Surgical pathology of the mouth and jaws

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4. Odontogenic tumours and tumour-like lesions

Odontogenic tissues, both ectodermal and mesenchymal, can give rise to true neoplasms, hamartomas (odontomas) and combinations of neoplasm and hamartomatous proliferation. Rather than trying to deal with these lesions in strict academic or histogenetic order, they are described in groups which have features in common.

Ameloblastoma

The ameloblastoma arises from odontogenic epithelium: it forms in the jaws or, exceedingly rarely, in the immediately adjacent soft tissues. It is the most common neoplasm of the jaws and its most striking histological feature is the ameloblastoma-like cells.

Clinical features

Most ameloblastomas are first recognized in patients aged between 30 and 60 years. They are rare in children. Eighty per cent form in the mandible; of these, 70% develop in the molar region, and often involve the ramus. They are symptomless until the swelling becomes noticeable. Small tumours may occasionally first be seen in routine radiographs. Even large tumours rarely cause paraesthesia or pain. If neglected, the tumour can perforate the bone and, ultimately, spread into the soft tissues making subsequent excision difficult.

Very rarely (fewer than 1% of cases), ameloblastomas form superficially to the alveolar ridge and cause little or no bone erosion. Extraosseous ameloblastomas typically affect middle-aged patients and resemble fibrous nodules clinically: their prominence usually leads the patient to seek treatment before they grow beyond 2 cm in diameter (El-Mofty et al, 1991).

Radiography

At least 85% of ameloblastomas form rounded, well-defined, multilocular, cyst-like radiolucent areas with well-defined margins. Variants are a honeycomb pattern of radiolucency, a soap-bubble appearance, a monolocular well-defined cavity (unicystic ameloblastoma) or rarely a few large radiolucent areas with small daughter cysts nearby. The bony margins are typically scalloped.

There is buccal and lingual expansion. The latter, seen on an occlusal radiograph, is not pathognomonic of ameloblastoma. The roots of adjacent teeth are frequently resorbed.

Differentiation of typical ameloblastomas from keratocysts, other tumours of the jaws or hyperparathyroidism by radiography alone is not possible, though the appearances are often highly suggestive.

About 13% of ameloblastomas are unicystic. They are more likely to be seen in persons under 20 years old, particularly involve the posterior part of the body of the mandible and may surround the crown of a displaced molar tooth. The radiolucent area is frequently well circumscribed, with the result that the tumour may not be distinguishable radiographically from a radicular, dentigerous or other non-neoplastic cyst.

Microscopy

Several subtypes have been described and some of these variations may be seen within limited areas of a single tumour. However, it has not been confirmed that these microscopic variations affect the behaviour.

Follicular ameloblastomas are the most readily recognizable pattern and consist of islands or trabeculae of epithelial cells in a connective tissue stroma. These epithelial processes consist of a core of loosely arranged polygonal or angular cells, resembling stellate reticulum, surrounded by a well-organized single layer of tall, columnar, ameloblast-like cells with nuclei at the opposite pole to the basement membrane. In other specimens, or other parts of the same tumour, the peripheral cells may be cut obliquely or be more cuboidal and resemble basal cells.

Cyst formation is common and varies from microcysts within a predominantly solid to a predominantly cystic tumour. Cysts may develop either within the epithelial islands or as a consequence of cystic degeneration of the connective tissue stroma.

In unicystic ameloblastomas, the cystic area expands to surround the remainder of the tumour. The latter has the structure of a typical ameloblastoma, but the lining of the cystic area becomes flattened and can closely resemble that of a non-neoplastic cyst. However, ameloblast-like columnar cells can sometimes be seen in the basal layer of the cyst lining or the latter may show budding or actual infiltration of the fibrous wall. Biopsy of the cyst wall alone may, nevertheless, lead to it being misdiagnosed as a simple cyst. This cystic overgrowth of ameloblastomas, with obliteration of the normal microscopic features of the tumour in the flattened cyst wall, is the reason for earlier beliefs that ameloblastomas can originate in non-neoplastic cysts. Only rare examples of this phenomenon have been authenticated. As a consequence, all cystic lesions should be enucleated, and the entire cyst wall, particularly any mural thickenings, examined for tumour. However, Saku et al (1991) report that cystic as well as solid ameloblastomas bind the lectins UEA-1 and BSA-1, enabling them to be differentiated from non-neoplastic odontogenic cysts which do not.

Plexiform ameloblastomas consist largely of thin trabeculae bordered by ameloblast-like or columnar cells, in a loose, vascular sparsely cellular connective tissue stroma. Cyst formation is common and frequently due to stromal degeneration.

