Surgical pathology of the mouth and jaws

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3. Cysts and cyst-like lesions of the jaws

Most jaw cysts fulfil the criteria of being pathological, fluid-filled cavities lined by epithelium. Only a few (simple and aneurysmal bone cysts) lack an epithelial lining or fluid contents, but occasionally a keratocyst has semi-solid contents of keratin. Cysts are the most common cause of chronic swellings of the jaws but few pose significant diagnostic or management problems to the competent surgeon.

The great majority of jaw cysts are odontogenic and usually radicular (periodontal) and can be recognized by the history and clinical and radiographic features. It is only rarely that the microscopic findings fail to confirm this assessment. However, it must always be borne in mind, as shown in Chapter 4, that ameloblastoma is the great deceiver. Important features of the main types of jaw cysts are summarized in Table 3.1 and the main points affecting the differential diagnosis of cysts from other cyst-like radiolucencies are summarized in Table 3.2.

Cysts of the jaws originate in several ways, as discussed in the text. However, inflammatory (periodontal) cysts, which are by far the most common, will be discussed first.

Relative frequency of different types of cyst

Several large series of cysts have been published and there is a moderate degree of agreement as follows:

Radicular	65-70%
Dentigerous	15-18%
Keratocysts	3-10%
Nasopalatine	2-5%.

Keratocysts show the widest variation because of differences in diagnostic criteria in earlier series.

Mechanisms of cyst formation

The main factors responsible for cyst development include (in varying degree):

- proliferation of the epithelial lining and connective tissue capsule

- accumulation of fluid within the cyst

- resorption of the surrounding bone and incomplete compensatory repair.

Epithelial proliferation. Young cysts in particular often show active proliferation with a thick irregular epithelium. In the case of radicular cysts, infection from the pulp chamber is the source of irritation. Oehlers (1970) confirmed that the majority of radicular cysts,

regardless of size, regress without surgical treatment once infection has been eliminated from the root canal or the tooth extracted. However, a few large cysts (residual cysts) persist after extraction of the causative tooth. Though they may ultimately regress, in terms of practical politics it is quicker to remove them surgically.

Table 3.1 Major characteristics of jaw cysts

Cyst type	*Site/relationship to teeth*	*M/F ratio*	*Age range*	*Radiographic features*
Radicular periodontal	At apex of non-vital tooth	3:2	20-50	Unilocular
Residual	Causative tooth extracted	Equal	50+	Unilocular
Paradental	Related to inflamed third molar follicle	4:1	20-29	Unilocular, over roots of third molar
Dentigerous	Upper3s. Lower 8s.	2:1	10-40	Unilocular. Contains crown of tooth.
Keratocyst (parakeratinized)	Frequently molar region. Impacted tooth in 50%±	2:1	40± (mean)	Multilocular
Keratocyst (othokeratinized)	Frequently molar region	3:2	34± (mean)	

Usually unilocular. 'Dentigerous' in 40%±

Nasopalatine
Midline. Anterior maxilla
Almost equal
40± (mean)
Unilocular. Sometimes heart-shaped

Nasolabial
Soft tissues, deep to alae nasi
1:4
30-50 (peak)
Unilocular depression of bone of labial surface of maxilla

Median mandibular
Midline between roots of vital incisors
?
?
Unilocular. Teeth vital

Calcifying odognogenic cyst
Majority in incisor-canine region
Equal
1-30 (peak)
Usually unilocular, sometimes flecked with calcifications. Associated odontoma in 25%.

As discussed later, persistent epithelial proliferation appears to be the major factor in the growth of keratocysts.

Table 3.2 Differential diagnosis of radiolucent cyst-like lesions of jaws

1. Cysts	Odontogenic
	Non-odontogenic
2. Tumours	Odontogenic
	Non-odontogenic (including metastases)
3. Tumour-like lesions	Giant cell granuloma
4. Hyperparathyroidism	Osteitis fibrosa cystica
5. Cherubism	
6. Stafne bone cavity	(Accessory salivary tissue)
7. Solitary bone cyst	
8. Aneurysmal bone cyst	
9. Anatomical structures	(Antrum, nasal airways, incisive fossa).

Hydrostatic effects of cyst fluids. Radicular and many other types of cyst tend to grow expansively in a balloon-like manner. The hydrostatic pressure within cysts appears to be about 70 mm of water and is therefore higher than the capillary blood pressure.

Earlier views were that cyst fluid contained mainly low molecular weight proteins and that the cyst wall, by acting as a semi-permeable membrane, maintained osmotic tension high enough to cause cyst expansion. More recent experiments have shown that cyst fluid is largely inflammatory exudate containing high concentrations of proteins, some (such as immunoglobulins) of high molecular weight together with cholesterol, breakdown products of erythrocytes, inflammatory cells, exfoliated epithelial cells, and fibrin. These findings are consistent with the usual presence of inflammation in cyst walls, as a consequence of which, capillaries in the cyst wall are more permeable and exude fluid into the cavity. The net effect is that hydrostatic pressure causes expansion of the cyst cavity.

Bone resorbing factors. In vitro, cyst tissue in culture can be induced to release bone resorbing factors. These are predominantly prostaglandings E2 and E3. Various cysts and tumours possibly differ in the quantities of prostaglandins produced, but it is uncertain to what extent this affects the mode of growth of cysts. Collagenase may be found in the walls of keratocysts, but its contribution to cyst growth is also unclear.

Most cysts of the jaw show a similar pattern of slow expansive growth. They differ mainly in their relationship to a tooth; this and the radiographic features are usually an adequate guide as to their nature. Even when it is impossible to decide the precise nature of a cyst, this rarely affects treatment. However, it is essential to distinguish odontogenic keratocysts and cystic ameloblastomas from other cysts. These occasionally have identical radiological appearances and diagnosis ultimately depends on microscopy.

