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CLINICAL RECORD

Cochlear implantation is a therapeutic option for superficial siderosis patients with sensorineural hearing loss

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Abstract

Background: Superficial siderosis is a rare disease that results from chronic bleeding in the subarachnoid space. Haemosiderin deposits throughout the subpial layers of the brain and spinal cord lead to progressive sensorineural hearing loss, which is seen in 95 per cent of patients with superficial siderosis. The impact of cochlear implantation on the quality of life of superficial siderosis patients is under debate.

Case report: A 38-year-old male with superficial siderosis presented with bilateral progressive sensorineural hearing loss. The patient underwent cochlear implantation and his quality of life was improved as evaluated by the Abbreviated Profile of Hearing Aid Benefit inventory.

Conclusion: The remarkable improvement in Abbreviated Profile of Hearing Aid Benefit scores shown in this study indicates that cochlear implantation leads to a better quality of life in superficial siderosis patients.

Key words: Siderosis; Hearing Loss; Cochlear Implantations; Quality Of Life; Questionnaires

Introduction

Superficial siderosis, which was first reported by Hamill in 1908,[1] is a rare progressive disease of the central nervous system resulting from subarachnoid haemorrhage. Trauma and/or neurosurgery are the major causes of subarachnoid haemorrhage, although almost 50 per cent of superficial siderosis cases have unknown aetiology.[2] The male-to-female ratio of superficial siderosis is 3:1. The age at onset of the symptoms varies from 14 to 77 years, with a mean age at onset of 44 years.[2] Superficial siderosis can cause: sensorineural hearing loss (SNHL), in 95 per cent of patients; cerebellar ataxia, in 88 per cent; pyramidal signs, in 76 per cent; dementia, in 24 per cent; bladder disturbances, in 24 per cent; anosmia, in 17 per cent; and anisocoria, in 10 per cent. In addition, 13 per cent of superficial siderosis patients have sensory signs such as lower limb formication, diminished vibration sense and isolated loss of joint position sense.[3]

Superficial siderosis is caused by haemosiderin deposits on the cerebellar surface, brainstem surface and ventricular margins. They are shown as low-density regions on T2-weighted magnetic resonance imaging (MRI) scans because of the magnetic susceptibility effects of iron.[4,5] In the histopathological study of human temporal bone, hearing loss in a patient with superficial siderosis is most likely correlated with severe degeneration of spiral ganglion cells, despite the presence of remaining hair cells in the middle and apical turns of the cochlea.[6]

Sensorineural hearing loss in superficial siderosis patients is usually treated with hearing aids. However, hearing aids are not effective for patients with severe hearing loss, and cochlear implants may be called for in such patients. Only 31 cases of superficial siderosis treated with cochlear implantation for SNHL have been reported. Even so, there is an ongoing debate regarding the effectiveness of cochlear implantation in superficial siderosis patients.[7] In addition, to the best of our knowledge, there have been no reports evaluating the quality of life in superficial siderosis patients with a cochlear implant. This study assessed the quality of life of a superficial siderosis patient with bilateral progressive SNHL based on a Japanese-language translation of the Abbreviated Profile of Hearing Aid Benefit inventory,[8] and shows the clinical course of cochlear implantation.

Case report

The patient was a 38-year-old male. From his early twenties, he had bilateral progressive SNHL. At age 28 years, he began to use hearing aids in both ears. At age 29 years, the patient presented to a hospital with a chief complaint of bilateral motor disturbances in his extremities, and was subsequently diagnosed with superficial siderosis based on an MRI scan of the head. At age 37 years, the patient experienced an acute exacerbation of hearing loss in his right ear. He visited a hospital and was administered prednisolone, loop diuretic and vitamin B12 for 10 days. However, the hearing loss continued to progress, and an audiometric evaluation showed

