

A Study of Aetiological Profile of Unilateral Sensorineural Hearing Loss

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ABSTRACT

Introduction

It is necessary to identify the aetiology of sudden sensorineural hearing loss for proper management. Despite advancement in the field of diagnostic radiology, immunochemistry and genetic studies, there are still many cases of USNHL where a definite cause cannot be identified. In this study, an attempt has been made to identify the aetiological factors responsible for unilateral sensorineural hearing loss (USNHL).

Materials and Methods

Seventyfive patients with acquired unilateral sensorineural hearing loss without chronic otitis media were included in the study. Hearing was assessed by tuning fork test and pure tone audiometry. Special tests were done to differentiate between cochlear and retrocochlear pathology, to objectively assess hearing loss and to diagnose nonorganic hearing loss. Radio-imaging was done to diagnose skull bone fractures, space occupying lesions in internal auditory meatus and cerebellopontine angle or intracranial pathologies. Biochemical analysis of blood, immunoglobulin and serological assay were done to rule out specific causes.

Results

Majority of patients were within 21-30 years of age. 37 patients presented with sudden and remaining with progressive hearing loss. Majority (57.3%) were found to have idiopathic cochlear pathology. Amongst the remaining, there were 8 cases of acoustic trauma, 4 each of Meniere's disease and cerebellopontine angle tumour and 3 each of Noise-induced hearing loss, labyrinthitis and Non-organic hearing loss. There were 2 cases of meningitis and 1 each of barotrauma, connective tissue disorder, iatrogenic trauma, fracture of petrous bone and cerebrovascular accident.

Conclusion

Most of the studies have been done on sudden sensorineural hearing loss. Current study, including sudden and progressive cases, is hence an attempt to ascertain aetiological profile of unilateral SNHL.

Keywords

Hearing Loss, Sensorineural; Unilateral; Idiopathic; Aetiology

Unilateral sensorineural hearing loss (USNHL) is one of the most intriguing situation faced by Otolaryngologists among the patients attending out-patient clinics. The condition is not only very difficult to treat for the doctors but can be embarrassing and frustrating for patients also. For proper management of this condition, it is necessary to identify the underlying cause.

In recent years there has been a huge advancement in the field of diagnostic radiology, immunochemistry and genetic studies. A sea change has come in the audiological battery of tests with the introduction of newer sophisticated audiological tests. Because of this, definite aetiological factors are now found in a good number of cases having USNHL. However, despite all

possible investigations, there are still many cases of USNHL where a definite cause cannot be identified.

In this study, an attempt has been made to identify the aetiological factors responsible for USNHL, guide the treatment protocol for individual cases and to ascertain the prognosis in each case. Cases of USNHL based on history and clinical finding have been studied and data

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has been analysed statistically using standard statistical methods.

The aim of this study was to establish the diagnosis of USNHL, determine its aetiology and locate the site of pathology. The cases were then managed accordingly.

Material and Methods

This prospective observational study was carried out in the ENT department of a tertiary care centre over a period of 2 years. Seventy five consecutive patients with USNHL who attended our out-patient department (OPD) or were admitted in various wards of the hospital were included in the study, were included in the study and the data was analyzed using standard statistical methods. Cases of chronic otitis media (COM) with mixed hearing loss, congenital SNHL, hereditary causes and those who were unwilling to participate in the study were excluded from it.

A detailed history was taken, general physical examination and thorough ENT evaluation was done. Hearing assessment was done using tuning fork test, free field hearing (FFH) and pure tone audiometry (PTA) in all cases. Special audiological tests like short increment sensitivity index (SISI), alternate loudness balance test (ALB), tone decay test were done to differentiate between cochlear and retro cochlear pathology. Brainstem evoked response audiometry (BERA) was done to objectively assess the hearing loss and to diagnose nonorganic hearing loss. Electrocochleography (ECoG) was done to estimate the ratio between summation potential and action potential (SP:AP) to diagnose Meniere's disease. Caloric test was done to see the involvement of peripheral vestibular system.

