Operative Techniques in Otolaryngology (2015) 26, 168–174

Operative Techniques in Otolaryngology

Management of ameloblastoma in the pediatric population

Sakeena J. Payne, MD,[a] Thomas W. Albert, DMD, MD,[b] Jessyka G. Lighthall, MD[a]

From the [a]Department of Surgery, The Pennsylvania State University Milton S. Hershey Medical Center, Hershey, Pennsylvania; and the [b]Department of Otolaryngology-Head and Neck Surgery, Oregon Health and Science University, Portland, Oregon

KEYWORDS
Ameloblastoma;
Pediatric jaw tumor;
Pediatric jaw cyst

Ameloblastomas are the most common benign tumors of odontogenic origin. They primarily occur in adults, with a minority occurring in the pediatric population (aged 18 and younger). Although imaging can suggest a diagnosis of ameloblastoma, they may be difficult to differentiate from other tumors or cysts of the maxillofacial skeleton and require histopathologic diagnosis. Treatment of these tumors in children is complex owing to the growing craniofacial skeleton. However, inadequate treatment results in a high recurrence rate. This paper reviews the epidemiology, histopathology, and treatment options for children with ameloblastoma.

Introduction

Tumors of the maxillofacial skeleton may be classified as either odontogenic or nonodontogenic and benign or malignant. Of the odontogenic tumors, ameloblastoma is the most common aggressive benign tumor, accounting for 1% of tumors and cysts of the jaw and 10% of odontogenic tumors overall.[1,2] Ameloblastomas may present as slowly enlarging facial swelling; however, most of them are found incidentally on radiographic evaluation. Although benign, these neoplasms are locally destructive and often recur if not entirely excised. Ameloblastomas typically occur within the third and fourth decade of life with the pediatric population accounting for approximately 10%-15% of all reported cases.[2]

Address reprint requests and correspondence: Sakeena J. Payne, MD, Division of Otolaryngology, Department of Surgery, Penn State Hershey Medical Center, 500 University Dr, Mailcode H091, Hershey, PA 17033.

E-mail address: Spayne1@hmc.psu.edu

http://dx.doi.org/10.1016/j.otot.2015.06.012
1043-1810/

Etiology

Ameloblastomas originate from odontogenic epithelium. They may develop from rests of dental lamina, the epithelial lining of a preexisting odontogenic cyst, oral mucosa basal cells, or developing enamel.[3] These tumors are unencapsulated and nonfunctional, representing arrested odontogenesis. In all, 3 subtypes have been described: conventional solid or multicystic intraosseous ameloblastoma, unicystic ameloblastoma, and peripheral or extraosseous ameloblastoma (Figure 1). Of these, the conventional solid variant is the most common. Previous studies claimed that unicystic ameloblastomas are most often seen in younger patients and that up to 50% of these lesions are diagnosed in the second decade of life.[1,4,5] More recently, it has been shown that even among the pediatric population, the solid multicystic type (63%-78%) predominates over the unicystic type (24%-37%).[2] This previous misconception is in part due to the original misdiagnosis of subtypes in older studies based on radiographic findings instead of pathologic evaluation.

