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Sensorineural hearing loss associated with psoriatic arthritis

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Abstract
Autoimmune inner ear disease is a well described entity. We report a case of sudden-onset sensorineural hearing loss in association with psoriatic arthritis, which has not been reported in the literature. The case satisfies the criteria for the presumptive diagnosis of autoimmune hearing loss. A high index of suspicion, with early diagnosis and aggressive treatment with steroids and/or immunosuppressive agents, is essential to prevent irreversible hearing loss. The condition of psoriatic arthritis must be added to the pantheon of autoimmune diseases that can lead to sensorineural hearing loss.

Key words: Autoimmune Disease; Hearing Loss, Sudden; Arthritis, Psoriatic

Introduction

McCabe first described the association of sensorineural hearing loss with autoimmune disorders in 1979.[1] There have since been several reports in the literature of sensorineural hearing loss (SNHL) with autoimmune diseases like ulcerative colitis, rheumatoid arthritis, Wegener's granulomatosis and giant cell arteritis.[2–5] We report a case of sudden-onset SNHL with psoriatic arthritis, another autoimmune disease, but one whose association with SNHL has not been previously reported. The literature is reviewed, and we recommend the acceptance of criteria to help identify autoimmune hearing loss.[6] Early diagnosis is essential in this treatable condition before hearing loss becomes irreversible.

Case report

A 62-year-old man presented with sudden-onset, right-sided hearing loss with poor speech discrimination and tinnitus. He had no vertigo. There was no preceding upper respiratory tract infection or head injury. He suffered from psoriatic arthritis, which had been diagnosed many years previously, and was well controlled with methotrexate. At presentation he was receiving methotrexate.

Clinical examination revealed normal tympanic membranes and a positive Rinne test in both the ears with the Weber lateralized to the left ear. Cranial nerve and neuro-otological examinations were normal. Pure tone audiometry (Figure 1) showed hearing thresholds of 50 dB. in the right ear (average thresholds at 0.5, 1, 2 kHz.) with normal hearing in the left ear.

Impedance audiometry showed normal compliance. Serum electrophoresis showed increased gamma globulins, and concentrations of C3 and C4 components of the complement system were raised, as was the erythroctye sedimentation rate (ESR) (73 mm/hr). The rheumatoid factor, antinuclear antibodies and ANCA tests were negative, as was the Otoblot test (antibodies to 68-kDa inner ear heat shock protein). A magnetic resonance imaging (MRI) scan of internal auditory meati, brain and cerebellopontine angle was normal.

The patient was treated with oral prednisolone (60 mg daily) for 3 weeks before tapering off and completion of treatment in the fourth week. At the end of the 4 weeks his hearing thresholds were nearly normal (Figure 2) and tinnitus had completely resolved. Three months later, the patient had not suffered any recurrence of hearing loss.

Discussion

The association between psoriatic arthritis and sensorineural hearing loss has not been described, although reports exist in the literature of toxic inner ear damage as a result of topical treatment of psoriatic arthritis with salicylates.[7] Our patient did not receive such treatment. Autoimmune inner ear disease is a recently recognized entity. It usually presents as bilateral sensorineural hearing loss occurring over weeks to months. It is also associated in some instances with balance disturbance and tinnitus and is more common among middle-aged women. It is seen to occur in association with autoimmune diseases. Early recognition and treatment with steroids and/or immunosuppressive agents leads to recovery of hearing loss, at least in part,[8] particularly in patients with mild hearing loss (<50 dB.).[6]

The inner ear is an immunoresponsive organ. These responses could be mediated via the immunocompetent cells recruited from either the endolymphatic sac or the systemic circulation. IgG is the predominant immunoglobulin found in the perilymph, with lesser amounts of IgM and IgA.[9] Following an inflammatory injury there is entry of leucocytes from the circulation into

