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Clinical Challenge | PATHOLOGY

Middle Ear Mass Causing Vertigo and Facial Nerve Weakness

Aron Kandinov, BS; Matthew Smith, MD; Syed F. Ahsan, MD

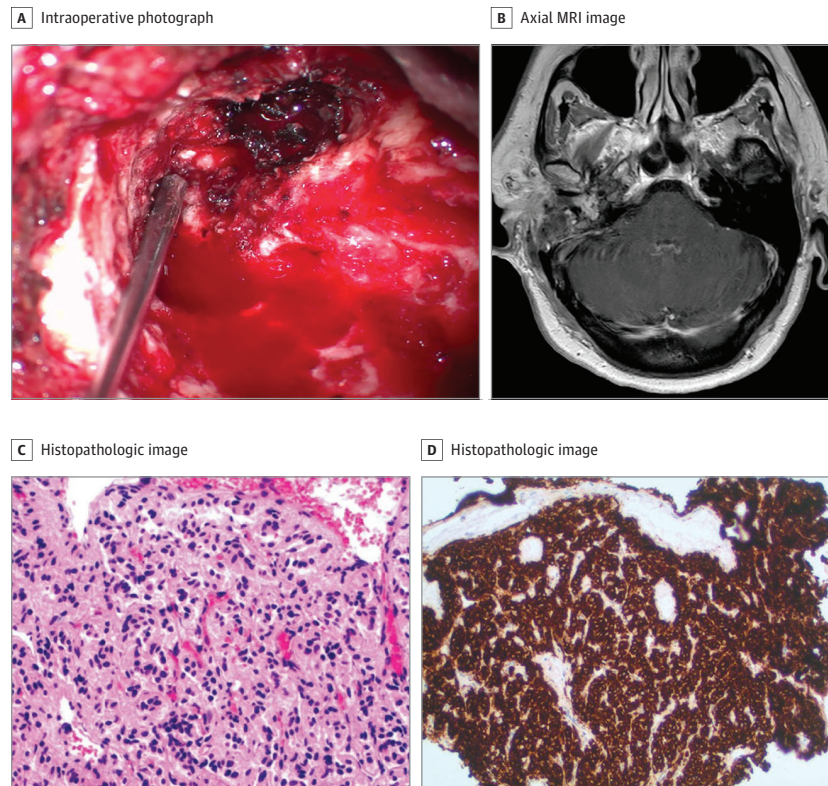


Figure. A, Mastoidectomy, visualization of vascular mass. B, T1-weighted axial magnetic resonance image (MRI) showing enhancement of right middle ear extending to external auditory canal. C, Hematoxylin-eosin, original magnification $\times 40$. Clusters of cells surrounded by vascular tissue. D, Synaptophysin stain, original magnification $\times 40$. Nests of strongly staining neuroendocrine cells separated by vasculature.

An elderly woman presented with 2 episodes of vertigo lasting a few minutes while stationary over 1 day and a several-year history of right-sided hearing loss. She had not experienced otalgia, otorrhea, or tinnitus. Physical examination revealed a mass obstructing the right external auditory canal and House-Brackmann grade 2 facial nerve weakness on the right. Audiogram revealed right-sided moderate to profound mixed hearing loss. Computed tomography revealed a soft-tissue mass emanating from the middle ear involving the facial canal with surrounding bony erosion, including dehiscence of the tegmen, cochlea, bony labyrinth, lateral semicircular canal, and jugular foramen. The patient underwent trans-temporal mastoidectomy and debulking. However, owing to the mass's vascularity and significant bleeding, the operation was aborted, and she was sent for embolization (Figure, A). Magnetic resonance imaging during embolization revealed intense enhancement in the right middle ear extending from the epitympanum to the hypotympanum with extension into the right external auditory canal (Figure, B). She returned to the operating room, and the vascular mass was found to extend inferiorly to the jugular bulb and carotid and anteriorly into the eustachian tube. The mass was adherent to the underlying bone and had invaded the facial canal but not the nerve itself. Both malleus and incus were eroded. Immunohistochemical stains of the specimen indicated cells positive for synaptophysin with a sustentacular pattern of cells positive for S-100 (Figure, C and D).

WHAT IS YOUR DIAGNOSIS?

