Operative Techniques in Otolaryngology (2014) 25, 96–109

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Operative Techniques in Otolaryngology

Imaging of fibro-osseous lesions of the temporal bone

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KEYWORDS
Fibro-osseus;
Fibrous dysplasia;
Osteoma;
Osteoblastoma;
Chondroblastoma;
Giant cell osteosarcoma;
Chondrosarcoma;
Otospongiosis

Fibro-osseus and cartilaginous lesions of the craniofacial region including the temporal bone are uncommon. Diagnosis of these lesions can be challenging due to overlapping clinical, radiologic, and pathologic features. This article discusses the diagnostic features of several fibro-osseus lesions of the temporal bone including: fibrous dysplasia, osteoma, osteoblastoma, chondroblastoma, giant cell tumor, osteosarcoma, chondrosarcoma, and otospongiosis.

Fibro-osseous (FO) and cartilaginous lesions of the craniofacial region, including the temporal bone, are relatively uncommon.[1,2] These lesions may share overlapping clinical, radiologic, and pathologic features causing potential difficulties in diagnosis. Included within this spectrum of temporal bone and other craniofacial FO and cartilaginous lesions are both nonneoplastic proliferations and neoplasms.

Included under the category of benign FO lesions are fibrous dysplasia (FD), ossifying fibroma which includes aggressive juvenile psammomatous active ossifying fibroma, osteoma, osteochondroma, and osteoblastoma (OB). In keeping with the purpose of this issue of the journal, selected benign and malignant lesions of the temporal bone are discussed. This article presents the clinical and radiographic criteria that may assist in differentiating these lesions from one another. The value of imaging appearance of these lesions in the pathologic diagnosis of FO and cartilaginous lesions cannot be overemphasized. The histopathologic diagnosis of such a lesion should not be rendered in the absence of imaging correlation.[2]

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http://dx.doi.org/10.1016/j.otot.2013.11.012

In contrast to similar lesions of the long bones, when these lesions are found in the head and neck, they are often curetted, thereby destroying their normal architecture and precluding any description of their gross pathologic appearance.[2]

The specific entities that are discussed in this article include FD, osteoma, exostosis, OB, chondroblastoma, giant cell tumor (GCT), osteosarcoma, chondrosarcoma (CS), and otospongiosis.

Fibrous dysplasia

FD is a nonneoplastic developmental FO disease affecting the bone in which normal medullary bone is replaced by structurally weak fibrous and osseous tissues.[2] It is most common in the second and third decades of life with slight female predilection.

The histology is a mixture of fibrous connective tissue and poorly formed trabecular bone replacing portions of normal bone. The histology of FD is not predictive of the biological behavior, and these lesions can be very vascular with significant bleeding at biopsy. Therefore, clinical history, examination, and imaging are usually adequate to make the diagnosis.[3]

It has been associated with a mutation in the guanine nucleotide stimulatory protein (GNAS1) gene.[4] FD becomes stable after skeletal maturity is reached, although it can become reactivated in various clinical settings including pregnancy. There are 3 subtypes of FD: monostotic, polyostotic, and craniofacial. Craniofacial FD is unique in that it involves multiple adjacent bones but is confined to the craniofacial complex. Polyostotic FD may present as a part of McCune-Albright-Sternberg syndrome.[2] The syndrome is characterized by the triad of polyostotic FD, endocrine dysfunction (hyperthyroidism and sexual precocity, the latter predominantly identified in women), and cutaneous hyperpigmentation.[1,2] Congenital FD, also known as cherubism, is an autosomal dominant disease characterized by bilateral swelling of the maxilla with involvement of the maxillary sinuses and infraorbital floor. This results in cherubic appearance as depicted in Renaissance art with a winged child looking upward toward heaven.

Craniofacial FD can be asymptomatic but can cause facial deformities, and if it compromises skull base foramina, it can cause cranial nerve dysfunction. With FD of the temporal bone, symptoms often vary, but most common presenting symptoms are conductive hearing loss (10%-67%), followed by sensorineural hearing loss (4%-15%), and rarely facial nerve weakness or paralysis.[3,5,6] Conductive hearing loss is caused by narrowing of the external auditory canal (EAC) and fixation of ossicles from the surrounding bone lesion. When it involves the course of the facial nerve, it can cause facial nerve weakness or paralysis.

