

Case Report

Idiopathic sequential bilateral profound sudden sensorineural hearing loss occurring 4 years after unilateral presentation

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Received: 23 June 2023

Accepted: 27 September 2023

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ABSTRACT

Sudden sensorineural hearing loss is defined as hearing loss of 30db or more at least 3 consecutive audiometric frequencies occurring within 72 hours period or less. Unilateral presentation of sudden sensorineural hearing loss is common, however bilateral sequential presentation of this disease is very rare. Clinically we couldn't anticipate the occurrence of sequential bilateral sudden sensorineural hearing loss at the first time. This bilateral sudden sensorineural hearing loss will affect the quality of life of patient. So, we report a case of 60 years old female patient with profound bilateral sudden sensorineural hearing loss with time interval of 4 years between involvement of both ears. Even though high dose steroid administered intravenously and intratympanic the patient's hearing was not restored and eventually require hearing aids. The purpose of presenting this article is that sequential profound bilateral sensorineural hearing loss may present without any warning sign, response to treatment is very poor and the rarity of this condition.

Keywords: Sensorineural, Profound, Bilateral

INTRODUCTION

Sudden sensorineural hearing loss, first reported by De Kleyn in 1944 is an otologic emergency which needs urgent intervention. Sudden sensorineural hearing loss is defined as hearing loss of 30db or more over at least 3 consecutive audiometric frequencies occurring within 72 hours period or less. Estimated annual incidence of sudden sensorineural hearing loss is 5-30 per 1,00,000 and 99% are unilateral.¹ Bilateral sudden sensorineural hearing loss is rare compared to unilateral (less than 5%).² According to time interval between onset of sudden sensorineural hearing loss in 1st affected ear and in subsequent ear, bilateral sudden sensorineural hearing loss is categorized into 2 types, simultaneous (less than 3 days) and sequential (more than 3 days interval).³ Bilateral sudden sensorineural hearing loss mostly associated with serious systemic pathologies than idiopathic aetiology. We could not anticipate occurrence of sequential bilateral sudden sensorineural hearing loss

when patient presented with unilateral sudden sensorineural hearing loss at 1st time. This type of bilateral sudden sensorineural hearing loss make patient more discomfort for verbal communications and that affect quality of life. So, we reported case of patient who had profound bilateral sudden sensorineural hearing loss with time interval of 4 years between involvement of both ears.

CASE REPORT

A 60 years old female came to our hospital with sudden onset of hearing loss on left side in 2019, with associated tinnitus and dizziness. She did not complain of any other otologic, and nasal symptoms and she had no relevant past and occupational history. On examination, tympanic membrane was normal and tuning fork test showed bilateral positive Rinne and weber lateralised to right with absolute conduction test reduced on left (Figure 1) shows the pure tone audiometry result which confirmed

sudden sensorineural hearing loss. She was admitted and started injection methylprednisolone along with antiviral.

Valacyclovir, vasodilator (xanthinol nicotinate) and multivitamin. On next day of admission, intratympanic steroid injection was administered into left ear. MRI brain with brain stem was done and normal. After 3 days pure tone audiometry was repeated, and it showed mild improvement in high frequencies. Then we discharged her with oral steroid, antiviral, vasodilator and multivitamin with a plan of intratympanic injection after 10 days. After 10 days she came with fresh pure tone audiometry, and it showed mild improvement in low and high frequencies and second dose of intratympanic steroid injection was administered. Then she reviewed after 6 weeks, and pure tone audiometry repeated it showed right hearing within normal limit and left moderate to severe mixed hearing loss with significant improvement in low frequencies (Figure 2).

