Cochlear implantation in superficial siderosis

RICHARD M. IRVING, M.D., F.R.C.S., JOHN M. GRAHAM, F.R.C.S.

Abstract

Superficial siderosis is a rare central nervous system disorder characterized by deafness, ataxia, and pyramidal signs. The hearing loss is believed to be predominantly neural and is usually progressive and bilateral. Careful assessment is therefore necessary to determine the best approach to hearing rehabilitation. A case is presented of superficial siderosis in a young woman who has benefited significantly from cochlear implantation using the Nucleus® device.

Key words: Siderosis; Hearing loss, sensorineural; Cochlear implant

Introduction

Superficial siderosis is a rare, ultimately fatal, central nervous system disorder. It occurs as a consequence of recurrent bleeding into the subarachnoid space. Neural damage results from the subsequent deposition of haemosiderin in the leptomeninges, in the subplial tissues of the brain and spinal cord, and in the cranial nerves (Tomlinson and Walton, 1964).

There is a predominance of males in the reported cases (M:F 3:1), and the age of onset of symptoms varies from 14 to 77 years, with a mean of 44 years (Fearnley *et al.*, 1995). The disease is ultimately fatal in most cases but it may progress slowly over many decades and the mean survival of reported cases is 11 years (Fearnley *et al.*, 1995). Superficial siderosis occurs secondary to repeated minor bleeds and has been reported as a consequence of spinal tumours such as ependymomas, aneurysms, arterio-venous malformations, chronic subdural haematomas, following hemispherectomy, and following traumatic cervical root avulsion (Fishman, 1993). In only 54 per cent of reported cases has a source of bleeding been found (Stevens *et al.*, 1991; Pribitkin *et al.*, 1994; Fearnley *et al.*, 1995).

The posterior cranial fossa, particularly the cerebellum and the eighth cranial nerve, and the spinal cord are most commonly involved. This predilection is thought to result from accelerated conversion of haem to ferritin and ultimately to haemosiderin in the Bergmann glia of the cerebellum and the glial sheath of the eighth nerve (Koeppen *et al.*, 1993). At post-mortem the appearance is of a rust-coloured and fibrotic leptomeninges. Microscopically haemosiderin deposition is seen in association with demyelination, neuraxonal dystrophy, gliosis and astrocytic proliferation.

Cerebellar ataxia and sensorineural hearing loss typically occur in the early stages of the illness. In a comprehensive literature review by Fearnley *et al.* (1995), the symptoms and signs in 63 cases have been collated (Table I). The diagnosis is confirmed by finding xanthochromoic CSF and characteristic magnetic resonance imaging (MRI) features. T2-weighted images reveal a marginal hypo-intensity due to the presence of ferric ions. This surrounds the midbrain, pons, medulla oblongata and spinal cord in the majority of cases (Fearnley *et al.*, 1995). Sadeh and Sandbach (1980) have suggested that the haem-iron complexes that are deposited catalyse lipid peroxidation which leads to breakdown of myelin and neuraxonal dystrophy. Antioxidants such as vitamin E may retard this process and have been proposed as a possible treatment. Treatment with iron chelating agents has also been suggested; however, their usefulness has not yet been established (Fishman, 1993). Treatment at present is frequently unrewarding and is aimed at identifying the bleeding source, and dealing with this appropriately. However in almost half the cases no underlying cause can be found. Hearing rehabilitation in these patients is initially with appropriate amplification. In some patients, however, the hearing loss progresses and an alternative is required. Options include vibrotactile stimulation, cochlear implantation and brainstem implantation.

Case report

A 33-year-old lady was referred for cochlear implant assessment with a three-year history of bilateral progressive hearing loss. Hearing had been lost initially in the left ear, and at the time of assessment she had some function on the right but this could only be improved minimally with amplification. She also complained of imbalance,

TABLE I
THE COMMONEST SYMPTOMS AND SIGNS IN 63 CASES OF SUPERFICIAL SIDEROSIS REPORTED IN THE WORLD LITERATURE UP TO 1995 (FEARNLEY *ET AL.*, 1995)

Sensorineural hearing loss	95%
Cerebellar ataxia	88%
Pyramidal signs	76%
Bladder disturbance	24%
Dementia	24%
Anosmia	17%
Sensory signs	13%

FIG. 1

MRI scan demonstrating a marginal hyopintensity surrounding the brainstem, fourth ventricle, and involving the eighth cranial nerve. The signal from the cochlea is within normal limits.

particularly in the dark, but did not complain of dizziness or vertigo. She had some humming tinnitus that affected her concentration.