The imaging features of FD depend on the stage of development and the amount of bony matrix within the lesion. Radiographic changes range from lucent zones to diffuse areas of sclerosis, at times referred to as "ground-glass" appearance (Figure 1A). The involved bone characteristically shows expansion. At times, expansion can be very prominent, particularly when the squamous portion of the temporal bone is involved (Figure 1B and C). Periosteal reaction and cortical break are not features of FD, and if present, should raise the possibility of malignant tumor such as osteosarcoma. The tympanic bone and squama of the temporal bone are preferentially involved by the sclerotic form of FD (Figure 1D). The lytic form or combined lytic and sclerotic form of FD usually involves the squama.

FD has intermediate signal intensity on T1-weighted (T1W) and heterogeneous hypointensity signal on T2-weighted (T2W) magnetic resonance images (Figure 1E). There may be areas of hyperintensity, particularly in early stages of the disease (lytic form). Following intravenous administration of gadolinium contrast material, there is often moderate to marked contrast enhancement (Figure 1F). This is owing to the fact that FD can be very vascular, which at times may cause significant bleeding at surgery. On imaging, FD of the temporal bone may be mistaken for meningioma, including en plaque meningioma, sclerotic metastases (breast and prostate), osteochondroma, OB, or osteosarcoma.

Management options include observation, medical therapy using bisphosphonates, and surgical excision with reconstruction. The mainstay of surgical management is aimed for reasonably acceptable esthetics. EAC lesions may need to be excised to prevent formation of cholesteatomas (Figure 1A). Surgical treatment is usually delayed until skeletal maturity to minimize the risk of regrowth and recurrence.

FD can undergo spontaneous malignant transformation in less than 1% of cases.[3,5] Malignant degeneration of FD is most commonly associated with radiation therapy, with transformation rates as high as 44%. The most common neoplasm is osteosarcoma, but it can also transform into fibrosarcoma, CS, and malignant fibrohistiocytoma.[5]

Osteoma and exostosis

Osteomas are benign bone-forming tumors that are almost exclusively identified in the craniofacial skeleton.[1,2] In the craniofacial regions, osteomas may be found in all sites but are most common in the fronto-ethmoid sinuses. Within the temporal bone, osteomas are most commonly seen involving the tympanic bone, mastoid, and petrous bone. Osteomas usually occur as single lesions but may be associated with Gardner syndrome, an inherited autosomal dominant disease that is characterized by colorectal polyposis, soft tissues lesions (fibromatosis, cutaneous epidermoid cysts, lipomas, and leiomyomas), and multiple craniofacial osteomas.[1,2] The radiographic appearance is that of a sharply delineated radiodense lesions arising in the external canal or petromastoid bone, protruding into the external ear (Figure 2A) or over the petromastoid region.

Exostoses are the most common bony tumors of the external ear (Figure 2B). Exostoses present as diffuse hyperostosis, often associated with water sports and are at times referred to as "swimmer's" or "surfer's" ear. Unlike osteomas of the EAC, which presents as a single, always unilateral bony mass, exostoses present often as bilateral, multiple bony excrescences. Histologically, exostoses show a laminated appearance of layers of subperiosteal cortical bone. Unlike osteomas, exostoses demonstrate no medullary cancellous bone, bone marrow, or fibrous tissue.[7]

Osteoblastoma

OB is a rare benign osteoblastic neoplasm that shares its histologic appearance with osteoid osteoma but is of larger size.[2] It accounts for 1% of all bone tumors. Most cases occur in individuals younger than 30 years, with the mean age being 20.4 years.[8] In the head and neck, the most common site of involvement is the jaw, with the mandible being more common than the maxilla, followed by the cervical spine and then the skull.[9] Overall, OB is more common in males, but in the head and neck region, it has female predominance.[10,11] OBs in temporal bone commonly involve the mastoid bone and zygomatic roots of its zygomatic process of the squamous portion of the temporal