Acanthomatous ameloblastomas show squamous metaplasia of the central core of epithelium of tumours which otherwise resemble the more common follicular type. Keratin formation may be prominent and the tumour may be mistaken for a squamous cell carcinoma.

Granular cell ameloblastomas are rare. They usually resemble the more common follicular type, but the epithelium, particularly in the centres of the tumour islands, forms

sheets of large eosinophilic granular cells resembling those of other granular cell tumours. This change may be so extensive that the peripheral columnar cells are replaced and the nature of the tumour may be difficult to recognize from a small biopsy specimen. Granular cell formation was thought to be an ageing or degenerative change but can be seen in ameloblastomas in young persons.

Desmoplastic ameloblastomas consist of only small islands of neoplastic epithelium, with few ameloblast-like cells, in a dense collagenous stroma. There is little or no cyst formation and this variant may be difficult to recognize as an ameloblastoma. It is unusual in that it has a strong tendency to form in the anterior part of the jaw and particularly in the maxilla. Most specimens are detected before they exceed 2 cm in diameter.

Basal cell ameloblastomas are the most uncommon variant and consist of more darkly staining cells predominantly in a trabecular pattern with little evidence of palisading at the periphery. Rare examples of extraosseous basal cell ameloblastomas have been mistaken for basal cell carcinomas.

These histological variations do not seem to affect the tumour's behaviour, except in so far as Hartman (1974), from a survey of 20 granular cell ameloblastomas, has suggested that they were more likely to recur than other types.

Management

The diagnosis should be confirmed by biopsy, but may only be made after operation in the case of cystic ameloblastomas, where tumour tissue may be found only in a limited area of mural thickening.

Treatment depends on the type and site of the tumour, but is usually by wide excision, preferably taking up to 1 cm of clinically normal bone around the magrin. Curettage leads to recurrence, though this may not be evident for several years. Excision can be curative, but may involve resection of the jaw and bone grafting. However, ameloblastomas are slow growing, typically infiltrate cancellous bone and only later invade compact bone. It may therefore be possible to preserve the lower border of the mandible, if free of tumour, to continue the dissection subperiosteally, and avoid complete resection and bone grafting. Bony repair follows, and a good deal of the jaw re-forms. Any residual tumour may take many years to recur, and regular radiographic follow-up should be carried out. A further limited operation can then be carried out if necessary. This approach is generally less unpleasant for the patient, who must be warned of the necessity of regular follow-up and of the possibility of further surgery. However even with *en bloc* resections, recurrence rates of up to 25% have been reported in the past.

The chief risk with mandibular ameloblastomas is extension into the soft tissues, when it becomes difficult to define the tumour margins and vital structures may be encroached.

Maxillary ameloblastomas can present considerable problems and, because of their proximity to the base of the skull and vital structures, are potentially lethal. Bredenkamp et al (1989) reported 5 such cases of which only 2 were known to be alive and disease-free after

periods of 14 months to 5 years. Four earlier cases were also reviewed: all had died or were presumed dead from their disease.

Maxillectomy should be regarded as the first requirement. Radical excision at the earliest possible stage is essential, as effective reoperation is unlikely to be possible and there is no satisfactory surgical protocol for the management of extensive tumours involving the base of the skull. Chemotherapy has not been shown to be of any value. Postoperative radiotherapy may slow the progress of recurrences, but is little more than palliative and has not been shown to be of any value as the primary treatment.

Unicystic ameloblastomas, by contrast, appear to have a considerably better prognosis. They are frequently enucleated as cysts before their neoplastic nature is appreciated, but even with this form of treatment, recurrence rates may be only 10-20%. The need for reoperation is determined by thorough examination of the specimen. Extension of the tumour through the fibrous cyst wall is an indication that recurrence is likely, but operation may be delayed until this appears. A local excision is frequently then adequate, but prolonged observation is essential.

Primary extraosseous ameloblastomas can be managed by local excision and though recurrence rates may be up to 25%, reoperation with wider local excision should be curative. By contrast, soft tissue extensions of intraosseous ameloblastomas require wide excision. However, the difficulties in defining their margins make them difficult to manage.

Odontoameloblastoma

This rare neoplasm is essentially an ameloblastoma combined with an odontoma.

Clinically, the chief differences from an amelobastoma is that it affects children and that the calcified odontomatous tissues are clearly visible in radiographs. However, its behaviour is that of an ameloblastoma, namely progressive infiltration and local destruction of surrounding tissues. The treatment is also the same as for an ameloblastoma. Too few cases have been reported to be certain about the ultimate prognosis, but rigorous long-term follow-up is clearly essential.

Malignant ameloblastoma and ameloblastic carcinoma

Both of these are exceptionally rare tumours and many pathologists may not see either in a lifetime's practice.

