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CASE REPORT

Neuro-endocrine adenoma of the middle ear: a case study

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Abstract Neuroendocrine adenomas are rare tumors, which can appear in the middle ear. Approximately a hundred cases have been reported in the literature. We report the case of a 58-year-old man who consulted for an abnormal sensation of fullness in the right ear. The otoscopic examination showed a retrotympanic tumefaction. The CT scan and MRI of the middle ear demonstrated a well-defined tissue mass without any osteolysis. We performed surgical exeresis by transcanal procedure with a cartilage graft tympanoplasty. Microscopic examination and immunohistochemistry revealed an endocrine adenoma of the middle ear. Neuroendocrine adenomas can develop in a number of different sites. When they appear in the middle ear they usually produce hypoacousia. The otoscopic examination shows non-specific findings with only retrotympanic swelling. Surgical exeresis enables histologic and immunohistochemically analysis of the surgical specimen. The adenoma is composed of two cellular types: neuroendocrine (which closely resemble carcinoid tumors) and glandular. Regular clinical and radiologic follow-up is necessary since recurrence is possible. The formal diagnosis of neuroendocrine adenomas of the middle ear requires histologic and immunohistochemically confirmations since the clinical symptoms are non-specific. Surgical excision with removal of the ossicular chain is the treatment of choice in order to prevent recurrence.

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Introduction

Neuroendocrine adenomas of the middle ear are rare benign tumors and approximately a hundred cases have been reported in the literature since they were first described by Derlacki [3] in 1976. Their clinical and paraclinical presentation is non-specific [1, 10]. Definitive diagnosis is based on histological and immunohistochemical examination [10].

We report the case of a 58-year-old man who underwent surgery for a neuroendocrine adenoma of the middle ear. Subsequently, we present a review of the literature and a description of the diagnostic and therapeutic aspects of these tumors.

Case report

A 58-year-old man consulted for an abnormal sensation of fullness in the right ear. The ENT specialist who examined him at that time noticed a tumefaction in the posterior wall of the right external auditory canal and recommended regular follow-up. The patient was seen in consultation ten years later because the impression of abnormal fullness in his ear had recently increased. Otoscopy revealed a postero-inferior, non-pulsatile tumefaction in the posterior wall of the right external auditory canal (Fig. 1), which displaced the tympanic membrane. The facial nerve function was normal. Audiometric evaluation showed normal hearing (Fig. 2). CT scan of the temporal bones (Fig. 3) revealed a 1 cm in diameter, homogenous, well circumscribed, and

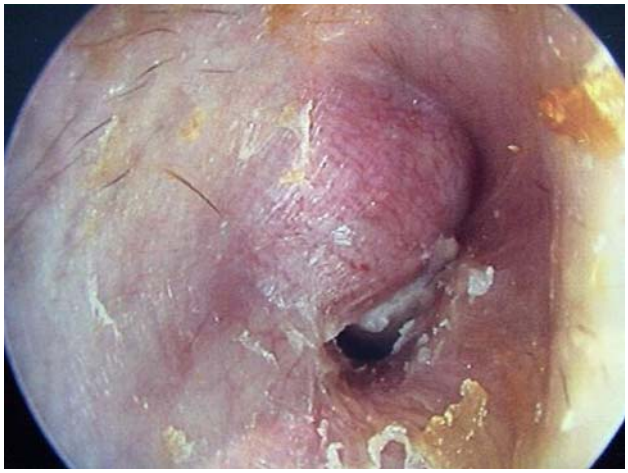

Fig. 1 Right otoscopy. Optique 0°, 4 mm Storz. Postero-inferior, non-pulsatile tumefaction in the posterior wall of the right external auditory canal

