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Case Report

Idiopathic Inflammatory Arthritis in the Auditory Canal in a Patient With Hearing Impairment: A Case Report and Literature Review

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Inflammatory arthritis can affect the auditory system during the disease course. Although most cases show asymptomatic hearing impairment, it can result in hearing loss. Here we describe the case of a 70-year-old female with hearing impairment associated with idiopathic inflammatory arthritis in her auditory system. She had suffered from hearing difficulties for decades; however, the causes of her hearing impairment had not been evaluated. Pure tone audiometry showed severe sensorineural hearing loss requiring a cochlear implant. The workup for the cochlear implant revealed erosive changes in the incudomalleolar and incudostapedial joints with soft tissue swelling on temporal bone computed tomography. Bone pathology revealed plasmacytic infiltration and granulomatous inflammation. Laboratory examinations showed elevated levels of inflammatory markers; otherwise, she had negative results for all autoantibodies. In patients with idiopathic hearing loss, inflammatory arthritis of the middle ear without peripheral arthritis can provide a clue regarding the cause of the hearing loss.

Keywords: Arthritis, Middle ear, Hearing loss, Cochlear implantation

INTRODUCTION

Hearing impairment has been reported in patients with autoimmune diseases including rheumatoid arthritis (RA), juvenile idiopathic arthritis (JIA), systemic lupus erythematosus (SLE), and vasculitis [1-10]. Among autoimmune diseases, previous studies have described the association between hearing impairment and RA [10-28]. While hearing impairment has been reported to occur in up to 60% of patients with RA, its pathogenesis is not clear [11]. Sensorineural hearing loss was the most common hearing impairment, occurring in up to 80% of cases, following by conductive hearing loss [13,15]. While conductive hearing loss can be caused by arthritis of the incudostapedial and incudomalleolar joints, sensorineural hearing loss can be associated with inflammation of the inner ear, as well as vasculitis, neuritis, or deposition of immune complex, which result in cochlear damage [15-18]. The risk factors for hearing impairment in RA include old age, male, long disease duration, and anti-citrullinated peptide antibody (ACPA) positivity [13-15]. However, to our knowledge, there has been no report of the association of hearing impairment with inflammatory arthritis in the ossicular chains without RA. Herein, we report a case in

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an older female with no peripheral symptoms of inflammatory arthritis who experienced hearing loss with joint abnormalities in both middle ears.

CASE REPORT

A-70-year-old female was referred to the otolaryngology clinic of a tertiary hospital in Seoul, South Korea, for worsening hearing difficulty in both ears and dizziness that had developed in the last month. She had hearing impairment in her right ear that had developed decades prior and had been listening only with her left ear with a hearing aid for more than 10 years; however, the cause of her hearing impairment had not been evaluated. Her current medications included amlodipine; candesartan; atorvastatin; and methimazole for hypertension, dyslipidemia, and hyperthyroidism, which she had taken for more than 1 year. She had never smoked and denied alcohol abuse. A history of noise trauma and injury in both ears was not reported, and there was no family history of hearing loss. She presented with non-whirling type vertigo with intermittent tinnitus, which was aggravated when walking. Caloric, head impulse, and rotational chair tests confirmed the bilateral loss of vestibular function, which could explain the aggravation of dizziness when walking. No symptoms including ear fullness, headache, nausea, vomiting, visual change, or dysarthria were found. Before visiting our hospital, high-dose corticosteroid treatment was tried for several days due to suspected sudden sensorineural hearing loss 6 months prior; however, the response was poor. The otoscopic examination did not reveal any evidence of chronic otitis media. Pure-tone audiometry indicated severe sensorineural hearing loss and reduced speech recognition in both ears (Figure 1A); therefore, cochlear implantation in her left ear was recommended. High-resolution magnetic resonance imaging for the assessment of the cochlear nerve was normal, however, temporal bone computed tomography revealed erosive change and deformities of both incudomalleolar and incudostapedial joints with soft tissue swelling (Figure 2). Based on these findings, the patient underwent consultation from a rheumatologist for underlying rheumatic disease. The patient had no tender joints or swollen joints suggestive of systemic inflammatory arthritis such as RA. The results of laboratory studies were within normal values, including complete blood cell count, C-reactive protein, and autoantibodies (rheumatoid factor, anti-nuclear and ACPA), except for an elevated erythrocyte sedimentation rate (ESR, 49 mm/

