Bilateral malignant melanoma metastases to the internal auditory canal/cerebellopontine angle: surgical management and preservation of function

Case report

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Although intracranial metastases of malignant melanomas are common, localization at the cerebellopontine angle (CPA) or in the internal auditory canal (IAC) is rare, and bilateral presentation especially so. We present the case of a 46-year-old Caucasian woman with bilateral IAC/CPA lesions and a prior history of malignant melanoma on the right leg. During preoperative investigations, the presence of the bilateral IAC/CPA lesions along with several radiologically identified lesions along the neural axis led to the suspicion that she had neurofibromatosis Type 2 despite her history of malignant melanoma and the lack of characteristic skin lesions and family history. Histopathological analysis of the resected lesion confirmed the intraoperative diagnosis of bilateral CPA malignant melanoma metastases. Surgical removal of the tumors via the retrosigmoid approach with preservation of normal bilateral facial nerve function and unilateral serviceable hearing, combined with control of the systemic disease, provided this patient with a near-normal quality of life for at least 42 months after the initial diagnosis of melanoma. (DOI: 10.3171/JNS/2008/108/4/0803)

KEY WORDS • cerebellopontine angle tumor • facial nerve • hearing preservation • internal auditory canal • melanoma • metastasis

Abbreviations used in this paper: CPA = cerebellopontine angle; CSF = cerebrospinal fluid; IAC = internal auditory canal; MR = magnetic resonance; NF2 = neurofibromatosis Type 2.
Presentation and Examination. At admission the patient’s status was largely unremarkable and her only complaints were of tinnitus and decreased hearing on the left side (50 dB in the main speech area). Auditory evoked potential testing, however, showed regular findings on both sides. The results of oncological staging studies were unremarkable with regard to metastases. A repeated MR imaging study confirmed the presence of bilateral intra- and extracanalicular space-occupying lesions measuring 1.2 × 0.9 × 0.7 cm on the left side and 1.3 × 0.7 × 0.6 cm on the right side (Fig. 1). A spine MR imaging study revealed lesions of the left T-12 and right L-3 spinal nerve roots, thus increasing the suspicion of NF2, despite the patient’s history of malignant melanoma.

Operation. Taking into consideration the existing hearing loss, the decision was made to initially resect the left-sided lesion. Brainstem auditory evoked potentials, somatosensory evoked potentials and electromyography of facial nerve function were constantly monitored during surgery, which was performed with the patient in a semi-seated position. A retrosigmoid suboccipital craniectomy was performed; the dura was incised in a curvilinear manner just medial to the sagittal and inferior to the transverse sinus, and CSF was allowed to egress by opening the lateral cerebellomedullary cistern. The CPA was inspected and instead of the expected vestibular schwannoma, the posterior surface of a black-colored tumor was seen infiltrating the facial and cochlear nerves and slightly protruding from the IAC. The posterior and superior walls of the IAC were drilled, thus exposing the whole tumor. Due to the considerable infiltration of the facial nerve and the result of the frozen section analysis, showing malignant melanoma, the lesion was partially removed.

Postoperative Course. The patient’s recovery from surgery was uneventful and and her facial nerve function was found to be completely preserved, but she was found to have complete loss of hearing in the left ear. Final histopathological analysis confirmed the lesion to be a lightly pigmented epithelioid malignant melanoma.

Considering the highly aggressive nature of malignant melanomas, the presence of multiple metastases along the neuraxis and the relatively poor prognosis with regard to life expectancy, it was decided that further surgery on the right side be postponed indefinitely in favor of palliative radiation of the entire neural axis. Although the patient decided not to undergo radiation treatment (continuing with interferon therapy and undergoing adjunctive chemotherapy instead), follow-up MR imaging showed that the intracranial and spinal lesions remained constant in size. Twenty four months postoperatively, the patient presented again with deteriorating hearing on the right side. Findings of MR imaging studies suggested slight enlargement of both intracanalicular tumors (Fig. 2) and showed the spinal lesions as unchanged.

Second Operation and Postoperative Course. The surgery was performed in an identical manner and a similar-looking tumor was found in the right IAC. This tumor, however, was not densely attached to the nerves and could be removed completely. The postoperative period was completely uneventful, with the patient experiencing normal facial nerve function and preserved serviceable hearing (Figs. 3 and 4). The histopathological analysis showed identical lightly pigmented epithelioid malignant melanoma (Fig. 5).

