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**TUMORS OF TEETH AND JAWS:
PATHOLOGY OF TUMOR-LIKE CONDITIONS
OF THE DENTOALVEOLAR SYSTEM**

Minsk BSMU 2018

МИНИСТЕРСТВО ЗДРАВООХРАНЕНИЯ РЕСПУБЛИКИ БЕЛАРУСЬ
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**ОПУХОЛИ ЗУБОЧЕЛЮСТНОЙ СИСТЕМЫ:
ПАТОЛОГИЧЕСКАЯ АНАТОМИЯ
ОПУХОЛЕПОДОБНЫХ ПРОЦЕССОВ
ЗУБОЧЕЛЮСТНОЙ СИСТЕМЫ**

**TUMORS OF TEETH AND JAWS: PATHOLOGY
OF TUMOR-LIKE CONDITIONS
OF THE DENTOALVEOLAR SYSTEM**

Учебно-методическое пособие



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INTRODUCTION

Tumor-like processes (benign processes) in the jaws represent a wide spectrum of conditions, many of which have cystic form. Tumor-like processes can be diagnosed with intraoral and panoramic radiography. Large tumor-like processes may expand jaw bone extensively and grow into neighboring structures. Since some types of tumor-like cystic processes may be locally aggressive and may even be difficult to distinguish from malignant neoplasms. Accordingly for accurate assessment of their structure and the determination of the biological potential of aggressiveness, it is necessary to use various visualizing instrumentations with an obligatory postoperative pathological study.

GENERAL CLINICAL PRESENTATION OF CYSTIC TUMOR-LIKE PROCESSES CYSTS OF THE JAWS

Traditionally, a cyst is defined as a pathological cavity lined wholly or in part by epithelium, having fluid or semi-fluid contents. However, as there are a few cysts which do not have an epithelial lining. The maxilla and mandible are more usually affected by disease arising from closely related structures such as the teeth and oral mucosa, rather than from primary bone disease. Disease of the teeth presents as pain, mobility or swelling although some conditions are detected as incidental findings at radiographic examination. Developmental cysts, odontogenic hamartomas and neoplasms are often painless but may present with bony swelling, facial asymmetry or a failure of teeth to erupt. Primary disease of the jawbones presents as bony swellings that may or may not involve overlying mucosa or adjacent teeth. Mobility is assessed in terms of buccolingual and vertical movement and is due to destruction of periodontal support, perhaps as a result of periodontal disease or because of an adjacent cyst or tumour. Plain radiographs are essential for the assessment of intrabony cystic lesions, particularly to determine the size, outline (unilocular or multilocular) and bone-lesion interface (sclerotic and well-defined implies a slow-growing lesion; ill-defined suggests a rapidly growing destructive lesion). CT scanning particularly with 3D-reconstruction is useful for large lesions and for assessing relationships with adjacent anatomical structures. MR images are not good at penetrating bone. Cysts may be detected because of clinical symptoms or signs. Occasionally an asymptomatic cyst may be discovered on a radiograph taken for another purpose. Symptoms may include: swelling; displacement or loosening of teeth; and pain (if infected). The most important clinical sign is expansion of bone. In some instances, this may result in an eggshell-like layer of periosteal new bone overlying the cyst. This can break on palpation, giving rise to the clinical sign of “eggshell cracking”.

If the cyst lies within soft tissue or has perforated the overlying bone, then the sign of fluctuance may be elicited by palpating with fingertips on each side of the swelling in two positions at right angles to each other. If a cyst becomes infected, the clinical presentation may be that of an abscess, the underlying cystic lesion only becoming apparent on radiographic examination.

CLASSIFICATION OF CYSTS OF THE JAWS

Cysts of the jaws are classified into several categories depending on histogenesis and etiology. Cysts of the jaws are more common than in any other bone, and the majority are lined wholly or in part by epithelium. Although the pathogenesis of many of these cysts is poorly understood, they are divided into two main groups depending on the origin of the lining epithelium. Those that arise from odontogenic epithelium are called odontogenic, those that have their source in other epithelial structures are known as non-odontogenic. Odontogenic cysts can be subdivided into developmental and inflammatory types depending on their etiology.

The classification of jaw cysts is based on that recommended by the World Health Organization.

Cysts of the jaws:

1. Odontogenic cysts.

1.1. Inflammatory:

- radicular cyst (apical and lateral);
- residual cyst;
- paradental cyst.

1.2. Developmental:

- dentigerous cyst (follicular cyst);
- lateral periodontal cyst;
- botryoid odontogenic cyst;
- glandular odontogenic cyst (sialo-odontogenic cyst);
- odontogenic keratocyst (“primordial” cyst, keratocystic odontogenic tumour);

- gingival cyst;
- eruption cyst;
- gingival cyst of adults;
- “gingival cysts” of infants (Epstein pearls).

2. Non-odontogenic cysts (developmental):

- nasopalatine duct cyst;
- nasolabial cyst;
- surgical ciliated cyst.

3. Pseudocysts:

- solitary bone cyst;

- aneurysmal bone cyst;
- focal bone marrow defect.

OTHER CYSTIC OR CYST-LIKE LESIONS

Cystic odontogenic tumours are not included in the above classification, but cystic ameloblastomas and calcifying odontogenic (ghost cell) cysts can be mistaken radiographically for radicular or other non-neoplastic cysts as discussed later.

Stafne's idiopathic bone cavity is discussed in this chapter since, while it is not a cyst, it may be mistaken for such on a radiograph.

Other cysts

Cysts associated with the maxillary antrum:

- benign mucosal cyst of the maxillary antrum;
- postoperative maxillary cyst (surgical ciliated cyst of the maxilla).

Cysts of the soft tissues of the mouth, face and neck:

- cysts of the salivary glands: mucous extravasation cyst, mucous retention cyst, ranula;
- dermoid and epidermoid cysts;
- lymphoepithelial (branchial cleft) cyst;
- thyroglossal duct cyst.

ODONTOGENIC CYSTS

Definition and origins.

By definition, the epithelial lining of these cysts originates from residues of the tooth-forming organ. There are three kinds of residue, each primarily responsible for the origin of a particular type of lesion:

1. The epithelial rests or glands of Serres persisting after dissolution of the dental lamina. These give rise to the odontogenic keratocyst. They may also be the origin of some developmental lateral periodontal and gingival cysts.

- Remnants of the dental lamina: odontogenic keratocyst, lateral periodontal cyst, gingival cyst of adult, glandular odontogenic cyst.

2. The reduced enamel epithelium which is derived from the enamel organ and covers the fully formed crown of the unerupted tooth. The dentigerous (follicular) and eruption cysts are derived from this tissue, as is the relatively uncommon inflammatory paradental cyst.

- Reduced enamel epithelium: dentigerous cyst, eruption cyst.

3. The rests of Malassez formed by fragmentation of the epithelial root sheath of Hertwig. All radicular cysts originate from these residues.

- Rests of Malassez: radicular cyst, residual cyst.

4. Unclassified: paradental cyst.

CYST GROWTH PATHOGENESIS

Several mechanisms are described for cyst growth, including:

- proliferation of epithelial lining and fibrous capsule;
- internal hydrostatic pressure of cyst fluid;
- resorption of surrounding bone.

RADIOLOGICAL EXAMINATION: GENERAL PRINCIPLES

As a basic principle, radiological examination should commence with intra-oral films of the affected region; for small cystic lesions, intra-oral films may be all that is needed for diagnosis, while for all cysts the fine detail of intra-oral radiography will help to clarify the relationship between lesion and teeth. For larger lesions, more extensive radiography is appropriate. Selection of films should take account of the value of having two views with differing perspectives.

Radiological signs. Classically, cysts appear as well-defined round or ovoid radiolucencies, surrounded by a well-defined margin.

Margins. Peripheral cortication (radio-opaque margin) is usual except in solitary bone cysts. 'Scalloped' margins are seen in larger lesions, particularly keratocysts. Infection of a cyst tends to cause loss of the well-defined margin.

Shape. Most cysts grow by hydrostatic mechanisms, resulting in the round shape. Odontogenic keratocysts and solitary bone cysts do not grow in this manner and have a tendency to grow through the medullary bone rather than to expand the jaw.

Locularity. True locularity (multiple cavities) is seen occasionally in odontogenic keratocysts. However, larger cysts of most types may have a multilocular appearance because of ridges in the bony wall.

