Cochlear Implant Patients with Superficial Siderosis

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Abstract

Background: Considered a rare disorder, superficial siderosis of the central nervous system (SSCN) has become more frequently diagnosed in recent years. As it is characterized by progressive sensorineural hearing loss, patients' needs may surpass the capability of hearing aid technology. Despite the retrocochlear nature of the disorder, patients have undergone cochlear implantation (CI) with varying success.

Purpose: To summarize the issues surrounding cochlear implant candidates with SSCN as well as highlight trends in performance postimplantation.

Research Design: Retrospective case reports of seven cochlear implant candidates detail the symptoms, typical audiologic presentation, and array of clinical issues for patients with this progressive and potentially fatal disease.

Results: Despite the retrocochlear component of a hearing loss caused by SSCN, cochlear implantation may be a viable option.

Conclusions: It is essential that the CI audiologist not only be aware of the disorder but also be well versed in the resulting implications for the cochlear implant process. A more thorough case history, an expanded candidacy test battery, and knowledge of the typical presentation of SSCN are critical. The diagnosis of SSCN will impact expectations for success with the cochlear implant, and counseling should be adjusted accordingly.

Key Words: Cochlear implant, hemosiderin, retrocochlear, superficial siderosis

Abbreviations: AN = auditory neuropathy; CI = cochlear implant or cochlear implantation; CID = Central Institute for the Deaf; CNC = consonant/nucleus/consonant; CSF = cerebrospinal fluid; HINT = Hearing in Noise Test; SSCN = superficial siderosis of the central nervous system

Superficial siderosis of the central nervous system (SSCN) is a rare disorder of the central nervous system in which repeated slow hemorrhaging into the subarachnoid space leads to an accumulation of hemosiderin, an insoluble form of iron oxide, throughout the subpial layers of the brain and spinal cord (Koeppen and Dentinger, 1988; Takasaki et al, 2000; Dhooge et al, 2002; Kobayashi et al, 2004). Ultimately, this process leads to gliosis, demyelination, and neuronal loss (Tomlinson and Walton, 1964; Kale et al, 2003; Vibert et al, 2004). The most susceptible cells are those of the auditory-vestibular nerve (95%) and cerebellum (88%) due to their significant glial components and extensive exposure to cerebrospinal fluid (CSF), although widespread cerebral involvement and intracochelear damage are possible (Koeppen and Dentinger, 1988; Fearnley et al, 1995; Kale et al, 2003; Kobayashi et al, 2004). The primary causes of superficial siderosis include dural pathology, tumors, vascular malformations, neurosurgical procedures, and trauma (Tomlinson and Walton, 1964; Dhooge et al, 2002).

The diagnosis of SSCN is complex, as the hemorrhage itself is often so minimal that the exact location...
of hemorrhage cannot be identified (Fearnley et al., 1995). Magnetic resonance imaging (MRI) is the most accurate diagnostic tool available and provides the characteristic finding of a hypo-intense signal in T2-weighted images particularly on the surface of the brainstem and cerebellum (Figure 1), due to deposition of hemosiderin. T2*-weighted MRI and/or gradient echo sequence MRI are especially sensitive for determining the presence of this disorder (Fearnley et al., 1995; Irving and Graham, 1996; Kale et al., 2003; Kobayashi et al., 2004). A supplementary CSF assessment may also confirm the diagnosis. SSCN patients’ CSF may exhibit xanthochromia (yellowish discoloration indicating the presence of bilirubin), increased red blood cells, the presence of iron-containing macrophages, and elevated protein levels (Fearnley et al., 1995; Dhooge et al., 2002), which are characteristic of the disorder.

The length of time from onset of hemorrhaging to onset of symptoms varies, ranging from four months to 37 years (Fearnley et al., 1995; McCarron et al., 2002). Characteristic symptoms (Table 1) develop as neuronal loss and demyelination progress (Kale et al., 2003). Sensorineural hearing loss and gait ataxia are the most prevalent features (Koeppen and Dentinger, 1988; Fearnley et al., 1995; Kale et al., 2003; Kobayashi et al., 2004). As hemorrhaging progresses, so does the severity of the symptoms. In addition to sensorineural hearing loss and gait ataxia, these symptoms may include dementia, anosmia (loss of smell), bladder disturbance, anisocoria (unequal pupil size), and various pyramidal and sensory signs (Fearnley et al., 1995; McCarron et al., 2002), among others. The only opportunity to slow or halt the disease progression is to locate and eliminate the source of the bleeding. However, the source cannot be located in 54 percent of cases, and even in cases where the hemorrhaging ceases, the accumulation of hemosiderin may continue to damage the central nervous system (Fearnley et al., 1995). Survival ranges from 1 to 37 years after the onset of symptoms (Fearnley et al., 1995). Ultimately, the disease may be fatal.

