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Sudden onset sensorineural hearing loss caused by meningeal carcinomatosis secondary to occult malignancy: Report of two cases

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Abstract

Meningeal carcinomatosis (MC) is an uncommon form of metastasis of solid tumors. In the absence of clinical meningeal or parenchymal involvements, the sensorineural hearing loss (SNHL) as the starting symptom of MC is very infrequent. We report the history of two patients affected by MC who presented first with progressive SNHL. In both cases the early magnetic resonance imaging (MRI) finding mimicked bilateral masses in the cerebellopontine angle (CPA). Only the histopathologic result and surgical biopsy in cases 1 and 2, respectively, identified masses secondary to occult malignancy. However the available investigations could not discover the primary site of metastatic carcinoma. Despite the poor prognosis, because of the rarity and severity of MC we consider important to make known our experience in order to consider metastatic tumors in the differential diagnosis for sudden SNHL.

Keywords: Sudden sensorineural hearing loss; Meningeal carcinomatosis; Occult cancer

1. Introduction

Meningeal carcinomatosis (MC) is characterized by multifocal spread of tumor cells in the leptomeninges from distant solid tumors, commonly adenocarcinomas of breast or lung [1,2]. In most of cases diagnosis is made when a variety of neurological symptoms appear in patients with a known malignancy but is not infrequently they are the first finding. There have been only few reports of sudden deafness as the presenting complaint of MC secondary to an occult malignancy.

2. Case report

2.1. Case 1

A 56-year-old man underwent on ENT consultation for a right sudden deafness associated with tinnitus, auricular fullness and vertigo. History-taking disclosed an intestinal bowel disease (IBD) diagnosed seven years ago and an accidental report of high level of CA-19-9 (esophagogastroduodenoscopy and colonoscopy were negative for neoplasm).

Head and neck clinical examination was unremarkable and any kind of nystagmus or cerebellar signs could not be observed. The audiogram demonstrated a moderate–severe sensorineural hearing loss in the right side (Fig. 1A) and normal pure-tone average in the left one. According to our protocol therapy for sudden hearing loss he underwent a systemic administration of steroids and plasma expander and an oral assumption of acid acetylsalicylate without improvement. Approximately 4 weeks later he returned complaining of beating headache, bilateral tinnitus and left fluctuanting hearing loss (Fig. 1B).

Hematological and biochemical investigations were normal. Any screening for bacterial, viral or autoimmune disease resulted to be negative. An MRI with contrast enhancement (CE) revealed soft tissue masses in both internal auditory canals and cerebellopontine (CPAs) angles leading to a differential diagnosis between an acoustic neuroma or a neurosarcoidosis (Fig. 2A). Total body computed tomography scan showed a focal pulmonary interstitial opacity suggesting a granulomatous or infective disease.

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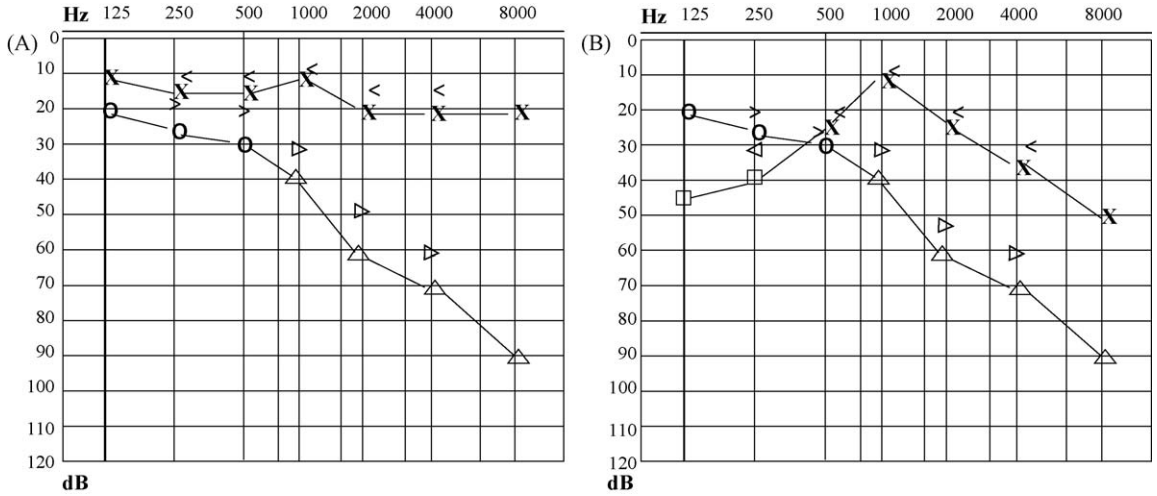

