Clinical Record

Benefits of magnetic resonance image scanning in progressive, bilateral, sensorineural hearing loss: a case of leptomeningeal haemosiderosis

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Abstract
Objective: Magnetic resonance imaging is a routine investigation in cases of asymmetric sensorineural hearing loss, but it is not routinely used to investigate bilateral sensorineural hearing loss.

Method: This case report illustrates the benefits of magnetic resonance image scanning in the latter patient group.

Results: A 53-year-old man with rapidly progressive, symmetrical, bilateral, sensorineural hearing loss was found also to have anosmia, imbalance and incoordination. Magnetic resonance image scanning demonstrated leptomeningeal haemosiderosis. Progressive, bilateral, sensorineural hearing loss is the most common presentation of this condition and magnetic resonance imaging is the diagnostic investigation of choice.

Conclusion: There are potential treatments for leptomeningeal haemosiderosis which prevent further irreversible damage, if a bleeding source can be found. Hearing loss may be due to cochlear or retrocochlear pathology. Cochlear implantation may be indicated.

Key words: Leptomeningeal Hemosiderosis; Superficial Siderosis; Sensorineural Hearing Loss; Magnetic Resonance Imaging

Introduction
Progressive, bilateral, sensorineural hearing loss (SNHL) is not routinely investigated with magnetic resonance imaging (MRI). This case report presents a patient with bilateral SNHL for whom MRI was diagnostic.

Case report
A 53-year-old man with bilateral, symmetrical SNHL had been followed at a rural hospital since early 2000 with serial audiometry (Figure 1). He had begun wearing hearing aids later that year. In April 2004, he had reported worsening tinnitus and rapid progression of his hearing loss, and hearing aids were no longer effective.

On close questioning, the patient reported that his balance had deteriorated over the previous 18 months. Several falls had prevented him from participating in his primary recreational activity, cross-country running. He had intermittent, right-sided, temporoparietal headaches and his sense of smell had deteriorated, but he had no other neurological symptoms.

Examination showed intact and mobile tympanic membranes with apparently normal middle ears. There was no spontaneous nystagmus. The patient fell to both sides during stressed Romberg testing and Unterberger’s stepping testing. Rapid alternating movements were slowed in the hands, right worse than left. Heel–shin coordination was moderately impaired bilaterally. Examination was otherwise unremarkable.

Audiometry confirmed rapid progression of the patient’s bilateral SNHL (Figure 1).

Due to the clinical presentation, a MRI scan was performed (Figures 2 to 4). On initial T2-weighted imaging, a very low signal was noted (particularly around the basal cisterns, brainstem and cerebellum) consistent with moderately pronounced leptomeningeal haemosiderosis. In view of these findings, we obtained axial T1-weighted and coronal gradient T2-weighted MRI sequences and an intracranial magnetic resonance angiography (MRA) scan. No abnormality was seen in either cerebellopontine angle or internal auditory canal. The VIIth and VIIIth cranial nerves were normal, as was the remainder of the central nervous system.

The MRA demonstrated a small (less than 2 mm) side wall aneurysm from the cavernous portion of the left internal carotid artery. No bleeding point was identified.

A neurology opinion was obtained. A cerebral angiogram and MRI scans of the temporal bones and spine were performed but did not pinpoint a bleeding source. The patient was commenced on tranexamic acid.

Further audiological testing showed absent distortion product otoacoustic emissions bilaterally. HINT (Hearing In Noise Testing) sentence testing was undertaken; the results were auditory visual 41 per cent, auditory alone 17 per cent and visual alone 12 per cent.

Cochlear implantation was decided upon, with appropriate counselling of the patient regarding a variable outcome of the procedure. The surgery was uneventful.

When the implant was switched on, the patient was able to distinguish sound and to achieve 25 per cent on HINT
Serial pure tone audiometry.

Axial, T2-weighted magnetic resonance image demonstrating a hypointense rim surrounding the cerebellum (arrow).

