CASE REPORT

Facial Nerve Neuroma: Clinical, Diagnostic, and Surgical Features

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ABSTRACT

Facial nerve neuromas are uncommon benign tumors. The diversity of tumor locations and their proximity to vital neurovascular structures create a variety of clinical situations, and the management strategy for each case differs. Two cases with tumors located at different sites and associated with different clinical presentations are discussed in terms of diagnosis and management.

KEYWORDS: Neuroma, facial nerve, chorda tympani, paralysis

Facial nerve neuromas are uncommon benign neoplasms of Schwann cells. They compose only 0.8% of all intrapetrous mass lesions.¹ More than 300 cases have been reported since they were described by Schmidt in 1930.²

These tumors can arise from any segment of the nerve from the cerebellopontine angle to the extratemporal peripheral portion.^{3,4} The diversity of locations involved with the tumor and their proximity to the auditory apparatus can create a variety of clinical situations.⁴ Thus, the site of the tumor, its extension, and the way it involves the surrounding structures determine the surgical approach.

This article presents two cases of facial nerve neuroma involving different locations and clinical manifestations, focusing on the diagnostic and therapeutic problems encountered.

CASE REPORTS

Case 1

A 38-year-old woman sought treatment for left facial paralysis, dizziness, and hearing loss and tinnitus in her left ear that had been present for about 1 year. Otoscopy revealed an intact tympanic membrane. A reddish mass was seen behind the tympanic mass. Neurological examination revealed total facial paralysis (i.e., House-Brackmann grade VI). Pure-tone audiometry indicated unilateral sensorineural hearing loss in the left ear with air- and bone-conduction thresholds elevated to 60 dB hearing loss. The tympanogram was type-B on the left side. The acoustic reflex was absent to acoustic stimuli on the left side. Computerized tomography

Figure 1 Preoperative axial CT of Case 1 showing the tumor in the tympanic cavity.

(CT) showed a soft-tissue mass within the left temporal bone corresponding to the location of the geniculate ganglion and the tympanic portion of the facial nerve. The mastoid cavity had also been invaded by the soft-tissue mass (Fig. 1). T2-weighted magnetic resonance imaging (MRI) revealed a mass lesion with high-signal intensity in the same location. The mass had extended to the posterior fossa by eroding the medial wall of the mastoid cavity. The patient underwent surgery performed by a team composed of both neurosurgeons and otorhinolaryngologists. Total tumor removal was achieved through a presigmoid retrosigmoid and transmastoid approach with anastomosis of the facial nerve (extratemporal portion distal to cranial nerve XI). The bony defect was obliterated with the temporal muscle flap. The external auditory canal and the tuba auditiva were stumped. Postoperative histopathological examination revealed the diagnosis of neuroma. The patient's facial palsy was grade II (Fig. 2), and MR images were free of disease in the fourth postoperative year (Fig. 3).

Case 2

A 49-year-old woman sought treatment for the sensation of a foreign body in her right external

Figure 2 Photograph of Case 1 in the fourth year after surgery.

auditory canal. She had neither tinnitus nor vertigo-dizziness. Facial functions were normal bilaterally. Otoscopy showed a skin-covered mass almost completely occluding the external auditory canal.

Figure 3 Control MRI images of Case 1 in the fourth postoperative year.

The mass originated from the posterior wall of the bony canal. The tympanic membrane could not be seen because of the mass. Pure-tone audiometry showed a conductive hearing loss with an air-bone gap of 40 dB. CT of the left temporal bone showed a soft-tissue mass in the mastoid portion of the facial nerve. The mass was eroding the posterior bony wall of the external auditory canal and occupying the lumen of the bony canal. The tympanic cavity was free of invasion (Fig. 4). MRI showed a mildly hyperintense mass lesion corresponding to the same location in T2-weighted MRIs. Electrophysiological tests evaluating the motor functions of the facial nerve revealed no abnormalities. Preoperatively performed transcanal punch biopsy from the mass was consistent with the findings of neuroma.

The otorhinolaryngology team performed a transmastoid approach. The tumor was identified on the mastoid portion of the facial nerve. It was eroding the posterior wall of the canal and invading the external auditory canal (Fig. 5). The trunk of the facial nerve was just distal to the second genu of the nerve and was intact along the posterior aspect of the tumor. The tumor arose from the initial portion of the chorda tympani at the junction with the main trunk of facial nerve. The tumor

Figure 4 Preoperative axial CT of Case 2 revealing tumor invasion from the mastoid cavity to the external auditory canal.

Figure 5 Intraoperative view of the tumor eroding the posterior canal wall and extending into the external auditory canal. *, body of incus.

was dissected while preserving the facial nerve. On exploration, the middle ear cavity and the ossicles were normal. Since the tympanic membrane was intact, a Bondy-type⁵ tympanomastoidectomy with partial mastoid cavity obliteration was performed. The postoperative period was uneventful. The patient's House-Brackmann grade III facial palsy regressed to grade I in 4 weeks (Fig. 6). Histopathological examination confirmed the initial diagnosis of neuroma.

DISCUSSION

The presentation of facial nerve neuroma tends to be insidious and depends on the location and extent of the lesion. Slowly progressive or sudden facial weakness, often preceded by facial twitching, is a common presenting complaint.⁶ In about 5% of patients with Bell's palsy, a facial nerve neuroma is found to be the cause.⁷ However, normal facial nerve function has been reported in 27% of patients.^{8,9} Because the tumor is near the auditory apparatus, hearing can be impaired. The loss may be sensorineural or conductive, depending on its origin, either proximal or distal to the geniculate ganglion.⁴ Although the tumor in Case 1 was

Figure 6 Photograph of Case 2 in the first month after surgery.

located distal to the geniculate ganglion, the hearing loss was also sensorineural, which indicates the invasion of the inner ear by this advanced lesion.

