A Rare Case of Middle Ear Myxoma

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A 59-year-old male patient complained of sensation of fullness in his right ear, intermittent otorrhea, and impaired hearing. Patient was admitted to our Hospital in June 2014. His medical history was significant for myringoplasty on the right ear performed in 1973. After initial surgery, he had no ear-related complaints until 8 months prior to this visit. Otoscopic examination revealed obstructive mass in the right ear canal. A pure tone audiogram showed severe mixed hearing loss on the right ear. Multislice computed tomography (MSCT) revealed osteodestructive mass obliterating external ear canal, tympanum, and mastoid with bone erosion of the lateral wall of epitympanum and tegmen tympani toward the middle cranial fossa. Magnetic resonance imaging (MRI) showed heterogeneous expansive mass with homogeneous opacification on postcontrast view in epitympanum and mastoid as well as erosion of tegmen tympani (Figures 1 and 2).

Intraoperatively, tumor mass was found in external ear canal, mastoid, and tympanum. A large bone defect of tegmen tympani toward the middle cranial fossa was found. There was neither dura invasion nor cerebrospinal fluid leak during surgery. We observed dehiscence of the lateral semicircular and facial canal, along with complete erosion of hearing ossicles. In order to remove the tumor completely, radical tympanomastoidectomy was performed. Postoperative recovery was uneventful, without signs of facial nerve palsy. There was no change in hearing level on operated ear nor balance disorder.

Histological examination demonstrated hypocellular myxoid tissue with stellate and spindle cells with no signs of nuclear atypia nor mitotic figures, covered by keratotic, reactively changed skin epithelium (Figure 3). Tumor cells were S-100 and desmin negative but CD34 and SMA positive. These findings strongly suggested superficial angio-myxoma. Myxomas are mesenchymal origin neoplasms that mostly affect heart but have been described in soft tissues and bone throughout the body.1

To the best of our knowledge, there are only 13 cases of temporal bones myxomas described so far.2,3 Clinical presentation can mimic chronic otitis media. Cases of facial nerve palsy, hearing loss, and vertigo as first symptoms have been described.1,3,4

References:

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Figure 1. A and B, MR coronal (A) and sagittal (B) scans of the temporal bone showing expansive mass of the temporal bone with destruction of the tegmen tympani (arrow).

Figure 2. Computed tomography (CT) scan showing expansive mass of the temporal bone.
Both MSCT and MRI can be useful, but definitive diagnosis cannot be based on imaging tests solely. Histological examination and immunohistochemical staining are crucial for a definitive diagnosis. Myxomas are usually insensitive to radiation therapy, with surgical resection being the treatment of choice.4 The etiology of myxomas occurring in the temporal bone is unclear. It has been hypothesized that they originate from primitive embryonic mesenchyme.1 Also, cases of middle ear myxomas with embolic origin from primary heart myxomas have been reported.3 External ear myxomas are often associated with Carney complex, a rare multiple neoplasia syndrome characterized by skin and mucosa pigmented lesions, cardiac, cutaneous and other myxomas, and multiple endocrine and other tumors.5,6 This association has not been observed in temporal bone myxomas. However, there are only a few cases described so far. The most common cause of death in patients with Carney complex is cardiac myxomas.6 Sporadic myxomas and Carney complex–related myxomas are histologically indistinguishable, and additional tests should be performed in patients with middle ear myxomas to rule out Carney complex. Given that heart and temporal bone myxomas can occur simultaneously, we performed heart echosonography to exclude this possibly fatal condition. Also, full skin examination was performed in order to exclude Carney complex. There were neither signs of heart myxoma nor skin changes characteristic for Carney complex.

In conclusion, middle ear myxomas are rare neoplasms but should be considered in differential diagnosis of temporal bone pathology. Radiologic imaging has proven to be a very useful diagnostic procedure in suspected middle ear disease.

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