Magnetic resonance imaging (MRI) of brain with gadolinium enhancement was done in all cases to assess the internal auditory meatus (IAM), cerebellopontine (CP) angle and other pathologies like meningitis, vascular anomaly and cerebrovascular accident (CVA). High resolution computed tomography (HRCT) of temporal bone was done in selected cases of head injury with suspected temporal bone fracture.

Haematological tests including complete haemogram

and biochemical parameters were done to rule out comorbidities in all cases. Double Sandwich ELISA test for detection of IgM antibodies of TORCH (Toxoplasma, Rubella, Cytomegalovirus, Herpes Simplex) pathogens was done to rule out infective causes. Other tests like Venereal Disease Research Laboratory test (VDRL), lipid profile, thyroid profile, Widal test for Typhoid, serum Angiotensin-converting enzyme (ACE) for Sarcoidosis, prothrombin time (PT), partial thromboplastin time (PTT) were done to rule out other pathology. Serological tests like Antinuclear antibody (ANA), double stranded DNA (dsDNA), cytoplasmic and perinuclear antineutrophil cytoplasmic antibody (cANCA and pANCA) were done to rule out connective tissue disorders. Human Immunodeficiency Virus (HIV) screening was done in all cases.

Wherever cause of hearing loss could be found, the treatment was directed at the definite diagnosis. In idiopathic cases, treatment was largely empirical constituting oral steroids, vasodilator and vitamins. They were also counselled regarding the nature of their illness and the measures they should take to protect the remaining hearing ability in the affected ear and to protect the normal ear.

Results

There were 52 male (69.3%) and 23 (30.7%) female. The majority of patients (34.7%) in our study were within the age group of 21-30 years, mean age being 38.6 years. Only one patient was under 20 years of age and two patients were more than seventy years of age (Fig. 1).

37 patients (49.3%) presented with sudden sensorineural hearing loss (SSNHL) which is defined as hearing loss of more than 30 dB, in three or more contiguous frequencies, occurring within a period of 72 hours.¹ Remaining 38 patients (50.7%) presented with progressive hearing loss. 15 patients (20%) attended for medical advice within one month of onset of hearing loss whereas 60 patients (80%) presented after a month.

In our study 57 patients (76%) had associated tinnitus which started along with hearing loss. No patient complained of pulsatile tinnitus. In 22% of the patients,

