

CASE REPORT

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A RARE CASE OF MIDDLE EAR ADENOMA

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ABSTRACT

Middle ear adenoma is a rare disease that is thought to originate in the middle ear mucosa. It occurs over a wide age range, has no gender predilection, and is not characterized by specific symptoms or findings. The most frequent complaints are unilateral hearing loss and ear fullness. We report a 48-year-old woman with middle ear adenoma who had a history of unilateral ear fullness and hearing loss on the left side. Middle ear adenoma was suspected following a biopsy performed under local anesthesia. To remove the tumor, the patient underwent a left postauricular canal wall-up tympanoplasty type IIIc. Microscopic examination and immunohistochemistry confirmed a middle ear adenoma. In this case, we diagnosed the lesion as middle ear adenoma with neuroendocrine differentiation on the basis of the pathological findings. The patient has shown no recurrence for almost 5 years, but since this adenoma showed neuroendocrine differentiation, long-term observation is required.

Key Words: middle ear adenoma, tympanoplasty

INTRODUCTION

Middle ear adenoma is a rare disease that is thought to originate in the middle ear mucosa. It is not characterized by any specific symptoms or findings, although some patients present with facial paralysis. The differential diagnosis of middle ear adenoma includes congenital cholesteatoma and benign middle ear tumors. The mass can have exocrine and/or neuroendocrine differentiation. We report here a case of middle ear adenoma with neuroendocrine differentiation.

CASE REPORT

A 48-year-old woman complaining of unilateral ear fullness and hearing loss on the left side first visited our otolaryngology clinic in August 2009. She has been suffering from the symptoms since the beginning of the year and had seen some otolaryngologists before her first visit to our hospital. One doctor had found “no problem” in the left ear and another doctor had diagnosed adhesive otitis and treated her for tympanic inflation. However, the symptoms did not abate

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and a doctor suspecting middle ear tumor referred her to us. Beside arthritis in her knee, the patient had no other medical or family history. Otoscopic examination revealed a retrotympenic tumefaction. (Fig. 1) Pure tone audiometry at 0.5, 1, and 2 KHz showed a conductive hearing loss of 31.2 dB in the left ear. Tympanometry showed type C findings in the left ear. Computed tomography (CT) of the temporal bone showed a shadow in the middle ear, specifically in the mesotympanum and hypotympanum, and mastoid cells appeared clear. There was no evidence of bone distraction. (Fig. 2)

Given the color of the tumor and that the tumefaction showed no growth, we diagnosed the mass not as a cholesteatoma but a benign middle ear tumor. We performed a biopsy under local anesthesia at the site of anterior superior quadrant lesion, and middle ear adenoma was suspected from the pathological findings. The patient underwent a left postauricular canal wall-up tympanoplasty type IIIc in December 2009 to remove and definitively diagnose the tumor. The yellowish-white tumefaction occupied the attic and mesotympanum, but had not damaged the auditory ossicle. We removed the head of the malleus and incus to enucleate the tumor. There was no invasion of the mastoid cells or facial canal. (Fig. 3) We reconstructed the ossicular chain with type IIIc with columella using auricular cartilage.

Microscopic examination of the tumor showed glandular and solid characteristic patterns of adenoma and a partial sheet-like pattern. The tumor cells were uniform and cuboidal or cylindrical without any atypical cells. Immunohistochemistry revealed the presence of cytokeratin (AE1/



Fig. 1 Finding of otoscopic examination
Retrotympenic tumefaction at the site of anterior superior quadrant lesion in the left ear

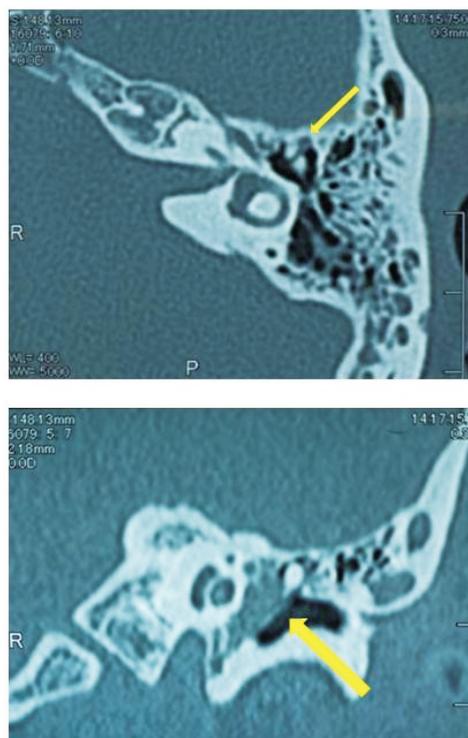


Fig. 2 Computed Tomography of the temporal bone
Shadow in the mesotympanum and hypotympanum, mastoid cells appear clear
(Upper: horizontal image, Lower: coronal image, Arrow shows tumefaction.)