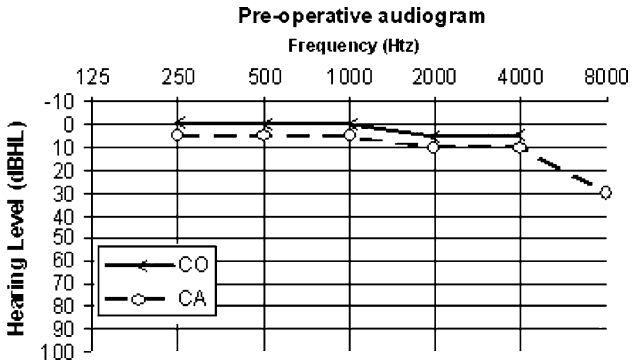

Fig. 2 Pure tone audiogram before surgery

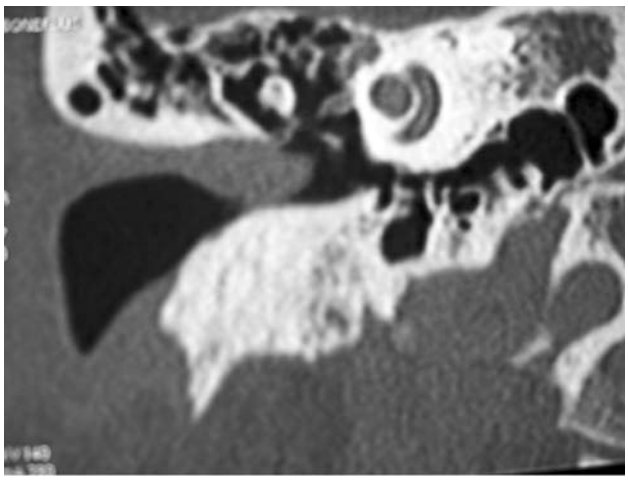

Fig. 3 Coronal computed tomography scan of the temporal bone: well-circumscribed, 1 cm in diameter, homogenous mass homogeneous mass of the tympanic membrane spreading to the external auditory canal

rounded tissue mass in the middle ear, surrounding the incus. No evidence bony erosion was observed. MR scan showed the same mass with similar morphological characteristics. The lesion was iso-intense during T1 with homogenous enhancement following the injection of gadolinium.

The patient underwent a right transcanal tympanotomy. The tumor was soft in consistency, beige and measured 1 cm in diameter. Part of the tumor was found to be adherent to the incus and there was lysis of the cochlearform process. The tumor was removed with the incus. The tympanic segment was grafted to give a type III stapes columella.

Microscopic examination revealed tumor formations dispersed in thin rows within dense conjunctive tissue without any atypical cells. The immunocytochemistry was positive for chromogranine (Fig. 4) and synaptophysine. The final diagnosis was neuroendocrine adenoma of the middle ear.

The post-operative period was uneventful. During follow-up consultations, pure tone audiometry showed a conductive hearing loss of 15 dB in the right ear (Fig. 5). The patient remained free of recurrence at 18 months.

Discussion

Neuroendocrine adenomas of the middle ear are benign epithelial tumors. The clinical presentation is non-specific [1, 10]. Consequently, the final diagnosis is established after histological and immunohistochemical examinations.

The microscopic findings are unencapsulated tumor and moderate cellularity. The tumors are arranged in glandular, trabecular and solid patterns composed of small cells with rounded, regular nuclei containing dense chromatin surrounded by eosinophilic cytoplasm [2, 4, 10]. There are no mitoses or atypical cells [2–4, 8, 10]. The immunohistochemestry we performed revealed the presence of the neuroendocrine markers synaptophysine and chromogranine. The literature describes the presence of these markers as well as vimentin, neuron specific enolase and human pancreatic polypeptid [1, 10]. The tumor cells react with antikeratine antibodies thereby confirming their epithelial nature [10]. When neuroendocrine elements are preponderant, carcinoid tumors have to be considered [7]. However, according to Torske et al. [10], these two entities represent the same tumor in different states of differentiation. Neuroendocrine adenomas are postulated to derive from pluripotent cells located in the mucosa of the middle ear [10].

Given the slow growth rate and the rarity of these tumors, the diagnosis of these tumors is often delayed [1]. The two sexes are affected equally, and the age range extends from 7 to 80 years [1, 2, 8, 10]. The patient described in our case report complaint a sense of fullness in the ear. But the chief complaint is unilateral hearing loss from a month to several years in duration [1–3, 5, 8, 10]. Other symptoms are otalgia, otorrhea, dizziness and tinnitus [1–3, 5, 8, 10]. The lesion can be totally asymptomatic [3, 4]. Otoscopy in most cases has demonstrated an intact tympanic membrane

