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Original Article

Cochlear Implantation: A Treatment Approach of Hearing Loss in Superficial Siderosis

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BACKGROUND: Superficial siderosis (SS) is a chronic condition characterized by progressive hemosiderin accumulation in the central nervous system (CNS) due to chronic subarachnoid hemorrhage. Common clinical features include progressive bilateral sensorineural hearing loss (SNHL), cerebellar ataxia and myelopathy. The aim of the study was to analyze the clinical presentation, management, and outcomes of patients affected by SS with bilateral severe-to-profound hearing loss and treated with cochlear implantation (CI), with a particular focus on the course of hearing benefit.

METHODS: A retrospective monocentric review was performed, including patients affected by CNS SS and profound SNHL that underwent CI between January 2012 and December 2021. Outcomes were assessed by comparing pre- and post-operative tonal and vocal hearing thresholds together with verbal perception tests.

RESULTS: A total of 4 patients were identified, with a mean follow-up time of 48.25 months. All patients showed significant improvement of hearing threshold and verbal perception in the first 6 months after surgery (preoperative mean aided PTA of 58.3 dB with a mean open-set speech perception score of 20.3% versus postoperative mean PTA in bimodal stimulation of 41.1 dB and open-set speech perception score of 60.1%). At the last follow-up evaluation, despite progressive deterioration of auditory performances, all patients were users and maintained subjective benefit from CIs.

CONCLUSION: Patients affected by SS who underwent CI showed a significant improvement in auditory and auditory-verbal performances in the first post-operative period. Despite a progressive decline of hearing benefit due to retrocochlear disease progression, CI represents valid support for lip-reading and spatial orientation even in the advanced stages of the disease.

KEYWORDS: Cochlear implantation, sensorineural hearing loss, superficial siderosis

INTRODUCTION

Superficial siderosis (SS) of the central nervous system (CNS) is a rare chronic condition characterized by progressive hemosiderin deposition in the subpial layers of the brain and spinal cord due to chronic or intermittent low-grade extravasation of blood into the subarachnoid space.[1,2] Hemosiderin accumulation occurs over several months, causing cytotoxic damage to the leptomeninges, brain surface, brainstem, cerebellum, cranial nerves, demyelination, and neuronal loss.[3,4] SS is generally idiopathic but can be associated with a history of neurosurgical procedures or CNS tumors and trauma.[3-5]

Before the introduction of magnetic resonance imaging (MRI), SS could only be diagnosed postmortem. Nowadays, thanks to T2-weighted and/or gradient echo sequence MRI, SS can be diagnosed by the presence of a hypointense rim around the brainstem, cerebellum, cranial nerves, and spinal cord (Figure 1).[5,6] Specific subarachnoid bleeding findings can also aid in the diagnosis, such as xanthochromia, elevated blood cell counts, and high protein levels. These signs are not constantly detectable in SS because of the intermittent nature of subarachnoid bleeding.[7]

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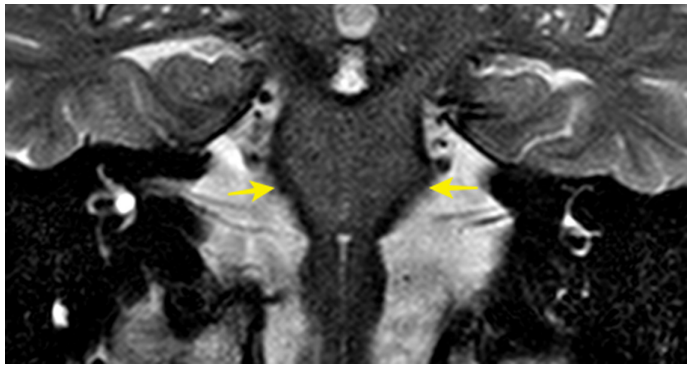

Figure 1. T2-weighted MRI images of patients with SS showing hemosiderin deposition (yellow arrows) along the brainstem at the CPA level in both axial (A) and coronal (B) views.

