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Sudden onset bilateral sensorineural hearing loss: a manifestation of occult breast carcinoma

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Abstract

Diffuse infiltration of the meninges by metastatic carcinoma (meningeal carcinomastosis) is a potential complication of systemic malignancy. It may present with a variety of neurological symptoms as any aspect of the neuraxis can be affected. Often there is a history of pre-existing malignancy.

The authors describe a case with an initial presentation of sudden onset profound bilateral sensorineural hearing loss. The underlying pathology was found to be an occult breast carcinoma, a previously unreported finding.

The role of cerebrospinal fluid cytology and radio-imaging in diagnosis is discussed. All previously reported cases of sudden hearing loss and meningeal carcinomatosis are reviewed.

Key words: Hearing Loss, Bilateral; Meningeal Neoplasms; Breast Neoplasms

Introduction

Meningeal carcinomas (leptomeningeal carcinomatosis, carcinomatous meningitis, malignant meningitis) is the diffuse or multifocal infiltration of the meninges by metastatic carcinoma. The primary tumours are usually stated to be adenocarcinomas of the breast or lung and malignant melanomas.[1]

Patients may present with a variety of symptoms and signs as any aspect of the neuraxis may be affected, the cerebral hemispheres, the cranial nerves and the spinal cord and roots.

Saenger[2] was the first to report sudden onset deafness as a consequence of meningeal carcinomatosis. Since then there have been several reports documenting deafness occurring as a result of metastatic disease. However, there have been only a few reports of sudden onset deafness as the presenting complaint of meningeal carcinomatosis, especially in cases of occult malignancy.

The authors present a case of an occult adenocarcinoma of the breast, in which the first manifestation of the neoplasm was sudden onset profound bilateral sensorineural hearing loss. The role of cerebrospinal fluid (CSF) cytology and neuroradiology in diagnosis is discussed. All previously reported cases of sudden hearing loss and meningeal carcinomatosis are reviewed.

Case report

A 61-year-old lady presented to the otolaryngology department with sudden onset bilateral hearing loss, clinical examination was unremarkable. An urgent magnetic resonance imaging (MRI) scan of the brain and temporal bones was requested. Approximately six weeks after presentation she was admitted to the ward complaining of anorexia, weight loss and vomiting. Her past medical history was unremarkable. Clinical examination revealed a cachexic lady with bilateral lower motor neurone facial nerve palsies (right side worse than left), an ataxic gait and a profound bilateral sensorineural hearing loss, confirmed on pure tone audiometry.

Haematological and biochemical investigations were normal. Syphilis serology, myeloma, vasculitis, virology and autoantibody screening was negative. The erythocyte sedimentation rate (ESR) was 28. Chest radiography was normal. Computerized tomography (CT) of the brain and internal acoustic meatus (IAMs) was reported as normal. The MRI scan was not performed as the patient was severely claustrophobic, subsequent attempts under sedation also failed.

An initial lumbar puncture revealed an elevated protein level of 2.02 g/L, a white cell count of 117, red blood cell count of 7 and an opening pressure of 8 cm of water. No

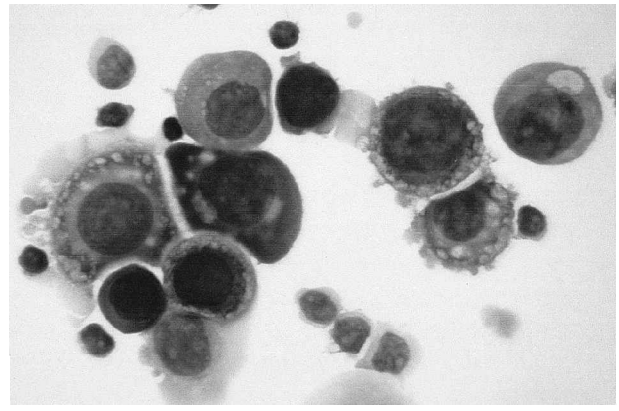

Fig. 1

Clusters and single large malignant cells with a high nucleocytoplasmic ratio and irregular nuclear contours (CSF, MGG; ×400).

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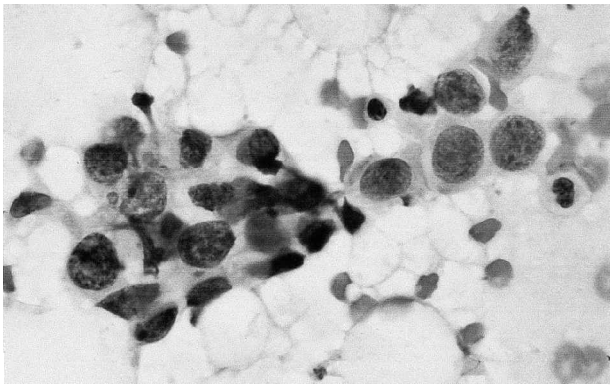

FIG. 2

Three-dimensional group of poorly cohesive epithelial cells with nuclear enlargement, abnormally clumped chromatin and multiple nucleoli (FNA breast, PAP; ×400).

malignant cells or organisms were seen. However, a repeat lumbar puncture revealed malignant cells in the CSF (Figure 1).

