

Cerebellopontine angle facial schwannoma relapsing towards middle cranial fossa

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Abstract

Facial nerve schwannomas involving posterior and middle fossas are quite rare. Here, we report an unusual case of cerebellopontine angle facial schwannoma that involved the middle cranial fossa, two years after the first operation. A 53-year-old woman presented with a 3-year history of a progressive left side hearing loss and 6-month history of a left facial spasm and palsy. Magnetic resonance imaging (MRI) revealed 4.5 cm diameter of left cerebellopontine angle and small middle fossa tumor. The tumor was subtotally removed via a suboccipital retrosigmoid approach. The tumor relapsed towards middle cranial fossa within a two-year period. By subtemporal approach with zygomatic arch osteotomy, the tumor was subtotally removed except that in the petrous bone involving the facial nerve. In both surgical procedures, intraoperative monitoring identified the facial nerve, resulting in preserved facial function. The tumor in the present case arose from broad segment of facial nerve encompassing cerebellopontine angle, meatus, geniculate/labyrinthine and possibly great petrosal nerve, in view of variable symptoms. Preservation of anatomic continuity of the facial nerve should be attempted, and the staged operation via retrosigmoid and middle fossa approaches using intraoperative facial monitoring, may result in preservation of the facial nerve.

Introduction

Facial nerve schwannomas are rare and constitute less than 1% of all intrapetrous mass lesions,^{1,2} and estimated at only 1.9% of all intracranial schwannomas.³ Facial schwannomas extending into both the cerebellopontine angle and middle cranial fossa are extremely rare and occurs in only 3% of all cases.⁴⁻⁶ Most of such tumors appear simultaneously in both sites. To our best knowledge, there have been no precise reports of cerebellopontine angle facial schwannoma relapsing towards middle cranial fossa. Here, we report an unusual case of cerebellopontine angle facial schwannoma that involved the middle cranial fossa, two years after the first operation.

Case Report

A 53-year-old woman presented with a 3year history of a progressive left side hearing loss and 6-month history of a left facial spasm and palsy. Neurological examination revealed left-side deafness and facial palsy [House-Brackmann (H-B) grade III] with disturbance in tear secretion on the left, and mild sensory impairment of the left trigeminal area. Magnetic resonance imaging (MRI) revealed 4.5 cm diameter of left cerebellopontine angle partially extending to the middle fossa (Figure 1). The tumor was subtotally removed via a suboccipital retrosigmoid approach under facial nerve monitoring. After drilling the meatus, the facial nerve proven by nerve monitoring was partially exposed. Immediately after surgery, the patient had worsened facial palsy (H-B grade IV), but this was resolved (H-B grade III) about one month after the operation. Postoperative MRI showed subtotal removal of the cerebellopontine angle tumor.

Two years after the operation, the patient had mild worsening of facial palsy (H-B grade IV) and occasional crocodile tears on the left side, and MRI examinations revealed tumor enlargement (Figure 2). Head Computerized tomography (CT) scan demonstrated erosion of the middle portion of the left petrous bone (Figure 3). The patient wanted the facial nerve preserved. By subtemporal approach with zygomatic arch osteotomy, the tumor was subtotally removed except that in the petrous bone involving the facial nerve. Nerve monitoring identified the facial nerve on the petrous bone. Postoperative MRI showed subtotal removal of the middle fossa tumor (Figure 4). The facial nerve improved to H-B grade III, and the crocodile eyes recovered two weeks after the operation.

Discussion

Preservation of facial nerve function

Surgical intervention is agreed treatment of facial schwannomas when required, however, the extent of tumor removal is still controversial because of vulnerable facial nerve function. After total removal of the tumor including petrous portion, direct facial nerve anastomosis or hypoglossal-facial nerve anastomosis are mandatory in most cases, but such intervention are not without risk, occasionally resultCorrespondence: Takafumi Nishizaki, Department of Neurosurgery, Ube Industries Central Hospital, 750, Nishikiwa, Ube, Yamaguchi, 755-0151, Japan. Tel: +81.836.51.9221 - Fax: +81.836.51.9252. E-mail: nishiza@jeans.ocn.ne.jp

Key words: facial schwannoma, cerebellopontine angle, middle cranial fossa, facial nerve preservation.

Received for publication: 5 April 2011. Accepted for publication: 19 April 2011.

