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Case Report

Middle Ear Carcinoid Tumor : Case Report and Literature Review

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SUMMARY

We herein report a rare case of a carcinoid tumor observed in the middle ear. The 51-year-old female patient presented to our hospital with an 8-year history of mild deafness and sensation of obstruction within the left ear. Otoscopic examination revealed a pinkish mass in the hypotympanum, which was visible through an intact tympanic membrane. Computed tomography (CT) of the temporal bone showed a well-circumscribed mass of soft tissue density filling the left hypotympanum. The provisional clinical diagnosis was cholesteatoma or another type of tumor (e.g. glomus jugulare tumor or facial nerve neurilemmoma). Transcanal tympanotomy was performed under general anesthesia to explore the lesion. At surgery, a well-delineated, encapsulated pinkish mass was found in the hypotympanum. It was not adherent to the tympanic membrane or ossicles. The mass arose, via a narrow pedicle, from the lining epithelium at the promontory. Complete resection of the mass was performed with careful clearance around the ossicles and epithelium at the promontory. Histological examination showed a carcinoid tumor composed of islands, cords, and nests of tumor cells in a fibrous stroma. Immunohistochemistry revealed granular cytoplasmic positivity for chromogranin A. No general symptoms of carcinoid syndrome were observed in this patient. Endoscopic examination revealed no evidence of other carcinoid tumors in the gastrointestinal and respiratory tracts. Eight years postoperatively, no evidence of recurrence or metastatic disease has been observed.

Key Words : Carcinoid tumor, middle ear, chromogranin A

INTRODUCTION

Carcinoid tumors have been reported in a wide range of organs, particularly within the gastrointestinal and respiratory tracts, although in the middle ear region, carcinoid tumor is thought to be a rare neoplasm apparently derived from the lining epithelium of the middle ear. It was first described in 1976 by Derlacki et al.[1], and further characterized in 1980 by Murphy et al.[2]. This neoplasm most likely originates from pre-existing neuroendocrine tissue or from primitive precursor cells in the middle ear[3]. The typical clinical manifestations are progressive conductive hearing loss, a sensation of obstruction of the ear, tinnitus, and pain[4]. This paper reports a case of middle ear carcinoid and describes the appearance on preoperative computed tomography (CT), detailed intraoperative findings, and pathological features, and reviews the relevant literature concerning this rare tumor.

