Etiology of 19 cases with rapidly progressive bilateral sensorineural hearing loss

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INTRODUCTION

Many people suffer from bilateral sensorineural hearing loss (bilateral SNHL), but it is not often that we encounter patients whose bilateral SNHL deteriorates rapidly within weeks or months. Although there are some case reports of rapidly progressive bilateral SNHL, their etiologies have not been well elucidated. In this study, we examined the etiologies of 19 cases with rapidly progressive bilateral SNHL.

METHODS AND MATERIALS

This study is a retrospective review of 19 patients who visited our department from January 2007 to December 2011 and met all of the following 4 criteria:

1. The Pure tone audiometry data shows bilateral sensorineural hearing loss.
2. The patient's history of deterioration was obtained in daily conversation developed within a year from the onset of symptom, with or without hearing aids.
3. The average of the pure-tone hearing threshold level increases with pace of 1 dB per month or more. (if the subject has received audiometric testing before or during the deterioration of hearing loss).
4. The patients with bilateral Meniere's disease were excluded.

Of all the 19 cases, 4 cases were caused by intracranial diseases: Cryptococcal meningitis, chronic viral meningitis with lymphocytes, meningeval metastasis of lymphoma, and superficial sialadenitis. Three cases were diagnosed as autoimmune inner ear disease (AIED) associated with systemic immune-mediated diseases: non-ANCA-associated vasculitis, Sjögren syndrome with Hashimoto disease, and Cogan's syndrome. One case was diagnosed as drug-induced (aminoglycoside) hearing loss and another was as auditory neuritis. Two cases were diagnosed as functional hearing loss (Figure 4).

RESULTS

DISCUSSION

In this study, rapidly progressive bilateral SNHL defined by our 4 criteria accounts for only 2.1% of bilateral SNHL (19 out of 900). The various kinds of diseases such as functional hearing loss, which is a psychogenic disease, and neuritis, which can be fatal, rapidly progressive bilateral SNHL. Using MRI, blood examination including autoantibody tests, and CSF analysis, we diagnosed 7 of the 19 cases (37%) as systemic or intracranial diseases. The appropriate treatments for these systemic and intracranial diseases improved the hearing loss in 4 of 7 cases. These results imply the importance of the evaluation of etiology of rapidly progressive bilateral SNHL.

CONCLUSIONS

Rapidly progressive bilateral SNHL is often caused by systemic or intracranial diseases, some of which, such as vasculitis and meningitis, could be fatal. Evaluation of systemic or intracranial diseases using MRI, blood test, and CSF analysis is important when you see patients who have rapidly progressive bilateral SNHL, which deteriorates over a period of weeks or months.

REFERENCES