Neuroendocrine Adenoma of Middle Ear Masquerading as a Middle Ear Vascular Lesion On Clinical Picture: Case Report and Review of Literature

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Abstract

Middle ear Neuroendocrine adenomas are rare tumors that can be easily confused as glomus tympanicum of the middle ear. 33-year-old man with no comorbidities who presented with complaints of right ear foreign body sensation and was later diagnosed as a neuroendocrine adenoma of the middle ear.

Introduction:

Neuro endocrine adenoma of the middle ear (NAME) is one of the rare tumors of the middle ear which was mentioned by Treitel in 1898.[8,9]Diagnosis of this tumor was under controversy over its histologic origin and have led to inappropriate diagnosis such as ceruminoma, ceruminous adenocarcinoma, ectopic salivary gland tumor or adenocarcinoma. Until recently when it was reported by Derlacki and Barney1 in 1976 as neuro-endocrine adenoma of middle ear.[6,7]

These tumors are very rare, and the diagnosis of middle ear adenoma is often delayed as patients present with vague or no symptoms to variable symptoms. The lesion can occur in different age groups and both sexes are equally affected. Very rare cases of facial nerve involvement have been reported in literature[4]. In our case, the patient did not have any specific symptoms other than mild discomfort in the left ear. On otoscopy there was whitish mass seen behind the tympanic membrane which was further shown on CT Temporal bone as a small round benign looking mass in hypotympanum.

Our objective with this case report is to show that neuroendocrine masses in the middle ear adenomas can be asymptomatic and diagnosis can be misled to congenital cholesteatoma or glomus jugulare as well these tumors can be excised with minimal damage to normal anatomy of the middle ear cleft.

Case Description:

33 years old male presented to clinic with complaint of left ear discomfort, He did not complaint of any ear pain, ear discharge or hearing loss. There was no dizziness or tinnitus. On examination of right ear, there was white lesion behind tympanic membrane posterio-inferior quadrant. Left ear was normal. PTA done revealed hearing within normal limits (Fig 1).

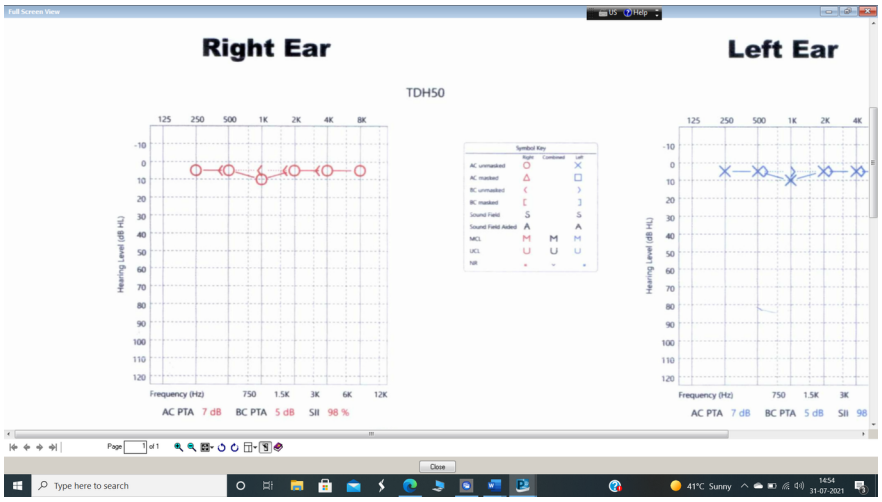

Fig 1 , *Pure Tone audiometry revealed normal hearing*.

Imaging:

CT petrous(FIG 3-4) bone displayed small oval shaped well defined mass in posterio-inferior aspect of the middle ear.

MRI Internal Auditory Meatus (fig 2) was done which showed nodular lesion of 3 mm within right middle ear cleft overlying dome of jugular bulb. Patient was planned for middle ear exploration and excision of middle ear mass. Transcanal approach was utilized ti expose the mass. During the surgery, we found a small whitish mass lying on the jugular bulb but easily separable from jugular bulb. ossicle chain was unaffected(fig 5,6). Middle ear cleft mucosa was normal and healthy with intact facial nerve.

