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Original Article

Association between Family History and Idiopathic Sudden Sensorineural Hearing Loss

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OBJECTIVE: Sudden sensorineural hearing loss (SSNHL) is defined as hearing loss of at least 30 dB occurring within three days over at least three contiguous frequencies. The etiology of SSNHL cannot always be precisely determined; in such cases, this condition is termed idiopathic SSNHL (ISSNHL). This unique study investigates the relationship between ISSNHL and positive family history for ISSNHL.

MATERIALS and METHODS: In total, 125 patients diagnosed with ISSNHL were retrospectively reviewed. The presence of ISSNHL in the family medical history and degree of kinship of family members diagnosed with ISSNHL were determined. For univariate analysis, a chi-squared test and/or Fisher's exact test was used for between-group comparisons of qualitative variables; a t-test was used for quantitative variables. Significant variables in the univariate analysis were introduced into stepwise logistic regression for multivariate analysis. $p<0.05$ indicated statistical significance.

RESULTS: Fifty-nine (47.2%) patients were male and 66 (52.8%) were female. Statistical analysis revealed no significant difference between sex and development of ISSNHL ($p=0.04$). Forty-two (33.6%) patients had a family medical history of ISSNHL, whereas 83 (66.4%) did not. A statistically significant association between the development of ISSNHL and a family history of ISSNHL was observed ($p<0.05$)

CONCLUSION: Our study supports an association between ISSNHL and genetic predisposition. Proving genetic susceptibility to ISSNHL will lead to improvements in the prediction, early diagnosis, and treatment of this disease.

KEYWORDS: Hearing loss, idiopathic sudden sensorineural hearing loss, family history, gender

INTRODUCTION

Sudden sensorineural hearing loss (SSNHL) is classically defined as hearing loss of at least 30 dB occurring within three days over at least three contiguous frequencies.[1] It is etiologically related to infection, trauma, immunological disease, toxic substances, use of ototoxic drugs, vascular disorders, neurological disorders, acoustic tumors, electrolyte disturbances, endolymphatic hydrops, and other conditions. [2-11] However, the etiology of SSNHL cannot always be precisely determined; in such cases, this condition is termed idiopathic SSNHL (ISSNHL). Because of this, ISSNHL should be considered a diagnosis of exclusion.

Estimates of the incidence of ISSNHL range from 5 to 20 cases per 100,000 people; it is a common otologic disease [12, 13]. In most cases, ISSNHL presents unilaterally; however, bilateral involvement may also be seen in rare cases. Hearing loss can be progressive, fluctuant, or steady. The male-to-female distribution appears to be equal [14]. The most common clinical presentation involves vertigo, tinnitus, a sensation of aural fullness, and vestibular symptoms [15].

The diagnosis of ISSNHL is made by the analysis of the detailed history, physical examination, blood tests, and radiological and audiological tests to exclude a number of possible etiologies [15, 16].

The treatment of sudden hearing loss is based on the underlying etiology. No strong evidence exists regarding the efficacy of any treatment options for patients with ISSNHL because of the uncertain etiology of this condition [17]. Management strategies may vary, but steroid administration is currently the most widely accepted treatment method.[17, 18] Bed rest, salt-free diet, and smoking and alcohol cessation can be added to steroid treatment.

There are some factors that affect the prognosis of ISSNHL. Negative prognostic factors include an initial hearing loss of >90 dB, a decline in the mid-frequencies of the audiogram, age of <15 or >60 years, an elevated erythrocyte sedimentation rate, accom-

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panying vertigo, and bilateral involvement [18-23]. On the other hand, tinnitus is an indicator of cochlear reserve and a good prognostic factor [24-25].

In the present study, we aimed to investigate whether there is any relationship between positive family history and ISSNHL.

MATERIALS and METHODS

The study was approved by the Ethical Committee of Marmara University Faculty of Medicine (ID number: 09.2014.0266, Issue date: 12.18.2014). In total, 125 patients who presented to Marmara University Hospital from 2010 to 2013 and who were diagnosed with ISSNHL were retrospectively reviewed. Informed consent was obtained from all the patients. The patients were classified according to age and sex. The presence of ISSNHL in the family medical history and the degree of kinship of family members diagnosed with ISSNHL were determined. The presence of ISSNHL in a family member was considered to be a positive family medical history. Family members were grouped as first- and second-degree relatives.

First-degree family members were mothers, fathers, sisters, brothers, daughters, and sons. Second-degree family members were maternal grandfathers and grandmothers, paternal grandfathers and grandmothers, maternal aunts and uncles, paternal aunts and uncles, and cousins. Statistical analysis of the obtained data was performed using SPSS 16 Software (SPSS Inc.; Chicago, IL, USA). For univariate analysis, a chi-squared test and/or Fisher's exact test was used for between-group comparisons of qualitative variables; a t-test was used for quantitative variables. Significant variables in the univariate analysis were introduced into stepwise logistic regression for multivariate analysis. P-values less than 0.05 indicated statistical significance.

