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This series provides an overview of current thinking in the more relevant areas of Oral Medicine, for primary care practitioners.
The series gives the detail necessary to assist the primary dental clinical team caring for patients with oral complaints that may be seen in general dental practice. Space precludes inclusion of illustrations of uncommon or rare disorders.
Approaching the subject mainly by the symptomatic approach, as it largely relates to the presenting complaint, was considered to be a more helpful approach for GDPs rather than taking a diagnostic category approach. The clinical aspects of the relevant disorders are discussed, including a brief overview of the aetiology, detail on the clinical features and how the diagnosis is made, along with guidance on management and when to refer, in addition to relevant websites which offer further detail.

Oral Medicine:16. Radiolucencies and Radio-Opacities. C. Odontogenic Tumours

Specialist referral may be indicated if the Practitioner feels:
- The diagnosis is unclear;
- A serious diagnosis is possible;
- Systemic disease may be present;
- Unclear as to investigations indicated;
- Complex investigations unavailable in primary care are indicated;
- Unclear as to treatment indicated;
- Treatment is complex;
- Treatment requires agents not readily available;
- Unclear as to the prognosis;
- The patient wishes this.

Odontogenic tumours

Odontogenic tumours are rare, are often asymptomatic, and discovered incidentally on imaging (Table 1). They are generally slow-growing and may reach a large size before becoming symptomatic, eg:

- Swelling, sometimes with cortical perforation. Despite some odontogenic tumours expanding rather than destroying bone, there may be local invasion of surrounding bone;
- Pain due to secondary infection or pathological fracture.

They usually appear as well-defined corticated unilocular or multilocular radiolucencies but, unlike cysts, they are more likely to cause root resorption and buccal and lingual cortical expansion.

The majority of odontogenic tumours are benign. Management is surgical and is dependent upon the type of tumour and varies from enucleation to resection.

Benign odontogenic tumours

Benign odontogenic tumours are 100 times more common than malignant ones: most (>50%) are odontomas, or ameloblastomas (around 10%).

Ameloblastomas are significant since they may recur or metastasize. Composed of ameloblast-like epithelial cells arranged as a peripheral layer around a central area resembling stellate reticulum, two main histological types exist. The *follicular* type contains discrete islands (follicles) of epithelial cells: the *plexiform* type consists of anastomosing strands. Ameloblastomas predominate in the posterior mandible, presenting typically in third to fifth decades as a slow-growing, painless, uni- or multi-locular mass ('soap-bubble' appearance on imaging) (Figures 1 and 2) usually

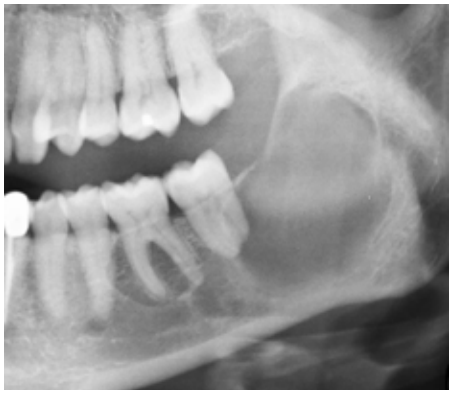

Figure 1. DPT of a well-defined multilocular radiolucency extending from the LL5 to the ramus of the mandible. Note the early apical resorption of the roots of the lower second molar. Diagnosis was an ameloblastoma.

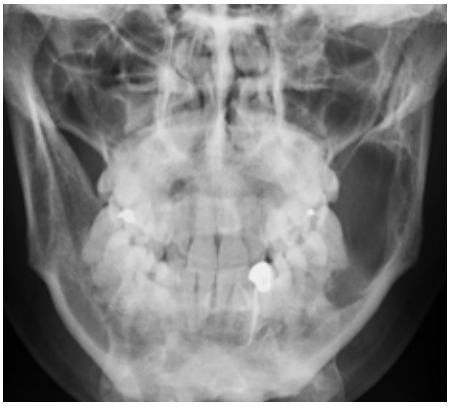

Figure 2. PA jaws of the ameloblastoma showing extensive lingual expansion consistent with a diagnosis of a solid rather than a cystic lesion.