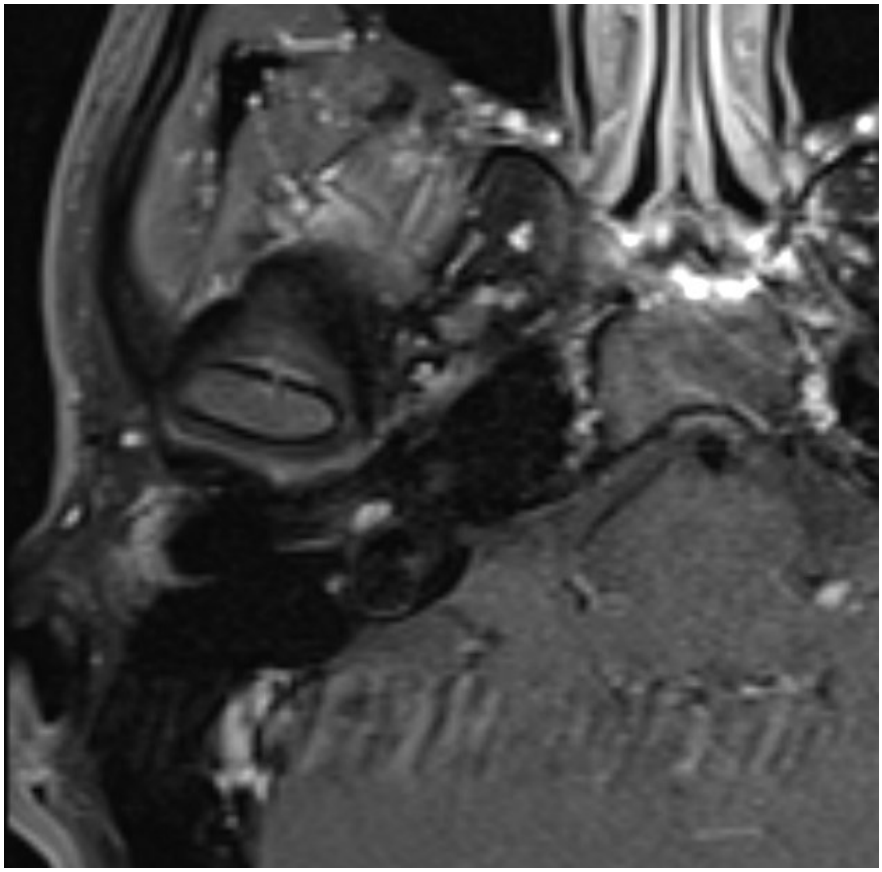

Fig 2, T2 MRI sagittal view showing tiny nodular lesion about 3 mm in diameter, within the inferolateral aspect of right middle ear cleft overlying the dome of the right internal jugular vein, showing intense postcontrast enhancement.

Fig3, CT temporal bone coronal view showing small mass in the hypotympanum. (arrow)

