

Brazilian Journal of
OTORHINOLARYNGOLOGY

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CASE REPORT

Neuroendocrine adenoma of the middle ear confused with congenital cholesteatoma[☆]

Adenoma neuroendócrino de orelha média confundido com colesteatoma congênito

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Received 29 October 2012; accepted 23 March 2013

Introduction

Neuroendocrine adenomas of the middle ear (NEAME) are rare tumors.¹⁻⁴ Whereas neuroendocrine cells are normally found in the lung and gastrointestinal tract, they are normally not found in the middle ear (ME) mucosa.¹⁻⁶ Although neuroendocrine tumors were initially believed to originate from the neuroendocrine cell system, it is now known that they may originate from tissue that lacks neuroendocrine cells, such as the ME mucosa, because these tumors are derived from pluripotent stem cells that are capable of neuroendocrine differentiation.^{1,5}

Case presentation

A 27-year-old female presented with a three-month history of fullness in the right ear. The patient visited a local hospital and was diagnosed as congenital cholesteatoma. The patient was referred to this hospital. Upon otoscopic examination, the right tympanic membrane (TM) was found to be bulging, with a whitish ME mass (fig. 1A). Pure tone audiometry revealed air and bone conduction thresholds of 20 and 12.5dB HL, respectively, on the right side. However, the thresholds at 4 kHz and 8 kHz were elevated. The computed tomography

(CT) scan showed soft tissue density in the ME cavity, and there was no evidence of bony destruction (fig. 1B).

A canal wall up mastoidectomy and tympanoplasty were performed. During the operation, a soft, pale, well-encapsulated mass was found filling the tympanum and encasing the stapes. The incus and stapes were extracted for complete removal of tumor. The footplate of the stapes was repositioned; the incus was remodeled and then positioned on the stapes footplate. The specimen was soft and relatively tougher than cholesteatoma, and there was no keratin material.

The histopathologic results revealed a well-differentiated neuroendocrine neoplasm. Routine light microscopy revealed a proliferation of small monotonous cells arranged in trabecular patterns. There were no findings of necrosis, nuclear atypia, and mitosis (fig. 1C). Immunohistochemical stainings were positive for chromogranin, synaptophysin, neuron specific enolase, vimentin, CD56, and cytokeratin (fig. 1D-E).

The final diagnosis was revised from congenital cholesteatoma to NEAME. The patient did not present any symptoms related to carcinoid syndrome. Chest and abdominal CT were performed, as well as an assessment of the urinary catecholamines and their metabolites for investigating distant metastases and simultaneous carcinoid tumor in other organs. The catecholamines and their metabolites were fou-

[☆]Please cite this article as: Kim YH, Lee SH, Jung DJ. Neuroendocrine adenoma of the middle ear confused with congenital cholesteatoma. Braz J Otorhinolaryngol. 2014;80:180-1.

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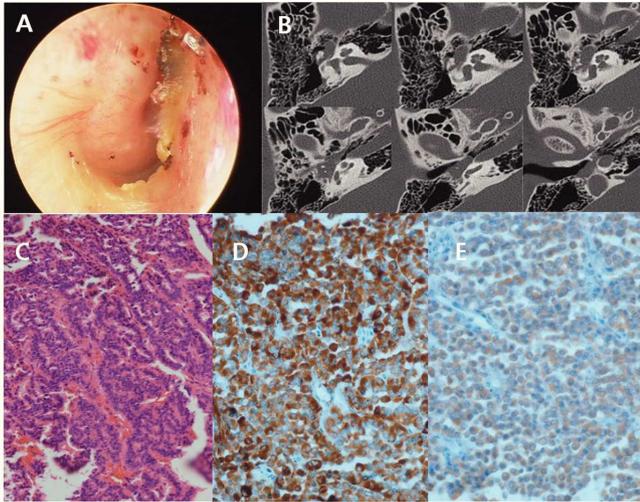


Figure 1 A, Preoperative otoscopic finding shows whitish mass behind the tympanic membrane. B, Preoperative axial view of temporal bone CT shows soft tissue density in the middle ear cavity. C, Tumor is composed of trabeculae lined by multiple layers of small and monotonous cells with an intraluminal eosinophilic secretion (H&E stain $\times 200$). D, Immunohistochemical stain for chromogranin shows strong positivity within tumor cells (chromogranin $\times 400$). E, Immunohistochemical stain for synaptophysin shows strong positivity within tumor cells (synaptophysin $\times 400$).

nd within the normal limits and chest and abdominal CT did not reveal any mass lesions. The patient has been followed for 30 months without evidence of recurrent disease.

Discussion

NEAME can be confused with glomus tympanicum and congenital cholesteatoma, as was the case with the present patient. It is difficult to diagnose NEAME before the final histological examination, since the clinical manifestations and radiographic findings are nonspecific.

The pathologic diagnosis of NEAME is primarily based on light microscopy with hematoxylin-eosin staining; the diagnosis is confirmed by immunohistochemical investigation.^{1,2} On immunohistochemical staining, epithelial markers include cytokeratins, and neuroendocrine markers include chro-

mogranin, synaptophysin, neuron specific enolase, vimentin, and CD56.⁵ Angouridakis et al. suggested that NEAME were positive for epithelial and neuroendocrine markers, while carcinoid tumors of the ME were negative for epithelial markers and positive for neuroendocrine markers.⁶ Saliba et al. proposed that NEAME were positive for immunohistochemistry and there was no evidence of metastasis, whereas carcinoid tumors of the ME were positive for immunohistochemistry and there was evidence of metastasis or carcinoid syndrome.⁵ In the present case, the tumor cells were positive for epithelial marker and neuroendocrine markers. There was no evidence of metastasis and carcinoid syndrome. Therefore, the tumor of present case is best described by the term NEAME.^{5,6}

Final remarks

NEAME are uncommon. The diagnosis of NEAME is made by the presence or absence of immunohistochemical markers, metastases, and carcinoid syndrome.

Conflicts of interest

The authors declare no conflicts of interest.

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