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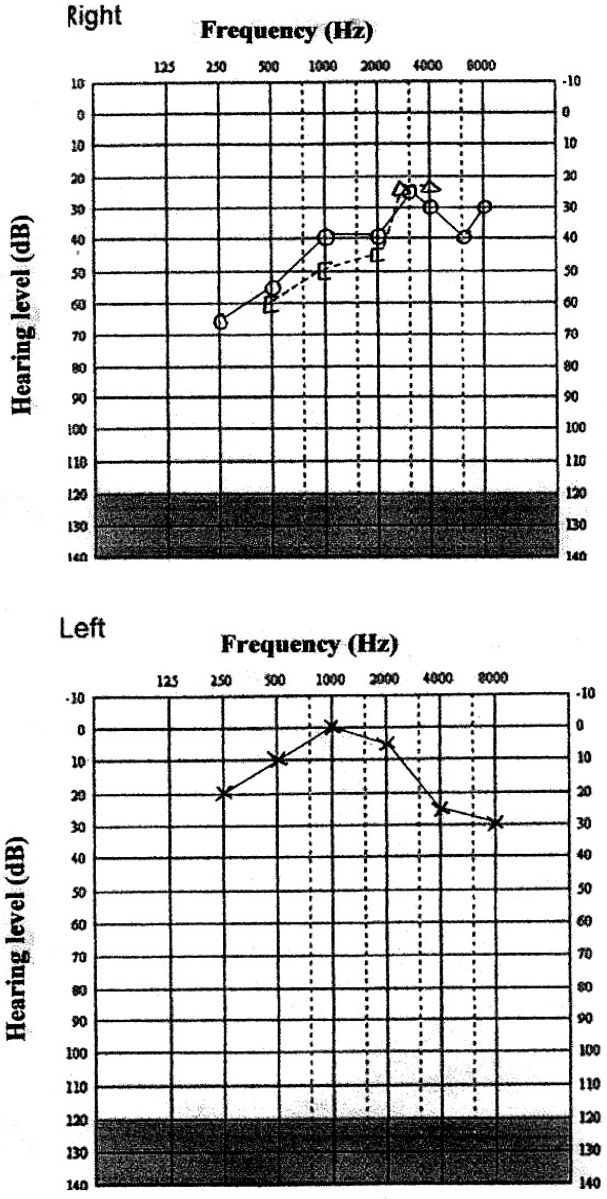

FIG. 1
Pre-treatment pure tone audiogram.

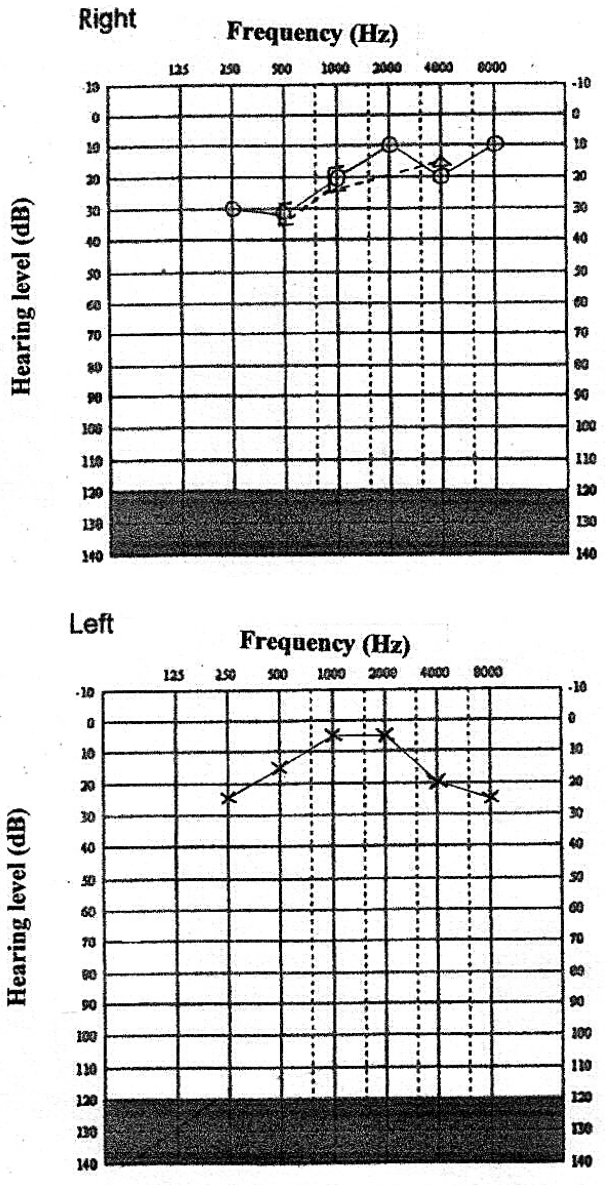

FIG. 2
Post-treatment audiogram showing hearing improvement in right ear.

