CASE REPORT

Sudden sensorineural hearing loss: A missed opportunity for treatment

Ali AH, Salahuddin Z, Salim R

Ali AH, Salahuddin Z, Salim R. Sudden sensorineural hearing loss: A missed opportunity for treatment . *Malays Fam Physician.* 2018;13(3); 29–31.

Keywords:
Anticardiolipin antibody;
Antiphospholipid syndrome;
Hearing loss, sudden;
Sensorineural hearing loss;

Authors:

Ahmad Hafiz Ali
(Corresponding author)
(MBBCh BAO)
Department of Otorhinolaryngology-Head & Neck Surgery
School of Medical Sciences
Universiti Sains Malaysia
16150 Kota Bharu, Kelantan
Malaysia
Email: matkurt@gmail.com

Zulkiflee bin Salahuddin
(MD, MS ORL HNS)
Department of Otorhinolaryngology-Head & Neck Surgery,
Hospital Raja Perempuan Zainab II
Kelantan, Malaysia

Rosdan Salim
(MD, MMED ORL-HNS)
Department of Otorhinolaryngology-Head & Neck Surgery
Universiti Sains Malaysia
Kelantan, Malaysia

Abstract

Sudden sensorineural hearing loss (SSNHL) is an otology emergency and carries significant morbidity if the diagnosis is missed. It can present to any specialty but in our local setting the patient usually presents to primary care as it is easily accessible. We present a case of SSNHL that was initially presented to a primary care centre and the patient was reassured without any investigation being carried out. SSNHL has many causes thus making diagnosis difficult. However, with knowledge of its possible, a diagnosis can be made and appropriate management can be advocated to the patient. Hence, we discuss the three main causes of SSNHL, while emphasizing the immune system-mediated mechanism as the main cause in this case.

Introduction

Sudden sensorineural hearing loss (SSNHL) is an otology emergency and the estimated incidence varies from 5 to 20 cases per 100,000 individuals per year[1]. However, there was a marked increase in the number of elderly Japanese receiving treatment for SSNHL in 1970s, 1980s, and 1990s.[1] It occurs equally among males and females, and affects the population between 50 to 60 years old. In most cases, the hearing loss is unilateral but about 5% of the cases are bilateral.[2]

Patient with SSNHL may present to different specialties, such as otorhinolaryngology, internal medicine, or primary care. In primary care or a non-otorhinolaryngology specialty, patient can be easily mis-diagnosed and inappropriately reassured without proper investigation.

The three main causes of SSNHL are viral infection, vascular occlusion and an immune system-mediated mechanism. In this case report, we want to highlight these causes and how the case was mis-diagnosed.

Case Report

A 35-year old woman presented to her obstetrician for per vaginal bleed. She was found to be 10 week pregnant and currently having a miscarriage. It was her second miscarriage. She was referred to our department for sudden-onset, right-sided hearing loss associated with intermittent tinnitus for the past two days. She denied any trauma, ear discharge, or vertigo. She also had a similar episode of sudden-onset hearing loss about six months prior to this presentation and sought treatment at a primary care centre but was reassured as the symptoms only lasted about 24 hours. Her obstetric history revealed that she had miscarried during her first pregnancy about a year ago at around 12 weeks gestation. She denied any history of infectious disease, any vascular abnormalities, bleeding disorders, rashes, or any symptoms that might suggesting connective tissue disease.

Her general physical examination was normal, with no rashes or bruises seen. Her ophthalmology and otoscopy examinations were also normal. Pure tone audiometry (Figure 1) was done and showed a moderate-to-profound mixed hearing loss on the right ear as there were conductive hearing loss element at 250 Hz and 500 Hz and severe-to-profound hearing losses at frequencies 1, 2, 4 and 8 kHz. The hearing threshold for the left ear was normal. A computed-tomography scan was done and produced normal findings. She was treated for sudden sensorineural hearing loss. She was started on one 60 mg tablet of prednisolone for one week followed by tapering doses.

Her blood test results, coagulation profile, renal and liver tests were all normal. However due to limitation of facilities, all the immunology investigations were referred to another centre and results were received two weeks later. All the immunology and serology tests including antinuclear antibody, complement (C3 and C4) tests, VDRL, anti-Ro/SSA, anti-La/SSB,

Rheumatoid factor and antilupus were negative except for the test for anti-cardiolipin antibodies, which was 60 (normal value < 16 UGPL). She was started on an anti-platelet agent in view of that positive result. During the follow-up 12 weeks later, the hearing symptoms and repeated pure tone audiogram remained the same.

