Fibro-osseous lesions of the head and neck

Gillian Hall

Abstract

This review aims to overview key histopathological features and provide diagnostic clues for a selection of the fibro-osseous lesions of the jaws and facial bones. These form a diverse group of conditions with differing aetiologies but similar histological appearances. Some may be part of more generalized systemic disease. It will become apparent that for this group of conditions, diagnosis cannot be made by examination of biopsy material by a pathologist in isolation, and both clinical and radiological correlation is needed. However, there are some subtle histopathological features that can allow one diagnosis to be favoured over all others, and these will be outlined. Identification of these features in material sent for histopathological analysis should assist the multidisciplinary team in making a definitive diagnosis.

Keywords craniofacial; fibro-osseous; fibrous dysplasia; jaws; ossifying fibroma

Introduction

The benign fibro-osseous lesions of the jaws and craniofacial complex encompass a diverse group of diseases with quite variable aetiologies and pathogenesis but which share similar and overlapping histological features that can make precise categorization difficult. As the name implies, common to all entities are replacement of normal mature bone firstly by a variably cellular fibroblastic stroma within which pathological ossification and/or calcification then occurs. It should be stressed at the outset that a definitive diagnosis is not really possible purely by examination of incisional or excisional biopsy material and there always exists the need for close clinical and, in particular, radiological correlation. The spectrum of diseases includes those with a clear developmental aetiology and known genetic mutations, through lesions which arise as a result of inflammation/infection plus some which are true neoplasms with potential for continued and progressive growth if not fully removed. Whilst there are no recognized malignancies within this spectrum of diseases, the anatomical location of the gnathic and craniofacial bones means that intracranial extension resulting in encephalitis and meningitis may occur with occasional fatal consequences.[1]

Although not strictly included in the fibro-osseous group of conditions, the odontogenic neoplasms of mesenchymal origin will also briefly be mentioned as these may be considered in the differential diagnosis of some of the lesions to be discussed and in fact, some earlier classifications included some of the cemento-osseous dysplasias in the odontogenic tumour section. (See ref.[2] for an overview of the evolving classification over time).

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Classification

The most up to date classification has very recently been published by the WHO, under a new heading of "Fibro-osseous and osteochondromatous lesions", with some of those included as "Bone-related lesions" in the 2005 edition now listed under a heading of "Giant cell lesions and bone cysts" (Box 1).[3] A much more comprehensive classification has previously been suggested by Eversole et al. in 2008[4] (Box 2). The classification of these diseases may evolve still further. Like many aspects of pathology, further splitting and lumping of entities may well rely on information derived from future genetic/molecular analyses.

Why do such a unique group of lesions exist in the jaws and are there comparable lesions elsewhere?

With the exception of fibrous dysplasia, the remaining true fibro-osseous lesions to be described are unique to the jaws. A lesion showing some similarities, osteofibrous dysplasia and confusingly also known as ossifying fibroma, is described in the long bones of very young children and is a cortical lesion.[5] This shows distinct features including presence of cytokeratin positive epithelial cells and cortical location.

Can the existence of this unique group of lesions be explained by the presence of the odontogenic apparatus and the epithelial and mesenchymal remnants of the tooth-forming process that we know to be involved in the formation of odontogenic cysts and neoplasms? The location of many of the cemento-osseous dysplasias around the apices of teeth and noted similarities to normal periodontal ligament tissue, along with the tendency for ossifying fibromas to occur in the tooth-bearing regions of the jaws, have led to speculation that both these lesions are of periodontal ligament origin. There is however no evidence to support this.[6] It is also notable that the psammomatoid type juvenile ossifying fibroma is most commonly encountered in the craniofacial bones rather than the tooth bearing jaws and hence it

WHO Classification of odontogenic and maxillofacial bone tumours 2017

Fibro-osseous and osteochondromatous lesions

- Ossifying fibroma
- Familial gigantiform cementoma
- Fibrous dysplasia
- Cemento-osseous dysplasia
- Osteochondroma

Giant cell lesions and bone cysts

- Central and peripheral giant cell granuloma
- Cherubism
- Aneurysmal bone cyst
- Simple bone cyst

Box 1

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Eversole 2008 classification