Malignant ameloblastoma is a histologically typical ameloblastoma which, nevertheless, has given rise to pulmonary metastases. These have retained the same microscopic appearances as the primary. Few of the reported cases have been adequately authenticated, and it has also been argued that even of the genuine cases some, at least, have resulted from aspiration implantation. If so, local excision of the secondary deposit should be curative.

Ameloblastic carcinoma refers to a tumour which initially has the microscopic features of an ameloblastoma, but which behaves in an aggressively malignant fashion, losing

differentiation and metastasizing. In the later stages the microscopic appearances can resemble a squamous cell carcinoma. Exceptionally rarely, the progressive loss of differentiation can be seen in successive specimens from the same patient and, when this happens, provides convincing evidence of the tumour's histogenesis. Also rarely, hypercalcaemia can be associated.

The problems of management are, therefore, those of an intraosseous carcinoma and if it has already metastasized, the prognosis is correspondingly poor.

Primary intra-alveolar carcinoma

Primary intra-alveolar carcinoma is a rare tumour which can arise in an odontogenic cyst. Müller and Waldron (1991) considered that this accounted for the majority of cases and traced 81 reported examples. The peak incidence is in the sixth and seventh decades and males are affected twice as frequently as females. Most cases have originated in residual cysts in the posterior mandible, but some have originated in keratocysts. Accelerated growth and pain are typical features, but neither may be obtrusive in the early stages. Radiographically, neoplastic change may be suspected in a cyst with a ragged margin, but sometimes the diagnosis is not made until after enucleation. Microscopically, the tumour is typically a well-differentiated squamous cell carcinoma.

Primary intraosseous carcinomas with no evidence of origin in odontogenic cysts are even more rare. They may originate from odontogenic rests, but it may be difficult to exclude the possibility that a carcinoma has entirely overgrown a cyst lining. To et al (1991) reported 3 cases but felt that several of 24 earlier cases were of dubious validity. They noted that, in 50% of them, diagnosis had been delayed by treatment of teeth thought to be causing the symptoms.

Clinically and radiographically, true primary intraosseous carcinomas cannot be reliably distinguished from infected cysts or other malignant tumours.

The diagnosis or primary intra-alveolar carcinoma can only be made by excluding (a) bone invasion by a mucosal carcinoma, (b) intraosseous salivary gland carcinoma (Chapter 12), (c) a metastasis, or (d) ameloblastic carcinoma.

From the viewpoint of management and prognosis, such distinctions are likely to be of considerably less significance than the extent of the tumour or the presence of metastases. However, metastases are slow to develop and are typically in the regional lymph nodes.

Treatment is preferably by radical excision, but many cases have been enucleated initially as cysts. If then carcinomatous change is found, radical excision should be carried out. The prognosis then appears to be relatively good and the 5-year survival rate appears to be 30-40%. However, data is limited by the rarity of this disease and the brevity of follow-up in many cases.

Adenomatoid odontogenic tumour

This uncommon lesion, previously known as adenoameloblastoma, is completely benign, and probably a hamartoma. It has little in common with the ameloblastoma in behaviour, and also has no glandular component.

Clinical features

Adenomatoid odontogenic tumour affects young people, either in late adolescence or as young adults, and is rare in older persons. Females are more frequently affected, in the ratio of 2 to 1. The tumour is most frequently found in the anterior maxilla and forms a very slow-growing swelling if not noticed by chance on a radiograph. It frequently has the appearances of a dental or dentigerous cyst and its nature may be unsuspected until found to be solid at operation. In an analysis of 499 examples retrieved from the world literature, Philipsen et al (1991) found that tumours containing a tooth (follicular variant) were three times as common as those that did not (extrafollicular variant), were recognized earlier at a mean age of 17 years and affected females twice as frequently as males. The extrafollcular variant was recognized at a mean age of 24 and over 50% of all types were diagnosed between the ages of 13 and 19 years. A rare extraosseous variant resembles a fibrous epulis clinically and accounted for less than 3% of cases.

Microscopy

There is a well-defined capsule and most specimens are only a few centimetres in diameter. The mass consists of whorls, strands and sheets of epithelium, among which are microcysts, resembling ducts cut in cross-section and lined by columnar cells similar to ameloblasts. These microcysts may contain homogeneous eosinophilic material. Fragments of amorphous or crystalline calcification may be seen among the sheets of epithelial cells.

Management

These lesions shell out readily and enucleation should be curative. There appear to be no authenticated recurrences.

