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Case Report

A Case of Gastric Meningeal Carcinomatosis Involving Bilateral Hearing Loss: The Difference between Clinical Images and Autopsy Findings

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We describe a rare case of meningeal carcinomatosis (MC) in a 66-year-old man who presented with bilateral deafness and vertigo, initially presumed to be neurofibromatosis type-2. Brain magnetic resonance imaging (MRI) of the patient revealed bilateral gadolinium enhanced masses at the cerebellopontine angle. However, multiple central nervous system symptoms, including loss of consciousness, gradually appeared. He had a history of gastric cancer; therefore, a lumbar puncture was performed. Cytological examination of the cerebrospinal fluid confirmed the presence of adenocarcinoma cells. The general condition of this patient worsened, and he died 46 days after the first onset of hearing loss. An autopsy was performed, and multiple infiltrations of adenocarcinoma cells in the brain were confirmed, though undetected by MRI. The prognosis of MC is extremely poor; therefore, rapid diagnosis is important to prevent mortality. Retrospectively, a lumbar puncture could have been conducted earlier to identify MC, especially in consideration of the clinical history of this patient.

KEYWORDS: Meningeal carcinomatosis, neurofibromatosis-2, gastric cancer, vestibulocochlear nerve, facial nerve

INTRODUCTION

Meningeal carcinomatosis (MC) is the infiltration of metastatic malignant cells into the meninges from a distant and solid tumor, commonly from melanoma, breast, lung, or colon [1, 2]. MC causes symptoms involving several levels of the central and peripheral nervous systems, most frequently affecting cranial nerves III (oculomotor), IV (trochlear), VI (abducens), and VII (facial) [3]. An initial diagnosis of MC is usually performed using magnetic resonance imaging (MRI), which shows diffuse meningeal enhancement. To establish the final diagnosis of MC, cerebrospinal fluid (CSF) analysis should show malignant cells with increased protein and decreased glucose levels. A hearing impairment caused by MC is a relatively rare pathology; vestibulocochlear (VIII) nerve involvement is reported in >10% of cases [4].

Herein, we report a case of MC caused by gastric cancer that involved bilateral hearing loss and vertigo. In this case, we established an initial diagnosis of neurofibromatosis type-2 (NF-2) because brain MRI and clinical findings were consistent with the symptomology of this disease. In this report, we retrospectively analyzed the clinical course, including cranial nerve symptoms and MRI findings, and pathological findings obtained via autopsy.

CASE PRESENTATION

A 66-year-old male patient presented with a sudden onset of hearing loss in the left ear. He had a history of gastric adenocarcinoma (poorly differentiated adenocarcinoma), and had undergone a total gastrectomy 1.5 years prior to the onset of hearing loss. Peritoneal dissemination was found via laparotomy, and he was diagnosed with T4a(SE) N2 CY1 P1 stage IV. No clinical symptoms