Classification of benign fibro-osseous lesions of the craniofacial complex

- Bone dysplasias
  - Fibrous dysplasia
    - Monostotic
    - Polyostotic
    - Polyostotic with endocrinopathy (McCune Albright)
  - Osteitis deformans
  - Pagetoid heritable bone diseases of childhood
  - Segmental odontomaxillary dysplasia
- Cemento-osseous dysplasias
  - Focal cemento-osseous dysplasia
  - Florid cemento-osseous dysplasia
- Inflammatory/reactive processes
  - Focal sclerosing osteomyelitis
  - Diffuse sclerosing osteomyelitis
  - Proliferative periostitis
- Metabolic bone disease: hyperparathyroidism
- Neoplastic lesions (ossifying fibromas)
  - Ossifying fibroma NOS
  - Juvenile ossifying fibroma
    - Trabecular type
    - Psammomatoid type
  - Gigantiform cementoma

Box 2

Comparison of bone and cementum

	Bone	Cementum
Collagen	Mostly type I collagen (90%) plus non-collagenous proteins	Mostly type I collagen (90%) plus non-collagenous proteins
Mineralization (approximate)	70%	50%
Intrinsic vascular and nerve supply	Present	Absent
Ability to remodel	Yes	
Staining characteristics	Lamellar pattern with Picosirius red histochemical stain. Green with modified Gallego iron fuschin stain	Apposition occurs during life and in reaction to inflammation/trauma but does not resorb Non-lamellar pattern with Picosirius red histochemical stain. Red with modified Gallego iron fuschin stain[7]
	Tends to be more eosinophilic on decalcified haematoxylin and eosin stained sections	Tends to be more basophilic on decalcified haematoxylin and eosin stained sections.

Table 1

seems that at least some of these entities have absolutely nothing to do with teeth. A further explanation for the existence of this group of diseases in the head and neck region might be drawn from knowledge of the development of this region and the fact that, with the exception of the condyle, the bones are formed by intramembranous ossification rather than endochondral ossification.

A further source of confusion relates to the precise nature of the mineralized tissue within these lesions. The name cemento-osseous dysplasia implies that both cementum and bone are present. Ossifying fibroma has previously been referred to as cementifying fibroma and cemento-ossifying fibroma, creating some confusion as to the nature of the hard tissue component.

So what is the nature of cementum and, in the context of fibro-osseous lesions, is there a way to differentiate it from bone. Cementum is perhaps the lesser understood and studied mineralized tissue. It is normally seen as a thin layer covering the outer aspect of the roots of the teeth. Periodontal ligament fibres attach into this layer and at their opposite end, into the bone of the maxillary or mandibular alveolus. Similarities and differences between bone and cementum are listed in Table 1. Cementum lacks the lamellar architecture of mature bone and has been described as resembling a primitive, fetal type woven bone. Cementum forming cells have been demonstrated to be phenotypically distinct from bone forming osteoblasts[8,9] and immunohistochemical staining differences have been reported, with cementocytes being positive for fibromodulin and lumican whilst the bone forming cells are negative.[8] Cementum Protein 1 is a further potential marker of interest.[10]

More recently, studies in animal models has proven differential gene expression between osteoblasts and cementoblasts, in particular in Wnt signalling pathways.[11]

Whilst there are clear differences between bone and cementum (Table 1), it seems that little attempt has been made to study the types of mineralized tissue within the benign fibro-osseous lesions of the head and neck. In descriptive terms, mineralized tissue showing trabecular morphology and usually with detectable osteoblasts and osteocytes seems to be regarded as bone. Rounded or irregularly shaped acellular mineralized material, resembling cementicles that are seen in the normal periodontal ligament, is often designated as cementum. The finding however of this latter material in the juvenile ossifying fibromas of the craniofacial bones, well away from tooth bearing areas, suggests that this may well just be an unusual type of bundle bone.[2] As a result, many advocate use of the term "psammomatoid" to describe the non-trabecular rounded, basophilic mineralized deposits, particularly when located at some distance from the teeth.[12] To date, the differential immuno staining and histochemical staining differences have not been utilized as a diagnostic tool in fibro-osseous lesions of the head and neck to assess the precise nature of the tissue. Some authors regard bone and cementum as essentially the same tissue.[13]

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