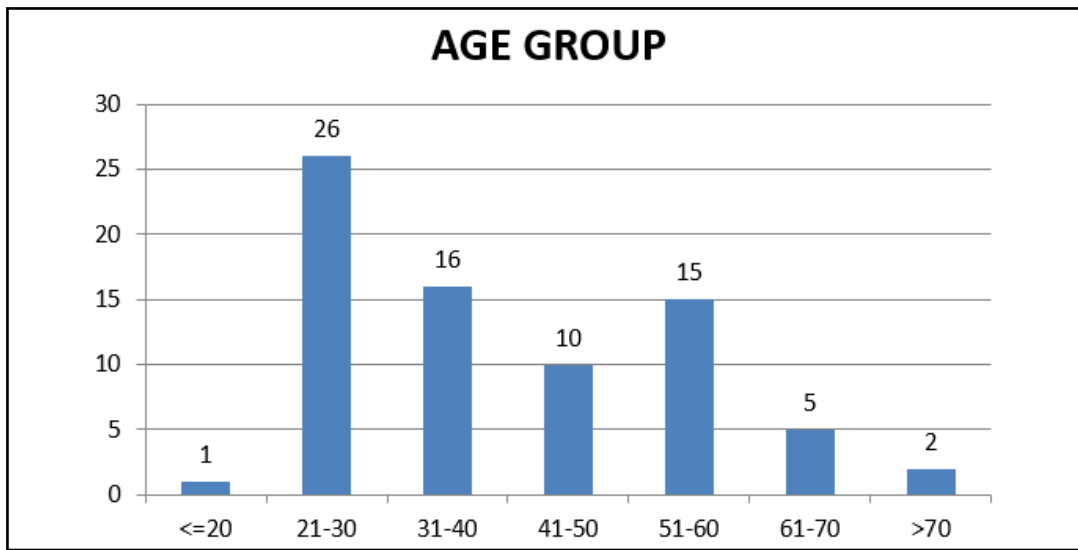


Fig. 1. Age distribution of patients

tinnitus interfered with their daily activities and sleep whereas in 78% of cases it was not troublesome. Giddiness was an associated symptom in 31 patients (41.3%). Among them, 4 patients had severe rotatory vertigo lasting for more than one hour but less than one day along with aural fullness and fluctuating hearing loss suggestive of Meniere’s disease. Three patients had severe vertigo with nausea and vomiting which lasted for more than one day associated with sudden onset hearing loss suggestive of labyrinthitis. Remaining patients

had nonspecific giddiness associated with hearing loss. 27 patients (36%) complained of aural fullness in ipsilateral ear during the onset of hearing loss. Three patients (4%) had the history of chronic exposure to loud noise whereas 8 patients (10.7%) had history of exposure to high intensity noise just before the onset of hearing loss. Out of 4 patients (5.3%) having preceding history of head injury, only one had fracture of temporal bone involving bony cochlea.

