Autoimmune inner ear disease associated with ankylosing spondylitis

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We report here a case study of a 24-year-old man suffering from autoimmune inner ear disease who also suffered from ankylosing spondylitis (AS). Although he was previously diagnosed with AS, he has now presented with sensorineural hearing loss and vertigo. It is the first case that a patient with AS displays symptoms of autoimmune inner ear disease. His symptoms responded well to prednisolone treatment. However, because weight gain is a common side effect of steroid treatment and may exacerbate the lower back pain, which is symptomatic of AS, he has so far been treated with a minimum dosage of prednisolone. In addition, intratympanic administration of dexamethasone was effective to suppress the exacerbation of deafness.

Keywords:
ankylosing spondylitis, autoimmune inner ear disease, hearing loss, prednisolone, vertigo

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Introduction

Autoimmune sensorineural hearing loss (SNHL) was first described by McCabe in 1979 [1]. Recently, this disease has become more widely known and specific cases have been reported [2,3]. Moreover, autoimmune SNHL with a vestibular dysfunction has been also reported and defined as autoimmune inner ear disease (AIED) [4]. It was reported that the patients with autoimmune SNHL often suffered from other autoimmune diseases [5]. However, there is no patient with both autoimmune SNHL and AS in these studies. There were however no examples of patients with ankylosing spondylitis (AS) among these 72 patients. In addition, the other such reports did not reveal any patients with AS [6,7]. It is very rare that a patient with AS displays symptoms of autoimmune SNHL or AIED.

We report the first case of a patient with AS presenting with AIED in the course of his disease.