Calcifying epithelial odontogenic tumour (CEOT)

This rare tumour, with bizarre cellular features, is often also referred to, for simplicity, as a Pindborg tumour after he first described it in 1955. Though exceedingly rare, this tumour is important, partly because of its unusual structure and because it has been mistaken, particularly in the past, for a poorly differentiated carcinoma.

Clinical features

Adults are mainly affected at an average age of about 40 years. The typical site is in the posterior body of the mandible which is twice as frequently involved as the maxilla.

Symptoms are usually lacking until a swelling becomes apparent. Radiographs show a translucent area with poorly defined margins and, usually, increasing calcification with areas of radiopacity within the tumour as it matures.

Microscopy

The tumour consists of sheets or strands of polygonal, slightly eosinophilic epithelial cells in a connective tissue stroma. The outlines of the epithelial cells are often distinct and intercellular bridges may be clearly seen. A striking feature is the gross variation in nuclear size including giant, frequently hyperchromatic nuclei and multinucleated cells. Though mitoses are rare, there is an alarming resemblance to a poorly differentiated carcinoma. However, unlike most epidermoid carcinomas, inflammatory infiltration of the stroma is typically absent.

Within the tumour, there are typically homogeneous hyaline areas, with the staining characteristics of amyloid, which may calcify. These calcifications form concentric masses in and around the epithelial cells, particularly those which appear to be degenerating, and may form large masses. Rarely, dentine or tooth-like structures may form but ameloblast-like cells are not seen.

The presence of amyloid is regarded by some as a criterion of diagnosis of calcifying epithelial odontogenic tumours. However, amyloid can be present in other epithelial tumours such as basal cell carcinomas, and it seems no more reasonable to regard a tumour product such as this as an essential feature than to dismiss the diagnosis of melanoma because of the absence of melanin production.

Calcifying epithelial odontogenic tumours are not encapsulated, are locally invasive and their behaviour seems to be similar to that of ameloblastomas.

A clear cell variant of CEOT tumour has been described, but may be difficult to distinguish from the clear cell odontogenic tumour discussed below.

Management

Diagnosis depends on histological examination and to make sure that there is no confusion with a poorly differentiated carcinoma.

Complete excision of the tumour with a border of normal bone appears to be curative. If excision is inadequate, recurrence follows.

Clear cell odontogenic carcinoma

This rare neoplasm was first described by Hansen et al (1985). It is locally invasive and destructive; lymph node and pulmonary metastases have been reported by Bang et al (1989).

Clinically, most cases have been in elderly patients (60-74 years) and have caused symptoms such as mild pain or tenderness, or loosening of teeth. The tumour causes

expansion of the jaw and a ragged area of radiolucency. Two of the three first reported cases were in the maxilla.

Microscopy

The tumour is poorly circumscribed and consists of clear cells, uniform in size with a central nucleus and a well-defined cell membrane. They form large sheets, interrupted by relatively thin strands of dense fibrous tissue stroma, in which there is no inflammatory infiltrate. There may also be lesser, dense areas of small basaloid epithelial cells with scanty cytoplasm.

A variable number of the epithelial cells may have a finely fibrillar rather than completely clear cytoplasm and may contain diastase-labile PAS=positive granules. There may be some nuclear pleomorphism but no more than occasional mitoses.

Differential diagnosis

Significant numbers of clear cells are rarely found in the CEOT, which also has a considerably more pleomorphic cellular picture than clear cell odontogenic carcinoma. The latter also lacks the calcifications and amyloid formation typical of CEOT.

Intraosseous salivary gland tumours are a well-recognized entity, though their rare clear cell variants (Chapter 12) do not appear to have been reported in the jaws and in any case salivary gland tumours affect the mandible. The most important tumour that needs to be distinguished from a clear cell odontogenic tumour is a metastasis of a renal cell carcinoma and, though these usually also involve the mandible, the differential diagnosis (Chapter 12) needs to be considered as it so seriously affects the prognosis.

Management

The behaviour of these tumours is potentially more aggressive than that of the CEOT. Wide excision may be effective in the short term, but recurrences or metastases or both have followed several years later.

Odontogenic ghost cell tumour (calcifying odontogenic cyst, Gorlin cyst)

The odontogenic ghost cell tumour is a rarity and though recognized in the jaws by Gorlin and colleagues only in 1964 (Gorlin et al, 1964), its cutaneous counterpart (benign calcifying epithelioma of Malherbe) was described in 1880 and is currently termed a pilomatrixoma. Though most frequently cystic (Chapter 3), this lesion can rarely be solid and has a potential for continued growth. The term *odontogenic ghost cell tumour* is therefore preferable. Its clinical features are similar to those of the cystic variant.

