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Review Article

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Evaluation of idiopathic sudden sensorineural hearing loss

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ABSTRACT

Idiopathic sudden sensorineural hearing loss (ISSNHL) is a medical emergency. The early diagnosis and treatment should improve the complete recovery. The critical issue is diagnosis.

Keywords: Sudden, Idiopathic, Sensorineural, Hearing loss

INTRODUCTION

Idiopathic sudden sensorineural hearing loss (ISSNHL), can be determined as 30 dB or more sensorineural hearing loss over at least three contiguous audiometric frequencies occuring within three days or less. It forms the 1 % of all cases of sensorineural hearing loss.[1,2]

The term 'idiopathic' sudden sensorineural hearing loss was called by De Kleyn in 1944.[3] Hearing loss may together with tinnitus in 70 % of all cases and vertigo or milder sensation of spatial disorientation is present in 50% of cases. The level of hearing loss may be mild in a restricted frequency or it may be total.[1,4] Sudden sensorineural hearing loss (SSNHL) is a critic, and an alarming symptom. It is a medical emergency. There are a number of SSNHL causes. On the other side most of SSNHL cases about 85-90% of cases, are idiopathic.[5,6] The estimated incidence is 5-20 cases per 100.000. The hearing loss is nearly always unilateral and is commonly associated with tinnitus and aural fullness. If the inner ear symptoms have together with the hearing loss, the hearing loss has poorer chance of recovery.[7] It has been reported that the frequency of sudden deafness is similar in males and females and similar on the right and left sides.[8] SSNHL can occur in both sexes equally. The average age is 30-50 years. It occurs commonly unilateral, 1.7% can be seen bilaterally.[9]

ETIOLOGY

The etiology of SSNHL is still remain unclear. It is idiopathic, viral infections, vascular compromise, discruption of cochlear membrane, immunological diseases and otological tumors have been accused for. One third of patients have upper respiratory prodrome. This underlies the role of the viruses in the etiology of SSNHL.[1,10] There are two potential mechanisms of viral infection may cause SSNHL. One mechanism is viral invasion of the fluid spaces and/or soft tissues of the cochlea (cochleitis) or invasion of the cochlear nerve (neuritis). The virus can reach the inner ear by hematogenous route, from the cerebrospinal fluid or from the inner ear. The second mechanism is reactivation of a virus that is latent within tissues of the inner ear. Neurotropic viruses could infect the cochlear neurons, remain dormant for varying lengths of time, and then become reactivated later in life to result in a viral neuritis and/or cochleitis, leading to SSNHL. There is another mechanism by which a virus could indirectly trigger SSNHL. This occurs as an antibody response that cross-reacts with an inner ear antigen (immune-mediated hypothesis). Also in the third mechanism; the virus could trigger a circulating ligand that causes pathologic activation of cellular stress pathways within the cochlea. Mumps virus has been accused for sudden deafness, IgM antibodies have been demonstrated by serologic studies.

Rubella virus has also been temporally or serologically associated with a few cases of sudden deafness. Immunization against mumps, measles and rubella has decreased the incidence of these diseases. The herpesviridae family of viruses have been proposed as causes of SSNHL. Herpes simplex type 1 and 2, Varicella zoster virus, CMV, Epstein-Barr virus, Human herpes - virus 6-7 and 8 can cause SSNHL by latent reactivation.[11]

Vascular insufficiency may cause sudden sensorineural hearing loss. Vascular insufficiency may be in conjunction with some systemic pathological process such as leukemia or intracranial lesions such as vestibular nerve schwannomas or meningiomas or surgical intervention.[12] The pathogenesis of hearing loss in patients with leukemia is complex. The changes in temporal bone can be determined into 4 main categories: 1) leukemic infiltration 2) hemorrhage 3) infection 4) hyperviscosity. Hematological malignencies should be considered one of the possible etiology in patients with SSNHL.[13]

The relationship between the abnormal cochlear microcirculation and SSNHL has been the issue of authors.[14] The alterations in microcirculation of cochlea may be cause of SSNHL. Cochlea is supplied by labyrinthine artery. Labyrinthine artery has no collateral vasculature. Vascular disorders may cause cochlear injury and this can be result in SSNHL.