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BENIGN **Odontogenic epithelium with mature, fibrous stroma without odontogenic ectomesenchyme**	MALIGNANT **Odontogenic carcinomas**
Ameloblastoma, solid/multicystic type	Metastasizing (malignant) ameloblastoma
Ameloblastoma, extra-osseous/peripheral type	Ameloblastic carcinoma – primary type
Ameloblastoma, desmoplastic type	Ameloblastic carcinoma – secondary type (dedifferentiated), intraosseous
Ameloblastoma, unicystic type	Ameloblastic carcinoma – secondary type (dedifferentiated), peripheral
Squamous odontogenic tumour	Primary intra-osseous squamous cell carcinoma – solid type
Calcifying epithelial odontogenic tumour	Primary intra-osseous squamous cell carcinoma derived from keratocystic odontogenic tumour
Adenomatoid odontogenic tumour	Primary intra-osseous squamous cell carcinoma derived from odontogenic cysts
Keratocystic odontogenic tumour (KCOT)	Clear cell odontogenic carcinoma
Odontogenic epithelium with odontogenic ectomesenchyme, with or without hard tissue formation	Ghost cell odontogenic carcinoma
Ameloblastic fibroma	**Odontogenic sarcomas**
Ameloblastic fibrodentinoma	Ameloblastoma fibrosarcoma
Ameloblastic fibro-odontoma	Ameloblastic fibrodentino and fibro-odonto sarcoma
Odontoma (odontome)	
Odontoma, complex type	
Odontoma, compound type	
Odontoameloblastoma	
Calcifying cystic odontogenic tumour	
Dentinogenic ghost cell tumour	
Mesenchyme and/or odontogenic ectomesenchyme with or without odontogenic epithelium	
Odontogenic fibroma	
Odontogenic myxoma/myxofibroma	
Cementoblastoma	

Table 1. WHO classification of odontogenic tumours.

Tumour type and % of odontogenic tumours	Age in years	Site	Association with unerupted teeth	Resorption of teeth	Other features
Ameloblastoma 11%	80%<40	90% mandible molar region	38% (predominantly third molar region)	39% knife edge	Multilocular; angle of mandible preserved.
Ameloblastic fibroma 2%	<20 40%<10	73% mandible molar/premolar	100%	Rare	Multilocular: less likely to destroy areas of expanded cortex than ameloblastoma.
Ameloblastic fibro-odontoma 2%	<20	Mandible = maxilla 25% anterior jaw	100%	Rare	Multilocular; very small lesions cause large amount of tooth displacement.
Adenomatoid odontogenic tumour 3%	5–50 Average 16 70% teenage	75% maxilla 90% in canine incisor region	74%	Rare	Snowflake opacities evenly arranged throughout lesion.
Calcifying epithelial odontogenic tumour 1%	30–50 peak 40	Mandible 2x maxilla Molar 3x premolar	52% rarely impacted Calcification begins	May occur knife edge	Maybe multilocular: lesion tends to extend into body rather than ramus. Less well defined than ameloblastoma.
Calcifying cystic odontogenic Tumour 2%	10–19	Mandible = maxilla 75% ant to 6	20–25%	Occasionally	Hydraulic bone expansion, expanded bone may appear perforated.
Keratocystic odontogenic tumour Commonest odontogenic tumour 3–10% of cyst like lesions	20–50	Mandible 3x maxilla			10% multiple. CT scan if sinus involved. Radiolucency in ramus not contacting any teeth most likely keratocyst. Minimal tooth displacement. Perforation of cortex may occur.
Odontogenic fibro/myxoma 3–5%	Mean 25–35	Mandible 3x maxilla Molar>premolar rare ramus	5% often congenitally absent tooth	Occasionally	May be multilocular; May destroy angle of mandible Septa intersect at 90°; may cross midline.
Cement-ossifying fibroma 40% of FCOL	15–50	80% mandible premolar and molar Maxilla, zygoma and canine		May occur	Expansion of bone equally in all directions Cortex remains intact Bowing of inferior border parallels tumour mass.
Benign cementoblastoma 9% of FCOL	<25	85% mandible 60% 1st molar 20–25% premolars		50% resorption of fusion	Only cemental lesion to be attached to root of involved tooth.
Odontomes 67%	<20	Compound: Anterior maxilla 62%. Complex: Mandible 1st 2nd molar 70%	48%	Rare	Compound 2x as common as complex.