Axial, three-dimensional magnetic resonance image demonstrating a hypointense rim surrounding the cerebellum and the right VIIth/VIIIth cranial nerve complex.
There have been just over 100 cases reported in ante-mortem diagnosis was made on surgical biopsy in leptomeningeal haemosiderosis is 0.05 per cent.2 The best estimate to date of the prevalence of symptomatic advent of non-invasive ante-mortem diagnosis with MRI. The literature to date, with more published since the present to the Chicago Neurological Society. The first described in 1908 when an autopsy case report was presented to the Chicago Neurological Society.1 The first hearing loss, and only two of these had cerebellar ataxia.9 Anosmia occurs in at least 17 per cent of cases.1 However, leptomeningeal haemosiderosis may have few symptoms or signs. In a review of 8843 brain MRI scans, Offenbacher et al. showed that only four of the 13 patients with evidence of leptomeningeal haemosiderosis on MRI scanning had hearing loss, and only two of these had cerebellar ataxia and myelopathy.3 The development of symptoms seems to be proportional to the amount of haemosiderin deposition. This supports the theory of a pre-symptomatic phase to the illness. The differential diagnosis of leptomeningeal haemosiderosis includes neurosyphilis, multiple sclerosis, autoimmune hearing loss and allergy.7,17 Rapidly progressive SNHL was the first symptom in our patient. His maximal progression of hearing loss was 13–24 dB per year loss reported by Weekamp et al.3 The differential diagnosis of progressive, bilateral SNHL in adults is shown in Table I. In our patient, the rapidity of progression, combined with neurological symptoms and signs, led to suspicion of intracranial pathlogy. Cerebrospinal fluid xanthochromia is present in approximately three-quarters of samples taken from patients with leptomeningeal haemosiderosis.2 Computed tomography scanning may show a hypodense rim over the brain surface or atrophy of the cerebellar vermis. However, the ‘gold standard’ investigative tool is now MRI. Gradient-echo and T2-weighted images show the haemosiderin as a pathognomonic, hypointense ring around the brainstem, cerebellum, corona, and midbrain.6,7,9,13,15 The VIIIth cranial nerve has a long glial segment, which may predispose it to involvement in leptomeningeal haemosiderosis, compared with other cranial nerves, which have their glial Schwann cell junction close to the brainstem.5,9,16 In contrast, systemic haemochromatosis (due to overloading of red blood cell breakdown products in the bloodstream) results in haemosiderin deposition in the epithelium of the choroid plexus, tuber cinereum and area postrema, but not in the subpial or leptomeninges.4

The most common clinical presentation of leptomeningeal haemosiderosis is a male patient with asymmetrical SNHL, progressing variably to profound deafness.17 Hearing loss is reported in 95 per cent of patients.1 Imbalance is the next most common feature (88 per cent), usually cerebellar ataxia.9 There are case reports of complete vestibular failure.3 Myelopathy, or pyramidal tract signs, such as weakness, spasticity, hyperreflexia and loss of discrete finger–hand movements, occurs in 76 per cent of patients.1 Anosmia occurs in at least 17 per cent of cases.1 However, leptomeningeal haemosiderosis may have few symptoms or signs. In a review of 8843 brain MRI scans, Offenbacher et al. showed that only four of the 13 patients with evidence of leptomeningeal haemosiderosis on MRI scanning had hearing loss, and only two of these had cerebellar ataxia and myelopathy.3 The development of symptoms seems to be proportional to the amount of haemosiderin deposition. This supports the theory of a pre-symptomatic phase to the illness. The differential diagnosis of leptomeningeal haemosiderosis includes neurosyphilis, multiple sclerosis, autoimmune hearing loss and allergy.7

Rapidly progressive SNHL was the first symptom in our patient. His maximal progression of hearing loss was 13–28 dB per year (for 250 Hz to 6 kHz), which compares with the 12–24 dB per year loss reported by Weekamp et al.3 The differential diagnosis of progressive, bilateral SNHL in adults is shown in Table I. In our patient, the rapidity of progression, combined with neurological symptoms and signs, led to suspicion of intracranial pathlogy. Cerebrospinal fluid xanthochromia is present in approximately three-quarters of samples taken from patients with leptomeningeal haemosiderosis.2 Computed tomography scanning may show a hypodense rim over the brain surface or atrophy of the cerebellar vermis. However, the ‘gold standard’ investigative tool is now MRI. Gradient-echo and T2-weighted images show the haemosiderin as a pathognomonic, hypointense ring around the brainstem, cerebellum, midbrain (arrow).
cranial nerves and spinal cord.2,10,17 This is secondary to susceptibility artefact from the haemosiderin, which is most evident on the gradient-echo images. The susceptibility artefact is less pronounced on T2-weighted images and is not usually visible on T1-weighted images.2,3,18,19 As our source of bleeding should be sought. Magnetic resonance imaging of the spine should be performed to exclude tumours, root avulsions and arteriovenous malformations. High resolution cerebral MRA should also be performed to screen for intracranial aneurysms.