Apical periodontal (radicular) cysts

Clinical features

The tern *radicular cyst* is used here because of possible confusion between the different types of periodontal cysts. They are the most common cause of chronic major swellings and the most common type of cyst of the jaws. They are rarely seen before the age of 10 years and are most frequent between the ages of 20 and 60 years. They are more common in men than women, roughly in the proportion of 3 to 2. More than three times as many cysts form in the maxilla as the mandible.

The vast majority of radicular cysts, like other cysts of the jaws, cause slowly progressive painless swellings. There are no symptoms until the cyst becomes large enough to be noticeable. If infected, the cyst becomes painful and may swell more rapidly partly due to peripheral inflammatory oedema.

The swelling is rounded and at first hard. Later, thinning of the bone may give rise to eggshell crackling on palpation. Finally, part of the wall can be resorbed entirely away leaving a soft, fluctuant, bluish swelling beneath the mucous membrane.

The dead tooth from which the cyst has originated is (by definition) present and its relationship to the cyst is usually obvious in radiographs.

Residual cysts

These were periapical cysts which have persisted after extraction of the causative tooth. Cysts of the jaws in older persons are usually residual cysts which are one of the most common causes of swelling of the edentulous jaw. They can cause trouble by interfering with the fit of dentures.

Residual cysts may slowly regress spontaneously if left untreated, and there is progressive thinning or even disappearance of their lining.

Lateral radicular cysts

Lateral radicular cysts are rare. They form at the side of the tooth as a result of the opening of a lateral branch of the root canal. Also rarely, the tooth is vital and the cyst appears to have resulted from inflammation in an adjacent gingival pocket; it then may be regarded as a type of paradental cyst.

Radiography

A radicular cyst appears as a rounded, clearly radiolucent area with a sharply defined outline intimately related to the apex of a tooth. There is sometimes a condensed peripheral radiopaque rim, but only if growth has been very slow and this is usually only seen in older patients.

The dead tooth from which a radicular cyst has arisen can be seen and may show an obvious carious cavity or discoloration due to pulpal devitalizagtion. Adjacent teeth usually remain vital but may become displaced a little or, occasionally, slightly mobile. Very large cysts in the maxilla may extend in any available direction, become irregular in shape and readily transgress the midline.

Infection of a cyst causes the outline to become hazy as a result of increased vascularity and resorption of the surrounding bone.

Microscopy

All stages can be seen from a periapical granuloma containing a few strands of proliferating epithelium to an enlarging cyst with a hyperplastic epithelial lining and dense inflammatory infiltrate.

The epithelial lining. The lining, derived from the epithelial rests of Malassez, is stratified squamous epithelium of variable thickness and is sometimes incomplete. Early, active proliferation of the lining epithelium is associated with inflammation and the epithelium may then be thick, irregular and hyperplastic or form rings and arcades to acquire a net-like appearance. As cysts mature, the epithelial lining becomes thinner and flatter and inflammatory cells become progressively fewer.

Though the epithelium is stratified squamous in type, it is somewhat poorly formed and lacks a defined basal cell layer. Rarely, as a result of metaplastic change, the epithelium

may keratinize or become respiratory in type with mucous and ciliated columnar cells, even in the mandible. Hyaline bodies may also be seen.

The cyst wall. The capsule consists of collagenous fibrous connective tissue. During active growth the capsule is vascular and shows a moderately heavy inflammatory infiltrate adjacent to and infiltrating the proliferating epithelium.

Plasma cells are often prominent or predominant and indicate defensive antibody production against microbial products leaking from the tooth.

Round the capsule there is osteoclastic activity and resorption of the bone of the cyst wall. Beyond the zone of resorption there is usually active bone replacement. The net consequence is that the cyst expands but retains a bony shell, even after it has extended beyond the normal contours of the jaw. Nevertheless, resorption outpaces apposition to cause the bony wall to become progressively thinner, until it forms a mere eggshell, then ultimately disappears altogether. The cyst then starts to distend the soft tissues. Long-standing cysts are often characterized by a thin-flattened epithelial lining, a thick fibrous wall and minimal inflammatory infiltrate.

Clefts. Within the cyst capsule or contents there are often needle-shaped spaces or clefts left by cholesterol dissolved out during specimen preparation. Small clefts are enclosed within attenuated foreign body giant cells and associated with extravasated red cells and blood pigment. Clefts form in the cyst wall but may extend into the cyst cavity.

Cyst fluid. The fluid is usually watery and opalescent or sometimes thicker, more viscid and yellowish. Cholesterol crystals may give it a shimmering appearance and their characteristic notched rhomboidal shape may be seen under the microscope in a smear of the fluid. In sections, the protein content of the fluid is usually seen as amorphous eosinophilic material, often containing broken-down leucocytes, foam cells distended with fat globules and clefts.

Differential diagnosis

The different radiolucent lesions which must be considered in the differential diagnosis of cysts have been summarized in Table 3.2. The main practical consideration is to differentiate radicular and dentigerous cysts from unilocular keratocysts or ameloblastomas, as discussed later.

Resorption of the apices of adjacent teeth is suggestive of a neoplasm rather than a cyst but is not diagnostic. Rarely, a metastatic deposit may produce a sharply defined area of radiolucency, though usually the outline is ill-defined and irregular. Tumours also tend to be painful and to grow more rapidly than cysts. Nevertheless, it may be difficult or impossible to distinguish them from an infected cyst in radiographs, until at operation the solid nature of a tumour becomes obvious and microscopy confirms the diagnosis.

Aspiration. The cystic nature of a radiolucent area can be confirmed by aspirating its contents by a needle inserted through the wall, under aseptic conditions. However, this fluid will not distinguish one cyst from another, nor a cystic neoplasm from a true cyst. The

presence of cholesterol crystals is of little diagnostic value. Rarely, a keratocyst may be filled with a semi-solid mass of squames and no aspirate is obtainable through a thin needle.

Treatment

Many different techniques have been advocated for the management of jaw cysts. Despite differences in detail they all depend on two basic principles - enucleation and wound closure, or marsupialization (decompression) with possible variations, namely:

- Enuclation with wound closure:

    (a) primary closure with or without bone grafting
    (b) secondary closure following initial marsupialization.

