Neuroendocrine Tumor of the Temporal Bone with Middle Cranial Fossa Spread

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Abstract

Introduction: Neuroendocrine tumors are rare neoplasms of the ear and temporal bone. These tumors are typically low grade and conventionally thought to have a small risk of invasiveness. This case study is a report of a patient who presented with a right middle ear mass and middle fossa lesion.

Methods
Retrospective case presentation

Results
A 35-year-old male with partially recovered right facial paralysis and progressive right-sided hearing loss, pain, and bloody otorrhea. The patient had a House-Brackmann Grade III facial function on the right and a soft tissue mass completely filling and extruding from the right external auditory canal (EAC). A temporal bone CT showed a soft tissue mass filling the middle ear and EAC and MRI revealed a middle fossa lesion. The patient underwent a right temporal bone resection and ear canal oversew. Pathology showed MeNET. The patient then underwent Dotatate PET/CT which showed focal uptake in the right middle cranial fossa at site of previously identified temporal lobe lesion. The patient subsequently underwent a right middle fossa craniotomy with intradural resection of this lesion. Intraoperative frozen section was consistent with neuroendocrine tumor.

Conclusions
MeNET is a rare primary ear tumor initially characterized as having low potential for metastasis, however several case series over the years have reported higher rates of local recurrence and distant metastasis. Surgical resection is the optimal primary treatment for these neoplasms, but adjuvant radiotherapy has been described in the literature for subtotal resection, recurrence, and distant metastasis. We present a young male initially thought to have right cholesteatoma with a separate right temporal lobe meningioma.

Introduction

- Neuroendocrine tumors, generally found in the digestive and respiratory tracts, are rare neoplasms of the ear or temporal bone.
- Middle Ear Neuroendocrine Tumors (MeNET) represent less than 2% of all primary ear tumors.[1]
- These tumors are typically low grade and conventionally thought to have a small risk of invasiveness and low metastatic potential.[2,3]
- We report a patient a MeNET of the right middle ear and the temporal lobe.

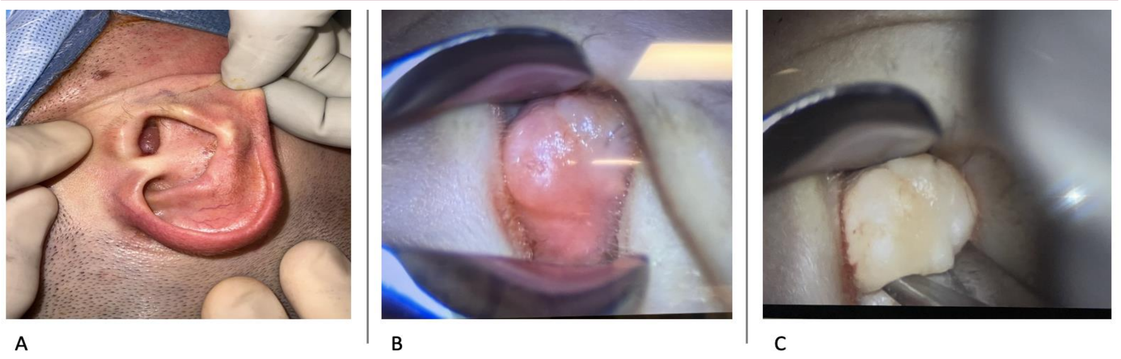
Methods and Materials

- Retrospective case presentation

Case Presentation

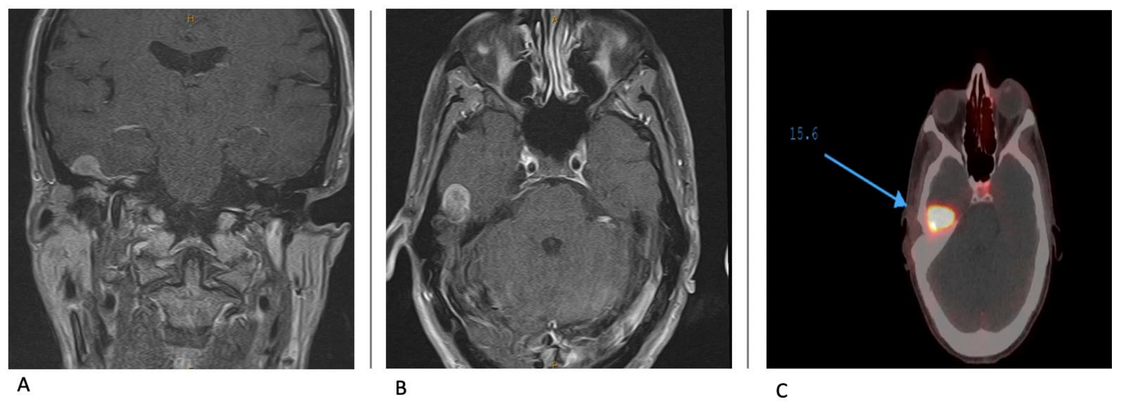
- A 35-year-old male presented with one episode of partially recovered facial paralysis, progressive right-sided hearing loss, pain, and bloody otorrhea.
- The patient had House-Brackmann Grade III facial function on the right and a soft tissue mass completely filling and extruding from the right external auditory canal (EAC).
- Initial CT showed soft tissue filling the mastoid antrum, middle ear, and EAC, thought to be cholesteatoma. MRI revealed a temporal lobe lesion, consistent with a meningioma.
- The patient underwent right temporal bone resection and ear canal oversew with findings including large polypoid mass in the EAC, soft tissue filling the middle ear attached to the promontory, and dehiscent tympanic segment of facial nerve. A cholesteatoma was also found in middle ear and mastoid antrum.
- Pathology confirmed a MeNET with lymphovascular invasion.
- Dotatate PET/CT showed focal uptake in the right temporal lobe. Additional uptake was noted in the left thyroid lobe, which may represent additional focus of disease.
- Interval MRI showed an increase in size of the right middle cranial fossa dural based tumor.
- The patient returned to the operating room for a right temporal craniotomy with intradural resection of the infratemporal lesion.
- Intraoperative frozen section was consistent with neuroendocrine tumor.
- Post-operative exams show stable House-Brackmann Grade III facial function and well-healed incisions.
- FNA of thyroid nodule seen on Dotatate scan was suboptimal for evaluation. Patient electing to proceed with close follow-up and repeat imaging
- The patient underwent adjuvant radiation therapy and received 54 Gy in 30 fractions.