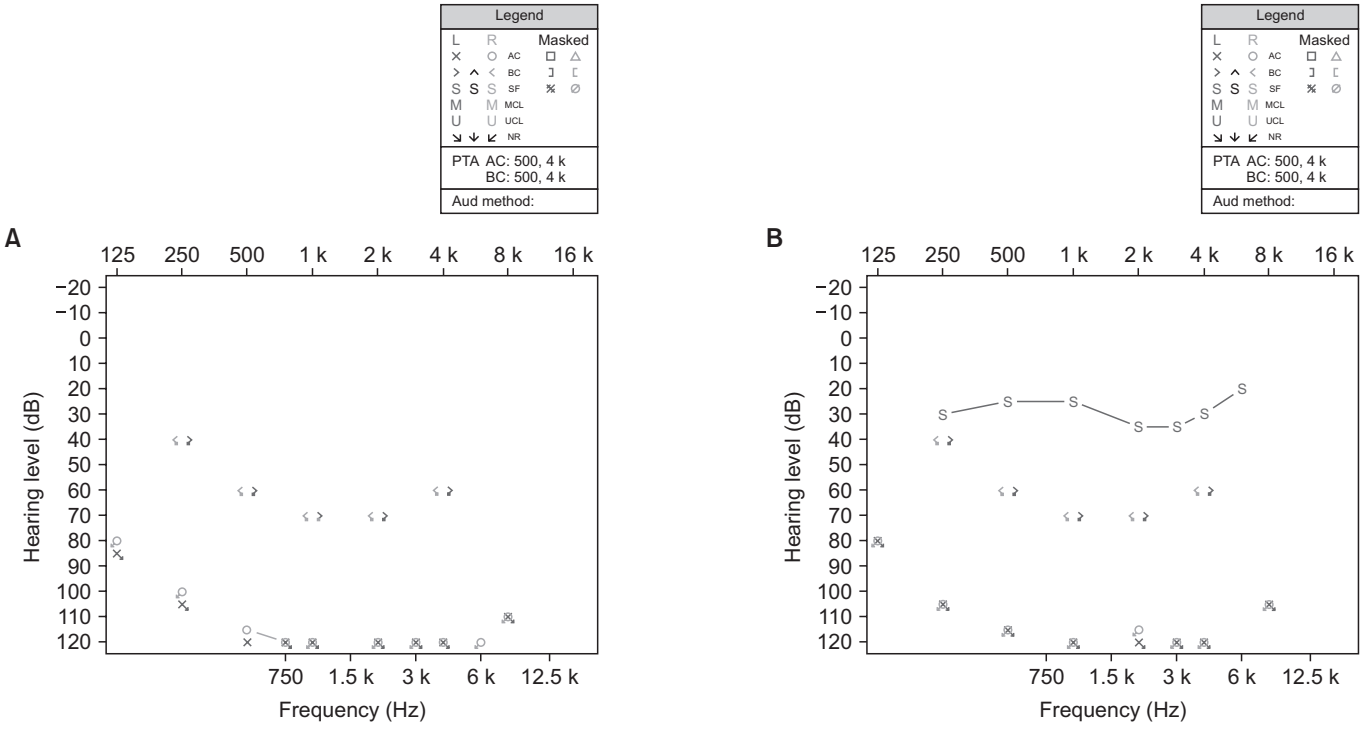

Figure 1. Pure-tone audiometry. (A) Before cochlear implant, severe sensorineural hearing loss of both ears observed. (B) Two months after cochlear implant, aided audiogram showed a 30 dB threshold through all frequencies.

