Case Report

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Monosymptomatic Manifestation of Clinically Isolated Syndrome with Sudden Sensory Neural Hearing Loss

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ABSTRACT

Clinical presentation of demyelinating disorders is very heterogeneous, but initially isolated cranial nerve involvement, especially eighth nerve lesions, is extremely rare as the manifestation of clinically isolated syndrome (CIS) or multiple sclerosis (MS), so sudden sensory neural hearing loss (SNHL) is rarely a sole and presenting symptom of MS. A 31-year-old female presented with a history of left-sided ear fullness sensation and sudden hearing loss (without any history of trauma or infection); the pure-tone audiometry (PTA) showed unilateral SNHL pattern (high-frequency zone). The neurological and otolaryngologic examination was normal. Magnetic resonance imaging of the brain revealed several disseminated typical demyelinating plaques. The oligoclonal bands had been detected in the sample of cerebrospinal fluid. All of the routine laboratory data and serum autoantibodies were within normal ranges. Administration of high-dose corticosteroid improved her hearing. SNHL is a rare and atypical manifestation of CIS. Our case is assigned to be a high-risk CIS and may be developed to MS. It may be due to acute inflammatory demyelinating lesions of the cochlear nerve which could be a manifestation of CIS at first.

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Introduction

Multiple sclerosis (MS) should be considered in the differential diagnosis of patients who present with isolated cranial nerve palsies. Clinicians should have a high index of suspicion when evaluating such patients particularly in low prevalence regions close to the equator. Early recognition and treatment of such a “clinically isolated syndrome” (CIS) may prevent early relapse (1). Sudden sensorineural hearing loss (SSNHL) during MS is a well-recognized

clinical manifestation (2). In one study in 2008, isolated cranial nerve involvement in MS was present in 10.4% of 483 patients, either as a presenting symptom (7.3%) or a symptom of disease relapse (3.1%), and the frequency of eighth nerve involvement, as a sole initial symptom, was almost 0% (1). High-resolution fast spin echo screening technique, used in conjunction with appropriate clinical prescreening and referral, can provide an equally sensitive method of evaluating unilateral SNHL compared to gadolinium-enhanced T1 magnetic resonance imaging (MRI) while reducing costs and providing distinct advantages in evaluating nonacoustic schwannoma causes of SNHL. Here, we present a case of CIS presenting with SSNHL which needs to be further discussed regarding auditory pathway involvement in MS.

Case Report

A 31-year-old female presented with a history of left-sided ear fullness sensation and sudden hearing loss. Her symptoms began abruptly when the patient first noticed tinnitus in her left ear upon awakening. She also experienced a distorted perception of sound, difficulty in following conversations, and sensitivity to loud sounds. These symptoms persisted throughout the day recurrently, and over the next few days, his tinnitus progressively worsened in intensity and became persistent. As she experienced sudden hearing loss (without any history of trauma or infection), pure-tone audiometry (PTA) was done and unilateral SNHL pattern (especially in the high-frequency zone) was observed and all the otology causes had been ruled out. The tympanogram was normal, and the acoustic reflex was absent in the left ear (Figure 1).

All other neurological examination was normal. Subsequently, the patient was referred to a neurologist for further evaluation. At 1 month from the onset of symptoms, a brain MRI was performed and it revealed several typical demyelinating plaques disseminated in different regions.

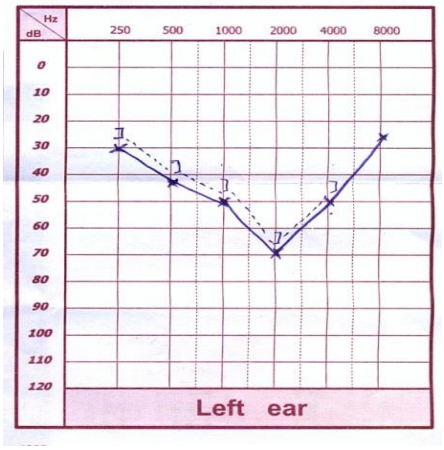

Figure 1. The pure-tone audiometry of the left ear showed sensory neural hearing loss in high-frequency zone

No lesions were detected along the auditory pathways and brainstem (Figure 2).

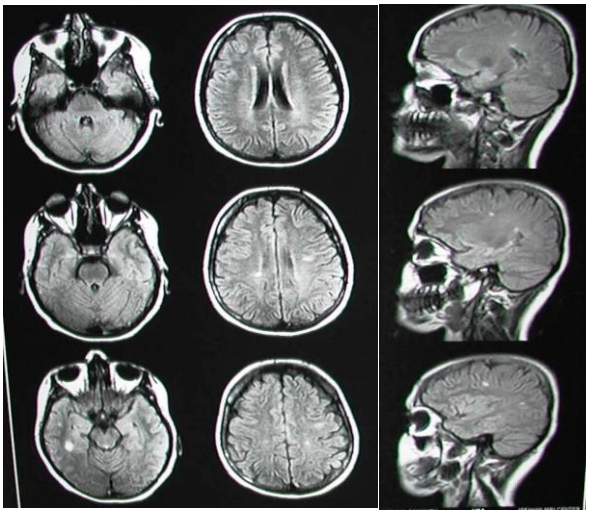

Figure 2. The brain magnetic resonance imaging revealed multiple disseminated oval-shaped demyelinating plaques in periventricular, temporal lobe and corpus callosum area

