Isolated Metastases of Adenocarcinoma in the Bilateral Internal Auditory Meatuses Mimicking Neurofibromatosis Type 2

—Case Report—

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Abstract

A 56-year-old male with a history of lung cancer presented with isolated metastases of adenocarcinoma in the bilateral internal auditory meatuses (IAMs), mimicking the bilateral acoustic schwannomas of neurofibromatosis type 2, and manifesting as rapidly worsening tinnitus and bilateral hearing loss. Magnetic resonance imaging showed small tumors in both IAMs with no sign of leptomeningeal metastasis. The preoperative diagnosis was neurofibromatosis type 2. Both tumors were removed and the histological diagnoses were adenocarcinoma. Neuroimaging differentiation of a solitary metastatic IAM tumor from a benign tumor is difficult, although rapidly progressive eighth cranial nerve dysfunction suggests a malignant process. Metastases should be considered as a rare diagnostic possibility in a patient with small tumors in both IAMs.

Key words: acoustic schwannoma, cerebellopontine angle, hearing, internal auditory meatus, metastasis, neurofibromatosis type 2

Introduction

Acoustic schwannomas usually occur unilaterally, whereas bilateral occurrence is associated with neurofibromatosis type 2, an autosomal dominant disorder related to a genetic defect on the long arm of chromosome 22.19,27) Sporadic occurrence of neurofibromatosis is not rare. The diagnosis is usually based on neuroimaging of tumors in the bilateral internal auditory meatuses (IAMs) or cerebellopontine angles (CPAs).5) Microsurgical techniques now permit resection of small acoustic schwannomas with preservation of hearing,6,10,20) so early diagnosis of neurofibromatosis type 2 is an important priority.4,21,23) Acoustic schwannomas originate at the glial-Schwann cell junction of the vestibular nerve coverings, and are confined to the IAM in the early stage. Magnetic resonance (MR) imaging has facilitated early detection of small acoustic schwannoma confined to the IAM,10,22) which is important to accomplish surgical removal of acoustic schwannoma with hearing preservation.4,21,23) Tumors in the IAM are mostly benign neoplasms of neuroectodermal origin; acoustic schwannoma comprises 70% to 90% of IAM tumors. Metastatic tumors in the IAM are very rare.1,6,20)

We report a case of isolated metastases of adenocarcinoma in the bilateral IAMs, mimicking neurofibromatosis type 2 in symptoms and neuroimaging appearance.

Case Report

A 56-year-old male developed right-sided tinnitus 4 months prior to admission. The tinnitus worsened rapidly, and hearing loss occurred suddenly on the right 2 months later. One month prior to admission, tinnitus and hearing loss also became evident on the left. MR imaging showed small tumors in the bilateral IAMs. The patient was referred to our hospital with a tentative diagnosis of neurofibromatosis type 2 on September 10, 1992.

On admission, the patient complained of hearing loss and tinnitus on both sides. He was alert. A pure-tone audiogram revealed deafness on the right and 35 dB hearing loss on the left. Neurological examination found only a mild peripheral-type right facial nerve paresis. T1-weighted MR imaging re-
revealed small isointense tumors in both IAMs. The lesions were enhanced after gadolinium administration. The tumor on the left was confined to the IAM, but the larger tumor on the right protruded slightly into the CPA beyond the porus acusticus (Fig. 1). The tumor on the right was slightly high intensity on the T2-weighted images (Fig. 2). No other abnormal enhancement was observed in the CPA or other intracranial sites (Fig. 3). Thin-slice bone-window computed tomography (CT) showed that the right IAM was eroded and enlarged, whereas the left IAM was normal (Fig. 4). The patient had undergone right upper lobectomy for adenocarcinoma of the lung 4 years previously, and total removal of a metastatic adenocarcinoma in the right temporal lobe 14 months previously (Fig. 5 upper row). Chest radiography and total body scintigraphy performed during the present hospital stay showed no recurrence or extracranial metastasis of lung cancer.

On September 19, 1992, the patient underwent removal of the tumor in the left IAM via a left lateral suboccipital approach. The tumor did not protrude
beyond the porus acusticus, and no abnormality was found in the CPA. The posterior wall of the IAM was drilled out. Opening the dura mater overlying the IAM exposed a reddish tumor adherent to the vestibular nerve. The tumor was removed subtotally, with the facial and cochlear nerves left intact (Fig. 6). After surgery, hearing on the left was preserved, and no new neurological signs developed. The histological diagnosis was adenocarcinoma (Fig. 5 lower left).