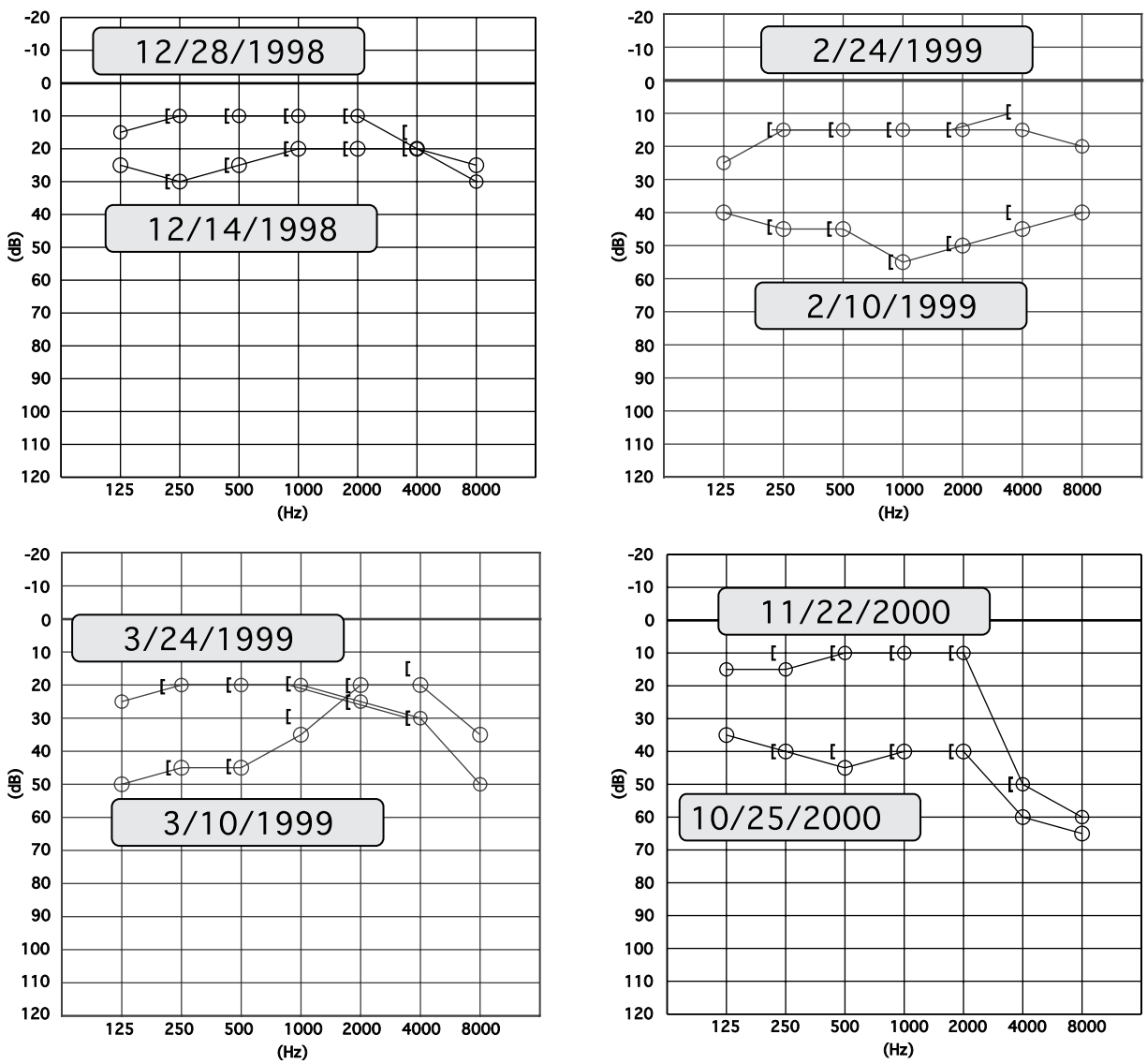
Case report

A 24-year-old man with a previously diagnosed AS was also suffering from repeated vertigo, tinnitus and ear fullness on the right side and was treated in our hospital. He suffered from general fatigue and a low-grade fever and had been treated by a general practitioner since February 1997. He was admitted to the Third Department of Internal Medicine in our hospital in November 1997. At that time, a physical examination revealed a limited mobility of the cervical and lumbar spine, and radiographs revealed bilateral sacroiliitis. The peripheral WBC count was 6900 cells/μl and the erythrocyte sedimentation rate was 90 mm/h. Laboratory tests revealed a C-reactive protein (CRP) level of 2.95 mg/dl. Immunoglobulin IgG fractions were elevated at 3230 mg/dl, and IgA and IgM fractions were within normal limits. The homolytic complement activity (CH50) was elevated at 60.6 U/ml and HLA typing was positive for B-27 and CW-7. On the basis of these findings, he was then diagnosed with AS [8]. Therefore, he started to take indomethacin (200 mg/day) orally. There was no patient suffering from the same symptoms in his family history.

During follow-up examinations in our outpatients' clinic in June 1998, he complained of a stuffed ear and tinnitus in his right ear. Laboratory tests revealed a CRP level of 3.48 mg/dl, and his drugs were changed to Salazosulfapyridine (1000 mg/day) and sodium diclofenac (200 mg/day). The symptoms in his ear did not disappear, and he was transported to the Department of Otolaryngology in our hospital by ambulance on 13 December 1998 after he had a vertigo attack. At the time of admission, a continuous right-beating nystagmus was seen using Frenzel goggles. Pure tone audiometry showed a mild right-side SNHL (Fig. 1a). The auditory brainstem response revealed the prolongation of a peak 1 latency and the elevation of the threshold on the right side. Distortion product otoacoustic emission amplitudes with frequencies of 1, 2 and 4 kHz were decreased on the right side. Caloric testing revealed a right canal paresis of 33%. There were no abnormal findings from magnetic resonance imaging of the head. The vertigo and hearing loss was ameliorated after the administration of ATP and vitamins, and he was discharged after 2 days at his own request.

