ABSTRACT

Objective: To understand the symptoms, signs and diagnostic management of two patients with bilateral metastatic disease to the internal auditory canal. Magnetic resonance imaging (MRI) findings mimic bilateral vestibular schwannoma. In addition, a literature review of bilateral metastatic disease to the temporal bone will be performed.

Methods: Retrospective case series at a tertiary referral center. Two patients with progressive bilateral sensorineural hearing loss. Both were evaluated through lumbar puncture with cerebrospinal fluid cytology and MRI of posterior fossa.

Results: Two patients presented with rapidly progressive, bilateral sensorineural hearing loss. One patient also had a complete unilateral facial paralysis. MRI of the brain demonstrated enhancing lesions in each internal auditory canal. Lumbar puncture with cerebrospinal fluid (CSF) cytology was performed for both patients. CSF cytology was positive for malignant cells in the patient with facial paralysis with worsening hearing in her right ear and intermittent, non-vertiginous dizziness. She also complained of a one-week history of progressive right-sided facial paralysis. Audiometric testing demonstrated a profound, bilateral sensorineural hearing loss. At the time of presentation, she complained of a four-week period of hearing loss. At the time of presentation, she complained of a four-week period of hearing loss.

Conclusions: The differential diagnosis of patients with progressive, bilateral sensorineural hearing loss should include metastatic disease. Facial paralysis in this situation is a poor prognostic sign. CSF cytologic analysis may help to determine if lesions found on MRI are due to malignant disease.

DISCUSSION

A benign neoplasm is the most common lesion of the cerebellopontine angle (CPA) and its complications. Lesions of the CPA are most commonly unilateral and typically present with gradual sensorineural hearing loss with or without tinnitus [1]. Approximately 3 – 10% of patients with vestibular schwannoma present with sudden, unilateral sensorineural hearing loss [7]. The differential diagnosis of progressive, bilateral sensorineural hearing loss includes metabolic, autoimmune, traumatic, ototoxic and metastatic causes. MRI findings of temporal bone metastatic disease may mimic those of Neurofibromatosis type 2 (NF2) when both internal auditory canals are involved. Some distinguishing features may help to distinguish these patients from those with metastatic disease. Patients with NF2 are typically diagnosed in their twenties, while patients with metastatic disease are more commonly identified in their fifties and older. In addition, facial paralysis is a very rare complication of NF2, whereas CPA lesion, except for facial nerve, may be present.

According to Albert and Terrence [11], hearing loss is the first manifestation of 10% of patients with meningeval carcinomatosis. Rapidly progressive, bilateral sensorineural hearing loss has been reported in association with metastatic malignancy to the eighth cranial nerve [2]. Metastatic spread can be through the hematogenous or leptomeningeal routes. Hearing loss is believed to be the result of direct tissue invasion and/or compromise of the blood supply to the eighth cranial nerve [2]. The presence of bilateral hearing loss on audiometric analysis is an uncommon finding in CPA lesions, even in large tumors. On the other hand, metastatic lesions have a relatively high incidence of FN involvement. As with SNHL, the cause of facial paralysis in metastatic CPA lesions is likely due to a decreased blood supply to the bone is believed to be due to invasion of the facial nerve and/or loss of blood supply.

CSF analysis and MRI in combination are considered the best means to diagnose leptomeningeval carcinomatosis. CSF analysis has low morbidity and, if positive, will typically alter the management of the patient. A false negative test, however, is common [6]. It is estimated that approximately 25% of patients with leptomeningeval carcinomatosis from breast cancer have a false negative CSF analysis [8]. Glantz et al. [12] suggested that false negative can be minimized by withdrawing at least 10.5 mL of CSF, processing the specimen immediately and obtaining the sample from a known site of leptomeningeval disease. Romp et al. [11] has proposed analyzing CSF sample for peptides expressed in breast cancer. Certainly these cases represent a diagnostic dilemma. However, as in one of our patients, given the rapid progression of symptoms and history of primary breast cancer, the most likely diagnosis is metastatic disease. This approach has been used by others in similar circumstances [6].

In Case 1, the patient neglected to keep up with age-appropriate cancer screening tests. A thorough history would uncover these deficiencies. This approach has proved productive more than once in our practice, consequently diagnosing bilateral and unilateral metastatic disease to the CPA.

In regards to our second case, approximately 5% of breast cancer patients will develop leptomeningeval metastasis during the course of their disease [8]. The fast presentation and the involvement of multiple cranial nerves should be considered as a red flag and a strong indication for malignancy.

CONCLUSIONS

MRI findings that demonstrate bilateral enhancing lesions of the internal auditory canal in cases of progressive, bilateral sensorineural hearing loss are highly suspicious for metastatic disease. These lesions may mimic the appearance of intracranial vestibular schwannoma. CNS metastasis of NF2 such lesion is uniformly due to metastatic disease. Evaluation of patients with progressive, bilateral sensorineural hearing loss should include MRI with and without contrast directed to the internal auditory canal. Identification of bilateral temporal bone lesions, or lesions suspicious for metastatic disease, should be evaluated with CSF cytologic analysis. If CSF analysis is negative, workup should include a total body metastatic analysis including age and sex appropriate cancer screenings.

REFERENCES