- Marsupialization (decompression):

    (a) incomplete removal of the lining and marsupialization into the mouth or antrum
    (b) complete removal of lining and marsupialization into the mouth or antrum.

Despite these various options, only enucleation and primary closure, without bone grafting, is commonly carried out. Marsupialization into the antrum may be used for large maxillary cysts with incomplete bony walls separating them from the antrum.

Enucleation and primary closure

This, the usual mode of treatment, is completely satisfactory in the vast majority of cases.

Only the affected (dead) tooth may need to be extracted or it may be root-filled and conserved. Access is made by raising a mucopeiosteal flap over the cyst and opening a window of adequate size in the overlying bone. The cyst wall is then carefully separated from the bone and the entire structure removed intact. The cyst lining should be sent complete in fixative, for histological examination.

The edges of the cavity are smoothed off and free bleeding stopped with swabs. The cavity is irrigated to check for cyst remnants. Dusting it with antibiotic powder to prevent infection of the clot is probably unnecessary. The mucoperiosteal flap is replaced and sutured in position to achieve a watertight closure. The sutures should be left in place for at least 10 days.

Advantages and disadvantages of enucleation of cysts. The cavity usually heals without complication and little after-care is necessary, and the complete lining is available for examination to confirm the diagnosis and to exclude a neoplasm.

Possible or theoretical disadvantages of enucleation include the following, but all are rare in competent hands:

- The clot filling the cavity may become infected.

- Incomplete removal of the lining may lead to recurrence.

- Serious haemorrhage, either primary or secondary, may follow.

- Apices of vital teeth or other structures may be damaged. When teeth project into a cyst cavity, covered only by the cyst lining, removal of the latter can damage the blood supply to, and kill, the teeth. Similarly, the inferior dental nerve and vessels may be directly associated with the cyst lining.

- The antrum may be opened if enucleation of a large cyst of the maxilla is attempted.

- With exceptionally large cysts, removal of sufficient bone may excessively weaken the jaw and lead to fracture. Under these circumstances the cyst can be decompressed by temporary marsupialization. When enough new bone has formed, the cyst can then be enucleated.

Generally speaking, enucleation is completely satisfactory and in competent hands can be applied even to very large cysts. There are few contraindications and they are rarely absolute.

Marsupialization

The cyst's extent must be estimated as accurately as possible using appropriately angled radiographs. With these as a guide, the cyst is opened by reflecting a mucoperiosteal flap, and a window made as large as the local anatomy allows but not such as to risk a fracture. The exposed cyst lining is excised, the cavity washed out and the lining sutured to the mucous membrane at the margins of the orifice. The object is to produce a self-cleansing cavity which becomes, in effect, an invagination of the oral cavity. The cavity is packed with ribbon gauze soaked in acriflavine emulsion, BIPP or Whitehead's varnish, or a temporary plug of gutta percha can be made. The sutures and pack are removed after a week and a permanent plug or extension to a denture is made for the same purpose. 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 bone. The operation has disadvantages, as follows:

- the orifice may close, allowing the cyst to reform

- the epithelial lining of the cyst may be friable or incomplete and cannot be sutured to the margins of the opening

- cleansing of the cavity depends on the patient, who has to be provided with a syringe to wash out the cavity after meals

- several visits are necessary to assess repair of the cavity and to decide when the plug can be removed

- the complete lining is not available for histological examination

- bony infilling may fail, particularly in the maxilla, and a permanent stagnating cavity is then left.

Paradental cysts

The paradental cyst is an uncommon inflammatory odontogenic cyst, first described by Craig (1976). It forms in relation to a partially erupted third molar tooth and is typically associated with pericoronitis. The cyst probably originates from the reduced enamel epithelium after unilateral expansion of the follicle as a result of inflammatory destruction of the adjacent alveolar bone. The cyst is attached to the amelocemental junction, usually on its buccal aspect, and rarely exceeds 2 cm in diameter. The microscopic features are not dinstinguishable from those of a radicular cyst but the tooth is vital. Most regard this cyst as an entity, but others term a variety of cysts developing beside a tooth, paradental.

Dentigerous (follicular) cysts

A dentigerous cyst surrounds the crown of an unerupted tooth and may displace it for a considerable distance. Dentigerous cysts are less common than radicular cysts, which preponderate in the ratio of at least 4 to 1. The lower third molars and upper canines are most often affected, but any permanent tooth can be involved.

Pathogenesis

Dentigeous cysts arise as a result of cystic change in the remains of the enamel organ after enamel formation is complete. Occasionally, the division between the remnants of the internal enamel epithelium and the external enamel epithelium forming the main cyst lining can be seen microscopically at the attachment of the cyst to the neck of the tooth. Though such a cyst appears to form between the layers of the reduced enamel epithelium, the layer which remains attached to the surface of the enamel is usually of negligible thickness and the enamel is virtually in direct contact with the cyst contents. The cyst lining originates from the major part of the reduced enamel epithelium and progressive growth of the cyst leads to dilatation of the dental follicle.

Factors which initiate dentigerous cyst development are not known. Inflammation does not play a significant role, but there is a strong association between failure of eruption of teeth and formation of dentigerous cysts. It is not merely that a dentigerous cyst may prevent a tooth from erupting, but dentigerous cysts predominantly affect maxillary canines and mandibular thirds molars which are particularly prone to failure of eruption.

Clinical features

Dentigerous cysts are uncommon in the first decade of life and are most often found between the ages of 20 and 50 years. They are more than twice as common in males as females.

Like other cysts, if uncomplicated they cause no symptoms until the swelling becomes obtrusive. Alternatively, a dentigerous cyst may be detected by chance in radiographs or when the cfause is sought for a missing tooth. Infection causes the usual symptoms of pain and increased swelling.

