Introduction

Meniere’s disease (MD) is a syndrome characterized by fluctuating vertigo, sensorineural hearing loss mostly confined to low frequencies, tinnitus, and aural fullness. Endolymphatic hydrops, a main pathologic finding of MD, has been proposed to have autoimmune, genetic, traumatic, viral, and allergic etiologies; however, no single cause has been validated. This may imply that endolymphatic hydrops is an epiphenomenon caused by an insult to inner ear homeostasis due to various etiologies. In this study, we investigated the frequency, inheritance patterns, and clinical characteristics of familial MD in a South Korean population. The results of this study can provide the basis for further genetic studies aimed at analyzing genetic mutations and mechanisms of MD in Asian populations.

Material and Methods

Direct and telephone interviews were performed for 286 definite MD patients and their family members who were suspected of having MD. The diagnosis of MD in family members was made by obtaining a detailed history, performing basic neurotological examinations and reviewing hearing test results. The clinical characteristics as well as the prevalence and inheritance patterns of familial MD were analyzed.

Results

Among the 331 definite MD patients, the prevalence of familial Meniere-like syndrome (at least one family member with definite MD and other members with probable MD) and definite familial MD (two or more family members with definite Meniere’s disease) were 9.8% and 6.3%, respectively. The most common inheritance pattern was autosomal dominant with incomplete penetrance (92.9% for familial MLS and 89.5% for definite familial MD). The preponderance of females (92.9% for familial MLS and 89.5% for definite familial MD) was significant. Also, maternal transmission was observed to be predominant. Two clinical factors were significantly different between familial and sporadic cases: the disease onset age and the prevalence of migraines. The disease onset was earlier and the prevalence of migraines was higher in familial cases than that of sporadic cases.

Conclusion

This is the first report describing the genetic aspects of MD in a single large Asian population. The prevalence of definite familial MD was 6.3% with an incomplete autosomal dominant inheritance pattern in most cases. Early onset age and a high prevalence of migraines were significant clinical features of familial MD in this South Korean population. These data could provide a basis for the analysis of the genetic mechanism of familial MD in Asian populations.

Reference