**SUMMARY OF FINDINGS**

A chart review of Mayo Clinic Arizona and Mayo Clinic Rochester patients was conducted. Between 1996 and 2006, 49 patients diagnosed with SSCN were evaluated. Most were initially evaluated by the Neurology Department; only a few came directly to the Audiology Department. Ninety-four percent of those patients reported hearing loss; 53 percent underwent audiologic evaluation at the Mayo Clinic. Of the 24 individuals who were evaluated audiologically, seven (29%) qualified for a cochlear implant using Medicare and/or Food and Drug Administration guidelines. Of the seven potential candidates, five (71%) underwent cochlear implantation.

The seven CI candidates included four females and three males who ranged in age from 42 to 71 years (mean = 57.86). Six of the candidates reported at least
one of the primary causes of SSCN in their comprehensive history: three had been involved in a serious motor vehicle accident, two had undergone a neurosurgical procedure, and one was involved in a motor vehicle accident followed by a neurosurgical procedure. Their symptoms were also consistent with the typical presentation of SSCN patients (Table 1). All of the patients were diagnosed following abnormal MRI. Six of the seven also had lumbar punctures. All of them exhibited elevated CSF protein levels, and half of the spinal taps were xanthochromic. Five patients reported “balance problems,” and four underwent vestibular testing (Table 2). All demonstrated sensorineural hearing loss (Figure 2) and extremely poor word recognition bilaterally. The average recognition score using isophoneme materials (Olson et al, 1996) was 11 percent. Nine of the 14 ears tested scored 0 percent. Because this was a retrospective review, speech testing protocols varied. Isophoneme materials were used and were presented at least 40 dB SL when possible; otherwise they were presented at the patient’s most comfortable level. Despite this variability in presentation, performance was consistently poor.

Postimplantation, speech-recognition results varied (Table 3). For the five current cases, results were generally poor, although one patient did achieve average speech recognition for both sentences and monosyllabic words. A review of the other four cases currently published in the literature revealed more positive results (Irving and Graham, 1996; Dhooge et al, 2002; Hathaway et al, 2006; Kim et al, 2006). Prior to implantation, all nine of these patients exhibited poor speech-recognition abilities. While some showed only limited improvement, all but patient 4 demonstrated at least minimal benefit from use of their cochlear implant.

**DISCUSSION**

Prior to the introduction of MRI scanning, SSCN could not be identified until autopsy (McCarron et al, 2002). With the advent of MRI, many more patients are being diagnosed in the early stages of the disorder (Kale et al, 2003). Understanding this disorder is important for audiologists, surgeons, and other members of any CI team. To date, only four cases of cochlear implantation in SSCN have been presented in the literature (Irving and Graham, 1996; Dhooge et al, 2002; Hathaway et al, 2006; Kim et al, 2006). As more SSCN patients receive cochlear implants, synthesis of presentation and performance data will shed more light on the potential for successful CI in this

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**Table 1. Signs and Symptoms of Superficial Siderosis in the Current and Previous Studies**

<table>
<thead>
<tr>
<th>Sign/Symptom</th>
<th>Percentage of Current Patients Displaying Symptom (N = 7)</th>
<th>Percentage of Patients in the Literature Displaying Symptom</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bilateral hearing</td>
<td></td>
<td></td>
</tr>
<tr>
<td>loss</td>
<td>100</td>
<td>95</td>
</tr>
<tr>
<td>Gait ataxia</td>
<td>86</td>
<td>88</td>
</tr>
<tr>
<td>“Balance problems”</td>
<td>71</td>
<td>Not reported</td>
</tr>
<tr>
<td>Wide-based gait</td>
<td>43</td>
<td>Not reported</td>
</tr>
<tr>
<td>Diplopia</td>
<td>43</td>
<td>Not reported</td>
</tr>
<tr>
<td>Seizures</td>
<td>43</td>
<td>&lt;10</td>
</tr>
<tr>
<td>Falls</td>
<td>43</td>
<td>Not reported</td>
</tr>
<tr>
<td>Spasticity</td>
<td>29</td>
<td>Not reported</td>
</tr>
<tr>
<td>Dysarthria</td>
<td>29</td>
<td>Not reported</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>29</td>
<td>Not reported</td>
</tr>
<tr>
<td>Bladder</td>
<td>29</td>
<td>24</td>
</tr>
<tr>
<td>Incontinence</td>
<td>29</td>
<td>10</td>
</tr>
<tr>
<td>Headaches</td>
<td>29</td>
<td>&lt;10</td>
</tr>
<tr>
<td>Hydrocephalus</td>
<td>14</td>
<td>&lt;10</td>
</tr>
<tr>
<td>Anosmia</td>
<td>14</td>
<td>10</td>
</tr>
<tr>
<td>Pyramidal signs</td>
<td>Not reported</td>
<td>76</td>
</tr>
<tr>
<td>Dementia</td>
<td>Not reported</td>
<td>24</td>
</tr>
<tr>
<td>Sensory signs</td>
<td>Not reported</td>
<td>13</td>
</tr>
<tr>
<td>Anisocoria</td>
<td>Not reported</td>
<td>10</td>
</tr>
</tbody>
</table>

