Cerebellopontine Angle Meningioma Associated with Cranial Accessory Nerve Neurinoma

—Case Report—

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Abstract

A 46-year-old female presented with a rare association of cerebellopontine (CP) angle meningioma with accessory nerve neurinoma manifesting as headache, occasional diplopia, speech disturbance, swallowing difficulty, and unsteady gait. Magnetic resonance imaging demonstrated a large tumor in the left CP angle. The tumor was totally removed through a lateral suboccipital approach. During the operation another smaller tumor was found originating from the cranial accessory nerve and was also totally removed. Histological examination found that the larger tumor was a meningotheliomatous meningioma and the smaller an Antoni type A neurinoma. The symptoms were apparently due to the larger tumor. Careful examination of neuroimages is necessary even after the main lesions responsible for the symptoms are identified.

Key words: multiple brain tumors, neurinoma, meningioma, accessory nerve, cerebellopontine angle, jugular foramen

Introduction

Multiple primary brain tumors of different histogenesis are not uncommon in patients with either phacomatosis or a history of cranial irradiation,²¹⁾ but are uncommon in the absence of such conditions.^{3,6,13)} The most frequent combination is meningioma and glioma in patients without neurofibromatosis, while meningioma and neurinoma is rare. Accessory nerve neurinoma is also an uncommon tumor. We present a rare case of cerebellopontine (CP) angle meningioma associated with accessory nerve neurinoma without neurofibromatosis, and discuss the clinical features of both multiple primary brain tumors and accessory nerve neurinomas.

Case Report

A 46-year-old female was admitted to our hospital on October 19, 1989 because of headache, occasion-

al double vision, speech disturbance, swallowing difficulty, and unsteady gait that had developed some 2 years prior to admission. There was no family history of neurofibromatosis or any other neurological disorders. Neurological examination on admission revealed Bruns' nystagmus with the larger component to the left, hypesthesia of the left face, diplopia on the left lateral gaze, left peripheral facial nerve paresis, decreased gag reflex and a deviation of the uvula and soft palate to the right on phonation, slurred speech, truncal ataxia, and discoordination of the upper and lower extremities predominantly to the left. No cutaneous signs of phacomatosis were observed. T₁-weighted magnetic resonance (MR) imaging revealed a large isointense mass in the left CP angle that measured 4 × 3.5 × 3 cm. The tumor was intensely and homogeneously enhanced with gadolinium-diethylenetriaminepentaacetic acid (Gd-DTPA) (Fig. 1).

She underwent a left lateral suboccipital craniectomy on October 31. A reddish-gray, nodular,

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Fig. 1 T₁-weighted MR images with (right) and without (left) Gd-DTPA enhancement, showing a large enhanced tumor in the left CP angle and a smaller one (arrow).

Fig. 2 Intraoperative photograph, showing the spherical tumor (T) arising from a rootlet of the left cranial accessory nerve. F: forceps, PICA: posterior inferior cerebellar artery, R: retractor, VA: vertebral artery, XI: eleventh cranial nerve.

moderately soft, and markedly vascular tumor was found in the left CP angle. The tumor was totally removed using a cavitron ultrasonic suction aspirator. The tumor arose from the dura over the posterior surface of the petrous bone halfway between Meckel's cave and the internal auditory meatus. The dural attachment of the tumor was extensively coagulated. During surgery another tumor was found originating from a rootlet of the left cranial accessory nerve (Fig. 2). The tumor was yellowish pink and round, measured 5 mm in diameter, and was located in the cerebellomedullary cistern near the jugular foramen. This tumor was

also removed by excising the rootlet on both sides of the tumor. Histological examination revealed that the larger tumor was a meningotheliomatous meningioma and the smaller an Antoni type A neurinoma (Fig. 3). The smaller tumor, although visible as a small mass, was not identified on the preoperative Gd-DTPA-enhanced MR image (Fig. 1 right).

Postoperatively, she developed fifth through tenth cranial nerve pareses on the left, which later improved gradually. She was doing well with only left hearing loss but no tumor recurrence 3 years 5 months after surgery.

Discussion

Only five cases of neurinoma associated with meningioma in patients without neurofibromatosis have been reported.^{1,7,13,24,30} Table 1 summarizes the

Fig. 3 upper: Photomicrograph of the larger CP angle tumor specimen, showing meningotheliomatous meningioma. HE stain, $\times 260$. lower: Photomicrograph of the specimen of the tumor arising from the accessory nerve, showing Antoni type A neurinoma. HE stain, $\times 260$.