the cochlea. The accumulation of leucocytes and local production of immunoglobulins incites an inflammatory reaction, resulting in degeneration of the organ of corti, stria vascularis and spiral ganglion, with resultant sensorineural hearing loss.[10] A postulated mechanism for autoimmune inner ear disease is reversible damage to cellular components of the organ of Corti.[6] Several other mechanisms have also been proposed. These include damage resulting from immune complex deposition and vasculitis, cross-reacting antibodies, T lymphocyte-mediated cytotoxicity, and low-grade tissue injury due to circulating pro-inflammatory cytokines in inflammatory bowel disease.[3,11–13]

The complement system is activated, either through the classic or alternate pathway. It has been suggested that activation of the complement system is part of the systemic inflammatory reaction in which the inner ear demonstrates the only clinical dysfunction.[10] This could explain the raised IgG, C3, C4 and ESR in our patient while also accounting for the absence of other systemic symptoms and negative immune markers.

It is interesting to note that laboratory diagnoses for autoimmune hearing loss are still inconclusive and non-specific. Serological markers for active and inactive immune processes include antinuclear antibodies, ANCA, RA factors, antibody against type II collagen, and other auto-antibodies, including non-specific haematological tests like ESR and CRP. However, none of these are specific to immune disorders, unlike the serological marker for allergy, which is readily identified by a raised titre of total and specific IgE. A recent study demonstrated that there is no correlation between autoimmune sensorineural hearing loss and heat shock protein (HSP) 70 (antibodies to KHRI-3 cochlear protein [68 kDa]).[14] This is unfortunate as this is the only test available for clinical use. Of course, HSP 70 is only one of many proteins cross-reacting against the inner ear in suspected immune-mediated hearing loss. Heat shock proteins can also be induced by a variety of stimuli, e.g. stress, ischaemia, free radicals and acoustic overstimulation.[10] Measurement of these proteins would

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only support the diagnosis of immune-mediated hearing loss if occurring in association with systemic autoimmune diseases. Various immunological studies have tried to identify various antibodies that specifically cross-react with inner ear antigens,[15–17] but this approach remains experimental at present. Unfortunately, as in the case described, the negative ANCA and HSP 70 (antibody to 68 kDa) tests do not preclude autoimmune mechanisms.

- **Case report of a patient with psoriatic arthropathy who developed sudden-onset mild-to-moderate sensorineural hearing loss**
- **The patient recovered on steroids**
- **The diagnosis is presumptive but based on existing criteria for immune-mediated sensorineural hearing loss**
- **Patients with psoriatic arthropathy may be prone to sudden or rapidly progressive sensorineural deafness; the prevalence of this is unknown**

Studies have shown that hearing loss can be the only initial manifestation of autoimmune disease, as noted in polyarteritis nodosa.[9] It can also occur in disease remission, as seen in ulcerative colitis.[2] Furthermore, studies have shown that in systemic autoimmune diseases such as ulcerative colitis, systemic lupus erythematosus and rheumatoid arthritis, there is no correlation between sensorineural hearing loss, age, sex, disease duration or activity, articular and extra-articular manifestations and presence of auto-antibodies.[9]

The diagnosis of autoimmune inner ear disease is still largely presumptive. However, recent papers have proposed a diagnostic profile for immune-mediated sensorineural hearing loss.[6] Major criteria proposed are: bilateral hearing loss, systemic autoimmune disease, raised titres of antinuclear or other antibodies, and complete recovery of hearing with steroid treatment. The minor criteria are: unilateral hearing loss, serum reactivity to HSP 70 and partial recovery of hearing with steroid treatment. The presence of three major criteria or two major and two minor criteria would support the diagnosis of immune-mediated hearing loss. The patient we report had two major criteria, with evidence of raised immune function (elevated IgG levels, ESR and complement) and the presence of psoriatic arthritis, a well known systemic immune disease. In addition, the presence of two minor criteria in our patient (partial hearing recovery, affected unilaterally) would fit the diagnosis of autoimmune hearing loss.