The patient was treated with a 5-day course of intravenous methylprednisolone (1 g/day), with partial improvement in hearing. High-dose corticosteroid improved her hearing but not recovered completely which had been confirmed by control PTA. The patient reported some residual tinnitus and hearing loss. In her medical history, she also described

episodes of dystonic cramps and causalgia in her legs, in the years preceding this attack. Neurological examination was normal. All of the routine laboratory data were normal. Autoantibodies of the serum were within normal ranges and autoimmune disorders or vasculitis had been ruled out. A sample of cerebrospinal fluid (CSF) obtained to measure the oligoclonal bands (OCBs) which had been detected and immunoglobulin G (IgG) index. IgG index was marginally elevated (0.65). All tests in serum and CSF were negative for herpes simplex virus 1-2, cytomegalovirus, Epstein–Barr, Coxsackie, hepatitis B and C, and also *Borrelia burgdorferi*. CSF cell count, protein, and glucose were normal. Treatment was intermittently continued with methylprednisolone 48 mg/day followed by tapering for the next 6 months. Clinical imaging findings were consistent with CIS at high risk of conversion to MS. A comprehensive neuropsychological assessment was also performed, covering multiple cognitive domains. The assessment showed impaired performance (≥ -1.5 standard deviation from normative data) in the tests of verbal episodic memory-immediate prose recall and rate of verbal learning and executive functioning. Her performance on the tests of attention and information processing speed, backward digit recall, retention and delayed recall of verbal and visual material, verbal fluency, visuoperceptual and constructional dexterities, verbal and visual reasoning, and concept formation remained within the average and high-average range, consistent with premorbid estimates. The patient was reevaluated and the neurological examination at that time showed increased tendon reflexes of both lower extremities, with left predominance, but no clear Babinski sign. Furthermore, he complained of generalized fatigue. The patient was followed up with brain MRI at 6 months, 1 year, and 2 years from the attack. MRI at 1 year included imaging of the cervical spinal cord with no demyelinating lesions and also brain diffusion tensor imaging with minor disturbances. New lesions were not detected and MRI scan of the brain after 1 year from the attack showed mild improvement of some lesions, including the sole lesion in the corpus callosum. Two years after the hearing loss attack, her hearing sense improved to a great extent but not completely. Follow-up neuropsychological assessment revealed a significant improvement in previously mentioned impairments, with the patient's scores in all the other neuropsychological tests remaining within the expected range of performance.

Discussion

MS can affect behavioral audiometric measures in many ways. Since the disease is characterized by multiple demyelinating lesions that occur throughout the white matter of the brainstem, the patterns of audiometric abnormality can vary noticeably among patients (3). In any case, the mean frequency of CIS/SSNHL in older and newer studies is < 1% of cases (2). The clinical course of hearing loss in CIS/SSNHL seems to be benign, with almost all patients fully recovering auditory function within a period of a few months. 62.5% of CIS patients, for whom the data are available, developed MS in the 1st year after their episode (4). Approximately 30% of patients with CIS and abnormal MRI will develop MS over the next year (5). It seems that presentation of CIS as SSNHL may be associated with increased risk of conversion to MS. This is consistent with evidence supporting that the presence of brainstem lesions at initial presentation confers a significantly increased risk of conversion to MS, as well as of increased future disability (6).

Most patients with CIS/SSNHL had concomitant tinnitus (86%) as did patients with definite MS/SSNHL (65%). In one study from all CIS patients with available MRI data, 50% of them had brainstem lesions, to which their SSNHL could be attributed (50%). There is a discrepancy here, which has been discussed by other authors (7). In our patient, no brainstem demyelinating lesion was

detected, but brain MRI was performed after a month of SSNHL initiation and after intratympanic dexamethasone administration. It is possible that a punctuate lesion along the auditory pathway may have disappeared in this time frame as has been described in other CIS/SSNHL cases (8). Although there are reports of lesions within the cochlear nerve itself, most of the series with available MRI imaging highlight eighth cranial nerve root-entry zone demyelination (2).

Neuropsychological studies in our patient showed impaired specific components of working memory. It is well known that working memory and attention are the cognitive domains that are specifically impaired early in the course of MS and are often related to corpus callosum lesions or atrophy (4).

As conclusion, this report describes a case of SSNHL in the context of MRI of the brain findings consistent with a demyelinating etiology. SSNHL is a rare and possibly under recognized manifestation of CIS. It may be due to acute inflammatory demyelinating lesions of the cochlear nerve (cochlear neuritis) which could be a manifestation of CIS at first.

Conflict of Interests

Authors have no conflict of interests.

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