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Imaging Case of the Month: Bilateral Internal Auditory Canal Melanoma

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Neurofibromatosis type 2 (NF2) is a rare, autosomal dominant disorder resulting from mutation of the NF2 tumor suppressor gene on chromosome 22. A mutated form of the gene product, merlin, leads to a predisposition for tumors in the central and peripheral nervous system. Though many sets of diagnostic criteria exist, all agree that the finding of bilateral vestibular schwannomas confirms the diagnosis. Patients may present with tinnitus, imbalance, hearing loss or facial weakness. However, many other lesions can affect the cerebellopontine angle (CPA) including, meningiomas, epidermoid cysts, ependymomas, melanomas and metastatic lesions. This case report outlines the importance of considering the differential diagnosis in patients with bilateral CPA lesions.

A 70-year-old male with long standing bilateral hearing loss and a history of cholesteatoma presented with worsened right-sided hearing. Audiometry revealed a profound right sensorineural hearing loss (SNHL) and high frequency left SNHL. Word recognition scores were 0% in the right ear and 72% in the left. Imbalance with head rotation, urinary incontinence, memory deficits and recent falls were also present. He had a history of a skin neurofibroma, but no malignancies. There was no family history of hearing loss, neurofibromatosis or cancer. Otoscopic examination and facial nerve function were normal. The head thrust test was positive with rightward rotation, but normal with rotation to the left. Magnetic resonance imaging (MRI) revealed bilateral enhancing lesions at the CPA and internal auditory canal (IAC) (Fig. 1A). The compilation of findings led to a presumptive diagnosis of NF2 with bilateral vestibular schwannomas and associated normal pressure hydrocephalus. A plan for ventriculoperitoneal (VP) shunt placement and repeat MRI to evaluate tumor growth rate was initiated.

Shortly after the presumed diagnosis, the patient presented with increasing confusion, unsteady shuffling gait and urinary incontinence. Imaging showed worsened ventriculomegaly with intraventricular hemorrhage, acute subdural hematoma, interval increase in right CPA lesion, an enhancing lesion of the right cerebellar hemisphere and chronic subdural hygroma (Fig. 1B). A VP shunt was placed which resolved his urinary incontinence but not the mental deficits or ataxia. Serial CT scans revealed extension of the hemorrhages, increasing edema, and mass effect. The patient died shortly afterwards from herniation due to massive acute subarachnoid hemorrhage.

At autopsy, foci of metastatic melanoma were identified at the CPA and left corpus callosum. Tumor cells expressed immunohistochemical markers for vimentin, S-100 and HMB45, and were negative for pankeratin cocktail, cytokeratin 7 and cytokeratin 20 (Fig. 2). No primary site of malignancy was identified.

Malignant melanoma is increasing in incidence. It comprises 1–8% of malignancies and is the third most common tumor to metastasize intracranially(1). Despite this, most metastases are to the gray matter, and CPA metastases are rare, especially without evidence of disseminated metastases. On MRI, melanoma is typically hyperintense on T1 and hypointense on T2. Amelanotic lesions, like this one, are often hypointense or isointense on T1 and hyperintense on T2, mimicking neuromas. Given their high vascularity, gadolinium enhancement is expected. These findings make it difficult to differentiate melanoma from typical benign CPA lesions on the basis of imaging alone. History of prior skin lesions, age at diagnosis and family history are crucial to proper diagnosis, and a high index of suspicion is necessary.

Brackmann et al report on 8 subjects with melanoma of the CPA(2), three of which were previously reported(3). In this series, 6/8 (75%) had a history of cutaneous melanoma and one had a primary CPA tumor. Four subjects had bilateral lesions. Jacob et al. report a case of bilateral CPA melanoma in a patient with a history of cutaneous melanoma 8 years prior(4). Subarachnoid hemorrhage is relatively common with melanoma metastatic to the brain(5), and the new lesion near the corpus callosum suggested metastatic disease. The patient's age at presentation and the negative family history of neurofibromatosis are further clues that, in hindsight, made the diagnosis of NF2 less likely. The current case is very unusual in that bilateral CPA metastatic melanoma was found without a prior melanoma history and without evidence of a primary lesion on autopsy. With increasing incidence of melanoma in society and better access to MRI, it is likely that we will also see an increased incidence in isolated CPA melanoma. Resection has proven successful in some cases; with several patients experiencing palliation of their CNS symptoms, and others, with primary lesions, experiencing a survival benefit(2). This identifies the importance of considering a full differential of CPA lesions when patients with bilateral lesions are identified. Melanoma should be considered with presentation at advanced age, rapid tumor growth, and subarachnoid hemorrhage.

References

1. Gerganov VM, Hore N, Herold C, et al. Bilateral malignant melanoma metastases to the internal auditory canal/cerebellopontine angle: surgical management and preservation of function. *J Neurosurg.* 2008; 108:803–7. [PubMed: 18377262]
2. Brackmann DE, Doherty JK. CPA melanoma: diagnosis and management. *Otol Neurotol.* 2007; 28:529–37. [PubMed: 17414177]
3. Arriaga MA, Lo WW, Brackmann DE. Metastatic melanoma to the cerebellopontine angle. Clinical and imaging characteristics. *Arch Otolaryngol Head Neck Surg.* 1995; 121:1052–6. [PubMed: 7646858]
4. Jacob A, Brightman RP, Welling DB. Bilateral cerebellopontine angle metastatic melanoma: a case report. *Ear Nose Throat J.* 2007; 86:388–90. [PubMed: 17702316]
5. Maiuri F, D'Andrea F, Gallicchio B, et al. Intracranial hemorrhages in metastatic brain tumors. *Journal of neurosurgical sciences.* 1985; 29:37–41. [PubMed: 4067634]

Figure 1.

Panel A: T1-weighted axial MRI with gadolinium showing a 15×10 mm enhancing lesion in the right cerebellopontine angle and a similar but smaller (8.7×5.4 mm) lesion in the left CPA (arrows). Panel B: CT scan done one month after the MRI. This revealed a lesion in the corpus collusum (C), which was not visualized on the prior MRI, subarachnoid hemorrhage, chronic subdural hygroma (H), and hydrocephalus.

Figure 2.

Panel A: H & E stained section of tumor at the cerebellopontine angle showed pleomorphic cells with mitotic activity (arrow) but lacking cytoplasmic pigmentation, consistent with poorly differentiated neoplasm and confirmed as melanoma by diffuse cytoplasmic immunoreactivity for HMB45 (Panel B). The lack of low grade primary leptomeningeal melanoma in the background and the presence of an additional separate metastatic focus were consistent with metastatic melanoma, rather than primary CNS melanocytic tumor. Panel B. Immunohistochemistry for HMB45 confirmed melanocytic differentiation.