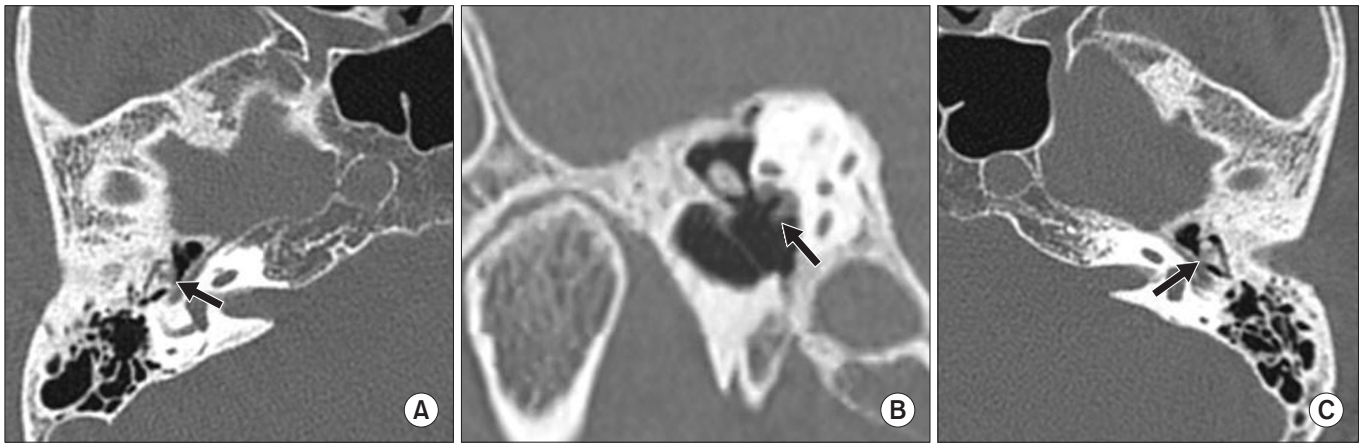

Figure 2. CT findings of the temporal bone. CT scan of the right middle ear cavity of a 70-year-old female with hearing loss. The arrows indicate the erosive change and deformity of the incudomalleolar joints with soft tissue infiltration (A, C) in the axial plane and soft tissue infiltration at the stapes in the stenvers plane (B). CT: computed tomography.

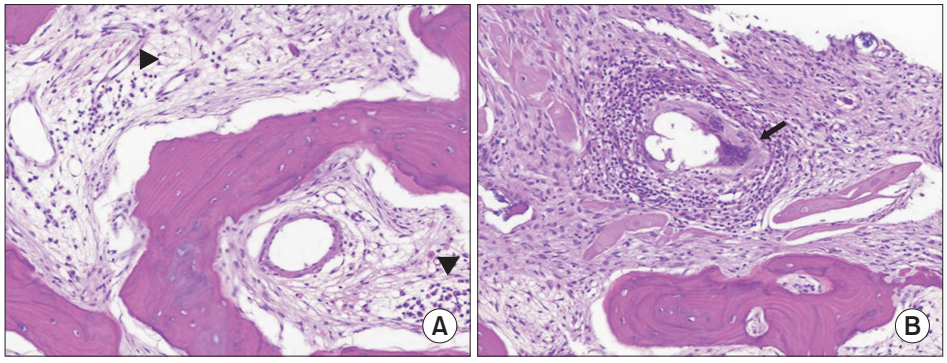

Figure 3. Pathology slides demonstrating chronic osteitis in the left incus (H&E, ×400). (A) Lymphoplasmacytic infiltration (triangles). (B) Granulomatous inflammation (arrow).

hr). To define the underlying cause of joint deformities in the middle ear, a biopsy was obtained from the arthritic part while performing cochlear implantation in the left ear. The pathologic findings of incus areas revealed plasmacytic infiltration and granulomatous inflammation (Figure 3). No acid-fast bacilli were identified on Ziehl–Neelsen staining and no fungal organisms were identified on Gomori's methenamine silver staining. As patients did not present any clinical manifestations suggesting granulomatosis with polyangiitis (GPA) such as sinusitis, chronic otitis media, and there was no evidence of vasculitis in pathologic examination. The patient was discharged on the sixth hospital day and visited a rheumatology outpatient clinic 4 weeks after discharge. Due to the patient's persistently high ESR, low-dose prednisolone (10 mg/day) and methotrexate (7.5 mg/week) were prescribed for non-infectious chronic inflammatory arthritis. Two months after the cochlear implantation, the patient's hearing level was improved, with an aided audiogram showing a 30 dB threshold through all frequencies (Figure 1B).

