Bilateral cerebellopontine angle metastatic melanoma: A case report

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Abstract
Although melanoma accounts for approximately 1% of all malignancies, melanoma metastases to the cerebellopontine angles (CPAs) are exceedingly rare. Here we describe a patient with melanoma metastases to the internal auditory canals and CPAs who presented with a remote history of cutaneous melanoma. This patient had a rapidly progressive hearing loss, vestibulopathy, and facial nerve dysfunction. Magnetic resonance imaging demonstrated bilateral, enhancing CPA lesions but was otherwise nonspecific. The diagnosis required a careful history, unilateral surgical resection for tissue acquisition, and histopathologic confirmation. A search for primary cutaneous melanoma at the time of presentation was negative. However, the history of cutaneous melanoma 8 years earlier distinguishes this patient’s metastatic disease from solitary primary intracranial melanoma, an equally rare disease. Treatment consists of surgical excision, radiation, chemotherapy, and immunotherapy. The prognosis for patients with melanoma metastases is generally poor, but isolated reports of long-term survival have been described. Metastatic disease to the CPAs must be included in the differential diagnosis for any patient presenting with rapid-onset VIIth or VIIIth cranial nerve symptoms.

Introduction
Malignant melanoma represents approximately 1% of all malignancies. The most common sites for spread are the lung, brain, liver, gastrointestinal tract, and bone, but melanoma is also the third most common metastatic lesion to the central nervous system (CNS). Autopsy studies on patients with metastatic melanoma have found melanoma metastases in the brain in 17.5 to 90% of cases.1 CNS melanoma tends to be diffuse, and patient survival is typically less than 3 months.2

In reviewing more than 1,000 cases of cerebellopontine angle (CPA) tumor, Brackmann and Bartels found only 3 lesions (0.2%) of metastatic origin.3 A search of the literature through March 2005 located 8 cases of metastatic melanoma isolated to the internal auditory canal (IAC) and CPA, 3 of which involved the IACs/CPAs bilaterally.2,4-12 Since that time, a review of the House Ear Institute experience identified 8 additional patients with CPA melanoma; 4 had bilateral disease.13 Therefore, we report the eighth such case with bilateral melanoma metastases filling the IACs and extending into the CPAs. Within 2 months of presentation, the CNS was diffusely involved.

Case report
A 56-year-old man presented to the local emergency department with a 1-day history of left facial weakness and numbness. He also described several days of generalized weakness, chills, and malaise before the onset of facial palsy. The patient’s medical history was significant for diabetes, hypertension, hyperlipidemia, hemorrhoidectomy, and a cutaneous melanoma excised from his back 8 years earlier.

Except for a mildly elevated blood glucose level, results of the patient’s laboratory tests were normal, as were his vital signs. Noncontrast computed tomography (CT) of the head demonstrated no acute pathology, and the patient was discharged from the hospital with a diagnosis of Bell’s palsy. He was started on oral steroids and acyclovir. Within 2 weeks, however, he progressed to bilateral facial weakness and hypesthesia.

Outpatient magnetic resonance imaging (MRI) of the brain (figure 1) was obtained by the consulting neurosurgical team. This study demonstrated bilateral enhancing masses in the patient’s IACs and CPAs. No other intracranial lesions were found. The patient was referred to our clinic for consultation and further management.

During our initial evaluation, the patient was found to have bilateral facial paresis and hypesthesia. An audiogram was obtained, and speech discrimination scores were 100%
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on both sides. Within 1 week, however, he progressed to bilateral, profound sensorineural hearing loss. Over the subsequent 2 weeks, he developed bilateral tinnitus, rapidly progressive balance dysfunction, and blurred vision. Physical examination now demonstrated anisocoria (right pupil 4 mm and left 2 mm) and complete bilateral facial paralysis (House-Brackmann grade VI).