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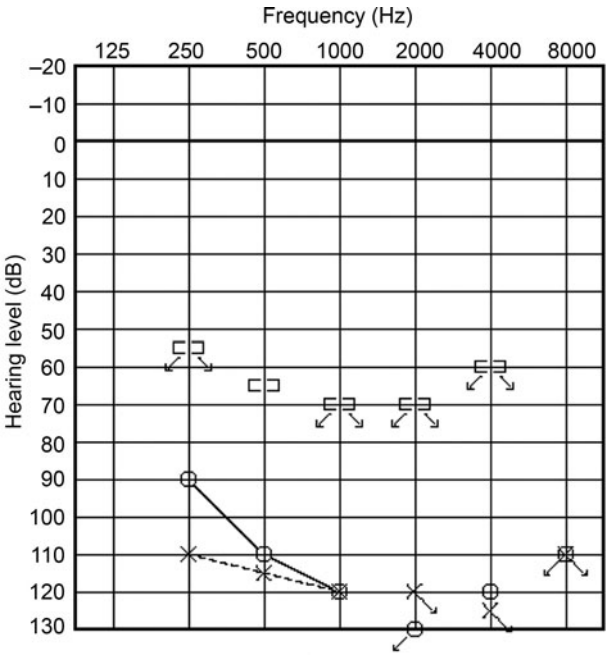

FIG. 1

Pre-operative audiogram. [= bone conduction (masked) right ear;] = bone conduction (masked) left ear; ○ = air conduction (unmasked) right ear; × = air conduction (unmasked) left ear

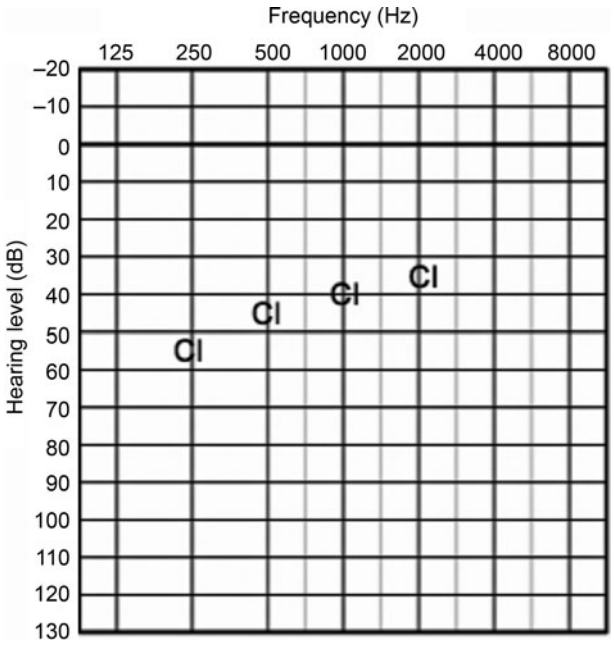

FIG. 2

Sound field threshold with a cochlear implant (CI) three months after surgery.

bilateral, nearly symmetric, severe SNHL. He was referred to our hospital for cochlear implantation.

An audiogram showed bilateral profound SNHL (Figure 1). The accuracy rate in a speech discrimination test using monosyllables was 0 per cent at 80 dB HL. There was no response (wave V disappears) in both ears at 90 dB HL in auditory brainstem response testing. There were no significant findings in computed tomography scans of the bilateral temporal bone. We could find the VIIIth cranial nerve by MRI. He had bilateral motor disturbances in his extremities, but no sensory disturbances. He could walk on crutches for short distances, but usually used a wheelchair. There was no cognitive disability, speech disability or other symptoms.

Cochlear implantation was performed using a Nucleus® CI422, with all active electrodes inserted into the right cochlea through the round window membrane. The patient had a good clinical course, with no complications following the procedure. There was significant improvement in his hearing three months after implantation (Figure 2).

Five months after the cochlear implant surgery, we evaluated the patient's quality of life based on a Japanese-language translation of the Abbreviated Profile of Hearing Aid Benefit inventory. The ease of communication, reverberation and background noise scores without a cochlear implant were 91.0 per cent, 84.8 per cent and 80.8 per cent, respectively. In contrast, the scores with a cochlear implant were 72.8 per cent, 60.7 per cent and 47.7 per cent, respectively. The aversiveness score was 18.5 per cent without a cochlear implant and 33.3 per cent with a cochlear implant (Figure 3).

Discussion

Superficial siderosis is a rare condition characterised by recurrent subarachnoid bleeding. Haemosiderin deposits in the leptomeninges, subpial tissue, spinal cord, brainstem and/or the VIIIth cranial nerve result in eventual demyelination.[9] Common clinical findings of superficial siderosis include SNHL, ataxia, dementia, anosmia and myelopathy; SNHL is one of the more frequent complaints.[3,10] The cause of the recurrent subarachnoid haemorrhages is not known in about 50 per cent of superficial siderosis cases.[3] Superficial siderosis is difficult to manage, and there is no established treatment for the progressive neurological conditions, including SNHL, caused by iron deposition in the central nervous system.

To the best of our knowledge, there have been 31 reported cases of cochlear implantation for superficial siderosis, including 1 bilateral cochlear implant case (Table I).[2,6,7,11–25] The

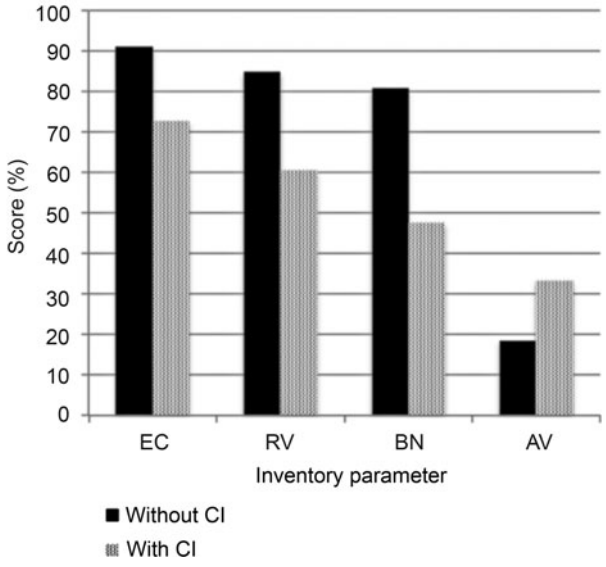

FIG. 3

Abbreviated Profile of Hearing Aid Benefit scores with and without a cochlear implant (CI). EC = ease of communication; RV = reverberation; BN = background noise; AV = aversiveness

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TABLE I
REPORTED CASES OF COCHLEAR IMPLANTATION FOR SENSORINEURAL HEARING LOSS IN SUPERFICIAL SIDEROSIS PATIENTS

Case	Study	Sex, age (y)	Cause	Presenting symptoms	Preserved effect of CI at last exam?	Follow-up duration (mth)
1	Irving & Graham[11]	F, 33	Idiopathic	SNHL, balance issues	Yes	24
2	Takahashi *et al.*[12]	F, 65	Idiopathic	SNHL	No	0.5
3	Dhooge *et al.*[13]	M, 50	Neurosurgery	SNHL, balance issues	No	9
4	Kim *et al.*[14]	M, 25	Head trauma	SNHL, balance issues	Yes	NR
5	Hathaway *et al.*[15] (update by Yoshikawa & Horsh[16])	F, 54	Head trauma	SNHL	No	72 (symptoms worse 5–6 y after implantation)
6	Wood *et al.*[17]	M, 53	Neurosurgery	SNHL, anosmia, balance issues	No	12 (symptoms worse 3–12 mth after implantation)
7	Wood *et al.*[17]	M, 50	Neurosurgery	SNHL, balance issues	No	9
8	Sydlowski *et al.*[18]	NR	NR	NR	No	NR
9	Sydlowski *et al.*[18]	NR	NR	NR	Yes	NR
10	Sydlowski *et al.*[18]	NR	NR	NR	Yes	NR
11	Sydlowski *et al.*[18]	NR	NR	NR	No	NR
12	Sydlowski *et al.*[18]	NR	NR	NR	Yes	NR
13	Bird *et al.*[19]	M	NR	NR	Yes	6
14	Vanat *et al.*[20]	M	Head trauma	NR	No	6
15	Nadol *et al.*[6]	M, 51	Neurosurgery	NR	Yes	6
16	Berrettini *et al.*[21]	M, 68	Head trauma	SNHL, anosmia	Yes	24
17	Berrettini *et al.*[21]	M, 73	Head trauma	SNHL, balance issues	Yes	36
18	Berrettini *et al.*[21]	M, 36	Neurosurgery	SNHL, anosmia	Yes	36
19	Sugimoto *et al.*[2]	F, 65	Neurosurgery	SNHL	Yes	8
20	Grover *et al.*[22]	M, 56	Head trauma	SNHL	Yes	84
21	Grover *et al.*[22]	M, 54	NR	SNHL, ataxia	No	9
22	Bittencourt *et al.*[23]	M, 62	NR	SNHL	Yes	12
23	Bittencourt *et al.*[23]	M, 39	NR	SNHL	Yes	12
24	Nogueira & Meehan[24]	M, 57	NR	SNHL	Yes	24
25	Ryan *et al.*[25]	M, 68	NR	SNHL, balance issues	Yes	11
26	Modest *et al.*[7]	M, 42	Neurosurgery	SNHL, balance issues	Yes	64
27	Modest *et al.*[7]	F, 53	Head trauma	SNHL, ataxia	Yes	11
28	Modest *et al.*[7]	M, 68	Neurosurgery	SNHL, balance issues	Yes	13
29	Modest *et al.*[7]	F, 70	Head trauma	SNHL, balance issues, myelopathy, vestibulopathy	No	18
30	Modest *et al.*[7]	M, 70	Head trauma	SNHL, balance issues, vestibulopathy	No	58
31 L	Modest *et al.*[7]	M, 11	Idiopathic	SNHL	No	3
31 R	Modest *et al.*[7]				Yes	64
32	Current case	M, 38	Idiopathic	SNHL	Yes	8