Radiography

The appearance is characteristic, namely, a well-defined cyst containing the crown of a displaced tooth. The cavity is rounded and unilocular, but occasionally, trabeculation or ridging of the bony walls appears as pseudoloculation. The slow, regular growth of these 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 example, may be pushed to the lower border of the mandible or more rarely into the neck of the condyle. Occasionally, the tooth within a dentigerous cyst is a supernumerary. A dentigerous cyst in the lower third molar region may become very large and extend into the coronoid process and condylar neck. Rarely, a keratocyst may envelop the crown of the tooth, as may an ameloblastoma, and either of these may produce a radiographic appearance exactly simulating a dentigerous cyst. Microscopy of the cyst lining is therefore essential.

Microscopy

The lining of dentigerous cyst typically consists of flattened stratified squamous epithelium in which mucous (goblet) cells are often present. The epithelium may rarely undergo metaplasia and keratinize. The structure of the cyst wall is otherwise similar to that of radicular cyst, but inflammatory changes are typically absent.

Management

Once the diagnosis has been established, it may occasionally be preferable to marsupialize a dentigerous cyst to allow the tooth to erupt if the tooth in in a favourable position and space is available. Alternatively, the tooth can be transplanted to the alveolar ridge or extracted, as appropriate, and the cyst enucleated. Usually, enucleation with removal of the tooth and primary closure is carried out.

Eruption cyst

An eruption cyst occasionally forms in the gingiva overlying a tooth about to erupt. Erutption cysts are extraosseous but are probably superficial dentigerous cysts. They are uncommon and probably account for fewer than 1% of cysts.

Clinically, eruption cysts are seen in children and involve teeth having no predecessors namely, deciduous or permanent molars. The cyst appears as a soft, rounded, bluish swelling in the gingiva overlying the unerupted tooth.

Microscopically, an eruption cyst is lined by thin, poorly formed stratified squamous epithelium separated from the oral mucosa by connective tissue.

Management

The tissue overlying the crown of the tooth may be removed, but most eruption cysts probably burst spontaneously as the tooth erupts.

Gingival cysts

Two types of gingival cysts are recognized, namely, infantile (dental lamina cyst of the newborn) and adult types.

Dental lamina cyst of the newborn (Bohn's nodules)

Up to 80% of neonates have small nodules or cysts in the gingivae, which have arisen from epithelial rests of Serres, from the dental lamina. These may enlarge sufficiently to become clinically obvious as creamy coloured swellings, a few millimetres in diameter, but rupture spontaneously and heal in a matter of months. Similar lesions (Epstein's pearls) around the junction of the hard and soft palates arise from non-odontogenic epithelium but behave in a similar manner.

Microscopically, dental lamina cysts are lined by thin stratified squamous epithelium and contain layers of desquamated keratin.

Gingival cysts of adults

Gingival cysts are exceedingly rare but typically form in the free or attached gingiva, particularly after the age of 40 years. They are also thought to arise from the rests of Serres, but their late appearance is difficult to explain. Alternatively, they may result from traumatic implantation of superficial epithelium.

Clinically, gingival cysts appear to affect males and females equally. The cysts form dome-shaped swellings, usually less than 1 cm in diameter. They may sometimes erode the underlying bone and may then be difficult to differentiate from a lateral periodontal cyst.

Microscopically, gingival cysts are lined by very thin, flat, stratified squamous epithelium and may contain fluid or layers of keratin.

Lateral periodontal cysts

These rare cysts are intraosseous and form in the bone beside a vital tooth. They are usually found in routine radiographs as they cause no symptoms unless they erode through the bone and extend into the gingiva. They differ from gingival cysts of adults in their situation and that they are twice as common in men as women.

The radiographic appearance is similar to that of other odontogenic cysts, apart from their position beside a tooth and near the crest of the alveolar ridge.

Microscopically, lateral periodontal cysts are lined by squamous or cuboidal epithelium, frequently only one or two cells thick, but sometimes with focal thickenings. Some of these cells may have clear cytoplasm and resemble those seen in the dental lamina.

Treatment

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

Botryoid and sialo-dontogenic cysts

Botryoid odontogenic cyst

This rare entity, described by Weathers and Waldron (1973), is regarded as a multilocular variant of the lateral periodontal cyst. It typically affects the mandibular premolar to canine region in adults over the age of 50 years.

Microscopically, the loculae are separated by fine fibrous septa and lined by flattened non-keratinized epithelium which forms sporadic bud-like proliferations protruding into the cyst cavity. Interspersed among the epithelial cells are clear, glycogen-containing cells.

Sialo-odontogenic cyst

This may be a variant of the botryoid odontogenic cyst. However, sialo-odontogenic cysts have been reported over a wide age range. Of the 8 patients reported by Gardner et al (1988), only three were over 50 and the youngest was 19 years.

Microscopy

Sialo-odontogenic cysts typically have:

1. An epithelial lining of variable thickness, but frequently much greater than that of a botryoid odontogenic cyst. It has a flat lower border with no inflammatory infiltrate.

2. An irregular or even papillary cystic surface to the epithelium, with a superficial layer of eosinophilic cuboidal cells and some ciliated cells.

3. Pools of mucicarmine-positive material and mucous cells in variable numbers within the epithelium.

4. Whorls or nodules of epithelial cells ('epithelial spheres') which may protrude into the cyst cavity as in a botryoid-odontogenic cyst.

Management

These cysts appear to be rather more aggressive than periodontal cysts. Two of them recurred among the 8 patients reported by Gardner et al (1988), but one responded to a further enucleation, the other to curettage of the cavity. It seems important therefore to make sure

that all traces of the epithelial lining are removed. Alternatively, the cyst can be resected, but it is uncertain whether such treatment is always necessary.

Odontogenic keratocysts

These cysts are uncommon, but are important because of their peculiarly infiltrative mode of growth and their strong tendency to recur after removal. Odontogenic keratocysts have been categorized as *orthokeratotic* and *parakeratotic* variants, as the latter, which form over 85% of keratocysts, have a considerably higher recurrence rate.