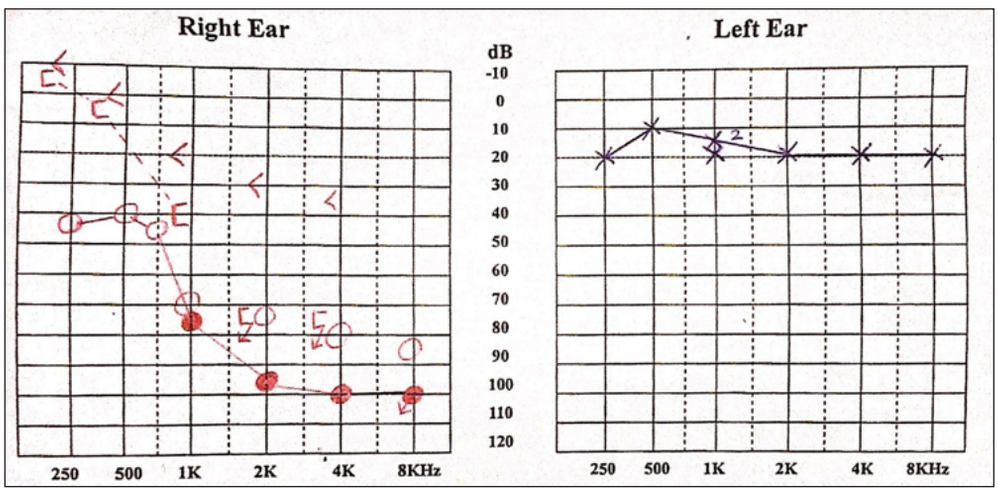

Figure 1 : Pure tone audiogram

Discussion

SSNHL is defined as acute hearing loss of 30 dB or more, in at least three consecutive frequencies, that develops within hours up to three days in duration.[1] The three main causes of SSNHL are viral infection, vascular occlusion and an immune system-mediated mechanism. It is very difficult to confirm the specific cause that contributes to each patient's hearing loss as detailed investigations reveal a specific cause only 10% of the time.[3] The remainder of the cases are labelled as idiopathic in aetiology.

In the case of viral infection causing SSNHL, the exact mechanisms are still unclear. Various viruses can cause this illness such as mumps, the Epstein-Barr virus, herpes simplex type I and II viruses or cytomegalovirus, and as well as enteroviruses.[3] Thus, any patient presenting to a primary care centre must be thoroughly examined, as only a small group of patients will have hearing loss, and SSNHL can be easily missed. As in our patient, frequently only an otoscopy examination is done at a primary care centre and no tuning fork test is conducted. It is also crucial to detect and treat SSNHL as early as possible because only 9% of patients will have significant recovery if treatment starts more than five days after a diagnosis of SSNHL is made.[4]

The cochlea is supplied by the terminal end of the vascular system. Any pathophysiological changes that lead to the narrowing of the vessel are thought to cause SSNHL. Therefore, general cardiovascular risk factors that a patient has a higher risk of SSNHL.[5] Other conditions, such as stroke and hypotension can also cause SSNHL because of the reduced blood flow to the cochlea.[3]

As far as our patient was concerned, the immune system-mediated mechanism causing her SSNHL was related specifically to her antiphospholipid syndrome. The pathogenesis of immune system-mediated hearing loss has been widely studied, but the exact mechanism remain unclear. Several mechanisms have been postulated which involves the amplification of autoantibodies production against the exposed phospholipid membrane lining the cell, therefore damaging the cell. The other mechanisms have been proposed include the hypercoagulable effect and also the increased complement activation in antiphospholipid syndrome, hence causing thrombosis in the cochlea vascular system resulting in SSNHL.[2] Our patient also had recurrent miscarriages, which it might suggest the presence of thrombosis in her decidua basalis.

In most of the immune system-mediated system hearing losses, the patient may have other immune-mediated diseases such as systemic lupus erythematous,[6] rheumatoid arthritis,[7] and relapsing polychondritis.[8] A study showed that 27% of patients with SSNHL had positive anticardiolipin antibodies, which indicates the possibility of ongoing immune system-mediated disease.[9] However, the cause

of sudden SSNHL in our patient can be safely be pinpointed as towards the primary antiphospholipid syndrome as there was no other manifestation of immune-mediated disease.

Conclusion

Patients with SSNHL will usually present to a primary care centre. It is important for the primary care physician to thoroughly examined and investigate the patient as this illness can be caused by many factors. Any possible diagnosis must be entertained, especially if the hearing loss is a sudden onset, occurs in a patient in a younger age group, is recurrent or occurs in a patient in any highly suspicious risk group. In a nutshell, early detection, treatment and proper referral is important in order to prevent further morbidity in these patients and preserve a better quality of life.

References

1. Wu CS, Lin HC, Chao PZ. Sudden sensorineural hearing loss: Evidence from Taiwan. *Audiol Neurootol*. 2006 Apr 13;11(3):151-6.
2. Greco A, Fusconi M, Gallo A, Marinelli C, Macri GF, De Vincentiis M. Sudden sensorineural hearing loss: An autoimmune disease?. *Autoimmune Rev*. 2011 Oct 31;10(12):756-61.
3. Schreiber BE, Agrup C, Haskard DO, Luxon LM. Sudden sensorineural hearing loss. *Lancet*. 2010 Apr 9;375(9721):1203-11.
4. Atay G, Kayahan B. Prognostic factors in sudden sensorineural hearing loss. *Balkan Med J*. 2016 Jan;33(1):87.
5. Aimoni C, Bianchini C, Borin M, Ciorba A, Fellin R, Martini A, Scanelli G, Volpato S. Diabetes, cardiovascular risk factors and idiopathic sudden sensorineural hearing loss: A case-control study. *Audiol Neurootol*. 2009 Aug 4;15(2):111-5.
6. Hisashi K, Komune S, Taira T, Uemura T, Sadoshima S, Tsuda H. Anticardiolipin antibody-induced sudden profound sensorineural hearing loss. *Am J Otolaryngol*. 1993 Aug 31;14(4):275-7.
7. Etherington J, Cooper A, Buchanan N. Case Report Antiphospholipid syndrome in rheumatoid arthritis presenting with sensorineural deafness. *Lupus*. 1994 Feb 1;3(1):63-4.
8. Kumakiri K, Sakamoto T, Karahashi T, Mineta H, Takebayashi S. A case of relapsing polychondritis preceded by inner ear involvement. *Auris Nasus Larynx*. 2005 Mar 31;32(1):71-6.
9. Toubi E, Ben-David J, Kessel A, Podoshin L, Golan TD. Autoimmune aberration in sudden sensorineural hearing loss: Association with anti-cardiolipin antibodies. *Lupus*. 1997 Jul 1;6(6):540-2.