After 4 years of first onset, she again came with sudden onset of hearing loss on right side, and she had discomfort in communicating with others. On examination tympanic membrane was normal and tuning fork test, Rinne positive bilaterally, weber could not appreciate, and absolute bone conduction test reduced bilaterally, and pure tone audiometry (Figure 3) showed bilateral severe to profound sudden sensorineural hearing loss (right>left). She was admitted and started injection methylprednisolone along with antiviral, vasodilator and multivitamin. Simultaneously intratympanic steroid injection also given in right ear. MRI was normal (Figure 5). Pure tone audiometry repeated after 3 days of admission it showed bilateral moderately severe to severe sensorineural hearing loss and discharged with oral medications. She reviewed on 10th day of discharge and pure tone audiometry repeated (Figure 4), showed right moderate to severe mixed hearing loss and left mild hearing impairment. However, there was not that much improvement in hearing we gave option of hearing aid.

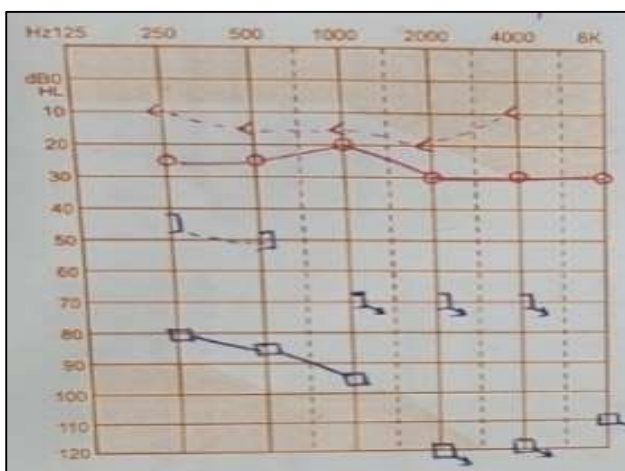


Figure 1: Pure tone audiometry report of patient at time of first onset of sudden sensorineural hearing loss on left side.

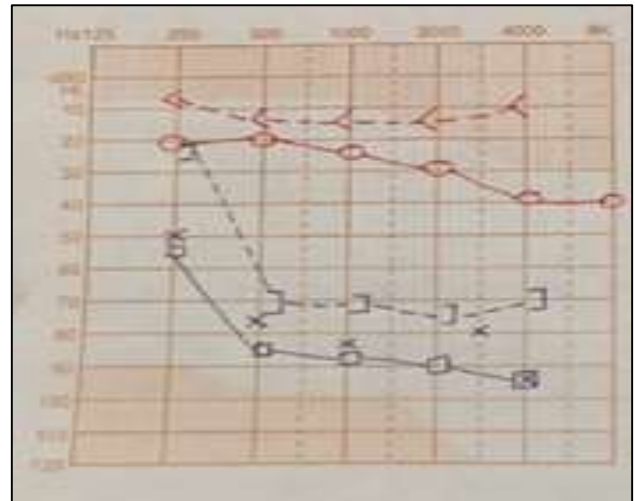


Figure 2: Pure tone audiometry report after treatment at the time of first onset of sudden sensorineural hearing loss on left side.

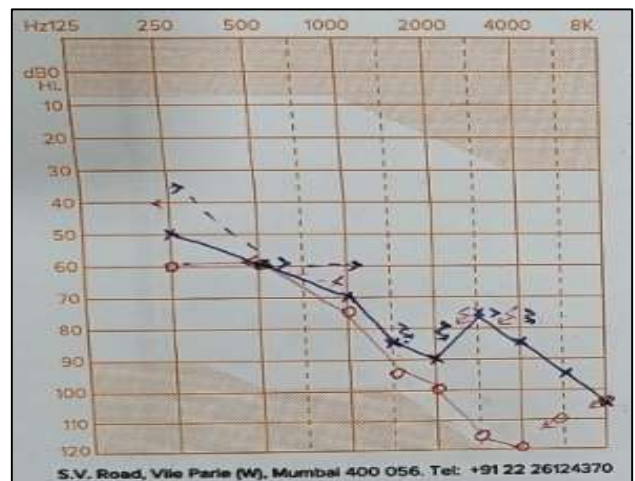


Figure 3: Pure tone audiometry report of patient after 4 years when presented with sequential onset of sudden sensorineural hearing loss on right side.

