An Intractable Case of MPO-ANCA-associated Middle Ear Disease

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We report herein on an intractable case of MPO-ANCA-associated middle ear disease with sensorineural hearing loss and facial palsy. A 73-year-old female complained of otitis media-like symptoms, but the treatment for otitis media with effusion was ineffective. She then showed bilateral progressive sensorineural hearing loss and facial palsy. PR3-ANCA was negative, but MPO-ANCA was positive in the serum. CT imaging revealed that the lesion was limited to the middle ear cavity without lesions in the lung and kidney. Non-specific findings were obtained in the histopathological examination of granular tissue of the mastoid cavity. She was diagnosed as having MPO-ANCA-associated middle ear disease with sensorineural hearing loss and facial palsy on the basis of the clinical findings and we administered cyclophosphamide in addition to a steroid. Discontinuance of the immunosuppressive therapy was, however, forced because of bacterial infection. Thereafter, steroid therapy alone failed to improve the left deafness, right moderate sensorineural hearing loss and right facial palsy.

Keywords : MPO-ANCA-associated middle ear disease, MPO-ANCA, PR3-ANCA, immunosuppressive therapy

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