Original Article

Neuro-otologic Manifestations of Multiple Sclerosis

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Abstract:

Background: Multiple sclerosis (MS) has numerous neurologic signs and symptoms, including hearing impairment. The reports concerning the type and severity of hearing loss in MS patients vary and little is known about an association of the disease characteristics with changes in hearing status.

Objective: To find the otologic and neuro-otologic manifestations of MS with the use of routine hearing assessment tools.

Methods: The presence of neuro-otologic signs and symptoms were studied in 30 patients diagnosed with MS. Patients underwent pure tone audiometry, speech audiometry and acoustic brain stem response (ABR) tests. The results were compared with 30 healthy age and sex matched controls.

Results: The most common finding was sensory-neural hearing loss followed by dizziness and nystagmus. The shape of the audiogram and severity of hearing loss was associated with both chronicity and disease activity. There were abnormal latencies in ABR waves which were more significant with high velocity stimulus.

Conclusion: Hearing status deserves careful attention in MS patients. It may be used as a diagnostic means or an indicator for evaluating the characteristics of the underlying disease.

Keywords: audiometry - auditory brain-stem response - multiple sclerosis - neuro-otologic signs and symptoms

Introduction

Multiple sclerosis (MS) is a chronic disease characterized by multiple areas of demyelination, inflammation, and glial scarring in the central nervous system. MS is the most common chronic and usually progressive neurologic disease whose clinical course varies from a benign and symptom-free disease to a rapidly progressive and disabling disorder.[1,2]

MS is more common in women. It usually occurs in the third and fourth decades and is uncommon before ten years of age.[2]

From 4% to 10% of patients diagnosed with MS develop sensory-neural hearing loss (SNHL). Hearing loss can be progressive or sudden in onset and can be bilateral, unilateral, symmetric, or asymmetric.[2,3] Often, MS patients do not complain about their hearing defect. Several studies have revealed that a mild decrease in acuity to pure tones is present in the majority of MS patients but no consistent pattern has been identified. Antonelli has found that although high frequency loss was the most common pattern in MS patients; however, the domed audiogram pattern, which is uncommon in the hearing impaired population, was the most distinctive feature.[4]

Reports on the amount and type of pure tone hearing loss associated with MS also vary considerably. Traditional speech testing, the determination of the speech reception threshold (SRT), and the discrimination score (SDS) have been considered to reveal only a few abnormalities in MS.[5,6] Grénman, in his study on MS patients, was unable to find any statistical significant differences in traditional speech tests administered to MS patients and a control group.[7]

Abnormal results for stapedial reflex thresholds elicited by pure tone stimuli have been reported in 13% to 69% of MS patients.[8] Bilaterally absent acoustic reflexes have been found in 20% of MS patients.[9]

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An abnormality of the acoustic brain-stem response (ABR) is assumed to be a diagnostic criterion for MS. Patterns of abnormality are variable and include prolonged latency of waves, absence or poor morphology of waveforms, and waveform abnormalities with an increased stimulus presentation rate.[9–11] It seems that 30 to 75% of all MS patients have an ABR abnormality.[12] Protti reported that in MS patients, the most distinctive feature of the ABR wave forms was poor morphology, particularly in waves III and V.[10] In the Chiappa studies on MS patients, the majority of ABR abnormalities were observed in wave V amplitude and the next frequent abnormality was an increased wave III-V interpeak latency.[13–15]

Materials and Methods

We observed 30 MS patients who were referred to Loghman Hakim Hospital, Shaheed Beheshti Medical University between April 2006 and May 2008. Patients all had a definitive diagnosis of MS by history, clinical examination, and paraclinics that included electrophysiological studies and magnetic resonance imaging (MRI). The exclusion criteria were a positive history of hearing loss, abnormal otoscopic examination, and recent history of medications with known otologic complications.

Patients were asked about the history and presence of hearing loss, vertigo, dizziness, tinnitus, and facial muscle activities in addition to neurologic and otologic examinations that were administered by a single examiner.

