The Journal of Laryngology & Otology (2009), ***123***, *1266–1270.*

doi:10.1017/S0022215109004551

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

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Accepted for publication: 23 October 2008. First published online 4 February 2009.

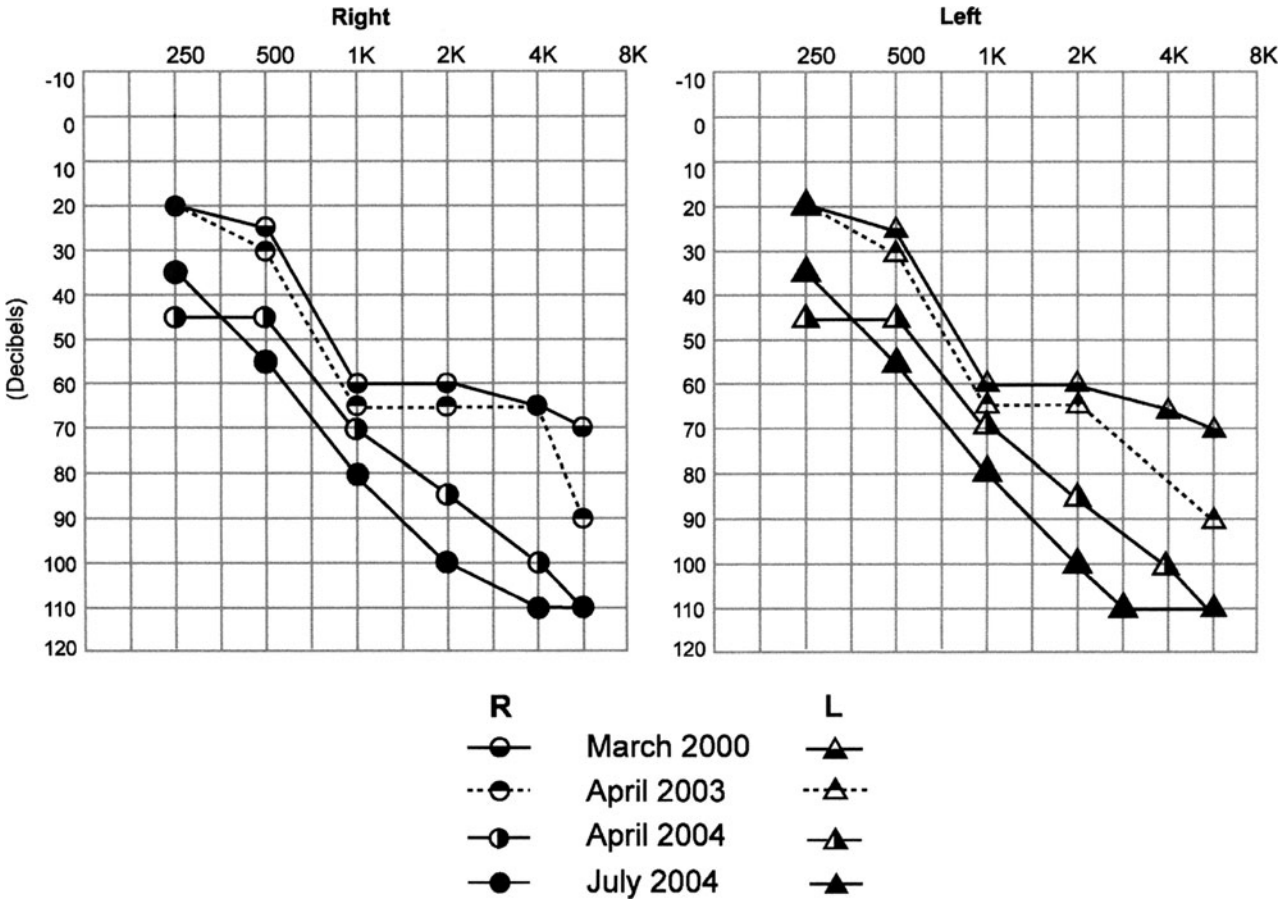

FIG. 1
Serial pure tone audiometry.

