from slowly to rapidly growing lesions. However, the degree of histologic differentiation, as determined by the relative degree of keratinization, is not correlated with behavior. As a group these tumors tend to infiltrate locally before they metastasize to other sites. The routes of extension depend on the primary site. The favored sites of local metastasis are the cervical lymph nodes, while the most common sites of distant metastasis are mediastinal lymph nodes, lungs, liver, and bones. Unfortunately, such distant metastases are often already present at the time of discovery of the primary lesion.

Odontogenic Cysts and Tumors

The overwhelming majority of odontogenic cysts are derived from remnants of odontogenic epithelium present within the jaws. In contrast to the rest of the skeleton, epithelial-lined cysts are quite common in the jaws. In general, these cysts are subclassified as either inflammatory or developmental (Table 16-2). Only the most common of these lesions are described later.

The *dentigerous cyst* is defined as a cyst that originates around the crown of an unerupted tooth and is thought to be the result of fluid accumulation between the developing tooth and the dental follicle. Radiographically, these are unilocular lesions most often associated with impacted third molar (wisdom) teeth. Histologically, they are lined by a thin layer of stratified squamous epithelium. Often, there is a dense chronic inflammatory cell infiltrate in the connective tissue stroma. Complete removal of the lesion is curative.

The *odontogenic keratocyst (OKC)*, now called *keratocystic odontogenic tumor*, is an important lesion that must be differentiated from other odontogenic cysts because of its aggressive behavior. OKCs can be seen at any age but are most often diagnosed in patients between ages 10 and 40. They occur most commonly in males within the posterior mandible. Radiographically, OKCs present as well-defined unilocular or multilocular radiolucencies. Histologically, the cyst lining consists of a thin layer of keratinized stratified squamous epithelium with a prominent basal cell layer and a corrugated epithelial surface. Treatment requires complete removal of the lesion, because OKCs are locally aggressive and recurrence rates for inadequately removed lesions can reach 60%. About 80% of the lesions are solitary, but multiple OKCs occur in roughly 20% of patients, who should be evaluated for nevoid basal cell carcinoma syndrome (Gorlin syndrome), which is associated with mutations in the tumor suppressor gene *PTCH* (Patched) located on chromosome 9q22 (Chapter 25).

Table 16-2 Histologic Classification of Odontogenic Cysts

Inflammatory
Periapical cyst
Residual cyst
Paradental cyst
Developmental
Dentigerous cyst
Odontogenic keratocyst
Gingival cyst of newborn
Gingival cyst of adult
Eruption cyst
Lateral periodontal cyst
Glandular odontogenic cyst
Calcifying epithelial odontogenic cyst (Gorlin cyst)

Table 16-3 Histologic Classification of Odontogenic Tumors

Tumors of odontogenic epithelium
Benign
Ameloblastoma
Calcifying epithelial odontogenic tumor (Pindborg tumor)
Squamous odontogenic tumor
Adenomatoid odontogenic tumor
Malignant
Ameloblastic carcinoma
Malignant ameloblastoma
Clear-cell odontogenic carcinoma
Ghost cell odontogenic carcinoma
Primary intraosseous squamous cell carcinoma
Tumors of odontogenic ectomesenchyme
Odontogenic fibroma
Odontogenic myxoma
Cementoblastoma
Tumors of odontogenic epithelium and ectomesenchyme
Benign
Ameloblastic fibroma
Ameloblastic fibro-odontoma
Adenomatoid odontogenic tumor
Odontoameloblastoma
Complex odontoma
Compound odontoma
Calcifying cystic odontogenic tumor (calcifying odontogenic cyst)
Dentinogenic ghost cell tumor
Malignant
Ameloblastic fibrosarcoma

The *periapical cyst,* in contrast to the developmental cysts described earlier, is inflammatory in origin. These are common lesions found at the apex of teeth. They develop as a result of long-standing inflammation of the tooth (pulpitis), which may be caused by advanced carious lesions or by trauma to the tooth in question. The inflammatory process may result in necrosis of the pulpal tissue, which can traverse the length of the root and exit at the apex of the tooth into the surrounding alveolar bone. Over time, like any chronic inflammatory process, a lesion with granulation tissue may develop, and subsequent epithelialization may lead to the formation of a radicular cyst. While the term *periapical granuloma* is not the most appropriate terminology (the lesion does not show true granulomatous inflammation), this older name is still used. Periapical inflammatory lesions persist as a result of the continued presence of bacteria or other irritating agents in the area. Successful treatment, therefore, necessitates the complete removal of offending material and appropriate restoration of the tooth or extraction.

Odontogenic tumors are a group of lesions with diverse histologic appearances and clinical behavior. Some are true neoplasms (both benign and malignant), while others are more likely hamartomas. Odontogenic tumors are derived from odontogenic epithelium, ectomesenchyme, or both (Table 16-3). The two most common and clinically significant tumors are the following: