Literature Review

SSCN mimics presbycusis and its results other retrocochlear disorders can be difficult using audiologic assessment alone. T2*-weighted gradient echo sequences on MRI remains the standard diagnostic tool. Disorders with identified otologic consequences include multiple sclerosis (MS), vestibular schwannoma (VS), and auditory neuropathy (ANSD). Pathologic mechanisms result in characteristic findings on audiologic assessment, imaging, and electro physiology of the auditory nerve system (SSCN) in an exorbitant disorder with devastating audiologic and neurologic consequences. The overall understanding of SSCN has advanced in recent years.

Major symptoms of SSCN include hearing loss, tinnitus, balance disorders, and facial or extremity weakness. These symptoms may result from subtle brainstem auditory conduction defects. In the current study, we offer new insights into three aspects of SSCN: (1) audiologic clinical presentation, (2) site of lesion, and (3) directions for future research.

SSCN remains a paucity of literature on its audiologic, vestibular, and neurologic consequences. Understanding the pathologic mechanism of superficial siderosis is unique and produces a phenotype that results in characteristic audiologic and neurologic presentations.

Otologic symptoms may be misidentified. The current study highlights the variability of clinical presentation in SSCN, but also offers what may be characteristic findings that could help to differentiate SSCN from other retrocochlear disorders. Understanding the pathologic mechanisms associated with SSCN can help in the development of prognosis and treatment options for patients with this disorder.

SSCN is an underrecognized retrocochlear disorder with paucity of audiologic vestibular consequences. There are few reports on the clinical presentation of SSCN. Previous reports have demonstrated that individuals may have cochlear involvement as shown by absent DPOAEs (100%), extra-axial lesions as shown by elevated or delayed ABR latencies, and increased or absent acoustic reflexes and delayed ABR latencies may also contribute to identification of inner- and extra-axial lesions.

The literature describes SSCN as a disorder with a long pre-symptomatic phase that usually presents with sensorineural hearing loss. Clinical presentations include hearing loss, tinnitus, vertigo/myokymia, and dysarthria. The current study highlights characteristic findings that could help in the differentiation of SSCN from other retrocochlear disorders.

Future research needs to evaluate the long-term impact of neurologic complications on the patient's otologic symptoms. Other studies could further investigate the role of MRI in diagnosis. The current study highlights characteristic findings that could help in the differentiation of SSCN from other retrocochlear disorders. Understanding the pathologic mechanisms associated with SSCN can help in the development of prognosis and treatment options for patients with this disorder.

Table 1: Characteristic audiologic findings based on site of lesion

<table>
<thead>
<tr>
<th>Site of Lesion</th>
<th>Characteristics</th>
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<tbody>
<tr>
<td>Vestibular schwannoma</td>
<td>Hearing loss, tinnitus, vertigo/myokymia, dysarthria</td>
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<tr>
<td>Auditory neuropathy</td>
<td>Hearing loss, tinnitus, vertigo/myokymia, dysarthria</td>
</tr>
<tr>
<td>Multiple sclerosis</td>
<td>Hearing loss, tinnitus, vertigo/myokymia, dysarthria</td>
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Research on SSCN is limited to mostly retrospective case studies with small sample sizes. Audiologic involvement has not been systematically investigated, but findings suggest that SSCN should be considered in the differential diagnosis of sensorineural hearing loss. Type and degree of hearing loss resulting from SSCN exceeds that which may be expected due to age or gender (Figure 1), although it may range from mild to profound in severity (Figure 2). The literature describes SSCN as a disorder with a long pre-symptomatic phase that usually presents with sensorineural hearing loss. Clinical presentations include hearing loss, tinnitus, vertigo/myokymia, and dysarthria. The current study highlights characteristic findings that could help in the differentiation of SSCN from other retrocochlear disorders. Understanding the pathologic mechanisms associated with SSCN can help in the development of prognosis and treatment options for patients with this disorder.

Methods

Two English-speaking adults (18 years or older) identified by SSCN by a physician and with MRI results. That supported this diagnosis were recruited for the study. Participants included: a case history, transtympanic imaging (MRI), and audiologic battery including pure-tone and speech discrimination. Otologic, vestibular, and neurologic symptoms were recorded.

Results

SSCN causes bilateral, asymmetric, progressive, sloping SNHL that is the primary clinical sign, the classic identifying triad includes gait disturbance, loss of balance, and myokymia. The current study highlights characteristic findings that could help in the differentiation of SSCN from other retrocochlear disorders. Understanding the pathologic mechanisms associated with SSCN can help in the development of prognosis and treatment options for patients with this disorder.