On further examination, a mobile 3 cm mass in the left breast associated with axillary lymphadenopathy was discovered. Fine needle aspiration of the mass yielded poorly differentiated malignant cells suggestive of adenocarcinoma (Figure 2). A diagnosis of meningeal carcinomatosis from a breast primary was made.

Dexamethasone and tamoxifen were commenced, but chemotherapy was withheld, due to her rapidly deteriorating condition. She eventually succumbed to her illness 11 weeks after presentation.

Discussion

Infiltration of the meninges by metastatic carcinoma was first described by Eberth in 1870.[3] The seeding of malignant cells in the meninges is an important complication of systemic cancers. Three levels of involvement are recognized within the neuraxis; the cerebral hemispheres, the cranial nerves and the spinal cord and roots. Multiple cranial nerve involvement is well documented, the most frequently affected nerves being the IIIrd, Vth and VIIth cranial nerves,[1] but isolated sudden onset hearing loss as a presenting symptom is unusual. Most cases occur in patients with known malignancies but occasionally the hearing loss can be the first manifestation of meningeal carcinomatosis.[1,4]

Albert and Terrence[5] in a review of five patients presenting with hearing loss as a result of meningeal carcinomatosis summarized the clinical features as: (1) initial unilateral hearing loss with tinnitus, (2) unilateral hearing loss rapidly progressing to severe bilateral involvement, (3) audiological and caloric evidence showing severe impairment of the VIIIth nerve and (4) facial nerve palsy, often seen at time of hearing loss. They found that 10 per cent of patients with meningeal carcinomatosis had VIIIth nerve involvement.

TABLE I

PUBLISHED CASES OF MENINGEAL CARCINOMAS PRESENTING WITH HEARING LOSS

Authors	Year	Age/Sex	Symptoms	CSF	Primary tumour
Oshiro and Pearlman[6]	1965	54/M	Bilateral hearing loss, tinnitus, facial palsy	−	Renal/adenocarcinoma
Hoshino *et al.*[12]	1972	57/F	Bilateral hearing loss, tinnitus, facial palsy	ND	Lung/adenocarcinoma
Alberts and Terrence[5]	1973	74/M	Bilateral hearing loss, bilateral facial palsies, right V nerve palsy	+	Unknown/adenocarcinoma
	1973	50/M	Bilateral hearing loss, tinnitus, left facial palsy, dysphonia	+	Lung/adenocarcinoma
	1973	62/M	Bilateral hearing loss, tinnitus, right facial nerve palsy	+	Unknown
	1974	64/F	Bilateral tinnitus, left-sided hearing loss, occipital headache	+	Unknown
	1976	48/M	Bilateral hearing loss, right tinnitus, right facial palsy, IX, X, XII nerve palsies	+	Lung/undifferentiated
Olson *et al.*[7]	1974	3 Patients	Hearing loss, no other details		
Nomura *et al.*[15]	1974	26/M	Hearing loss, vertigo, facial palsy	ND	Stomach/adenocarcinoma
Katsarkas and Seemayer[14]	1976	44/F	Bilateral hearing loss, facial palsy	+	Uterinecervix/epidermoid carcinoma
Bergstrom *et al.*[15]	1977	52/M	Bilateral hearing loss, vertigo, facial palsy	ND	Bronchus/large cell carcinoma
Miyata *et al.*[16]	1985	56/F	Bilateral hearing loss, vertigo	ND	Lung/adenocarcinoma
Saito *et al.*[17]	1985	55/F	Hearing loss, facial palsy, vertigo	ND	Lung/small cell carcinoma
Ohira *et al.*[18]	1991	25/M	Bilateral hearing loss, vertigo, facial palsy	ND	Pancreas/adenocarcinoma
Houck and Murphy[8]	1992	65/F	Bilateral hearing loss, tinnitus, unsteadiness	+	Breast/undifferentiated
Civantos *et al.*[19]	1992	68/M	Bilateral hearing loss, tinnitus	+	Oesophagus/squamous cell carcinoma
Imamura *et al.*[4]	1997	71/M	Bilateral hearing loss, tinnitus, bilateral facial palsies	+	Renal/transitional cell carcinoma
Shen and Young[20]	1997	57/M	Bilateral hearing loss, tinnitus	+	Lung/adenocarcinoma
Morgan *et al.*[1]	1998	49/M	Bilateral hearing loss, headaches	+	Stomach/adenocarcinoma
	1998	60/F	Right facial palsy, right sided hearing loss	+	Unknown/adenocarcinoma
Present study	2000	61/F	Bilateral hearing loss, bilateral facial nerve palsies	+	Breast/adenocarcinoma

ND = Not described; CSF = CSF cytology; + = positive for malignant cells; − = negative for malignant cells.