A diagnosis of superficial siderosis had been made on the basis of successive xanthochromoic CSF taps and a characteristic MRI appearance (Figure 1). No source of chronic bleeding could be identified despite internal carotid and spinal angiography and myelography.

She is married with a small child and prior to her illness had worked as a hairdresser. Her ability to communicate was reasonable, she had some lip reading ability and occasionally tried finger spelling, but tended to resort to pen and paper in most social circumstances. Her speech production was well preserved.

On examination both tympanic membranes were intact and healthy. She was Romberg negative, had a normal gait, and there was no spontaneous nystagmus. A caloric test with ice water demonstrated no response following left ear stimulation and minimal response following right ear stimulation.

Pure tone audiometry showed a bilateral profound hearing loss. There were no responses in the left ear; on the right ear there was a response to a 250 Hz tone at 70 dB HL but no response to frequencies higher than this. Aided responses on the right dropped from 50 dBA at 250 Hz to more than 100 dBA at 3 kHz. Speech perception using UCL-CUNY sentences revealed no discrimination with her right ear aided. Auditory brainstem response testing at 95 dB showed no repeatable wave form on either side. Computed tomography (CT) scans of the cochleas were normal bilaterally. Visual evoked potentials were normal.

In order to try and determine whether the problem stemmed from the cochlear or the acoustic nerve we attempted to record otoacoustic emissions, but none were detected. Electrocochleography demonstrated no compound action potential in either ear in response to wide band clicks at 100 dB HL.

Promontory stimulation testing on the left resulted in a feeling of vibration at 50 Hz and then pain. On the right a ringing and a buzzing sensation was experienced between 50 Hz and 400 Hz. A dynamic range was demonstrated. Gap detection and temporal difference limen were greater than 250 m sec on the right.

She was implanted with a right-sided Nucleus® multichannel device. At operation all 22 active and five of the inactive electrodes were inserted. She made excellent progress following initial programming and rehabilitation and all the electrodes were working. At nine months her

free field audiogram thresholds were 0.5 kHz-45 dB, 1.0 kHz-55 dB, 2.0 kHz-45 dB and 4 kHz-45 dB. She also achieved CID sentence scores of 66 per cent in the auditory alone test condition.

Throughout the period of rehabilitation her threshold and comfort levels for all electrodes have been repeatedly recorded, and there has been no indication that these are increasing. It is now two years following implantation.

Discussion

Superficial siderosis is extremely rare and in the past may have been underdiagnosed since conventional neuroradiology is almost always normal and prior to the introduction of MRI it was usually diagnosed at post mortem. The features in an elderly patient can mimic senile dementia combined with presbycusis.

Sensorineural hearing loss is the cardinal feature of the condition, it occurs early in the illness and ultimately will affect 95 per cent of patients. The characteristic picture is of a progressive, predominantly high tone hearing loss which initially may be asymmetric. One case has been reported (Kott et al., 1966) of sudden bilateral sensorineural loss, occurring at the time of a subarachnoid haemorrhage.

Vestibular function has only been evaluated in a minority of the reported cases and in all the vestibuloocular reflex is diminished or absent (Neuman, 1948; Revesz et al., 1988; Stevens et al, 1991).

The predilection for the eighth cranial nerve is largely due to its having a long glial segment. The eighth nerve also lies in the cerebellopontine angle, in which it is exposed to both a large pool of CSF and a greater flow of CSF than other cranial and spinal nerves. This may expose the nerve to a larger amount of iron and haem. Macroscopically the proximal eighth nerve appears darkly pigmented and atrophic (Neuman, 1956; Koeppen and Dentinger, 1988). Distal to the glial-schwann cell junction the nerve is unaffected (Koeppen and Dentinger, 1988; Revesz et al., 1988). The mechanism for this protective effect of the Schwann cells is unclear.