*Note: See Fearnley et al, 1995; McCarron et al, 2002.*

**Table 2. Vestibular Findings in Superficial Siderosis of the Central Nervous System Cochlear Implantation Candidates**

<table>
<thead>
<tr>
<th>Vestibular Finding</th>
<th>Number of Patients (N = 4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abnormal pursuit</td>
<td>2</td>
</tr>
<tr>
<td>Abnormal saccades</td>
<td>0</td>
</tr>
<tr>
<td>Abnormal fixation suppression</td>
<td>0</td>
</tr>
<tr>
<td>Abnormal vestibular evoked myogenic potentials</td>
<td>2</td>
</tr>
<tr>
<td>Unilateral weakness</td>
<td>3</td>
</tr>
<tr>
<td>Bilateral weakness</td>
<td>2</td>
</tr>
<tr>
<td>Central findings</td>
<td>2</td>
</tr>
<tr>
<td>Peripheral findings</td>
<td>3</td>
</tr>
</tbody>
</table>

*Figure 2. Average hearing level (in dB HL) prior to implantation for seven superficial siderosis of the central nervous system cochlear implantation candidates.*
challenging population. The seven cases presented here examine the current best practices for evaluation and follow-up care of cochlear implant patients.

SSCN patients typically present with progressive, sloping sensorineural hearing loss and very poor word recognition much like any other late-deafened adult CI candidate (Irving and Graham, 1996; Takasaki et al, 2000; Dhooge et al, 2002; Vibert et al, 2004; Hathaway et al, 2006; Kim et al, 2006). Based on an audiogram alone, they are indiscernible from a typical cochlear implant candidate. However, it is important to realize that the severity of the hearing loss is primarily due to demyelination of the eighth nerve, although cochlear damage may also play a role. The diagnosis of superficial siderosis critically impacts expectations and performance for a cochlear implant recipient. Due to the retrocochlear nature of the disorder, open-set speech recognition postimplantation is often poor; however, several studies have reported that patients may achieve good open-set speech recognition, scoring approximately 60–75% on sentence materials postimplantation (Irving and Graham, 1996; Hathaway et al, 2006; Kim et al, 2006). Based on an audiogram alone, they are indiscernible from a typical cochlear implant candidate. However, it is important to realize that the severity of the hearing loss is primarily due to demyelination of the eighth nerve, although cochlear damage may also play a role. The diagnosis of superficial siderosis critically impacts expectations and performance for a cochlear implant recipient. Due to the retrocochlear nature of the disorder, open-set speech recognition postimplantation is often poor; however, several studies have reported that patients may achieve good open-set speech recognition, scoring approximately 60–75% on sentence materials postimplantation (Irving and Graham, 1996; Hathaway et al, 2006; Kim et al, 2006). Based on an audiogram alone, they are indiscernible from a typical cochlear implant candidate. However, it is important to realize that the severity of the hearing loss is primarily due to demyelination of the eighth nerve, although cochlear damage may also play a role. The diagnosis of superficial siderosis critically impacts expectations and performance for a cochlear implant recipient. Due to the retrocochlear nature of the disorder, open-set speech recognition postimplantation is often poor; however, several studies have reported that patients may achieve good open-set speech recognition, scoring approximately 60–75% on sentence materials postimplantation (Irving and Graham, 1996; Hathaway et al, 2006; Kim et al, 2006).