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AE3) and neuroendocrine markers. Synaptophysin was positive and chromogranin was focally positive. Microscopic examination and immunohistochemistry confirmed a middle ear adenoma. It is widely held that when these neuroendocrine elements are dominant, a carcinoid tumor should be considered; however, the tumor showed a glandular pattern on histopathology, so we diagnosed it as middle ear adenoma with neuroendocrine differentiation. (Fig. 4)

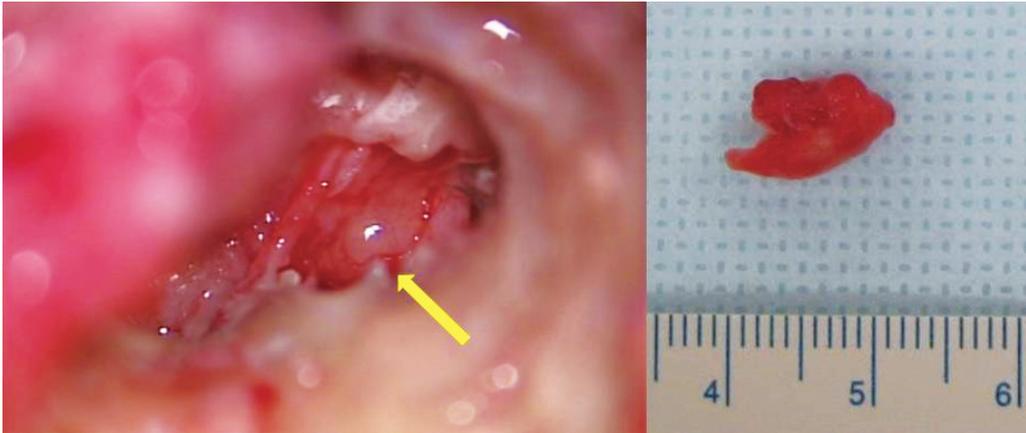


Fig. 3 Operative findings and extracted tumor

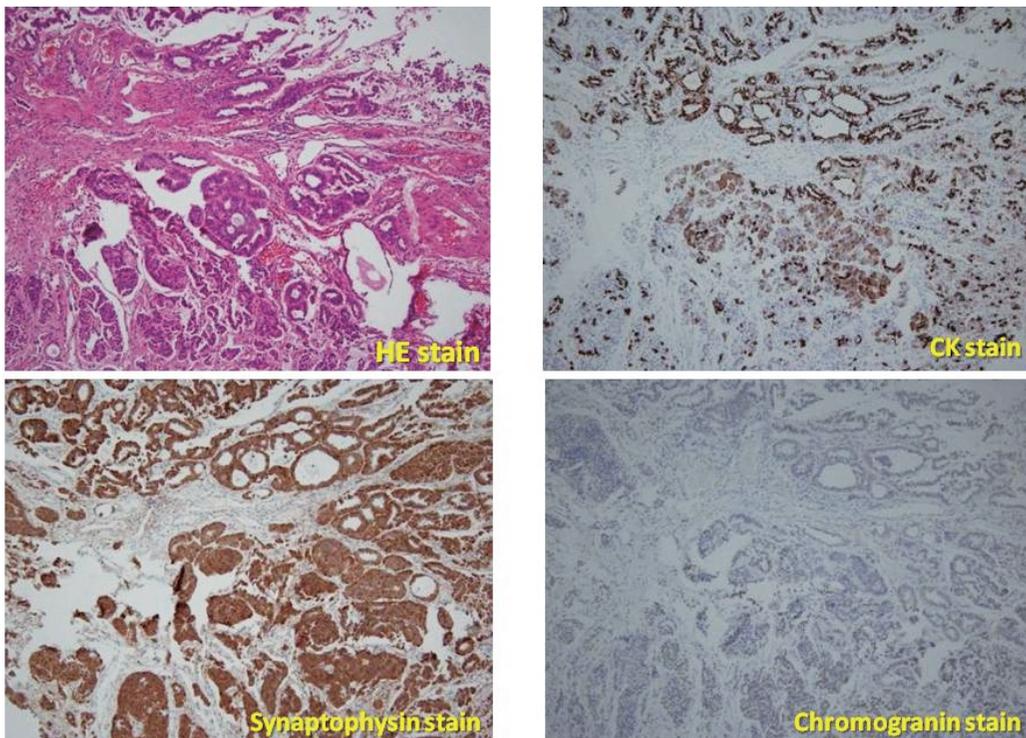


Fig. 4 Histopathological results of the tumor
(HE stain; hematoxylin-eosin stain, CK stain; cytokeratin stain)

The patient's postoperative course was uneventful and she has shown improvement of the hearing loss. Postoperative CT at 1 year showed no recurrence and pure tone audiometry at 0.5, 1, and 2 KHz showed a loss of 25.0 dB in the left ear. As of the time of writing, at almost 5 years, there has been no recurrence.

