

CASE REPORT

Isolated Acute Hearing Loss as the Presenting Symptom of Leptomeningeal Carcinomatosis

Tzu-Hsien Lai^{1,2,3}, Chien Chen^{1,2}, Der-Jen Yen^{1,2}, Hsiang-Yu Yu^{1,2}, Chun-Hing Yiu^{1,2}, Shang-Yeong Kwan^{1,2*}

¹Department of Neurology, The Neurological Institute, Taipei Veterans General Hospital, ²National Yang-Ming University School of Medicine, Taipei, and ³Department of Internal Medicine, Yuanshan Veterans Hospital, Yilan, Taiwan, R.O.C.

Leptomeningeal carcinomatosis is characterized by diffuse infiltration of the meninges by metastatic cancer. We report a rare case of leptomeningeal carcinomatosis with an initial presentation of isolated acute hearing loss. Progressive unsteady gait and multiple cranial nerve palsies ensued. Computed tomography of the chest with contrast revealed a 2 cm nodule over the right upper lung. Cytology of cerebrospinal fluid confirmed the diagnosis of adenocarcinoma. The patient was discharged after diagnosis. Isolated acute hearing loss is a rare initial presentation in leptomeningeal carcinomatosis, not necessarily with a known history of malignancy. A high index of suspicion is mandatory for accurate diagnosis. [*J Chin Med Assoc* 2006;69(10):496–498]

Key Words: carcinomatous meningitis, hearing loss, leptomeningeal carcinomatosis, multiple cranial nerve palsy

Introduction

Meningeal carcinomatosis is the diffuse or multifocal infiltration of the meninges by metastatic carcinoma, usually with a known history of malignancy. However, it could be the initial presentation of occult tumors. The symptoms and signs are protean, mainly headache, backache, polyradiculopathies, multiple cranial nerve (CN) palsies and a confusional state.¹ Saenger² was the first to describe hearing loss as a consequence of meningeal carcinomatosis. Since then, there have been only a few reports of sudden-onset deafness as the presenting complaint of meningeal carcinomatosis.³ We present a case of an occult adenocarcinoma with acute hearing loss. Tinnitus, vertigo, unsteady gait and multiple CN palsies ensued.

Case Report

A 66-year-old man presented to a local hospital in September 2003 with acute onset of right ear hearing loss. Mild tinnitus, vertigo and unsteady gait soon followed. The patient had no major medical history. The symptoms progressed, and he was admitted to

another hospital 3 weeks later. Internal auditory artery infarction was impressed but brain magnetic resonance imaging (MRI) revealed no evidence of ischemic lesion. Three days after admission, double vision and left eye ptosis developed. Seven more days later, he lost his left-ear hearing. Verbal communication was impossible, and he was then transferred to our hospital for further evaluation. Neurologic examinations at that time revealed bilateral CN VIII, right CN VII and left CN III palsies.

After the patient was admitted to our hospital, we noted a mass on his routine chest film. Computed tomography (CT) of the chest with contrast enhancement (Figure 1) disclosed a 2 cm nodule over the right upper lung. During bronchoscopic exam, the patient had a transient shock and the procedure was stopped, so attempts to get an adequate pathologic sample failed.

Lumbar punctures were performed on days 37 and 47 after onset of hearing loss. The opening pressures were 330 mmH₂O and 250 mmH₂O, respectively. Elevated white blood cell count (380/mm³ and 100/mm³) and protein (188.6 mg/dL and 356 mg/dL) with reduced glucose (7 mg/dL and 32 mg/dL) were noted in exams of the cerebrospinal fluid (CSF) on both days. Gram stain, acid-fast stain, Indian

*Correspondence to: Dr Shang-Yeong Kwan, Department of Neurology, The Neurological Institute, Taipei Veterans General Hospital, 201, Section 2, Shih-Pai Road, Taipei 112, Taiwan, R.O.C.

E-mail: cchien@pavo.seed.net.tw • Received: December 2, 2005 • Accepted: May 19, 2006

ink stain and cryptococcal antigen test were all negative. Malignant cells in the CSF were noted in the second lumbar puncture (cytology-adenocarcinoma). Brain MRI (Figure 2) revealed bilateral auditory tube enhancement. Meningeal carcinomatosis from a lung primary cancer was diagnosed, and the family refused further treatment after explanation and discussion. The patient was discharged the next day.

Discussion

Infiltration of the meninges by metastatic carcinoma was first described by Eberth in 1870.⁴ Notably, past history of known malignancy is absent in 6–38% of patients. The site of the primary tumor is never identified in 2–4% of patients.^{4–6} In a study of 126 cytology-confirmed patients, the symptoms of meningeal carcinomatosis included headache, spine or radicular limb

pain, nausea/vomiting, weakness, and sensory disturbance, and the signs included altered mental status, meningismus, cerebellar signs, and multiple CN palsies.² Although multiple CN palsies are common, isolated acute hearing loss as the presenting symptom is rare.⁷

Hearing loss in meningeal carcinomatosis was first described by Saenger in 1900.² Alberts and Terrance⁸ summarized the clinical features as follows: (1) hearing loss is initially unilateral and associated with tinnitus; (2) unilateral hearing loss rapidly progresses to severe bilateral involvement; (3) audiometric and caloric studies reveal severe CN VIII impairment; and (4) facial nerve palsy is commonly noted at the time of hearing loss.