The patient underwent a translabyrinthine excision of the left CPA mass. During surgery, the tumor was found to be soft and friable with no obvious capsule. The VIIth and VIIIth cranial nerves were engulfed by the mass except laterally in the IAC and again medially at the brainstem, where each could be identified distinctly. Involved portions of the facial nerve were resected in continuity with the tumor. The tympanic and labyrinthine segments of nerve were then rerouted and a primary anastomosis was performed on the brainstem remnant.

Histopathologic examination of the specimen (figure 2) revealed large, pleomorphic polygonal cells with prominent nuclei. These cells were positive for S-100 protein and demonstrated greater than a 50% proliferative index by Ki67 immunohistochemistry. These findings were consistent with the histopathology of the cutaneous melanoma excised from the patient’s back 8 years earlier.

By postoperative day 3, the patient began complaining of severe frontal headaches. He was hyponatremic as a result of SIADH (syndrome of inappropriate antidiuretic hormone secretion) and had mild pulmonary edema. CT of the chest, abdomen, and pelvis failed to reveal any obvious signs of malignancy. A bone scan was also negative. Deteriorating mental status prompted repeat head CT and MRI. The MRI now demonstrated lesions in both hypoglossal canals and several masses in the lateral ventricles in addition to the known lesion in the right CPA. A lumbar puncture was positive, with tumor cells present in the cerebrospinal fluid. Radiation and chemotherapy were recommended. Unfortunately, the patient deteriorated rapidly and died.

Discussion

It is important to distinguish melanoma metastatic to the CPA from primary intracranial melanoma. Primary intracranial melanoma is thought to arise from melanocyte cell rests of neural crest origin that are distributed within the leptomeninges. These cells can be precursors to malignant melanoma, melanocytoma, pigmented schwannoma, and pigmented medulloblastoma.15 Patients with primary intracranial melanoma do not have a history of cutaneous melanoma, and a thorough metastatic workup shows no evidence of a primary lesion. Mean survival ranges from 6 months in patients with diffuse leptomeningeal melanoma to 19 months for those with solitary primary intracranial melanomas.14

The typical presentation of metastatic melanoma to the IAC and CPA is rapidly progressive auditory, vestibular, and facial nerve dysfunction. These tumors infiltrate nerves rather than compressing them. Patients typically have a remote history of cutaneous melanoma, and most reported cases of solitary IAC/CPA metastases have latency periods of more than 5 years. Our patient’s primary lesion—a cutaneous melanoma on his back—had been resected 8 years earlier. Unlike benign lesions of the CPA, these lesions are commonly associated with headaches, nausea, emesis, and the involvement of other cranial nerves, such the Vth, IXth, Xth, and XIth.5

Metastasis likely results from hematogenous seeding of the IAC with subsequent growth into the CPA.5 Surgically obtained tissue is necessary for a histologic diagnosis, and special stains such as proliferation assays, S-100 protein, and HMB-45 are needed. A complete metastatic workup—including examination of the skin and mucous membranes; CT scans of the chest, abdomen, and pelvis; and a bone scan—should be performed. There is an emerging role for positron-emission tomography in the workup of malignant melanoma.16,17

The literature indicates that intracranial melanoma has
varied MRI findings. Shortening of T1 and T2 relaxation times may be most characteristic, but these tumors have been found to be hypointense, hyperintense, or isointense to surrounding brain tissues. Most lesions enhance with gadolinium. Variations in melanin content and the presence of blood within the tumor may affect imaging characteristics on MRI. Whether solitary brain melanoma lesions are metastatic or primary, surgical resection palliates CNS symptoms and is thought to improve survival. Radiation, chemotherapy, and immunotherapy are also recommended. While prognosis is usually poor, Arriaga and colleagues reported 1 case with greater than 5-year survival. Metastatic disease to the IAC/CPA must be included in the differential diagnosis for any patient presenting with rapidly evolving cranial neuropathies of the lateral skull base.

References