Fig. 1. Audiogram recorded at the beginning of the symptoms (A) and 2 months later (B) (symbols: *circle*: right ear air-conduction; c*ross*: left ear air-conduction; *open tip of the arrow to the right*: right ear bone-conduction; *open tip of the arrow to the left*: left ear bone-conduction; *triangle*: masked right ear air-conduction; *square*: masked left ear air-conduction; *closed tip of the arrow to the right*: masked right ear bone-conduction).

A lumbar puncture revealed normal protein and glucose levels within the cerebrospinal fluid (CSF) and the absence of neurotropic infections. However numerous unusual cells were found.

Basing upon radiological features an autoimmune disorder, namely sarcoidosis was hypothesized. For this reason systemic steroids were re-administered.

Nevertheless a right oculomotor nerve paresis and a visual impairment appeared.

A second brain MRI scan performed few days later showed a diffuse dural thickening, subaracnoid enhancement and nodular lesions within the internal auditory canal and on the right parietal region (Fig. 2B). A percutaneous biopsy of a lung lesion revealed a pulmonary aspergilloma (mycetoma, most likely iatrogenic). The new biochemical investigation revealed an increase of pancreatic markers (amylase and lipase). A new lumbar puncture showed malignant cells strongly evocative for adenocarcinoma. He died few days after the diagnosis of meningeal carcinomatosis. According to the autoptic results, death has been caused by diffuse distant metastasis of a pancreatic adenocarcinoma.

2.2. Case 2

A 64-year-old woman developed a left sudden hearing loss, tinnitus, auricular fullness and vertigo. Her past medical history was unremarkable. ENT clinical assessment was normal. A left deafness and a right moderate–severe sensorineural hearing loss could be detected (Fig. 3A). She underwent the same pharmacological treatment as case 1 without functional improvement. Hematological and bio-

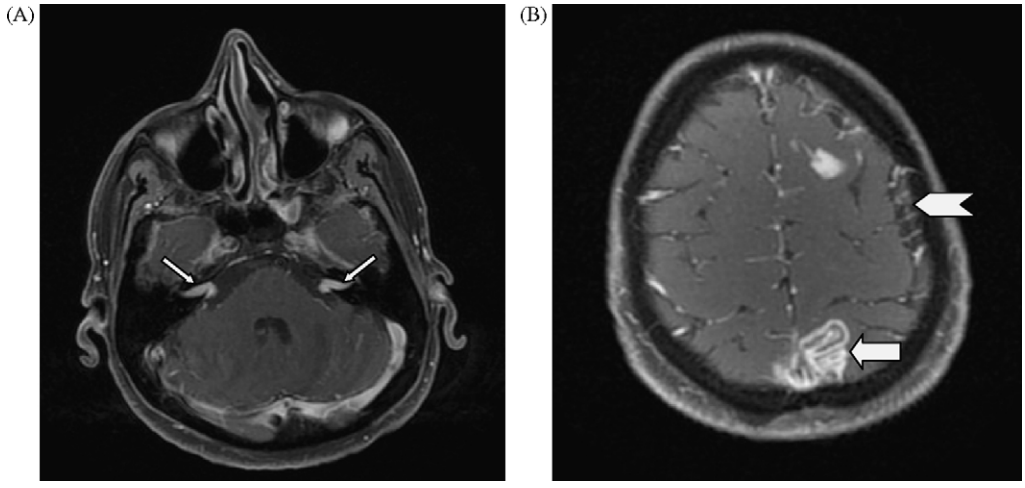

Fig. 2. (A) Brain MRI with contrast enhancement revealing soft tissue masses in both internal auditory canals and CPAs (white arrows). (B) The second brain MRI scan, performed few day later, showed a leptomeningeal dissemination characterized by dural thickening diffuse subaracnoid enhancement (white arrow) and enhancing nodular lesions on the right parietal region (white tip of the arrow).