Histopathology of human spiral ganglia in Sjogren’s syndrome

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LEARNING OBJECTIVES
1. Understand histopathologic features of Sjogren’s syndrome in the human inner ear.
2. Correlate human histopathology with available animal models.
3. Gain a better understanding of the histopathologic basis of inner ear disease.

INTRODUCTION
Sjogren’s Syndrome (SS) is the second most common autoimmune rheumatic disease affecting approximately 500,000 to 2 million patients in the US. It is characterized by lymphocytic infiltration of the lacrimal and salivary glands (1). Some patients develop hearing loss, with a reported prevalence of 20% to 90% (2). However, the incidence of hearing loss is thought to be the first otologic manifestation of SS. In a study of 40 female SS patients, 45% of patients demonstrated conductive sensorineural hearing loss (SNHL) mainly in the high frequencies and was associated with disease duration (2). Subclinical SNHL is likely more common in patients with SS. SS is one of several autoimmune disorders in which hearing loss has been described. This group of “autoimmune inner ear disorders” (AIED) was first described by McCabe in 1979 (4). AIED are believed to be associated with immunoreactivity to inner ear components (5,6). The pathogenesis of immune-mediated SNHL is unclear but may include immunocomplex-mediated vasculitis in the inner ear or autoantibodies directed against inner-ear antigenic epitopes (3).

METHODS AND MATERIALS
The histopathology of the inner ear in two patients with SS and correlate these findings to known mouse models of autoimmunity.

RESULTS
There was severe loss of the intermediate cells of the stria vascularis bilaterally in specimen 1 (Fig. 1). Figure 2 shows a higher magnification view of the stria vascularis with signs of hydrops in the apical, middle and basal turns along with collapse of Reissner’s membrane as compared to a normal cochlea in Figure 2B. The organization of the organ of Corti along with the inner and outer hair cells were preserved in both SS specimens (Fig. 3). The BM was thickened under the stria marginal cells in both specimens (Fig. 4). Figure 5 shows Immunohistochemistry showing IgG antibody deposition within capillaries of stria vascularis. There was IgG antibody deposition in the capillaries within the stria vascularis in both patients (Fig. 7). Figure 7B shows the stria vascularis incubated with the same antibodies in a normal specimen.

REFERENCES