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CASE REPORT

A 51-year-old female patient presented in 1997 with

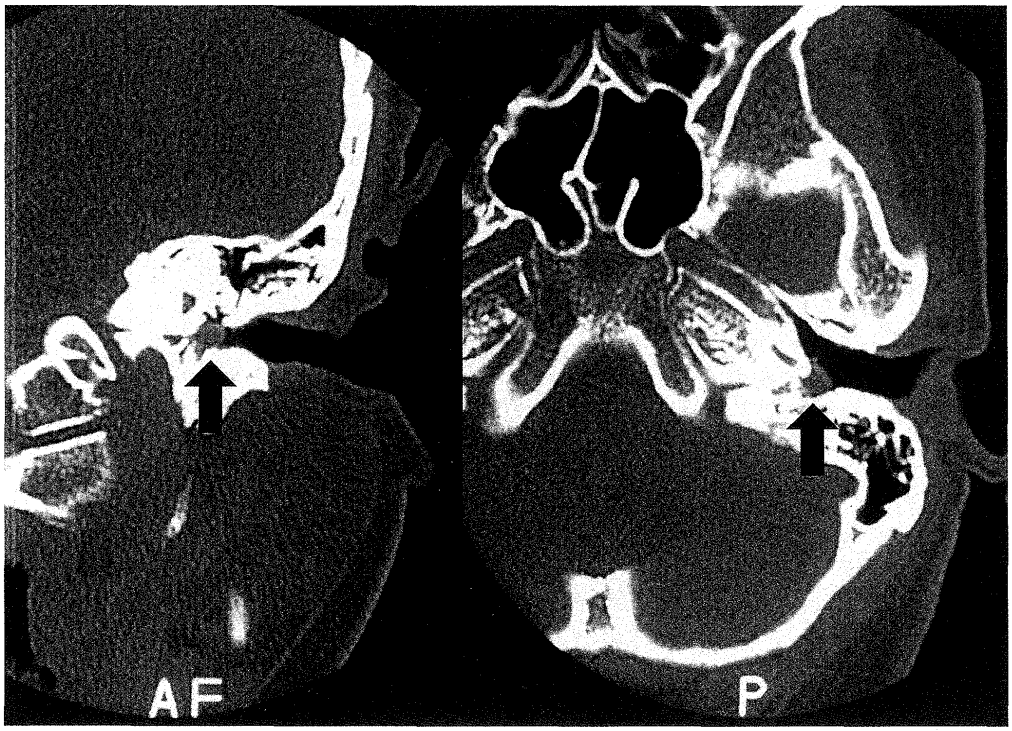

Fig. 1 CT images : coronal view (left) and horizontal view (right) revealed a mass of soft tissue density in the left hypotympanum (arrow). No evidence of bone destruction or erosion was visible adjacent to this lesion.

an 8-year history of mild left-sided hearing loss and sensation of obstruction. Otoscopic examination revealed a pinkish, pearly mass in the left hypotympanum. This lesion was visible through an intact and thin tympanic membrane. Pure tone audiometry revealed a 30 dB mixed hearing loss in the left ear. Computed tomography (CT) of the temporal bone demonstrated a well-circumscribed mass of soft tissue density almost completely filling the left hypotympanum (Fig. 1, arrow). No evidence of bone destruction or erosion was observed adjacent to this lesion. The mastoid air cells were intact and well-developed. A provisional clinical diagnosis of cholesteatoma or another tumor (such as glomus jugulare tumor or facial nerve neurilemmoma) was made.

Transcanal tympanotomy was performed under general anesthesia for exploration of this lesion. When the tympanic membrane was carefully elevated with the malleus handle from the annulus (Fig. 2A, asterisk), a well-delineated, encapsulated pinkish mass measuring about 0.8 × 0.5 cm was found in the hypotympanum (Fig. 2A, arrow) . It was not adherent to the tympanic membrane or ossicles. Operative findings suggested that the lesion was not a typical cholesteatoma because of the absence of a distinct lamellar keratin structure. The tympanic segment of the facial nerve was not involved. The possibility of glomus jugulare tumor was thought negligible as low vascularity was visible under the operating microscope. The mass, which had a narrow pedicle, arose from the lining epithelium at the promontory. Complete resection of the mass was performed with careful clearance around the ossicles and epithelium of the promontory (Fig. 2B). The ossicular connection was confirmed to be intact and the operation was completed successfully.

The pathological diagnosis was a middle ear carcinoid tumor, based on the following histological and immunohistochemical features. Microscopy revealed a carcinoid tumor composed of islands, cords, and nests of tumor cells in a fibrous stroma. Typical structures of this tumor seemed indistinctive since the tissue was affected with crushing damages of surgical procedure (Fig. 3A). Tumor cells exhibited minimal pleomorphism and had round to oval shaped dark nuclei and eosinophilic granular cytoplasm. Mitotic figures and necrotic foci were absent. Immunohistochemistry revealed distinct positivity for chromogranin A corresponding to granular cytoplasm (Fig. 3B).

