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INTRACOCHLEAR SCHWANNOMA AND COCHLEAR IMPLANTATION

JONA KRONENBERG, MD
ZEEV HOROWITZ, MD MINKA HILDESHEIMER, PHD
TEL HASHOMER, ISRAEL

A case of intracochlear schwannoma in a 58-year-old candidate for cochlear implantation is described. The tumor was located in the basal turn of the cochlea and was discovered only during surgery. Computed tomography and magnetic resonance imaging obtained prior to surgery failed to detect the tumor. Intralabyrinthine schwannomas are rare tumors that grow either in the vestibule, as intravestibular schwannomas, or in the cochlea, as intracochlear schwannomas. Complete removal of this tumor was achieved through a posterior tympanotomy approach. Cochlear implantation, which resulted in good hearing, was successfully performed 3 years later.

KEY WORDS — cochlear implant, intracochlear schwannoma.

INTRODUCTION

Acoustic neuromas are benign tumors derived from the perineural Schwann cells of the eighth cranial nerve. These tumors usually arise from the vestibular nerve within the internal auditory canal and expand into the cerebellopontine angle. Intralabyrinthine schwannomas are uncommon, and only 23 cases have been reported in the world literature.[1-7] These were diagnosed accidentally, either during labyrinthectomy and vestibular neurectomy or at autopsy. These intralabyrinthine tumors were described as either intravestibular[1] or intracochlear schwannomas.[2]

This report adds a unique case of an intracochlear schwannoma found during cochlear implantation surgery. To the best of our knowledge, this is the first case of surgical removal of an intracochlear schwannoma prior to cochlear implantation.

CASE REPORT

A 58-year-old patient with complete bilateral deafness was referred to our department for cochlear implantation. On the left side, he had had an acoustic neuroma removed 24 years prior to admission. On the right side, his hearing had gradually deteriorated for unknown reasons, resulting in complete hearing loss. In May 1993, he was admitted for cochlear implantation on the right side. Computed tomography performed prior to surgery revealed no abnormalities on the right side, and on the left side, bony changes consistent with the surgical removal of the acoustic neuroma were found. The patient had neither vestibular complaints nor tinnitus. A posterior tympanotomy was performed, and the scala tympani of the basal turn was opened through a cochleotomy. A white, firm mass was discovered in the basal turn of the cochlea, leaving the apex of the cochlea free of tumor. No bony erosion was noted anywhere in the labyrinth. The tumor was completely resected. As the histologic frozen section proved to be a schwannoma, the procedure was terminated. Magnetic resonance studies obtained after surgery showed no residual tumor. During 3 years of follow-up, there was no recurrence.

In July 1996, a cochlear implantation was undertaken with a 22-channel Nucleus Spectra device. The facial recess was found to be obliterated, and the opening in the cochlea was plugged by connective tissue. This plug was easily removed, and the entire length of the active electrode array was inserted. Hearing rehabilitation commenced 4 weeks after implantation. Only the 10 apical electrodes could be used for electrical stimulation. The remaining electrodes caused an unpleasant sensation in the lower part of the face during activation, probably caused by spread of the electrical current through the connective tissue. Nevertheless, the results of the patient's auditory speech perception were fairly good. The score on syllables of Hebrew words presented in closed set was 100%. The score was 30% on open-set presentation of topic-related, everyday sentences developed at the City University of New York for this and related studies adapted to Hebrew. The patient managed to use the telephone when talking with people known to him.

DISCUSSION

Acoustic neuromas (vestibular schwannomas) usually arise from the vestibular nerve in the internal

From the Departments of Otolaryngology–Head and Neck Surgery (Kronenberg, Horowitz) and Communication Disorders (Hildesheimer), Sheba Medical Center, Tel Hashomer, and Sackler Faculty of Medicine, Tel-Aviv University, Tel-Aviv, Israel.
CORRESPONDENCE — Jona Kronenberg, MD, Dept of Otolaryngology–Head and Neck Surgery, Sheba Medical Center, Tel Hashomer, Israel.

auditory canal, but they can be found anywhere along the path of the eighth nerve. They rarely present within the labyrinth, arising from the terminal fibers of the vestibular or cochlear sensory end organs. The extension of intralabyrinthine schwannoma in the inner ear varies. Miyamoto et al[1] described an isolated tumor in the vestibule, while others described the tumor in the cochlea only.[2] When the tumor originated in the cochlea, the scala tympani of the basal turn was the common site of growth.[3] More extensive tumors have been found, in both the vestibule and cochlea.[4] In most cases there was no extension of the tumor into the internal auditory canal.[5] In our case, the schwannoma developed in the basal turn only, leaving the apical part of the cochlea free of any tumor.

Intralabyrinthine schwannomas are difficult to diagnose. In most cases, the patient history, otoneurological tests, and imaging including computed tomography and magnetic resonance imaging fail to demonstrate the existence of a tumor. It is usually diagnosed accidentally during labyrinthectomy for Meniere's disease,[5] vestibular neurectomy,[6] or autopsy.[2] Only 1 case has been discovered radiographically.[4]

In early reports, intralabyrinthine schwannoma was usually combined with neurofibromatosis.[7] More recent reports have described intralabyrinthine schwannoma in patients without neurofibromatosis.[8] Most of the recent cases have been diagnosed as Meniere's disease.[6] Among the 23 cases reported, 7 patients suffered from complete deafness, but patients with nearly normal hearing were also described.[6] In our case, the patient had complete hearing loss that had become progressively worse during the 10 years prior to surgery.

Although intralabyrinthine schwannoma is a rare occurrence and cannot usually be diagnosed without surgery, it is important to question its existence, especially in deaf patients who are candidates for cochlear implantation surgery and in whom no explanation for deafness was found. As cochlear implantation has become popular nowadays, we can assume that more cases of intralabyrinthine schwannoma will be found.

Complete removal of small tumors via the original cochlear implantation approach seems to be sufficient for small intralabyrinthine schwannomas. If this proves to be effective, implantation can proceed.

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