We report three cases of myeloperoxidase-perinuclear anti-neutrophil cytoplasmic antibodies (MPO-ANCA)-positive otitis media with ANCA associated vasculitis (OMAAV). Case 1 was a 75-year-old female. She had right hearing loss and kidney involvement. Treatment with oral prednisolone was initiated and the MPO-ANCA titers decreased. Sensorineural hearing loss remained. Case 2 was a 77-year-old male. He had bilateral hearing loss and lung involvement. Treatment with oral prednisolone and oral cyclophosphamide was initiated. Serum MPO-ANCA became negative, and both hearing loss and lung involvement improved. However, serum PR3-ANCA became positive and he developed myelitis. Case 3 was a 71-year-old male. He had bilateral hearing loss but no other sites of involvement, such as lung and kidney. Treatment with oral prednisolone and oral cyclophosphamide was initiated. The MPO-ANCA titers became negative. He recovered from his hearing loss, however, he developed multiple cranial nerve palsy.

**Keywords** : ANCA, OMAAV, hearing loss, myelitis, cranial nerve palsy

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The clinical course of case 1

The MPO-ANCA titers decreased, however, sensorineural hearing loss remained.

(A) The tympanic membrane was cloudy and the middle ear cavity was filled with granulation on both sides.
(B) CT revealed a soft tissue lesion in the middle ear cavity and mastoid antrum. An infiltrative shadow was found in the left lung.
(C) The clinical course of case 2

(A) The tympanic membrane was cloudy and the middle ear cavity was filled with granulation on both sides.
(B) CT revealed a soft tissue lesion in the middle ear cavity and mastoid antrum.
(C) The clinical course of case 3

(A) The tympanic membrane was cloudy and the middle ear cavity was filled with granulation on both sides.
(B) CT revealed a soft tissue lesion in the middle ear cavity and mastoid antrum.
(C) The clinical course of case 3