The possible mechanism for vascular cause of SSNHL includes hemorrhage, thrombosis, embolism, vasospasm and hypercoagulability.[14] The inner ear has been long known to be vulnerable to hypoxia. Interuption of blood supply to the cochlea for 30 minutes causes cochlear damage.

The partial vascular occlusion results in loss of spiral ganglion, and total occlusion results in labyrinthine ossificans.[3] A sudden break within the labyrinthine membranes of the cochlea with or without an accompanying rupture of the oval or round window membrane has been hypothesized to be the cause of SSNHL.[11]

Physical exertion, the valsalva maneuver, nose blowing changes the intralabyrinthine pressure and these situations may cause breaks within the labyrinthine membranes. However; patients presenting clinically with SSNHL do not describe a precipitating physical event leading to the hearing loss. Most of patients report deafness upon awakening or while being sedantery . Also weight lifters, women in childbirth do not experience an increased incidence of SSNHL.[11]

Cardiovascular disease, cigarette smoking and hypertension are the most common risk factors associated with SSNHL.[15-17] Advanced age, severe hearing loss, heredity, audiogram shape, and presence of vertigo are negative prognostic factors.[15,17-18]

HISTORY AND DIAGNOSIS

SSNHL is nearly always unilateral, but bilateral SSNHL can be seen. Bilateral SSNHL is rare, and could be caused by autoimmune disease, syphilis, trauma, neoplasia and vascular causes. The kind of activity being undertaken at the time of the hearing loss may give information about the cause. Excessive strainingi, especially result in perilymphatic fistula and physical or acustic trauma can result in SSNHL. A detailed past medical history may include potential causes; such as autoimmune disease, diabetes mellitus, sarcoidosis or vascular disease. Previous otological surgery may point to disease recurence or failure of outcome. Ototoxic drugs, such as aminoglycosides, furosemide, Non-Steroidal Anti-İnflammatory Drugs (NSAIDs), chemotherapetics should be asked. And the patient should be sought for any relevant family history such as for otosclerosis.[5] Aural fullness, vertigo may point at Meniere's disease.[3] The severity of initial hearing loss is a bad prognostic factor. Also the initial symptom of vertigo is a bad prognostic indicator. The duration of the onset of hearing loss to treatment is very important. The hearing improvement commonly has been seen in one week period.[19]

EXAMINATION

A routine cranial nerve examination should be perform to exclude cranial nerve abnormalities. Cranial nerve abnormalities may suggest intracranial lesions (acoustic neuromas or malignancy) or multiple sclerosis. Otoscopy should be performed carefully. Foreign bodies, wax can cause conductive hearing loss. Pure tone audiogram is the first step audiological test used to differentiate conductive hearing loss from sensorineural hearing loss by assessing both air and bone conduction thresholds. Pure tone audiogram will determine hearing loss, the degree and type of loss (Figure 1).[5]

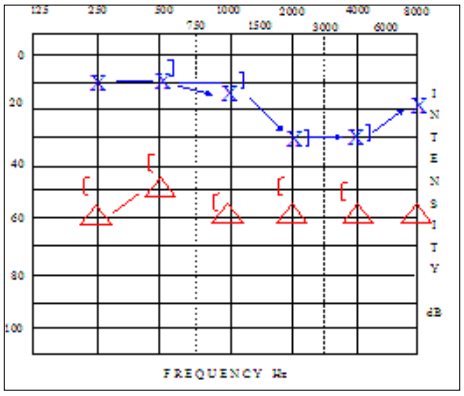

Figure 1: A right ear SSNHL (Pure tone audiogram).

