Neuroendocrine adenoma of the middle ear

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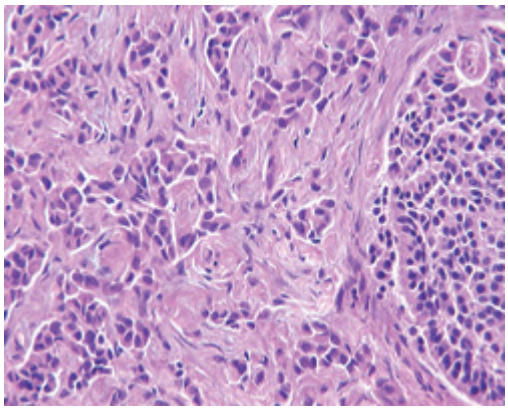

Figure 1. A variety of patterns of growth are seen in this neuroendocrine adenoma of the middle ear. Note the "infiltrative" growth pattern of the plasmacytoid cells. An organoid growth is seen on the right.

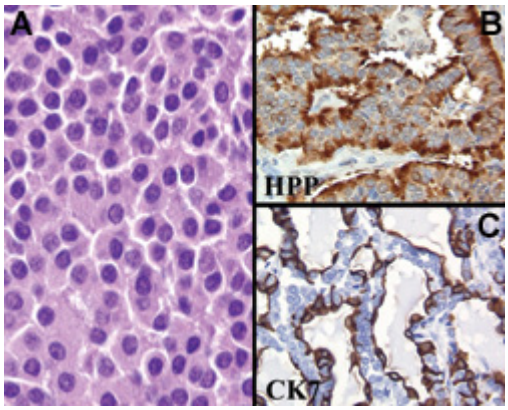

*Figure 2. Cells are often quite plasmacytoid with a delicate "salt-and-pepper" nuclear chromatin distribution (**A**). Neoplastic cells are positive with chromogranin, human pancreatic polypeptide (**B**), and CK7 (**C**).*

Neuroendocrine adenomas of the middle ear, also known as *middle ear adenomas* and *carcinoids*, are rare neoplasms. These tumors occur equally in the genders, and they usually affect middle-aged patients. Patients present clinically with hearing loss and pain; tinnitus, equilibrium changes, and nerve paralysis might also be identified. Tumors are usually smaller than 1 cm in their greatest dimension, and they occasionally extend into the external or internal auditory canal.

Microscopically, neuroendocrine adenomas of the middle ear are unencapsulated and "pseudoinvasive" with moderate cellularity (figure 1). They display several growth characteristics: glandular spaces, trabeculae, festoons, ribbonlike patterns, anastomosing cords, and solid sheets with variable cohesiveness. The glandular spaces frequently contain an amorphous secretory product. The predominant architectural pattern tends to vary among tumors and even within a single tumor. The cells are cuboidal-to-columnar and uniform in size, and they feature eosinophilic, finely granular, and homogenous cytoplasm. Cytoplasmic borders are indistinct. The nuclei are round-to-oval with finely dispersed "salt-and-pepper" chromatin, and they may be centrally or eccentrically placed with only minimal pleomorphism. A plasmacytoid appearance may be noted (figure 2). Nucleoli are inconspicuous, and mitoses are essentially absent. The gland-duct spaces are lined with a dual-cell population made up of an inner (luminal), flattened, and slightly more intensely eosinophilic cell surrounded by a basally positioned cuboidal-to-columnar cell. The tumor cells are immunoreactive with cytokeratin, CK7, and CAM 5.2; CK7 specifically highlights the inner (luminal) layer of the glandular cells. Neuroendocrine marker immunoreactivity includes chromogranin, neuron-specific enolase, human pancreatic polypeptide, synaptophysin, and serotonin.

All tumors are managed by surgery (simple or modified mastoidectomy); radical mastoidectomy is reserved for tumors that encase the ossicles. Recurrences or regrowths occur in as many as 20% of cases. Removal of the ossicular chain is usually associated with a reduction in recurrence.

Suggested reading

Mills SE, Fechner RE. Middle ear adenoma. A cytologically uniform neoplasm displaying a variety of architectural patterns. Am J Surg Pathol 1984;8:677-85.
Torske KR, Thompson LD. Adenoma versus carcinoid tumor of the middle ear: A study of 48 cases and a review of the literature. Mod Pathol 2002;15:543-55.