# **Clinical Case Reports**

## CASE REPORT

## **Reiter's syndrome and hearing loss: a possible association?**

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#### **Funding Information**

No funding information provided.

Received: 5 May 2014; Accepted: 5 May 2014

#### Clinical Case Reports 2014; 2(6): 310-312

doi: 10.1002/ccr3.119

## Introduction

Reiter's syndrome is a spondyloarthropathy, which is one of the diseases known as reactive arthritis [1]. It occurs as a nonsuppurative complication of an infection by lipopolysaccharide-producing bacteria, such as, *Yersinia enterocolitica, Campylobacter jejuni*, and *Chlamydia trachomatis* [1]. Its pathogenesis is unknown, but Reiter's syndrome is associated with the HLA-B27 gene. Its classic triad is defined as arthritis, conjunctivitis, and urethritis. Other manifestations may also occur such as central and peripheral neuropathy, mucocutaneous lesions and heart issues such as pericarditis and valvular lesions [1].

Autoimmune sensorineural hearing loss (SNHL) associated with autoimmune disorders such as polyarteritis nodosa, systemic lupus erythematosus, and Cogan's disease have appeared over the years [2, 3]. Some spondyloarthropathies associated with the HLA-B27 gene have been associated with ear issues such as Menière's disease and SNHL as well. The mechanism of human leukocyte antigen (HLA)-associated autoimmunity is complex; HLA-B27 antigen plays a role in several nonorgan-specific autoimmune disease states, including ankylosing spondylitis, acute anterior uveitis, and Reiter syndrome [3]. In white populations and most other groups, 90% of

#### Key clinical message

Patient complained of hearing loss and tinnitus after the onset of Reiter's syndrome. Audiometry confirmed the hearing loss on the left ear; blood work showed increased erythrocyte sedimentation rate and C3 fraction of the complement. Genotyping for HLA-B27 was positive. Treatment with prednisolone did not improve the hearing levels.

#### Keywords

Autoimmune disease, autoimmunity, hearing loss, reactive arthritis, reiter syndrome.

patients with ankylosing spondylitis carry HLA-B27, as opposed to 8% of the general population [4].

## **Case Report**

AFT, 57 years old, presented with a complaint of rapidly progressive hearing loss and an intense, waterfall-like, tinnitus on the left ear, that started 1 year before the consult. The patient did not indicate any other previous diseases that could affect hearing. In clinical evaluation, the outer ear, tympanic membrane, and middle ear were found to be normal. Audiometry was performed and showed mildly severe SNHL in the left ear (Fig. 1); the speech recognition threshold (SRT) was 30 dB on the right year and 70 dB on the left ear; speech recognition percentage index (SRPI) on the right ear was 84% at 70 dB and 40% at 100 dB on the left. Brain evoked response audiometry (BERA) demonstrated increased absolute latency on the left side and the absence of the wave III. The patient referred that the auditory complaints started just after the onset of joint pain in the lumbar column, left knee and left foot; conjunctivitis; and urethral discharge. By then, immunofluorescence of the material collected from the urethra showed Chlamydia infection; therefore, Reiter's syndrome was diagnosed.



**Figure 1.** Audiometry showing an assymetric bilateral neurosensorial hearing loss. Speech recognition threshold of 30 dB on the right ear and 70 dB on the left ear. Speech recognition percentage index on the right ear was 84% at 70 dB and 40% at 100 dB on the left ear.

Bone scintigraphy showed synovitis in the lumbar column and in the left knee (Fig. 2). Blood work showed and increased erythrocyte sedimentation rate (54 mm/h) and an elevated C3 fraction of the complement (170 mg/dL). Immunoglobulin levels were within normal range, and the tests for antinuclear factor, anti-DNA factor, cryoglobulin, c-Anca, and p-Anca, were negative. The HLA genotyping resulted in a positive response to HLA-B27.

A magnetic resonance imaging (MRI) of the inner ear was performed 6 months after the initial complaint of hearing loss, and it did not show any noteworthy findings.

## Discussion

Autoimmune inner ear disease can be diagnosed by clinical manifestations plus positive immune laboratory testing and beneficial treatment response. The typical patient profile includes unexplained, progressive, asymmetric, bilateral SNHL [5, 6].

The association between ankylosing spondylitis and neurosensorial hearing loss has been described in several studies in the past years [1-4, 7-10], even though it is



Figure 2. Bone Scintigraphy showing synovitis in lumbar column and in the left knee.

not yet recognized as an extra-articular feature of the disease. Recently published studies show a possible association between HLA-B27-related spondyloarthropathies and Menière's disease, causing autoimmune hearing loss [3]. That complication is assumed to occur because of autoantibodies, deposition of antigen–antibody complexes with activation of the complement system or an inflammatory reaction mediated by sensitized T lymphocytes. The hearing loss could be conductive, caused by middle ear involvement [11], or sensorineural, secondary to inner ear involvement [7, 8, 12–15]. There are also reports of cochlear dysfunction as a cause of ear involvement in patients with ankylosing spondylitis [16].

The patient complained of rapidly progressive hearing loss and the onset of an intense tinnitus on the left ear just after the initial symptoms of Reiter's syndrome. When autoimmune cause of the hearing loss was suspected, the patient was treated with a 15-day regimen of prednisolone, starting at 60 mg/day and tapering the dosage to zero by reducing 20 mg/day every 5 days, but the symptoms of hearing loss or tinnitus did not improve, and the audiometry did not show any improvements as well.

## Conclusion

The possibility of SNHL as an extra-articular feature of Reiter's syndrome should be considered, since the association between HLA-B27-related spondyloarthropathies and SNHL has been suggested by several studies.

## **Conflict of Interest**

None declared.

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