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Case Report

# Audiovestibular Manifestations in Patients with Ankylosing Spondylitis – A Case Report and Review of the Literature

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Ankylosing spondylitis is a chronic systemic inflammatory disease of unknown origin affecting up to 1% of the population. Audiovestibular impairment has been observed in ankylosing spondylitis and sensorineural hearing loss (SNHL) is the most common form. The cause of SNHL is still unknown but the possible causes are as follows: vascular inflammation (obliterative vasculitis) of small vessels, ossification of the articular tissue of the middle ear, and use of non-steroid anti-inflammatory drugs. This is a case report of a patient with ankylosing spondylitis and SNHL along with a discussion of the literature regarding cochleovestibular impairment in ankylosing spondylitis.

**KEYWORDS:** Ankylosing spondylitis, sensorineural hearing loss, inflammatory disease

## INTRODUCTION

Ankylosing spondylitis (AS) is a chronic systemic inflammatory disease. The disease primarily affects the axial skeleton and its cause is unknown; it is known to affect up to 1% of the population. The onset is usually in the 30s and affects men three times more often than women. Autoimmune etiology characterized by inflammation is the suspected etiology. AS typically affects the spine and sacroiliac joints. Peripheral joints are the other sides of the skeleton involvement. Inflammation of the joints leads to synovitis, fusion, and a tendency to ankylosis. Osteoporosis and vertebral fractures can also be seen. This rheumatic disease also involves extraskeletal organs, such as the eyes, heart, lungs, and gut, along with manifestations such as anterior uveitis, cardiac conduction defect, aortic valve incompetence, inflammatory bowel disease, and renal and lung abnormalities. The diagnosis depends on the following New York criteria: the presence of back pain (because of the inflammation of the spine and sacroiliac joints), the restriction of lumbar flexion and chest expansion, and radiological findings of sacroiliitis and the human leukocyte antigen (HLA) B27 positivity. The disease is strongly associated with the HLA B27. Sensorineural hearing loss (SNHL) is the most common form of cochleovestibular impairment of AS; however, the cause of SNHL still remains unknown.

In this report, a patient who has AS with SNHL was presented and the literature of cochleovestibular impairment in AS was discussed.

## CASE PRESENTATION

A 52-year-old man who was a bank clerk visited our clinic as an outpatient; he had a 2-year history of slowly progressing hearing impairment. He was diagnosed with AS at the age of 34 years. With bilateral sacroiliitis on plain radiograph and magnetic resonance imaging (MRI) as well as positivity for B27 on HLA typing according to the modified New York criteria, he was diagnosed with ankylosing spondylitis. Previous episodes of peripheral inflammatory arthritis had been treated with sulphasalazine and some non-steroid anti-inflammatory drugs (NSAID). He has been using naproxen since the last 2 years. The respiratory, cardiovascular, and other systems as well as the eyes were normal. In his laboratory investigations, hemoglobin was 13.2 g/dL with normal white cell and platelet counts. The urea, electrolyte, and calcium levels as well as the liver and thyroid function tests were normal. The erythrocyte sedimentation rate (ESR) was 20 mm in the first hour. The patient had a normal complete ear, nose, and throat (ENT) examination; normal facial nerve function; no complaints of tinnitus and vertigo; and no family history for hearing loss or no history of acoustic or head trauma as well as otologic or neurotologic operation. Pure tone audiometry revealed bilateral SNHL and 50-dB hearing level in both ears, particularly increasing after 6000 Hz. With impedance audiometry (tympanogram), there was a type A curve for each ear. Speech discrimination scores were 92% for each ear. With videonystagmography, he had no pathologic nystagmus. Written informed consent was obtained from the patient.

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## DISCUSSION

Audiovestibular manifestations believed to be depended on vasculitis or autoimmunity have been observed in autoimmune-mediated diseases or rheumatoid connective tissue diseases. SNHL is the most prominent audiovestibular symptom in these diseases. Despite the unknown underlying pathophysiological mechanism, there are some theories for explaining the emergence of SNHL such as immune complex-dependent vasculitis in the inner ear, antibody formation against the inner ear, and neuritic as well as ototoxic effects of drugs. The most accepted theory is the autoimmune theory (autoimmune inner ear disease) [1].

Sensorineural hearing loss, conductive hearing loss, and vestibular symptoms were described in patients with AS. A discussion on the characteristic ear manifestation of AS is still ongoing. Some authors advocate that middle ear involvement leading to conductive hearing loss is the typical form but others think that SNHL because of inner ear involvement [2, 3]. The following three mechanisms may be responsible for otologic involvement: 1) pathology of inner ear epithelium and vascular tissue because of autoimmune reaction, 2) impaired mobility of middle ear ossicles, and 3) NSAID ototoxicity [4].