Imaging



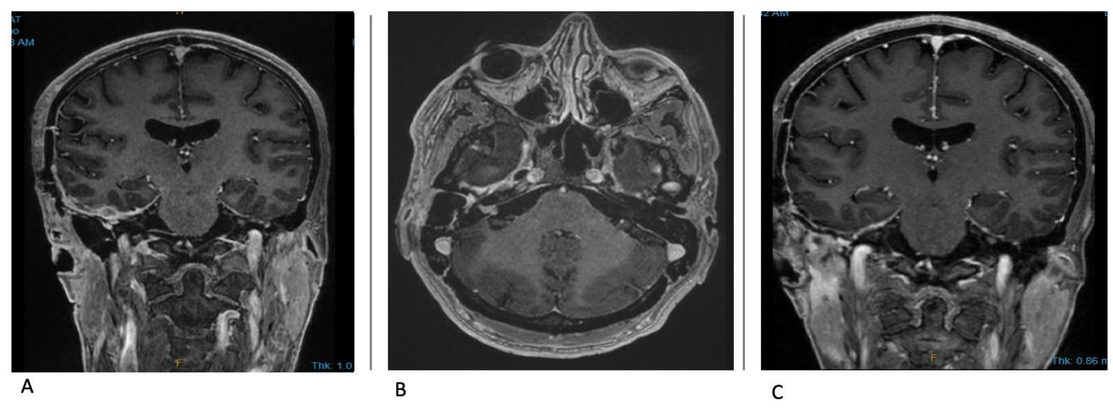
A B C

Figure 1. Intraoperative photos (A), (B), (C) demonstrating large, fleshy, polypoid mas emanating from external auditory meatus.



A B C

Figure 2. Coronal (A) and Axial (B) T1 post-contrast pre-operative images. (A) and (B) demonstrate an enhancing right middle cranial fossa dural based tumor. Axial (C) Dotatate PET/CT after initial surgery with focal uptake in right middle cranial fossa.



A B C

Figure 3. Coronal (A) and Axial (B) T1 post-contrast images at 1 month post-resection. Coronal (C) T1 post-contrast imaging at 6 months post-resection and 3 months post-radiation therapy. Image (A) demonstrates resection of mass with some residual dural enhancement/thickening. Image (B) 7 mm nodular enhancement associated with the 7th and 8th nerve in right IAC. Image (C) shows decreased dural thickening and enhancement.

Discussion

- MeNET is a rare clinical entity, with approximately 150 cases reported in the literature.[4,5]
- These tumors are generally thought to be indolent with slow, local growth, but can be aggressive and metastatic lesions are reported in primary and recurrent tumors.[2,3]
- Generally, MeNET presents as nonspecific ear mass.
- The most common symptoms include conductive or mixed hearing loss, aural fullness, otalgia, tinnitus, and otorrhea. Less commonly, patients present with facial nerve palsy.[1,2,3]
- These tumors mimic more common pathology. Differential diagnosis must include cholesteatoma, jugulotympanic paraganglioma, acoustic neuroma, meningioma, endolymphatic sac papillary tumor, rhabdomyosarcoma, and adenocarcinoma. [5]
- Complete surgical resection is the primary treatment, but there are varying opinions in the literature regarding optimal surgical method.[4,5]
- Despite adequate surgical resection, there is a 15-25 % chance of persistent or recurrent disease and 7-10% rate of metastatic disease.[4,5]
- In patients with metastatic disease or aggressive cases, the treatment should involve a combination of surgical resection and comprehensive therapy, including postoperative adjuvant radiotherapy.[4,5]

Conclusions

- We report a rare case of a MeNET with spread to the middle cranial fossa on initial presentation. This patient underwent surgical resection and adjuvant radiation therapy, but will need long-term follow-up given the relatively high rate of recurrent disease reported in the literature

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