In tuning fork tests, the findings were consistent in 72

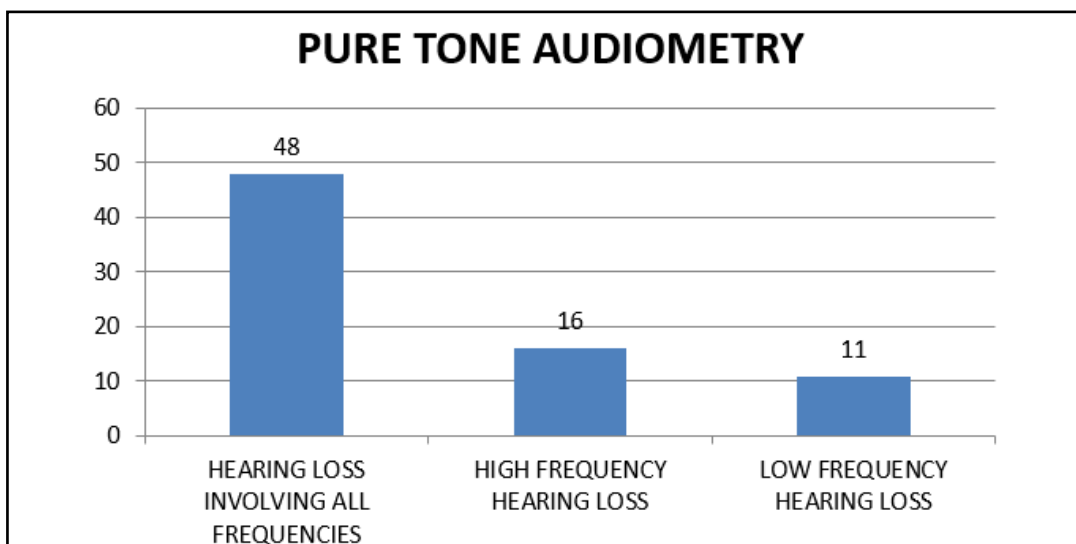


Fig. 2. Frequency related hearing loss in pure tone audiometry

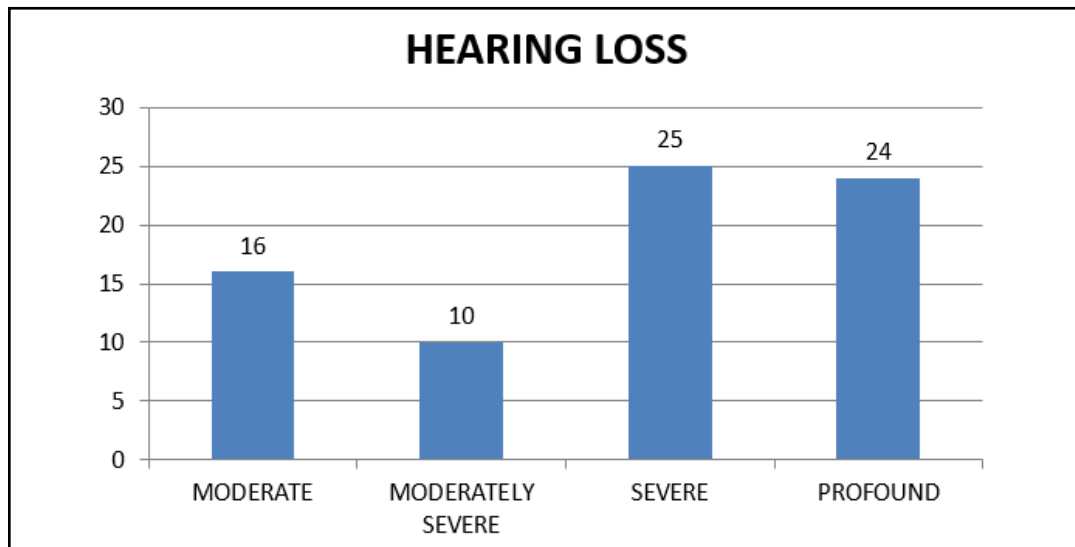


Fig. 3. Distribution of severity of hearing loss

patients (96%) with their history and clinical findings. Remaining 3 patients were later diagnosed as cases of non organic hearing loss (NOHL). PTA was done in all cases. 48 patients (64%) had hearing loss involving all frequencies, 16 patients (21.3%) had high frequency hearing loss whereas 11 patients (14.7%) had low frequency hearing loss (Fig. 2).

16 patients (21.3%) had moderate hearing loss, 10 patients (13.3%) had moderately severe hearing loss, 25 patients (33.3%) had severe hearing loss and 24 patients

(32%) had profound hearing loss (Fig. 3).

Bi-thermal caloric test was done in all cases. If found absent, it was repeated with ice cold water to see any response before labelling the case as dead labyrinth. Caloric response was absent on the side of hearing loss in 20 patients (26.7%), reduced compare to healthy side in 28 patients (37.3%) and absolutely normal in 27 patients (37%) (Fig. 4).

Otoneurologic examination was normal in 67 patients (89.3%) whereas some abnormality was detected in 8