Effects upon adjacent structures. Where a lesion abuts another structure, such as a tooth or the inferior dental canal, it may cause displacement. Roots of teeth may be resorbed. When a cyst reaches a certain size, the cortex of the bone often becomes thinned and expanded. In posterior maxillary lesions the antral floor may be raised. Perforation of the cortical plates may be recognized as a localised area of greater radiolucency overlying the lesion.

Effect on unerupted teeth. Unerupted teeth may become enveloped by any cyst, a feature which may lead to erroneous diagnosis as a dentigerous cyst.

Most jaw cysts behave similarly and usually grow slowly and expansively. They differ mainly in their relationship to a tooth, and the radiographic features are usually a good guide as to their nature. These occasionally have identical radiographic appearances, and diagnosis ultimately depends on histopathology.

Key features of jaw cysts:

1. Form sharply-defined radiolucencies with smooth borders.
2. Fluid may be aspirated and thin-walled cysts may be transilluminated.

3. Grow slowly, displacing rather than resorbing teeth.
4. Symptomless unless infected and are frequently chance radiographic findings.
5. Rarely large enough to cause pathological fracture.
6. Form compressible and fluctuant swellings if extending into soft tissues.
7. Appear bluish when close to the mucosal surface.

DIFFERENTIAL DIAGNOSIS

There are many causes of circumscribed areas of **cyst-like areas of radiolucency** in the jaws: anatomical structures (maxillary antrum and foramina); pseudocysts; neoplasms, particularly ameloblastoma; giant cell granuloma of the jaws; hyperparathyroidism; cherubism.

True cysts are usually readily recognised by their clinical and radiographic features. The cystic nature of a lesion can be confirmed by aspirating its contents through a needle inserted through the wall under aseptic conditions. The detection of fluid does not distinguish one cyst from another or a cystic neoplasm from a true cyst. The presence of cholesterol crystals is not of diagnostic significance. Very large radiolucencies tend to be cysts rather than abscesses, although they may be infected.

Histologically, the lining of simple stratified squamous epithelium with an inflammatory infiltrate confirms the diagnosis.

Neoplasms. Benign (odontogenic) tumours or occasionally an ameloblastoma may resemble a simple cyst radiographically. Resorption of adjacent teeth suggests a neoplasm rather than a cyst but is not diagnostic in itself. It is essential that the whole of a cyst lining be available for examination, since part of the lining, even though neoplastic, may appear as a thin layer of flattened stratified squamous epithelium like that of a simple cyst. However, it is particularly important to distinguish keratocysts and unicystic ameloblastomas from common types.

Rarely a metastasis in the jaw may cause a sharply defined area of radiolucency. More often it causes a lesion with a hazy outline and irregular shape. Tumours also tend to be painful and to grow more rapidly than cysts. Nevertheless, it may be difficult or impossible to distinguish them from infected cysts radiographically, but at operation the solid nature of a tumour will be obvious and histological examination will confirm the diagnosis.

INCIDENCE OF CYSTS OF THE JAWS

Cysts are the most common cause of chronic swellings of the jaws. They are more common in the jaws than in any other bone because of the many rests

of odontogenic epithelium remaining in the tissues. Odontogenic cysts (not surprisingly) affect the tooth-bearing region of the jaws. Most non-odontogenic, true cysts are developmental, and form in the region of the anterior maxilla. The incidence of each type of jaw cyst varies slightly from series to series and the figures given in Table 3 are approximations. At least 90 per cent of all jaw cysts are of odontogenic origin.

Relative frequency of different types of jaw cysts:

- radicular (periodontal) — 65–70 %;
- dentigerous — 15-18 %;
- odontogenic keratocysts — 3–5 %;
- nasopalatine — 5–10 %;
- lateral periodontal < 1 %;
- paradental < 1 %.

TREATMENT

Enucleation and primary closure

Enucleation of a cyst involves the removal of the whole cyst, including the epithelial and capsular layers from the bony walls of the cavity. Enucleation is the usual method and usually entirely effective (table 1). The affected (dead) tooth may be extracted or rootfilled and preserved. A mucoperiosteal flap over the cyst is raised and a window is opened in the bone to give adequate access. The cyst is then carefully separated from its bony wall. The entire cyst is removed intact and should be sent in fixative for histological examination. The edges of the bone cavity are smoothed off, free bleeding is controlled and the cavity is irrigated to remove debris. The mucoperiosteal flap is replaced and sutured in place. The sutures should be left for at least 10 days.

Table 1

Advantages and limitations of enucleation of cysts

Advantages	Possible disadvantages
The cavity usually heals without complications. Little aftercare is necessary. The complete lining is available for histological examination	Infection of the clot filling the cavity. Recurrence due to incomplete removal of the lining. Serious haemorrhage (primary or secondary). Damage to apices of vital teeth projecting into the cyst cavity. Damage to the inferior dental nerve. Opening the antrum when enucleating a large maxillary cyst. Fracture of the jaw if an exceptionally large mandibular cyst is enucleated

Any disadvantages are largely theoretical and in competent hands even very large cysts can be enucleated safely. Recurrence is remarkably rare. There are few contraindications: they are relative rather than absolute.

Marsupialisation

Marsupialisation is a simple operation that may be performed under local anaesthesia in which a window is cut and removed from the cyst lining. This allows decompression of the cyst, which then slowly heals by bone deposition in the base of the cavity. Marsupialisation is a largely outmoded treatment for radicular cysts. The cyst is opened essentially as for enucleation but the lining is sutured to the mucous membrane at the margins of the opening. The aim is to produce a self-cleansing cavity which becomes in effect, an invagination of the oral tissues. However, considerable aftercare is needed to keep the cavity clean. The cavity is initially packed with ribbon gauze and after the margins have healed a plug or extension to a denture is made to close the opening. Food debris has to be regularly washed out, but once the cavity has filled up from the base and sides sufficiently to become self-cleansing the plug can be removed. The cavity usually becomes closed by regrowth of the surrounding tissue and restoration of the normal contour of the part. However, the orifice may close and the cyst re-form. Also, the complete lining is not available for histological examination. The main application of marsupialisation is for temporary decompression of exceptionally large cysts where fracture of the jaw is a risk. When enough new bone has formed, the cyst can be enucleated. Occasionally retention of the tooth in a dentigerous cyst is needed and marsupialisation may allow it to erupt. Marsupialization (partial removal) requires considerable aftercare and patient cooperation in keeping the cavity clean — syringing after meals. Healing may take up to six months and not all cyst lining is available for histopathology.

Radicular cysts are discussed first as being the most common type, whose treatment forms the basis for that of most other jaw cysts.

RADICULAR CYSTS

Radicular cysts (also known as *apical periodontal cyst*, *dental cyst*) are located at the root tips of teeth in which the pulp has become necrotic, mostly due to advanced tooth caries. These inflammatory lesions form a spectrum of changes related to the apical region of a non-vital tooth (usually a consequence of dental caries), with considerable overlap in clinical, radiological and pathological findings. Radicular cysts are subdivided into apical, lateral, and residual types depending on the anatomical relationship of the cyst to the root of the tooth. Apical radicular cysts are the most common cystic lesions in the jaws and are always associated with the apices of non-vital teeth. They account for about 75 per cent of all radicular cysts.

Aetiology. Pulp death, apical periodontitis, proliferation of epithelial rests of Malassez, cystic change in epithelium; expansion of cyst by hydrostatic pressure; resorption of surrounding bone.

Incidence. Radicular cysts are the most common cause of chronic swellings of the jaws.

Very common, 70 % of jaw cysts; 60 % occur in maxilla; all ages but rare in children and with deciduous teeth. They are rarely seen before the age of 10 and are most frequent between the ages of 20 and 60 years. They are more common in males than females, roughly in the proportion of 3 to 2. The maxilla is affected more than three times as frequently as the mandible.

Clinical Presentation. Radicular cysts, like other cysts of the jaws, cause slowly progressive painless swellings, with no symptoms until they become large enough to be conspicuous. When small they are frequently symptomless and are usually discovered during routine radiological examination. As they enlarge they produce expansion of the alveolar bone and ultimately may discharge through a sinus. If infection enters, the swelling becomes painful and may rapidly expand, partly due to inflammatory oedema. The swelling is rounded and at first hard. Later, when the bone has been reduced to eggshell thickness, a crackling sensation may be felt on pressure. Finally, part of the wall is resorbed entirely away, leaving a soft fluctuant swelling, bluish in colour, beneath the mucous membrane. The rate of expansion of radicular cysts has been estimated at approximately 5 mm in diameter per year. Pain is seldom a feature unless there is an acute exacerbation which may rapidly progress to abscess formation.