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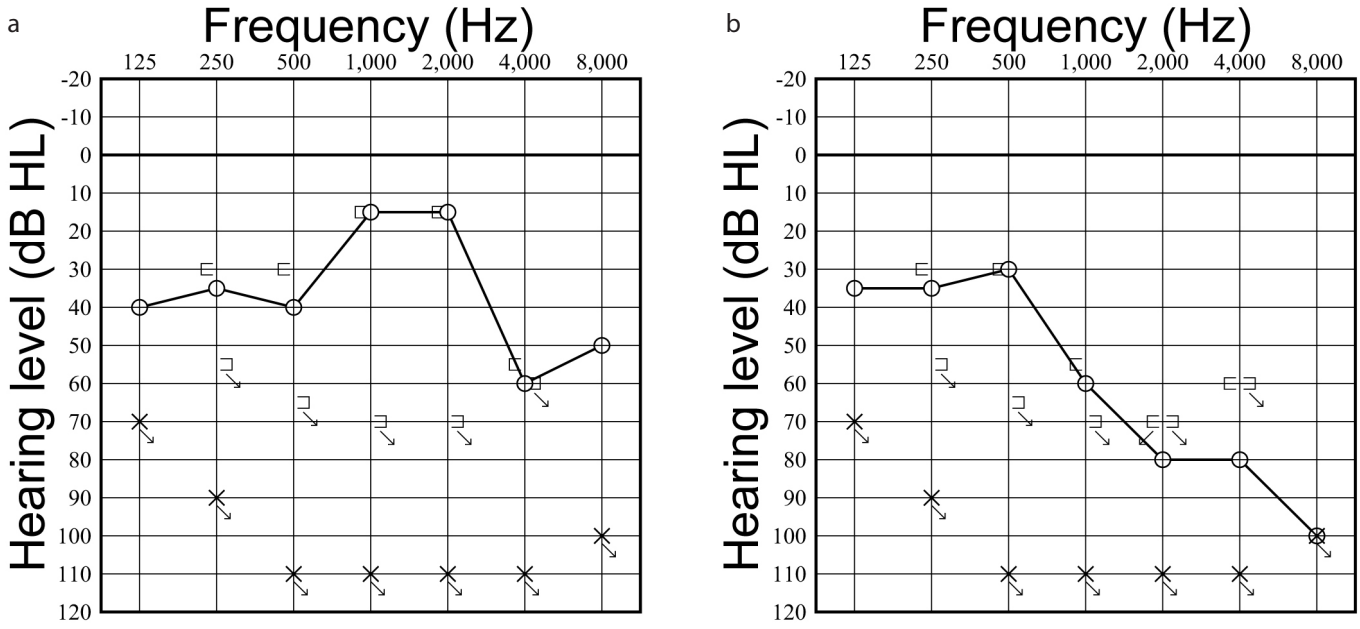

Figure 1. a, b. Changes observed in the audiogram. The patients' hearing level at first visit (a). The left hearing level was out of range at all frequencies. This is compared with the patient's hearing level 17 days after his first visit (b). Right hearing loss at high frequencies progressed; however, speech perception worsened.

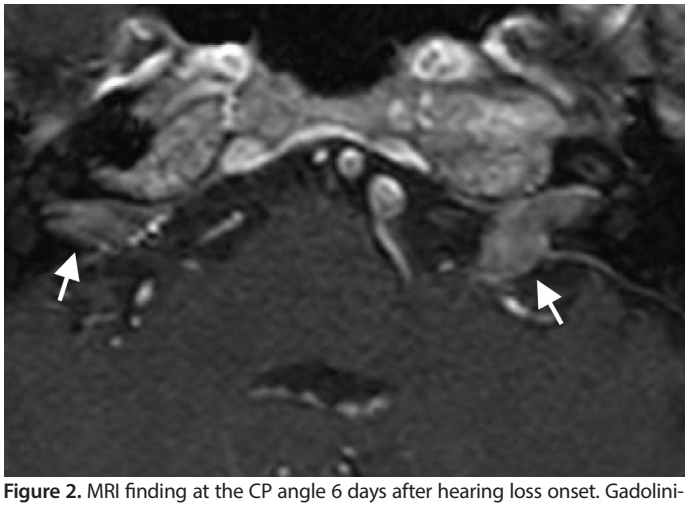

Figure 2. MRI finding at the CP angle 6 days after hearing loss onset. Gadolinium-enhanced T1-weighted image showing enhanced tumor lesions at bilateral internal auditory meati (white arrows). There were no tumor lesions at other brain sites, even after retrospective analysis.

were observed after surgery; therefore, no additional treatment was performed. The hearing deficit in the left ear was profound and out of range of the audiometer [>110 dB (HL) at all frequencies tested, as shown in Figure 1]. He also experienced slight dizziness and spontaneous rightward beating nystagmus. There were no other cranial nerve symptoms; therefore, he was diagnosed with sudden deafness in the left ear, and he was started on prednisolone treatment. However, his hearing in the left ear did not improve.

Four days after the onset of left hearing impairment, he began to experience left facial palsy. Therefore, we conducted a brain MRI that revealed bilateral gadolinium-enhanced masses at the cerebellopontine angle (CP angle), indicating that the most probable diagnosis was NF-2 (Figure 2). There were no other masses, which had smooth and localized edges, consistent with the typical presentation of NF-2. The lack of other neurological symptoms further supported this diagnosis. The neurosurgeon suggested gamma-knife irradiation for this case.