Despite the term *odontogenic keratocyst*, the majority of keratocysts, though they have a characteristic type of epithelial lining, fail to form significant amounts of keratin, while even radicular or dentigerous cysts can occasionally show metaplastic keratinization. However, the latter lack any significant tendency to recur after enucleation.

Keratocysts are thought to have arisen from parts of the dental lamina. Nevertheless, if this is so, it is difficult to reconcile the frequent appearance of keratocysts in middle age unless their growth is quite remarkably slow. The factors determining the development of these cysts are also unknown. In particular, keratocysts differ from other cysts in that proliferation of their lining rather than hydrostatic pressure is the main determinant of their growth. They also tend to extend by finger-like processes along the lines of least resistance, namely the cancellous spaces. Hence these cysts may be a considerable size before they expand the jaw and become clinically apparent. To accommodate this overgrowth, the cyst lining becomes much folded. Radioactive labelling suggests that, despite their slow growth, the lining may be prolifetating more actively than cells of the oral mucosa. This epithelial proliferation is probably a factor determining the frequency with which keratocysts recur and the difficulties in eradicating them if allowed to extend into the soft tissue. As a consequence, the lesion has been likened to a 'benign cystic neoplasm'.

Clinical features

Keratocysts account for about 5% of jaw cysts. The parakeratotic variant is considerably more common in men than women. Most series record a peak incidence in the second and third decades, but other large series have shown peaks between the fifth and seventh decades. The variation in these reported findings may be partly accounted for by different diagnostic criteria and possibly also the findings may genuinely differ from one country to another.

They are often symptomless until the bone is expanded or they become infected. The main difference is that expansion of the jaw is much less than would be expected from the radiographic extent of the cyst. Hence there are often no clinical signs until the cysts are found by chance on routine radiographs in asymptomatic patients.

The most common site, accounting for at least 80% of keratocysts, is the angle of the mandible, extending for variable distances into the ramus and forwards into the body. Extension of the cavity up the ramus, even into the neck of the condyle, is characteristic.

Keratocysts, once they have penetrated the cortex of the jaw, can continue to grow in the soft tissues and are then even more difficult to eradicate.

Radiography

Keratocysts appear as well-defined radiolucent areas. Parakeratotic cysts are typically multilocular or rounded with a scalloped margin. Orthokeratotic keratocysts are frequently unilocular. A multilocular keratocyst may be difficult to differentiate from an ameloblastoma radiographically, but the bony wall of a keratocyst is typically sclerotic and appears as a sharply demarcated cortex.

Rarely a keratocyst may envelop an unerupted tooth and simulate a dentigerous cyst. As they enlarge, keratocysts tend to displace the roots of adjacent teeth.

Microscopy

The cyst wall is thin and the lining has the following characteristic features:

1. The epithelium is of uniform thickness, typically about 7-10 cells thick without rete ridges.

2. The squamous epithelium has a clearly defined, palisaded layer of tall basal cells in the parakeratotic type, a prickle cell layer which forms the bulk of the epithelium, which is often much folded. In the orthokeratotic type the basal cell layer is cuboidal or flattened.

3. Keratin formation amounts to no more than a thin eosinophilic layer of parakeratin in the parakeratotic variant. In the orthokeratotic variant there is a well-defined granular cell layer and keratin formation may be so abundant as to fill the cyst cavity with semi-solid material.

4. Occasionally, daughter cysts or epithelial islands are present in the cyst wall, particularly when the patient has the jaw cysts, naevoid basal cell carcinoma (Gorlin-Goltz) syndrome.

5. The fibrous wall is thin and often loose or myxoid. Unlike other odontogenic cysts, the epithelial lining is weakly attached to and readily separates from the underlying connective tissue.

6. Inflammatory cells are typically absent or scanty. If significant inflammation is superimposed, the characteristic appearances of the epithelium are lost, so that the lining resembles that of a radicular cyst. Confirmation then depends on finding uninflamed areas of typical epithelium.

The chief clinical and radiographic features of these two types of cysts are summarized in Table 3.3, but in essence the main differences are in the microscopic appearances and in the fact that the parakeratotic variant appears to have the strongest tendency to recur.

Table 3.3 Important features of keratocysts

	Parakeratotic	*Orthokeratotic*
Relative frequency (approximately)	88%	12%
Sex:		
male = 61.5% overall	62%	57%
female = 38.5%	38%	43%
Age at presentation	34 yrs	40 yrs
Association with impacted tooth	48%	76%
Midline location	6%	16%
Pain	15%	9%
Radiographic appearance	Usually multilocular	Frequently monolocular
Recurrence	43%	4%.

Recurrence

The overriding clinical problem with keratocysts is their strong tendency to recur after treatment, and recurrence rates of up to 60% have been reported. In a survey of 24 orthokeratotic cysts, Wright (1981) found only a single recurrence and that the parakeratotic variant was more likely to recur. His conclusions were abundantly confirmed by Crowley et al (1992) who, in a survey of 449 keratocysts, found a recurrence rate of at least 46% in 387 parakeratinized cysts and only a single recurrence among 55 orthokeratinized cysts. Crowley et al (1992) found that 2.2% of these cysts had linings partly ortho- and partly parakeratinized and that these had a similar recurrence rate to the entirely parakeratinized cysts.

The reasons for the high recurrence rate of parakeratinized cysts are not clear, but factors which may contribhute include the following:

- keratocyst lining are thin and fragile and, particularly when the cysts are large, are very difficult to enucleate intact

- extension of the cyst into cancellous bone increases the difficulty of removing the lining

- keratocysts may have, in their periphery, satellite daughter cysts which may be left behind after enucleation of the main cyst

- evidence that the epithelial lining, particularly of parakeratinized cysts, is more vigorously proliferative than that of otbher cyst linings suggests that if a few epithelial cells remain they can readily form another cyst after enucleation of the main lesion

- if, as seems likely, keratocysts develop from remnants of dental lamina, then other remnants of dental lamina might contribute to the formation of a second lesion which appears to be recurrence.

