Intratemporal Facial Nerve Neurinoma
—Case Report—

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Abstract

A 59-year-old female had episodes of vertigo for 13 years, right tinnitus for 6 years, and right hearing difficulty for 2 years. She had no facial nerve dysfunction or other neurological deficits. Postcontrast computed tomography (CT) did not show abnormalities, but a wide, high-window CT scan revealed erosion of the petrous pyramid on the right side. Magnetic resonance imaging clearly delineated the entirety of a small tumor transecting the petrous bone. At operation a neurinoma was found to originate from the facial nerve proximal to the geniculate ganglion; it was totally removed. This case is unique in that she had a long history of signs and symptoms of acoustic nerve disturbance, but no facial nerve dysfunction whatsoever.

Key words: facial nerve, geniculate ganglion, neurinoma, magnetic resonance imaging, petrous bone

Introduction

Neurinomas of the facial nerve are reportedly rare, especially within the temporal bone. However, since the first report by Schmidt in 1930, at least 295 cases of intracranial and intratemporal facial nerve neurinomas have been reported, mostly by otolaryngologists. Facial nerve paresis is the most frequent presenting symptom. It is, however, absent in 27% of cases, and in a few cases tinnitus and hearing difficulty were the only presenting symptoms, which lead to an erroneous diagnosis of acoustic neurinoma. The case reported here is one such rare case in which a facial nerve neurinoma presented with no facial nerve dysfunction at all.

Case Report

A 59-year-old female was referred to our department with a 13-year history of vertiginous attacks. Initially, the attacks occurred only once every year or two. After 7 years, she developed tinnitus in the right ear, and the episodes of vertigo increased in frequency and were accompanied by vomiting. The tinnitus progressively worsened and hearing difficulty developed in the right ear 2 years prior to admission. She had been treated for Ménière's disease, without success.

On admission to the Department of Otolaryngology of this institution, she exhibited right sensorineural hearing loss of 75 dB. Caloric and auditory evoked responses were absent on the right side. Tomography of the petrous pyramid indicated slight enlargement of the internal auditory canal on the right side, the diameter being 7 mm on the right and 5 mm on the left. Pre- and postcontrast computed tomographic (CT) scans were interpreted as normal. However, a CT scan set at a wide, high-window level revealed widening of the internal auditory canal and erosion of the petrous bone on the right side (Fig. 1). T1-weighted magnetic resonance (MR) imaging disclosed a small mass, slightly but heterogeneously isointense compared with the brain tissue, filling the eroded portion of the petrous bone, and the dilated internal auditory canal. The mass extended from the internal auditory porus to the middle fossa, transecting the petrous bone (Fig. 2).

When she was referred to us with a diagnosis of right acoustic neurinoma, she had tinnitus and hearing loss on the right side. There were no facial motor or sensory disturbances or other neurological deficits. She had no cutaneous stigmata suggestive of neurofibromatosis. We re-evaluated the CT scans
and MR images and arrived at a diagnosis of facial nerve neurinoma rather than acoustic neurinoma, since the petrous bone was eroded along the facial canal toward the geniculate ganglion.

At surgery, the petrous portion of the middle fossa was reached extradurally through a right subtemporal craniotomy. The small tumor at the midpoint of the petrous pyramid (Fig. 3) was totally removed by means of microsurgery. The tumor originated from the facial nerve and involved the geniculate ganglion from its proximal portion to the internal auditory canal. The exact site of origin could not be verified. The facial nerve could not be identified as such, except at the internal auditory porus. The greater superficial petrosal nerve was not involved. The acoustic nerve was compressed against the lower wall of the internal auditory canal. The cochlea and the semicircular canals were apparently not affected. The bony defect was sealed with a piece of the temporalis muscle.

Postoperatively, she had right peripheral facial nerve paralysis, with a May score of 30, and a 50% decrease in taste sensation in the anterior two thirds of the tongue on the right side. The right-side tinnitus and hearing loss remained unchanged. The histological diagnosis was neurinoma, mainly of Antoni type A. The specimen included some ganglia which were probably geniculate (Fig. 4).
Discussion

Neurinomas may originate from any portion of the facial nerve and involve multiple segments.\(^{10,13}\) The tympanic segment of the facial nerve is most frequently involved (58%), followed by the vertical (48%) and the labyrinthine/geniculate segments (42%). Meatal involvement occurs in 30% of cases.\(^{10}\)

Although it is said that the clinical manifestations are related to the site of the tumor and the direction of its growth,\(^{5,13}\) very little correlation has actually been found between the symptoms and the portions of the nerve affected.\(^{10}\)

Usually, the earliest clinical manifestations are facial nerve paresis, tinnitus, and/or hearing difficulty. The most common presenting symptom is facial nerve paresis,\(^{10}\) which is often accompanied or preceded by hemifacial spasm or fasciculation.\(^{1,10,13}\)

Remissions and recurrences of the initial facial nerve paresis have been observed over periods as long as 35 years,\(^{12,17}\) with a resultant misdiagnosis of Bell's palsy.\(^{13}\) The facial nerve paresis may improve when the neurinoma extends through the eroded bony wall, thereby reducing the pressure within the canal.\(^{17}\)

Although quite a few patients report auditory symptoms of a few to 15 years' duration prior to the onset of facial weakness,\(^{2,4,5,9,13}\) some degree of facial nerve paresis eventually develops in 73% of patients with facial nerve neurinoma.\(^{10}\) The remaining 27% have no facial paresis whatsoever,\(^{10}\) and this may lead to an erroneous diagnosis of acoustic neurinoma.\(^{13}\)

Facial nerve neurinomas do not always cause facial nerve paralysis if the site of origin allows for tumor expansion; such sites include the internal auditory canal, middle ear, and parotid gland.\(^{1,11}\) In our case, enlargement of the internal auditory canal and erosion of the petrous bone along the facial nerve proximal to the geniculate ganglion might have prevented compression of the facial nerve and the consequent impairment of its function. In nearly all of the reported cases in which facial nerve dysfunction was absent, there was bony erosion in the internal auditory canal and/or the geniculate ganglion region.\(^{8,11}\)

A few asymptomatic facial nerve neurinomas have been found incidentally at autopsy or operation,\(^{5,15}\) but these tumors were very small, having diameters of only 2–3 mm.

When auditory symptoms precede peripheral facial nerve paresis, the site of origin is thought to be in the middle ear at the height of the horizontal portion of the facial canal, and hearing loss is of the conductive type.\(^{12}\) Our patient's hearing loss was sensorineural in nature and there was no bony erosion in the peripheral portion beyond the geniculate ganglion. Tumors that originate in the internal auditory canal produce sensorineural hearing loss secondary to acoustic nerve compression, and this usually follows the onset of the facial nerve paresis.\(^{12}\) Our patient's tumor appeared to have originated in the meatal portion. It caused compression of the cochlear and vestibular nerves, leading to vestibular dysfunction of many years' duration and to sensorineural hearing loss of a few years' duration. As mentioned, the absence of facial nerve dysfunction is probably attributable to the bony erosion, which allowed adequate space for the facial nerve.

In this case, postcontrast CT did not demonstrate the tumor because of its small size. However, a CT scan with a wide, high-window setting disclosed the bony erosion. MR imaging was extremely helpful, delineating the entire tumor within the temporal bone.

References

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