Sudden Sensorineural Hearing Loss (SSNHL): Case Report

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Abstract

Background
Sudden sensorineural hearing loss (SSNHL) commonly known as sudden deafness, is an unexplained, rapid loss of hearing either all at once or over a few days in which patients experience a sudden drop in hearing. It is commonly linked to infections, head trauma, autoimmune diseases, iatrogenic, severe infections, blood circulation problems, neurological disorders, such as multiple sclerosis, disorders of the inner ear, such as Ménière’s disease, and even metabolic disorders such as diabetes mellitus. We presented a case of sudden sensorineural hearing loss in a secondary hospital in Indonesia to add more knowledge on this topic.

Case presentation
A 76-year-old male, had chief complain of a sudden right sided hearing loss. There were no other symptoms and he had no history of trauma to the ear. The patient had hypertension and consumed amlodipine and clonidine regularly. Patient had normal otoscopic and rhinoscopy examination results. Audiometry shown a hearing threshold of 37 dB in the left ear and 117 dB in the right ear, both in air and bone conducted pure-tone stimuli and tympanogram showed type As in both ears. A diagnosis of right ear sudden sensorineural hearing loss was made, and medical treatment consisted of methylprednisolone, acyclovir, mecobalamin, vitamin B6, ranitidine and omeprazole were given to the patient.

Conclusion
Pathophysiology of this disease are labyrinthine viral infection, labyrinthine vascular compromise, intracochlear membrane ruptures, and immune-mediated inner ear disease. In our case, we suspect that both ischemic vascular disease and viral infection as the cause of ISHHL. If there is no definitive or treatable etiology found, treatment regimen should be dictated by the most likely factors involved, which were systemic steroids and acyclovir in our case.

Keywords: Hearing loss, sensorineural, sudden deafness

Background
The cochlea is the part of the inner ear which function is to convert mechanical energy (vibrations) into nerve impulses sent to the brain. Vestibulocochlear nerve is a nerve that is responsible for the sense of hearing and that is also pertinent to the senses of balance and body position. Problems in both cochlea and vestibulocochlear nerves can cause sensorineural hearing loss.
Incidence of SSNHL is 5-20 per 100,000 cases in the United States. The number of the incidence may be higher than these estimates because there are patients who recover quickly that they do not seek for medical care. All ages can be affected, although the peak incidence is between the fifth to sixth decade of life. Most of the cases have unilateral damage, while bilateral involvement is only 2%. Symptoms including tinnitus and dizziness are 41 to 90% and 29 to 56%.

Sudden sensorineural hearing loss is commonly linked to infections, head trauma, autoimmune diseases, exposure to certain drugs that treat cancer or severe infections, blood circulation problems, neurological disorders, such as multiple sclerosis, disorders of the inner ear, such as Ménière’s disease, and even metabolic disorders such as diabetes mellitus.

Sudden sensorineural hearing loss could decrease quality of life, so early diagnosis and prompt treatment is required in this disease. We presented a case of sudden sensorineural hearing loss in a secondary hospital in Indonesia to add more knowledge on this topic.

**Case presentation**

A 76-year-old male, working as an entrepreneur, came to the Unggul Karsa Medika Hospital and complained of a sudden right-sided hearing loss. From history taking, there were no other general nor neuro-otological symptoms such as cold, vertigo or instability, tinnitus, ear pain, otorrhea, and ear fullness. The patient had no history of trauma to the ear. The patient had a medical history of hypertension and taking amlodipine and clonidine regularly. While on physical examination, the otoscopic examination was normal, anterior rhinoscopy was normal, audiometry showed a hearing threshold of 37 dB in the left ear and 117 dB in the right ear, both in air and bone-conducted pure-tone stimuli and tympanogram showed type As in both ears. The routine blood examination was normal. The random blood sugar test was normal. Lipid profile examination showed an elevated triglycerides (167 mg/dL) and cholesterol LDL (109 mg/dL).

A diagnosis of right ear sudden sensorineural hearing loss was made. Medical treatment consisted of methylprednisolone 8 mg 4x1 for 7 days, acyclovir 400mg 3x1 for 7 days, mecobalamin 500mcg 2x1 for 7 days, vitamin B6 10 mg 2x1 for 7 days, ranitidine 150 mg 2x1 for 7 days and omeprazole 20 mg 2x1 for 7 days.

**Discussion**

There are several causes for SSNHL but the majority cases have no etiologic factors identified.
complications of the nervous system and later can cause a sudden deafness. In our case, we also suspected the patient to have viral infection or viral reactivation. Thus, we gave the patient acyclovir and methylprednisolone.

Immunologically mediated vasculitis also has a relation with the ISSHL pathogenesis. A study demonstrated that there is a role of endothelial cells promoting vasculitis by secreting cytokines. ISSHL seems to be considered as the result of abnormal activation of endocochlear nuclear factor κB. It is a molecular transcription factor that responds to pathogenic stress such as infection, mechanical, or osmotic so it helps to mediate a cellular response, accompanied with the stimulation of synthesis of cytokines and alterations of homeostatic balance of the inner ear.