The fact that recurrences have occasionally appeared several decades after the original operation suggests that new lesions of this sort can develop. However, the fact that cysts with mixed type linings have a recurrence rate consistent with the parakeratinized component suggests that the latter's proliferative capacity is a major determinant of recurrence.

The time of recurrence is very variable. It is typically within the first 5 years, but may, as already mentioned, be after many years. The parakeratinized variant is more likely to recur, but it seems probable, also, that the more conservarive the enucleation the greater the likelihood of recurrence.

Rigorous treatment and, whenever possible, removal intact of every trace of the lining are likely to reduce the risk of recurrence, but there is not absolute certainty of a complete cure and rarely recurrence of keratocysts has been reported in bone grafts after excision. Clearly, it is important to warn patients of the possible need for further operations and they should be followed up with regular radiological examinations at intervals for an indefinite period.

Treatment

It is desirable to confirm the diagnosis of keratocysts before operation, to dispel any doubts as to whether the lesion is an ameloblastoma. The quickest and most reliable method is by biopsy. However, once the cavity has been opened, careful inspection is usually adequate to settle the diagnosis in most cases.

Once the diagnois has been confirmed, treatment should be by complete enucleation. This is usually difficult as the lining is friable (particularly if inflamed), but treatment should be thorough, even aggressive, to try to ensure that every fragment of cyst lining has been removed. Because of these difficulties, recommendations have ranged from simple enucleation with prolonged follow-up, to jaw resection and reconstruction, but several studies have found that the recurrence rate of keratocysts is not significantly affected by the mode of treatment. The present consensus, based on several recent series, is that the cyst should be opened widely, the lining carefully enucleated and then the bony margins cut back by at least 5 mm all round with an acrylic burr. Though it appears that the orthokeratotic variant is less likely to recur, there is probably no justification for less thorough treatment than for the parakeratotic type. Prolonged radiographic follow-up is essential and recurrences are treated in a similar manner as they develop. This is the policy advocated by the present authors.

Syndrome of multiple jaw chysts, skeletal anomalies and multiple basal cell carcinomas

This syndrome, often called the Gorlin-Goltz syndrome, is inherited as an autosomal dominant trait and has the following main features:

1. Multiple keratocysts of the jaws in approximately 85% of patients.

2. Multiple, early onset basal cell carcinomas of the skin in over 50% of patients.

3. Skeletal anomalies (usually of a minor nature) such as bifid ribs and abnormalities of the vertebrae.

4. Characteristic facies with frontal and temporoparietal bossing and a broad nasal root in about 70% of patients.

5. Intracranial anomalies frequently include a characteristic lamellar calcification of the falx cerebri, an abnormally shaped sella turcica and occasionally intracranial tumours.

Many other findings have been reported and the effects on the patient depend on the syndrome's predominant manifestation. Thus in some cases there are inumerable basal cell carcinomas which can be disfiguring or troublesome in other ways. Though termed 'naevoid', these tumours differ from typical basal cell carcinomas only in their early onset and have been reported even in a child of 2.

Other patients have a great many jaw cysts necessitating repeated operations.

Microscopy

The cysts, in the great majority of patients, are of the parakeratotic type but more frequently have daughter cysts or islands of epithelium in the walls (Woolgar et al, 1987). Like keratocysts in normal persons, they are more common in the body or ramus of the mandible.

Management

Patients should be warned of the possible need for repeated operations to remove jaw cysts and of other features of the syndrome. Referral to a dermatologist or other specialists may also be necessary.

The cysts must be managed like other keratocysts and enucleated intact if possible. If large, preliminary marsupializagtion to allow shrinkage of the cyst, before enucleation, has been recommended. If neglected, these cysts can give rise to any of the complications of other jaw cysts.

Primordial cysts

Odontogenic keratocysts were originally termed 'primordial cysts', but the term has been revived to describe what appears to be a different entity. However, not all pathologists recognize the latter and regard the terms primordial and keratocysts as synonymous.

The existence of these rare cysts, as mentioned earlier, is somewhat controversial. They have been categorized as a form of follicular cyst and are believed to result from cystic degeneration of the stellate reticulum before the start of enamel formation. A tooth (usually a lower third molar) is therefore missing.

Clinically, primordial cysts are usually found early in life, often during a routine radigraphic examination, when they are seen as a sharply defined unilocular area of

radilucency close under the crest of the alveolar ridge. Growth of these cysts is slow and produces little expansion of the bone.

Microscopically, these cysts are lined by stratified squamous epithelium which may or may not keratinize, and there are cords or strands of odontogenic epithelium in the cyst wall. However, primordial cysts may be indistinguishable from the keratocysts described above - hence the doubt about their being a distinct entity.

Treatment of primordial cysts should be the same as for keratocysts, namely, thorough enucleation and curettage of the bony wall.

Calcifying odontogenic cyst

The calcifying odontogenic cyst frequently has a propensity for continued growth and forms a solid tumour in about 14% of cases.

Clinically, almost any age can be affected, but the peak incidence is between the ages of 10 and 19 years. There is no sex predilection and the maxilla or mandible are equally frequently affected (Buchner, 1991). The lesion most often forms anterior to the first molar, particularly in the incisor/canine region. Though typically intraosseous, approximately 20% of cases form in the gingival mucosa and may merely indent the underlying bone. On radiographs, the appearance is usually that of a cyst, but it may be multilocular or flecks of calcification may be seen. Erosion of the roots of adjacent teeth is an occasional feature.

Microscopy

The lining consists of squamous epithelium with cuboidal or ameloblast-like basal cells which may be palisaded. This epithelium is sometimes many cells thick and can contain areas resembling stellate reticulum. The most striking feature, however, is abnormal keratinization, producing areas of swollen, eosinophilic cells whose cytoplasm and nuclei become progressively paler (ghost cells) and eventually disappear to leave hyaline masses. The extent of this change may be such as to fill the cyst cavity. The ghost cells typically calcify in patchy fashion and in areas where this keratin-like material comes into contact with the connective tissue capsule, it excigtes a foreign body reaction.