Microscopy

The appearances have much in common with the cystic variant, but tend to resemble an ameloblastoma in that processes of ameloblastoma-like odontogenic epithelium infiltrate a connective tissue stroma and frequently surround stellate reticulum-like cells. There are

variable numbers of ghost cells and deposits of dentinoid. In approximately 10% of cases, odontomas or other odontogenic tumours are associated.

Management

The capability for continued growth leads to recurrence after incomplete removal. Excision is required and is effective.

Odontogenic ghost cell carcinoma

The malignant variant of the odontogenic cell tumour is rare. An example which metastasized to lungs and other sites to cause the death of the patient was reported by Grodjesk et al (1987) who reviewed previous reports of aggressive behaviour of these tumours.

Clinically this carcinoma grows more rapidly than the benign variant.

Microscopically, the cell types in the benign and malignant ghost cell tumours are similar, but in the latter, the epithelial cells are pleomorphic, and the nuclei are frequently hyperchromatic, show mitotic activity and are retained in some of the ghost cells.

Wide excision of odontogenic ghost cell carcinoma and careful follow-up is necessary, but there is as yet no definitive protocol for the treatment of these rare tumours.

Squamous odontogenic tumour

The squamous odontogenic tumour, first described by Pullon et al (1975), is another rare entity.

Clinically, the squamous odontogenic tumour mainly affects young adults and involves the alveolar process of either the maxilla or mandible, close to the roots of erupted teeth which it may loosen. It can also expand the jaw. Radiographically also, squamous odontogenic tumour can mimic severe bone loss due to periodontal disease, or can produce a cyst-like area of radiolucency.

Microscopy

The characteristic features are circumscribed, rounded or more irregular islands of squamous epithelium in a fibrous stroma. Foci of keratin or parakeratin and calcifications or globular eosinophilic structures may form in the epithelial islands.

Management

This tumour appears generally to be benign, though invasion of adjacent structures by maxillary lesions has been reported (Goldblatt et al, 1982). Curettage or conservative resection and extraction of any teeth involved appears frequently to have been effective treatment, but some, particularly in the maxilla, may require wider resection.

Ameloblastic fibroma

This rare tumour is somewhat similar histologically to the ameloblastoma.

Clinically, ameloblastic fibroma, unlike most ameloblastomas, usually affects young adults or adolescents, but the radiographic appearances may be very similar, namely a multi- or unilocular cyst, most frequently in the posterior part of the body of the mandible. Otherwise, ameloblastic fibromas are slow-growing and usually asymptomatic, apart from eventual expansion of the jaw.

Microscopy

This neoplasm has both epithelial and connective tissue components. The epithelium consists of ameloblast-like or more cuboidal cells surrounding others resembling stellate reticulum or are sometimes more compactly arranged. The epithelium is sharply circumscribed by a basal membrane and forms islands, strands or cauliflower-like proliferations in a loose but cellular, fibromyxoid connective tissue, which forms little collagen and resembles the immature dental papilla.

An exceedingly rare granular cell variant, in which large eosinophilic granular cells replace the fibromyxoid connective tissue component, has been described and termed *granular cell ameloblastic fibroma.*

Ameloblastic fibro-odontoma

This lesion consists of an ameloblastic fibroma together with mixed calcifying or calcified dental tissues. Clinically, it is essentially similar to an ameloblastic fibroma, but radiographs typically show the opaque odontomatous tissues in a circumscribed but otherwise radiolucent area.

Management

The ameloblastic fibroma and fibro-odontoma are benign, and though they may separate readily from their bony walls, Trodahl (1972) reported a recurrence rate of over 40% in the material submitted to the Armed Forces Institute of Pathology. Conservative resection with a margin of sound bone therefore appears to be the treatment of choice.

In the past there has been confusion between the ameloblastic fibro-odontoma and the odonto-ameloblastoma. The latter, as described earlier, is an aggressive tumour for which more radical surgery is essential.

Ameloblastic sarcoma (ameloblastic fibrosarcoma)

Ameloblastic sarcoma is an exceedingly rare malignant counterpart of ameloblastic fibroma - itself a rare tumour.

Clinical features

Wood et al (1988) found reports of 43 cases. In these, the mandible was twice as frequently affected as the maxilla. Young adults of either sex were equally affected, though the reported age incidence ranges from 13 to 78 years.

Some cases appear to arise in ameloblastic fibromas. They initially have similar symptoms, but recur and become obviously aggressive. Others are obviously malignant from the start, grow rapidly and cause either pain or swelling or both.

Radiography

Typically an ill-defined area of radiolucency may be associated, in maxillary lesions, with extension into the antrum to produce an opacity as well as destruction of the walls.