Because the causes of SSNHL have many different etiologies and factors, there is no examination that can accurately determine its etiology. That is why it is still considered as idiopathic in 71-85% cases and therefore there is not a treatment that targets the etiology. Over time, MR imaging has provided new insights for the etiology because of high-resolution sequences that are able to detect subtle changes in the inner ear. However, it is still unclear how MR imaging could change therapeutic strategies. A study revealed MR imaging with 3D-FLAIR sequences provides new insights and may change current clinical and therapeutic practices. It should be performed as soon as possible and preferably before the masking effect of steroids becomes apparent because the hypothesis is that pre-contrast and contrast-enhanced MR imaging abnormalities are more easily detected before steroid treatment is started. MR imaging was not available in our hospital; thus we could not perform it.

Treatment can be based on rational approaches such as from the history of the patient, physical examination findings, and laboratory results. If there is no definitive or treatable etiology found, treatment regimen should be dictated by the most likely factors involved. The treatment regimens are varied, it can be regulated by the mechanism of action. Several therapies for these are vasodilators to improve the blood supply to cochlea, preserving hypoxia. Rheologic agents functioned by altering blood viscosity so that better oxygen delivery might be achieved. Anticoagulants are useful to interfere with coagulation cascade as a mechanism to avoid formation of blood clots. Antiviral agents such as Acyclovir and Amantadine can be used. Surgical care is usually done in repairing oval and round window perilymph fistulae with history of recent trauma or barotrauma. For addition, Antiviral therapy, thrombolytic therapy, vasodilator therapy, and vasoactive substances should not be routinely prescribed to patients with SSNHL. Hyperbaric Oxygen Therapy can also be done in combination with steroid therapy to patients with SSNHL within 2 weeks of onset. It is thought to protect hair cells from ischemic damage by the increasing of oxygenation.

For the inflammatory agents, corticosteroids are the primary anti-inflammatory agents used for ISSHL but the mechanism is still unknown. It is effective in controlling clinical trials using high doses of prednisone taper. However, a study says that the safety and efficacy of intratympanic steroids therapy (IST) have not been studied in a randomized clinical trial. There is evidence that suggests IST improves treatment success by increasing intracochlear corticosteroid and reducing incidence of toxic side effects. Still, the mechanism through which steroids may improve hearing remains unknown. The presence of both glucocorticoid and mineralocorticoid receptors in the inner ear may be the clue of steroids mechanism of action. The known roles and main roles are to protect cochlea from harmful effects of inflammatory mediators such as tumor necrosis factors (TNF-α and NF-κB) and cytokines (interleukin 1 and 6), which elevated in infection and flogosis, increasing cochlear blood flow, so to avoid cochlear ischemia, avoiding noise-induced hearing loss, and regulate protein synthesis in the inner ear. The vascular stria regulates Na/ K secretion to maintain endocochlear potential and it is the most frequent site of injury in ISSHL. Systemic steroid therapy improves vascular stria function and may preserve its morphology and so it has the potential for recovering from ISHHL. In our patient, there were hypertension and hyperlipidemia, both which are known as risk factors for ischemic vascular diseases. The role of systemic steroid therapy in improving vascular stria function may help our patient recovering from his ISHHL.

Prognosis for hearing recovery for ISSHL is dependent on a number of factors such as the severity of the hearing loss, age, presence of vertigo, and shape of the audiogram. Indicators of better prognosis include low frequency hearing loss, a less severe hearing loss at presentation, and early commencement of treatment.
Conclusion
In our case, we suspect that both ischemic vascular disease and viral infection as the cause of ISHHL. Hypertension and hyperlipidemia were present in our patient, as the risk factor of ischemic vascular disease. Treatment can be based on rational approaches such as from the history of the patient, physical examination findings, and laboratory results. If there is no definitive or treatable etiology found, treatment regimen should be dictated by the most likely factors involved, which were systemic steroids and acyclovir in our case. Prognosis for hearing recovery for ISSHL is dependent on a number of factors such as the severity of the hearing loss, age, presence of vertigo, and shape of the audiogram.

List of abbreviations
SSNHL - Sudden sensorineural hearing loss  
ISHHL - Idiopathic Sudden Sensory Hearing Loss  
VZV - varicella zoster virus  
MR - Magnetic Resonance  
IST - Intratympanic Steroids Therapy  
TNF-α - Tumor Necrosis Factors alpha  
NF-kB - Nuclear Factor Kappa Beta

Declarations
Ethics approval and consent to participate
Informed consent from the patient has been obtained before the study.

Consent for publication
Consent for publication regarding patient data has been obtained before the study. All the patient identity has been kept secret.

Availability of data and materials
Not Applicable

Competing interests
The authors declare that they have no competing interests.

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References