DISCUSSION

Middle ear adenoma, a benign epithelial tumor first described by Hyams and Michaels in 1976, is thought to originate in the middle ear mucosa.¹⁾ It is a rare disease, and only 29 cases have been reported in Japan to date. It occurs over a wide age range, has no gender predilection, and is not characterized by any specific symptoms or findings.²⁾ The most frequent complaints are unilateral conductive hearing loss, ear fullness, tinnitus, and dizziness. The overwhelming majority of middle ear adenomas do not invade or erode the temporal bone or infiltrate the facial nerve. Some patients experience facial paralysis and there have been three such cases reported in Japan.³⁻⁵⁾ Otoloscopic examination is useful for finding retrotympanic tumefaction, but it is hard to distinguish from the colors.⁶⁾ Temporal bone imaging tests will show a localized tumor in the middle ear without bone destruction. Differential diagnosis of middle ear adenoma includes congenital cholesteatoma, carcinoid tumor, schwannoma, teratoma, meningioma, and paraganglioma of the middle ear. A pathological examination is required to differentiate middle ear adenoma, and the recommended treatment is total exploration of the tumor and extirpation.^{7,8)} Immunohistochemical findings may indicate an adenoma that is an aggressive papillary middle ear tumor with an aggressive growth pattern (e.g. causing bone destruction and cranial nerve involvement), which is classified as a low-grade malignant tumor. Middle ear adenomas and aggressive papillary tumors are both extremely difficult to diagnose preoperatively.⁹⁾

The World Health Organization criteria for benign neoplasms of the middle ear give middle ear adenoma and middle ear carcinoid as the same category of disease. Middle ear carcinoid is described as showing more endocrine differentiation than middle ear adenoma. Carcinoid tumors arise from the neuroendocrine cells normally found in the lung and gastrointestinal tract, and these endoderm-originated cells are usually scattered within the epithelium. Carcinoid tumors have been reported in a wide range of organs, most commonly in the gastrointestinal tract, lung, and pancreas.¹⁰⁾ Many authors consider carcinoid tumors of the lung to behave as low-grade malignant tumors, with significant metastatic potential. Carcinoid tumors secrete serotonin and/or prostaglandin leading to carcinoid syndrome, such as flushing, diarrhea, and cramping. Unlike carcinoid tumors of other organs, middle ear carcinoids do not manifest as typical carcinoid syndrome. In 2006, Suzukawa et al. reported there have been 35 cases of middle ear carcinoid in Japan.¹¹⁾ Ramsay et al. reported metastasis to the lymph nodes and other organs in 9% of cases and recurrence in 22%¹²⁾, although Suzukawa et al. reported such cases to be rare. Microscopic examination of carcinoid tumors shows solid glandular and trabecular patterns, forming a ribbon-like tubular pattern. Immunohistochemical staining is usually positive for cytokeratin, and the neuroendocrine markers synaptophysin, chromogranin, and neuron specific enolase. Microscopic examination of middle ear adenoma also shows solid glandular and tubercular patterns. The tumor cells are usually uniform and cuboidal or cylindrical without any atypical cells. They have a moderate amount of acidophilic cytoplasm and may assume a plasmacytoid appearance. Again similar to carcinoid tumors, immunohistochemical staining is usually positive for cytokeratin and neuroendocrine markers such as chromogranin and neuron specific enolase.¹³⁾ Aggressive papillary tumors show a papillary pattern on histological examination.

In the present case, we arrived at the final diagnosis through a number of clinical steps. We

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had suspected middle ear adenoma from the preoperative otoscopic and CT findings and then from the pathological findings on biopsy. As the recommended treatment is total exploration and extirpation, we performed left postauricular canal wall-up tympanoplasty type IIIc to remove the tumor. Intraoperatively, the tumefaction was not found to have damaged the auditory ossicle, and there was no invasion of the mastoid cells or facial canal. Microscopic examination of the tumor showed a solid glandular adenoma pattern, but also in part a sheet-like pattern characteristic of neuroendocrine differentiation, and immunohistochemistry revealed the presence of the epithelial marker cytokeratin (AE1/AE3) and neuroendocrine markers. Chromogranin-A, synaptophysin, and neuron-specific enolase were used as neuroendocrine markers, and synaptophysin was positive and chromogranin was focally positive. However, we arrived at a final diagnosis of middle ear adenoma with neuroendocrine differentiation rather than carcinoid tumor because most of the tumor showed an adenoma pattern on microscopic examination and because chromogranin was only focally positive.¹⁴⁾

Middle ear adenoma and carcinoid tumor are both rare neoplasms of the middle ear, and it is difficult to diagnose one from the other. Some authors consider middle ear adenoma and carcinoid to be the same disease in a different state of differentiation. Patients must be followed for possible recurrence, although recurrence is rare. Regular follow-up with otoscopy, audiometry, and/or CT are recommended. We diagnosed this case as a middle ear adenoma, but since the adenoma showed neuroendocrine differentiation, long-term observation is required.

CONCLUSION

The middle ear adenomas are rare neoplasms and have no specific symptoms or clinical presentation. Total exploration and extirpation is required for treatment, and microscopic and immunohistochemical examinations are required for a definitive diagnosis. Middle ear adenomas can show neuroendocrine differentiation, as in the present case. Recurrence and metastasis are considered rare, but long-term observation is required.

The authors declare that they have no conflict of interest.

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