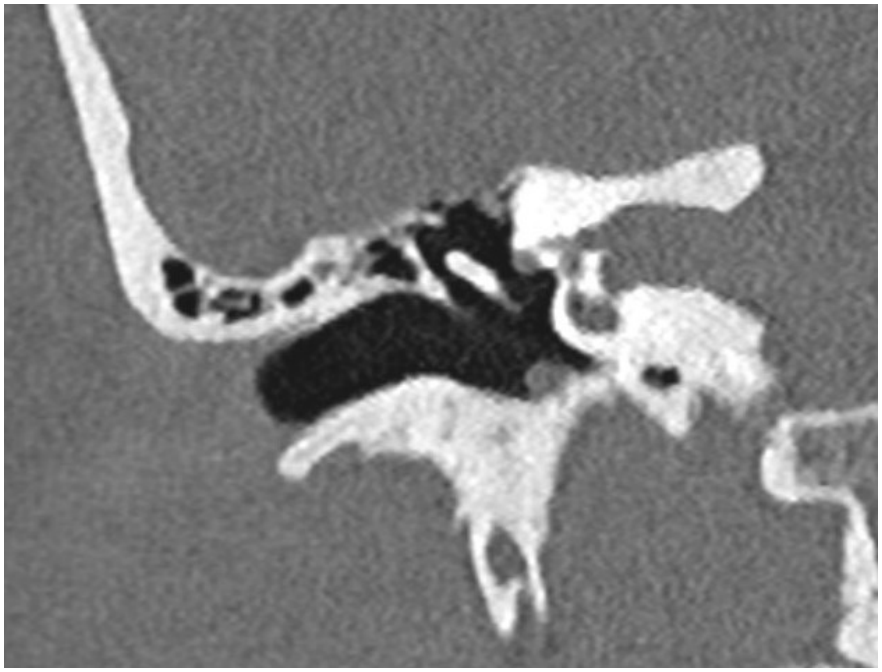

Fig4, CT temporal bone coronal view showing adenoma in middle ear over jugular bulb in hypotympanum, not affecting ossicular chain

Intra-operative images:

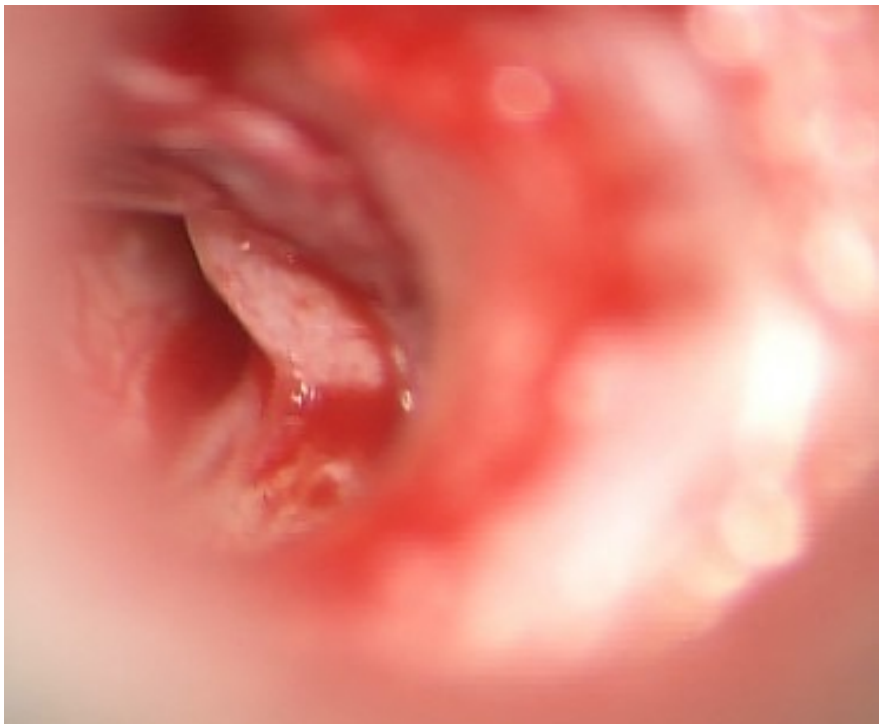

Fig5, Large arrow showed mass in hypotympanum.

Small arrow showed jugular bulb.