RESULTS

Dextran, acyclovir, and steroid treatments (loading dose, 250 mg intravenously; maintenance dose was obtained by tapering at 1 mg/kg/day) were administered to all the 125 retrospectively diagnosed patients with ISSNHL. In total, 59 (47.2%) patients were male and 66 (52.8%) were female. Statistical analysis revealed no significant difference between sex and development of ISSNHL (p=0.04). The mean age of the patients was 45 years [min-max, 5-82; standard deviation (SD), 18.825; standard error mean (SEM), 1.584].

The statistical analysis also revealed a positive correlation between ISSNHL and an age of ≥ 45 years (p=0.04). SHL was accompanied by vestibular symptoms in 12 patients (9.6%). Involvement of the right ear, left ear, and both ears was observed in 64 (51.2%), 60 (48.0%), and 1 (0.8%) patient, respectively. Thus, ISSNHL affected both ears at the same rate.

Two patients (1.6%) had a history of previous ISSNHL. The mother of eight patients (6.4%), father of six (4.8%), sister of six (4.8%), brother of nine (7.2%), maternal grandfather/grandmother of three (2.4%), paternal grandfather/grandmother of three (2.4%), maternal aunt/uncle of two (1.6%), paternal aunt/uncle of three (2.4%), cousin of one (0.8%), and both mother and sister of one (0.8%) had a history of ISSNHL. A total of 29 patients (23.22%) with a positive family medical history had first-degree relatives diagnosed with ISSNHL, and 12 (9.6%) patients had second-degree relatives diagnosed with ISSNHL.

In total, 42 (33.6%) patients had a family medical history of ISSNHL, whereas 83 (66.4%) did not. A statistically significant association between the development of ISSNHL and a family history of ISSNHL was observed (p<0.05).

DISCUSSION

The incidence of ISSNHL is estimated to be 5-20 cases per 100,000 people, although it comprises 90% of the etiologies of SSNHL. However, recent studies have indicated that this ratio is approximately 150/100,000 [26]. Literature shows no difference in the incidence of this disease between sexes [27]. In our study, 59 (47.2%) of the 125 patients diagnosed with ISSNHL were male and 66 (52.8%) were female. The data indicated no effect of sex on the development of ISSNHL; this finding is consistent with that found in literature. ISSNHL can occur at any age, but it most commonly affects patients who are approximately 45 years of age [22]. The mean age of the patients in the current study was 45 years, which is consistent with that found in literature (min-max, 5-82; SD, 18.825; SEM, 1.584).

According to the available data, ISSNHL affects both the right and left ears at the same rate; however, SHL in association with perilymphatic fistulae is more frequently seen in the left ear [28]. In the present study, ISSNHL affected the right and left ear at the same rate.

The incidence of bilateral involvement was reportedly 4% in Shaia and Sheehy's 1220-case series, and 50% of those patients exhibited simultaneous bilateral involvement [27]. Increased serum antinuclear antibody autoantibody titers among patients with bilateral ISSNHL were reported in a study performed by Fetteman et al. [28] For this reason, serum autoantibody titers of patients with bilateral ISSNHL must be checked under the consideration of possible autoimmunity.

Huges [29] stated that the presence of syphilis should also be considered in patients with ISSNHL. In our study, one patient exhibited bilateral involvement of ISSNHL with negative results for serum syphilis serology and serum antibody testing. Inconsistency with previous studies was observed because of the presence of only one patient with bilateral SNHL in the present study. In an analysis of 809 patients with ISSNHL, Park et al. [30] reported that only 11 patients (1.3%) had a history of recurrent ISSNHL.

In our study, two patients had a history of recurrent ISSNHL, and its incidence was found to be consistent with that found in literature. Gäckler et al. [31] reported a positive family medical history rate of 21.4% in their study of 186 patients diagnosed with ISSNHL.

In the same study, the authors found that 10 patients had two relatives with a positive ISSNHL history [31]. In our study, the first-degree relatives of 29 (23.22%) patients and second-degree relatives of 12 (9.6%) patients had a history of ISSNHL. One patient had both a mother and sister with a positive ISSNHL history. When all data were statistically evaluated, a significant relationship was observed between ISSNHL and both the family medical history and the degree of kinship. The results of the present study are consistent with the data of Gäckler et al. [31] in terms of the presence of ISSNHL and degree of kinship. A few previous studies have been published on this subject. Our study is the first in our country. However, larger studies should be performed to obtain more accurate data. We believe that family

members of patients with ISSNHL should also be involved to these studies. Such studies will determine whether the presence of ISSNHL in the family medical history increases the possibility of ISSNHL.

Our study supports the presence of an association between ISSNHL and genetic predisposition. In the future, the relationship between ISSNHL and genetic susceptibility can be demonstrated by examining environmental and genetic factors in more detail. Proving genetic susceptibility to ISSNHL will lead to improvements in the prognosis, early diagnosis, and treatment of this disease.

Ethics Committee Approval: This study was approved by the Ethical Committee of Marmara University Faculty of Medicine (ID number: 09.2014.0266, Issue date: 18.12.2014).

Informed Consent: Written informed consent was not obtained due to the retrospective nature of this study.

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