

Sudden hearing loss in a patient with rheumatoid arthritis; a case report and review of the literature

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Dear Sir,

There is considerable evidence suggesting a relationship between autoimmune diseases and hearing impairment. As a specific entity, autoimmune sensorineural hearing loss (ASHL) has been reported to occur in association with autoimmune diseases such as rheumatoid arthritis (RA)^{1,2}. We wanted to draw attention to this relationship by a case with RA presented sudden hearing loss.

Our case was a 37-year-old woman with a 12-year history of seropositive RA who had good response to methotrexate. Bilateral sudden sensorineural hearing loss developed in the follow-up of the patient. Her past medical history was unremarkable, she had no history of vertigo or ocular problems. On physical examination, systemic evaluation was normal except from mild limitations in the extension of right elbow and wrists bilaterally. Neurological examination was unremarkable and in particular there were no cerebellar or vestibular findings. Otoloscopic examination was also normal. A magnetic resonance image of her brain revealed no pathologic findings. The audiogram showed symmetric sensorineural hearing loss predominantly for the high frequencies. ASHL was considered as a diagnosis based on bilateral rapidly progressive sensorineural hearing loss and the presence of RA. Prednisone 1 mg/kg per day was administered to the patient in the otorhinolaryngology review and she responded well with a subtotal recovery.

AISH is a well-described clinical entity that has a relationship with a number of systemic autoimmune disorders¹. It was defined by McCabe in a case series with sudden sensorineural hearing loss who responded to corticosteroids and/or cyclophosphamide³. This favorable response to corticosteroids suggested that hearing loss in these patients was caused by an autoimmune mechanism in spite of its uncertain etiopathoge-

nesis. Typically it presents with an idiopathic, rapidly progressive predominantly bilateral sensorineural hearing loss⁴. One of the important features of this entity is its association with systemic immune-mediated diseases. A systemic autoimmune disorder may be present in approximately one-third of cases¹. Several autoimmune diseases such as RA, ankylosing spondylitis, systemic lupus erythematosus, Sjogrens syndrome, polyarteritis nodosa, relapsing polychondritis, Cogans disease and Crohns disease have been reported as associated disorders with ASHL⁵. In addition, recent reports have also described associations with primary antiphospholipid syndrome and ankylosing spondylitis^{6,7}. Since diagnostic criteria for AISH are currently not defined, diagnosis based on clinical findings. Similar with our case, the typical high-risk clinical profile was suggested as middle-aged patients (often female) with bilateral, asymmetric, progressive sensorineural hearing loss, with or without dizziness, and occasional systemic immune disease such as RA⁸. Its increased prevalence among middle-aged females may be consistent with other autoimmune diseases. Treatment strategies for ASHL are usually controversial; corticosteroids and/or immunosuppressants such as cyclophosphamide suggested to be effective³. Responsiveness to steroid can be seen and hearing loss may be reversible with prompt treatment. The efficacy of adalimumab was reported in the management of a RA patient with ASHL that did not respond to steroids¹. In the contrary, sensorineural hearing loss temporally related to adalimumab in two patients with RA was also reported⁹. In previous investigations there were positive outcomes with Infliximab and contrasting results with Etanercept in this setting¹⁰⁻¹². Since tumor necrosis factor (TNF) blockade by specific antibodies may offer an additional treatment option in some reports and possible anti-TNF associated sensorineural hearing loss was reported in others, further studies on anti-TNF agents in this situation are needed.

In conclusion, with this case presentation we wan-

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ted to draw attention to ASHL that may be in association with autoimmune diseases such as RA. Although it is unusual as a cause of hearing loss, is important to recognize because early diagnosis and treatment can have a marked effect on the clinical outcome.

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