An aggressive papillary tumor of middle ear: A case report

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ABSTRACT

Aggressive papillary tumors of the middle ear constitute a small percent of adenomatous middle ear tumors. Patients usually present with long-lasting tinnitus, hearing loss, and vertigo and occasionally facial paralysis. Computed tomography findings predominantly suggest bone invasion. Thus, it is commonly misdiagnosed with chronic otitis media with a cholesteatoma. A 72-year-old man presented with chronic ear pain and ear drainage. The patient had a 63 decibels (dB) mixed-type hearing loss and a mass lesion at the temporal bone high resolution computed tomography. Explorative tympanotomy revealed a tumor with thick mucoid secretion and no bone erosion, for which an incisional biopsy revealed an adenoma. Radical modified mastoidectomy was performed and the definite pathology result was compatible with an aggressive papillary tumor. The patient was free of disease at six months of follow-up.

Keywords: Ear neoplasm, mastoidectomy, middle ear, surgery.

Adenomatous tumors of the middle ear are very rare entities¹,² accounting for only a small portion of the primary neoplasms of the ear, which mostly originate from the neuroectoderm such as paragangliomas, meningiomas, and neurinomas.³ In 1988, Gaffey et al.⁴ first described an aggressive papillary middle ear tumor in a series of 10 patients with an aggressive papillary growth pattern with frequent bone destruction and endolymphatic sac and posterior dura invasion. In 1989, Heffner⁵ described the possible origin of this tumor as the endolymphatic sac. Lastly, in the tumor classification published by the World Health Organization (WHO), an aggressive papillary middle ear tumor is classified as the same entity with the Heffner’s tumor and endolymphatic sac tumor (ELST).⁶

The majority of patients mostly present with slowly progressing and long-lasting history of tinnitus, conductive hearing loss, vertigo and, rarely, facial paralysis.⁷ Patients are often misdiagnosed and mistreated with chronic otitis media. Furthermore, accompanying von Hippel-Lindau (VHL) disease is frequent in patients with endolymphatic sac tumors, particularly in those with bilateral diseases.⁸

In this case report, we present a case of a recurrent, unilateral aggressive papillary middle ear tumor with detailed radiological and histopathological findings in the light of the literature.
CASE REPORTS

A 72-year-old man was admitted to our tertiary otology clinic with ear pain and ear drainage for last three years which progressed within the last year with the initial diagnosis of an otic polyp. Otomicroscopic examination revealed a polypoid mass fulfilling the entire left external ear canal. The left tympanic membrane was not visible, and the pure tone audiogram revealed a 63 decibels (dB) mixed-type hearing loss at 500-2,000 Hertz (Hz). Physical examination and the hearing thresholds of the right ear were within the normal ranges. Bilateral facial nerve functions were normal. High-resolution computed tomography (HRCT) scan of the temporal bone showed a mass originating from the middle ear extending to the external ear canal. Mastoid air cells were normal and there was no erosion or sclerosis of the mastoid cortex present, which was not compatible with a cholesteatoma (Figure 1a, b). An explorative tympanotomy procedure was planned due to conflicting physical examination and radiological findings. A written informed consent was obtained from the patient. Intraoperatively, an incisional biopsy was taken from the hemorrhagic mass. Postoperative pathology analysis reported glandular structures standing back to back with an initial diagnosis of adenoma. Thus, radical mastoidectomy was performed to eradicate the disease. During the mastoidectomy procedure, a highly vascular granulation tissue was present with thick and highly mucoid secretion. The lesion was filling the entire middle ear cavity extending to both hypotympanum and epitympanum. Postoperative pathological examination reported an aggressive papillary tumor (Figure 2).

During the postoperative follow-up, the patient was disease-free with no complaints. However, at the second-year follow-up, recurrence of the disease was observed and the patient underwent revision mastoidectomy. At six months of follow-up after the revision surgery, the patient was free of disease with no complaints.

Figure 1. (a, b) Temporal high-resolution computed tomography showing a mass lesion originating from middle ear extending to external ear canal with no prominent erosion or sclerosis of mastoid cortex.

Figure 2. Papillary glandular pattern showing complex branching (H-E ×10).
DISCUSSION

Aggressive papillary tumor of the middle is a very rare tumor classified in the papillary tumors of the middle ear by the WHO.\[9\] Histopathologically, the structure of the tumor resembles a renal cell carcinoma, thyroid carcinoma, and papillary variant of choroid plexus tumor.\[10\] In 1989, Heffner\[4\] described the similarity between the histopathology of papillary tumors of the middle ear and endolymphatic sac tumors and co-incidence of VHL syndrome, and indicated that aggressive papillary tumors are originally adenocarcinomas of endolymphatic sac epithelium. In 1995, Pollak et al.\[11\] reported that the endolymphatic sac was not involved and originated from the pneumatic cell mucosa around jugular bulbus. After a long debate on whether the tumor originates from the endolymphatic sac, and even though the tumor is named as the Heffner’s tumor the disease is classified apart from an endolymphatic tumor as an aggressive papillary tumor of middle ear at the fourth edition of the Head and Neck Classification by the WHO in 2017.\[9\]

Aggressive tumors are most commonly seen in middle-aged women, and ear drainage refractory to local treatments is the most common clinical presentation of the tumor.\[2\] Ear drainage is mostly due to tympanic membrane perforation secondary to mass effects of the tumor. Occasionally, the tumor may present with progressive conductive hearing loss, tinnitus, and vertigo such as the other middle ear masses.\[1\] Similar to the literature, our patient presented to our clinic with ear drainage unresponsive to local and systemic antibody treatment and progressive conductive hearing loss. Computed tomography (CT) scan showed a soft tissue density with expansive growth patterns in the middle ear and rare bony erosion. These findings are compatible with a cholesteatoma which is the frequent cause of misdiagnosis.\[2\] The differential diagnosis of the tumor can be only made with pathological assessment of the biopsy taken via explorative tympanotomy. Similarly, our patient was misdiagnosed with a cholesteatoma and referred to our clinic for cholesteatoma surgery.

Surgery is the main treatment modality for aggressive papillary tumors. Although early diagnosis and modified radical mastoidectomy or radical mastoidectomy is curative, due to the late onset of audiovestibular symptoms, diagnosis is delayed in the majority of cases. Therefore, complete resection is difficulty and recurrence may occur. In a case report, Elhefnawy\[12\] presented a case with multiple recurrences. In addition, although metastasis is rare, intracranial extension may be seen.\[13\] Similarly, a modified radical mastoidectomy procedure was performed and local recurrence was observed two years later in our patient, which was treated with radical mastoidectomy. The patient was disease-free at six months of follow-up after second surgery.

In conclusion, aggressive papillary tumors of the middle ear are rare entities which account for only a small percentage of middle ear tumors. It typically presents with slowly-progressing, conductive-type hearing loss, tinnitus, and vertigo; however, in most cases, patients apply to physicians with ear drainage due to secondary tympanic membrane perforation due to the mass effect of the tumor. Definite diagnosis is made based on pathological examination of the biopsy specimens obtained from the mass during explorative tympanotomy. Although the mass is locally invasive, distant metastasis is not expected. Early diagnosis and mastoidectomy is curative in most cases, but local recurrence is common. Patients with middle ear tumors such as aggressive papillary tumors may be misdiagnosed with chronic otitis media; therefore, unusual tissue on the external ear or middle ear should be sent to pathological examination to confirm the diagnosis.

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REFERENCES