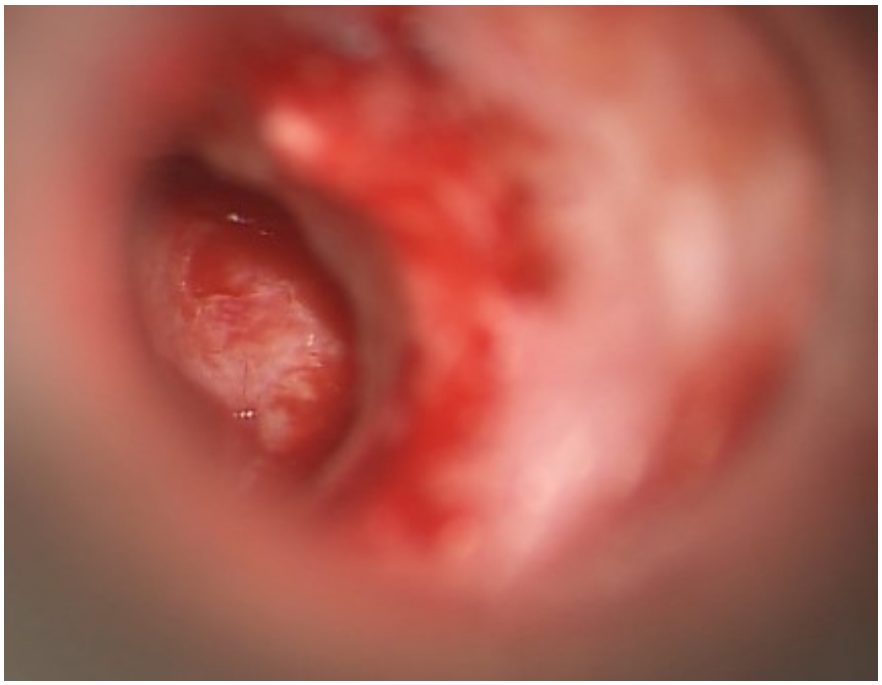

Fig 6, After Curetting the bone (inferior wall), mass is more visible in hypotympanum.

The patient had an uneventful postoperative period and was discharged the next day. The patient was followed up monthly for 3 months post-surgery which revealed no recurrence.

Histopathology:

Histopathology showed tubular and cribriform proliferation of cuboidal cells as an epithelium with granular amphophilic cytoplasm with round dark blue nuclei. There was no evidence of necrosis, atypia or increased mitotic activity.

The tumor cells demonstrated positive staining for synaptophysin, and cytokeratin (AE1/AE3) also display patchy positivity for CD 56 and EMA and No sustentacular cells were identified are negative for S100. The tumor had a low proliferation index of 1%.

Discussion:

Neuroendocrine adenomas of the middle ear are benign epithelial tumors. Clinical presentation is nonspecific and vary in each patient. Consequently, the diagnosis is established after histological and immunohistochemical examinations[5-7].

Some of the important differential diagnosis for Neuroendocrine adenoma of middle ear includes glomus tympanicum, glomus Jugulare, congenital cholesteatoma which are difficult to exclude on imaging or clinically. Thus, they are usually diagnosed by histopathology. In Our case, the Radiological imaging was suggestive of glomus tumor. However, on Histopathology, it was proved otherwise.

The pathogenesis of this tumor is still under discussion as different authors have proposed multiple origins. Some have a view that they are originated from the cells of the mucosa of the middle ear. Some are suggesting that they developed from off-site embryonic nests of glandular cells in the middle ear mucosa.[7,8,17]Recently Katabi and Torske and Thompson proposed that it may originate from undifferentiated pluripotential endodermal stem cells since epithelial cells with neuroendocrine differentiation do not exist in the middle ear.[16,17]

The most preferred treatment option is surgical excision with Middle ear exploration. [10-15] If Ossicular chain is involved, tympanoplasty with ossicular chain removal is treatment of choice, depending upon the involvement of the ossicle. If lesion is small, localized to middle ear cleft and is not invading surrounding structures like in our case, excision with trans-canal approach can be performed. If the lesion involves most of the middle ear cleft, tympanoplasty with mastoidectomy is recommended. [10,11]

Conclusion

Neuroendocrine adenoma of middle ear is a rare tumor. It is slow growing, benign epithelial tumor. It can be asymptomatic, or symptoms may be non-specific and can be misdiagnosed as congenital cholesteatoma or glomus tympanicum or glomus jugulare. The definite diagnosis is established by histopathology and immunohistochemistry.[14,16]The definite treatment is surgical excision. Surgical Approaches depend upon lesion involvement in the middle ear cleft. If the tumor is small and not invading surrounding structures, there is no need for radical surgery. Complete excision of tumor can be possible without disturbing normal anatomy of middle ear cleft. However, Intra-operative use of endoscope 30 degree or 45 degrees to exclude any remnant is important. In addition, post-operative follow up plays significant role to rule out recurrence. It can be combined clinically with CT/MRI remains the best available method for controlling recurrence. [13-15]

Consent Statement:

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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