Pure tone audiometry (PTA), speech audiometry which included speech recognition threshold (SRT), the Speech Discrimination Score (SDS), and ultimately Acoustic Brain Stem Response (ABR) were administered. The same tests were given to 30 age and sex matched healthy controls. None of the controls had a significant past medical history that included neuro-otologic and hearing problems.

The ABR test was performed by a Madsen 2250 system in a standard acoustic environment. The stimulant was an 85 dB click and presented to the patient via an ear phone. Presentation rates of 10, 30, 50, and 70 clicks per minute were used. Silver chloride surface electrodes were used for picking up the electro-neurologic signals.

The time delay of the initiation of waves I, III, and V as well as I-III, I-V and III-V interpeak intervals were measured and a delay of more than 2.5 SD was considered abnormal. The amplitude of wave V was considered abnormal when the proportion of V/I was below 1. The results were analyzed by independent *t*-test using SPSS 15.0 software.

Results

There were 30 patients (15 males and 15 females) whose ages were between 17 to 45 years with a mean of 30.8±7.4 years enrolled in this study. Patients all exhibited different disease characteristics. In 12 patients (40%) the course of MS was progressive, and 18 patients (60%) had a relapsing course. The mean length of disease was 35.6 (1 – 120) months in this group. The disease was in remission in 22 patients (73.3%) and active in 8 (26.6%).

The most common neuro-otologic findings in the MS patients were mild hearing loss (23.3%) and dizziness (63.3%). True vertigo was present in 6.6% of the patients. No cases of facial nerve dysfunction were found (Table 1).

Table 1 . Neuro-otologic signs and symptoms in MS patients

Signs and Symptoms	Count	%
Dizziness	19	63.3
Vertigo	6	20
Nystagmus	9	30
Hearing loss	27	90
Tinnitus	3	10
Facial nerve disorders	0	0

The average pure tone thresholds in frequency ranges from 250 to 8000 Hz were compared to those obtained from the control group. The results are shown in Table 2.

Sensorineural hearing loss was seen in 27 patients (90%) from which 3 (10%) had mixed hearing loss. No cases of pure conductive hearing loss were diagnosed. The mean time lag between MS diagnosis and initiation of hearing loss was 4.2±2.7 months. The results showed a statistically significant hearing loss, particularly in high frequencies in the MS group.

Table 2. The means of pure tone thresholds obtained in the three frequency ranges in both case and control groups

Frequency Range (Hz)	Case (dB)	Control (dB)	*P*-value
250	15.1±5.4	8.3±4.8	<0.001
500, 1000, 2000	9.7±4.2	7.6±3.5	0.035
4000, 8000	16.2±6.5	9.1±3.6	<0.001

The association between the elapsed time after initiation of MS symptoms and hearing thresholds in different frequencies as well as the association between disease activity and hearing status are noted in Tables 3 and 4, respectively.

Table 3. Mean hearing thresholds in low, middle, and high frequencies in different times after MS diagnosis

Time (months)	Count	%	250 Hz	500 – 2000 Hz	4000 – 8000 Hz
<2	3	10.0	13.2±5.3	8.2±4.7	10.4±4.9
2 to 6	6	20.0	14.7± 6.4	8.6±5.8	12.8±5.8
6 to 12	8	26.6	15.2±5.2	9.6±3.5	14.8±5.3
12 to 24	9	30.0	17.1± 6.1	10.3±6.2	18.4±6.3
>24	4	13.3	17.4±5.9	10.8±5.7	22.5±6.8

Table 4. Association between activity of disease and hearing status

Disease Activity	Count	%	250 Hz	500 – 2000 Hz	4000 – 8000 Hz
Active	8	26.6	16.7±5.2	10.4±3.4	16.9±5.8
In remission	22	73.3	14.4±4.5	9.1±4.1	16.0±7.9
P-value	—	—	<0.001	<0.001	0.002

The mean Speech Discrimination Scores in cases and controls were 79.45±19.4 and 92.2±6.4, respectively. The results of the ABR test are shown in Table 4.
A 10-clicks-per-second stimulus was applied and we noticed that 24 patients (80%) had an abnormality in latency or amplitudes of the waves. In more rapid stimulants, the abnormal results increased to 83%.