Clinical presentation is variable, often requiring years before demonstrating a symptomatic onset.[6] The characteristic triad includes progressive bilateral sensorineural hearing loss (SNHL), cerebellar ataxia, and myelopathy. Pyramidal signs, dementia, and anosmia can also occur.[8,9] If the source of hemorrhage can be detected, surgery can be an option, but SS is likely to progress despite surgical procedures when the hemosiderin deposition has already occurred. Nonsurgical approaches include steroids and iron-chelating medications, both with limited success on disease progression.[7]

The first option in auditory rehabilitation of SS-related SNHL is represented by conventional hearing aids, but a progressive loss of their benefit may be expected due to the retrocochlear nature of the disease. Timing and appropriateness of cochlear implantation (CI) are still debated, and less than 50 cases of CI in patients affected by SSHL have been reported.[5] The present study describes auditory rehabilitation with CI in 4 additional cases of SSHL. Disease presentation, clinical course, and serial post-implantation auditory perception scores are analyzed.

METHODS

A retrospective chart review of all the patients diagnosed with SS and severe-to-profound SNHL that underwent CI between 2012 and 2021 at "B. Ramazzini" Hospital of Carpi was performed. Inclusion criteria included a diagnosis of SS based on MRI and clinical signs and a minimum follow-up of 12 months. Each patient underwent a standard preoperative medical, radiological, and audiological CI candidacy evaluation. Data collection included patient demographics, clinical presentation, imaging studies, surgical details, and complications.

All patients underwent pre-operative pure tone audiometry along with speech discrimination testing via the Italian "Common Evaluation Protocol Results in Rehabilitation Audiology." Moreover, pre-operative transient evoked otoacoustic emissions (TEOAE) and auditory brainstem response (ABR) were performed. Outcomes were assessed by comparing pre-operative and post-operative tonal hearing thresholds and verbal perception test averages during the entire follow-up.

Statistical analysis was performed using Jamovi 2.5 statistical software (Computer Software, Sydney, Australia) for Microsoft Windows. For continuous variables, descriptive statistics such as means, median, mode, interquartile range, minimum, maximum, and range were assessed. For categorical variables, absolute frequencies, relative frequencies, cumulative frequencies, and percentages were reported. Postoperative results were analyzed, and a paired samples *t*-test was performed.

The study was performed according to the Declaration of Helsinki. A written informed consent was obtained.

RESULTS

Overall, 4 patients underwent CI over a 9-year period, 3 males and 1 female, with a mean age of 69 years (range 66-72). In all patients, MRI confirmed the presence of SS deposits in the CNS (Figure 1); the etiology of SS was unknown in all cases.

Progressive SNHL was the most common clinical feature (n = 4; 100%), followed by imbalance (n = 3; 75%) and cerebellar ataxia (n = 2; 50%) (Table 1). In 1 case, a mild attention and linguistic deficit (n = 1; 25%) was diagnosed.

All patients were initially fitted with hearing aids and underwent CI evaluation after experiencing a progressive decline in auditory performance. The average time from the first hearing aid to CI was 3 years (range 1-7 years).

Preoperative ipsilateral hearing levels ranged from severe to profound for all 4 ears (mean PTA of 92.5 dB; range 85-103) and open set speech perception scores with bilateral hearing aid ranged from 0 to 29.3% (mean 11.3%). Transient evoked otoacoustic emissions were absent in all cases, while preoperative ABRs of the implanted ears showed no identifiable waves at a stimulation of 100 dB HL. Demographics and clinical data are resumed in Table 1.

Considering speech audiometry outcomes, in each patient, the worst ear was implanted. In case of symmetric hearing loss, the right ear was chosen, as patients were all right-handed. All patients underwent unilateral CI (details in Table 1). All patients had standard transmastoid CI surgery with complete insertion of the electrode array through the round window. Patient number 2 was the object of a previous report, as a tension pneumocephalus developed in the early

MAIN POINTS

- Superficial siderosis (SS) is a chronic condition characterized by progressive hemosiderin accumulation in the central nervous system (CNS).
- Progressive bilateral sensorineural hearing loss (SNHL) is a common clinical feature, and hearing aids can provide partial and short-lasting benefit.
- A retrospective monocentric review, including patients affected by CNS SS and profound SNHL that underwent cochlear implantation (CI), identified 4 cases.
- All patients showed significant improvement of hearing threshold and verbal perception in the first 6 months after surgery. At the last follow-up evaluation, despite progressive deterioration of auditory performances, all patients were users and maintained subjective benefits from cochlear implants.
- Despite the progressive decline of hearing benefit due to retrocochlear disease progression, CI represents a valid support for lip-reading and spatial orientation even in the advanced stages of the disease.