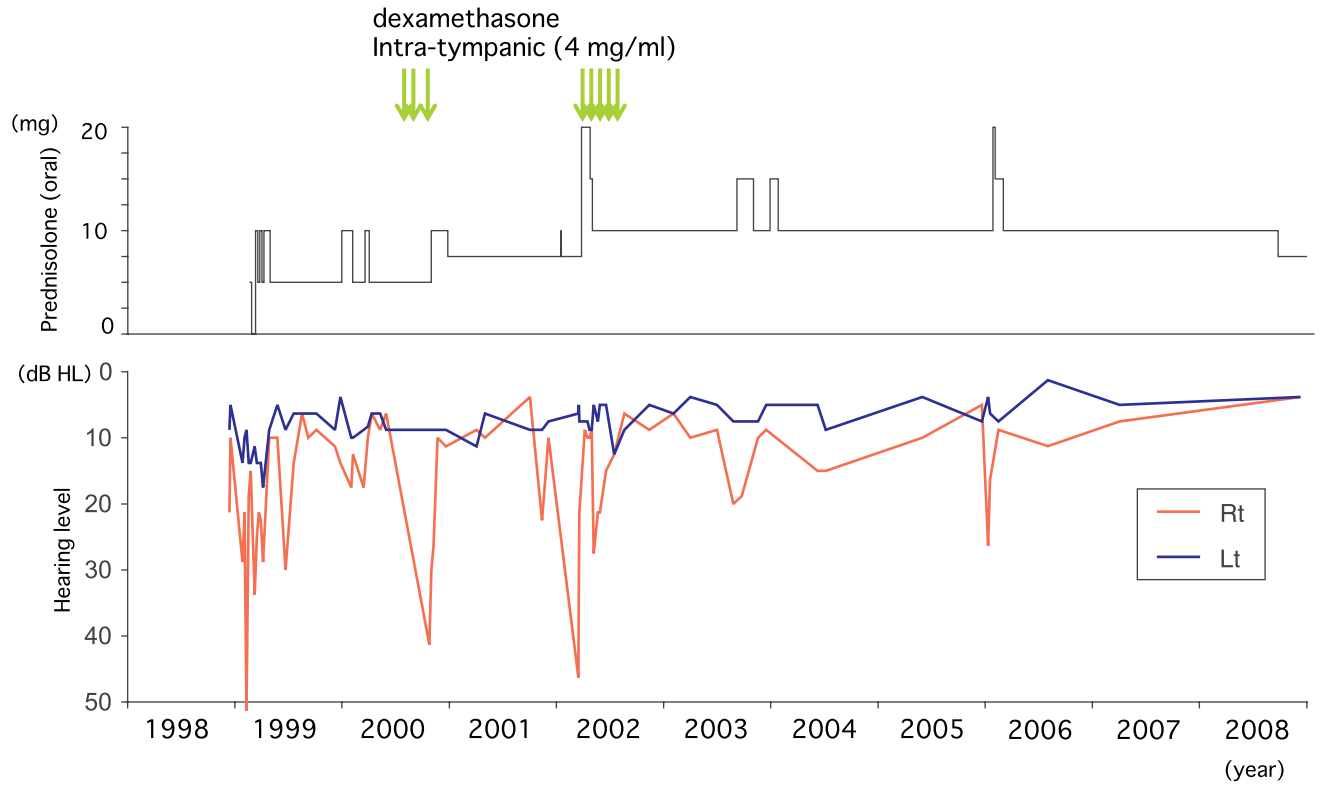
The symptoms however were recrudescent and he visited our outpatient clinic again in January 1999. He was prescribed ATP and vitamins, and his symptoms did not disappeared. When he was prescribed prednisolone (10 mg/day), his symptoms were ameliorated for 1 week and his hearing improved to almost normal levels (Fig. 1b). Prednisolone dosage was therefore decreased gradually and discontinued on 24 February 1999. Two weeks later, however, he suffered a repeat of hearing loss and another attack of vertigo. The administration of prednisolone was restarted and the patient's hearing improved on 24 March 1999 (Fig. 1c). At this time, an AIED was suspected on the basis of his steroid-responsive symptoms and the presence of AS. Since this diagnosis, he has continued to take prednisolone up to the present time, the dosage being adjusted at different times depending on the severity of his symptoms. The patient presented again with hearing loss in October 2000, and his prescribed dose of prednisolone was increased to 15 mg/day, but hearing level did not improved. Therefore, the intratympanic infusions of dexamethasone were performed, resulting in improved hearing for a period of 2 weeks (Fig. 1d). Since then, the dosage of prednisolone ranged from 5 to 15 mg/day, while the auditory function of the patient was monitored carefully. When the control of hearing was difficult only with the increasing dosage of prednisolone, the intratympanic administrations of dexamethasone were used in 2002. His symptoms have subsided with a continuing internal dosage of prednisolone (7.5 mg/day) up to the present time (August 2012) and the CRP level stays under 1.0 mg/dl (Fig. 2).

Figure 1



Pure tone audiogram of the right ear air conduction at the time of admission, 14 December 1998, and at discharge, 17 December 1998; at the maximum hearing deterioration, 10 February 1999, and at the recovery 2 weeks after steroid treatment commenced, 24 February 1999.

Figure 2



Pure tone audiogram of the right ear air conduction following cessation of steroid administration, 10 March 1999, and 2 weeks following the commencement of steroid treatment, 24 March 1999; before increased steroid dosage, 25 October 2000, and 4 weeks after increased steroid dosage, 22 November 2000.