On October 3, 1992, removal of the tumor in the right IAM was performed via a right lateral suboccipital approach. The tumor protruded beyond the porus acusticus into the CPA, with no other CPA abnormality. After opening the IAM, a reddish intrameatal tumor was removed subtotally (Fig. 7). The tumor was tightly adherent to the vestibulocochlear nerve in the IAM. The histological diagnosis was adenocarcinoma (Fig. 5 lower right). The patient underwent radiotherapy after surgery and did well as an outpatient until January 1993 when he developed back pain. Lumbar puncture findings indicated cerebrospinal fluid (CSF) dissemination of adenocarcinoma, of which he died on June 10, 1993. Autopsy was not permitted.

Discussion

In the present case, preoperative MR imaging and intraoperative observation indicated that the tumor on the left was confined to the IAM. The tumor on the right protruded into the CPA beyond the porus acusticus, but the eroded, enlarged IAM suggested that the tumor had originated from the IAM rather than the CPA. Origin of the tumor in the IAM was also suggested by the adhesion of tumor to the vestibulocochlear and facial nerves in the IAM but not in the CPA. Both metastatic tumors in the present case initially involved the IAM.

Metastatic tumors in the IAM usually result from either direct temporal bone involvement by the malignant tumor or leptomeningeal metastasis. Solitary metastasis in the IAM not associated with the temporal bone involvement or leptomeningeal metastasis is very rare. Only one previous case of isolated IAM metastasis has been reported. The present case involved metastases to both IAMs with no temporal bone involvement or leptomeningeal metastasis, despite the previous history of left temporal lobe metastasis. The minimal extent of subarachnoid space in the IAM makes dissemination of malignant cells through the CSF to the IAM relatively unlikely, so the isolated IAM metastases were more likely to have a hematogenous origin.

Metastatic tumors in the CPA are more common, accounting for 0.2% to 2% of all CPA tumors. Cytological examination of the CSF or histological study of a meningeal biopsy specimen has shown malignancy in reported cases of CPA metastasis. Metastasis to the CPA is considered to have a leptomeningeal, not a hematogenous origin. The CPA is a common site of leptomeningeal metastasis because of the abundant CSF in the large cisternal space. Previous cases of CPA metastasis have not shown enlargement of the IAM. This observation supports the idea that these CPA metastases originated in the CPA, not the IAM. Although IAM metastasis of hematogenous origin may expand into the CPA in late stages of progression, most CPA metastases are of leptomeningeal origin and do not result from IAM metastasis.

Progressive bilateral eighth cranial nerve dysfunction is the most common symptom of neurofibromatosis type 2, but the auditory function of our patient deteriorated exceptionally rapidly,
which is uncommon in patients with benign tumors in the IAM. Our patient also developed right facial nerve paresis. Small acoustic schwannomas rarely cause facial nerve paresis. Rapidly progressive eighth cranial nerve dysfunction accompanied by facial nerve paresis suggests malignant tumors in the IAM rather than acoustic schwannomas. However, schwannomas of the facial nerve occurring near the geniculate ganglion can cause both facial nerve paresis and eighth cranial nerve dysfunction even at an early stage. The characteristic sign of IAM metastasis is severe, unremitting pain localized to the mastoid and retromastoid areas, requiring narcotics for control. Our patient did not complain of mastoid pain. Considering the above possibilities and manifestations, the clinical differentiation of an IAM metastasis from a benign tumor can be very difficult.

In the present case, neuroimaging examinations including MR imaging and CT failed to differentiate IAM metastases from acoustic schwannomas. The features of the metastases on T1-weighted images obtained before and after gadolinium administration were similar to those of small acoustic schwannomas. CPA metastasis can be differentiated from acoustic schwannomas because the metastasis shows isointensity on T2-weighted images, which is uncommon in acoustic schwannomas. However, the IAM metastases showed a slightly hyperintense signal on T2-weighted images. The eroded, enlarged IAM observed in the present case is often associated with small acoustic schwannomas. Thus, radiological differentiation of the IAM metastases from small acoustic schwannomas was difficult in this case.

Metastases should be considered as a rare but possible diagnosis for small bilateral IAM tumors, especially when the patient presents with rapidly progressive eighth cranial nerve dysfunction.

References

Fig. 6  Intraoperative photographs of the tumor in the left internal auditory meatus (IAM) as seen from the left lateral suboccipital approach. No abnormality is seen in the cerebellopontine angle (upper; arrow, porus acusticus). After opening the dura mater overlying the IAM (lower left; double arrow), the tumor can be seen in the IAM. The facial and cochlear nerves remain intact after subtotal tumor removal (lower right).

Fig. 7  Intraoperative photographs of the tumor in the right internal auditory meatus (IAM) as seen from the right lateral suboccipital approach. The tumor (upper; arrow) protrudes beyond the porus acusticus into the cerebellopontine angle. After opening the IAM, the intra- and extrameatal tumor is subtotally removed (lower).

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