Table 1. Patient Demographics and Clinical Data

Patient Number	Age at Implantation	Gender	Date of CI	Date of First Hearing Aid	Laterality of CI	Type of IC	Presenting Symptoms	Identifiable Cause	Follow-Up Time (Months)
1	68	F	September 13, 2012	2009	Right	Digisonic Neurelec	SNHL, mild ataxia, and imbalance	None	115
2	68	M	October 22, 2015	2013	Right	Digisonic Neurelec	SNHL, imbalance	None	48
3	72	M	February 12, 2020	2019	Left	Oticon Neuro Zti	SNHL	None	18
4	66	M	March 03, 2021	2014	Left	Oticon Neuro Zti	SNHL, mild ataxia, and mild attentional-linguistic deficit	None	12
Mean	68.5								48.25

CI, cochlear implant; SNHL, sensorineural hearing loss.

postoperative period due to repeated nose blowing in the presence of a small bony defect of the tegmen mastoideum; surgical revision was required to close the defect without removing the implant.[10] No other postoperative complications were reported.

All patients initially achieved significant speech recognition in the bimodal condition. However, in patient 2 bimodal stimulation was no longer effective after 12 months and abandoned using the contralateral hearing aid. Preoperative and postoperative audiological data are detailed in Table 2.

Audiological evaluation 6 months after implantation showed that all patients enjoyed a relevant benefit from their implant (paired *t*-test demonstrated significant differences; $P < .01$). In the bimodal stimulation setting, the auditory threshold average was 41.1 dB (range 36.6-46.6 dB) and the average postoperative open set speech perception score was 57.4% (range 37-72.3), compared to the preoperative mean aided PTA of 58.3 dB and preoperative mean open set speech perception score of 20.3%.

Progressive deterioration of auditory performance over time was noticed in all patients. At the last follow-up evaluation (mean 48.25 months, range 12-115), all patients were users and maintained significant subjective benefit from their implant.

DISCUSSION

In most cases, the presenting symptom of SS is SNHL, occurring in the early phases of the disease.[11] A progressive, bilateral high-frequency hearing loss with excessive loss of speech understanding in aided conditions is characteristically reported, with an estimated annual threshold deterioration rate ranging from 7 to 24 dB/year.[12] Both retrocochlear and cochlear damage mechanisms have been implicated in the genesis of hearing loss. The long glial segment along its course in the cerebellopontine cistern favors considerable exposure of the VIII nerve to subarachnoid hemosiderin accumulation and cytotoxic neural damage.[13,14] Moreover, histopathology studies on temporal bones of patients affected by SS showed degeneration of the cochlear epithelium associated with ferritin deposits.[14,15]

Hearing aids remain the first therapeutic choice in patients affected by SS-related hearing loss. Nevertheless, the combination of cochlear and retrocochlear damage leads to partial and short-lasting benefits. Superficial siderosis patients frequently experience sudden worsening of hearing, with an impact on speech perception in aided conditions and consequent struggles in hearing aid fitting and use.

Retrocochlear diseases have traditionally been considered a contraindication to CI; however, indications for CIs have gradually extended to some retrocochlear conditions, i.e., auditory nerve hypoplasia, auditory neuropathy, and acoustic neuroma with an intact cochlear nerve.[16,17]

The first CI in a patient affected by SS has been performed in 1996 by Irving and Graham.[11] Since then, 49 cases have been described in English Literature.[1-4] However, some criticisms remain. The CNS progression of the disease limits the benefit duration of CI. Therefore, during preoperative counseling, it is mandatory to establish realistic expectations regarding auditory performance, predicting a gradual worsening of CI benefit due to SS-related global neurologic decline.[18]

