Research Article

Single-fiber EMG in Meniere's Disease
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Summary

Objective: To assess neuromuscular transmission (NMT) in patients with Meniere's disease (MD), in which the precise etiology is unknown and a channelopathy hypothesis has been proposed.

Methods: Single fiber electromyography (SFEMG) during voluntary contraction of the extensor digitorum communis muscle, nerve conduction studies of upper and lower extremities and concentric needle electromyography of the extensor digitorum communis were performed on 20 patients (13 female, 7 male) with MD and 12 age-matched normal controls (4 female, 8 male). Mean jitter value was calculated for each subject and was compared with the values recorded from normal controls.

Results: The mean jitter values of the MD group were not different from the mean jitter values of normal control subjects.

Conclusion: Even though our findings are not consistent with neuromuscular conduction defect, this doesn’t exclude presence of channelopathy in the pathogenesis of MD.

Key words: Meniere's disease, neuromuscular transmission, channelopathy

INTRODUCTION

Meniere's disease (MD) is a disease of the inner ear characterized by spontaneous attacks of vertigo, sensorineural hearing loss, aural fullness and tinnitus. The American Academy of Otolaryngology-Head and Neck Surgery has published guidelines for making a diagnosis of MD[12]. Distortion of the membranous labyrinth, characterized by endolymphatic hydrops, is thought to be the pathologic...
basis of MD\(^{(16)}\). However, the precise etiology is unknown. Anatomic, genetic, immunologic, infectious, metabolic and vascular causes have been proposed\(^{(8,10,13,17)}\). Patients with MD have been reported to have an increased prevalence of migraine during life\(^{(15)}\). A common vascular mechanism for migraine and MD has been suggested\(^{(14)}\). Another hypothesis is their both being channelopathies\(^{(7,14)}\).

Channelopathies have been identified as the cause of various paroxysmal disorders such as hypokalemic paralysis, episodic ataxias or familial hemiplegic migraine. Like many of the channelopathies MD is a paroxysmal disorder. Dysfunction of neuromuscular transmission (NMT) detected by single fiber electromyography (SFEMG) has been reported in migraine\(^{(1-3)}\). In this study, we investigated NMT in patients with MD which is known to be commonly associated with migraine and which shares several features of other paroxysmal disorders, some being channelopathies.

**MATERIAL AND METHODS**

The study was conducted in the specialized neurootology and electrophysiology clinics in Ege University Medical School Department of Neurology, İzmir, Turkey. The study protocol was approved by the local ethic committee. Informed consent was obtained from all the participants. 20 patients (13 women and 7 men; aged 33 to 74 years; mean age 53.9 years, SD, 14.4 years) diagnosed as definite MD according to the criteria of American Academy of Otolaryngology-Head and Neck Surgery and 12 age-matched healthy normal subjects (4 women, 8 men; aged 46 to 61 years; mean age, 54 years; SD, 4.5 years) were included in the study.

**Electrophysiological Study:**

SFEMG was performed during voluntary contraction of the extensor digitorum communis (EDC) muscle, and a concentric needle electrode (Medtronic DCF-25, diameter 0.30 mm, ref: 9013S0011) was used for the SFEMG recording. None of the patients or control subjects used any drug within 3 days of SFEMG examination. A Medelec synergy electromyograph (Medelec synergy Oxford Old Woking, Surrey, U.K.) was used for recording, saving data, and analysis. Low-cut filter was set at 500 Hz, and high-cut filter was set at 10 kHz. Only potentials with a stable shape, a rise time of less than 0.3 ms, and amplitude of more than 150 µV were accepted for jitter analysis. For each jitter analysis, 60 to 100 consecutive traces were recorded. Eleven different potential pairs were recorded from each subject, and jitters of these potential pairs were calculated. All subjects were required to have normal motor nerve conduction on routine screening tests of radial, median, posterior tibial and peroneal nerves and normal sensory nerve conduction of sural nerves for inclusion in the study. The concentric needle EMG of the EDC muscle was also performed. During the entire electrophysiological examinations, the skin temperature was maintained above 31ºC in the upper extremity and above 30ºC in the lower extremity. Mean value of consecutive differences (MCD) was used as the jitter value.

**RESULTS**

Both controls and patients with MD had normal nerve conduction studies. Concentric needle EMG studies of the EDC muscle were also normal. Mean jitter values of the MD group did not differ from the normal control subjects (25.4 ± 3.4 ms versus 27.2 ± 2.6 ms, respectively; p=0.06) (Table 1).
Table 1: Age, average number of fibers studied and mean value of consecutive differences (MCD) used as the jitter value of the normal control subjects and patients with Meniere’s disease.

<table>
<thead>
<tr>
<th></th>
<th>Meniere’s Disease (Mean±SD)</th>
<th>Normal control (Mean±SD)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n=20</td>
<td>n=12</td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td>53,9 ± 14,4</td>
<td>54,0 ± 4,5</td>
<td></td>
</tr>
<tr>
<td>Average number of fibers studied</td>
<td>11,2 ± 2,4</td>
<td>9,8 ± 0,4</td>
<td></td>
</tr>
<tr>
<td>MCD (μs)</td>
<td>25,4 ± 3,4</td>
<td>27,2 ± 2,6</td>
<td>0,06</td>
</tr>
</tbody>
</table>

**DISCUSSION**

SFEMG is the most sensitive neurophysiological method of assessing the neuromuscular junction in humans. NMT abnormalities have been detected in migraineurs with or without aura\(^1\-\(^3\)). Recently same abnormalities have been reported in patients with cluster headache\(^4\,5\). To explain this subclinical NMT abnormality in migraineurs it was thought that some migraineurs share similar genetic abnormalities with FHM patients and have abnormalities of P/Q-type Ca\(^{2+}\) channels which are present in motor nerve endings and in the brain\(^1\(^1\). However, SFEMG performed in patients with FHM revealed that NMT is normal in FHM\(^1\(^8\). It was postulated that mechanisms involving other proteins or channels can be the cause of subclinical NMT abnormality in migraineurs\(^4\).

The importance of genetic factors in MD has been suggested on many occasions. Autosomal dominant nonsyndromic progressive sensorineural hearing loss associated with vestibular dysfunction having clinical features very similar to MD has been mapped to the DFNA9 locus on chromosome 14, the COCH gene; probably encoding an extracellular matrix protein\(^6\). This raised the possibility of a COCH mutation in patients with MD. On the other hand, sensory region of the membranous labyrinth, inner hair cells have at least two distinct K+ channels in their basolateral membrane and they also express calcium channels. Outer hair cells have at least three types of K+ channels. In the cochlea the hair cell channel is KCNQ4. This is a voltage gated potassium channel expressed prominently in outer hair cells and mutations result in non-syndromic autosomal dominant progressive hearing loss\(^9\). Mutations in two other genes encoding K+ channel subunits, KCNQ1 and KCNE1, have been found in syndromic hereditary deafness\(^9\). These data strongly suggest that MD has genetic bases and either accumulation of insoluble deposits formed by COCH mutations or channel disorders are the underlying etiologies. Clinical features such as sensitivity to dietary ions and reduction of the vertigo attacks with acetazolamide are on behalf of the channelopathy hypothesis. We wanted to examine the NMT in patients with MD. A subclinical NMT deficit detected could strengthen our suspicions about a channel disorder as found in some patients with migraine. However, no such deficit could be revealed. This doesn't mean that normal NMT excludes the channelopathy.
hypothesis as no transmission deficit has been found in patients with FHM. Genetic studies are the only way of confirming the channelopathy hypothesis.

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REFERENCES