Pathogenesis. Arise when the contents of the necrotic pulp canal leak out of the apical foramina and set up an inflammatory reaction at the apex. The persistent inflammatory stimulus induces granulation tissue formation to help wall off the necrotic debris. Epithelial rests around the root (“cell rests of Malassez”) proliferate, initially as complex strands and arcades then as a well-defined lining; when present the epithelium allows the term radicular cyst to be used. Cysts enlarge by a hydrostatic mechanism — the high protein content of the inflammatory exudate in the lumen draws water into the cyst while the lack of lymphatics in the wall prevents it draining away — producing a rounded radiolucency usually with a sclerotic border. May resorb the apical portion of the tooth. Most are located apically but 10 % are seen in lateral relationship (accessory apical foramina).

Major factors in the pathogenesis of cyst formation

Epithelial proliferation. Infection from the pulp chamber induces inflammation and proliferation of the epithelial rests of Malassez. If infection can be eliminated from the root canal, small radicular cysts (up to 1 or 2 cm diameter) may regress without surgery.

Hydrostatic effects of cyst fluids. That radicular and many other cysts expand in balloon-like fashion, wherever the local anatomy permits, indicates that internal pressure is a factor in their growth. The hydrostatic pressure within cysts is about 70 cm of water and therefore higher than the capillary blood

pressure. Cyst fluid is largely inflammatory exudate and contains high concentrations of proteins, some of high molecular weight which can exert osmotic pressure. Consistent with the inflammation usually present in cyst walls, cyst fluid may contain cholesterol, breakdown products of blood cells, exfoliated epithelial cells, and fibrin. The cyst wall does not seem to act entirely as a simple semipermeable membrane. Low-molecular-weight proteins are present in similar concentrations to those in the plasma but there are smaller amounts of high-molecular-weight proteins. The capillaries in the cyst wall are more permeable as a result of inflammation and contribute varying amounts of immunoglobulins and other proteins. The net effect is that pressure is created by osmotic tension within the cyst cavity.

Bone-resorbing factors. Experimentally, cyst tissues in culture release bone-resorbing factors. These are predominantly prostaglandins E2 and E3. Different types of cysts and tumours may produce different quantities of prostaglandins but if so, it is unclear whether this affects the mode of growth of the cyst. Collagenases are present in the walls of keratocysts, but their contribution to cyst growth is also unclear.

Radiography. A well-defined outline, round or ovoid radiolucency is associated with the root apex or, less commonly in the lateral position, of a heavily restored or grossly carious tooth. A condensed radiopaque periphery is present only if growth is unusually slow and is usually more prominent in long-standing cysts. The dead tooth from which the cyst has arisen can be seen and often has a large carious cavity. Adjacent teeth may be tilted or displaced a little or occasionally become slightly mobile. Very large cysts in the maxilla may extend in any available direction and become irregular. Infection of a cyst causes the outline to become hazy as a result of greater vascularity and resorption of the surrounding bone.

However, the clinical presentation, location at the root tip of a nonvital tooth, leaves little room for other diagnostic possibilities.

Pathology. The histologic appearance of the radicular cyst is rather unspecific. All stages can be seen from a periapical granuloma containing a few strands of proliferating epithelium derived from the epithelial rests of Malassez, to an enlarging cyst with a hyperplastic epithelial lining and dense inflammatory infiltrate. Epithelial proliferation results from irritant products leaking from an infected root canal to cause chronic periapical inflammation.

The epithelial lining consists of stratified nonkeratinising squamous epithelium of variable thickness. It lacks a well-defined basal cell layer and is sometimes incomplete. Early, active epithelial proliferation is associated with obvious chronic inflammation and may then be thick, irregular and hyperplastic or appear net-like, forming rings and arcades. Hyperplasia is a prominent feature resulting in long anastomosing cords of epithelium forming complex arcades extending into the surrounding capsule. Breaks in the lining —

epithelial discontinuities — are common. Within the cyst epithelium, in approximately 10 per cent of cases the lining contains hyaline eosinophilic bodies — Rushton bodies — of varying size and shape. The specific nature of these so-called Rushton bodies is unclear. Hyaline (Rushton) bodies are thin refractile rod-like or hair-pin or other shapes. Staining is variable. Their nature is unknown. They may be an epithelial product or haematogenous in origin.

Occasionally, the lining squamous cells are admixed with mucous cells or ciliated cells. Metaplasia of the epithelial lining may give rise to mucous cells, found in about 40 per cent of radicular cyst linings and, more rarely, ciliated cells and areas of respiratory-type epithelium. May be present in the cyst lining and capsule foamy macrophages and haemosiderin. Long-standing cysts typically have a thin flattened epithelial lining, a thick fibrous wall and minimal inflammatory infiltrate.

The cyst capsule and wall. The capsule consists of collagenous fibrous connective tissue. During active growth the capsule is vascular and infiltrated by chronic inflammatory cells adjacent to the proliferating epithelium. Plasma cells are often prominent or predominant, and are a response to antigens leaking through the tooth apex.

In *the bony wall* there is osteoclastic activity and resorption. Beyond the zone of resorption there is usually active bone formation. The net effect is that a cyst expands but retains a bony wall, even after it has extended beyond the normal bony contours. This bony wall nevertheless becomes progressively thinner since repair is slower than resorption, until it forms a mere eggshell, then ultimately disappears altogether. The cyst then starts to distend the soft tissues and appears as a soft bluish swelling.

Clefts. Cholesterol from the breakdown of blood cells is frequently seen in cysts as needle-shaped clefts. These are left by cholesterol dissolved-out during preparation for sectioning. The cholesterol is derived from breakdown of blood cells. Small clefts are enclosed by foreign body giant cells, and extravasated red cells and blood pigment are associated. Clefts may also be seen extending into the cyst contents but are formed in the cyst wall.

Cyst fluid. The fluid is usually watery and opalescent but sometimes more viscid and yellowish, and sometimes shimmers with cholesterol crystals. Aspiration of cyst fluid typically also shows cholesterol as flat, rhomboid, notched crystals often with many inflammatory cells. Histologically, the protein content of the fluid is usually seen as amorphous eosinophilic material, often containing breakdown leucocytes and cells distended with fat globules. A smear of this fluid may show typical notched cholesterol crystals microscopically. Cholesterol crystals often seen in the cyst fluid but not specific to radicular cysts.

The composition of cyst fluid is complex and variable. It is hypertonic compared with serum and contains:

1. Breakdown products of degenerating epithelial and inflammatory cells, and connective tissue components.
2. Serum proteins. All groups of serum proteins are present in cyst fluid and the soluble protein level is 5–11 g/dl. Most are derived as an inflammatory exudate. Compared with serum the fluid contains higher levels of immunoglobulin which probably reflects local production by plasma cells in the capsule.
3. Water and electrolytes.
4. Cholesterol crystals.

RESIDUAL RADICULAR CYSTS

The other varieties of radicular cyst are less common. A radicular cyst may persist after extraction of the causative tooth. The residual cyst is a radicular cyst that has remained in the jaw and failed to resolve following extraction of the involved tooth. Residual cysts may interfere with the fit of dentures, but may slowly regress spontaneously. This is suggested by the progressive thinning of the lining. Residual cysts are a common cause of swelling of the edentulous jaw in older persons. About 20 per cent of radicular cysts are of this type. However, it should be noted that most periapical inflammation will resolve after removal of the causative agent. The reasons why some lesions persist as residual cysts are unknown.

LATERAL RADICULAR CYSTS

The lateral type of **radicular cyst** is very uncommon. Lateral cysts can occasionally form at the side of a nonvital tooth and arises as a result of extension of inflammation from the pulp into the lateral periodontium along a lateral root canal. Others are developmental and form beside a vital tooth. *Treatment* usually involves endodontic therapy (root canal treatment), apicectomy (removing the apical 2 mm of the tooth root via a surgical approach and sealing off the pulp canal) or removal of the tooth. Recurrence is uncommon but relates to a failure to control the contents of the pulp canal.

PARADENTAL CYSTS

Paradental cysts occasionally result from inflammation round partially erupted teeth. The paradental cyst is located on the lateral side of the tooth at the border between the enamel and root cementum. This cyst is secondary to an inflammatory process in the adjacent periodontal tissues that induces

proliferation of neighbouring odontogenic epithelial rests, similar to the pathogenesis of the radicular cyst. It is a rare lesion. They affect males predominantly, usually between the ages of 20–25 years, particularly mandibular third molars. The affected tooth is vital but typically shows pericoronitis. Radiographically, they appear as well-defined radiolucencies related to the neck of the tooth and the coronal third of the root.