Table 2. Pre- and Postoperative Audiological Data

Preoperative Data		Postoperative Data			
	Mean		Mean 6 Months	Mean 12 Months	Mean at Last Available FU 48.25 months
PTA (right ear) (dB)	90.5 dB	PTA (CI) (dB)	40.5 (range 36.6-46.6)	47 (range 38.3-63.3)	50 (range 38.3-63.3)
PTA (left ear) (dB)	87.1 dB	Bimodal PTA (CI + HA) (dB)	41.1 (range 36.6-46.6)	46.6 (range 36.3-63.3)	45 (range 36.3-58.3)
PTA with HA (dB)	58.3 dB	Speech perception score (CI) (%)	53.8 (range 31.2-73.6)	51.3 (range 33.6-66.8)	45.2 (range 33.6-66.8)
Speech perception score with HA (%)	20.3	Bimodal Speech perception score (CI + HA) (%)	60.1(range 39-72.2)	50 (range 24.4-72.6)	48.1 (range 24.4-72.6)
Open set speech perception score (%)	11.3	Open set speech perception score (CI) (%)	47.2 (range 24.3-77)	41.7 (range 17.3-53.3)	32.3 (range 26.3-53.3)
		Bimodal Open set (CI + HA) (%)	57.4 (range 37-72.3)	50 (range 8.7-71.3)	38.5 (range 8.7-70)

CI, cochlear implant; HA, hearing aid; PTA, pure tone average.

As remarked by Modest et al,[19] accurate preoperative evaluation together with long-term follow-up and HA/CI adjustment are of paramount importance.

It can be difficult to preoperatively identify whether the hearing loss etiology is due to cochlear or retrocochlear damage, and even though this aspect can severely affect hearing outcomes after CI surgery. Specific delayed latencies or abnormal morphology in ABR can help to detect the correct site of the damage.[4] Nevertheless, ABR is likely to show delayed peak I (characteristic of cochlear disease) together with an increased peak I-V range (retrocochlear pathologies), as both damaging components often coexist.[20] In these situations, and in case of profound hearing loss or complete deafness, a trans-tympanic promontory stimulation test may represent a useful tool, yet it lacks normative data.[21] Despite the interest in this technique, promontory test presents several concerns.[22] According to Kileny et al and Kuo et al,[23, 24] auditory responses after trans-tympanic promontory stimulation can be achieved in 89%-92% of patients[23,24] with false-negative results possibly due to the lack of diffusion of the electrical stimulation through the otic capsule.[25] Promontory stimulation test may lack sufficient sensitivity in identifying potentially good performers with cochlear implants and nowadays it is limited to patients with long auditory deprivation.[22]

In all the patients included in the present study, a tailored follow-up and mapping strategy has been adopted. The presence in the team of an expert hearing care professional is crucial, as these patients present rapid adaptation to cochlear maps and may need frequent shifts and stimulation frequency variations due to the retrocochlear component of hearing loss. In the present study, a mean postoperative follow-up of 48.25 months was available (range 12-115 months). All cases showed an evident and immediate benefit from implantation but also experienced a progressive decrease in hearing performance as already described by Tyler et al.[12] Yoshikawa et al[26] described the case of a patient who received significant benefit during 5 years. However, at a 6-year postoperative evaluation, CI performance showed relevant deterioration. Wood et al[8] reported 2 cases with had poor outcomes at 1 week and 6 months post-implantation. In 2021, Artukarslan et al[1] reported a series of 5 patients: 3 of them showed sustained benefit from their CI with a 12 month mean follow-up, 1 patient had no benefit and 1 showed initial improvement with subsequent decline. In the present study population, at the last follow-up evaluation (mean 48.25 months, range 12-115), despite a progressive decrease in hearing performance, all patients maintained subjective benefit from their implant. The patient with the longest follow-up (115 months) presented a global neuro-cognitive deterioration secondary to SS, with a subsequent drop in hearing performance. However, she kept using CI for auditory orientation and to support lip-reading. To the best of our knowledge, this is the longest follow-up period reported in the English literature.

In order to make the present findings stronger and more statistically relevant, further multi-center and expanded patient cohort studies are needed. Future research may focus on the best-tailored CI mapping strategy and neuro-cognitive deterioration rate in the postoperative years, together with its possible deceleration due to the presence of CI support.

CONCLUSION

Cochlear implantation is a reliable strategy for hearing rehabilitation in patients with severe-to-profound SNHL caused by SS. The present study shows satisfying CI outcomes in the very first postoperative period. However, a progressive decline due to CNS deterioration should be expected. For this reason, accurate preoperative counseling is mandatory. Anyway, even after several years, CI seems to preserve useful benefits in auditory spatial orientation and in supporting lip-reading.

Availability of Data and Materials: Data that support the findings of the present study are available on request from the corresponding author.

Ethics Committee Approval: N/A.

Informed Consent: Written informedconsent was obtained from the patients who agreed to take part in the study.

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