Management

Though calcifying odontogenic cysts have a potential for continued growth, Buchner (1991) could find few reporgts of recurrence after enucleation. Enucleation should therefore be considered as the first choice, but follow-up should be maintained for a decade.

Nasopalatine, incisive canal and related cysts

These uncommon cysts form in the midline of the anterior part of the maxilla. The nasopalatine, median palatine, palatine papilla and median alveolar cysts are probably all variants of the same entity, but differ slightly in their relation to the postulated line of the nasopalatine canal.

Pathogenesis

Nasopalatine canal cysts arise from the epithelium of the nasopalatine ducts in the incisive canal. In cats, for example, there is an organ of smell communicating with the mouth through the palatine canal, and cats inhale through the mouth when identifying an odour. In man, only 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.

Clinical features

Nasopalatine cysts are slow-growing and resemble other jaw cysts clinically, apart from their site. Occasionally, they cause intermitent discharge with a salty taste. If neglected, they can grow large enough to produce a swelling in the midline of the anterior part of the palate or perforate the palate anteriorly, and form a prolabial swelling. If very superficial, they give rise to the so-called palatine papilla cyst.

Radiography shows a well-defined rounded, ovoid or occasionally heart-shaped radiolucent area, and a well-defined, often sclerotic margin in the anterior part of the midline of the maxilla. These cysts are usually symmetrical but may be slightly larger to one side.

The anterior palatine fossa must be distinguished from a small nasopalatine cyst. The maximum size of a normal fossa is up to 6 or 7 mm.

Microscopy

The lining of nasopalatine cysts is variable in character, but is usually either stratified squamous epithelium or ciliated columnar (respiratory) epithelium or both together. A few scattered chronic inflammatory cells are often seen beneath the epithelium in some parts of the cyst wall, but are not a prominent feature. Mucous glands are often also present in the wall.

A characteristic feature is the frequent presence of neurovascular bundles in the wall. These are the long nasopalatine nerve and vessels which pass through the nasopalatine canal and are often removed with the cyst.

Treatment

Nasopalatine cysts should be enucleated and recurrence is then unlikely. With exceptionally large examples, enucleation brings with it the risk of perforation of the nasal floor or formation of a palatal fistula. The site of these cysts also makes possible damage to the nerve supply to, with resulting anaesthesia of, the anterior palate or devitalization of the anterior teeth. The upper aspect of these rare, large nasopalatine cysts (or other anterior maxillary cysts) can be exposed by a Le Fort I down-fracture osteotomy to gain access to the floor of the nose and this enables complete enucleation to be carried out.

Globulomaxillary cyst

This exceedingly rare lesion has been traditionally ascribed to proliferation of sequestrated epithelium along the line of fusion of embryonic processes. It is now accepted that this view of embryological development is incorrect and there is no evidence that epithelium becomes buried in this fashion. Most so-called globulomaxillary cysts appear to be lateral periodontal cysts or keratocysts.

Nasolabial cyst

This exceedingly rare cyst is in the soft tissues outside the bone, deep to the nasolabial fold. The aetiology is unknown. The lining is pseudo-stratified columnar epithelium often with some stratified squamous epithelium. If allowed to grow sufficiently large, the cyst produces a swelling of the upper lip and distorts the nostril. Characteristically, the nasolabial fold is obliterated. Tangential radiographs often reveal a saucer-like concavity underlying the cyst.

Treatment of a nasolabial cyst is usually by simple excision, but occasionally may be complicated if the cyst has perforated the nasal mucosa and discharged into the nose.

Median mandibular cyst

This cyst was thought to be a fissural cyst which originated from entrapped epithelium during fusion of the halves of the mandible or a developmental cyst that resulted from inclusion of epithelium trapped in the central groove of the mandibular process in the 10-14 mm embryo. These concepts are untenable. The mandible forms as a single unit within the mandibular process, there is no fusion and it is not possible for epithelium to become entrapped. The median mandibular cyst may be a primordial cyst that develops from a supernumerary tooth bud or remnants of the dental lamina, but many are found to be other types of cyst on microscopy. The cyst forms in the symphyseal region and is asymptomatic unless infected. Typically, the adjacent teeth are vital.

The treatment of choice for medial mandibular cysts is enucleation, but care must be taken not to endanger the vitality of the related anterior teeth. If, therefore, the cyst is exceptionally large, marsupialization may be necessary.

Cyst-like lesions without epithelial lining

Siomple (solitary: haemorrhagic) bone cyst

Simple bone cyst is a preferable term to *solitary* as they can be multiple. It is a bony cavity, but has no epithelial lining and often no fluid content.

The pathogenesis of solitary bone cysts is unknown. The idea that these cysts ('traumatic' or 'haemorrhagic' bone cysts) resulted from injury to, and haemorrhage within, the bone of the jaw, followed by a failure of organization of the clot and of bony repair, is untenable. There is no evidence that a blow, insufficient to cause a fracture, can cause extensive bleeding within the bone nor that anything other than normal repair would follow. Blood-filled bone cavities in the jaws are left by the enucleation of cysts, but solitary bone

cysts do not arise as a complication. Further, a common form of treatment for solitary bone cysts is to open them and to allow bleeding into the cavity. Normal healing usually follows.

Clinical features

Simple bone cysts are mostly seen in teenagers, and are rare after the age of 25 years. They mainly form in the mandible, but are typically painless and frequently a chance radiographic finding, particularly when panoramic radiographs are taken. Significant expansion of the jaw is unusual. Females are affected more often than males in a ratio of about 3 to 2.

Radiography

These cavities form rounded, radiolucent areas which generally tend to be less sharply defined than odontogenic cysts, and have two distinctive features:

- the area of radiolucency is typically much larger than the size of any swelling suggests

- the cavity can arch up between the roots of the teeth and may, as a consequence, be first seen on a bite-wing radiograph.

They may also extend across the midline of the anterior mandible.

Microscopy

The cavity has a rough bony wall and such lining as there is may be thin connective tissue or only a few red cells, blood pigment or osteoclasts adhering to the surface of the bone.

