Metastatic Esthesioneuroblastoma of the Temporal Bone

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ABSTRACT

Objectives: Metastasis to the temporal bone from an olfactory esthesioneuroblastoma is extremely rare. We report a case of a metastatic esthesioneuroblastoma lesion to the temporal bone.

Study Design: Case report and review of the literature.

Methods: We reviewed the patients clinical information including presenting symptoms, physical exam findings, imaging, intraoperative findings, and pathologic examination of resected tumor specimen of a patient with esthesioneuroblastoma of the temporal bone are described in detail.

Results: A 48 year old male had a history of esthesioneuroblastoma status post-endoscopic resection and adjuvant radiation in 2012 presented with right otalgia of 4 months duration that had been treated unsuccessfully with multiple rounds of antibiotics. He also reported right sided aural fullness, hearing loss and intermittent vertigo. Shortly thereafter he developed right sided neck masses and House-Brackmann II/VI facial weakness. A large EAC mass was noted on physical examination. CT revealed a large mass in the right temporal bone encasing the facial nerve with associated bony destruction. There was diffuse enhancement of the mass on MRI with adjacent dural enhancement. PET/CT was also performed and was notable for pathologic parotid and bilateral neck nodes. An excisional biopsy of the EAC mass returned positive for esthesioneuroblastoma. A transotic approach to the posterior fossa with extradural resection of tumor was performed in conjunction with a superficial parotidectomy and bilateral neck dissections. Permanent pathology returned esthesioneuroblastoma. The patient underwent postoperative adjuvant radiation.

Conclusions: Esthesioneuroblastoma has the potential for locoregional metastasis and can affect the temporal bone. Treatment consists of surgical resection, adjuvant radiation and in some cases chemotherapy.

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CASE DESCRIPTION

A 48-year-old male had a history of esthesioneuroblastoma (ENB) status post-endoscopic resection and adjuvant radiation in 2012. Four years after completion of therapy he presented with right otalgia of 4 months duration that had been treated unsuccessfully with multiple rounds of antibiotics. He also reported right-sided aural fullness, hearing loss and intermittent vertigo. Shortly thereafter he developed right-sided neck masses and House-Brackmann II/VI facial weakness.

A large external auditory canal (EAC) mass was noted on physical examination. An audiogram was performed and the patient was noted to have a profound asymmetric right sided sensorineural hearing loss with a 70 db pure tone average and normal hearing on the left side. CT revealed a large mass in the right temporal bone encasing the facial nerve with associated bony destruction (Figures 1 and 2). MRI imaging showed diffuse enhancement of the mass with adjacent dural enhancement. PET/CT was also performed and was notable for FDG-avid, pathologic right parotid and bilateral neck nodes. An excisional biopsy of the EAC mass returned positive for esthesioneuroblastoma.

A transcochlear approach to the posterior fossa with extradural resection of tumor was performed in conjunction with a superficial parotidectomy and bilateral neck dissections. Permanent pathology confirmed the initial diagnosis of esthesioneuroblastoma (see Figures 3 and 4). The patient underwent postoperative chemotherapy and adjuvant radiation.

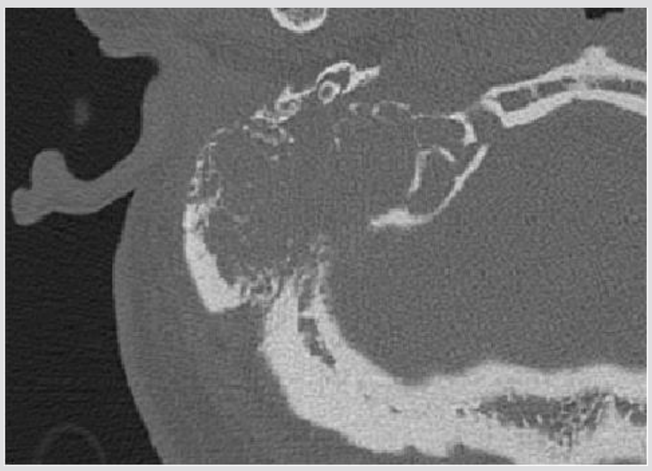

Figure 1. Representative axial CT scan image from initial presentation.

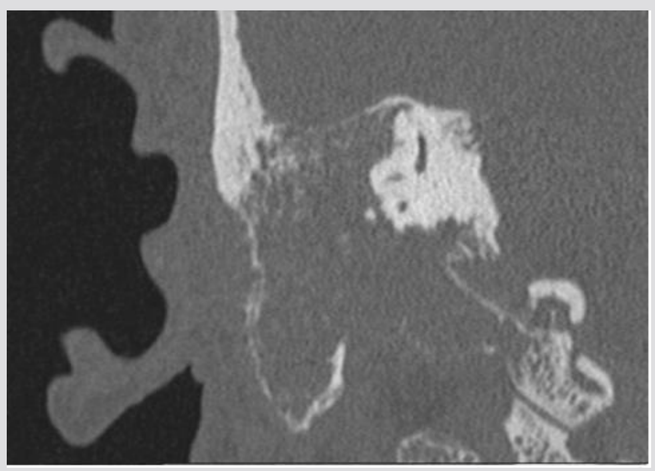

Figure 2. Representative coronal CT scan image from initial presentation.