Discussion

Patients with autoimmune SNHL often suffer from other autoimmune diseases [5]. There were few reports on patients with AS presenting autoimmune SNHL, although many cases of autoimmune SNHL were reported previously. Djupesland *et al.* [9] reported the cases of SNHL associated with AS. However, they could not confirm that the hearing loss was related to the mechanisms of autoimmunity. Only three patients with AS who were diagnosed as autoimmune SNHL were reported previously [10–12]. In these three patients, the fact that hearing improved after the treatment with steroids suggested that their hearing losses were concerned with immunological mechanism. In our case study presented here, the patient developed not only hearing loss, but also vertigo, and we diagnosed the patient as having AIED [4]. This present case is therefore the first report of AIED in AS. Previous reports showed that autoimmune SNHL often accompanied autoimmune diseases such as Cogan's syndrome and rheumatoid arthritis [5–7]. On the basis of our findings in this report, we propose that there should be careful monitoring of both hearing and vestibular functions when treating patients with other autoimmune diseases.

A few patients suffering from AS also suffer from conductive hearing loss, and the mobility of ear ossicles of these patients was assumed to be limited with the arthritis of the ear ossicles [13,14]. The reason why the arthritis of the ear ossicles appeared in some patients was unclear. In addition, it was reported that the middle ear involvement in patients with AS is very uncommon (<1% of ears in patient with AS) [15]. In our case, the hearing examinations of the patient have never shown conductive hearing loss, although he underwent the frequent hearing examinations.

AS is an autoimmune disease that affects the articulations and the ambient tissues near the trunk. Almost all patients present with consistent symptoms such as pain and stiffness of the sacroiliac joints at the back of the pelvis in their 30s, and these symptoms gradually become more severe. Nonsteroidal anti-inflammatory drugs were often administered as a first-line drug to patients with AS to reduce inflammation and relieve pain, whereas steroids were rarely used because weight gain, a common side effect of steroid use, may exacerbate the symptoms. In contrast, patients with an AIED should be treated immediately with a high dose of steroids, once a diagnosis of AIED is considered likely [4]. In three patients who were reported as autoimmune SNHL with AS, the maximum doses of

prednisolone were 50 and 60 mg/day, respectively [10,11]. When we treated the patient in this case study, we tried to prescribe a low dose of prednisolone (15 mg/day) so as not to promote weight gain. Although we had to increase the dose of prednisolone temporarily because of further deterioration in his hearing, we tried to ensure that the lowest dose possible was administered. Because his symptoms had not yet advanced, we believe that our choice of treatment was appropriate and successful in this case.

It is often difficult to reduce the maintenance dose of prednisolone. In the previous three cases of autoimmune SNHL in patients with AS, the physicians had trouble in reducing the dosage of prednisolone because of the relapse of symptoms [10–12]. Their final maintenance doses were 12.5 and 20 mg/day. In one of the cases, the hearing was impaired after a few deteriorations of symptoms [10]. Therefore, we cannot reduce the dosage furthermore while we monitor his auditory function.

Cytotoxic drugs [15–17] were used with prednisolone for autoimmune diseases. Some otolaryngologists reported the effect of azathioprine on the hearing function of the patients suffering from autoimmune SNHL [18,19]. In addition, azathioprine was often used as a steroid-sparing agent, and many cases of steroid-sparing effects were reported in autoimmune diseases [20,21]. Therefore, we should ponder whether we administer cytotoxic drugs in combination with steroids, when the maintenance dose of prednisolone is getting more.

Recently, intratympanic steroids have been used for the treatment of inner ear diseases. Intratympanic dexamethasone application results in significantly higher perilymph steroid concentrations than those achieved by means of a systemic route [22]. Dodson *et al.* [23] reported that the topical application of steroids could control the vertigo attack in patients suffering from Ménière's disease. Gouveris *et al.* [24] reported that intratympanic administration of steroids could be the therapeutic option for the treatment of patients with idiopathic sudden SNHL who do not respond to intravenous steroid and vasoactive therapy. In addition, the topical administration of steroids was used to protect the auditory function of the patients with AIEDs [25,26]. In our case, we should reduce the dose of systemic administration of prednisolone to prevent the weight gain of the patient. Therefore, the combination of the treatment, topical and systemic administration of steroids, was useful therapy.

Acknowledgements

Conflicts of interest

None declared.

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