Ear and temporal bone meningioma

Lester D.R. Thompson, MD

Meningiomas account for 30% of all intracranial neoplasms, but primary extracranial (ectopic) meningiomas of the ear and temporal bone are less common, accounting for about 10% of all ear and temporal bone tumors. Meningiomas are derived from the arachnoid cap cells, also called *pacchionian bodies.*

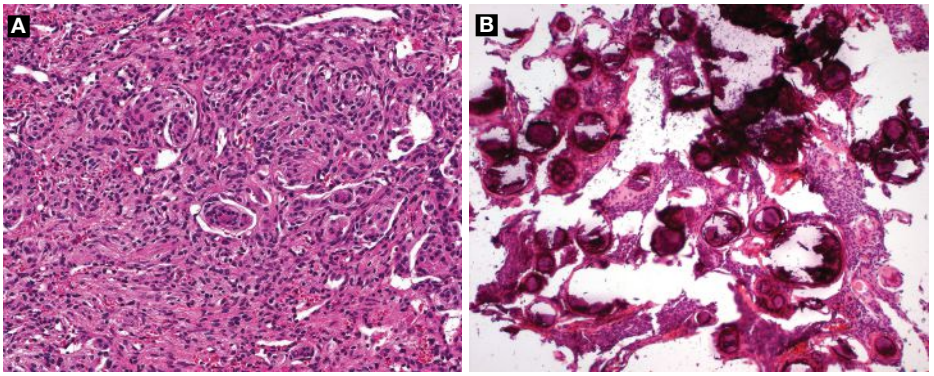

Figure. ***A:*** *Image shows a vague whorling to the proliferation that features a spindled to epithelioid appearance. A fibrous stroma is focally noted.* ***B:*** *Innumerable psammoma bodies are seen in this image, along with concentrically layered calcified material. The meningothelial growth is noted between the dark blue calcium.*

More often women than men (2:1), patients typically present with hearing loss, tinnitus, otitis media, pain, headaches, dizziness, and/or vertigo, often of many years' duration. The overall mean age at presentation is 50 years (range: 10 to 90); women are often older than men. Tumors tend to involve the middle ear more often than the internal auditory meatus, but direct extension from the central nervous system must be radiographically or clinically excluded before a diagnosis of ectopic or primary ear and temporal bone meningioma is rendered. Computed tomography or magnetic resonance imaging helps to document an intracranial component and tumor extent, especially when detecting en plaque tumors.

Patients usually experience a good long-term outcome, with a 5-year survival of approximately 85%. The incidence of recurrence or persistence after surgery ranges as high as 20%.

Because of anatomic restrictions, most tumors are smaller than 1.5 cm, removed in multiple, gritty tissue fragments. The tumor cells infiltrate the interstices of bone and are arranged in a meningothelial to whorled architecture. The tumor cells may blend with surface squamous epithelium. The lobules and nests of the tumor exhibit a syncytial, epithelioid appearance (figure, A). The cells are usually bland with round nuclei, often with intranuclear cytoplasmic inclusions. Psammoma bodies may be seen (figure, B). Within the ear and temporal bone, meningothelial, psammomatous, and fibroblastic types are the most common.

Occasionally, immunohistochemistry may be needed to confirm the nature of the cells by identifying positive reactions for EMA, CAM5.2, and CK7 (the latter in a pre–psammoma-body pattern). The tumor cells are negative with GFAP, SMA, synaptophysin, and chromogranin. The histologic differential diagnosis includes schwannoma, paraganglioma, middle ear adenoma, and meningocele.

Suggested reading

Rushing EJ, Bouffard JP, McCall S, et al. Primary extracranial meningiomas: An analysis of 146 cases. Head Neck Pathol 2009;3(2):116-30.

Thompson LD, Bouffard JP, Sandberg GD, Mena H. Primary ear and temporal bone meningiomas: A clinicopathologic study of 36 cases with a review of the literature. Mod Pathol 2003;16(3):236-45.

From the Department of Pathology, Southern California Permanente Medical Group, Woodland Hills Medical Center, Woodland Hills, Calif.