Histologically, it resembles the other inflammatory odontogenic cysts or a radicular cyst but for a more intense inflammatory infiltrate in the wall, the distinction being made by the specific clinical presentation. Treatment consists of excision with or without concomitant extraction of the involved tooth. Enucleation is effective.

DENTIGEROUS CYSTS

Dentigerous cyst (follicular cyst): a developmental cyst that surrounds the crown of an unerupted tooth and is attached at the cervical region and is a dilatation of the follicle. A dentigerous cyst encloses part or all of the crown of an unerupted tooth. It is attached to the amelocemental junction and arises in the follicular tissues covering the fully formed crown of the unerupted tooth.

Aetiology. Cystic change in remains of enamel organ after completion of enamel formation. This is a developmental defect of unknown cause.

Incidence. Common, 15–18 % of jaw cysts. Dentigerous cysts occur over a wide age range and although many are detected in adolescents and young adults there is an increasing prevalence up to the fifth decade. They are uncommon in children. They are about twice as common in males than in females and twice as common in the mandible than in the maxilla. The cysts most frequently involve teeth which are commonly impacted or erupt late. The majority are associated with the mandibular third molar and then, in order of decreasing frequency, the maxillary permanent canines, maxillary third molars, and mandibular premolars. Uncommonly, they are associated with supernumerary teeth or with complex and compound odontomes.

Clinical features. Like other cysts, uncomplicated dentigerous cysts cause no symptoms until the swelling becomes noticeable. Alternatively a dentigerous cyst may be a chance radiographic finding or found when the cause is sought for a missing tooth. Pain is not a feature unless there is secondary inflammation. Infection of a dentigerous cyst accelerated swelling.

Pathogenesis and expansion. Dentigerous cysts develop from the follicular tissues, but the stimulus is unknown and the mechanism of cyst formation unclear. Enlargement is by hydrostatic mechanisms but what generates the forces is not clear.

The cyst develops between the crown of the unerupted tooth and the reduced enamel epithelium, but the mechanisms of cyst formation are

unknown. One hypothesis suggests that compression of the follicle by a potentially erupting but impacted tooth increases the venous pressure in the follicle, leading to increased transudation of fluid. Pooling of this transudate separates the follicle from the crown, resulting in cyst formation. Another hypothesis suggests that the cysts arise as a result of proliferation of the outer layers of the reduced enamel epithelium, as would normally occur in tooth eruption, followed by breakdown of cells within the epithelial islands, leading to cyst formation. Attachment of the cyst lining at or near the amelocemental junction suggests that dentigerous cysts arise as a result of cystic change in the remains of the enamel organ after enamel formation is complete. Division between the remnants of the internal enamel epithelium covering the enamel and the external enamel epithelium, forming the greater part of the cyst lining, can occasionally be seen at the attachment of the cyst to the neck of tooth. In a few cases a cyst may arise as a result of spread of periapical inflammation from a deciduous predecessor to involve the follicle of the permanent successor, accumulation of inflammatory exudate leading to cyst formation. These inflammatory dentigerous cysts most often involve premolar teeth and are much more common in the mandible than in the maxilla. The mechanism of expansion of dentigerous cysts is probably similar to that of radicular cysts. Bone resorbing factors, including prostaglandin E2 and interleukin-1, are produced by the cysts and the contents are hypertonic compared with serum. The rate of cyst expansion in children may be rapid but enlargement is much slower in adults. Progressive growth of the cyst leads to dilatation of the dental follicle. Factors triggering these changes are not known. However, there is a strong association between failure of eruption of teeth and formation of dentigerous cysts which predominantly affect teeth which are particularly prone to failure of eruption, namely, maxillary canines and mandibular third molars in particular.

Radiography. Well-defined radiolucency, unilocular in form with a sclerotic border surrounding the crown of an unerupted tooth (so-called “dentigerous relationship”). May resorb roots of adjacent teeth.

The cavity is well circumscribed and contains the crown of a tooth displaced from its normal position. Occasionally there may be pseudoloculation as a result of trabeculation or ridging of the bony wall. The slow growth of dentigerous cysts usually results in a sclerotic bony outline and a well-defined cortex. The affected tooth is often displaced a considerable distance, and a third molar, for instance, may be pushed to the lower border of the mandible. Rarely, if the cyst remains unrecognised for a long period the enclosed tooth may become resorbed.

These features readily distinguish a dentigerous cyst from a radicular cyst. However, a keratocyst or ameloblastoma may occasionally envelop the crown of the tooth, and either of these may create the radiographic appearance of

a dentigerous cyst. The diagnosis ultimately therefore depends on histological examination.

Radiographically, other cysts may present in apparent dentigerous relationship. For example, an odontogenic keratocyst may envelop the crown of an impacted third molar and these extrafollicular lesions may be difficult to distinguish from true dentigerous cysts.

In most instances, dentigerous cysts are a fortuitous finding on oral radiographs. Only when excessively large may they cause swelling of the involved part of the jaw. If there is inflammation, they will cause pain and swelling. Removal of the cyst wall and the tooth involved will yield a permanent cure.

Pathology. Macroscopic examination of intact specimens reveals a cyst attached to the amelocemental junction. In most cases the cyst completely surrounds the crown of the associated tooth (central type). Less frequently the cyst projects laterally from the side of the tooth and does not completely enclose the crown (lateral type).

The lining of dentigerous cysts (probably originating from external enamel epithelium) is typically a thin, regular layer, some two to five cells thick, of nonkeratinized stratified squamous or flattened/low cuboidal epithelium without a defined layer of basal cells. The inner enamel epithelium covering the crown of the tooth is usually lost. It resembles the reduced enamel epithelium from which it is derived. Also, mucous-producing cells as well as ciliated cells may be observed. Mucous cell metaplasia is common and increases with age, and epithelial discontinuities are frequently observed. The lining is supported by a fibrous or fibromyxomatous connective tissue capsule free from inflammatory cell infiltration, unless there has been secondary inflammation. Cholesterol clefts may be present and islands of odontogenic epithelial nests representing remnants of the dental lamina are occasionally observed.

Occasionally, a cyst which clinically and radiographically appears to be a typical dentigerous cyst is lined by ameloblastomatous epithelium which proliferates into the cyst lumen. Such lesions are classified as unicystic ameloblastomas.

The cyst contains a proteinaceous, yellowish fluid, and cholesterol crystals are common.

Histologic examination, however, will be decisive in ruling out these possibilities among which keratocyst and unicystic ameloblastoma are the most prevalent. Moreover, the radiologic picture of the dentigerous cyst may be mimicked by hyperplasia of the dental follicle, the connective tissue capsule that surrounds the unerupted tooth. Fibromyxomatous areas in the connective tissue wall of the dentigerous cyst may resemble the odontogenic myxoma. The presence of odontogenic epithelial rests may lead to the erroneous

diagnosis of one or another type of epithelial odontogenic tumour. However, identification of the epithelial cyst lining will rule out these alternatives.

Management principles. Treatment requires removal of the unerupted tooth, the cyst being delivered at the same time. Recurrence is rare. If the tooth is in a favourable position and space is available, it may occasionally be possible to marsupialise a dentigerous cyst to allow the tooth to erupt. Alternatively, the tooth can be transplanted to the alveolar ridge or extracted, as appropriate, and the cyst enucleated.

ERUPTION CYSTS

An eruption cyst occasionally forms over a tooth about to erupt. The eruption cyst is a specific type of dentigerous cyst located in the gingival soft tissues. An eruption cyst is, strictly, a soft tissue cyst in the gingiva overlying the crown of an unerupted tooth. It is probably a true superficial dentigerous cyst which arises in an extra-alveolar location. It arises from enamel organ epithelium after enamel formation is complete and is, in effect, a superficial dentigerous cyst. Eruption cysts involve both the deciduous and permanent dentitions. Because they arise in an extra-alveolar location they present as fluctuant swellings on the alveolar mucosa and are often bluish in colour. Mostly, these cysts are short-lived, rupturing with the progressive eruption of the associated tooth.

Clinical features. Eruption cysts affect children and involve teeth having no predecessors (deciduous teeth or permanent molars). Clinically, they appear as blue-stained blisters of the oral mucosa, the blood-stained cyst content being visible through the thin membrane separating cyst content from oral cavity. The cyst lies superficially in the gingiva overlying the unerupted tooth and appears as a soft, rounded, bluish swelling. Mostly, these cysts are short-lived, rupturing with progressive eruption of the associated tooth.