Lumbar puncture has long been recognized as the single most important diagnostic study, and the importance of repeated punctures has been stressed in several reports.^{1,6,8} Olson et al⁶ reported that over 90% of their 50 patients with meningeal carcinomatosis required 2 or more lumbar punctures, and only 37 of 50 had positive cytology even after repeated punctures. Other findings in the CSF included raised protein level, mild cellular pleocytosis, hypoglycorrhachia and increasing opening pressure.

Imaging studies, including myelography, CT and MRI, can aid in diagnosis. Of these, gadolinium-enhanced MRI is the most helpful.^{9,10} The typical findings include abnormal enhancement of the leptomeninges or CNs, intraventricular nodular enhancement, or communicating hydrocephalus.

Our patient experienced an initially isolated unilateral hearing loss, followed by mild tinnitus, vertigo, and unsteady gait. The hearing loss progressed to bilateral involvement and the patient became clinically deaf. Right facial nerve and left oculomotor nerve involvements were present later. CSF cytology was positive in the second lumbar puncture. Brain MRI

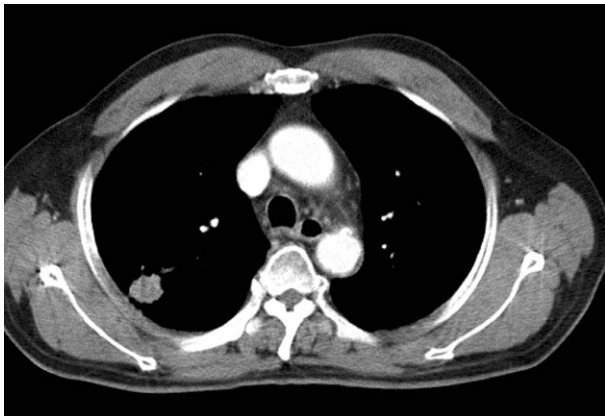


Figure 1. Chest computed tomography with gadolinium enhancement shows an enhanced 2 cm mass lesion over the posterior segment of the right upper lobe.

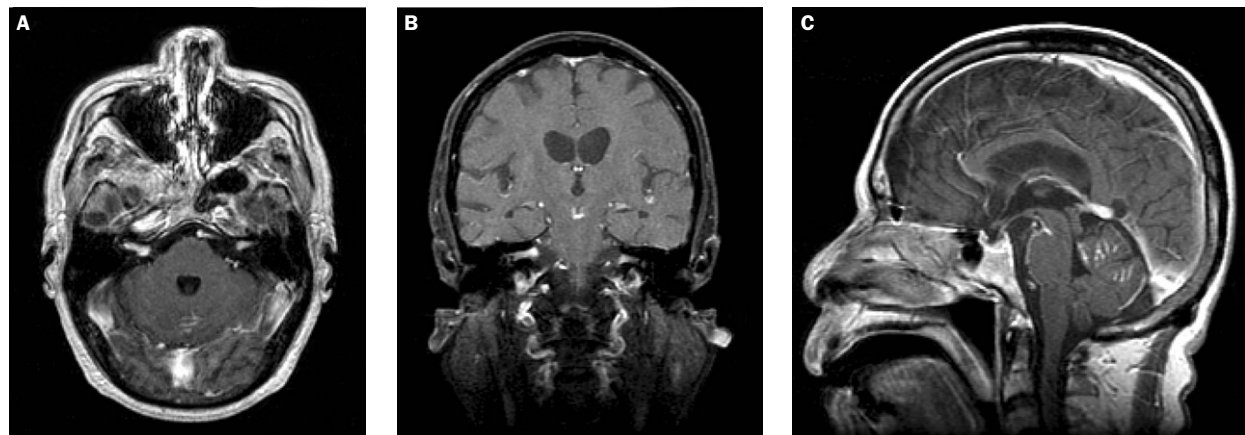


Figure 2. Gadolinium-enhanced magnetic resonance imaging shows strong enhancement of bilateral auditory tubes. Leptomeningeal enhancement is also shown.

disclosed characteristic gadolinium-enhanced bilateral auditory tubes. The diagnosis was adenocarcinoma with meningeal carcinomatosis, most likely from the lung.

Although rare, isolated hearing loss could be the first presentation of an occult malignancy.⁷ It could be unilateral with rapid progression to bilateral, or could be bilateral initially. A high index of suspicion is required for accurate diagnosis.

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