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Intracochlear Glandular Schwannoma

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Benign glandular schwannomas are rare, with 8 cases having been reported in the literature.¹ The tumors have been found on the extremities, trunk, and scalp, but none have been reported as being associated with the ear, a common location for sporadic schwannomas.² Intralabyrinthine and cochlear schwannomas, such as illustrated in this case, were rarely described until the advent of magnetic resonance imaging, when reports of the tumor became quite common.³ However, no cases of benign glandular schwannomas in the ear have been described previously.

Case Report

The patient was first examined at age 43 years because of a progressive hearing loss in the right ear that began 13 years prior to his otologic examination. Audiometric tests indicated a loss of 81 dB and a discrimination score of 60%. He had had a total loss of hearing in the left ear dating from an episode of scarlet fever and subsequent mastoidectomy at 4 years of age. The loss in the right ear continued to progress and was total 7 years after the initial examination.

Electronystagmography showed no response in either ear. Computerized axial tomography demonstrated a distortion of the inner ear on the left side but normal findings on the right.

A single-electrode cochlear implant electrode was introduced through the round window of the right ear, after the removal of some ossification in the first millimeter of the basal cochlear segment, to a depth of 6 mm. The patient’s perceptual performance was average for single-electrode implant users. He died of chronic obstructive pulmonary disease 3 years after implantation.

The right implanted temporal bone was removed for microscopic evaluation. The bone was fixed in 10% buffered formalin for 2 weeks, decalcified in ethylenediaminetetraacetic acid (EDTA), embedded in celloidin, and cut into 20-micron sections using a sledge microtome. Every tenth section was stained with hematoxylin and eosin and mounted on coverslipped slides.

Microscopic examination revealed that the vestibule was filled with tissue with elongated nuclei arranged in whorls and palisading characteristic of Antoni type A schwannomas. The tumor extended into the proximal segments of the semicircular canals and had subluxated the stapedial footplate to protrude slightly into the oval window niche. It also invaded the cochlea and violated the basilar membrane, in the hook area, so that it occupied both the scala tympani and vestibule for about 8 mm into the basal cochlear segment.

Scattered throughout the tumor in the vestibule and cochlea were islands of tissue with a typical glandular morphology and consisting of cuboidal cells surrounding a lumen filled with eosinophilic staining material or arranged in sheets (Figure 1). These areas labeled positive with anti-cytokeratin monoclonal antibody (CAM 5.2; Becton Dickinson, San Jose, California) followed by goat antimouse secondary antibody conjugated to fluorochrome Dylight 649 (Jackson Immunoresearch, West Grove, Pennsylvania), which was indicative of glandular tissue (Figure 2). It was negative with S-100 monoclonal antibody (Santa Cruz Biotechnology, Santa Cruz, California) and the same secondary antibody specific for Schwann cells.

The electrode path traversed the tumor in the scala tympani and was not contiguous with any of the glandular areas. There was distension of Reissner membrane halfway into the scala vestibule in all cochlear segments and filamentous fibrosis in the perilymph in the inferior cochlear segment. This distension was most likely caused by blockage of the endolymphatic duct by the tumor and consequent cochlear hydrops. The spiral ligament was somewhat atrophic; however, the stria vascularis appeared normal. About 75% of the outer hair cells and dendrites were missing.

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but the ganglion cell population was near normal (22,527 vs normal of 25,270 ± 6385) for the patient’s age.4

Discussion
This is the first report of a benign glandular schwannoma occurring in the temporal bone, specifically in the inner ear. The commonly proposed hypothesis of the origin of these tumors is that of Yoshida and Toot,5 who propose that they arise from pluripotential neural crest cells that can differentiate into various phenotypes.

This report was approved by the St Vincent’s Medical Center IRB.

Author Contributions
Jose N. Fayad, supervised; Adam Markaryan, immunohistochemistry; Fred H. Linthicum Jr, performed the analysis; Richard T. Miyamoto, performed surgery, obtained temporal bones, maintained clinical records.

Disclosures
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