- A. Middle ear paraganglioma
- B. Facial nerve hemangioma
- C. Facial nerve schwannoma
- D. Middle ear adenoma

Diagnosis**A. Middle ear paraganglioma****Discussion**

Middle ear paragangliomas are parasympathetic neoplasms of naturally occurring paraganglionic bodies. Paragangliomas are slow-growing, invasive, vascularized neoplasms arising from either the middle ear or the jugular bulb, with a small percentage derived from the facial nerve.¹ Middle ear paragangliomas predominate in women, usually presenting in the fifth and sixth decades with biphasic peaks in the fourth and seventh decades.^{2,3} Though most go unreported, the incidence of head and neck paragangliomas has been estimated at 1 per 100 000.^{2,4} Most of these are sporadic, although 10% to 30% are familial and associated with mutations in the succinate dehydrogenase complex.^{3,5}

The hallmark presentation for middle ear paraganglioma includes pulsatile tinnitus (81%-90%), hearing loss (77%-81%), and aural fullness (12%-71%).^{6,7} They often abut or invade nearby cranial nerves, with patients occasionally presenting with facial paresis (0%-3%), dysphasia, hoarseness, and even shoulder or tongue weakness.^{6,7} Physical examination may reveal a fleshy red mass in the middle ear that blanches secondary to pressure from pneumatic otoscopy (Brown sign). Biopsy is contraindicated owing to the mass's vascularity and possible catecholamine secretion.

Parasympathetic paragangliomas in general have a low incidence of catecholamine secretion, with only 1% to 3% of patients presenting with symptoms of catecholamine excess.^{3,8} Patients should nevertheless be questioned regarding symptoms of catecholamine secretion such as palpitations, headaches, and hypertension. While all suspicions for middle ear paraganglioma should be tested for urine metanephrine and vanillylmandelic acid preoperatively, positive results should prompt investigation for pheochromocytoma owing to the low incidence of functional glomus tumors.² Serum epinephrine and norepinephrine levels are also useful in diagnosis, as parasympathetic paragangliomas lack the enzyme phenylethanolamine N-methyltransferase, which converts norepinephrine to epinephrine in their sympathetic counterparts.

Computed tomography with contrast classically demonstrates a homogeneous mass with intense enhancement, and is particularly sensitive for signs of bony involvement. Magnetic resonance imaging with gadolinium enhances soft-tissue detail, resulting in a lower false-negative rate⁹ and better delineating dural and neurovascular involvement. Magnetic resonance images demonstrate T1-isointense signaling and T2-hyperintense signaling with a possible "salt and pepper" pattern in larger lesions owing to flow voids.⁸ Magnetic resonance angiography can further delineate the relationship to the carotid artery, sigmoid sinus and jugular bulb, and identify potential blood supply—often the ascending pharyngeal artery in the case of jugular bulb paragangliomas. The differential diagnosis for a vascular lesion near the geniculate ganglion includes facial nerve hemangioma, which also presents with significant facial nerve paresis and bony erosion disproportionate to its size.

Histologically, paragangliomas present as nests of chief cells—polygonal cells with round nuclei and granular, eosinophilic cytoplasm—surrounded by spindle-shaped sustentacular cells in a Zellballen pattern.³ Higher-grade tumors have been shown to progressively lose the relationship between the 2 cell types.¹⁰ Immunohistochemical staining of chief cells demonstrates positive staining for neuroendocrine markers, such as neuron-specific enolase, synaptophysin and/or chromogranin, and negative staining for keratins. This lack of keratin immunostaining in paragangliomas distinguishes them from middle ear adenomas. Sustentacular cells typically stain positive for S-100.

Middle ear paragangliomas are often locally destructive but rarely malignant. Although features such as an increase in mitotic activity, central necrosis and vascular invasion may increase suspicion of malignancy, only metastasis to a lymph node or distant organ confirms malignancy.² Surgical resection is indicated owing to their progressive growth pattern and involvement of major vessels and cranial nerves, although radiotherapy presents a growing option.⁶ While preoperative embolization may be helpful in reducing bleeding, our practice has also found the use of carbon dioxide laser extremely helpful in achieving hemostasis.

ARTICLE INFORMATION

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