Y = years; CI = cochlear implant; mth = month; F = female; SNHL = sensorineural hearing loss; M = male; NR = not reported; L = left; R = right

existing literature indicates uncertainty regarding the efficacy of cochlear implantation in superficial siderosis patients who have SNHL. Wood *et al*. reported that two patients with superficial siderosis had limited benefit from cochlear implants one week and six months post-implantation.[17] Previously, retrocochlear diseases affecting the central auditory pathway were considered a contraindication to cochlear implantation. However, a recent study reported on the clinical utility of cochlear implants in patients with superficial siderosis despite central nervous system involvement.[7] Superficial siderosis is considered a retrocochlear disease, but, as is shown in Table I, many cases indicate the positive benefits of cochlear implants. In the present study, cochlear implantation resulted in remarkable improvement of the patient's audiometric threshold, and improvement in quality of life was quantified by his Abbreviated Profile of Hearing Aid Benefit score.

The Abbreviated Profile of Hearing Aid Benefit inventory, a shortened version of the Profile of Hearing Aid Benefit inventory, is a 24-item self-assessment questionnaire in which patients report on their difficulties with communication and/or noise in various everyday situations. Although the Abbreviated Profile of Hearing Aid Benefit inventory was originally used to measure the effect of hearing aids, it is also useful in evaluating cochlear implants.[26] The Abbreviated Profile of Hearing Aid Benefit inventory contains four subscales: ease of communication, reverberation, background noise and aversiveness. Lower ease of communication, reverberation and background noise scores, and a higher aversiveness score indicate a better quality of life.

- **Superficial siderosis is a very rare disease that often leads to progressive sensorineural hearing=loss**
- **A superficial siderosis patient's quality of life was quantitatively evaluated**
- **Cochlear implantation improved their quality of life**
- **Although hearing loss related to superficial siderosis is progressive, cochlear implantation can improve such patients' quality of life**

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Differences in Abbreviated Profile of Hearing Aid Benefit scores for this patient, associated with whether the implant was turned on or off, were checked for significance using the rules developed by Cox and Alexander.[8] The changes (more than 11 per cent) in each score (ease of communication, 91.0 per cent without a cochlear implant, 72.8 per cent with a cochlear implant; reverberation, 84.8 per cent without a cochlear implant, 60.7 per cent with a cochlear implant; background noise, 80.8 per cent without a cochlear implant, 47.7 per cent with a cochlear implant; and aversiveness, 18.5 per cent without a cochlear implant, 33.3 per cent with a cochlear implant) indicated that the cochlear implant made a very positive contribution to his quality of life. He is satisfied with the effect of the cochlear implant, and now enjoys musical concerts. Improvements to his quality of life associated with the cochlear implant could be quantitatively assessed using the Abbreviated Profile of Hearing Aid Benefit inventory. Although some unsuccessful cases of implantation have been reported, both audiological and social benefits can be expected from cochlear implants for superficial siderosis patients with SNHL.[25]

Conclusion

To the best of our knowledge, this is the first report to assess the quality of life of a superficial siderosis patient with profound, bilateral, acquired hearing loss with and without a cochlear implant. Cochlear implantation is a promising therapeutic approach for progressive SNHL in superficial siderosis patients. It is also useful for enhancing quality of life. Wood *et al*. reported that patients with progressive neural deterioration of retrocochlear pathways might lose the benefits of cochlear implantation.[17] Although gliosis, demyelination and neuronal loss in the central nervous system are usually progressive, cochlear implants may play a beneficial role in the management of SNHL in superficial siderosis patients.

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