Microscopy

The essential features are islands of epithelium with the same features as those of the ameloblastic fibroma, but surrounded by highly cellular and pleomorphic fibrous tissue. Pleomorphism may be gross and mitoses frequent, but there can be all grades of transition from the benign variant, so that it may be difficult to be certain, histologically, whether or not the mesenchymal component is malignant. In some areas, the sarcomatous element can overwhelm the epithelial component, but in the absence of the latter, the diagnosis cannot be confirmed.

Management

The ameloblastic fibrosarcoma is aggressive, with invasion of adjacent soft and hard tissues, including the base of the skull. Wide radical excision seems to offer the most promising approach as, in many of the reported cases treated more conservatively, there have been multiple recurrences, and sometimes death from local spread of the tumour. However, it is not certain whether this tumour metastasizes. Successful treatment by chemotherapy has been claimed, but its value has not been widely confirmed.

Odontogenic myxoma

The myxoma is peculiar to the jaws. Myxoid tumours elsewhere in the skeleton are the result of myxomatous degeneration in fibrous tumours. Myxomas of the jaws, by contrast, show a close structural resemblance to dental mesenchyme, occasionally contain epithelial rests, affect the tooth-bearing areas of the jaws and a tooth is frequently absent. Growth of this tumour is often rapid at first.

Clinically, young people are predominantly affected, though the age incidence is wide. A fusiform swelling of the jaw and loosening of teeth are typical. Radiographically, there is a radiolucent area with scalloped margins or a soap-bubble appearance which may be similar to that of an ameloblastoma.

Microscopy

Myxomas consist of spindle-shaped or angular cells with long, fine, anastomosing processes. Moshiti er al (1992) reported positive reactions to antibodies to vimentin and actin and negative reactions to S-100 protein: ultrastructural studies also suggested that myxoma cells were myofibroblasts. These cells are scantily distributed in loose mucoid intercellular material. Small amounts of collagen fibres may also be formed and there may be small, scattered epithelial rests. The tumour margins are ill-defined and peripheral bone is progressively resorbed.

Occasional variants on these appearances are mainly due to formation of greater amounts of collagen when the lesion may be termed a fibromyxoma.

Management

Myxomas are benign, but can infiltrate widely so that recurrence after excision is common. Wide excision is desirable in the hope of preventing recurrence, but is not always successful. Irradiation and other methods used to destroy residual tumour tissue have not proved to be of significant value. In spite of aggressive treatment, some tumours can persist for more than 30 years (Cawson, 1972) after the original operations. By this time the tumour may appear inactive and be symptomless.

Rare variants, with a more cellular and pleomorphic microscopic picture, can behave aggressively and may be categorized as myxosarcomas. However, though locally destructive, myxosarcomas appear to have little potential for metastasis.

Odontogenic fibroma

The odontogenic origin of this rare endosteal tumour is confirmed by the presence of epithelial rests, but they are not invariably found.

Clinically, odontogenic fibroma more frequently affects the mandible, but there seems to be no significant predilection for either sex or any age. It forms a slow-growing, asymptomatic mass and usually, therefore, remains unrecognized until it causes a swelling. Alternatively, it may be seen by chance in a radiograph as a sharply defined, rounded area of lucency in a tooth-bearing area.

Microscopy

The odontogenic fibroma consists of spindle-shaped fibroblasts with variable amounts of collagen fibres which may have a whorled arrangement. Strands of epithelium resembling the rests of Malassez may be present in very variable amounts.

Management

The odontogenic fibroma does not infiltrate or adhere to the surrounding bone which merely undergoes pressure resorption. It shells out from the bone so that conservative excision is curative.

Cemental tumours and dysplasias

Cementum, being a modified form of bone, is subject to changes comparable to the dysplasias of bone or to neoplastic changes. Cemental tumours and dysplasias are a group of lesions in which there is continued proliferation of cementum, but they form a difficult group with characteristics that are sometimes controversial. In making a diagnosis, account should be taken of the clinical and radiographic features, the microscopic features and the behaviour. These lesions are relatively uncommon, so that there is too little knowledge to reach any firm conclusions about all of them. Four main types are usually described, but some cases do not fit precisely into these categories. They are all rare except in Blacks.

Cementoblastoma

This is probably a benign neoplasm, which forms a mass of cementum-like tissue as an irregular or rounded mass attached to, and often resorbing, the root of a tooth.

Clinically, cementoblastomas are mainly seen in young adult males, typically below the age of 25 years. The tumour is usually slow growing and most frequently attached to a mandibular first molar root. The jaw is not usually expanded, but occasionally a cementoblastoma can produce gross bony swelling and pain.

Radiography

There is a radiopaque mass with a thin but well-defined radiolucent periphery, attached to the roots of a tooth. The mass is usually densely radiopaque, but may appear mottled. It is typically rounded, but may be more irregular. Resorption of the roots to which the mass is attached is common, but the tooth remains vital.

