Odontogenic tumours

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Abstract

Odontogenic tumours are rare lesions and many non-specialist histopathologists will not have seen many during their working lifetime. The purpose of this review is to give an overview of odontogenic tumours, with an emphasis on differential diagnoses and common diagnostic pitfalls, especially for the more common tumours. It is not an exhaustive review but is intended to complement specialist texts, focussing on helpful features which pathologists may employ to increase confidence when reporting odontogenic tumours.

Keywords jaws; odontogenic; review; tumour

Introduction

Odontogenic tumours are rare and arise from the tooth-forming tissues in the jaws. The majority of odontogenic tumours are reported by specialist Oral and Maxillofacial Pathologists, compounding the rarity of these tumours in routine practice. The common odontogenic tumours, including odontoma and ameloblastomas, are largely straight-forward to diagnose. Difficulties arise when there is increased proliferation, inflammation masking the characteristic features, where lesions show features common to several odontogenic tumours, and so-called hamartomatous lesions which may reflect residual odontogenic tissues in the tooth-bearing regions of the jaws. Indeed, many odontogenic tumours mimic different stages of tooth development. This review will address these issues and give some pointers for the assessment of these lesions when they appear on the desk of a non-specialist pathologist.

Odontogenic tumours are derived from epithelial and mesenchymal elements of the tooth-forming apparatus and are found exclusively within the maxillofacial skeleton.[1] Intraosseous (central) and soft tissue (peripheral) lesions in the gingivae and alveolar mucosa may occur over a wide age range.[2] The aetiology is largely unknown. Benign lesions may present slowly with non-specific symptoms, whereas malignant lesions often cause pain and rapid swelling. Radiological imaging should always be used in conjunction with histology when making a diagnosis of an odontogenic tumour as it shows the proportions of hard and soft tissues, the borders of the lesion and its size and relationship to surrounding structures. There are few diagnostically useful immunohistochemical markers that can be utilized (especially if the immunophenotype has been affected by inflammation), but in most cases the clinical history, radiology and H&E appearance is sufficient to reach the correct diagnosis.[3]

Great care must be taken when assessing tissue from a jaw area still containing developing teeth and not showing any clinical or radiographic signs of tumour being present. Immature odontogenic tissues in these areas and at this age are plentiful and an erroneous diagnosis of an odontogenic tumour is easily made.[2]

This paper is subdivided to mirror the 2005 World Health Organisation (WHO) classification of odontogenic tumours (see Table 1).[1] As our knowledge has evolved, the current classification requires updating as the histogenesis of some tumours is controversial or unhelpful. Various authors have suggested that the classification of benign versus malignant does not reflect the reported range of behaviours and it may be better to classify lesions similar to the WHO Bone and Soft Tissue Tumour classification[4,5] which includes intermediate (locally aggressive) and intermediate (rarely metastasizing) categories. Only time will tell if this approach is adopted.

Benign

Odontogenic epithelium with mature, fibrous stroma without odontogenic ectomesenchyme

Ameloblastoma (see Table 2)

Solid/multi-cystic type (SMA) — Ameloblastoma is a slow-growing, locally aggressive tumour which has a predilection for the posterior mandible, no gender predilection and occurs most commonly in middle age (30—60 years). Large lesions may cause pain or paraesthesia. Radiographically, these lesions are often multilocular radiolucencies, sometimes causing resorption of adjacent teeth. Cortical bone resorption allows the tumour to spread freely into the surrounding soft tissues. Two main histological patterns are recognised, but this has no influence on behaviour or patient management. The follicular pattern is characterised by islands of odontogenic epithelium scattered within a mature fibrous stroma. The cells at the periphery of these islands are columnar and often palisaded, resembling ameloblasts. The central portions resemble the stellate reticulum and may become cystic (Figure 1). Several sub-types of follicular ameloblastoma occur, including spindle cell, basal cell, granular and acanthomatous. In the acanthomatous type, focal keratinization may be present which may mimic intraosseous squamous cell carcinoma. However, mitoses and pleomorphism are absent. The second major pattern is plexiform, consisting of odontogenic epithelium arranged in anastomosing cords. In both types, mitotic activity and pleomorphism are rare.

Odontogenic epithelial rests may mimic an ameloblastoma. However, they almost always lack a stellate reticulum centre that is typical for an epithelial island in ameloblastoma.[2] Moreover, they may show squamous metaplasia and dystrophic calcification and usually, the peripheral cells are not as columnar as in ameloblastoma with less nuclear polarization.[2]

In cases where distinction from an odontogenic keratocyst (OK) is difficult (such as small, cystic or cross-cut biopsies), one study found that ameloblastomas express CD56 in 97% (37/38) of cases, but was only present in 5% (1/19) of OKs.[6] In cystic lesions, the combination of widespread CD56 expression in peripheral cells and calretinin expression in the superficial cells

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Summary of benign and malignant odontogenic tumours

Odontogenic tumours				
Benign			**Malignant**	
Odontogenic epithelium with mature, fibrous stroma without odontogenic ectomesenchyme	Odontogenic epithelium with odontogenic ectomesenchyme, with or without hard tissue formation	Mesenchyme and/or ectomesenchyme with or without odontogenic epithelium	Carcinomas	Sarcomas
Ameloblastoma - solid - peripheral - desmoplastic - unicystic	Ameloblastic fibroma/ fibrodentinoma	Odontogenic fibroma	Metastasizing ameloblastoma	Ameloblastic fibrosarcoma
Squamous odontogenic tumour	Odontoma - complex - compound	Odontogenic myxoma	Ameloblastic carcinoma - primary - secondary	Ameloblastic fibrodentino- and fibro-odontosarcoma
Adenomatoid odontogenic tumour	Calcifying odontogenic cyst	Cementoblastoma	Primary intraosseous squamous cell carcinoma	
Calcifying epithelial odontogenic tumour	Odontoameloblastoma		Clear cell odontogenic carcinoma	
Keratocystic odontogenic tumour	Dentinogenic ghost cell tumour		Ghost cell odontogenic carcinoma	

Table 1

Sub-types of ameloblastoma

Histological type	Age at presentation (years)	Gender/Ethnicity	Site	Behaviour/Management
Solid/multi-cystic	30—60.	—	Posterior mandible.	Benign but locally aggressive. Excision, ensuring margin of uninvolved tissue.
Peripheral	Mean of 52.	Twice as common in males.	Gingival and alveolar mucosa. Mandible > maxilla.	Benign but less aggressive than solid/multi-cystic type. Conservative excision.
Desmoplastic	30—60.	—	Anterior mandible.	Benign but can behave more aggressively than solid/multi-cystic type. Excision with margin.
Unicystic	Mean of 16 (associated with unerupted tooth). Mean of 35 (absence of unerupted tooth).	More common in black individuals.	Posterior mandible. 80% associated with unerupted third molar tooth.	Conventional A-UT and its luminal variant are slow-growing and non infiltrative. Treat with enucleation only. The mural variant may behave in a similar way to solid/multi-cystic ameloblastoma. Management is dependent on depth of invasion into cyst wall.

Table 2

helps support a diagnosis of ameloblastoma over OK. Future studies are critical to determine which therapies will have the greatest efficacy alone or in combination, and to identify which markers carry prognostic value.[7]

Ameloblastomas should be treated with excision, ensuring an appropriate margin of uninvolved tissue. Recurrences are common especially in the posterior maxilla, where prognosis is worst.