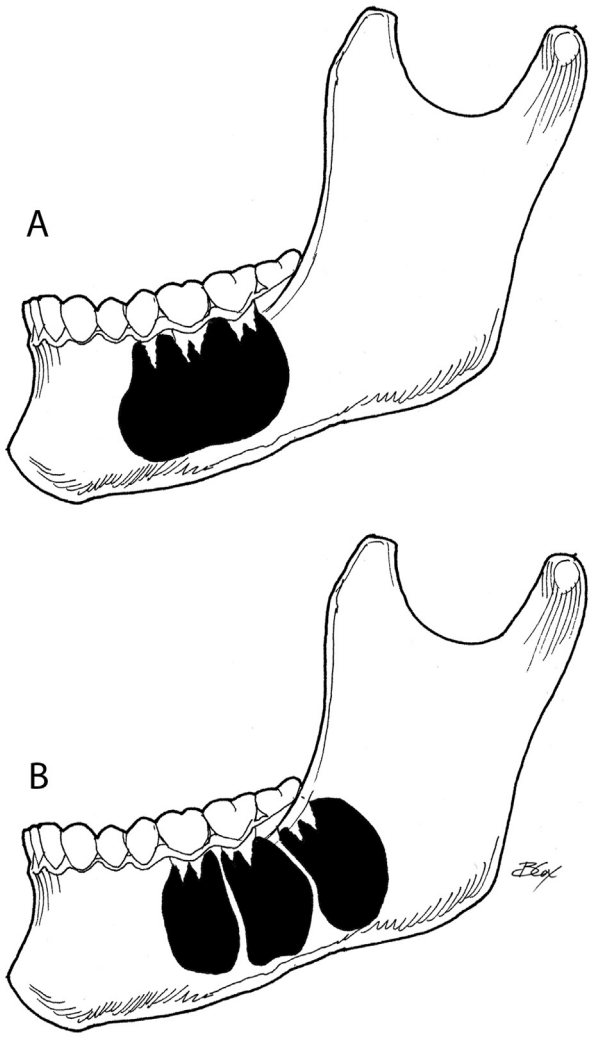

Figure 1 Examples of ameloblastoma: (A) Unilocular ameloblastoma and (B) Multilocular ameloblastoma.

Approximately 86% of all ameloblastomas fall within the conventional solid or multicystic subtype. Histopathologically these tumors have a combination of cystic and solid features, and several microscopic subtypes have been described although no difference in clinical behavior among the identified patterns has been found. The unicystic subtype accounts for 10%-46% of all intraosseous ameloblastomas. The pathogenesis of this variant is still a topic of debate. The first theory is that the lesion originates as a de novo neoplasm, whereas the second is that nonneoplastic cyst epithelium undergoes neoplastic transformation.

Unicystic ameloblastomas are further divided into luminal, intraluminal, and mural unicystic ameloblastoma based on infiltration of the cyst lumen. Although the unicystic variant is thought to be overall less aggressive compared with the solid subtype, mural unicystic ameloblastomas have proven to be as aggressive as the solid variant, warranting a more radical resection.

The peripheral ameloblastoma is the least common subtype, accounting for only 1%-10% of all ameloblastic tumors. They are often misdiagnosed as fibromas or pyogenic granulomas until histologically found to have the same features as those of the intraosseous forms.[6]

Clinical presentation

The incidence of ameloblastoma in the pediatric population is estimated to be between 7% and 39% of all cases.[2,7-10] The average age at presentation is 14.8 years and less than 10% of childhood cases occur under the age of 10 years.[1,2] There is a slight male predominance. Approximately 85%-90% of intraosseous ameloblastomas arise in the mandible and peripheral ameloblastomas are usually found on the posterior alveolar and gingival mucosa with some predilection for mandibular subsites.[6] Of the ameloblastomas that originate in the mandible, the molar-ramus region is the most common site of occurrence followed by the symphyseal region.[2]

Although ameloblastomas are typically asymptomatic, mass effect on surrounding structures may lead to symptoms and signs including pain, malocclusion, root resorption, loose teeth, paresthesias, ulceration, and trismus. Painless, progressive facial swelling or bony expansion of the jaw is the most common presentation. Smaller lesions are often discovered on routine surveillance panoramic radiography.

Evaluation

Evaluation of any child with a tumor of the maxillofacial skeleton should begin with a complete history and detailed head and neck physical examination. Careful evaluation of the face most often reveals a painless expansion or swelling of the jaw. Intraoral findings may include expansion of the mandibular or maxillary alveoli. Peripheral ameloblastomas usually present as a sessile gingival mass. Plain film panoramic radiography may suffice for diagnostic evaluation; however, computed tomography and magnetic resonance imaging provide more information on the extent of the lesion for operative planning.

Radiographic evaluation of multicystic ameloblastomas reveals radiolucent multilocular lesions, which are often described as having a soap bubble or honeycomb appearance. Expansion of the cortical bone as well as tooth root resorption can be seen with irregular scalloping of lesion margins. Solid ameloblastomas appear very similar to unilocular lesions on radiographic evaluation. Unicystic ameloblastomas are similar in appearance to odontogenic cysts and appear as a solitary and well-defined lucency typically surrounding the crown of an unerupted molar.[3,11] Peripheral ameloblastomas rarely display significant bony involvement, although in some cases the superficial alveolar bone can become eroded.[6] Ameloblastomas with an ill-defined radiographic boundary have been found to have the highest proliferative ability and poorest prognosis.[12]

Though physical examination and radiographic evaluation provide important clues, diagnosis and the eventual treatment plan are ultimately dependent on histopathologic