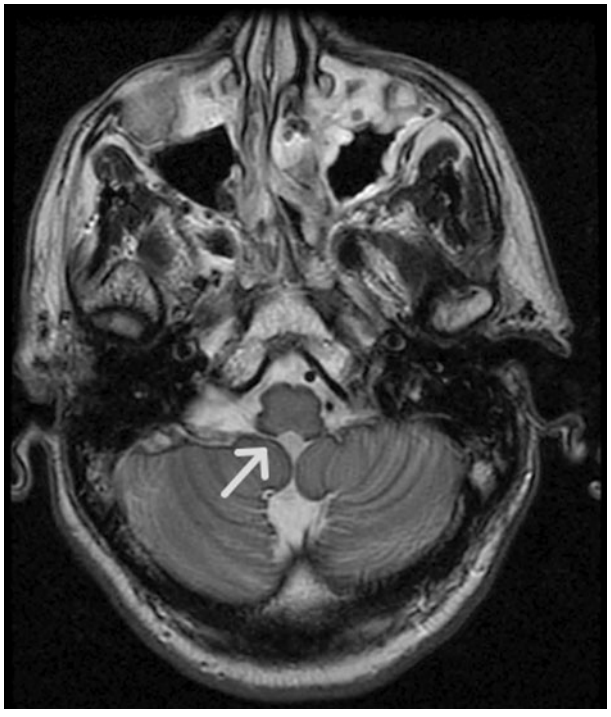

FIG. 2
Axial, T2-weighted magnetic resonance image demonstrating a hypointense rim surrounding the cerebellum (arrow).

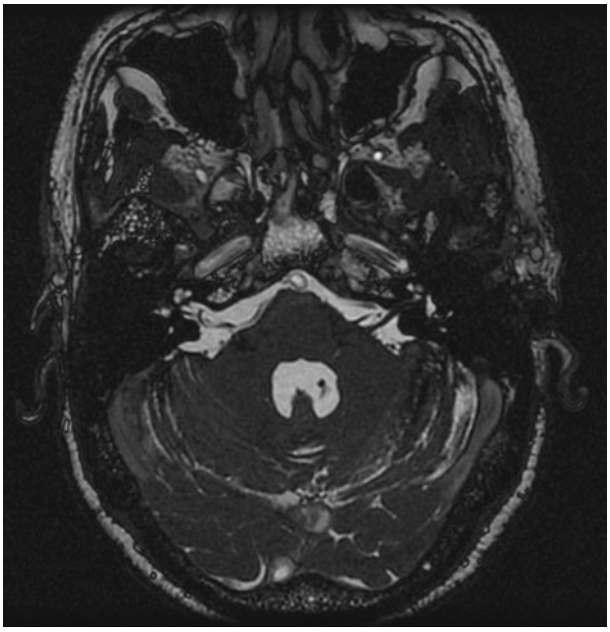

FIG. 3
Axial, three-dimensional magnetic resonance image demonstrating a hypointense rim surrounding the cerebellum and the right VIIth/VIIIth cranial nerve complex.

https://doi.org/10.1017/S0022215109004551 Published online by Cambridge University Press