Table 2. Summary of odontogenic tumours. Hamartomas, FCOL = Fibrous cemento-osseous lesions. Note multilocular lesions may also give rise to unilocular radiolucencies.

replacing a tooth and producing more buccolingual expansion and knife edge root resorption than does KCOT (but differentiation is difficult by plain radiography or CT). MRI may then help.

Squamous odontogenic tumour is rare and usually presents as a painless swelling and radiolucency between teeth which become mobile. It may mimic periodontal disease.

Calcifying epithelial odontogenic tumour (CEOT: Pindborg tumour) – rare, benign but aggressive. Three distinct histological features are:

- Sheets of pleomorphic epithelial cells, in places, characterized by a clear cytoplasm ('clear cells');
- Amyloid;
- Concentric masses of calcified tissues.

Usually seen in mandibular premolar or molar region associated with the crown of an impacted tooth, CEOT is radiolucent with scattered calcified components.

Adenomatoid Odontogenic Tumour – is the 'two-thirds tumour' – most commonly noted in the second and third decades of life and two-thirds of cases:

- Are in females;
- Occur in the anterior maxilla;
- Are associated with an impacted tooth (usually canine).

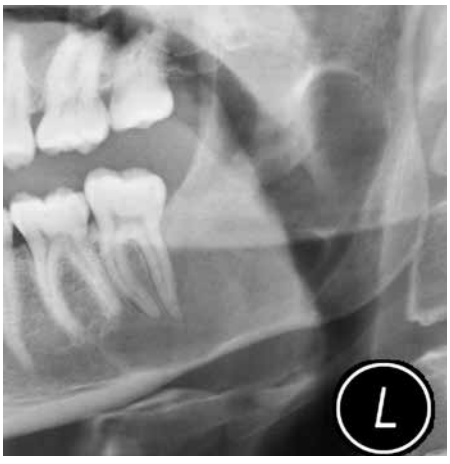

Figure 3. Sectional DPT of an odontogenic keratocyst; note the similarity in appearance to the ameloblastoma in Figure 1 but absence of apical root resorption.

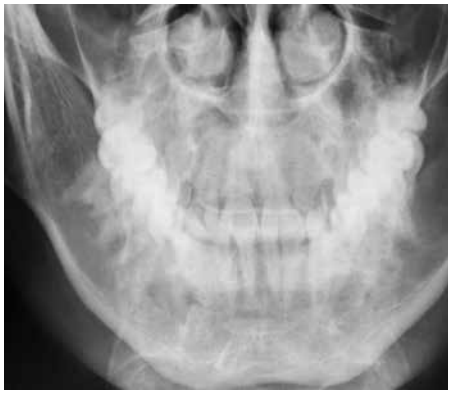

Figure 4. Section of PA jaws showing the keratocyst with little bucco-lingual expansion in relation to its size compared to the ameloblastoma in Figure 2.

Sheets and strands of epithelial cells are arranged as convoluted bands and tubular structures, in which ameloblast-like cells are arranged radially around a homogeneous eosinophilic material. It presents as a well-demarcated unilocular radiolucent lesion, often with punctate calcifications. It rarely recurs after excision.

Keratocystic odontogenic tumour (KCOT) has a propensity for destruction and recurrence locally. Radiologically, KCOT usually presents as a well-defined corticated unilocular or multilocular radiolucency which enlarges through cancellous bone, giving rise to late cortical expansion, and for its size the amount of bucco-lingual expansion is small compared to other odontogenic tumours (Figures 3 and 4). The lining has a regular keratinized stratified squamous epithelium, five to eight cell layers thick and without rete pegs. Desquamated keratin is often present within the lumen and the fibrous wall is usually thin. KCOT are often multilocular, well-defined, radiolucent, usually without an associated tooth. The keratin-rich debris shows a characteristic central signal drop on MRI T2-weighted images.