DISCUSSION

Hearing impairment occurs in 37%~60% of adults aged 61~80 years [29]. Among them, age-related sensorineural hearing loss followed by noise trauma, Meniere disease, infection, medication, and autoimmune disease, is the most common [29]. The patient in the present case thought that her hearing loss was simply a sensorineural defect. However, her evaluation for a cochlear implant incidentally revealed erosive arthritis of the middle ear and the pathology findings in incus showed granulomatous inflammation. Granulation tissue in middle ear can be observed in various disease such as tuberculosis, fungal infection, Langerhans cell histiocytosis, and chronic inflammatory disease [30]. Because there was no evidence of infection or chronic otitis media, we considered that autoimmune disease might contribute to patient's hearing dysfunction. In autoimmune disease, sarcoidosis, GPA, RA can present granulation tissue [31-33]. However, the inflammation is limited to the ear,

Table 1. Literature on hearing impairment in patients with systemic inflammatory arthritis

Author	Disease	Number of patients	Mean age (yr)	Hearing impairment, number (%)	Abnormal tympanogram, number (%)	Pathology findings of the middle ear	Findings
Goodwill et al. [22]	RA	76	52.8	7 (9.2) : SNHL 5, CHL 2	None	Bone absorption, fibrous tissue	HL was not related with RA duration or activity
Rosenberg et al. [23]	RA	68	N/A	None	16 (42)	(-)	The conducting system was abnormal in RA but HL is rare
Poorey and Khatri [24]	RA	25	N/A	16 (64) : SNHL 13, CHL 3	8 (32)	(-)	CHL was common in early and active RA
Ozcan et al. [11]	RA	37	47.7	19 (51.4) : SNHL 10, MHL 4, CHL 5	14 (37.8)	(-)	HL could be caused by multifocal involvement of the audiologic system in RA
Takatsu et al. [17]	RA	36	54.6	13 (36.1) : SNHL 13	18 (25)	(-)	SNHL is related to ESR and plasma IL-6, and MMP-3. Latent-type CHL can be caused by stiffness of the middle ear
Halligan et al. [25]	RA	29	63 (median)	17 (59) : SNHL 13, MHL 1, CHL 3	5 (17)	(-)	No difference in audiometric measurement between patients with RA and controls
Milisavljevic et al. [18]	RA	9	66.9	N/A	N/A	(-)	SEM showed increased lysis and surface degeneration of the auditory ossicles in patients with RA
Tavernier and Ranfaing [26]	RA	1	59.0	MHL	N/A	Mass consisting of fibrous tissue riddled with inflammatory elements, primarily mononucleates	Inflammatory fibrous tissue in the middle ear could result in HL in patients with RA
Pascual-Ramos et al. [27]	RA	113	43.3	27 (24) : SNHL 25, MHL 2	20 (18)	(-)	SNHL the most common type of HL in patients with RA, and older age was a risk factor for HL
Lobo et al. [14]	RA	43	48.9	20 (46.5) : SNHL 16, MHL 2, CHL 2	6 (11.9)	(-)	SNHL was the main HL in patients with RA and was associated with anti-citrullinated peptide antibodies
Ahmadzadeh et al. [28]	RA	42	53.0	9 (21.3) : SNHL 5, MHL 1, CHL 3	13/84 ears (15.4)	(-)	SNHL was more prevalent in patients with refractory RA
Magarò et al. [8]	AS	1	56	CHL	+	(-)	CHL can be induced by ear arthritis in patients with AS
Casellini et al. [5]	RA/AS	RA 19 AS 22	RA 56 AS 45.5 (median)	RA 13 (68.4) : SNHL 11, MHL 1, CHL 1 AS 15 (68.2) : SNHL 13, CHL 2	RA 4 (14.3) AS 3 (15.8)	(-)	HL can occur in patients with AS patients as well as those with RA. Small numbers of patients with otosclerosis among those with AS
Dagli et al. [4]	AS	28	34.3	10 (35) : SNHL 10	None	(-)	Damage of the outer hair cells in patients with AS