Radiology. The extra-bony position of the eruption cyst means that the only radiological sign is likely to be a soft tissue mass.

Pathology. An eruption cyst is basically a dentigerous cyst in soft tissue over an erupting tooth. The histological features are similar to those of the dentigerous cyst, though reduced enamel epithelium is often seen. Thin fibrous wall with thin squamous epithelial lining deeply and oral mucosal epithelium superficially. There is variable inflammatory infiltrate in the wall. Haemorrhage into the cyst cavity is common as a result of trauma. The lining of eruption cysts may be similar to that described above but is usually modified by chronic inflammation, possibly as a result of trauma. The latter also explains why many contain blood.

Management. The cyst roof may be removed to allow the tooth to erupt, but most eruption cysts probably burst spontaneously and never come to surgery.

ODONTOGENIC KERATOCYST

The odontogenic keratocyst is a relatively uncommon lesion which has aroused much interest because of its unusual growth pattern and tendency to recur — unlike most other cysts. Odontogenic keratocyst, formerly also called *primordial cyst* but currently known as keratocystic odontogenic tumour, is defined by the presence of keratinizing stratified squamous epithelium not exceeding 10 cell layers in thickness, palisading of the basal cells, and a parakeratinised, corrugated surface.

Aetiology. Unknown. Presumably the cyst originates from primordial odontogenic epithelium (any part of dental lamina or remnants thereof) or enamel organ before the start of amelogenesis. A tooth is sometimes missing. The aetiology of keratocysts is therefore speculative.

Incidence. About 10 % of odontogenic cysts. Odontogenic keratocysts occur over a wide age range, but there is a pronounced peak incidence in the second and third decades with a second smaller peak in the fifth decade (bimodal age distribution — 2nd–3rd decades and 5th decade). The cysts are more common in males than females (the male to female ratio is about 1.5 to 1). The mandible is usually affected. 70–80 per cent of keratocysts occur in the mandible (especially near angle of mandible). At least 50 % of keratocysts form in the angle of the mandible, extending forwards into the body and upwards into the ramus.

Pathogenesis. Keratocysts probably arise from any of the primordial epithelia (the dental lamina or its remains) or, as originally believed, from the enamel organ before tooth formation. However, it is difficult to reconcile such origins with the appearance of keratocysts in middle age and their relatively rapid epithelial turnover. Radioactive labelling used to estimate mitotic activity suggests that keratocyst lining may be proliferating more actively than mucosal epithelium. Mural growth therefore appears to be more important in the enlargement of keratocysts than osmotic pressure exerted by the cyst's contents. This active mural growth and epithelial proliferation is probably a factor determining the frequency with which keratocysts recur.

Two types of keratocyst are recognised. They have slightly different demographic characteristics (table 2), but the important difference is that parakeratotic cysts are far more likely to recur than orthokeratotic cysts.

Important features of keratocysts

Feature	Parakeratotic	Orthokeratotic
Relative frequency, %	88	12
Sex, %		
Male = 6 1.5 % overall	62	57
Female = 38.5 % overall	38	43
Age at presentation, years	34	40
Association with impacted tooth, %	48	76
Midline location, %	6	16
Pain, %	15	9
Radiographic appearance	Usually multilocular	Frequently monolocular
Recurrence rate, %	43	4

Growth of the odontogenic keratocyst. The odontogenic keratocyst has an aggressive pattern of growth, burrowing through cancellous bone in a predominantly anteroposterior direction. Growth of keratocysts is by extension of finger-like processes into marrow spaces rather than by expansion. As noted previously, this distinguishes it from the other odontogenic cysts which tend to expand equally in all directions, that is, in a unicentric ballooning pattern. The unique pattern of growth of the odontogenic keratocyst, in comparison to the other cysts, suggests that different mechanisms of enlargement are involved. The major factors are:

1. *Active epithelial growth.* The epithelial lining of keratocysts shows a higher rate of mitotic activity than other odontogenic cysts. Growth of the wall being greater than expansion of the cyst cavity, causes the lining to become much folded. The proliferation is not uniform but tends to occur in clusters, which may account for foldings in the cyst lining and projections of the cyst into cancellous spaces resulting in a multicentric pattern of growth.

2. *Cellular activity in the connective tissue capsule.* Active growth of the capsule occurs in association with the proliferating areas of the epithelium. Osteoclasts tend to be located around the tips of the projections of the lining which are proliferating into the cancellous spaces.

3. *Production of bone resorbing factors.* Like the radicular cysts the odontogenic keratocyst releases bone resorbing factors, including prostaglandins, collagenase, and interleukins-1 and -6. However, it is likely that this is the result of the focal rather than uniform pattern of growth activity of the cyst wall, described above. The aggressive behaviour of keratocysts indicates that they are effective resorbers of bone.

Increase in intracystic pressure, due to the hypertonic nature of the contents of keratocysts or to the accumulation of squamous debris within the lumen, is unlikely to be a significant factor in cyst expansion, and would not account for the biological behaviour of the lesion.

Clinical features. Keratocysts, like other jaw cysts, are symptomless until the bone is expanded or they become secondarily infected, and this probably accounts for why some do not present until the fifth decade. Unlike radicular and dentigerous cysts which tend to expand in a unicentric ballooning pattern, keratocysts enlarge predominantly in an anteroposterior direction and can reach large sizes without causing gross bony expansion. They are often discovered fortuitously on routine radiographic examination.

The majority of keratocysts arise sporadically and present as solitary lesions, although in a few patients two or more cysts may develop. The main difference is that expansion of the jaw is much less than the radiographic extent of the cyst. Hence clinical signs often fail to appear until the cyst is well advanced, and relatively extensive cysts are occasionally found by chance in radiographs.

An important clinical feature of keratocysts is their tendency to recur after surgical treatment. Recurrence rates vary in different reported series from around 3 per cent to about 60 per cent. It is likely that the rate is decreasing with improved management following recognition of this problem. Possible factors related to recurrence are discussed later.

Radiographic features. Radiographically, keratocysts appear as well-defined radiolucencies, either more or less rounded, often with densely corticated margins. The bony wall appears sharply demarcated. The shape margins may be “scalloped” in shape. That may be unilocular or multilocular and simulating an ameloblastoma. Occasionally, a keratocyst may envelop an unerupted tooth and be indistinguishable radiographically from a dentigerous cyst. May resorb or displace roots of adjacent teeth. Keratocysts may also present as developmental lateral periodontal cysts. Expansion typically limited, with a propensity to grow along the medullary cavity

Histopathology. The cyst wall is usually thin and often folded and is lined by a regular continuous layer of stratified squamous epithelium some five to ten cells thick. The basal cell layer is well defined and consists of palisaded columnar or occasionally cuboidal cells. The suprabasal cells resemble those of the stratum spinosum of oral epithelium and there is an abrupt transition between these and the surface layers which differentiate towards keratin production. Parakeratosis predominates but areas of orthokeratinization are occasionally seen. The cells desquamate into the cyst lumen. Mitotic activity is higher than in other types of odontogenic cysts and mitotic figures are found in basal and suprabasal cells.

The fibrous capsule of the cyst is usually thin and generally free from inflammatory cell infiltration. If the cyst becomes secondarily inflamed the epithelial lining loses its characteristic histology and comes to resemble that of a radicular cyst. Small groups of epithelial cells resembling dental lamina rests are often found in the capsule and these can give rise to independent

satellite cysts (daughter cysts) around the main lesion. Satellite cysts are usually small and often microscopic in size.

Keratocysts contain thick, grey/white cheesy material consisting of keratinous debris. There is little free fluid. The term odontogenic keratocyst must not be used to describe any odontogenic cyst producing keratin, it refers to a specific clinicopathological entity. Other jaw cysts such as radicular and dentigerous cysts may, rarely, produce keratin by metaplasia, but the epithelial linings of such cysts are usually orthokeratinized and do not show the regular and ordered epithelial differentiation that characterize the odontogenic keratocyst.

Orthokeratinization is seen in a minority (about 30 %), occasionally with keratin forming semisolid cyst contents. The epithelium is typically much folded and tends to separate from the fibrous wall. Daughter cysts are occasionally seen in the cyst wall and may account for some recurrences. An inflammatory infiltrate is typically absent but infection and inflammation cause the lining to resemble that of a periodontal cyst.

