Meningiomas Intrinsic to the Geniculate Ganglion

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ABSTRACT

Geniculate ganglion meningiomas are extremely rare lesions—only 14 cases have been reported in the literature. Two new cases of these tumors are described. On computed tomography and magnetic resonance imaging, both lesions appeared centered on the area of the geniculate ganglion, extending to the tympanic cleft and eroding the middle cranial fossa floor. The first case was treated through a middle cranial fossa approach. Because the tumor was so large in the second case, a subtotal petrosectomy was used. The authors review the literature to clarify the clinical and radiological characteristics of these tumors and their surgical treatment.

KEYWORDS: Geniculate ganglion, meningioma, temporal bone, petrosectomy

Geniculate ganglion meningiomas are extremely rare lesions with only 14 cases previously reported in the literature.¹⁻³ The tumors are characterized by a slowly progressive facial deficit, with or without hearing deterioration. Radiological features

are nonspecific, and preoperative differential diagnosis with facial nerve neuromas or hemangiomas is practically impossible. Surgical removal with hearing preservation and facial nerve reconstruction is the treatment option most frequently adopted.

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CASE REPORTS

Two new cases, operated on at the Gruppo Otológico, are presented, and their findings and therapeutic aspects are analyzed.

Case 1

A 56-year-old woman complained of a slowly progressive left facial nerve paralysis for 5 years and left-sided tinnitus for 3 years. At clinical examination the facial paralysis was classified as grade VI according to the House-Brackmann (H-B) scale. Otoscopy was normal bilaterally. The audiological tests revealed a left sensorineural hearing loss between 4000 and 8000 Hz.

High-resolution computed tomography (CT) showed a mass occupying the area of the geniculate ganglion, with erosion of the tegmen tympani and extension to the epitympanum and on the promontorium.

Magnetic resonance imaging (MRI) confirmed the presence of a lesion in the area of the geniculate ganglion. It was hypointense on T1- and hyperintense on T2-weighted sequences; enhance-

ment after gadolinium infusion was heterogeneous (Fig. 1A,B). The tumor protruded slightly in the middle cranial fossa. Based on the clinical and radiological characteristics, the preoperative diagnosis was a facial nerve neuroma.

On March 3, 1994, the patient underwent a middle cranial fossa approach. A reddish bleeding mass was identified in the area of the geniculate ganglion. The lesion did not infiltrate the dura of the middle cranial fossa, which was easily elevated from the superior surface of the petrous bone. The facial nerve was involved by the tumor from the fundus of the internal auditory canal to the tympanic portion. Total tumor removal was achieved, followed by sural nerve grafting.

The postoperative period was uneventful, but the patient experienced a deterioration in sensorineural hearing, with the threshold dropping to a mean of 78 dB for the four main frequencies. On histological examination the lesion was diagnosed as an angiomatous meningioma.

At the patient's last follow-up examination, 3 years after surgery, facial nerve function had recovered to a grade D according to the repaired facial nerve recovery scale.⁴ Control MRI and high-resolution CT scans were negative for a recurrence (Fig. 2).

Figure 1 Case 1. (A) Axial and (B) coronal preoperative MRIs showing a lesion with heterogeneous gadolinium enhancement occupying the area of the geniculate ganglion.

Figure 2 Case 1. Postoperative high-resolution CT showing the area of surgical removal with no sign of recurrence.

Case 2

A 22-year-old boy was affected by a slowly progressive weakness of the left facial nerve of 8 years' duration and left-sided hearing loss. At clinical observation the patient had a grade IV H-B deficit. Otoscopy revealed a reddish retrotympanic mass causing the posterosuperior quadrant of the tympanic membrane to bulge. The audiological examination showed a left-sided conductive hearing loss with a mean air threshold of 50 dB.

High-resolution CT revealed a lesion with indistinct boundaries centered on the area of the geniculate ganglion with erosion of the floor of the

middle cranial fossa and extension in the epitympanum and mesotympanum (Fig. 3A,B). The MRI confirmed the presence of a mass that appeared hypointense on T1-weighted images and hyperintense on T2-weighted images, with heterogeneous gadolinium enhancement (Fig. 4A,B). Based on the radiological findings, a facial nerve hemangioma was the presumptive diagnosis.

On November 11, 1998, the patient underwent a subtotal petrosectomy. To control the superior portion of the lesion, the approach was enlarged through a middle cranial fossa craniotomy. The mass entirely occupied the epitympanum and the tympanic cleft, reaching the Eustachian tube and the temporomandibular joint anteriorly. The incus and stapes were completely eroded. The copiously bleeding lesion was carefully detached from the middle cranial fossa dura, which showed no signs of infiltration. The facial nerve was involved by the lesion from the labyrinthine portion to the second genu. Total tumor removal was followed by placement of a sural nerve graft to reconstruct the facial nerve. The final cavity was obliterated with abdominal fat, and the external auditory canal was closed as a blind sac.

