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Sensorineural hearing loss as the initial manifestation of polyarteritis nodosa

Koichi Tsunoda, M.D., Jun Akaogi, M.D.*, Naoko Ohya, M.D.*, Toshihisa Murofushi, M.D.†

Abstract

A 74-year-old male was referred for the sudden onset of bilateral sudden deafness. The patient had no history of any disease or trauma to the head. Pure tone audiometry revealed bilateral moderate, to severe, sensorineural hearing loss. Auditory brain stem responses (ABRs) showed normal peak and interpeak latencies. These audiological findings suggested that his hearing loss could be attributed to inner ear lesions. However, we felt an alternative explanation for this sudden deafness was likely to exist because the patient also had a month-long fever of unknown origin (FUO) and weight loss of 5 kg/month. Using the criteria of The American College of Rheumatology, we made the diagnosis of polyarteritis nodosa (PAN). Serum MPO-ANCA was positive ($\times$ 661).

For treatment, the patient was begun on prednisolone and cyclophosphamide. Nine months later, fever, hypertension, nephritis, pneumonitis, and arthritis had completely resolved, the MPO-ANCA became negative (MPO-ANCA $< \times$ 10). Furthermore, his hearing improved.

Key words: Hearing Loss, Sensorineural; Polyarteritis Nodosa

Case report

A 74-year-old male was referred for the sudden onset of bilateral sudden deafness. For one month prior to admission, the patient had noted rapidly progressive bilateral hearing loss. The patient had no history of any disease or trauma to the head. Pure tone audiometry revealed bilateral moderate, to severe, sensorineural hearing loss. Auditory brain stem responses (ABRs) showed normal peak and interpeak latencies. These audiological findings suggested that his hearing loss could be attributed to inner ear lesions (Figure 1). Vestibular tests were normal. Roentgenograms of the nasal sinus and temporal bones showed no significant findings, but chest studies showed infiltrates.

However, we felt an alternative explanation for this sudden deafness was likely to exist because the patient also had a month-long fever of unknown origin (FUO) and weight loss of 5 kg/month. Therefore we referred the patient to the rheumatology service to rule-out the presence of a collagen-vascular, or rheumatological disease. At the time of admission the patient was discovered to be hypertensive (> 165/95 mmHg), and have renal insufficiency (BUN 48.4 mg/dl, Cr 4.0 mg/dl, urine protein 1.5 g/day), and polyarthritis. Using the criteria of The American College of Rheumatology,[1] we made the diagnosis of polyarteritis nodosa (PAN). Serum MPO-ANCA was positive ($\times$ 661).

For treatment, the patient was begun on prednisolone 40 mg/day and cyclophosphamide 50 mg/day. Nine months later, fever, hypertension, nephritis, pneumonitis, and arthritis had completely resolved, the MPO-ANCA became negative (MPO-ANCA $< \times$ 10). Furthermore, his hearing improved (Figure 1). The improvement in pure tone hearing was most remarkable at frequencies lower than 1000 Hz.

Discussion

One autopsy report revealed the evidence of PAN of the temporal bone, including endolymphatic hydrops of the basal turn of the cochlea, and fibrosis and ossification of the cochlea and vestibular system which obstructed the

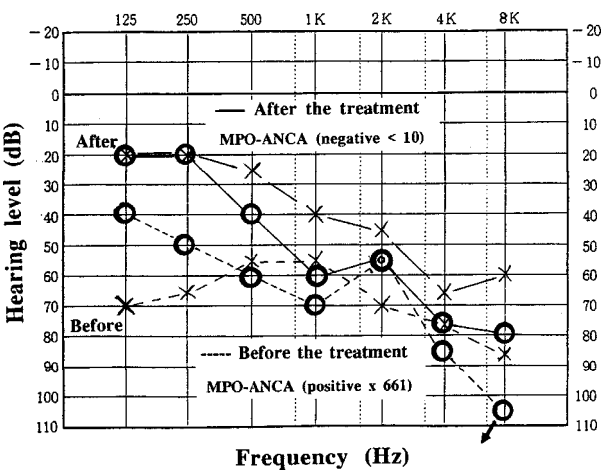

Fig. 1

Left side (X) and right side (O) pure-tone audiometry. The dotted line shows the results before treatment. The solid line shows the results nine months after the treatment.

From the Department of Otolaryngology and Rheumatology*, Nissan Tamagawa Hospital, Tokyo, and the Department of Otolaryngology†, Faculty of Medicine, University of Tokyo, Tokyo, Japan.
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auditory vessels.[2] In our patient, ABRs indicated an inner ear lesion, and fractuation of hearing at low frequencies suggested endolymphatics hydrops. Other autopsy studies have supported the role of vascular sequelae of PAN affecting the temporal bone.[3,4] There is one previous case report of bilateral, sudden onset deafness which proved to be the presenting complaint of PAN. As in our case, deafness improved with steroids and immunosuppressive treatment.[5]

Sudden onset sensorineural hearing loss is an otolaryngological emergency. In most cases, the patient's general condition is normal. However, when other findings are present, it is necessary to fully explore systemic causes of the otolaryngological disease. The early diagnosis of collagen-vascular diseases may enhance the response to systemic treatment. The MPO-ANCA study was helpful not only for diagnosis but also paralleled the vascular events of polyarteritis nodosa affecting the inner ear.

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Address for correspondence:
Koichi Tsunoda, MD,
Chief, Department of Otolaryngology,
Nissan Tamagawa Hospital,
4-8-1 Seta,
Setagaya-ku,
Tokyo, 158, Japan.

Fax: 00+ 81-3-3700-2090

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