Fourteen days after the onset of left-sided hearing loss, vertigo and right-sided (contralateral) hearing loss appeared. The hearing level in the right ear was approximately 60 dB (HL; Figure 1); however, conversation was impossible due to poor speech perception. Additionally, loss of consciousness gradually appeared. Based on these new and severe symptoms, a lumbar puncture was performed. Cytological examination of the CSF revealed adenocarcinoma cells consistent with a diagnosis of MC, secondary to gastric cancer. The general condition of this patient began to deteriorate, and he died 46 days after the first onset of hearing loss.

On the day of death, an autopsy was performed with the family's consent. Pathological analysis showed poorly differentiated gastric cancer, and tumor metastasis to the abdominal lymph nodes, liver, pancreas, and peritoneum with dissemination. In the brain, bilateral metastasis of the adenocarcinoma was found at the CP angles, as indicated on MR images. Moreover, metastasis was observed in the pituitary gland, left frontal lobe, left basal ganglia, left hippocampus, midbrain, and medulla oblongata spreading along the subarachnoid space, despite obvious mass detection with MRI. The CP angle metastatic tumor infiltrated into the granular and Purkinje cell layers at the cerebellar cortex, and the structure of the vestibulocochlear and facial nerves were entirely replaced by metastatic cancer cells (Figure 3).

DISCUSSION

The symptoms of MC are typically widespread, and they often involve multiple components of the central nervous system. The most frequent symptoms of MC are headache, general weakness, altered mental status, nausea, and vomiting. Cranial nerve palsies are often apparent but usually not the only visible symptom [5]. A large autopsy study revealed that the incidence of MC is approximately 5%-8% in patients with can-

