance of development of these odontogenic tumors occurs in the pediatric population after the age of 6 years. It is during this time of secondary tooth development that the dental crown is being formed. Alteration in these particular cells can lead to the formation of odontogenic tumors.

EPITHELIAL ODONTOGENIC TUMORS

Odontogenic tumors of this category usually originate from the epithelial cells of tooth development. As the secondary teeth begin to develop and grow, cells from the rest of the dental lamina, developing enamel organ, epithelial lining of odontogenic cysts, and the basilar epithelial cells of the gingival surface epithelium, can be altered in their development.⁴ Most of these tumors occurring in the pediatric population are ameloblastomas. KCOTs are also seen frequently in syndromic children (eg, basal cell nevus syndrome).

Ameloblastoma

Epidemiology

Ameloblastoma is the most common aggressive benign tumor of the mandible and maxilla. However, its occurrence in the pediatric population is rare. Most case series and retrospective reviews across multiple institutions report that 10% to 15% of cases present in the pediatric age group.⁵ Ameloblastomas are classified into 3 major histologic subtypes: solid or multicystic, unicystic, and peripheral.

Unlike adult ameloblastomas, the pediatric population has a higher percentage of unicystic ameloblastomas. Most involve unerupted teeth, because it is thought that the lesions are produced de novo by neoplastic transformation of the non-neoplastic cyst lining.⁴ Ackerman, and colleagues⁶ described 3 histologic subtypes for the unicystic ameloblastoma. Type 1 and 2 have epithelium with no invasion into the fibrous cyst wall. Type 3 invades into the cyst wall either in a follicular or plexiform pattern, thereby having the capacity to invade adjacent bone (Fig. 1). Type 3 should be considered as aggressive a lesion as the conventional ameloblastoma.

The unicystic ameloblastoma mimics a dentigerous cyst clinically and radiographically, and therefore simple enucleation is often the treatment that is recommended and performed. The pathologic diagnosis of unicystic ameloblastoma is often surprising, with the expectation of a

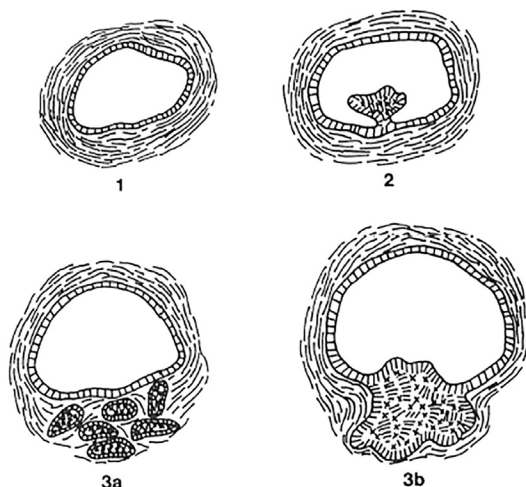


Fig. 1. Histologic subtypes: 1 and 2. Type 1 and 2 show no invasion into the fibrous cyst wall. 3. Type 3 shows invasion into the cystic wall in type 3a (follicular) or type 3b (plexiform) pattern. (From Ackermann GL, Altini M, Shear M. The unicystic ameloblastoma: a clinicopathological study of 57 cases. *J Oral Pathol* 1988;17:541; with permission.)

dentigerous cyst, leading the surgeon to question whether further treatment is indicated.⁷

Location

- Similar to adults, almost all occur in the body and angle of the mandible
- Location in the maxilla is very rare in the pediatric population; single case reports are noted in the literature

Symptoms

- Slow-growing facial deformity (Fig. 2)
- Tooth mobility and buccal expansion seen intraorally
- Complaints of pain and paresthesia are rare

Radiographic features

- Most have a unicystic appearance (Fig. 3)
 - Even solid ameloblastomas can appear unicystic on radiographs in pediatric patients

Treatment

Treatment of pediatric ameloblastoma can be controversial. As stated earlier, most pediatric ameloblastomas appear to be dentigerous cysts on clinical and radiographic examination. The diagnosis of unicystic ameloblastoma after enucleation can lead practitioners to undertreat patients in this population. Fearing the potential interruption of facial growth and loss of function, treating physicians have patients undergo the

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