A Patient with Myeloperoxidase Antineutrophil Cytoplasmic Antibody-Positive Polyangiitis Who Developed Sensorineural Hearing Loss and Scleritis

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Abstract

A 70-year-old woman was admitted to our hospital because of sudden hearing loss. She was treated with intratympanic dexamethasone, but her hearing impairment progressed. After admission, she developed scleritis of her left eye. Laboratory findings included elevated white blood cell count and C-reactive protein level, microhematuria, and proteinuria. Serology was positive for myeloperoxidase antineutrophil cytoplasmic antibody (MPO-ANCA), but negative for proteinase 3 (PR3)-ANCA. Renal biopsy revealed a single glomerulus with extensive glomerular tuft necrosis, indicating necrotizing vasculitis. She was diagnosed with MPO-ANCA-associated polyangiitis. ANCA-related polyangiitis should be considered in the differential diagnosis of sudden deafness or scleritis.

Key words: myeloperoxidase antineutrophil cytoplasmic antibody (MPO-ANCA), polyangiitis, sensorineural hearing loss, scleritis

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Introduction

Myeloperoxidase antineutrophil cytoplasmic antibody (MPO-ANCA)-positive microscopic polyangiitis is a necrotizing small-vessel vasculitis that commonly manifests as a rapidly progressive glomerular nephritis (RPGN), with necrotizing glomerular tufts, alveolar hemorrhage, or interstitial pneumonia (1-3). Most patients with ANCA-associated pauci-immune glomerulonephritis present with a rapid loss of renal function associated with hematuria, proteinuria, and hypertension (4). Here we report a rare case of MPO-ANCA-related microscopic polyangiitis in which the first manifestation was sensorineural hearing loss, which was followed by scleritis.

Case Report

A 70-year-old woman with a 20-year history of hypertension and a 7-year history of diabetes mellitus was admitted to the Department of Otorhinolaryngology at Ehime University Hospital because of progressive left ear hearing loss. Ten years previously, she suddenly experienced impaired hearing in her right ear. Her hypertension had been well controlled by treatments with amlodipine 5 mg/day and candesartan 4 mg/day and diabetes by diet and exercise. Her height and weight were 150 cm and 41 kg, respectively. Skin lesions, peripheral neuropathy and arthralgia were not seen. Because during the current therapy admission neither otitis media nor mastoiditis was found, she was diagnosed with sudden deafness in her left ear and treated with intratympanic dexamethasone. Despite this treatment, her left ear hearing impairment progressed. On day 20 after admission, she developed scleritis of her left eye (Fig. 1). She was afebrile, and the results of laboratory investigations included white blood cell count, 15,200 cells/μL; serum creatinine, 0.57 mg/dL; blood urea nitrogen, 14 mg/dL; and C-reactive protein, 4.38 mg/dL. Serology was positive for MPO-ANCA (127 ELISA units [EU]), but negative for proteinase 3 (PR3)-ANCA (<10 EU) and antinuclear antibody. Microscopic

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ANCAs are autoantibodies directed against neutrophils. Enzyme immunoassay research has revealed that ANCAs are directed mainly against proteinase 3 (PR3-ANCA) or myeloperoxidase (MPO-ANCA) (5). ANCAs were initially found in patients with certain types of glomerulonephritis, such as RPGN and segmental necrotizing glomerulonephritis in histopathological diagnosis obtained from renal biopsy (1, 3, 4). The three major ANCA-associated vasculitides are microscopic polyangiitis, Wegener’s granulomatosis, and the Churg-Strauss syndrome (6, 7). The present patient was diagnosed with microscopic polyangiitis in ANCA-associated syndromes. When ANCA-related polyangiitis is associated with a systemic vasculitis, patients may present with pulmonary-renal, dermal-renal, or multisystem disease. Common affected sites include the lungs, upper airways, sinuses, ears, eyes, gastrointestinal tract, skin, peripheral nerves, joints, central nervous system, and kidneys. Even in patients with no clinical extrarenal manifestations of active vasculitis, systemic symptoms consisting of fever, fatigue, myalgias, and arthralgias are common (8). The present patient with MPO-ANCA-related polyangiitis developed very rare symptoms, consisting of hearing loss as a first manifestation, followed by scleritis.

Hearing loss is associated with several vasculitides, most commonly with Wegener’s granulomatosis (9, 10) and Cogan’s syndrome (11, 12). In Wegener’s granulomatosis, the most frequent affected organs are skin, upper and lower respiratory tracts, the peripheral and central nervous system joints, and kidneys. The main cause of hearing loss in Wegener’s granulomatosis is due to otitis media or mastoiditis (13, 14). The present patient did not have skin, respiratory tract or nervous system lesions and otitis media or mastoiditis. Typical Cogan’s syndrome is defined as a chronic inflammatory disease of unknown origin, and autoimmune disease, characterized by bilateral sensorineural hearing loss, inflammatory ocular manifestations mainly due to nonsyphilitic interstitial keratitis and vestibular symptoms resembling Ménière’s disease (15, 16). The present patient did not have vestibular symptoms such as vertigo or dizziness and interstitial keratitis. The clinical manifestations of our patient differed somewhat from these two syndromes. Sensorineural hearing loss can be a presenting symptom of MPO-ANCA related polyangiitis; however, this presentation has been reported to be very rare (17-19).

An ocular surface manifestation such as scleritis is a major eye presentation in patients with PR3-ANCA- and MPO-ANCA-associated vasculitides. In addition, scleritis is a hallmark of Wegener’s granulomatosis (19). Watkins et al reported on 59 ANCA-positive patients with ocular inflammations, which included scleritis (75%), uveitis (17.9%), and other ocular inflammatory conditions (33.9%) such as peripheral ulcerative keratitis and orbital pseudotumor (20).
Matsumoto also reported on 31 MPO-ANCA-positive patients with eye manifestations (19). There were 19 women and 12 men, with ages ranging from 11 to 79 years. The most common ocular presentations were ocular surface manifestations, including episcleritis and scleritis, the latter of which was seen in our patient.

Most patients with ANCA-associated pauci-immune necrotizing glomerulonephritis have RPGN with rapid loss of renal function associated with hematuria and proteinuria. Although the present patient had abnormal urinary findings, including hematuria and proteinuria, she did not have progressive renal insufficiency. She presented with hearing loss, an easily recognized extrarenal symptom which might have allowed the diagnosis of MPO-ANCA-related polyangiitis at an early stage. As a histological evidence of necrotizing vasculitis, it is necessary to show the fibrin or fibrinogen deposition inside the affected glomerulus by immunohistochemistry. For this reason, a small number of glomeruli might be affected (only one glomerulus was affected in fifteen glomeruli obtained by renal biopsy) at early diagnosis.

This case illustrates why ANCA-related polyangiitis should be considered in the differential diagnosis of sudden deafness or scleritis.

The authors state that they have no Conflict of Interest (COI).

References

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