Animal studies, inducing superficial siderosis by repeated CSF injections of red blood cells (Koeppen et al., 1993), have elegantly shown the pathological changes in the cerebellum but neither the eighth nerve nor the cochlea have been examined. The effect on the cochlea of repeated minor subarachnoid haemorrhage is unknown, and it is quite possible that this could damage the organ of Corti particularly if the cochlear aqueduct is patent. Cochlear involvement may help to explain why cochlear implantation provided so much benefit in our patient.

Involvement of the brainstem is very common in superficial siderosis, and the CNS parenchyma can be involved for a depth of up to 3 mm. This could result in damage to the cochlear nuclei in the floor of the fourth ventricle. In addition widespread cerebral involvement has been described including destruction of the auditory cortex. Superficial siderosis is therefore a condition that could theoretically involve the auditory pathway at multiple sites.

The initial management of a patient with hearing loss secondary to superficial siderosis is by amplification with hearing aids. Despite the fact that the hearing loss is thought to be predominantly neural, we feel that cochlear implantation should be considered when the patient develops bilateral profound hearing loss and no longer gains a significant benefit from hearing aids. Our patient has certainly gained significant and prolonged benefit. The condition is however progressive and it is possible that any benefit from cochlear stimulation may not last. A

brainstem implant might be indicated if eighth nerve function deteriorates. However this may give little benefit since the disease process also involves the surface of the brain and could possibly involve the cochlear nucleus and temporal lobe cortex in some cases.

References

Fearnley, J. M., Stevens, J. M., Rudge, P. (1995) Superficial siderosis of the central nervous system. *Brain* **118:** 1051–1066.

Fishman, R. A. (1993) Superficial siderosis. *Annals of Neurology* **34:** 635–636.

Koeppen, A. H., Dentinger, M. P. (1988) Brain haemosiderin and superficial siderosis of the central nervous system. *Journal of Neuropathology and Experimental Neurology* **47:** 249–270.

Koeppen, A. H., Dickson, A. C., Chu, R. C., Thach, R. E. (1993) The pathogenesis of superficial siderosis of the central nervous system. *Annals of Neurology* **34:** 646–653.

Kott, E., Bechar, M., Bornstein, B., Askenasy, H. M., Sandbank, U. (1966) Superficial haemosiderosis of the central nervous system. *Acta Neurochirugica* **14:** 287–298.

Neuman, M. A. (1948) Hemochromotosis of the central nervous system. *Journal of Neuropathology and Experimental Neurology* **7:** 19–34.

Neuman, M. A. (1956) Hemochromatic pigmentation of the central nervous system. *Archives of Neurology and Psychiatry* **76:** 355–368.

Pribitkin, E. A., Rondinella, L., Rosenberg, S. I., Yousem, D. M. (1994) Superficial siderosis of the central nervous system: An underdiagnosed cause of sensorineural hearing loss and ataxia. *American Journal of Otology* **15:** 415–418.

Revesz, T., Earl, C. T., Barnard, R. O. (1988) *Superficial siderosis of the central nervous system presenting with longstanding deafness. Journal of the Royal Society of Medicine* **81:** 479–481.

Sadeh, M., Sandbach, U. (1980) Neuraxonal dystrophy and hemosiderin in the central nervous system. *Annals of Neurology* **7:** 286–287.

Stevens, I., Peterson, D., Grodd, W., Poremba, M., Dichgans, J. (1991) Superficial siderosis of the central nervous system. A 37 follow up of a case and review of the literature. *European Archives of Psychiatry and Clinical Neurosciences* **241:** 57–60.

Tomlinson, B. E., Walton, J. N. (1964) Superficial haemosiderosis of the central nervous system. *Journal of Neurology, Neurosurgery and Psychiatry* **27:** 332–339.

Address for correspondence:
Mr Richard M. Irving, M.D., F.R.C.S.,
Department of Otolaryngology – Head and Neck Surgery,
University of California, San Francisco,
350 Parnassus Avenue, Suite 210,
San Francisco, California 94117,
USA.

Fax: 00 1 415 476 0666