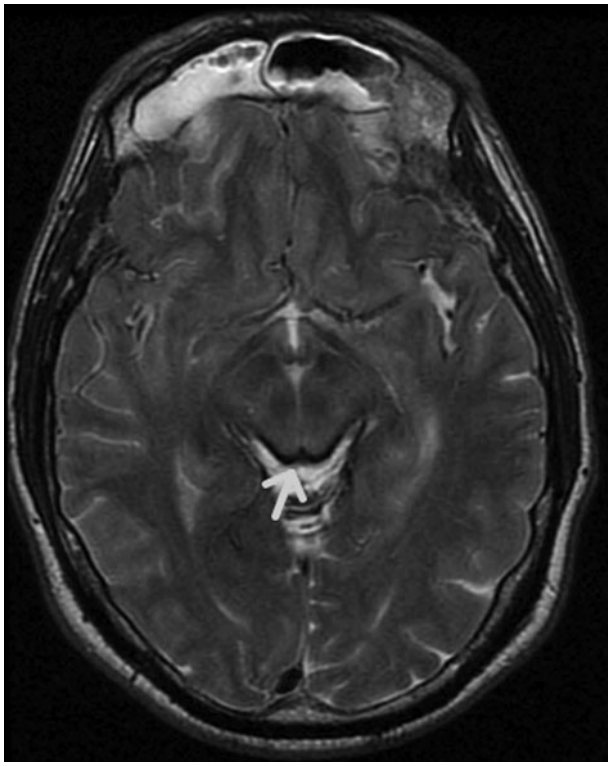

FIG. 4

Coronal, gradient-echo magnetic resonance image demonstrating a hypointense rim on the posterior aspect of the midbrain (arrow).

sentences (audiovisual). This improved to 46 per cent two months later.

Unfortunately, approximately four months after the procedure there was a significant reduction in implant performance. This coincided with a deterioration in the patient's balance and a fall from his bicycle.

At six months post-surgery, he had a further fall from his bicycle, and noticed further hearing deterioration at around this time. For some time, he was unable to detect sound at all. Integrity testing at this time confirmed a functioning implant, and imaging confirmed no change in electrode position. Progressive neural deterioration was assumed to be the aetiology of the hearing loss.

At the patient's most recent follow up, it was possible to map six electrodes at the basal end of the array only, and he was able to use sound as an adjunct to lip-reading.

Discussion

Haemosiderosis is an uncommon disease characterised by the deposition of haemosiderin in body tissues. When it occurs in the central nervous system (CNS), it is known as leptomeningeal haemosiderosis or superficial siderosis of the CNS. Leptomeningeal haemosiderosis was first described in 1908 when an autopsy case report was presented to the Chicago Neurological Society.[1] The first ante-mortem diagnosis was made on surgical biopsy in 1965.[1] There have been just over 100 cases reported in the literature to date, with more published since the advent of non-invasive ante-mortem diagnosis with MRI. The best estimate to date of the prevalence of symptomatic leptomeningeal haemosiderosis is 0.05 per cent.[2]

It is generally held that leptomeningeal haemosiderosis results from recurrent or chronic bleeding into the subarachnoid space.[3–7] This is only symptomatic in 37 per cent of cases.[1] Some authors hypothesise that leptomeningeal haemosiderosis may occur due to an inability to metabolise blood in the cerebrospinal fluid (CSF).[2,8] The condition is potentially treatable when a bleeding point is located; however, approximately half the reported cases had no known bleeding source.[1,4,9,10] The most commonly identified aetiologies are dural pathology, tumours and vascular anomalies.[1,3,9,11–13]

The leptomeninges comprise the pia and arachnoid mater. The pia mater is firmly adherent to the glial capsule of the CNS. In leptomeningeal haemosiderosis, microscopic changes (including both intracellular and extracellular haemosiderin deposition) occur on the surface of CNS structures, with resulting neuronal loss, reactive gliosis and demyelination.[1,4,5,14] Haemosiderin formation occurs mainly within microglia as they synthesise ferritin, so Haemosiderin is taken up selectively by CNS areas rich in microglia and areas close to CSF flow.[9]

This accounts for the particular vulnerability of the leptomeninges, subpial and subependymal tissue of the cerebral hemispheres, cerebellum, brainstem, spinal cord and cranial nerves.[4,6,7,9,14,15] 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.[6,9,16] 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 tissue or leptomeninges.[4]

The most common clinical presentation of leptomeningeal haemosiderosis is a male patient[1] with asymmetrical SNHL, progressing variably to profound deafness.[9,17] Hearing loss is reported in 95 per cent of patients.[1] Imbalance is the next most common feature (88 per cent), usually cerebellar ataxia.[1] There are case reports of complete vestibular failure.[9] 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.[2] 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.[17]

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.*[9]

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 pathology.