Table 1. Continued

Author	Disease	Number of patients	Mean age (yr)	Hearing impairment, number (%)	Abnormal tympanogram, number (%)	Pathology findings of the middle ear	Findings
Adam et al. [3]	AS	45	39.6	32 (71.1) : SNHL 32	N/A	(-)	High-frequency SNHL was common in AS; risk factors include disease duration and extraspinal involvement
Amor-Dorado et al. [9]	AS	50	52.5	29 (58) : SNHL 29	4 (8)	(-)	High-frequency SNHL and abnormal vestibular tests more common in patients with AS than controls.
Amor-Dorado et al. [2]	PsA	60	52.9	36 (60) : SNHL 36	N/A	(-)	High-frequency SNHL was the predominant pattern of HL in patients with PsA
Siamopoulou-Mavridou et al. [7]	JIA	18	10.6	N/A	17/36 ears (47.2)	(-)	JIA changes middle ear function by stiffness of the tympanic membrane and ossicular chain
Takatsu et al. [17]	JIA	19	12.2	None	12/38 ears (31.6)	(-)	A dual effect of JIA on both the middle and inner ears

RA: rheumatoid arthritis, SNHL: sensorineural hearing loss, CHL: conductive hearing loss, HL: hearing loss, N/A: not available, MHL: mixed hearing loss, ESR: erythrocyte sedimentation rate, IL-6: interleukin-6, MMP-3: matrix metalloproteinase-3, SEM: scanning electron microscope, AS: ankylosing spondylitis, PsA: psoriatic arthritis, JIA: juvenile idiopathic arthritis.

and clinical symptoms and signs suggesting sarcoidosis, RA, and GPA were not presented. Furthermore, pathological findings revealed no evidence of vasculitis. Therefore, the patient's arthritis could not be defined yet as a specific rheumatic disease.

Hearing impairment caused by autoimmune diseases is mostly discussed in patients with RA, followed occasionally by SLE, ankylosing spondylitis, and JIA [2-10]. We reviewed articles on hearing loss related to inflammatory arthritis (Table 1). Although hearing impairment occurs frequently in patients with autoimmune disease, the pathogenesis of hearing impairment remains controversial [13,15]. Bone defects in the middle ear were observed in murine models of RA and destruction of the incudostapedial and incudomalleolar joints can result in impaired conduction [18,34]. However, it does not necessarily lead to a hearing impairment [6,17,23]. Previous studies showed that sensorineural hearing loss, rather than conductive hearing loss, is the predominant type of hearing impairment in autoimmune disease [2,5,27,28]. Although the As type-tympanogram was more observed in RA patients than the controls, all the patients with hearing impairment presented sensorineural hearing loss [17]. Sensorineural hearing loss can be caused by damage to the inner ear and cochlear hair cell and can also be impaired by vasculitis, neuritis, or the deposition of immune complexes in patients with RA [4,5]. As patients with RA have various forms of hearing impairment, joint damage in the middle ear and inflammatory changes in the inner ear may cause conductive hearing impairment, sensorineural hearing loss, and mixed-type loss. The present case showed sensorineural hearing impairment on pure-tone audiometry, and conductive impairment was not found even with inflammatory arthritis in both the middle ears simultaneously. Thus, it was difficult to determine the impact of the joint damage in her middle ear on her hearing impairment. However, the damage was evidence of an antecedent inflammatory process that mimicked RA and might be related to hearing impairment.

The development and severity of hearing impairment could be related to disease activity [14,35]. The patient was administered methotrexate and prednisolone to control the inflammation in her ear. However, the roles of disease-modifying antirheumatic drugs (DMARDs) and corticosteroids in the recovery of hearing function are uncertain [1,15,16]. Therefore, further studies are needed to assess the role of DMARDs on hearing recovery in patients with inflammatory arthritis who receive cochlear implants.

