Bone-Related Lesions of the Jaws

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KEYWORDS

• Osseous dysplasia • Ossifying fibroma • Fibrous dysplasia • Giant cell granuloma
• Aneurysmal bone cyst • Osteosarcoma

Key points

- Not all bone tumors or tumorlike lesions also occur in the jaws.
- Not all bone-related gnathic lesions occur in the peripheral skeleton.
- Some bone tumors behave differently when developing at this site.
- Radiologic and clinical correlations are mandatory for making adequate diagnoses.

ABSTRACT

The jaws combine several unique properties that mainly result from their distinct embryonic development and their role in providing anchorage for the teeth and their supporting structures. As a consequence, several bone-related lesions almost exclusively develop in the jaws (eg, osseous dysplasias, ossifying fibromas), have distinct clinical features (eg, osteosarcoma), or hardly ever occur at this location (eg, osteochondroma, enchondroma). The specific characteristics of these tumors and tumorlike lesions are outlined in this article.

OVERVIEW AND DEVELOPMENTAL CONSIDERATIONS

During early embryogenesis, the first pharyngeal arch develops a maxillary and a mandibular prominence. Intramembranous ossification inside the maxillary prominence forms the squamous temporal, maxillary, and zygomatic bones. The ventral parts of the first arch cartilage form a horseshoe-shaped primordium of the mandible, which later disappears after the mandible develops because of intramembranous ossification of the mandibular prominence–related mesenchyme. Only in the median plane of the chin and in the mandibular condyle is there also some enchondral ossification contributing to the normal development.[1]

Although it remains to be elucidated how and to what extent these developmental peculiarities influence the development of tumors and tumorlike lesions in this region, their contribution seems obvious. Enchondromas generally do not occur in the jaws and, despite well-documented cases of chondrosarcomas, no convincing studies have correlated morphology and IDH1/2 mutation status, which might be important to rule out osteosarcoma (OS) with a predominant chondroblastic differentiation. Giant cell tumor of bone is another example of a tumor that is common in the peripheral skeleton and virtually nonexistent in the jaws. Improvements in the molecular understanding of gnathic tumors will help to clarify the reasons for morphologic and clinical differences but are difficult to achieve because of the rarity of most lesions.

This article discusses bone-related lesions commonly occurring in the jaws, including fibro-osseous and giant cell–containing subtypes, and discusses their main differences and differential diagnoses. Furthermore, the distinct clinical behaviors of bone-forming tumors and particularly of

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OSs are outlined and compared with lesions of the peripheral skeleton.

FIBRO-OSSEOUS LESIONS

This descriptively defined group of lesions comprises 3 distinct entities, of which some can be further divided into subtypes with characteristic clinical, radiologic, and histologic presentation:

1. Cemento-osseous dysplasia (COD), including periapical, focal, and florid subtypes
2. Cemento-ossifying fibroma (COF), including conventional, juvenile trabecular, and juvenile psammomatoid subtypes
3. Fibrous dysplasia (FD)

All lesions share a monomorphic fibroblastic stroma embedding various combinations of bone and cementumlike material (**Table 1**).

CEMENTO-OSSEOUS DYSPLASIA

COD constitutes the most common fibro-osseous lesion in the jaws and develops exclusively in the tooth-bearing areas of the gnathic bones. They are considered to derive from cells of the periodontal membrane and to represent nonneoplastic lesions of limited growth potential.[2] However, the molecular pathogenesis is largely unknown. In contrast with other fibro-osseous lesions, the matrix of COD matures into a dense mass over time, resulting in a characteristic radiologic and histologic appearance. There is a striking predilection for middle-aged (black) women.[2] Three distinct clinical variants can be distinguished:

1. Periapical COD: associated with a single or few mandibular incisors
2. Focal COD: single lesion not involving the anterior mandible, most prevalent in the posterior mandible
3. Florid COD: multifocal, generally present in multiple quadrants

Patients are usually asymptomatic and remain undiagnosed until routine dental radiographs are performed.[3] The radiology varies according to the stage of maturation. Early lesions appear as small periapical radiolucencies (**Fig. 1**A) that over time transform into heavily calcified and radio-opaque masses surrounded by a thin lytic rim. The periodontal ligament remains unaffected. Expansive growth and cortical thinning are not typical features of periapical and focal COD but can occur in the florid subtype.[4] Because classic COD does not require treatment and the clinical/radiographic constellation is highly characteristic, biopsy should not be required for the diagnosis. Surgical intervention is even considered contraindicated because it can result in persistent infection that is difficult to treat.[5]

On histology, COD shows trabecular woven bone formation that typically fuses with the preexisting bone of the adjacent cortex. Osteoblastic rimming is not a prominent feature. In addition, a hypocellular or cell-free basophilic matrix can be observed that is generally regarded as cementum/cementicles[2] (see **Fig. 1**B). Because cementum is physiologically formed by cementoblasts after follicular ectomesenchymal cells have been stimulated by exposed dentin, some investigators questioned whether these globular deposits with concentric patterns of mineralization represent cementum and suggested to use “psammoma bodies” or at least “cementum-like material” as more descriptive terms. The World Health Organization (WHO) classification of head and neck tumors from 2005 omitted “Cemento-” in the definition of COD and COF, stating that the distinction between cementum and bone would be equivocal and without clinical relevance.[6] However, the new edition of the WHO classification reintroduced the term, which seems reasonable because it underlines a highly characteristic histologic feature of those lesions irrespective of its true nature.[2] The matrix of COD is enclosed by a moderately cellular and monomorphic proliferation of fibroblastlike spindle cells without atypia and very low (if any) mitotic activity. Over time, the matrix mineralizes and coalesces to form calcified masses resembling ginger roots. Florid COD may be associated with pseudocystic changes that can resemble simple bone cysts. Malignant transformation does not occur.

CEMENTO-OSSIFYING FIBROMA

COFs comprise a group of benign fibro-osseous neoplasms of which 3 variants can be distinguished:

1. Conventional cemento-ossifying fibroma (CCOF): a tumor of odontogenic origin exclusively developing in the tooth-bearing areas of the jaws, most frequently in the premolar/molar region of the mandible
2. Juvenile trabecular ossifying fibroma (JTOF): a rare bone tumor occurring most commonly in the maxilla but also in the mandible and rarely in extragnathic sites
3. Juvenile psammomatoid ossifying fibroma (JPOF): a rare tumor only infrequently affecting the jaws but more commonly developing in extragnathic sites, preferentially in the periorbital frontal and ethmoid bones

COF is rare and, similar to COD, primarily affects women in the third and fourth decades of life.[7] JTOF and JPOF can occur at younger ages (8.5–12 years mean age for JTOF and 16–33 years for