Management. The diagnosis should preferably be confirmed before operation, both to exclude an ameloblastoma and also because of the problem of removing keratocysts entirely. Aspiration is unlikely to be helpful. Squames in the cyst fluid are not in themselves diagnostic as other cysts can form keratin and, very rarely, a carcinoma with cystic change may shed squames into its interior. Keratocyst fluid has a low soluble protein content but, in practice, biopsy confirms the diagnosis more reliably and quickly.

On confirmation of the diagnosis, treatment should be by complete enucleation. This is usually difficult as the lining is friable (particularly if inflamed) but treatment should be thorough, to try to be certain that every fragment of lining has been removed. Large cysts are treated by marsupialisation and packing; over time the cyst shrinks in size and may disappear completely. Resection provides the lowest recurrence rate but at the cost of considerable morbidity. Enucleation alone is likely to lead to a recurrence rate of up to 60 %. However, treating the cyst lining with Carnoy's fluid improves the results of enucleation. The purpose is to fix the lining and make it easier to remove intact.

Preliminary decompression also improves the prognosis.

Recurrence. High recurrence rates are reported (up to 60 %) because of technical difficulty in removing all of the cyst lining, including projections into cancellous bone. A major factor leading to recurrence is the difficulty in removing every trace of the epithelial lining. Its more vigorous proliferative activity than that of other cyst linings may allow a few remaining epithelial cells to readily form another cyst. Formation of another cyst from other dental lamina remnants is not strictly a recurrence but the fact that recurrences can appear up to 40 years after enucleation suggests this may happen. Otherwise

recurrence is often within the first 5 years after treatment. Vigorous treatment is likely to reduce the risk of recurrence, but there is no absolute certainty of a cure, and patients should be followed up with regular radiographic examinations. Recurrence rates are affected by various factors.

Possible reasons for recurrence of keratocysts:

1. Thin, fragile linings, difficult to enucleate intact.
2. Fingerlike cyst extensions into cancellous bone.
3. Daughter cysts sometimes present in the wall.
4. More rapid proliferation of keratocyst epithelium.
5. Other dental lamina remnants may produce another keratocyst (pseudo-recurrence).
6. Inferior standard of surgical treatment.

Epithelial residues and satellite cysts are more common in cysts associated with the naevoid basal cell carcinoma syndrome. Retention of epithelial residues or satellite cysts when the main lesion is enucleated is one of the factors associated with the high recurrence rate of keratocysts. The thinness of the cyst wall and its low tensile and rupture strength compared with radicular cysts make enucleation more difficult and recurrence may thus follow retention of fragments of torn lining. Recurrences (20 %) are due to small pieces of lining and/or daughter cysts that remain following curettage.

As there is sufficient evidence that this lesion actually represents a cystic neoplasm, the most recent WHO classification proposes the diagnostic designation *keratocystic odontogenic tumour*.

GINGIVAL CYSTS

Gingivalcysts are of little clinical significance. Gingival cysts are divided in those occurring in adults and in infants. Both, as the name already implies, are located in the gingival tissues.

Gingival cysts of infants (Dental lamina cysts of the newborn, Epstein's pearls, Bonn's nodules).

Up to 80 % of newborn infants have small nodules or cysts in the gingivae, due to proliferation of the epithelial rests of Serres. Most resolve spontaneously. They are common in neonates when they are often referred to as Bohn's nodules or Epstein's pearls. Gingival cysts of infants occur either as single or multiple cysts on the edentulous alveolar ridge of the newborn infant. Most disappear spontaneously by 3 months of age. They arise from remnants of the dental lamina which proliferate to form small keratinizing cysts.

Cysts may also arise from nonodontogenic epithelium along the midpalatine raphe. When occurring at the midpalatal raphe, they are known as palatal cysts of infants. These tiny lesions, usually not larger than 3 mm, arise from dental lamina rests and disappear spontaneously within a short time.

Histologically, they resemble epidermoid cysts. These may enlarge sufficiently to appear as creamy-coloured swellings a few millimeters in diameter, but also resolve spontaneously in a matter of months.

GINGIVAL CYSTS OF ADULTS

Gingival cysts are exceedingly rare. Possibly they arise from dental lamina rests. They may be multiple. They usually form after the age of approximately 40. They occur most frequently in females and in the interpremolar region of the mandible. Clinically, they form dome-shaped swellings less than 1 cm in diameter and sometimes erode the underlying bone. Histologically, they are lined by very thin of one to three cell layers, flat, stratified squamous epithelium and may contain fluid or layers of keratin. It is likely that most represent developmental lateral periodontal cysts (see below) that have arisen in an extra-alveolar location. Possibly, both lesions are the same, only differing in location. They are unlikely to recur after enucleation.

LATERAL PERIODONTAL CYSTS

These uncommon intraosseous cysts are developmental and form beside a vital tooth or between the roots of vital teeth. They arise from odontogenic epithelial remnants: fragments of the dental lamina, reduced enamel epithelium, or rests of Malassez. Inflammation does not play a role in their development. They are usually seen by chance in routine radiographs and resemble other odontogenic cysts radiographically, apart from their position beside a tooth, near the crest of the ridge. They cause no symptoms unless they erode through the bone to extend into the gingiva. The developmental lateral periodontal cyst is an uncommon lesion that must be distinguished from a lateral radicular cyst associated with a non-vital tooth and from an odontogenic keratocyst arising alongside the root of a tooth.

Microscopically, the lining is squamous or cuboidal epithelium, frequently only one or two cells thick, but sometimes with focal thickenings to five cell layers. Its most typical feature are focal, plaque-like thickenings that consist of the cells may have clear cytoplasm that may contain glycogen and resemble those seen in the dental lamina. The underlying connective tissue does not show prominent inflammatory changes. The lateral periodontal cyst may be confused with other cysts that may lie lateral to the root of the tooth. The most important of them is a radicular cyst not occurring at the root tip but at the opening of an accessory canal. However, in case of lateral periodontal cysts, the involved tooth is vital and in case of radicular cysts by definition not. Moreover, acknowledging the specific histology of the lateral periodontal cyst will allow separation from the radicular cyst. The same histology serves to

distinguish the lateral periodontal cyst from other cystic or solid lesions that may manifest themselves as paradental radiolucencies. Occasionally, developmental lateral periodontal cysts are multilocular and may be described by the adjective “botryoid” because of their resemblance to a bunch of grapes (botryoid odontogenic cyst).

The cyst should be enucleated and the related tooth can be retained if healthy.

BOTRYOID ODONTOGENIC CYSTS

The botryoid odontogenic cyst is probably a variant of, and even more rare than, the lateral periodontal cyst with the same clinical and histologic appearance. It typically affects the mandibular premolar to canine region in adults over 50.

Microscopically, it is typically multilocular with fine fibrous septa. The lining consists of flattened non-keratinised epithelium interspersed with clear, glycogen-containing cells and sporadic budlike proliferations protruding into the cyst.

Treatment by curettage is the most appropriate treatment. It should be enucleated or conservatively excised as it has a strong tendency to recur, sometimes after a long time interval.

GLANDULAR ODONTOGENIC CYST

The glandular odontogenic cyst, also called sialo-odontogenic cyst, is a rare, developmental odontogenic cyst. Most have occurred in the anterior part of the mandible where they present as a slow-growing, painless unilocular or multilocular radiolucency.

The glandular odontogenic cyst is another rare entity with many features in common with botryoid odontogenic cysts but there are pools of mucin and mucous cells in variable numbers within the epithelium. Glandular odontogenic cyst represents a cystic lesion that is characterised by an epithelial lining with cuboidal or columnar cells both at the surface and lining crypts or cyst-like spaces or ductal lumina within the thickness of the epithelium. The lesion is very rare, accounting for only 0.04 % in a large series of jaw cysts. The lining epithelium partly consists of nonkeratinising squamous epithelium of varying thickness with focal thickenings similar to the plaques in the lateral periodontal cyst and the botryoid odontogenic cyst. This stratified epithelium may have a surface layer of eosinophilic cuboidal or columnar cells that can have cilia and sometimes form papillary projections. Some of these superficial cells may assume an apocrine appearance with cytoplasmic fragments budding off into

the cyst lumen. Also, mucus-producing cells may be present, either at the epithelial surface or as clusters within the epithelial lining.

Glandular odontogenic cyst is rare but may superficially mimic a central mucoepidermoid carcinoma. Features include epithelial whorls, cuboidal eosinophilic cells, goblet cells, ciliated cells and mucous pools which, with expression of p53 and Ki67, may aid the diagnosis. It is frequently also multilocular, tend to be aggressive or locally invasive nature and has a strong tendency to recur. Multilocularity not necessarily visible in radiographs.