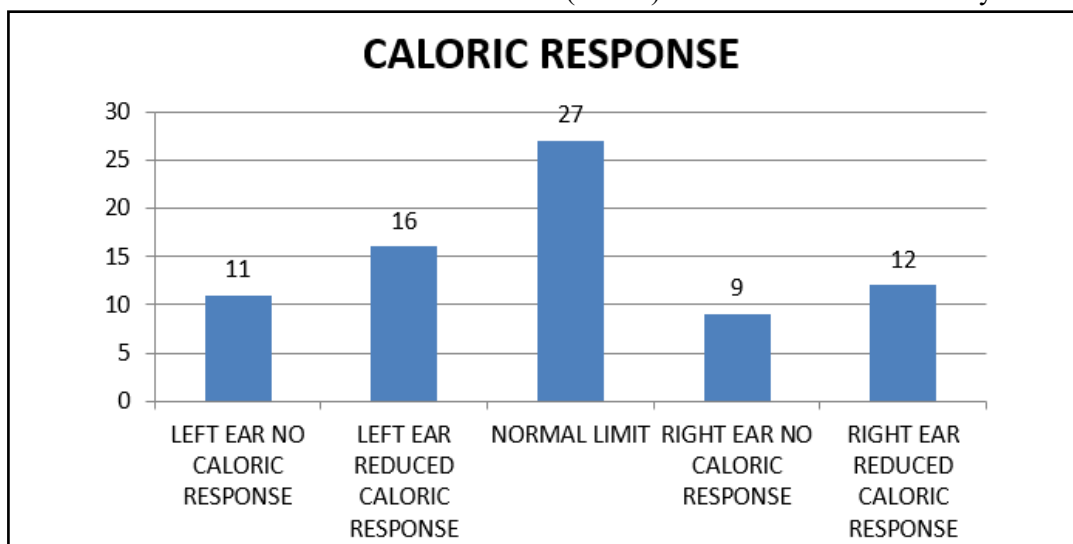


Fig. 4. Distribution of caloric response

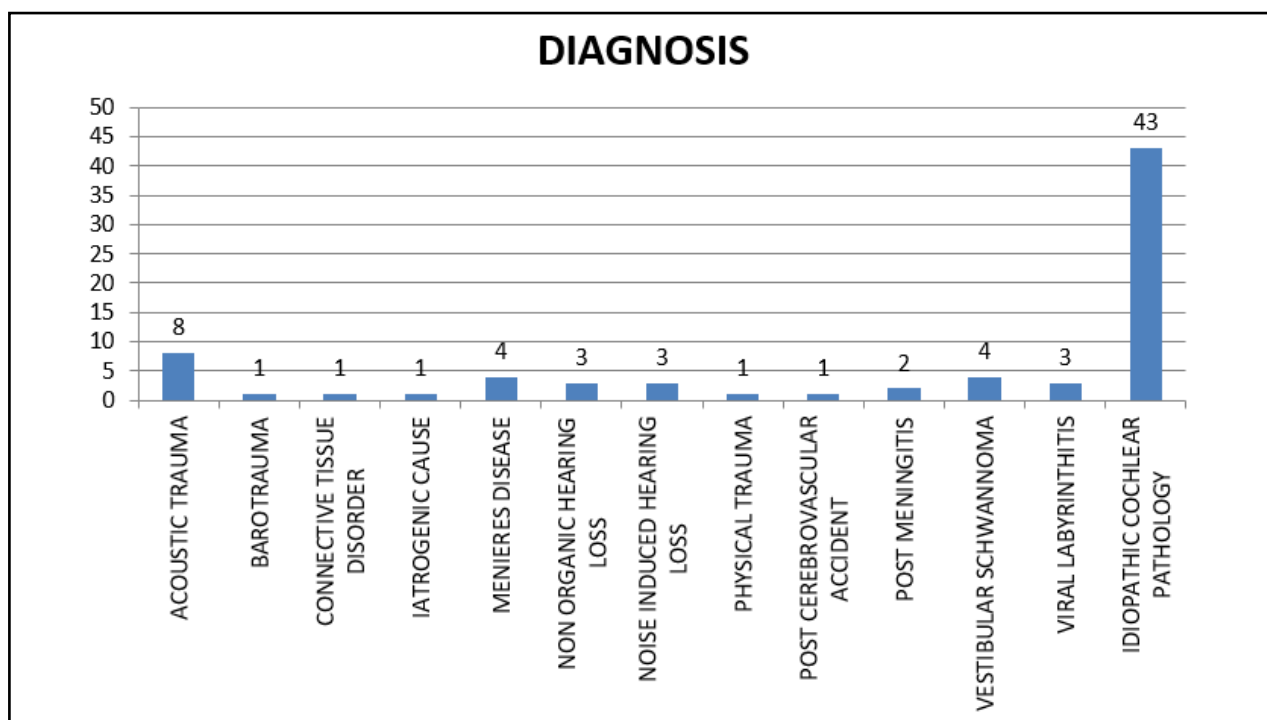


Fig. 5. Aetiological pattern of USNHL

cases (10.7%). Among those cases, cerebellar tests were abnormal in 5 cases, 2 patients had nystagmus towards the side of pathology and one patient had nystagmus opposite to the side of hearing loss. 66 patients (88%) had cochlear pathology (SISI score >90 and tone decay <5dB) and 6 patients (8%) had retro cochlear pathology (SISI score <20% and tone decay >30dB). ECoG showed increased SP:AP ratio only in 4 patients (5.3%). Findings in BERA were consistent with hearing loss in 72 patients (96%), whereas 3 patients were found to be malingerers.

HRCT of temporal bone was done in 4 cases with history of head injury and showed fracture line across cochlea in only one case. Gadolinium enhanced MRI of brain, CP angle and IAM was abnormal in 8 cases (10.7%). 4 of them were found to have vestibular schwannoma at CP angle, two had communicating hydrocephalus following meningitis, one had cerebellar infarction and one patient had multiple cerebral infarcts.