Table 5. Means of absolute and relative latencies of ABR waves by 10 and 70 clicks per second in cases and controls

ABR Mode	Intervals	Case (mS)	Control (mS)	*P* Value
10 clicks/s	Absolute wave I latency	2.232±0.022	2.261±0.072	0.001
	Absolute wave III latency	4.331±0.023	4.281±0.010	<0.001
	Absolute wave V latency	6.448±.0059	6.292±.0260	<0.001
	IPL I-III	2.099±0.098	2.020±0.121	<0.001
	IPL III-V	2.117±0.106	2.011±0.083	<0.001
	IPL I-V	4.216±0.122	4.031±0.102	<0.001
70 clicks/s	Absolute wave I latency	2.234±0.031	2.263±0.024	<0.001
	Absolute wave III latency	4.331±0.017	4.281±0.019	<0.001
	Absolute wave V latency	6.461±0.041	6.295±0.031	<0.001
	IPL I-III	2.113±0.023	2.019±0.014	<0.001
	IPL III-V	2.114±0.142	2.013±0.067	<0.001
	IPL I-V	4.227±0.143	4.032±0.095	<0.001

The most common abnormal patterns were simultaneous abnormalities of amplitude and latency (58.3%) followed by an abnormality in amplitude alone (29.1%) and eventually abnormality in latency, alone (12.5%).

Discussion

With regard to the results of other studies that reported different percentages of hearing loss in the different frequency ranges, our audiologic findings in MS patients confirmed the presence of threshold abnormalities in all frequency ranges. Our findings showed that the dome shaped audiogram was the most common pattern of hearing loss in this group of patients. This was in accordance with the results of numerous previous studies. However, it was in contrast with the findings of other authors5 who have shown low tone loss to be the most common pattern of hearing loss in MS patients. Considering the association between the chronicity of MS and hearing loss characteristics, this study showed that patients in the earlier stages of disease had mild low frequency hearing loss which progressed to a more obvious hearing impairment that involved all frequency ranges, particularly higher frequencies. This resulted in a dome shaped audiogram in patients diagnosed with MS for greater than two years.

There were statistically significant higher pure tone thresholds in patients with active disease. It seems that hearing loss is dependent on disease activity. There were only 8 patients with active disease in our study, therefore, confirmation of this hypothesis and evaluation of the role of audiometry in estimating treatment competency needs further studies with larger sample sizes.

Cochlear neuritis as well as involvement of more distal neural pathways of hearing is a proposed mechanism of hearing loss in MS patients.16 Relatively low speech discrimination scores in the patients confirmed this hypothesis, however, this is in contrast with the findings of other investigators5–7 who have stated that MS does not significantly alter speech audiometry results. In the patients, hearing loss was a dormant and progressive process. This is also in contrast with other studies17,18 that noted sudden sensory-neural hearing loss in the course of MS.

The ABR test, in the patients, showed a significant abnormality when compared with normal controls. It is well known that ABR has an obvious role in the diagnostic evaluation of MS. Our results suggested a significant abnormality in auditory pathways that affect the shape and delay time of ABR waves. The measured values of ABR in our study were similar with the results of other investigators10–12 and this study also demonstrated a concept regarding relative improvement in diagnostic abilities of ABR by the increase in presentation rate of the imported signals from 10 clicks/s to 70 clicks/s.

Conclusion

The results of otologic and neuro-otologic tests including PTA, SDS, and ABR can be used as complementary tools to confirm the diagnosis of MS. This study shows that the severity and shape of hearing loss is associated with disease chronicity. Furthermore, audiological evaluation of MS patients may be useful for estimation of treatment competency. Electro-physiological studies of hearing pathways are also useful in the diagnosis and monitoring of MS; especially when ABR is performed with a higher click presentation rate protocol.

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