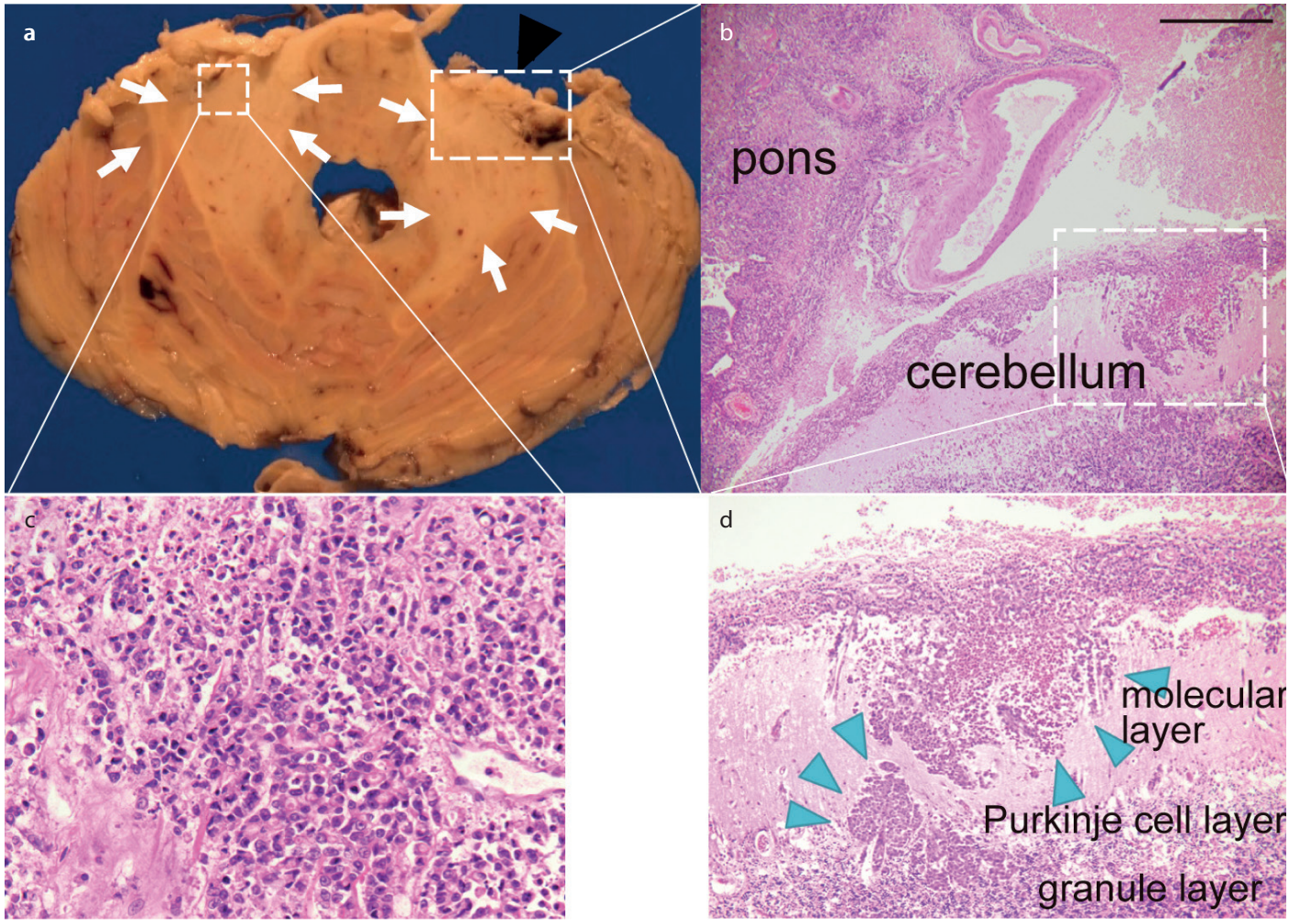

Figure 3. a-d. Autopsy findings at the CP angle. A macroscopic view of the cerebellum and pons, where bilateral CP angles (white arrows) were replaced by tumor tissue (a). The structure of vestibulocochlear and facial nerves is completely replaced by metastatic cancer cells (black arrow head). A low magnification view around the CP angle showing invasion of carcinoma cells in cerebellum tissue (white dotted square; b). By comparison, a high magnification view of tumor infiltration at the right CP angle shows the infiltration of poorly differentiated adenocarcinoma with a high nucleus-cytoplasm ratio (c). Tumor cell invasion in the cerebellum is observed at high magnification (d). The metastatic tumor (green arrow heads) has infiltrated into the granular and Purkinje cell layers at the cerebellar cortex along the subarachnoid space.

cer [2]. A multi-center retrospective analysis of gastric leptomeningeal carcinoma cases revealed that in patients with gastric MC, hearing loss (3.7%) and facial palsy (1.95%) are rare symptoms, while headache (85.1%) and nausea (59.2%) are the most common symptoms [4]. Isolated vestibulocochlear nerve symptoms are very rare in MC[6].

In this case, we confirmed the infiltration of neoplastic cells into the internal auditory meatus through the subarachnoid space and CSF. According to these pathological findings, the etiology of this patients' hearing loss was the direct invasion of metastatic cancer cells into the cranial nerves. The left hearing threshold, as measured on the audiogram, was preserved at lower frequencies; however, speech perception was extremely poor. This suggests that the left cochlear structure was preserved, but the cochlear nerve was destroyed by invasive cancer cells. Furthermore, the left-sided hearing loss and vertigo were sudden and profound. This was possibly due to vascular compromise, in addition to direct invasion and axonal destruction [7]. The facial nerve is often involved in cases of MC with hearing impairments [8]. In this case, hearing loss preceded facial palsy, which may indicate that sensory nerves are more vulnerable than motor nerves [9].

We initially identified this case as NF-2 because we did not observe any metastatic lesions in the brain, except at the CP angle, using MRI. Hearing loss and vertigo caused by vestibulocochlear nerve dysfunction and facial palsy caused by facial nerve dysfunction are common symptoms of NF-2, which occurred at the internal auditory meatus involving both cranial nerves. There were multiple metastatic lesions in the brain, which were confirmed by autopsy. However, detecting the metastatic lesions using MRI remained difficult, even retrospectively. However, sudden and bilateral onset of hearing loss, vertigo, and facial palsy are rarely encountered in cases of NF-2. The prognosis of MC is extremely poor, indicating the need for rapid diagnosis. Because of the clinical history of this patient, it would have been advantageous to conduct the lumbar puncture earlier to detect MC.

CONCLUSION

We reported a rare case of MC involving bilateral vestibulocochlear and facial nerve dysfunction, initially identified as NF-2, based on the MRI findings. In this case, although establishing the exact diagnosis was difficult, a lumbar puncture should have been performed earlier to enable detection of MC.

Informed Consent: Written informed consent was obtained from the parents of the patient's.

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