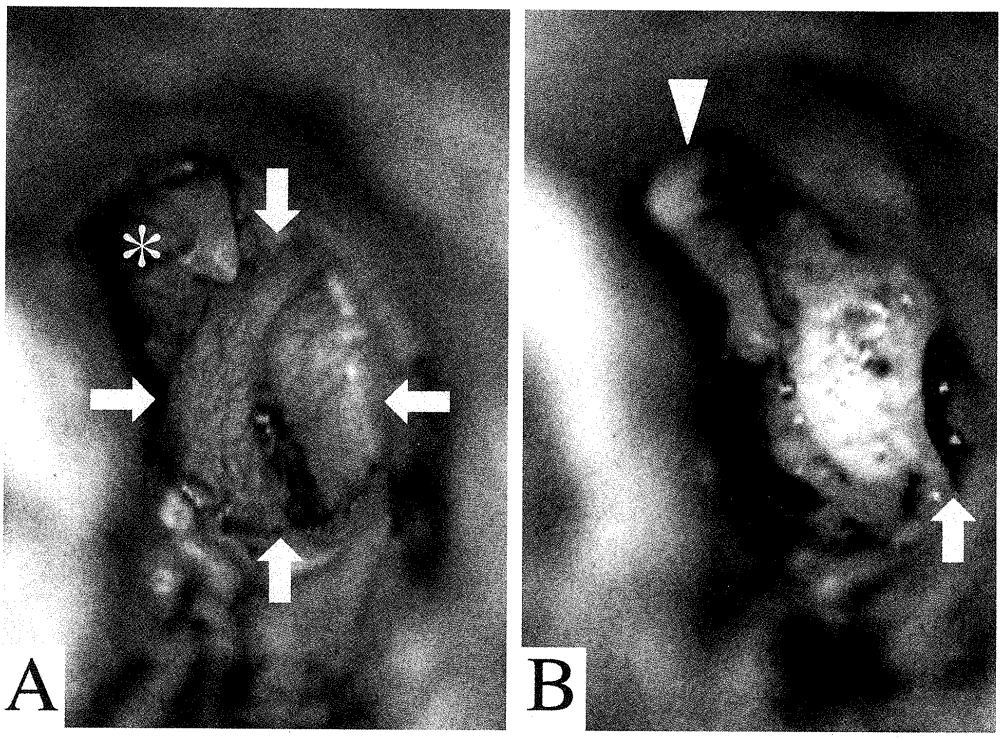

Fig. 2 A) Elevated tympanic membrane with malleus handle from the annulus (asterisk). A well-delineated, encapsulated mass measuring about 0.8 × 0.5 cm was found in the hypotympanum (arrow).
B) After removing the lesion. The bright promontory was observed between the round window niche (arrow) and malleus (arrowhead).

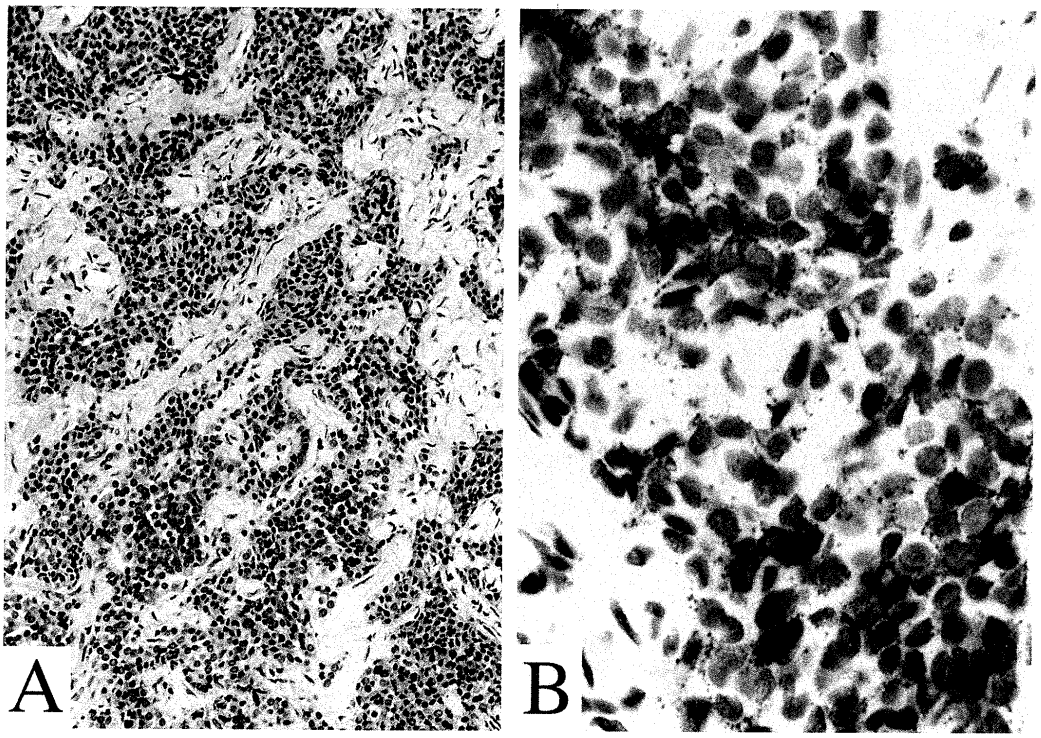

Fig. 3 A) Histological examination showed a carcinoid tumor composed of islands, cords, and nests of tumor cells in a fibrous stroma (HE stain, × 100).
B) Immunohistochemistry revealed granular positivity for chromogranin A corresponding to granular cytoplasm (× 400)

No general symptoms of carcinoid syndrome (e.g. tachycardia, diarrhea, facial rash, generalized edema, or ascites) were observed in this patient. Endoscopic examination was performed to screen for other carcinoid tumors in the gastrointestinal and respiratory tracts, but none were observed. No evidence of recurrence or metastatic disease has been noted eight years postoperatively.

