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MINI-SYMPOSIUM: HEAD AND NECK PATHOLOGY

Maxillofacial fibro-osseous lesions

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Summary Fibro-osseous lesions are a poorly defined group of lesions affecting the jaws and craniofacial bones. All are characterized by the replacement of bone by cellular fibrous tissue containing foci of mineralization that vary in amount and appearance. Classification and, therefore, diagnosis of these lesions is difficult because there is significant overlap of clinical and histological features. The group includes developmental and reactive or dysplastic lesions as well as neoplasms. Recently a new terminology has emerged that has culminated in the latest WHO classification. The core of this classification is the concept of a spectrum of clinicopathological entities in which the diagnosis can only be made on the basis of a full consideration of clinical, histological and radiological features. This review will describe the salient features of these lesions in an attempt to provide practical guidance for the surgical pathologist.

Introduction

Fibro-osseous lesions are a poorly defined group of lesions affecting the jaws and craniofacial bones. All are characterized by the replacement of bone by cellular fibrous tissue containing foci of mineralization that vary in amount and appearance. Classification and, therefore, diagnosis of these lesions is problematical, partly because of a lack of agreement about terminology, but also because of a significant overlap in histological features. The group includes developmental and reactive or dysplastic lesions as well as neoplasms. A number of workers have tried to clarify the classification of these lesions[1–7] and although they may not have agreed on an exact terminology, a concept has emerged which has culminated in the latest WHO classification.[8] The core of this classification is the concept of a spectrum of clinicopathological entities in which the diagnosis can only be made on the basis of a full consideration of clinical, histological and radiological features. Although the terminology is still problematic, this review will use this new classification (Table 1) and will concentrate on the histopathological features that may guide the working surgical pathologist towards a

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diagnosis. It is emphasized again that diagnosis must take account of radiological features and for these the reader is referred to more expert reviews.[9,10]

Fibrous dysplasia

Fibrous dysplasia is a developmental or possibly hamartomatous condition of unknown aetiology. In over 80% of cases it affects only one bone (monostotic) but it may also be polyostotic and affect multiple bones. The polyostotic type may also be associated with café-au-lait skin pigmentation and precocious puberty in the McCune Albright syndrome, which almost exclusively affects young females. The jaws and skull are often affected by monostotic fibrous dysplasia and may be involved in the polyostotic disease. In addition, the craniofacial region may be affected by a form of fibrous dysplasia that is not restricted to a single bone, but may be confined to a single anatomical site. These lesions affect primarily the maxilla but may also cross sutures into the sphenoid, zygoma, frontonasal bones and base of the skull. This type of fibrous dysplasia does not meet the precise criteria for the monostotic or polyostotic forms and has been termed *craniofacial fibrous dysplasia*.[1,9]

In the head and neck, radiology and computed tomography (CT) imaging of fibrous dysplasia show a poorly defined lesion that merges with adjacent bone. Early lesions may be radiolucent, but they become increasingly radiopaque and typically show a diffuse radiopacity or 'ground glass' appearance. Lesions are usually first diagnosed in children and young adults and present as a painless diffuse enlargement of the jaws. The monostotic form has an equal gender distribution, but polyostotic lesions are usually found in females. The maxilla and paranasal regions are more frequently affected than the mandible.

There are no distinguishing histological features between the three types of fibrous dysplasia. The normal bone is replaced by cellular fibrous tissue composed of spindled fibroblasts in a moderate amount of collagen. This contains fine branching, curvilinear trabeculae of woven bone with little evidence of osteoblast rimming (Fig. 1), although osteoid seams and osteoclast activity may be seen. A characteristic feature of fibrous dysplasia that may help distinguish it from ossifying fibroma is that the lesional bone merges imperceptibly with adjacent cancellous bone or with the overlying cortex (Fig. 2). Also, in fibrous dysplasia the fibrous tissue has a monotonous cellularity and the fine pattern of bony trabeculae is repeated throughout the entire lesion.[11] Spherical, cementicle or psammomatoid calcifications may be seen in a minority of lesions, but they are never prominent. In older or mature lesions there may be lamellar bone with mature trabeculae arranged in elongated parallel arrays (Fig. 3). In an ideal biopsy the margins of the lesion and the overall pattern may be clearly apparent, allowing some confidence in a histological diagnosis. Most often, however, the pathologist is faced with curetted fragments making distinction from other fibro-osseous lesions impossible. Only correlation with clinical and radiographic information can lead to a final diagnosis.

It should be noted that fibrous dysplasia affecting the jaws and craniofacial bones may differ in two important ways from lesions affecting the axial skeleton. First, head and neck lesions are diffuse

Table 1 Classification of fibro-osseous lesions of the maxillofacial region.

Fibrous dyplasia
Monostotic fibrous dysplasia
Polyostotic fibrous dysplasia
Craniofacial fibrous dysplasia
Osseous dysplasias
Periapical osseous dysplasia
Focal osseous dysplasia
Florid osseous dysplasia
Familial gigantiform cementoma
Ossifying fibroma
Conventional ossifying fibroma
Juvenile trabecular ossifying fibroma
Juvenile psammomatoid ossifying fibroma

Based on the new WHO classification[8] and also from Waldron,[1] Slootweg,[3] Brannon and Fowler[6] and El Mofty.[7]

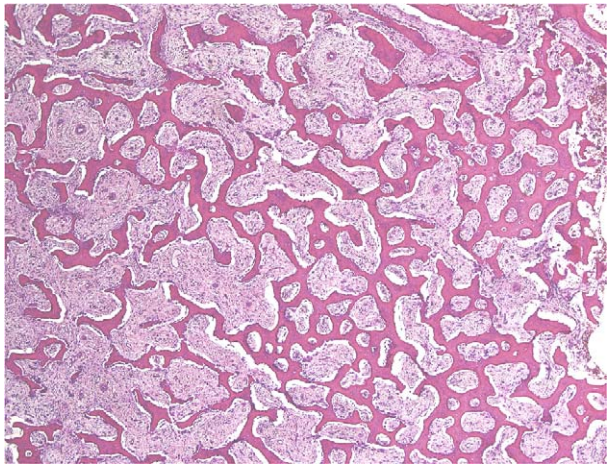

Figure 1 Fibrous dysplasia, the typical histology shows quite evenly distributed fine branching trabeculae of woven bone.