A recent review of autoimmune hearing loss by the senior author (BNK)[9] listed a variety of systemic diseases, including rheumatoid arthritis, but there is no mention in the literature of hearing loss with psoriatic arthritis. This case report illustrates the association of sensorineural hearing loss with psoriatic arthritis and this should be borne in mind by the otolaryngologist, as early and aggressive treatment with steroids may result in recovery of hearing.

References

1 McCabe BF. Autoimmune sensorineural hearing loss. *Ann Otol Rhinol Laryngol* 1979;**88**:585–9
2 Kumar BN, Walsh RM, Wilson PS, Carlin WV. Sensorineural hearing loss and ulcerative colitis. *J Laryngol Otol* 1997;**11**:277–8
3 Weber RS, Jenkins HA, Coker NJ. Sensorineural hearing loss associated with ulcerative colitis. *Arch Otolaryngol* 1984;**110**:810–12
4 Luqmani R, Jubb R, Reid A, Adu D. Inner ear deafness in Wegener's granulomatosis. *J Rheumatol* 1991;**18**:766–8
5 Jacob A, Ledingham JG, Kerr AIG, Ford MJ. Ulcerative colitis and giant cell arteritis associated with sensorinerual deafness. *J Laryngol Otol* 1990;**104**:889–90
6 García Berrocal JR, Ramírez Camacho R, Millán I, Górriz C, Trinidad A, Arellano B, *et al*. Sudden presentation of immune-mediated inner ear disease: characterization and acceptance of a cochleovestibular dysfunction. *J Laryngol Otol* 2003;**117**:775–9
7 Maune S, Frese KA, Mrowietz U, Reker U. Toxic inner ear damage in topical treatment of psoriasis with salicylates. *Laryngorhinootologie* 1997;**76**:368–70
8 Summers RW, Harker L. Ulcerative colitis and sensorineural hearing loss: is there a relationship? *J Clin Gastroenterol* 1982;**4**:251–2
9 Mathews J, Kumar BN. Autoimmune sensorineural hearing loss. *Clin Otolaryngol* 2003;**28**:479–88
10 García Berrocal JR, Ramírez-Camacho, R. Sudden sensorineural hearing loss: supporting the immunological theory. *Ann Otol Rhinol Laryngol* 2002;**111**:989–97
11 Kumar BN, Smith MSH, Walsh RM, Green JR. Sensorineural hearing loss in ulcerative colitis. *Clin Otolaryngol* 2000;**25**:143–5
12 Kanzaki J, O-Uchi T. Circulating immune complexes in steroid-responsive sensorineural hearing loss and the long term observation. *Acta Otolaryngol Suppl Stockh* 1983; **393**:77–84
13 Brookes GB. Immune complex deafness: preliminary communication. *J Roy Soc Med* 1985;**78**:47–55
14 Yeom K, Gray J, Nair TS, Arts HA, Tellian AS, Disher MJ, *et al*. Antibodies to HSP-70 in normal donors and autoimmune hearing loss patients. *Laryngoscope* 2003; **113**:1770–6
15 Mathews J, Rao S, Kumar BN. Autoimmune sensorineural hearing loss: is it still a clinical diagnosis. *J Laryngol Otol* 2003;**117**:212–4
16 Harris JP, Sharp PA. Inner ear autoantibodies in patients with rapidly progressive hearing loss. *Laryngoscope* 1990; **100**:516–24
17 Kastanioudakis I, Skevas A, Danielidis V, Tsiakou E, Drosos AA, Moustopoulos MH. Inner ear involvement in rhematoid arthritis: a prospective clinical study. *J Laryngol Otol* 1995;**109**:213–18

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