There are often no cyst contents, but there may sometimes be little fluid.

Treatment

It may only be necessary to open the cavity to confirm the diagnosis. The characteristic lack of cyst fluid and the unlined bony wall are then usually enough to indicate the diagnosis, but it is preferable if possible to remove some of the connective tissue lining for confirmatory purposes. Opening of the cavity is followed by healing, probably as a result of bleeding, but natural regression is also a possibility. The fact that progressive solitary bone cysts are not seen in elderly patients also suggests that these lesions can resolve spontaneously. Spontaneous bone regeneration was seen by Saito et al (1992) in two excised specimens.

Aneurysmal bone cyst

Aneurysmal bone cysts are traditionally included with other jaw cysts, but are cysts mainly in terms of their radiographic appearance. They rarely affect the jaws. Nothing is known of the aetiology and the most acceptable possibility is that the aneurysmal bone cyst

is a vascular malformation, but they are secondary to other bone lesions in a significant minority of cases.

Clinical features

Most patients are between 10 and 20 years of age and there is a slight female preponderance. The mandible is usually affected.

The main manifestation is usually a painless swelling and a radiolucent area, which may be balloon-like or occasionally show a suggestion of trabeculation or loculation. It may expand rapidly.

Microscopy

The aneurysmal bone cyst consists of a highly cellular mass of blood-filled spaces which has been likened to a blood-filled sponge. Despite this appearance, the blood-filled spaces lack an endothelial lining identifiable by immunocytochemistry or electron microscopy. Alles and Schulz (1986) therefore suggest that the appearance is due to extravasation of red cells from the septal capillaries which lack a basal membrane to their endothelium. However, the extreme cellularity, mitotic activity and frequent presence of giant cells may lead to confusion with a sarcoma. It is essential that this mistake should not be made and the patient subjected to unnecessarily radical and possibly hazardous treatment.

An ossifying fibroma or other bone tumour may be associated. In 123 aneurysmal bone cysts below the head and neck region, Martinez and Sissons (1988) found that 36 were part of some other bone lesion, particularly a giant cell tumour or chondroblastoma.

Treatment of an aneurysmal bone cyst usually consists of thorough curettage, but excision may be preferable as the lesion occasionally recurs. The blood appears to be static, and in contrast to juvenile angiofibroma, which can appear considerably less vascular microscopically, there is little risk of haemorrhage. Cryotherapy is an alternative approach, but experience of its value is limited.

Stafne's idiopathic bone cavity

Stafne's bone cavities are symptomless and only found by chance in radiographs. They may be mistaken for cysts in trhe mandible.

They are seen as clearly demarcated round or oval radiolucencies, nearly always below the inferior dental canal in the premolar, molar or angle regions of the mandible. These cavities are concave depressions in the lingual aspect of the mandible and contain accessory lobes of the submandibular salivary gland. Very occasionally, a similar lesion is seen above the inferior dental canal and these are found to contain accessory sublingual gland tissue. Even more rarely, similar enclaved islands of salivary tissue may be found in other parts of the mandible. Intraosseous salivary gland tumours which probably arise from this ectopic tissue are discussed in Chapter 12.

No treatment is necessary for Stafne's idiopathic bone cavities and their diagnosis may be confirmed by sialography.

Cystic neoplasms

Cysts frequently form within ameloblastomas but are usually microscopic. In a few cases such a cyst can expand rapidly (probably by hydrostatic pressure) and, as a result, evelop the tumour. In some cases the gross specimen therefore appears to be a cyst in which the tumour forms no more than mural thickenings. Once fluid has accumulated within such cysts, the epithelial lining becomes flattened and may be indistinguishable from that of a non-neoplastic cyst histologically. Radiographically too, an ameloblastoma may sometimes exactly simulate a dental or dentigerous cyst. It is not surprising, therefore, that the idea has spread that neoplasms can arise within cyst linings.

Though this is an uncommon cause of confusion, it emphasizes the importance of enucleating cysts completely. This makes the whole lining available for examination to exclude the presence of any neoplastic tissue.

Neoplastic change in cyst linings

Rarely, carcinomas can arise from the epithelial ling of cysts. Eversole et al (1975) considered that 36 cases in the world literature were authentic. These are currently termed *primary intraosseous carcinoma ex odontogenic cyst* and are discussed in Chapter 4.

Benign mucosal cysts of the antrum

The benign mucosal cyst of the antrum is sometimes referred to as a mucocele or retention cyst. With the widespread use of panoramic radiography, it has become apparent that these csts are common. Many authors have reported a 5% incidence in orthopantomograms surveyed at random. The peak incidence is in the third decade and 1.5 times more males than females are affected.

Symptoms are nearly always absent until the patient becomes aware of a sensation of fullness of the side of the face, numbness or nasal obstruction.

Radiography

Antral cysts are usually single and appear as smooth, spherical or dome-shaped opacities with either a narrow or a broad base. They vary in size from 10 mm to lesions filling most of the antral cavity. Characteristically, the thin cortical line of the antral floor remains intact and is not displaced.

Radiographic follow-up shows that many of these cysts persist for a long time without enlarging, while others resolve spontaneously.

Microscopically, antral mucous cysts may either be secretory (retention) cysts and lined by respiratory or less well differentiated epithelium, or non-secretory and, like a mucous extravasation cyst of the oral mucosa, be lined by fibroblasts.

Surgical intervention of antral mucosal cysts is only necessary if they cause symptoms. They may be removed via a Caldwell-Luc approach or endoscopically through the lateral wall of the nose.

Sublingual dermoids

Dermoid cysts lined by epidermis and skin appendages are found in the floor of the mouth lingual to the mandible, usually in the midline. They produce swellings of the floor of the mouth and neck. It has been postulated that they arise from epithelium trapped between the two embryonic lingual swellings which enlarge backwards from the mandibular arch to form the anterior two-thirds of the tongue.

Enucleation is curative and recurrence rare.