SUMMARY

This case is the first report of suspected hearing loss related to inflammatory arthritis in the middle ear without evident RA. Our experience supported the theory that hearing impairment occurs through a multifocal involvement of the auditory system, including cochlear hair cells and ossicles. Therefore, additional studies are required on inflammatory arthritis of the auditory canal without features of systemic autoimmune in patients with unknown causes of hearing impairment.

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CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

AUTHOR CONTRIBUTIONS

Conceptualization: H.J.P. and Y.G.K. Data acquisition: S.H.N. and G.H.K. Writing—original draft: S.H.N. Writing—review & editing: H.J.P. and Y.G.K.

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REFERENCES

1. Rahne T, Clauß F, Plontke SK, Keyßer G. Prevalence of hearing impairment in patients with rheumatoid arthritis, granulomatosis with polyangiitis (GPA, Wegener's granulomatosis), or systemic lupus erythematosus. Clin Rheumatol 2017;36:1501-10.
2. Amor-Dorado JC, Barreira-Fernandez MP, Pina T, Vázquez-Rodríguez TR, Llorca J, González-Gay MA. Investigations into audiovestibular manifestations in patients with psoriatic arthritis. J Rheumatol 2014;41:2018-26.
3. Adam M, Erkan AN, Arslan D, Leblebici B, Ozlüoglu L, Nafiz Akman M. High-frequency sensorineural hearing loss in patients with ankylosing spondylitis:is it an extrarticuler feature of disease? Rheumatol Int 2008;28:413-7.
4. Dagli M, Sivas Acar F, Karabulut H, Eryilmaz A, Erkol Inal E. Evaluation of hearing and cochlear function by DPOAE and audiometric tests in patients with ankylosing spondilitis. Rheumatol Int 2007;27:511-6.
5. Casellini C, Citera G, Rosemffet M, Ruggeri S, Saviotti A, Maldonado Cocco JA. Audiovestibular disorders in patients with ankylosing spondylitis. J Clin Rheumatol 2005;11:81-5.
6. Ikiz AO, Unsal E, Kirkim G, Erdag TK, Guneri EA. Hearing loss and middle ear involvement in patients with juvenile idiopathic arthritis. Int J Pediatr Otorhinolaryngol 2007;71:1079-85.
7. Siamopoulou-Mavridou A, Asimakopoulos D, Mavridis A, Skevas A, Moutsopoulos HM. Middle ear function in patients with juvenile chronic arthritis. Ann Rheum Dis 1990;49:620-3.
8. Magarò M, Ceresia G, Frustaci A. Arthritis of the middle ear in ankylosing spondylitis. Ann Rheum Dis 1984;43:658-9.
9. Amor-Dorado JC, Barreira-Fernandez MP, Vazquez-Rodriguez TR, Gomez-Acebo I, Miranda-Filloy JA, Diaz de Teran T, et al. Audiovestibular manifestations in patients with ankylosing spondylitis. Medicine (Baltimore) 2011;90:99-109.
10. Mancini P, Atturo F, Di Mario A, Portanova G, Ralli M, De Virgilio A, et al. Hearing loss in autoimmune disorders: prevalence and therapeutic options. Autoimmun Rev 2018; 17:644-52.
11. Ozcan M, Karakus MF, Gündüz OH, Tuncel U, Sahin H. Hearing loss and middle ear involvement in rheumatoid arthritis. Rheumatol Int 2002;22:16-9.
12. Lee SY, Kong IG, Oh DJ, Choi HG. Increased risk of sudden sensory neural hearing loss in patients with rheumatoid arthritis: a longitudinal follow-up study using a national sample cohort. Clin Rheumatol 2019;38:683-9.
13. Emamifar A, Bjoerndal K, Hansen IM. Is hearing impairment associated with rheumatoid arthritis? A review. Open Rheumatol J 2016;10:26-32.
14. Lobo FS, Dossi MO, Batista L, Shinzato MM. Hearing impairment in patients with rheumatoid arthritis: association with anti-citrullinated protein antibodies. Clin Rheumatol 2016;35:2327-32.
15. Emamifar A, Hansen IMJ. An update on hearing impairment in patients with rheumatoid arthritis. J Otol 2018;13:1-4.
16. Chaitidis N, Theocharis P, Festas C, Aritzi I. Association of rheumatoid arthritis with hearing loss: a systematic review and meta-analysis. Rheumatol Int 2020;40:1771-9.
17. Takatsu M, Higaki M, Kinoshita H, Mizushima Y, Koizuka I. Ear involvement in patients with rheumatoid arthritis. Otol Neurotol 2005;26:755-61.
18. Milisavljevic D, Stankovic M, Zivic M, Radovanovic Z, Stankovic P. Changes in auditory ossicles in rheumatoid arthritis: scanning electron microscopic study. Eur Arch Otorhinolaryngol 2010;267:363-6.
19. Jeong H, Eun YH, Park EJ, Chae JY, Kim H, Lee J, et al. Rheumatoid arthritis is associated with low-/mid-frequency hearing impairment: data from the Korean National Health and Nutrition Examination Survey. Arthritis Rheumatol 2016;68(10 Suppl):1551.
20. Oztürk A, Yalçin S, Kaygusuz I, Sahin S, Gök U, Karlidag T, et al. High-frequency hearing loss and middle ear involvement in rheumatoid arthritis. Am J Otolaryngol 2004;25:411-7.
21. Salvinelli F, Cancilleri F, Casale M, Luccarelli V, Di Peco V, D'Ascanio

