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Ramsay Hunt Syndrome: A Histopathology Observation of A Facial Sequelae:

Temporal Bone Histopathology Case of the Month

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Abstract

In 1907, James Ramsay Hunt (1) described a syndrome characterized by facial nerve paralysis associated with herpetic eruption of the pinna and suggested that herpes zoster oticus resulted from a geniculate ganglionitis. However, many contemporary authors agree that this condition represents a polycranial neuronitis and not simply a geniculate ganglionitis. Thus, it is a more complex disease, and the precise pathogenesis in the temporal bone that leads to facial palsy is not yet known.

Case Report

The patient was a 76-year-old woman with a 2-month history of left facial paralysis when she was first seen in the office. Before the onset of her facial paralysis, she experienced a typical vesicular eruption of her left concha, severe pain, vertigo, and sudden hearing loss. She was treated with prednisone without improvement and then underwent surgical transmastoid facial nerve decompression (2). During surgery, moderate swelling of the facial nerve was found from the beginning of the vertical segment of the facial nerve to the stylomastoid foramen. After surgery, she had partial improvement of her facial paralysis and died 7 years later from other causes.

Histopathologic Findings

The facial nerve at its horizontal and labyrinthine portions seems abnormal with pronounced segmental atrophy (Figs. 1 and 2). The superior vestibular nerve shows partial atrophy. The crista of the superior semicircular canal has undergone severe degeneration, and there are no fibers in its substance. It was not possible to determine the histopathologic cause of the sensorineural component of the hearing loss.

References

1. Ramsay Hunt J. On herpetic inflammations of the geniculate ganglion. A new syndrome and its complications. *J Nerve Ment Dis* 1907;34:73–96.
2. Fisch U. Total intratemporal exposure of the facial nerve. *Arch Otolaryngol Head Neck Surg* 1972;95:335–41.

Fig. 1. Horizontal section just below the geniculate ganglion showing the internal auditory canal and the horizontal segment of the facial nerve. a, Posterior half of the facial nerve is atrophied. b, Superior vestibular nerve is atrophied (hematoxylin-eosin, $\times 40$).

Fig. 2.
High magnification of the facial nerve showing degeneration of the posterior half of the nerve (hematoxylin-eosin, $\times 200$).