Discussion

Although malignant melanomas have a tendency for intracranial metastasis, the CPA and the IAC remain relatively rare sites. Seventeen cases of such metastases have been presented in the literature; the average age of the patients in these cases was 49.8 years, and their neurological signs and symptoms at presentation were related to cochlear or vestibular nerves dysfunction. The mean interval from primary melanoma treatment to CPA metastasis was 7.7 years, and the mean duration of survival after the diagnosis of metastasis was 11 months.3

Bilateral CPA masses are typical for NF2: vestibular...
schwannomas or facial nerve neuromas. Few authors report on different bilateral lesions: bilateral choroid plexus papillomas, bilateral endolymphatic sac tumors presenting as CPA masses, or bilateral CPA metastases. Of the 3 intracanalicular metastases presented by Falcioni et al., 1 was bilateral.

In the published reports on bilateral CPA melanoma metastases, the diagnosis was based on examination of the CSF, neuroimaging findings, or postmortem evaluation, rather on histopathological examination of resected tumor. Lee and Weber report on a patient whose MR imaging findings of enhancement of the meninges around the brainstem and posterior fossa were consistent with carcinomatous meningitis, and the diagnosis of bilateral CPA melanoma metastases was based on the detection of melanoma cells in the CSF sample. Tu et al. describe a patient with bilateral intracanalicular melanoma metastases, but the diagnosis was based purely on neuroimaging findings as the patient had multiple intracerebral melanoma metastases and did not undergo surgery. Arriaga et al. present the case of a 60-year-old patient with presumably bilateral amelanotic melanomas—only the right tumor was treated surgically, and the patient survived only 5 months postoperatively. Brackmann and Doherty present a series of 14 cases of melanoma metastases to the CPA, including 4 involving bilateral tumors. The diagnosis in 2 of these cases was made based on the basis of cytological analysis of CSF and a positive history of primary melanoma, and the patients were treated with whole-brain radiation therapy and intrathecal chemotherapy. In the other 2 cases only 1 of the CPA tumors was removed surgically, and this lesion was proven to be a metastatic melanoma on histopathological analysis.

**Fig. 2.** Axial (left) and coronal (right) T1-weighted contrast-enhanced MR images obtained prior to the second surgery revealing slight enlargement of both tumors.

**Fig. 3.** Axial (left) and coronal (right) T1-weighted contrast-enhanced MR images after the second surgery showing complete removal of the right-sided melanoma metastasis.
Preservation of hearing was not an issue in these cases and the translabyrinthine approach was applied, because some degree of hearing loss already existed.

Some authors state that metastatic tumors in the CPA are clinically distinct from benign CPA tumors and cause rapidly progressive sensorineural hearing loss, followed by onset of progressive facial nerve weakness through direct nerve involvement or edema. Typically, CPA melanomas are hyperintense on T1-weighted images and hypo-, iso-, or hyperintense on T2-weighted images and enhanced after the administration of gadolinium. Amelanotic melanomas, however, are isointense on T1-weighted images and hyperintense on FLAIR sequences. The neuroimaging features are not conclusive and cannot exclude the diagnosis of metastatic melanoma. The only characteristic specific to metastasis is the presence of multifocal cerebral lesions.

Although in the case presented in this paper, the existence of a primary malignant melanoma lesion elsewhere in the body increased the suspicion that the intracranial lesions could be metastases, the clinical course did not give us clues to the diagnosis. The typical images of bilateral intracanalicular lesions, along with the spinal nerve root lesions, led us to assume the presumptive diagnosis of NF2, despite the patient’s relatively advanced age for a first diagnosis of an NF2-associated lesion, the lack of other characteristic symptoms, and the absence of a family history of NF2. Surgical removal of the CPA tumors via the retrosigmoid approach with preservation of normal bilateral facial nerve function and unilateral serviceable hearing, combined with control of the systemic disease, provided a near-normal quality of life for our patient for at least 42 months after the diagnosis of the primary melanoma.

Conclusions

The diagnosis of melanoma metastases should be considered in cases of bilateral IAC/CPA tumors. Preservation of function in such cases is possible but greatly depends on the invasiveness of the tumor with regard to the facial and vestibulocochlear nerves.

References

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