Cerebrospinal fluid xanthochromia is present in approximately three-quarters of samples taken from patients with leptomeningeal haemosiderosis.[1] Computed tomography scanning may show a hyperdense 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 rim around the brainstem, cerebellum,

https://doi.org/10.1017/S0022215109004551 Published online by Cambridge University Press

TABLE I

DIFFERENTIAL DIAGNOSIS OF PROGRESSIVE, BILATERAL, SENSORINEURAL HEARING LOSS IN ADULTS

Diagnosis	History	Investigations
Presbycusis	Elderly Lifelong noise exposure	Audiogram: symmetrical loss of high frequency & speech discrimination
Noise-induced HL	Noise exposure Tinnitus	Audiogram: notch at 4–5 kHz
Autoimmune HL	May have vertigo Primary autoimmune inner-ear disease Systemic autoimmune disorders (e.g. relapsing polychondritis, Wegener's granulomatosis, lupus erythematosis, rheumatoid arthritis) May respond to prednisone or methotrexate	Audiogram: poor speech discrimination ESR ANA Antimicrosomal & thyroglobulin Abs Rheumatoid factor Complement C1Q Smooth muscle Ab Antigliadin & antiendomysial Abs HLA-B27 & HbALC Syphilis tests (e.g. VDRL)
Ototoxicity	Exposure to ototoxins (e.g. aminoglycosides, chemotherapeutic agents, heavy metals) Infections (e.g. meningitis, syphilis)	
Other	Hyperviscosity syndrome (e.g. sickle cell Ca) Waldenstrom's macroglobulinaemia Kawasaki's disease Leptomeningeal haemosiderosis Ménière's disease/endolymphatic hydrops	MRI Standard blood profile
Development & hereditary disorders	Family Hx of non-syndromic hereditary hearing loss Family Hx of syndromic hereditary hearing loss + features (e.g. Alports, Usher's type III, large vestibular aqueduct syndrome, ?Waardenburg's syndrome)	CT

HL = hearing loss; ESR = erythrocyte sedimentation rate; ANA = antinuclear antibody; Abs = antibodies; HLA = human leukocyte antigen; HB = Haemoglobin; VDRL = venereal disease research laboratory test; Ca = carcinoma; MRI = magnetic resonance imaging; Hx = history; CT = computed tomography

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] 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.

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.[1,20] Other authors have found evidence of isolated or concurrent cochlear pathology, in the form of increased cochlear microphonic thresholds or absent stapedial reflexes.[1,9] As yet, MRI scanning is not detailed enough to detect localised uptake of haemosiderin by the cochlea. Some argue that involvement of the cochlear aqueduct, alteration in perilymph composition or hypoxia of the cochlea may be present.[9] Symptomatic treatment of hearing loss may be possible through cochlear implantation.[21] However, many patients are so neurologically debilitated at the time of diagnosis that implantation is not appropriate.

Irving and Graham reported significant benefit from cochlear implantation in a case of leptomeningeal haemosiderosis, up to two years following implantation.[22] Further, informal contact with this patient more than 10 years after insertion confirmed that the implant was still functioning.

Although our patient had an encouraging start, his implant performance progressively deteriorated over a six-month period, presumably secondary to ongoing neural deterioration.