Table 3. Summary of Speech-Recognition Abilities Postimplantation for Five Current Superficial Siderosis of the Central Nervous System Cochlear Implantation (SSCN CI) Patients and Four SSCN CI Patients Previously Reported in the Literature

<table>
<thead>
<tr>
<th>Patient</th>
<th>Maximum Postimplantation Auditory-Only Speech Recognition Score</th>
</tr>
</thead>
</table>
| Patient 1 (current case) | 8% consonant/nucleus/consonant (CNC) words  
16% CNC phonemes  
11% Hearing in Noise Test (HINT) sentences  
74% CNC words  
96% Central Institute for the Deaf (CID) sentences |
| Patient 2 (current case) | 48% CNC words  
74% CNC phonemes |
| Patient 3 (current case) | 39% City University of New York sentences  
Not reported; unable to complete speech testing |
| Patient 4 (current case) | 33% Overlearned Speech Randomization Test  
75% Four-choice spondees  
2% HINT sentences |
| Patient 5 (current case) | 66% CID sentences  
Not reported  
71% HINT sentences  
76% unspecified sentence test  
60% unspecified monosyllabic word test  
50% unspecified multisyllabic word test |
| Patient 6 (Irving and Graham, 1996) | 66% CID sentences |
| Patient 7 (Dhooge et al, 2002) | Not reported |
| Patient 8 (Hathaway et al, 2006) | 71% HINT sentences |
| Patient 9 (Kim et al, 2006) | 76% unspecified sentence test  
60% unspecified monosyllabic word test  
50% unspecified multisyllabic word test |

Although retrocochlear involvement may seem to be a contraindication for cochlear implantation, studies have demonstrated the effectiveness of CI in patients with auditory neuropathy (AN), another retrocochlear disorder (Shallop et al, 2001; Katada et al, 2005). The similarity of AN and SSCN symptoms suggests that SSCN patients may benefit from CI as well. Cochlear implantation is a viable option for SSCN patients, although a thorough understanding of both the disease and its audiologic implications is essential (Irving and Graham, 1996; Dhooge et al, 2002; Hathaway et al, 2006; Kim et al, 2006; Sydlowski et al, 2007).

Review of these seven cases highlights several interesting trends in SSCN patients. While many SSCN patients may present for audiologic evaluation or cochlear implant candidacy testing with a diagnosis of SSCN, many may have the disorder and have not yet been diagnosed. Because the onset of symptoms may develop years after onset of hemorrhaging, SSCN is commonly an incidental finding. That reason, combined with the fact that hearing loss is the most common symptom of SSCN, supports the conclusion that members of the cochlear implant team should be aware of the potential for previously undiagnosed neurologic disorders. Hearing loss, coupled with other neurologic symptoms and certain historical features, may indicate a condition such as SSCN.

In addition to the standard audiologic evaluation, the importance of a thorough case history cannot be overemphasized. Every CI evaluation should include a thorough medical and surgical history. It is particularly important to ascertain whether the patient has a history of any of the following:

- Head trauma and/or neurosurgical procedures
- Gait and/or balance concerns
- Bladder disturbances
- Headaches or seizures
- Loss of taste or smell
Additionally, a comprehensive audiologic and vestibular assessment should be conducted. For many centers, the following battery may be an expanded version of the standard protocol:

- Pure-tone air- and bone-conduction audiometry
- Unaided speech reception and recognition
- Acoustic immittance measurements
- Aided speech-recognition testing with appropriate hearing aids
- Otoacoustic emission testing
- Auditory brain stem response testing (if history is significant)
- Videonystagmography or electroneystagmography
- Vestibular evoked myogenic potentials assessment
- Computerized dynamic posturography testing

Finally, if a patient’s history or evaluation is suggestive of SSCN or other neurologic condition, radiological testing should be considered. In the course of a cochlear implant evaluation, a computed tomographic scan is usually completed, but an MRI scan may not be ordered. It is important to remember that following cochlear implantation, MRI is no longer an option for the patient without magnet removal. Because MRI is the only way to diagnose SSCN, the opportunity for radiological evaluation prior to surgery should not be overlooked.

Further research is needed to determine whether there may be a situation that would automatically preclude the recommendation of cochlear implantation for superficial siderosis patients. Additionally, future research should investigate optimal mapping strategies for these individuals.

**CONCLUSION**

Despite the retrocochlear component of a hearing loss caused by SSCN, cochlear implantation is a viable option. Therefore, it is essential that the CI audiologist not only be aware of the disorder but also be well versed in the resulting implications for the cochlear implant process. A more thorough case history, an expanded candidacy test battery, and knowledge of the typical presentation of SSCN are critical. The diagnosis of SSCN will impact expectations for success with the cochlear implant, and counseling should be adjusted accordingly.

**REFERENCES**