Focally, the epithelium shows areas of increased thickness in which glandular spaces form which may be lined by the various above mentioned cell types.

Focally, the epithelial cells may be arranged into spherical structures with a whorled appearance. Mucous cells and cuboidal cells with cilia may occur in many other jaw cysts, in particular in the dentigerous cyst. Therefore, in the absence of the other histologic features, their mere presence should not lead to the diagnosis of glandular odontogenic cyst.

Mucoepidermoid carcinoma is the major alternative that should be considered. The combination of nonkeratinising squamous epithelium and mucus-producing cells also characterizes this salivary gland neoplasm that may occur intraosseously. However, epithelial plaques consisting of clear cells are not a feature of this latter lesion. When they are not found, it may be impossible to make the distinction. The glandular odontogenic cyst most commonly affects the body of the mandible and the most prominent symptom is painless swelling. Treatment may be conservative but recurrence may occur.

Other cysts: a large variety of cysts can occur in the jaws. Some will be developmental cysts unrelated to teeth (nasopalatine duct cyst, nasolabial cyst, dermoid cyst), others will be associated intimately with the odontogenic apparatus and will be developmental (lateral periodontal cyst, gingival cyst of adults, glandular odontogenic cyst) or inflammatory in nature (paradental cyst). In addition, samples from a periodontal pocket or inflamed dental follicle can mimic cystic lesions. Of these only the glandular odontogenic cyst is likely to recur because of the presence of daughter cysts in its wall.

NON-ODONTOGENIC CYSTS

Nasopalatine duct cyst (incisive canal cyst; median palatine cyst, midline cyst of anterior maxilla).

The nasopalatine duct cyst is a distinct clinicopathological entity and is the commonest of the nonodontogenic cysts. The nasopalatine duct connects the organ of Jacobson in the nasal septum to the palate in many animals. Jacobson's organ is joined centrally to an accessory olfactory bulb. Jacobson's organ has disappeared in man. In man, vestiges of a primitive organ of smell in

the incisive canal can be found in the form of incomplete epithelium-lined ducts, cords of epithelial cells, or merely epithelial rests. These cells can give rise to nasopalatine duct cysts.

Aetiology. It is a developmental lesion thought to arise from epithelial remnants of the nasopalatine duct which connects the oral and nasal cavities in the embryo. The stimulus for cystic change is unknown.

Incidence. Nasopalatine cysts are uncommon. The cyst presents most commonly in the fifth and sixth decades and occurs more frequently in males than in females.

Clinical features. It may be asymptomatic and be discovered on routine radiographic examination, or present as a slowly bluish enlarging swelling in the anterior region of the midline of the palate. Occasionally, it discharges into the mouth when the patient may complain of a salty taste. Pain may occur if the cyst becomes secondarily inflamed. Although cysts may arise at any point along the nasopalatine canal, most originate in the lower part and some arise entirely within the soft tissue of the incisive papilla. If allowed to grow sufficiently large, nasopalatine cysts may cause a swelling in the midline of the anterior part of the palate, particularly when superficial (so-called palatine papilla cyst). Radiologically, they present as radiolucent lesions situated

Radiographic features. Radiographically, nasopalatine duct cysts present as well-defined round or ovoid radiolucencies, often with a sclerotic rim in the anterior part of the midline between the roots of both maxillary central incisor teeth. Sometimes it appears to be “heart-shaped” because of superimposition of the anterior nasal spine. They are usually symmetrical about the midline but some are displaced to one side. The cyst must be distinguished from the normal incisive fossa and although precise limits cannot be placed on the maximum size of the latter, it is generally accepted that a radiolucency not greater than 6–8 mm wide may be considered within normal limits. Radiological assessment should include examination of the lamina dura of the central incisors (to exclude a radicular cyst) and assessment of size (the nasopalatine foramen may reach a width of as much as 8 mm). Where there are standing teeth, the lesion must also be differentiated from a radicular cyst.

Histopathology. The cysts may be lined by a variety of different types of epithelium. Stratified squamous epithelium, pseudostratified ciliated columnar (respiratory) epithelium often containing mucous cells, cuboidal epithelium, or columnar epithelium may be seen alone or in any combination. Sometimes salivary acini may be found in the cyst wall. The epithelium is supported by a connective tissue capsule which usually includes prominent neurovascular bundles from the terminal branches of the long sphenopalatine nerve and vessels. Collections of mucous glands and a scattered chronic inflammatory cell infiltrate are frequently present.

Treatment. Nasopalatine duct cysts should be enucleated. As surgical treatment comprises emptying the nasopalatine canal, the specimen always includes the artery and nerve that run in this anatomic structure. These are seen within the fibrous cyst wall and form the most convincing diagnostic feature, as the specific epithelial structures may be obscured by inflammatory changes. Recurrences are rarely seen, and are probably due to incomplete removal.

NASOLABIAL CYST

The nasolabial cyst is a rare lesion which forms outside the bone and arises in the soft tissue of the upper lip just below the ala of the nose, deep to the nasolabial fold. It probably arises from remnants of the lower part of the embryonic nasolacrimal duct and is occasionally bilateral. It is traditionally grouped with the jaw cysts previously regarded as fissural lesions. The majority of cases present in the fourth decade and over 75 per cent occur in women.

Clinically, it presents as a slowly enlarging soft-tissue swelling obliterating the nasolabial fold. If allowed to grow sufficiently large, the cyst produces a swelling of the upper lip and distorts the nostril.

Radiography may reveal nothing, as the nasolabial cyst is a soft tissue lesion. However, radiography will be performed to exclude other causes of the swelling.

The cysts are usually lined by non-ciliated pseudo-stratified columnar epithelium but stratified squamous epithelium (through squamous metaplasia), mucous cells, and rarely ciliated cells may also be present. Apocrine metaplasia of the cyst lining has also been reported.

Treatment consists of enucleation, but simple excision occasionally may be complicated if the cyst has perforated the nasal mucosa and discharged into the nose.

SURGICAL CILIATED CYST

Surgical ciliated cysts arise from detached portions of the mucosa that line the maxillary antrum and are buried within the maxillary bone. This may occur after trauma or surgical intervention in this area. Mostly, the cyst is an incidental radiographic finding, observed as a well-defined unilocular radiolucency adjacent to the maxillary antrum. The cyst lining is similar to the normal mucosal surface of the paranasal cavities: pseudostratified ciliated columnar epithelium with interspersed mucous cells. Treatment consists of simple enucleation.

PSEUDOCYSTS OR NON-EPITHELIALIZED PRIMARY BONE CYSTS

Non-epithelialized bone cysts occur most often in long bones but are occasionally seen in the jaws, almost exclusively in the mandible.

SOLITARY BONE CYST

The solitary bone cyst, also known as simple bone cyst, traumatic bone cyst, haemorrhagic bone cyst, and unicameral bone cyst. Its pathogenesis is ill-understood; a remnant of intraosseous haemorrhage is the most favoured hypothesis.

Pathogenesis. The pathogenesis of the solitary bone cyst is unknown. It is commonly believed that there is a relationship to trauma, but the evidence is not convincing. Although a history of trauma can be elicited in about 50 per cent of cases, the interval between trauma and discovery of the lesion can range from months to years and the apparent relationship may be purely fortuitous. It has been suggested that the solitary bone cyst, aneurysmal bone cyst, and central giant cell granuloma of bone are related lesions reflecting some haemodynamic disturbance in medullary bone. In the case of the solitary bone cyst it has been argued that trauma produces intramedullary haemorrhage which, for unknown reasons, fails to organize and that cavitation occurs by subsequent haemolysis and resorption of the clot.

Clinical features. The solitary bone cyst occurs predominantly in children and adolescents with a peak incidence in the second decade. There is no definite sex predilection although some series have shown a slightly higher incidence in males. The cyst arises most frequently in the premolar and molar regions of the mandible. Maxillary lesions are rare. The majority of solitary bone cysts are asymptomatic and are chance radiographic findings; some degree of bony expansion occurs in about 25 per cent of cases.

Radiographic features. Radiographs show a cavity that varies from less than 1 cm in diameter to one that occupies the entire mandibular body and ramus. Radiographically, the lesion presents as a radiolucency of variable size and irregular outline. The solitary bone cyst appears as a well-defined but non-corticated radiolucency. Typically, it has little effect on adjacent structures and 'arches' up between the roots of standing teeth. The margins of the lesion are usually well defined. The inferior dental canal may not be displaced, but the cortical margins of the canal may be lost where it overlies the lesion. Expansion is rare.

