Bilateral Cochleovestibulopathy Due to Internal Auditory Canal Metastasis in a Patient with Stomach Cancer

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INTRODUCTION
Sudden onset bilateral vestibulopathy is a rare condition; it is one of the most difficult balance problems to treat because nearly all patients experience long-term unsteadiness and oscillopsia due to disturbance of the bilateral vestibulo-ocular response [1]. Compensatory spontaneous healing seldom occurs. Systemic application of ototoxic drugs, trauma, neurological problems such as migraine, multiple sclerosis, bilateral inner ear problems such as cochlear otosclerosis, immune mediated inner ear disease, bilateral Meniere’s syndrome, and congenital abnormalities such as large vestibular aqueduct syndrome may cause gradual or sudden bilateral loss of vestibular function [2]. Bilateral cochleovestibulopathy due to bilateral metastasis of the internal auditory canal (IAC) is extremely rare.

CASE PRESENTATION
A 44-year-old man with stage III stomach carcinoma developed sudden hearing loss in the right and then in the left ear within a few days. He also experienced sudden and severe loss of balance with severe headache. A verbal informed consent was obtained from the patient who participated in this study. An audiogram showed bilateral neurosensorial hearing loss that was profound in the left ear and moderate in the right ear (Figure 1). Videonystagmography with caloric testing (Micromedical Technologies, Inc., USA) revealed severe loss of excitability on both sides. Facial function was normal on both sides. Magnetic resonance imaging (MRI) of the temporal bone demonstrated symmetrical and linear contrast enhancement in T1- and T2-weighted images (Figure 2). Contrast enhancement was evident in the posterior fossa along the eighth nerves outside the IACs. Two diagnostic possibilities, bilateral IAC metastasis and bilateral cochleovestibular neurinitis, emerged after radiological consultation. The patient refused to undergo a lumbar puncture for cerebrospinal sampling to investigate tumor cells. He had received multiple intratympanic injections of steroids for a week because the consulting oncologist was not in favor of high doses of systemic steroids. One month later, the patient’s hearing showed partial recovery on both sides, and his balance was better due to vestibular exercises and medical support. However, he developed mild facial paralysis on the right side. The clinical picture was progressive and indicative of meningitis carcinomatosis. A control MRI showed no change in the masses on both sides of the IAC. The patient died in six months despite undergoing whole brain radiotherapy and chemotherapy.

DISCUSSION
Metastatic occurrence within the IAC is rare; the primary site is sometimes unknown. Rohfls et al. [3] reported an interesting case of a patient with sudden onset vertigo and hearing loss who underwent an operation due to the appearance of a mass in the IAC. It
was discovered that the patient had metastatic adenocarcinoma of unknown primary. Chang and Michaelidis reviewed literature reports of metastatic carcinoma of the IAC between 1946 and 2015. They found 102 cases; 52.9% of these were bilateral. Lung (21.2%), skin (18.6%) and breast (16.7%) metastases showed the highest rates of bilateral cases. Meningeal metastasis occurred at a much higher rate in bilateral cases (47.2%) compared to unilateral cases (8.5%). Brain parenchymal metastasis also occurred at a higher rate in bilateral cases (38.2%) versus unilateral cases (19.2%). Outcomes for cases of IAC metastases were generally poor, with 56.3% of unilateral cases and 86.1% of bilateral cases reporting patient death within 5 years of diagnosis.

Most lesions within the IAC are benign tumors; vestibular schwannomas account for about 90% of these cases. Bilateral presentation of vestibular tumor is always associated with clinical aspects of neurofibromatosis type II. Primary bilateral malignant occurrence inside the auditory canal is low, and these lesions are usually metastatic. The spread of tumor cells in the cerebrospinal fluid may result in a distinct condition known as meningitis carcinomatosis, which has extensive involvement of the meningeal membranes and indicates poor prognosis. Demonstration of tumor cells circulating in the cerebrospinal fluid confirms the diagnosis. The IAC is an unusual location for metastatic involvement. Gloria-Cruz et al. found only 2 cases of isolated IAC metastasis among 76 cases of temporal bones with metastasis. Possible routes for the spread of the tumor from the primary site to the IAC could be direct extension, leptomeningeal extension, leukemic infiltration, cerebrospinal fluid dissemination, and hematogenous spread. The hematogenous route seems to be the most likely

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**Figure 1.** Pure tone audiogram showing moderate to severe hearing loss in the right ear and almost total hearing loss in the left ear

<table>
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<th>Frequency (Hz)</th>
<th>Hearing threshold (dB)</th>
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<tr>
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<tr>
<td>250</td>
<td>10</td>
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<td>4K</td>
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<td>8K</td>
<td>60</td>
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**Figure 2.** a-d. Magnetic resonance imaging of the temporal bone (a-d). Axial images of the right (a) and left (b) IACs in T2-weighted view show the appearance of masses in both IACs. The fundus of the canal appears intact on both sides. Coronal (c) and axial (d) T1-weighted images with contrast enhancement showing linear enhancement of both IACs.
one in this case. However, bilateral occurrence is difficult to explain. Tumor cells reaching the cerebrospinal space by the hematogenous route may be spread by cerebrospinal fluid.

The mechanism of hearing loss and vestibular dysfunction in this case may be compression of the rapidly growing mass inside the canal. Intervention into the vascular supply of the inner ear is also probable because tumor cells infiltrate the dura first, unlike acoustic tumors, which originate from the vestibular nerve sheath. Sone et al. [7] reported auditory follow-up of a patient with tumor metastasis to both IACs. Auditory brainstem response eventually disappeared as the patient’s hearing gradually worsened. However, distortion product otoacoustic emissions were present at low frequencies, suggesting that the upper turn of the cochlea was intact. Image-guided differentiation based on magnetic resonance studies is poor [8]. These tumors present contrast enhancement in T1-weighted images as well. However, thick linear and extranodular contrast enhancement may be indicative of meningeal infiltration [9]. Rapidly evolving bilateral audiovestibular symptoms, headache, facial paralysis, and magnetic resonance images with the presence of cancer in the patient’s history are highly suggestive of meningitis carcinomatosis.

Treatment of bilateral loss of vestibular function is always difficult. Residual function of the vestibular system on both sides is not sufficient to promote spontaneous recovery. Medical support and vestibular rehabilitation can only partially restore the deficit. The patient had some relief after intratympanic steroid injection, medical support, and vestibular rehabilitation. However, there is no evidence of effective treatment for patients with bilateral vestibulopathy [10]. Patients experience lifetime chronic postural instability.

Informed Consent: Verbal informed consent was obtained from the patient who participated in this case.

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REFERENCES