Treatment outcomes have been demonstrated to be better in patients with a flat or ascending type audiogram than in those with a descending or profound type. Patients with low frequency deafness experience fast hearing

recovery.[8,20] The prognosis of patients with descending type of hearing loss has poor recovery in children.[8,21]

Tympanometry can help assessing tympanic membrane mobility and middle ear function. Especially in middle ear problems, tympanometry is helpfull. Flexible nasoendoscopy visualises the postnasal space for possible masses. Blood tests are not urgent and can be guided by clinical findings.[5]

Diabetes mellitus, renal and/or cardiovascular diseases should be reminded. Routine blood studies should be done to rule out systemic illness and metabolic diseases. Magnetic Resonance İmaging (MRI) should be performed against the possibility of neurological lesions. Acoustic tumors, minimal Mondini dysplasia of the cochlea, ear anomalies like vestibular aqueduct syndrome, vascular anevrymsms of the AICA and basilar artery can be ruled out by the help of MRI scans. In cases of bilateral SSNHL, systemic immune disease mediated hearing loss should be remembered and the treatment protocol should be different.[3]

TREATMENT

The treatment of SSNHL should be started immediately. The time is limited. The treatment is not helpful after 30 days because of the permanent damage. Systemic steroids are the major treatment agents for their high anti-inflamatory effect.[1,22] They reduce cytotoxic immune response, increase the microvascular blood flow in the cochlea and decrease the endolymphatic hydrops.[1] Steroid treatments restrain the inflammatory reaction, activate ion transport in the stria vascularis and spiral ligament within the cochlear duct, control homeostasis of the endolymph, and increase blood circulation in the cochlea.[9,23]

In secondary hydrops, like cogans, and immune mediated diseases, salt restricted diet and small dose of diuretics thiazide may be benefical.[3]

The treatment protocol for steroid is; intravenous hydrocortisone; prednisolone 1 mg/kg or methylprednisolone 0.8 mg/kg. This dosage should be decreased gradually.[1]

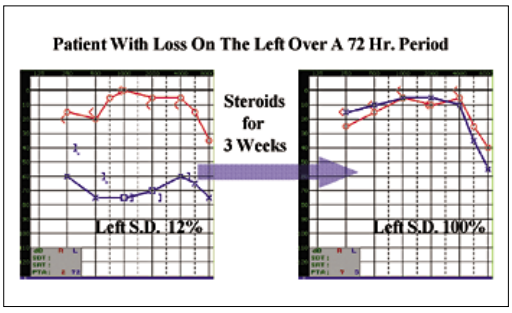

Figure 2: A left ear ISSNHL pure tone audiogram and the audiogram after steroid theraphy.

Systemic steroids, orally or intravenously, are the major treatment of SSNHL (Figure 2). Steroids are potent anti-inflammatory agents. It is believed that, steroids may cause vasodilatation with increased microvascular blood flow in the cochlea resulting in decreasing endolymphatic hydrops and inflammation. To maintain this process; steroids should be higher concentrations in the inner ear. Especially intravenously they could reach the adequate concentrations in the inner ear. If there is no improvement of hearing after a period of 2-3 months, with or without treatment amplification and supportive audiologic rehabilitation to the patient should be considered. In diabetics the ENT specialist should be in a close relationship with a diabetologist . Diabetologist should strictly monitore the blood sugar level daily, while the patient is an iv steroid therapy , and appropriate glycemic control should be achieved with an insulin sliding scale. For the hypothyroidic patients, appropriate administrations of oral thyroid hormones is necessary in order to maintain the euthyroid status, while iv steroid treatment. Iv steroid may affect the blood pressure level. Hypertensive patients also should be observed closer against the possibility of hypertension induced endolymphatic hydrops and micro-angiopathy within the inner ear. According to the most of authors, administration of high dose intravenous corticosteroids to patients with SSNHL is highly recommended in the literature. Comorbidities such as diabetes, hypertension reduce the recovery chance. Iv steroid therapy has best results when commenced within complete recovery of cochleopathy. High frequency hearing preservation have better hearing improvement at the end of treatment. Stomach ulcer, left sided heart insufficiency, renal/ adrenal disorders, active systemic sepsis are the steroid treatment limitations.[7]