Another rare presentation, which is typical for a subgroup of facial nerve neuromas, is a mass obstructing the external auditory canal, causing conductive hearing loss. This finding is associated with a rare entity, a chorda tympani neuroma.¹⁰ The chorda tympani alone was involved in nine reported cases.^{10–18} Our Case 2 adds the tenth such case to the English literature. These tumors can grow large without causing facial paralysis.¹¹ With their typical and defined characteristics, the diagnosis is less complex compared with that of facial nerve neuromas. However, this subgroup of neuromas can be difficult to distinguish from tumors in the external auditory canal. Clinical suspicion requires confirmation with preoperative histopathological examination. Thus, preoperative fine-needle aspiration cytology can be extremely useful.¹⁰ Alternatively, as in our Case 2, a micro-otoscopic transcanal incisional biopsy can provide histopathological data. CT and MRI are required to plan surgery.¹⁹

As for the diagnosis of facial nerve neuroma, the presence and extension of the tumor can be seen most accurately by radiologic examination. Bone-targeted high-resolution CT of the temporal bone is believed to be superior to MRI. Enlargement of the facial nerve suggests involvement of a neoplastic process.⁴ MRI is complementary, providing information about the nerve itself. Abnormal enhancement indicates the presence of the tumor. High-resolution CT should be performed first to visualize the facial canal followed by MRI to demonstrate the actual tumor. The clue to diagnosis is an intact facial canal on CT and high-signal intensity on T2-weighted MRIs.²⁰ Histologically, however, facial nerve neuromas are identical to acoustic neuromas and have the same enhancement characteristics. Therefore, it is seldom possible to identify the nerve of origin of a neuroma in the internal acoustic canal.³ Anterosuperior erosion of the internal acoustic canal or erosion of the labyrinthine facial nerve canal, if present, may be the only diagnostic clue.²¹

Topographic tests of facial nerve function are of little value because involvement of the facial nerve may not immediately impair the electrical activity of the nerve. Electromyography is helpful for quantifying the nerve's residual motor function and therefore for predicting postoperative prognosis of facial recovery after nerve reconstruction.⁴ Electroneurography can help predict the prognosis of postoperative facial palsy. It is believed that better postoperative results are achieved when the nerve is repaired if compound action potentials decrease no more than 50%.²²

Surgical excision is the definitive treatment for facial nerve neuromas. Because they are encapsulated tumors usually attached to the nerve and appear to push axons aside as they grow, theoretically they can be stripped free of the nerve without sacrificing it.²³ There are isolated reports of dissecting the tumor from the nerve and obtaining normal or adequate functional outcomes.^{2,9,24,25} In our Case 2, the facial trunk was preserved. The facial nerve was easily identified because the tumor originated from the chorda tympani. However, these

tumors usually must be resected with the affected portion of the nerve because they are diagnosed after facial palsy is present, or in advanced stages because of their insidious progression. Thus, the nerve should be repaired immediately by primary repair or by a nerve interposition graft. Regardless of the type of repair, patients can expect no better recovery than an eventual House-Brackmann Grade III palsy.^{4,26}

Timing for surgery is controversial. Surgical resection of facial nerve neuromas is indicated without delay for patients with progressive facial palsy or paralysis, for large cerebellopontine angle tumors compressing the brainstem or producing hydrocephalus, and for tumors invading the inner ear.²² However, patients without facial dysfunction pose the greatest dilemma because facial palsy is usually inevitable after surgery.

Because facial nerve neuromas almost always grow slowly,²² waiting to pursue surgery until the facial nerve either shows progressive deterioration or becomes paralyzed in patients with normal facial nerve function has been advocated.^{27,28} To delay the need for complete resection further, wide tumor decompression to allow the tumor to expand outside the natural bony confines has also been advocated.²²

However, some authors believe that the surgical approach becomes more difficult, the likelihood of postoperative complications increases, and the recovery of facial function is poorer if the tumor continues to grow.^{4,16,29} The risk of surrounding structures such as the inner ear being invaded increases,¹⁶ and eventually the brainstem may become compressed.^{22,27}

The surgical approach should be chosen based on the anatomic location and extension of the tumor. A middle fossa craniotomy is used for patients with intracanalicular neuromas and useful hearing. The translabyrinthine or transpetrosal route is useful for patients without useful hearing and a facial nerve neuroma in the internal acoustic canal.^{4,22} The indications for a retrosigmoid-suboccipital approach for facial nerve neuromas are limited because it does not afford access to the labyrinthine facial nerve. Moreover, anatomic and

functional preservation of the eighth cranial nerve is technically unlikely because the nerve is usually stretched across the posterior surface of the tumor and at risk of injury during resection.²²

Tympanoplasty should be performed in patients with serviceable hearing.⁴ As for chorda tympani neuromas, which typically involve the tympanomastoid cavity, an early tympanomastoid approach is indicated to remove the lesion and to permit reconstruction of the sound mechanisms when the ossicular chain is destroyed before surgery.¹⁶⁻¹⁸

Because each case of facial nerve neuroma manifests with different clinical features, surgical treatment of these tumors is challenging. The surgical strategy should be designed based on the location and the extension of the lesion and on the involvement of the auditory apparatus. Because the surgical procedure usually consists of tumor resection and of facial nerve and auditory reconstruction, a multidisciplinary approach with collaborating neurosurgeons and otorhinolaryngologists is essential to optimize outcomes.

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