Table 1 Summary of the six reported cases of meningioma and neurinoma without association of neurofibromatosis

Author (Year)	Age/ Sex	Location (Histology)
Gardner and Turner (1939) ⁷⁾	48/F	lt CP angle meningioma (fibroblastic), lt acoustic neurinoma
Kitamura <i>et al.</i> (1965) ¹³⁾	24/F	lt frontal meningioma (meningothelial), lt acoustic neurinoma (Antoni type A)
Akagi <i>et al.</i> (1973) ¹⁾	23/F	lateral ventricle meningioma (transitional), lt acoustic neurinoma (Antoni type A and B)
Schnegg and Tribollet (1984) ²⁴⁾	39/M	lt parietal meningioma (transitional), lt facial nerve neurinoma
Wilms <i>et al.</i> (1992) ³⁰⁾	47/F	rt CP angle meningioma (fibromatous), rt acoustic neurinoma
Present case	46/F	lt CP angle meningioma (meningothelial), lt accessory nerve neurinoma (Antoni type A)

location and histology of these cases. Cases of bilateral acoustic neurinomas associated with meningioma are omitted because they were considered to be neurofibromatosis II.¹¹⁾ The coexistence of meningioma and neurinoma is more common in middle-aged women, but ours is the first reported combination of a CP angle meningioma and an accessory nerve neurinoma.

Several etiologies for multiple primary brain tumors of different histogenesis have been proposed. Embryonic residue with potential malignancy may subsequently develop into multiple brain tumors,²⁾ while carcinogenic stimuli could also produce multiple tumors of different histogenesis.¹⁶⁾ A subsequent brain tumor, especially meningioma, could also occur in a patient after cranial irradiation for a previous brain tumor,²⁹⁾ or as the result of a local irritative effect from a preexisting tumor.²³⁾ Phacomatoses are often associated genetically with multiple brain tumors of both different and same histogeneses.²¹⁾ Multiple brain tumors with a different histogenesis might also occur purely coincidentally.^{13,23)} The tumors of our patient were not considered to be of embryonic origin, nor was there evidence of either phacomatosis or a history of carcinogenic stimuli including cranial irradiation. Since the two tumors were located proximally on the same side of the posterior fossa but not in direct contact, local irritation was unlikely.

Tumorigenesis in man has recently been analyzed by the molecular genetic approach. Casalone *et al.*⁴⁾

found that a suppression gene of meningioma is located on the long arm of chromosome 22, while Seizinger *et al.*²⁵⁾ demonstrated that a specific loss of a gene on chromosome 22 is frequently present in patients with unilateral acoustic neurinoma, as well as bilateral acoustic neurofibromatoses. Therefore, the genes responsible for the occurrence of meningioma or neurinoma are both located on chromosome 22. Consequently, the combination of meningioma and neurinoma might have a common origin in affected sites on chromosome 22. The association of these tumors in our patient might have been determined genetically, but no gene analysis was done.

A neurinoma arising in the ninth, tenth, or eleventh cranial nerve is often described as a jugular foramen neurinoma because of its proximity. Such a neurinoma has usually become quite large before detection, so it is often difficult to identify the nerve of origin. The neurinoma in our patient was small and clearly revealed the anatomical relationships between the neurinoma and the adjacent structures (Fig. 2). Iwasaki and Kondo⁹⁾ have also described such a small accessory nerve neurinoma within the jugular foramen.

Intracranial neurinomas usually arise from sensory nerves and seldom from pure motor nerves. Since the eleventh cranial nerve is a pure motor nerve, neurinoma arising from the accessory nerve is rare, with only 13 cases reported previously.^{5,8,10,12,15,17-20,22,27,28)} The patients were aged from 18 to 67 years old (mean 44 yrs). Accessory nerve neurinomas occurred more frequently in middle age, as for other intracranial neurinomas in general.³¹⁾ There were eight males and six females, apparently showing no female predominance in contrast to other neurinomas.³¹⁾ The duration of symptoms varied, but was usually lengthy, from 5 months to 5 years.

The clinical manifestations of accessory nerve neurinoma are combinations of fifth to twelfth cranial nerve pareses, nystagmus, cerebellar dysfunctions, and myelopathy.^{5,8,10,12,15,17-20,22,27,28)} Patients with tumors originating from the spinal root exhibit upper cervical myelopathy associated with eleventh cranial nerve dysfunction. The most frequent sign of accessory nerve neurinoma is eleventh cranial nerve paresis, although that of jugular foramen neurinoma is reported as hearing disturbance.^{8,9)} Hearing disturbance must be considered an important initial sign or symptom of accessory nerve neurinoma.^{15,19)} Only one case has demonstrated purely jugular foramen syndrome.⁹⁾

Small acoustic neurinomas of 1-5 mm size are reported to be asymptomatic.^{14,26)} The neurinoma in

our patient measured 5 mm in diameter, and caused little compression of adjacent structures (Fig. 2). Therefore, the preoperative lower cranial nerve pareses were caused by the larger CP angle meningioma.

The smaller tumor was not identified on the preoperative MR image. We recommend a more careful examination of neuroimaging even after the main lesions responsible for the neurological signs are found.

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