The intratympanic steroid injection may reduces the success of oral steroid theraphy, and may be a choice of late treatment if the response to systemic steroid is poor.[5] Combined theraphy with intratympanic and systemic steroids is more effective than systemic steroids alone in the treatment of poor prognosis SSNHL. It has been shown that the chance of salvaging hearing decreases if the time interval between the insult and the administration of intratympanic steroid theraphy after oral steroid failure increases. If intratympanic steroids are to be used, therefore, they should be used as soon as possible after it becomes clear that systemic steroids are not effective, preferably within 2 weeks of the original insult.[24]

Antiviral agents like acyclovir or valacyclovir should be added to the treatment protocol for the probable viral etiology. Acyclovir and valacyclovir are used commonly. Valacyclovir is a prodrug of acyclovir that produces serum acyclovir levels that are 3-5 times as high as those achieved with oral acyclovir theraphy, and similar to levels achieved by intravenous acyclovir. None showed any benefit in using these drugs to treat SSNHL.

Acyclovir or valacyclovir are active against HSV or VZV.[11]

Vascular occlusion or ischemia has been accused for SSNHL. The labyrinthine artery, which supplies the cochlea and vestibular sense organs, is an end artery.[11] For this low molecular weight dextrans used to maintain the expansion of plasma volume and increase in cardiac output, increase in vascular perfusion and microcirculation, reduction in blood viscosity; platelet adhesiveness.[1]

The treatment protocol is for intravenous dextrans: in 5% dextrose 500 cc in 4-6 hours (patients should be kept under continuous supervision against a cardiac overload possibility) and this protocol may be repeated if there is a subjective improvement.[1]

ROLE OF SURGERY

Surgery is necessary for the hearing loss secondary to trauma, rupture of the round window or oval window membrane or spontaneous labyrinthine fistula. A perilymphate leak occurs especially following barotrauma or traumatic event. Rarely it occurs spontaneously due to Mondini dysplasia with oval window fistula or vestibular aquaduct syndrome or from Hyrtle's fissure. The diagnosis of the microfistula is difficult. The use of rigid endoscope and beta 2 transferrin assays have improved the sensitivity of finding the fistula. Exploratory tympanotomy should be carried out in these cases. Emergency stapedektomy is required in cases with fracture footplate and stapes suprastructure. In doubtful leaks around footplate or round window membrane, sealing with the fat graft or adipose tissue is a treatment method for the further deterioration of hearing loss.[3]

HYPERBARIC OXYGEN

The use of hyperbaric oxygen in the treatment of SSNHL is intended to increase the partial oxygen pressure and oxygen concentration in the inner ear and to improve the microcirculation and blood profile. Oxygen inhalation increases the oxygen tension of the perilymphatic fluid by 450% of its initial value, and this state remains for 1 h after termination of the HBO. Perilymphatic oxygen tension decreases in patients with ISSNHL. Best results are achieved if the treatment is started early after the onset of deafness. Aslan et al., reported the successful outcomes of HBO theraphy in additional SSNHL theraphy. In patients older than 50 years, the benefical feature of the HBO is limited and no benefit in patients older than 60 years. HBO theraphy is benefical against the unsuccessful conventional treatment. Especially HBO treatment has been recommended in high frequencies of ISSNHL. In summary, the HBO theraphy additional to conventional tretament modalities improves the outcome of ISSNHL, especially at the frequencies of 250, 500, 1000 and 4000 Hz and in hearing loss of above 61 dB.[25]

The incidence of SSNHL in children is less than that in adults and the low incidence in children has resulted in very limited data for pediatric age group. The positive and negative prognostic factors are similar in children to adults. Corticosteroid theraphy should be managed carefully with a close relationship with a pediatrician not to cause growth failure.[26]

In long term care for SSNHL, especially in children; permanent deafness can lead additional delays in language development and dysarthia. Therefore, in proper patients cochlear implant surgery is recommended. Hearing aids and speech training are very important for speech production after cochlear implantation. Therefore, it is necessary to diagnose bilateral SSNHL and to surgically intervene with a cochlear implant as soon as possible.[27]

CONCLUSION

In conclusion, the treatment of SSNHL has remained one of the most conversial issues in otolaryngology. The critical process is true and immediate diagnosis.

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