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In Reply: IgG4 Related Disease and Sensorineural Hearing Loss

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We thank the authors for their thoughtful letter which raised important consideration concerning establishment of the inner ear involvement in IgG4-related disease.

As seen in the literature, IgG4-related disease is also a rare and emerging disease entity with autoimmune mechanisms, and hearing loss is not a frequent manifestation in IgG4-related disease [1,2]. Usual presentations of hearing loss associated with autoimmune diseases are bilateral and symmetric sensorineural type [3]. However, our case showed mixed hearing loss with middle ear involvement. Little is known about the pathophysiology and clinical manifestation of the otologic involvement in IgG4-related systemic disease. It usually involves interstitial tissue rather than mucosa [4,5]. Middle ear mucosa may not be claimed the target organ of IgG4-related systemic disease since we did not perform middle ear mucosal biopsy. However, we suspected that middle ear mucosa in this patient was also affected by this autoimmune disease based on the following two points. Firstly, histopathologic evaluation of the bronchial mucosa showed dense lymphoplasma cell infiltration. Considering nasopharyngeal and middle ear mucosa, a continuum of respiratory epithelium, can be involved in IgG4-related disease, conductive component of hearing loss can be accompanied due to the result of middle ear inflammation. Secondly, air-bone gap was not improved with ventilation tube insertion and antibiotics during initial treatment, but was improved with immunosuppressive therapy. Interestingly, fluctuation of air-bone gap paralleled with bone conduction threshold (Fig. 1) [1]. This means that the disease activity of IgG4-related disease paralleled in the middle ear and in the inner ear depending on the immunosuppressive therapy.

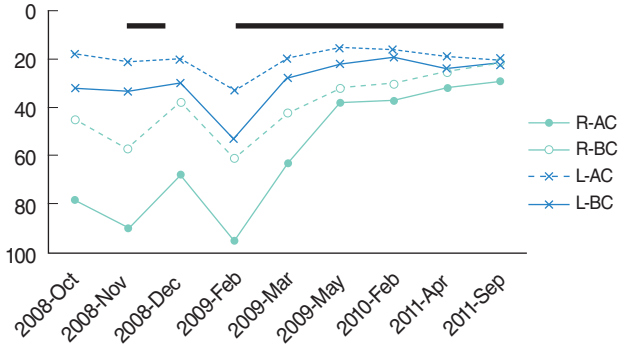

Fig. 1. Pure tone audiogram demonstrating fluctuation of bilateral mixed hearing loss. Air-bone gap was improved during the period of immunosuppressive therapy (black bars). R, right; BC, bone conduction threshold; AC, air conduction threshold; L, left.

Finally, we appreciate the authors for their in-depth observations on inner ear involvements in this emerging autoimmune disease.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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