DISCUSSION

In this report, we discuss the pre and postoperative diagnosis, histogenesis, and management of middle ear carcinoid tumor. This rare tumor was first described in 1976 by Derlacki et al.[1] as an adenomatous tumor of the middle ear and was further characterized in 1980 by Murphy et al.[2]. Carcinoid tumors have been reported in a wide range of organs, most commonly in the aero-digestive tract[5, 6]. To the best of our knowledge, more than twenty cases of middle ear carcinoid tumor have been reported in the literature[7~16]. However, the true incidence is hard to assess since this tumor is difficult to distinguish from other middle ear adenomatous tumors by histopathological appearance[3, 10].

The diagnosis of middle ear carcinoid tumor was difficult to make preoperatively. The present case was visualized as a pinkish, pearly mass in the hypotympanum through an intact tympanic membrane. Although most patients with this tumor present with a variety of otological symptoms such as hearing loss, obstruction of the ear, tinnitus and pain, these symptoms are also observed in various other otic diseases including cholesteatoma. On the basis of the present clinical and radiographic findings, our previous experience led us to a provisional preoperative diagnosis of cholesteatoma or other tumor (such as glomus jugulare tumor or facial nerve neurilemmoma). Derlacki et al. reported a similar case[1], which was preoperatively diagnosed as cholesteatoma. Unlike carcinoid tumor of other regions, most cases involving the middle ear do not present with typical carcinoid syndrome, probably due to the small size of the primary tumor[11]. Latif et al.[9] reported a unique case in which systemic symptoms of carcinoid syndrome disappeared after surgery. Since carcinoid usually appears as a mass, detailed inspection of intraoperative findings through an operating microscope is so important to assess the lesion.

Histopathological examination is required for definitive diagnosis. Carcinoid tumor demonstrates islands, cords, and nests of tumor cells with minimal pleomorphism. Carcinoid tumor is thought to originate from neuroendocrine cells (Kulchitsky cells), which are considered to develop from endodermally derived pluripotential stem cells. Middle ear carcinoid tumor is actually histopathologically similar to carcinoid tumor of other organs[5] ; while the mucosa of the middle ear is also derived from endoderm, neuroendocrine cells are not noted within the healthy or inflamed middle ear cavity. The existence of undifferentiated and unidentified pluripotential cells in the middle ear has been hypothesized to explain the origin of middle ear carcinoid tumor[3]. In the present case, surgery revealed that the mass had arisen, via a narrow pedicle, from the lining epithelium at the promontory. This suggested that the tumor may have developed in the mucosa of the middle ear, thus endorsing the above-mentioned hypothesis.

Various reviews of the treatment and prognosis of middle ear carcinoid tumor have been published[4, 11, 12]. Manni et al.[4] reported that conservative surgery with radical removal of the tumor was the treatment of choice because this strategy had resulted in disease-free survival for two to 16 years for their patient series. To the best of our knowledge, most cases of middle ear carcinoid tumor have been treated surgically and have exhibited clinically benign behavior (slow growth and very low tendency to invade). In our case, the tumor was considered to have developed over several years because the patient presented with an 8-year history of deafness and obstruction. On the other hand, metastatic potential has been reported in certain cases. Mooney et al.[16] reported a case of middle ear carcinoid tumor with cervical lymph node metastasis that recurred 9 years after initial treatment. Menezes et al.[14] reported a case with metastases in the parotid gland and cervical lymph nodes. Hence, this tumor may be capable of invasion and metastasis, and therefore warrants long-term follow up. Moreover, Mandigers et al.[13] described a case of typical carcinoid tumor in the jugulotympanic region, in which the patient's clinical condition deteriorated. Histological and cytological appearance are not a reliable criteria for determining malignant potential[12]. In the present case, careful

and complete removal was possible because the tumor was not adherent to the ossicles or tympanic membrane. Histology showed a typical carcinoid tumor with positive chromogranin A staining, and no evidence of recurrence or metastasis was observed for eight years postoperatively. The above-mentioned reports suggest that long-term follow-up is very important in order to clarify the biological behaviour of middle ear carcinoid tumor.

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