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ing in worse facial paralysis.5-8 In a large series of facial nerve schwannoma McMonagle suggested that facial nerve function were better in patients in whom no removal, decompression or subtotal removal was undertaken.9 Huge facial schwannoma extending to both cerebellopontine angle and middle fossa is extremely rare.^{4,6} Especially in those patients, postoperative facial function seems worse. In McMonagle series, we could not find any similar huge tumor, however, they had 4 surgicallytreated tumors arising from cerebellopontine angle and geniculate and labyrinthine segments. Three of them had worse H-B grade postoperatively despite of nerve reconstruction. In the present case, facial function was favorable after the both operations. It may be due to the staged operation via retrosigmoid and middle fossa approaches using intraoperative facial monitoring, resulting in preservation of the facial nerve. Sherman et al. suggested that preservation of anatomic continuity of the facial nerve should be attempted, and it does not seem to lead to frequent recurrence.10

The preservation of the facial function is difficult at posterior fossa, because the facial nerve is completely encased by the tumor in facial schwannoma, unlikely in vestibular schwannoma. Special attention must be paid to preserve the facial function by repeatedly monitoring the facial fascicles. While it is easy to remove the tumor at the middle cranial fossa, when the procedure reaches to the petrous bone where the facial nerve is present. frequent facial monitoring is mandatory. Middle temporal approaches after drilling petrous bone enable easy access to the cerebellopontine angle. However, there has been a significant concern that this involves the risk impairing facial function.

Preoperative diagnosis

Preoperative definite diagnosis is also impor-

tant for surgical strategy to preserve the facial function. Nadeau reported that patient presenting with gradual or persistent facial nerve dysfunction with or without gradual hearing loss should be suspected of facial nerve schwanno-

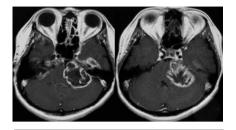


Figure 1. Magnetic resonance imaging revealing a 4.5 cm diameter left cerebellopontine angle tumor extending to middle fossa.

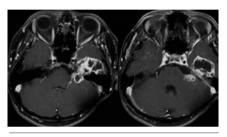


Figure 2. Two years after the first operation, magnetic resonance imaging demonstrating 4 cm diameter left middle fossa tumor.

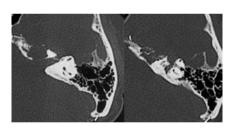


Figure 3. Computed tomography scan demonstrating erosion of the middle portion of the left petrous bone.

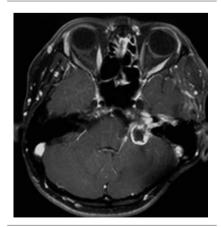


Figure 4. Postoperative magnetic resonance imaging revealing subtotal removal of the tumor except for that in the petrous bone.

ma.¹¹ Cerebellopontine angle tumors are sometimes indistinguishable radiographically from vestibular schwannomas, however, when tumor is extending into the geniculate ganglion of the facial nerve or distal to it, facial nerve schwannoma is more likely in facial schwannoma.¹¹ Bone CT may demonstrate the presence of a mass expanding or remodeling its bony structures.⁹ Involvement of the geniculate ganglion or labyrinthine segment is also clue to distinguish facial from vestibular schwannoma.⁹

Tumor origin

Facial nerve segments that are frequently involved with tumor are labyrinthine/geniculate, tympanic, canalicular, and cerebellopontine angle.9,10 Seventy-four percent of the schwannoma extend along more than one segment of the facial nerve.9 Facial schwannomas extending into both the cerebellopontine angle and middle cranial fossa are extremely rare.5,6 The clinical manifestation of facial schwannoma is variable depending on segments involved.11 Tumors distal to and including geniculate ganglion are involved, the patient suffers from facial nerve dysfunction with conductive hearing loss. Tumor involving cerebellopontine angle do not usually present with facial nerve dysfunction. The hearing disturbance is conductive or sensorineural, depending on the tumor origin is proximal or distal to the geniculate ganglion.8 Facial spasm has been shown in 2.1% of the patients.¹⁰ Facial tic or twitching can be seen in 17% of the patients,9 and tumors arising in the labyrinthine segment of the facial nerve are often associated with facial spasms.¹⁰ Taken together the fact that our patient had facial dysfunction and spasm/tic, and complete hearing loss, this tumor seemed to arise from broad segment of facial nerve encompassing cerebellopontine angle, meatus, geniculate/labyrinthine. Impairment of lacrimal gland and taste is associated with tumor involvement into great petrosal nerve and/or tympanic segment.^{8,12} When this tumor relapsed towards middle fossa, the patient suffered from crocodile eyes. The tumor might involve the great petrosal nerve or tympanic segment partially.

Stereotactic radiosurgery

Stereotactic radiosurgery (SRS) may be one of important alternatives or complementary to surgery in the treatment of facial schwannoma. However, there is little information on the use of SRS in the management of this tumor. Sharman *et al.* reported on two elderly patients treated with SRS whose tumor growth was arrested, and facial and hearing functions were preserved.⁸ However, Shirazi *et al.* reported a case of facial schwannoma undergoing malignant degeneration 10 years after SRS.¹³ The patient required a total temporal bone resection resulting in facial



and vocal paralysis. They suggested that SRS should be attempted in symptomatic patients with poor surgical candidates. Our patient did not irradiate after subtotal resection of the tumor, because there is inadequate data regarding tumor control or deterioration in facial function on such young patient. Further investigation on growth rates and facial nerve function after SRS is required, particularly in young patients with facial schwannoma.

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