Complete hemogram was found to be deranged in 8 patients (10.7%); 2 patients with SSNHL onset in high altitude area showed features of polycythaemia,

two patients had reduced platelet count, three had leucocytosis and one patient had increased INR. Blood glucose estimation in 14 patients (18.7%) showed hyperglycaemia. Deranged lipid profile was found in 24 (32%) cases. In 13 cases both cholesterol and triglycerides were elevated, in 4 cases only cholesterol level was elevated and in 7 cases serum triglycerides were increased. 9.3% of cases had associated Hypothyroidism.

Anti DsDNA antibody was found in 4 (5.3%) cases. Among them, only one patient had full blown features of connective tissue disorder along with unilateral sensorineural hearing loss. pANCA and cANCA were positive in 3 of cases (4%). In our study, majority (57.3%) were found to have idiopathic cochlear pathology (Fig. 5).

4 patients (5.3%) had complete recovery of hearing – hearing threshold being 20dB, 23 patients (30.7%) had partial recovery of hearing – hearing gain more than 11dB but threshold remaining higher than 20dB and remaining 48 patients (64%) had shown no recovery of hearing - hearing gain between 0 to 10dB only.

Discussion

Just as we use two eyes to see in three dimensions, we use two ears to perceive the world of sound around us. This “dimensional hearing” is made possible by binaural hearing.

A person with monaural hearing loses the ability to ‘map’ the sound in space, pick out soft sounds and to separate a single voice from the surrounding background noise. USNHL is relatively uncommon as compared to bilateral SNHL. Audiometric data of 252 patients suffering from hearing loss in an Indian retrospective study showed that 163 patients (66.5%) had SNHL, out of which only 6 cases (3.7%) had USNHL.²

Age distribution:

Unilateral hearing loss, though not common in paediatric age group, is found in both children and adults. In children, it is mostly detected on screening. They do not, as a routine, present as OPD cases. In our study age of patients ranged from 15-73 years, mean age being 38.6 years; where 34.7% were within the age group of 21-30 years. Bansal et al in their study found that age of patients ranged from 9 years to 76 years (mean age being 41.5 years), maximum patients were in 3rd decade (23.87%), followed by 2nd (21.29%) and 4th decade (21.29%).³

Sex distribution:

In our study, 52 cases (69.3%) were male and 23 cases (30.7%) were female. Our finding showed a slight variance with Bansal et al, who in their study group of 155 pure USNHL cases, found that 88 patients (56.77%) were males and 67 (43.23%) were females. This is in sync with WHO Global Estimate on Prevalence of Hearing Loss, 2012, which states out of 328 million adult with disabling hearing loss, 183 million are male and 145 million are female.⁴

Laterality:

Sharma et al, among pure unilateral cases, reported right ear involvement in 48.2% of cases and left ear involvement in 51.8% of cases,⁵ which is similar to the

findings in our study where right ear was affected in 45.3% of cases and left ear in 54.7% of cases.

Onset:

SSNHL was found in 37 (49.3%) patients while the rest (50.7%) presented with gradual onset of hearing loss. In the study done by Bansal et al, only 12 cases (7.74%) had sudden onset of USNHL while 143 (92.26%) had gradual onset.³

Duration of hearing loss:

In our study, 15 (20%) patients sought ENT consultation within a month of onset of hearing loss whereas 60 (80%) patients presented later. It is one of the known prognostic factors because initiation of definitive treatment after ten days of onset of hearing loss has been related to poorer recovery of hearing compared to those who had been treated promptly.⁶

Associated tinnitus:

In our study 76% of cases had associated tinnitus which started along with hearing loss. Tinnitus has been reported to be only an accompanying symptom in a study by Edizer et al,⁷ without influencing the prognosis. On the other hand, tinnitus has been reported as a prognostic factor correlated with better recovery rates in other studies.^{8,9}

Associated giddiness:

41.3% cases in our study had vertigo as an associated symptom along with hearing loss. Presence of vertigo, which has been reported to occur in as much as 40% of patients with sudden hearing loss, has been shown to be a negative prognostic factor.⁹

Audiological tests:

Table 3 shows that hearing loss in our patients was from moderate (21.3%) to profound (32%). There was no case of mild hearing loss suggesting that mild unilateral hearing loss was not noticed by the patients or was ignored by them. This finding was in direct contrast with the findings of Bansal et al where 34.2% of USNHL

cases were of mild degree.³ Tone decay test and SISI, done to differentiate between cochlear and retro cochlear pathology, were suggestive of cochlear pathology in 66 patients (88%) and retro cochlear pathology in 6 patients (8%). BERA, done to objectively measure the hearing loss, was corroborative with findings in PTA in 72 patients (96%). 3 patients were found to be malingerers. ECoG showed increased SP:AP ratio only in 4 patients

Imaging studies:

MRI brain, in our study, showed abnormal findings in 8 cases (10.7%) with 4 patients (5.3%) diagnosed to have vestibular schwannoma. Findings are similar to the study conducted by Robert et al (2000) where high resolution fast spin echo MRI revealed abnormality in 11.8% cases in a series of 1070 patients of USNHL. Among them, 5.2% cases were vestibular schwannomas and the rest were other pathologies including vascular causes.¹⁰

Relevant blood tests:

14 patients (18.7%) had hyperglycaemia in our study. Pre-existing microvascular lesions in diabetic patients has poor prognosis in SSNHL and raised post prandial blood glucose level could be a risk factor indicator for cochlear dysfunction in them.¹¹ Lipid profiles were deranged in 24 patients (32%) in our study. In a study conducted by Cadoni G et al (2005) regarding the diagnosis, treatment and outcome of SSNHL, deranged lipid profile was not found to have any bearing in hearing recovery.¹² Hypothyroidism was found in 7 patients (9.3%) in our study. In a study conducted by Narozny W et al (2006) regarding the prognostic factors in SSNHL, hypothyroidism was associated with poorer hearing recovery compared to those who were euthyroid.⁶ Anti DsDNA antibody was found in 4 patients (5.3%) whereas pANCA and cANCA were positive in 3 patients (4%).

Among them, only one patient had full blown features of connective tissue disorder along with USNHL without any other co-morbidities. Serum ACE estimation, screening for HIV, TORCH antibody, VDRL test were negative in all cases. None of the blood tests led to the aetiology of hearing loss except one case

of connective tissue disorder. Although there was one case of polycythemia with the onset of hearing loss occurring in high altitude location, hypercoagulability state could not be ascertained. It is, therefore, debatable whether blood tests, due to their low diagnostic yield, are essential in the work up of SNHL or not.

Aetiological profile:

On completion of our study of 75 cases of unilateral hearing loss, though cochlear pathology was identified in 43 patients (57.3%), no specific cause was found to be directly related to the pathogenesis of hearing loss. They have been classified as that of idiopathic cochlear pathology. 8 patients (10.7%) were diagnosed as hearing loss due to acoustic trauma. There were 4 cases each (5.3%) of Meniere's disease and Vestibular schwannoma, 3 cases each (4%) of noise induced unilateral hearing loss, viral labyrinthitis and Non-organic hearing loss. Two patients (2.7%) had communicating hydrocephalous following meningitis. One case (1.3%) each could be attributed to barotrauma, connective tissue disorder, iatrogenic sensorineural hearing loss following surgery for chronic otitis media, head injury with fracture of petrous apex involving bony cochlea and cerebrovascular accident with infarction in ipsilateral posterior inferior cerebellar artery.

The comparative study of aetiologies of unilateral SNHL, as found in our study, in studies done by Sharma et al,⁵ Usami et al¹³ and the meta-analysis done by Chau JK¹⁴ et al throws interesting facts (Table I). Where most of the studies find Idiopathic SNHL as commonest cause, Sharma et al reported only 7.09% cases to be idiopathic. Presbycusis, which did not even feature as a cause in USNHL in most of the studies, was reported to constitute 78.72% in the study by Sharma et al, which also reported 1.41% cases of otosclerosis and 2.13% cases of ototoxicity leading to USNHL.