Microscopy

The mass consists of cementum which typically contains many reversal lines, and has a pagetoid appearance. Cells are enclosed with the hard tissue like osteocytes in bone, while in the larger irregular spaces in the calcified tissue are many osteoclast- and osteoblast-like cells. At the periphery and in areas of active growth, there is a broad zone of unmineralized tissue, and the mass has a fibrous capsule.

These appearances are similar to those of an osteoblastoma and they are only dinstinguishable by the cementoblastoma's relationship to teeth. However, if not recognized by its clinical and other features, the highly active cellular elements may be mistaken for an osteosarcoma.

Management

Cementoblastomas are benign and if the related tooth is extracted and the mass completely excised, there should be no recurrence. If incompletely removed, the mass will continue to grow. More aggressive variants, for which more radical treatment may be needed, have also been described.

Cemento-ossifying fibroma

No practical distinction can be made between so-called cementifying and ossifying fibromas.

Clinically, these uncommon tumours are well circumscribed and characterized by slow expansile growth, usually in the mandibular premolar or molar region. The most common complaint is a painless swelling, unless seen by chance in a routine radiographic examination. They can affect patients over a wide age range, but most commonly between 20 and 40 years, with a reported female predilections.

Radiography

These tumours usually have well-defined borders, and are radiolucent or with varying degrees of calcification. Calcifications tend to be concentrated at the centre of the lesion and some specimens appear largely radiopaque with a narrow radiolucent rim. Sometimes there is divergence or occasionally resorption of related roots. Large mandibular tuours are rare but can cause a characteristic downward bowing of the lower border of the jaw.

Microscopy

Cemento-ossifying fibromas consist of spindle-shaped fibroblasts, with a widely variable degree of cellularity. Some specimens show moderate amounts of collagen, while others may have less intercellular matrix and the cells may be in whorled or storiform patterns.

The types of calcifications within the tumour also vary widely. Trabeculae of woven bone with osteoblastic rimming are often prominent and frequently form an interconnecting reticular pattern. Thicker trabeculae of lamellar bone as well as dystrophic calcifications may also be seen. Specimens which have a predominance of acellular ovoid or spherical calcifications resembling cementicles are commonly termed cementifying fibromas. These nodules are minute at first, but gradually grow, fuse and ultimately form a dense mass. However, similar amorphous spherical calcifications can be found in fibro-osseous lesions of the skull or other bones far distant from any odontogenic tissue. In any cases, most so-called cementifying fibromas show admixed spherical calcifications and bone trabeculae, so that the different patterns of calcification are clearly only minor variants of the same pathological process.

Histologically, cemento-ossifying fibroma may be indistinguishable from many cases of fibrous dysplasia except by the clinical and radiographic findings. Important differences are that ossifying fibroma more frequently affects the mandible, is well circumscribed and can have a fibrous capsule. Fibrous dysplasia, by contrast, merges with normal bone at its margins. It can also extent to the alveolar margin, but in so doing does not fuse with or otherwise distort the roots of teeth. Microscopically, the trabeculae in fibrous dysplasia, though variably shaped, tend to be slender and arcuate or branched with osteoblasts throughout their substance. This contrasts with the more conspicuous osteoblastic rimming typical of ossifying fibroma, where the bone is typically lamellar rather than woven.

A biopsy of an ossifying fibroma also may not be distinguishable microscopically from periapical cemental dysplasia or from other localized cemento-osseous lesions. The latter, which are often found in the mandible at the site of previous extractions, have little or no potential for continued growth and are probably reactive rather than neoplastic.

Though there may be conceptual arguments for distinguishing small ossifying fibromas from cemento-osseous dysplasia, it is not of practical surgical importance.

Management

Cemento-ossifying fibromas may have a definable capsule and can usually be enucleated. Occasionally, large tumours which have distorted the jaw require local resection and bone grafting. The prognosis is good and recurrence rare. However, if an associated tooth is extracted, a densely calcified cemento-ossifying fibroma can become a focus for chronic osteomyelitis. If this happens, complete excision becomes necessary.

Periapical cemental dysplasia

As its name implies, this lesion is probably dysplastic and seems to lack any potential for progressive growth.

Periapical cemental dysplasia affects women, particularly the middle-aged, 10-15 times more frequently than men: it is also more common in Blacks. It usually affects the mandibular incisor region, but can affect several sites or be generalized. It is asymptomatic, but can be seen in radiographs in its early stages as rounded radiolucent areas simulating periapical granulomas, but the related teeth are vital. Increasing radiopacity starts centrally until the masses become densely radiopaque, and all stages of development may be seen in multiple lesions.