Amor-Dorado et al. [5] found SNHL in 58% of the 50 patients with AS. Casellini et al. [6] determined higher SNHL rates in 22 patients with AS than those in the control group. Alatas et al. [4] reported the ratio of SNHL to be 28.6% in 28 patients with AS, whereas it was reported to be 4.3% in the control group. Dagli et al. [7] studied 28 patients with AS and 25 controls and found SNHL in 10 patients (35%). Eryılmaz et al. [1] reported the ratio of SNHL to be 28.6%.

Sensorineural hearing loss can be observed in all frequencies but high frequencies are most affected. Eryılmaz et al. [1] observed significant deterioration at higher frequencies in 59 patients with AS. In the study by Adam et al. [2], there was no statistically significant difference between the AS group (45 patients) and the control group (31 patients) with respect to conventional frequency thresholds. However, in this study, there was a statistically significant difference at a frequency of 14–16000 Hz in 32 patients with AS (71%) as compared with 12 controls (40%) [2]. Alatas et al. [4] observed SNHL both at low and high frequencies in patients with AS. Erbek et al. [8] found statistically significant difference at high frequencies in 32 patients with AS.

Acoustic brainstem responses (ABR) and otoacoustic emission testing results were also reported in patients with AS having SNHL. Alatas et al. [4], did not find any statistically significant difference in the ABR results between patients and the control group. Erbek et al. [8] found that the rates of reproducibility in transient evoked otoacoustic emission testing (TEOAE) were significantly lower in patients with AS than those in controls. Daglı et al. [7] determined the same result with distortion product otoacoustic emission testing (DPOAE). According to Daglı et al. [7], high frequencies are organized in the basal part of the cochlea; therefore, the damage is in the outer hairy cells of basal and intermediate parts of the cochlea in patients with AS. Nevertheless, Kahveci et al. [9] did not find any cochlear dysfunction in patients with AS with regard to the audiological and DPOAE results. They explained this with the multifactority of SNHL in AS (age, drugs, duration of the disease, duration of medication, acoustic trauma, and previous middle and inner ear infections). They added that another possible reason for obtaining normal DPOAE results might be owing to SNHL because of hairy cell dysfunction in the inner ear and synaptic or retro cochlear pathology [9].

During the course of AS, immunologic vasculitis, autoantibodies against the inner ear, and ototoxic effects of drugs may cause SNHL. Vasculitis, which was observed in autoimmune diseases, can cause degenerative changes in tissues. Because of autoimmune and vascular causes, degeneration or loss of hairy cells can be observed in the patients with AS. Stria vascularis may be the affected area in AS. Stria vascularis is an epithelial tissue that is rich in vascularization, and its main function is to arrange the electrical potentials of endocochlea and to supply the hairy cells [4, 10].

The most common cause of renal involvement in patients with AS is secondary amyloidosis (abnormal accumulation of amyloid proteins in various tissues of the body). Amyloidosis can cause polyneuropathies. According to Kahveci et al. [9], the involvement of cochlear and vestibular nerves as well as the central pathways of cochleovestibular system with amyloidosis may cause SNHL. Drugs used for the medical treatment of AS were discussed as a possible etiological factor for SNHL. The most used medications in AS are as follows: NSAID and sulfasalazine (an antibacterial from sulfonamides). Active metabolites of sulfasalazine are 5-aminosalisilic acid and sulfapiridine. These drugs are immune suppressants. Ototoxic effects of these immunosuppressant drugs are well discussed in the literature. Savastano et al. [10] argued that the use of salicylates may cause SNHL but cochlear improvement can be seen after the cessation of treatment. Besides, SNHL was observed to be greater in patients treated with anti-TNF∝+methotrexate than in patients treated with anti-TNF∝ alone [10].

There was no correlation determined between hearing levels and the clinical or laboratory parameters of patients with AS. Amor-Dorado et al. [5], considered that the risk of SNHL was higher in patients with the involvement of the hip, anterior uveitis, and presence of the HLA B27.

Conductive hearing loss because of the damage of ossicular chain was also defined besides SNHL in patients with AS. Amor-Dorado et al. [5] reported abnormal tympanometry results in 8% of the patients despite normal otomicroscopic examination and absence of any middle ear effusion. Amor-Dorado et al. [5] explained this statement with the stiffness of the ossicular chain.

Vestibular dysfunction is also seen in patients with AS. In the study by Amor-Dorado et al. [5], the incidence of abnormal results in vestibular tests was high. Erbek et al. [8] determined vestibular pathology in 34% of the patients and statistically significant abnormal results in caloric tests.

Similar to other rheumatologic diseases, inner ear involvement must be kept in mind for patients with AS; these patients must be followed up with audiological and vestibular examinations.

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