Acoustic trauma, which was the second most common aetiology in our study, did not feature in other studies. Usami et al reported 15.7% cases of USNHL due to otological diseases out of which 12.5% were due to chronic otitis media. Other studies reported only 4.7% to 8% cases due to otological causes, that to none due to COM. The 3rd most common aetiology was quite

Table I: Comparative study of aetiological pattern in USNHL

AETIOLOGY		OUR STUDY (N=75)	SHARMA ET AL (N=141)	USAMI ET AL (N=172)	CHAU JK ET AL
Idiopathic		43(57.3%)	10 (7.09%)	94 (54.6%)	71%
Infectious diseases	Viral: Mumps, Measles, Herpes	-	-	1 (0.58%)	12.80%
	Bacterial Labyrinthitis	3 (4%)	-	-	
	Meningitis	2 (2.7)	-	-	
Otologic disease	COM	-	-	22 (12.8)	4.70%
	Meniere's disease	4 (5.3%)	5 (3.54%)	-	
	Ear surgery	1 (1.3%)		4 (2.3%)	
	Otosclerosis	-	2 (1.42%)	-	
	Ototoxicity	-	3 (2.13%)	-	
	Autoimmune	1 (1.3%)	2(1.42%)	1(0.58%)	
Trauma	Temporal bone fracture	1 (1.3%)	6 (4.25%)	3 (1.7%)	4.20%
	Barotrauma	1 (1.3%)	-		
Vascular/ Haematologic		1	-	-	2.80%
Neoplastic		4 (5.3%)	1 (0.7%)	9 (5.2%)	2.30%
Other causes	Acoustic trauma	8(10.7%)	-	-	2.20%
	Noise induced HL	3 (4%)	1(0.7%)	-	
	Non-organic HL	3 (4%)	-	10 (5.8%)	
	Presbycusis	-	111	-	

Table II: Statistical analysis of causative factors in USNHL

AETIOLOGY	% OF CASES IN THE PRESENT STUDY	% OF CASES IN STUDY BY JUSTIN K CHAU ET AL (2010)	P VALUE	SIGNIFICANCE
Infectious disease	2.7	12.8	0.008	Significant
Trauma	3.9	4.2	0.947	Not significant
Vascular cause	1.3	2.8	0.488	Not significant
Neoplastic causes	5.3	2.3	0.311	Not significant
Other causes	5.3	2.2	0.293	Not significant
Idiopathic	57.3	71	0.061	Not significant

intriguing. It was CP angle tumour in our study, where as in the study by Usami et al it was NOHL in the study by Usami et al and Temporal bone fracture in the study by Sharma et al and in Meta-analysis by Chau JK et al. The study by Fetterman et al identified a definitive cause in only 10% of cases which is in contrast with our finding of 42.7%.¹⁵

‘Test for equality of Population Proportion’ was used for analyzing data. An α level of 5% has been taken and hence any p value <0.05 has been taken as significant. SPSS software version 16 has been used for the analysis. Statistical analysis done after comparing our result of causative factors in USNHL with the result of study conducted by Chau JK et al (2010)¹⁴ for sudden USNHL showed statistically significant difference only in infectious disease as an aetiology. There was no statistical significant difference in idiopathic, traumatic, vascular, neoplastic or other etiological factors (Table II).

Conclusion

Unilateral SNHL can be of sudden or progressive. Scientific data is lacking in etiological study of USNHL where both sudden as well as progressive hearing loss

cases have been evaluated. Most of the studies have been done on sudden SHHL and some have been done on paediatric unilateral SNHL. Authors could find only one study by Bansal et al³ which, though included unilateral SNHL in the age group of 9 years to 72 years, did not elaborate on etiological factors. Our study, which included both sudden and progressive cases, is hence a unique attempt to ascertain aetiological profile of unilateral sensorineural hearing loss.

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