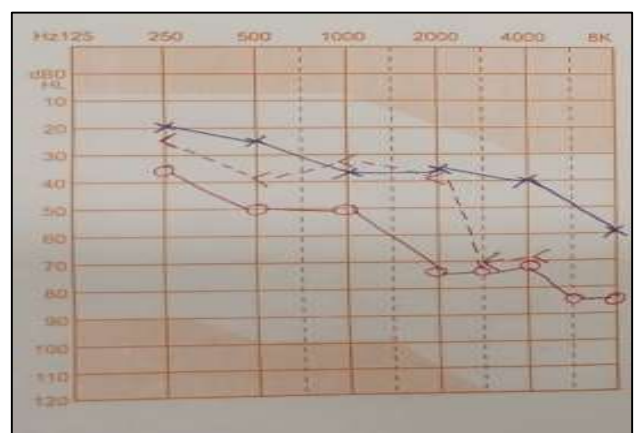


Figure 4: Pure tone audiometry report after treatment of sequential presentation of sudden sensorineural hearing loss on right side



Figure 5: Magnetic resonance image of cerebellopontine angle.

DISCUSSION

Sudden sensorineural hearing loss is an otological emergency for which a definitive aetiology and treatment remains controversial. The possible causes of sudden sensorineural hearing loss can be infection, traumatic, neoplastic, autoimmune, toxic, circulatory, neurologic, metabolic.¹ Despite the long list of possible cause, majority of cases are idiopathic. Possible causes of idiopathic sudden sensorineural hearing loss include labyrinthine viral infection, vascular insult, intracochlear membrane rupture, autoimmune inner ear disease. Majority of these cases are unilateral in their occurrence and bilateral involvement is very rare. Based on the time interval between onset of hearing loss on each side. Bilateral sudden sensorineural hearing loss classified as simultaneous and sequential, before 2007 it was not defined and was considered as a single entity. Xenellis et al first defined simultaneous and sequential sudden sensorineural hearing loss, if it is affecting both ears in less than 3 days and 3 days to one month time interval respectively and they reported 7 cases of sequential bilateral sudden sensorineural hearing loss in 2007.³ His study reported the severity of hearing loss at presentation in simultaneous bilateral disease was significantly higher than one in unilateral or sequential bilateral disease. Simultaneous bilateral sudden sensorineural hearing loss, sequential bilateral sensorineural hearing loss and unilateral sudden sensorineural hearing loss may have completely different profile and should not be investigated or managed as one disease. Some authors included the case of more than 3 months interval as sequential.^{2,4,5} It is unknown whether pathophysiology of sequential bilateral sudden sensorineural hearing loss differ from that of simultaneous bilateral sudden

sensorineural loss. We could hypothesize that certain factors could impact the cochlea bilaterally, that making the course of disease simultaneous in nature and these factors could continue to progress with a more sequential pathological course. If the onset of time interval is longer, we could consider the possibility that different kind of insults affected each side.² There are four factors that affect the prognosis of idiopathic sudden sensorineural hearing loss, age more than 60 years, associated vertigo, time since onset, audiogram with profound hearing loss and downward sloping audiogram -all of these have poor prognosis.

Many treatments regimen have been proposed like systemic or intratympanic steroid injection, hyperbaric oxygen therapy. Oh et al reported that bilateral sudden sensorineural hearing loss demonstrated a strong association with pre-existing conditions like diabetes mellitus, hypercholesterolemia.⁶ But our patient did not have any pre-existing disease. we searched the PubMed database for similar reports and found one case report of sequential idiopathic bilateral sudden sensorineural hearing loss of 37 days of first occurrence, but in our case involvement of opposite ear was 4 years after the onset of first incidence.

CONCLUSION

It is very difficult to anticipate the involvement of opposite ear when patient presented with sudden sensorineural hearing loss. So, patient should be informed about the possibility of bilateral involvement and necessary precaution to be taken and reassure about the prognosis of this condition.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Patil AA, Suchina EK. Idiopathic sequential bilateral profound sudden sensorineural hearing loss occurring 4 years after unilateral presentation. *Int J Otorhinolaryngol Head Neck Surg* 2023;9:884-7.