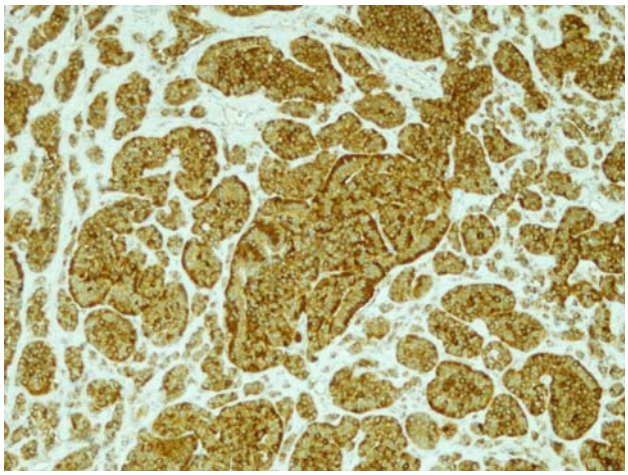

Fig. 4 Immunohistochemical staining revealing positive reaction of chromogranin

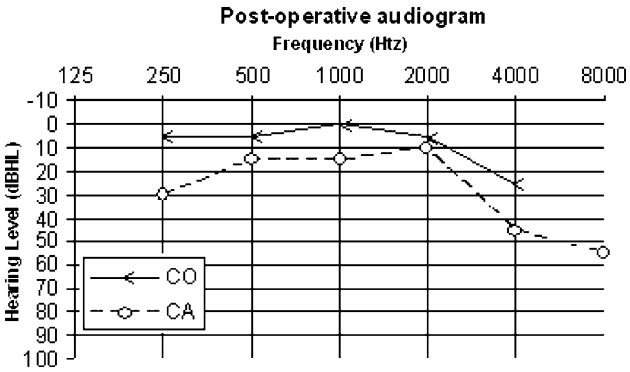

Fig. 5 Pure tone audiometry 10 months after surgery

displaced medially by a retrotympanic mass [2–4]. Several authors have described facial paralysis, which is probably due to nerve compression without invasion [10]. Audiometry reveals a conductive hearing loss [2, 5].

CT scan of the temporal bones usually shows a homogenous, hypodense lesion well limited to the middle ear [6]. The radiologist verifies the integrity of the ossicular chain. Ossicles can be embedded in the mass [6]. Sometimes bone or ossicular erosion can be observed [2, 5]. Indeed, osteolysis and facial nerve involvement seem to indicate a poor prognosis. When the temporal bone is involved, the clinician should eliminate a number of differential diagnosis including paraganglioma, meningioma, cholesteatoma, adenocarcinoma and neurinoma of the facial nerve. MRI reveals an iso-intense mass during T1 with homogenous enhancement following the injection of gadolinium but does not provide any additional information than that provided by a CT scan of the temporal bone. However, in case with voluminous tumors, it can reveal an extension to the posterior cerebral fossa and the cerebello-pontile angle [6].

Surgical excision is the only curative treatment of the lesion [2, 4, 10]. The treatment consists of complete surgical excision with removal of the ossicular chain, if involved [2, 10]. It can be monitoring by rigid optics 0°, 30°, and 70° [2, 5]. The exploration usually discovers an elastic, avascular, pale yellow mass measuring an average of 8 mm [10]. Surgery should be tailored on the basis of the clinical and radiological findings and generalizations about treatment are difficult. But, as suggested by Jahrsdoerfer et al. [4], we think a transcanal tympanotomy may be performed if the lesion is small and confined to the middle ear cleft. If the lesion fills most of the middle ear, a facial recess approach mastoidectomy is recommended [4, 5]. Torske et al. [10] reported a recurrence rate of 18% in their series and underlined the fact that the ossicular chain was left intact in all their patients. [2]. So complete surgical removal of the neoplasm, to include encased ossicles, should be the treatment of choice. Tympanoplasty can be immediately performed during the same operating time. In our case, we performed a type III tympanoplasty since we removed the incus and had a good functional result. No adjuvant treatment is necessary [2, 5].

The patient must be followed closely for possible recurrent disease. However, recurrence is rare with initial complete excision, it frequently appears after incomplete removal of the tumor. The average interval for the recurrence is 158 months [9]. Metastasis are extremely rare and even controversial [1, 9, 10]. A regular long term patient follow-up with otoscopy and audiometry is recommended. Some authors performed systematically CT or MRI examination in order to reveal recurrences [8].

Conclusion

Neuroendocrine adenomas of the middle ear have a nonspecific clinical presentation and the diagnosis should be considered when any retrotympanic tumor is discovered. The final diagnosis is based on microscopic and immunohistochemical examination. In general, complete excision is required and the patient must be followed closely for possible recurrent disease.

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