Histological examination showed a "transitional meningioma with prevalent fibrous tissue and a lot of vessels. Psammoma bodies were occasionally included." The patient was discharged with no complications and the bony threshold was

Figure 3 Case 2. (A) Axial and (B) coronal preoperative high-resolution CTs showing a lesion centered on the area of the geniculate ganglion with erosion of the middle cranial fossa floor and extension to the tympanic cleft.

Figure 4 Case 2. (A) Axial and (B) coronal preoperative MRIs after gadolinium infusion showing heterogeneous enhancement of a mass involving the geniculate ganglion.

preserved unchanged. The patient was subsequently lost to follow-up.

DISCUSSION

The most frequent meningioma locations correspond closely to the sites where arachnoid villi are usually found, namely, along the venous sinuses, at the foramina of the cranial nerves, and at the exits of the spinal nerves from the meningeal sleeves.⁵ The temporal bone can be involved with meningiomas originating not only from adjacent structures but also within the petrous pyramid.^{6,7} Four locations have been reported: the internal acoustic meatus, the jugular foramen, the region of the geniculate ganglion, and the sulcus of the greater and lesser superficial petrous nerve.^{5,8-10}

It is assumed that meningiomas of the geniculate ganglion may originate from arachnoid cell rests displaced during embryogenesis and lying within the sheath of the facial nerve.⁵ These tumors usually affect young patients (our first patient, a 56-year-old, represents the oldest reported case) and slowly progressive facial nerve weakness is the most common symptom. Auditory function may or may not be normal, depending on the size of the tumor and its involvement with the middle or inner ear.

High-resolution CT and MRI are complementary for a detailed radiological examination of

other lesions of the facial nerve. On CT, meningiomas of the geniculate ganglion appear as a mass occupying the superior portion of the tympanic cleft with erosion of the bony floor of the middle cranial fossa. The bony margins of these lesions are usually poorly defined, and sometimes the presence of intratumoral bone is suggested.¹¹ On MRI these tumors appear as a mass centered in the area of the geniculate ganglion, hypointense on T1-weighted and hyperintense on T2-weighted sequences. After gadolinium infusion, enhancement is heterogeneous. The clinical presentation and radiological appearance of meningiomas of the geniculate ganglion strongly resemble those of facial nerve neuromas, particularly hemangiomas. Thus, it is not easy to differentiate between these lesions preoperatively. Moreover, the differential diagnosis is of limited importance because their surgical treatment is not the same.

CONCLUSION

In the literature the combined transmastoid/middle cranial fossa approach is reported as the approach most used for these tumors.³ However, size and location of the tumor as well as preoperative hearing must be considered when this approach is planned. Most of the tumors reported previously were small and patients' hearing was intact at pre-

sensation. In our first patient, we performed the middle cranial fossa approach to remove the mass and also to preserve hearing. Our second patient's tumor was large, occupying the epitympanum and the mesotympanum with erosion of the ossicular chain and extending anteriorly to the temporomandibular joint and Eustachian tube. Its unusually large size was probably related to the long delay between the onset of the patient's facial nerve symptoms and diagnosis. Thus, a subtotal petrosectomy enlarged with a middle cranial fossa craniotomy was performed to resect the tumor completely.

Meningiomas of the geniculate ganglion should be distinguished from meningiomas arising from the overlying dura. The origin of these tumors is easily verified during surgery because in true geniculate ganglion meningiomas, the dura of the middle cranial fossa is easily elevated from the surface of the tumor.

Establishment of a cleavage plane between the facial nerve and tumor with anatomical preservation of the nerve has rarely been reported. At the end of the operation, a cable nerve graft is needed in most cases. The postoperative function of the facial nerve is related to the duration of its preoperative weakness. No results better than H-B grade III are achievable after cable nerve graft. In the case of facial nerve grafting, the H-B scale seems difficult to use. Functional recovery is better defined according to the repaired facial nerve recovery scale.⁴ Prolonged radiological follow-up is strongly recommended because the potential risk for development of neurofibromatosis has been reported.¹

NOTE

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COMMENTS FOR PUBLICATION

The authors report two patients with meningiomas of the geniculate ganglion. The diagnoses were confirmed pathologically after surgical exploration and resection. The authors detail the patients' clinical presentations as well as their diagnostic evalua-

tions, including the high-resolution CT and MRI necessary to evaluate these lesions.

Tumors arising from this region are exceedingly rare; only 14 cases have previously been reported. These two new cases add to the growing body of literature regarding the natural history and presentation of these lesions. The discussion also nicely illustrates the available range of surgical options depending on the size of the tumor.

These case reports should be read with interest by neurosurgeons and otorhinolaryngologists alike. Case 2, in particular, illustrates the need for a middle fossa craniotomy with a subtotal petrosectomy to access a large lesion in this region.

We applaud the authors on their case illustrations. Future publications reviewing all previous reports of meningiomas of the geniculate ganglion will no doubt be valuable and anticipated by many.

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