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Several pathways for tumour dissemination to the meninges have been suggested; haematogenous metastases to the choroid plexus and thence to the meninges, primary haematogenous spread through meningeal vessels, spread through Batson's venous plexus, retrograde spread of tumour along perineural lymphatics and sheaths, extension along perivascular lymphatics and secondary spread from epidural or parenchymal metastases.[5]

Direct tumour invasion and ischaemia are thought to be the causative factors leading to hearing loss. Oshiro and Perlman[6] described a case of bilateral hearing loss and facial palsy in a patient with renal adenocarcinoma. Tumour infiltrate had filled both internal auditory canals and invaded the facial nerves to the geniculate ganglions. The middle ear and mastoid air systems were free of tumour.

Imamura *et al.*[4] in a review of temporal bone histopathology associated with meningeal carcinomatosis found that, in the nine cases reviewed, eight had tumour invasion of the IAM with destruction of the vestibulocochlear nerve, but the middle ears were free of disease. In the majority of the reviewed cases the otological symptoms preceded the development of facial paralysis. This may be explained by differences in vulnerability between the facial nerve (motor neurone) and vestibulocochlear nerve (sensory neurone) as well as the location of tumour cells in the IAM. In the review of cases (Table I) the vast majority of patients present with bilateral hearing loss, or rapidly progress to a bilateral hearing loss. It is suggested that tumour cells invade the IAM's of both ears synchronously from the CSF thus affecting the VIIth and VIIIth nerves. Of interest is the fact that few patients present with vertigo, although unsteadiness is not uncommon. This could be explained by the simultaneous infiltration and destruction of both vestibular nerves.[4]

The primary tumours responsible for meningeal carcinomatosis are often cited to arise from the lung, breast and from melanomas. Adenocarcinoma is the most frequently identified histological type.[7] The authors found this to be consistent with the reviewed cases (Table I). Most large series,[7] however, cite breast and lung tumours to be the main originators of meningeal carcinomatosis. In our review this was the case for lung tumours but not for breast tumours, in which only one other reported case presenting with bilateral deafness was found.[8] In that case, the patient had a past history of undifferentiated breast carcinoma before presenting with hearing loss. Our case is the first presenting with sudden bilateral hearing loss due to an occult breast tumour.

CSF cytology remains the most important aid to diagnosis in meningeal carcinomatosis, but multiple lumbar punctures may have to be performed before detecting malignant cells. An initial negative result does not rule out the diagnosis, as demonstrated in our case. Olson *et al.*[7] stated that over 90 per cent of their patients required two or more lumbar punctures and only 37 of 50 had positive cytology even after multiple lumbar punctures. In this large series only three patients had hearing loss at presentation, however, no further information was provided.

Other characteristic findings in the CSF include: (1) raised protein levels, (2) mild cellular pleocytosis, predominantly lymphocytic, (3) hypoglycorrhachia and (4) increased opening pressure.[5] However, these findings are not specific and can also be found in conditions such as tuberculosis, sarcoidosis and fungal infections.[9]

Imaging has a questionable role in the diagnosis of meningeal carcinomatosis. CT of the brain and IAMs is usually normal, but is useful in excluding other abnormalities. Gadolinium enhanced magnetic resonance imaging (MRI) is the most sensitive imaging technique available for diagnosis, revealing abnormal leptomeningeal enhancement. Von Campe *et al.*[10] stated in their paper that in 70 per cent of cases the scan was diagnostic.

The treatment of meningeal carcinomatosis is palliative, its treatment cannot be the same as that for intracerebral metastasis since the whole neuraxis is involved. There do not appear to be any well-established regimens. The options are whole brain radiotherapy and/or intrathecal chemotherapy.[8] Adjunctive steroid therapy may help reduce cerebral oedema.

The prognosis is poor, untreated patients generally succumbing within six weeks and treated patients within 12 weeks of diagnosis.[11] Deterioration is due to a number of factors including the natural course of the primary tumour and the multifocal nature of meningeal carcinomatosis. The hearing loss rarely improves even with treatment.

Although rare, sudden onset bilateral deafness may be the first manifestation of an occult malignancy. Diagnosis however can be difficult and a high index of suspicion is required.

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