KCOT is associated with chromosome 9 patch gene mutations. Multiple KCOTs in young patients should suggest the basal cell naevus (Gorlin-Goltz) syndrome – an autosomal dominant disorder also with midface hypoplasia, frontal bossing and prognathism, falx cerebri calcification and skeletal anomalies.

Ameloblastic fibroma consists of islands, elongated strands, or terminal buds of ameloblast-like cells and central stellate reticulum cells, surrounded by a cellular hyaline material. It is usually well-defined, pericoronal multiloculated radiolucent and associated with an impacted tooth, often in the posterior mandible.

Odontogenic myxoma is clinically and radiographically indistinguishable from ameloblastoma.

Odontoma (*odontome*) – a 'hamartoma' consisting of dentine and enamel, is often associated with an impacted tooth, classified as follows:

- Compound type (Compound composite odontomes) – multiple small simple denticles embedded in fibrous connective tissue within a capsule. Multiple lesions may be seen in Gardner syndrome;
- Complex type (Complex composite odontomes) – an irregular mass of all dental tissues.

Odontomes typically present during the second decade, are more common in females than males, and often in the mandibular premolar–molar region. Typically, they can behave like teeth: they can grow and tend to erupt, or may displace adjacent teeth. Failure of a tooth to erupt is usually the justifying reason for radiographic exposure which identifies the odontome.

Cementoblastoma is a neoplasm of cementum typically seen in patients under 25 years. Usually fused to a root (typically mandibular premolar or first molar), it is a well-defined radio-opacity with a radiolucent margin. It may cause pain which responds to NSAIDs. Hypercementosis, in contrast, is smoother, less nodular, and has a thin radiolucent margin continuous with the periodontal ligament space.

Malignant odontogenic tumours

These are generally considered as the malignant counterparts of the benign categories (Table 2).

Some useful points with regard to odontogenic tumours are given in Table 3.

Normal follicle space 2.5 mm intra-oral 3 mm DPT	>2.5 80% cystic The canine has larger follicle space
Radicular cyst or granuloma	Diameter >1.6 cm or Area 200 mm
Dentigerous cysts	Lingual expansion rare
Keratocystic odontogenic tumour	10% multiple CT scan if sinus involved. Radioluency in ramus not contacting any teeth most likely keratocyst. Minimal tooth displacement. Perforation of cortex may occur.
Nasopalatine cyst Normal size of nasopalatine duct 6 x 7 mm	4:1 M:F Diameter >1cm ?cystic Normal well-defined lateral walls only
Central Giant Cell Granuloma	F:M 2:1 60% < 20 years of age. Associated with Paget's disease. Rare in ramus; usually anterior to first molar; uneven buccolingual expansion.
Aneurysmal bone cyst	90% < 30 years. Cortex remains intact even when large. Marked buccolingual expansion compared to AP.
Central haemangioma	Premature exfoliation of 1^{o} and delayed eruption of 2^{o} teeth. Phleboliths. Vertical expansion important feature
Arteriovenous malformation	Cortical thinning without perforation Sunray appearance: Phleboliths
Solitary bone cysts	50% > 3 cm. Few extend into ramus 70% have scalloped upper margin
Ameloblastoma ***The Great Mimicker***	80% < 40 years. 10–15% unilocular 38% associated with unerupted teeth MRI better for recurrence high signal on T2 weighted images Angle usually preserved Locules larger centrally
Ameloblastic fibroma	Even when small causes buccolingual expansion
Ameloblastic odontofibroma	Small lesion large tooth displacement
Adenomatoid odontogenic tumour	74% unerupted tooth. 68% canines, 70% < 20 years, F:M 2:1
Calcifying epithelial odontogenic tumour	Frequently scalloped margin. Variable definition. Expands into body rather than ramus.
Calcifying cystic odontogenic tumour	Hydrostatic expansion. Cortical perforation 96% unilocular
Odontogenic myxoma	Soap bubble appearance; expansion and perforation May destroy angle
Benign cementoblastoma	Attaches to tooth root Occlusal film sunray appearance
Cement-ossifying fibroma	Expansion inferior cortex of the mandible

Table 3. Useful points in diagnosis of radiolucent lesions of the jaws.