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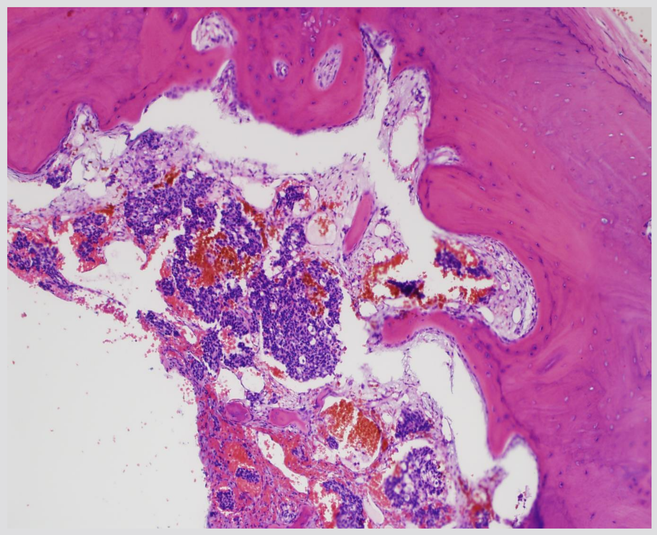

Figure 3. Low power (4x) H+E image of olfactory neuroblastoma within the medullary aspect of the mastoid bone. The nested, small round tumor deposits are morphologically in keeping with this disease recurrence..

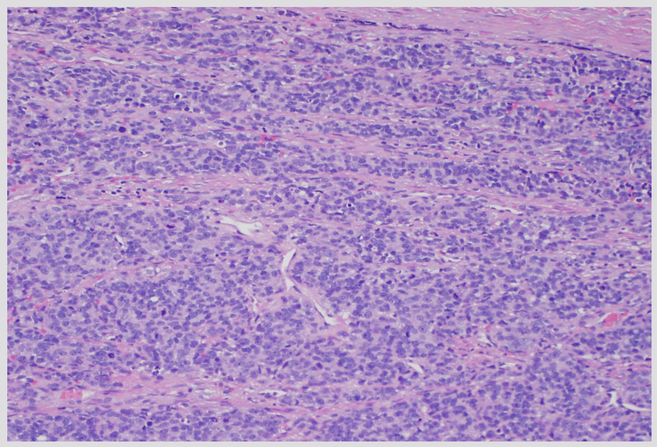

Figure 4. High power (20x) H+E image of the same lesion showing the small round cell malignancy with an appreciable mitotic rate.

HISTOPATHOLOGY

The lesion extended from the temporal lobe dura superiorly, the posterior fossa dura posteriorly, into the upper neck inferiorly, to the jugular foramen medially and to the carotid canal anteriorly. This large lesion was removed in a piecemeal fashion and sent for histopathological analysis. Tumor cells consistent with ENB were identified within the mastoid bone. The tumor involving the medullary aspect of the mastoid bone exhibits a relatively monomorphic proliferation of small round cells with minimal cytoplasm in a nested pattern. (Figure 3) High power histologic examination of tumor-involved dura demonstrates a highly cellular proliferation of similar nested small round cells, morphologically consistent with an esthesioneuroblastoma. (Figure 4) An appreciable mitotic rate was noted, supporting a WHO grade 2 tumor. Not included were supportive immunohistochemical stains revealing positivity for synaptophysin (neuroendocrine marker), while staining for cytokeratins was negative.

DISCUSSION

Esthesioneuroblastoma (ENB) in an uncommon tumor originating from the olfactory neuroepithelium and has a reported incident of 0.4 per million.[1] ENB is responsible for approximately 5% of all malignant sinonasal tumors.[2] There is a wide age range and both males and females are affected equally.[2] Diagnosis is typically made late in the disease course as many patients are typically asymptomatic and symptoms, when present, are generally nonspecific.[3]

ENB is typically treated with surgery and adjuvant radiation. Advances in skull bases surgery and radiotherapy techniques have resulted in improved survival.[2] Consequently, more patients have been observed to develop regional and distant metastases.[4] Distant metastases is reported in approximately 7% of patients on initial presentation; however up to 39% of patients will develop metastatic lesions on follow-up.[4] The most common metastatic sites are cervical lymph nodes, lungs, brain and spine. Metastasis to the temporal bone has not been previously reported.

Definitive diagnosis of temporal bone ENB is made by histopathologic and immunohistochemical (IHC) studies. Microscopic evaluation reveals nests or sheets of small blue cells with scant cytoplasm. IHC analysis typically reveals positivity for S-100 (in a peripheral, sustentacular pattern), as well as neuroendocrine markers chromogranin and synaptophysin.[2] Given the morphologic overlap, a pertinent negative immunohistochemical result for pancytokeratins is usually indicated to exclude a neuroendocrine carcinoma.

Since ENB of the temporal bone is an extremely rare entity there is no agreed upon treatment modality. Tumor size, hearing status, involvement of facial nerve, as well as patients' age and overall health should be taken into careful consideration when selecting treatment modality and surgical approach.

CONCLUSIONS

Esthesioneuroblastoma has the potential for locoregional metastasis and can affect the temporal bone. Treatment consists of surgical resection, adjuvant radiation and in some cases chemotherapy.

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3. Banuchi VE, Dooley L, Lee NYet al. Patterns of regional and distant metastasis in esthesioneuroblastoma. The Laryngoscope 2016; 126:1556-1561.