Pathological features. Surgical exploration is undertaken to confirm the clinical diagnosis and characteristically reveals a rough bony-walled cavity devoid of any detectable soft-tissue lining. At surgical exploration, one encounters a fluid-filled cavity. In many cases the cavity appears empty, but in

others there is a little clear or blood-stained fluid. Rapid healing follows surgical exploration but even without surgical intervention the cyst will resolve spontaneously with time.

Material for histologic examination may be difficult to obtain as a soft tissue lining of the bony cavity may be entirely absent or very thin. If present, it usually consists only of loose fibrovascular tissue, although it may also contain granulation tissue with signs of previous haemorrhage such as cholesterol clefts, macrophages loaded with iron pigment (haemosiderin) and multinucleate giant cells. Sometimes, this cyst develops simultaneously with a variety of fibro-osseous cemental lesions. There is no epithelial lining.

Treatment. These bone cysts are often incidental findings on radiographs. Unlike true cysts, solitary bone cysts probably heal spontaneously. The cavity should be opened only to confirm the diagnosis. Aspiration may reveal clear fluid or air indicating that no further intervention is necessary. The resulting bleeding into the cavity causes it to heal, i.e. these cysts are not caused by bleeding into the bone.

ANEURYSMAL BONE CYST

The aneurysmal bone cyst is rare in the jaws. It arises either as a primary lesion, or as a secondary change in some other preexisting disorder of bone. Secondary aneurysmal bone cysts in the jaws possibly is a developmental defect or a result of bleeding into, or vascularization of, a pre-existing lesion such as a fibro-osseous lesions and central giant cell granuloma.

Clinical features. Most primary aneurysmal bone cysts have arisen in the mandible, usually the posterior part of the body or angle, and have occurred in children or young adults. It presents as a firm expansile swelling causing facial deformity and may be associated with pain.

Radiology. The aneurysmal bone cyst typically presents as a fairly well-defined uni- or multilocular radiolucency which may have a ballooned-out appearance due to gross cortical expansion. Sometimes it has a multilocular appearance because of the occurrence of internal bony septa and opacification. Marked expansion is a feature.

Pathology. Grossly, the cyst resembles a blood-filled sponge. Microscopically, it consists of numerous, non-endothelial-lined, blood-filled spaces of varying size lined by flattened cells and separated by highly vascular connective tissue septa and similar solid areas often with many multinucleated giant cells and evidence of old and recent haemorrhage are common. Sometimes the solid areas may calcify and resemble ossifying fibroma. Deposits of osteoid are also seen.

Treatment. These cysts benefit from curettage. However, they may be associated with a second pathological lesion such as a vascular malformation

which may lead to profound haemorrhage. Patients with this cyst need to be managed in hospital.

FOCAL BONE MARROW DEFECT

The focal bone marrow defect represents an asymptomatic radiolucent lesion of the jaws that contains normal hematopoietic and fatty bone marrow. It is also called *osteoporotic bone marrow defect*. This condition is mostly seen at the angle of the mandible where it reveals its presence as a radiolucency with more or less well-defined borders. Due to the lack of radiographic specificity, the lesion is usually biopsied. Then, histologic examination will reveal the presence of normal hematopoietic marrow. Of course, further treatment is superfluous.

STAFNE'S IDIOPATHIC BONE CAVITY

This is an uncommon developmental anomaly of the mandible that is included here for convenience since it may be mistaken for a cyst on a radiograph. It is a symptomless chance finding which appears as a round or oval, well-demarcated radiolucency between the premolar region and angle of the jaw, and is usually located beneath the inferior dental canal. Occasionally, the anomaly is bilateral.

The radiographic appearances are due to a saucer-shaped depression or concavity of varying depth on the lingual aspect of the mandible, which, in the great majority of cases, contains ectopic salivary tissue in continuity with the submandibular salivary gland. Sialography may be useful in identifying such salivary inclusions.

CYSTS OF THE SOFT TISSUES

With the exception of the salivary mucoceles, cysts of the oral soft tissues are uncommon. Although strictly speaking gingival and nasolabial cysts are soft-tissue lesions they are traditionally grouped with cysts of the jaws. The main types of soft-tissue cysts, including those occurring in the neck.

SALIVARY MUCOCELES

Cysts arising in connection with minor salivary glands are common. About 90 per cent of cases are of the mucous extravasation type.

EXTRAVASATION MUCOCELES

Over 70 per cent of all mucous extravasation cysts arise in the lower lip, followed by the cheek and floor of mouth. They are extremely uncommon in the upper lip. (In contrast, salivary tumours occur much more frequently in the upper lip than in the lower lip.) The cyst occurs over a wide age range but most patients are under 30 years of age and there is a peak incidence in the second decade.

Clinically, the lesion presents as a bluish or translucent submucosal swelling and there may be a history of rupture, collapse, and refilling which may be repeated. It arises as a result of extravasation of mucus from a ruptured duct and a history of trauma can often be elicited from the patient.

Microscopically, the lesion typically consists of a mucin-filled cystic cavity or cavities lined by inflamed granulation tissue. There is no epithelial lining. The extravasated mucus evokes a chronic inflammatory reaction and the wall of the cyst is infiltrated by large numbers of macrophages with vacuolated cytoplasm containing phagocytosed mucin. Similar cells are seen within the cyst lumen. The torn duct may be seen running into the lesion. In some cases the mucus is present as diffuse pools rather than being contained within a more or less discrete cyst-like space.

RETENTION MUCOCELES

In contrast to extravasation mucoceles, retention mucoceles occur most frequently in patients over 50 years of age and are almost never found in the lower lip.

They are derived from cystic dilatation of a duct and are lined by epithelium of ductal type. Because the mucus is still contained within the duct there is no surrounding chronic inflammatory reaction. Their pathogenesis is unknown but progressive ballooning of a partially obstructed duct or even spontaneous cystic change have been suggested.

RANULA

Ranula is a clinical term used to describe a swelling of the floor of the mouth which is said to resemble a frog's belly. It is not a pathological diagnosis. Histologically, most ranulae are mucous extravasation cysts. Occasionally, a ranula may extend through the mylohyoid muscle and present in the submandibular area or neck, referred to clinically as a plunging ranula.

DERMOID AND EPIDERMOID CYSTS

Dermoid cysts are developmental lesions which occur at a variety of sites in the head and neck including, occasionally, the floor of the mouth. They may present as intraoral or submental swellings. The cyst is presumed to arise from enclavement of epithelium in the midline as a result of deranged fusion of the mandibular and hyoid branchial arches.

Histologically, the cyst is lined by a regular layer of orthokeratinized stratified squamous epithelium resembling epidermis. The lumen contains keratinous debris. To be designated as dermoid, skin appendages, such as hair follicles, sebaceous and sweat glands, and erector pili muscles, must be identified in the wall of the cyst. In the absence of skin appendages the cysts are designated as epidermoid. Epidermoid cysts occurring elsewhere in the oral soft tissues are acquired rather than developmental lesions. They arise as a result of traumatic implantation of epithelium into the deeper tissues, with subsequent cystic change and expansion.

LYMPHOEPITHELIAL CYST

Lymphoepithelial cyst is the term now used to describe lesions previously classified as branchial cysts. The majority occur deep to sternomastoid or along its anterior border at the level of the angle of the mandible. It is an unusual lesion in the oral cavity, generally arising in the floor of the mouth. Histologically, the cyst is lined by stratified squamous epithelium and its wall contains wellorganized lymphoid tissue.

The cysts are of developmental origin but their pathogenesis is uncertain. Although they may be derived from remnants of the branchial arches or pharyngeal pouches it is likely that most arise from epithelium, probably of salivary origin, that becomes entrapped by lymphoid tissue. An origin from tonsillar tissue has also been suggested.

THYROGLOSSAL CYST

The thyroglossal cyst is a developmental lesion derived from residues of the embryonic thyroglossal duct, the vestigial remains of which are represented by the foramen caecum on the tongue. Intraoral cysts, in the midline of the tongue or floor of the mouth, are very rare. Most thyroglossal cysts arise in the region of the hyoid bone.

It is convenient here to mention that functioning thyroid tissue may also occur in the tongue, although examples are rare. Before excision of ectopic lingual thyroid it is important to establish that the patient has functioning thyroid tissue.

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