Microscopy

The appearances resemble those of cemento-ossifying fibroma. In the early stages, cellular fibrous tissue contains foci of cementum-like tissue which grows and fuses to form a solid, bone-like mass.

Management

The main consideration is to distinguish early lesions from periapical granulomas by routine dental investigation. Once this has been confirmed, further treatment is usually unnecessary.

Florid cemento-osseous dysplasia (gigantiform cementoma)

Women of middle age, particularly Blacks, are mostly affected. The calcifications are frequently symmetrical, may involve all four quadrants and are asymptomatic unless infected. There is a rare familial type.

Radiography

Gigantiform cementoma appears as radiopaque, irregular masses without radiolucent borders and in the past these have been interpreted as diffuse sclerosing osteomyelitis. However, any infection of these lesions is a secondary event.

Microscopy

The masses consist of densely calcified material resembling secondary cementum and containing few lacunae and are sometimes fused to the roots of teeth.

Treatment is difficult in that complete removal of these lesions may require more extensive surgery than their nature justifies and observation by regular follow-up with radiographs at intervals is more appropriate. If infection supervenes, as a result of extraction of an associated tooth or from any other source, chronic osteomyelitis may develop in the dense calcified tissue. In such circumstances the mass may eventually sequestrate or the entire lesion may need to be excised to allow the infection to resolve.

Odontomas

Odontomas are hamartomas or malformations of dental tissues not neoplasms. Even when the morphology is grossly distorted, as in complex odontomas, the pulp, dentine, enamel and cementum form in correct anatomical relationship with one another. Like teeth, they do not develop further, once fully calcified. They also tend to erupt, but once exposed to saliva can become carious and lead to abscess formation. Odontomas may also prevent normal teeth from erupting, or displace them. Cyst formation is another occasional complication.

Odontomas affect the maxilla slightly more frequently than the mandible and are frequently detected in early adolescence, in routine radiographs.

Compound odontomas

These consist of many separate, small denticles. The malformation may be produced by repeated divisions of a tooth germ or by overgrowth of, and multiple budding-off from, the dental lamina with the formation of many tooth germs.

The denticles are embedded in fibrous connective tissue, and have a fibrous capsule. Inflammatory or cystic changes may involve the mass.

Clinically, a compound odontoma most frequently forms in the anterior part of the jaws, where it usually gives rise to a painless swelling. The denticles may be seen radiologically as separate calcified bodies.

The mass should be enucleated surgically as a potential source of obstruction to erupting teeth.

Complex odontoma

This odontoma consists of an irregular mass of hard and soft dental tissues, having no morphological resemblance to a tooth and frequently forming a cauliflower-like mass of hard dental tissues.

Clinically, this malformation is usually seen in young persons, but may escape diagnosis until late in life. Typically, a hard painless swelling is present, but the mass may start to erupt and its many stagnation areas encourage infection.

Radiographically, when calcification is complete, an irregular radiopaque mass is seen with areas of dense radiopacity due to the enamel. Should the mass become infected, the calcified tissues may be mistaken for a sequestrum or an area of sclerotic bone.

Histologically, the mass consists of enamel, dentine and cementum, together with pulp and periodontal ligament in varying amounts. The arrangement is disordered, but frequently has a radial pattern. The pulp is usually finely branched so that the mass is perforated by small vascular channels like a sponge. If seen before calcification starts, the mass may be mistaken for one of the several types of mixed odontogenic tumours.

The odontoma should be removed by as conservative surgery as possible.

Other types of odontomas

In the past, complex classifications have been devised to include such anomalies as dilated, gestant (invaginated) and geminated odontomas. Frequently, part of these malformations is obviously tooth-like. Dilated and gestant odontomas arise by invagination of cells of the enamel organ or of the epithelial sheath of Hertwig, which actively proliferate to expand the developing tooth, or extend through the opposite pole. Gestant odontomas range in severity from a cingulum pit in an otherwise normal upper lateral incisor to the so-called dens in dente.

These malformations require to be removed as potential obstructions to the eruption of other teeth, as a focus for infection or for cosmetic reasons.

Enamel pearls are uncommon, minor abnormalities, which are formed on otherwise normal teeth by displaced ameloblasts or by proliferation of these cells beyond the normal limit of the amelocemental junction.

The pearl may consist only of a nodule of enamel attached to the dentine, or may have a core of dentine which sometimes contains a horn of pulp. Enamel pears are usually rounded, a few millimetres in diameter, and often form near the bifurcation of first molar roots. An enamel pearl can cause a stagnation area at the gingival margin, but its removal may expose the pulp and necessitate extraction.