L, et al. Hearing thresholds in patients affected by rheumatoid arthritis. Clin Otolaryngol Allied Sci 2004;29:75-9.
22. Goodwill CJ, Lord IJ, Jones RP. Hearing in rheumatoid arthritis. A clinical and audiometric survey. Ann Rheum Dis 1972;31:170-3.
23. Rosenberg JN, Moffat DA, Ramsden RT, Gibson WP, Booth JB. Middle ear function in rheumatoid arthritis. Ann Rheum Dis 1978;37:522-4.
24. Poorey VK, Khatri R. Study of auditory function in rheumatoid arthritis. Indian J Otolaryngol Head Neck Surg 2001;53:261-3.
25. Halligan CS, Bauch CD, Brey RH, Achenbach SJ, Bamlet WR, McDonald TJ, et al. Hearing loss in rheumatoid arthritis. Laryngoscope 2006;116:2044-9.
26. Tavernier L, Ranfaing E. Rheumatoid arthritis involved in the ear drum. Otol Neurotol 2011;32:e5-6.
27. Pascual-Ramos V, Contreras-Yáñez I, Enríquez L, Valdés S, Ramírez-Anguiano J. Hearing impairment in a tertiary- care-level population of Mexican rheumatoid arthritis patients. J Clin Rheumatol 2012;18:393-8.
28. Ahmadzadeh A, Daraei M, Jalessi M, Peyvandi AA, Amini E, Ranjbar LA, et al. Hearing status in patients with rheumatoid arthritis. J Laryngol Otol 2017;131:895-9.
29. Walling AD, Dickson GM. Hearing loss in older adults. Am Fam Physician 2012;85:1150-6.
30. Magliocca KR, Vivas EX, Griffith CC. Idiopathic, infectious and reactive lesions of the ear and temporal bone. Head Neck Pathol 2018;12:328-49.
31. Jacobs RP, Moore M, Brower A. Wegener's granulomatosis presenting with erosive arthritis. Arthritis Rheum 1987;30:943-6.
32. Sodhi A, Naik S, Pai A, Anuradha A. Rheumatoid arthritis affecting temporomandibular joint. Contemp Clin Dent 2015;6:124-7.
33. Tyndel FJ, Davidson GS, Birman H, Modzelewski ZA, Acker JJ. Sarcoidosis of the middle ear. Chest 1994;105:1582-3.
34. Chen R, Schwander M, Barbe MF, Chan MM. Ossicular bone damage and hearing loss in rheumatoid arthritis: a correlated functional and high resolution morphometric study in collagen-induced arthritic mice. PLoS One 2016;11:e0164078.
35. Yildirim A, Surucu G, Dogan S, Karabiber M. Relationship between disease activity and hearing impairment in patients with rheumatoid arthritis compared with controls. Clin Rheumatol 2016;35:309-14.