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**

https://doi.org/10.1017/S0022215109004551 Published online by Cambridge University Press

References

1 Fearnley JM, Stevens JM, Rudge P. Superficial siderosis of the central nervous system. *Brain* 1995;**118**:1051–66
2 Offenbacher H, Fazekas F, Schmidt R, Alexandra Muthu, D Scott Stevenson, Philip A Bird. Superficial siderosis of the central nervous system: MRI findings and clinical significance. *Neuroradiology* 1996;**38**:551–6
3 Grunshaw ND, Blanshard KS, Hussain SSM, Grace ARH. Superficial siderosis of the central nervous system – diagnosis by magnetic resonance imaging. *Clin Radiol* 1993;**48**:186–8
4 Hughes JT, Oppenheimer DR. Superficial siderosis of the central nervous system: a report on nine cases with autopsies. *Acta Neuropathol (Berl)* 1969;**13**:56–74
5 Koeppen AHW, Barron KD. Superficial siderosis of the central nervous system: a histological, histochemical and chemical study. *J Neuropathol Exp Neurol* 1971;**30**:448–9
6 Koeppem AH, Dentinger MP. Brain haemosiderosis and superficial siderosis of the central nervous system. *J Neuropathol Exp Neurol* 1988;**47**:249–70
7 Koeppen AH, Dickson AC, Chu RD, Thach RE. The pathogenesis of superficial siderosis of the central nervous system. *Ann Neurol* 1993;**34**:646–53
8 Longridge NS. Invited comment. *Otol Neurotol* 2003;**24**: 742
9 Weekamp HH, Huygen PL, Merx JL, Kremer HP, Cremers CW. Longitudinal analysis of hearing loss in a case of hemosiderosis of the central nervous system. *Otol Neurotol* 2003;**24**:738–42
10 Stevens I, Peterson D, Grodd W, Poremba M, Dichgans J. Superficial siderosis of the central nervous system: a 37 year followup of a case and review of the literature. *Eur Arch Psychiatry Clin Neurosci* 1991;**241**:57–60
11 Mascalchi M, Salvi F, Pirini MG, D'Errico A, Ferlini A, Colli F *et al*. Transthyretin amyloidosis and superficial siderosis of the CNS. *Neurology* 1999;**53**:1498–503
12 Ellie E, Camou F, Vital A, Rummens C, Grateau G, Delpech M *et al*. Recurrent subarachnoid haemorrhage associated with a new transthyretin variant (Gly53Glu). *Neurology* 2001;**57**:135–7
13 Li KW, Haroun RI, Clatterbuck RE, Murphy K, Rigamonti D. Superficial siderosis associated with multiple cavernous malformations: report of three cases. *Neurosurgery* 2001;**48**:1147–51
14 Revesz T, Earl CJ, Barnard RO. Superficial siderosis of the central nervous system present with longstanding deafness. *J R Soc Med* 1988;**81**:479–81
15 Anderson NE, Sheffield S, Hope JKA. Superficial siderosis of the central nervous system: a late complication of cerebellar tumours. *Neurology* 1999;**52**:163–9
16 Courville CB. Applied anatomy of the VIIIth nerve and its environs, the cerebellopontine angle. *Laryngoscope* 1932; **42**:415–31
17 Kwartlet JA, De La Cruz A, Lo WWM. Imaging case study of the month: superficial siderosis of the central nervous system. *Ann Otol Rhinol Laryngol* 1991;**100**:249–50
18 Patel MR, Edelman R, Warach S. Detection of hyperacute primary intraparenchymal hemorrhage by magnetic resonance imaging. *Stroke* 1996;**27**:2321–4
19 Wehrli FW. Fast imaging: principles, techniques and clinical applications. In: Atlas SW, ed. *Magnetic Resonance Imaging of the Brain and Spine*, 2nd edn. Philadelphia: Lippincott-Raven, 1996:1413–99
20 River Y, Honigman SW, Gomori JM, Reches A. Superficial hemosiderosis of the central nervous system. *Mov Disord* 1994;**9**:559–62
21 Haferkamp C, Schabet M, Plinkert PK. Cochlear prosthesis implantation in superficial leptomeningeal hemosiderosis [in German]. *HNO* 1999;**47**:642–6
22 Irving RM, Graham JM. Cochlear implantation in superficial siderosis. *J Laryngol Otol* 1996;**110**:1051–3

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Mr S Stevenson takes responsibility for the integrity of the content of the paper.
Competing interests: None declared