Where possible, treatment should be directed at eliminating the bleeding source. The benefit of iron chelation therapy has been debated.1,9,10,20 The anatomical aetiology of hearing loss in leptomeningeal haemosiderosis is controversial and affects potential therapeutic options. Retrocochlear pathology is suggested by delayed brainstem auditory evoked potentials.2,3,18,15 A source of bleeding should be sought. Magnetic resonance imaging of the spine should be performed to exclude tumours, root avulsions and arteriovenous malformations. High resolution cerebral MRA should also be performed to screen for intracranial aneurysms.

When presented with a patient who has progressive, bilateral SNHL, features such as cerebellar ataxia and spinal cord dysfunction should be sought. Leptomeningeal haemosiderosis should be included in the differential diagnosis, and MRI scans of the CNS should be obtained.

Conclusion
When presented with a patient who has progressive, bilateral SNHL, features such as cerebellar ataxia and spinal cord dysfunction should be sought. Leptomeningeal haemosiderosis should be included in the differential diagnosis, and MRI scans of the CNS should be obtained.

- Haemosiderosis is an uncommon disease characterised by the deposition of haemosiderin in body tissues
- When the condition occurs in the central nervous system, it is known as leptomeningeal haemosiderosis or superficial siderosis
- Progressive, bilateral, sensorineural deafness is the most common presentation of this condition, in association with central neurological symptoms and signs
- Early diagnosis with magnetic resonance imaging is important in order to facilitate treatment

<table>
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<tr>
<th>Diagnosis</th>
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<tr>
<td>Presbycusis</td>
<td>Elderly</td>
<td>Audiogram: symmetrical loss of high frequency &amp; speech discrimination</td>
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<td>Noise-induced HL.</td>
<td>Lifelong noise exposure</td>
<td>Audiogram: notch at 4–5 kHz</td>
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<tr>
<td>Autoimmune HL.</td>
<td>Noise exposure</td>
<td>Audiogram: poor speech discrimination</td>
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<td></td>
<td>Tinnitus</td>
<td>ESR</td>
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<td></td>
<td>May have vertigo</td>
<td>ANA</td>
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<td></td>
<td>Primary autoimmune inner-ear disease</td>
<td>Anti-microsomal &amp; thyroglobulin Abs</td>
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<td></td>
<td>Systemic autoimmune disorders (e.g. relapsing polychondritis, Wegener’s granulomatosis, lupus erythematosis, rheumatoid arthritis)</td>
<td>Rheumatoid factor</td>
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<td>May respond to prednisone or methotrexate</td>
<td>Complement C1Q</td>
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<tr>
<td>Ototoxicity</td>
<td>Exposure to ototoxins (e.g. aminoglycosides, chemotherapeutic agents, heavy metals)</td>
<td>Smooth muscle Ab</td>
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<td></td>
<td>Infections (e.g. meningitis, syphilis)</td>
<td>Antigliadin &amp; antiendomysial Abs</td>
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<tr>
<td>Other</td>
<td>Hyperviscosity syndrome (e.g. sickle cell Ca)</td>
<td>Syphilis tests (e.g. VDRL)</td>
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<td>Waldenstrom’s macroglobulinemia</td>
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<td>Kawasaki’s disease</td>
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<td>Leptomeningeal haemosiderosis</td>
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<td>Me´nie`re’s disease/endolymphatic hydrops</td>
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<td>Development &amp; hereditary disorders</td>
<td>Family Hx of non-syndromic hereditary hearing loss</td>
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<td>Family Hx of syndromic hereditary hearing loss</td>
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<td>Family Hx of syndrome + features (e.g. Alports, Usher’s type III, large vestibular aqueduct syndrome, Waardenburg’s syndrome)</td>
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HL = hearing loss; ESR = erythrocyte sedimentation rate; ANA = antinuclear antibody; Abs = antibodies; HLA = human leucocyte antigen; HB = Haemoglobin; VDRL = venereal disease research laboratory test; Ca = carcinoma; MRI = magnetic resonance imaging; Hx = history; CT = computed tomography
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