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## Sudden Asymmetric Sensorineural Hearing loss: A Diagnostic Challenge.

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### ABSTRACT

Sensorineural hearing loss (SNHL) results from dysfunction at the level of the vestibulocochlear nerve, inner ear, or central processing centers of the brain. WHO lists hearing impairment at 15<sup>th</sup> rank in its report entitled "Global Burden of Disease" and ranks 2<sup>nd</sup> in its "Disability-adjusted life years" report. The majority of patients affected by hearing loss (>80%) suffer from sensorineural hearing loss. Asymmetric Sensorineural Hearing Loss often presents as diagnostic challenge. Incidence of unilateral SNHL varies in different studies and range from 3.24% to 19.3%. Occasionally Asymmetric Sensorineural Hearing Loss (ASNHL) may be indicative of an underlying retrocochlear disease. In all these cases magnetic resonance imaging (MRI) with cerebellopontine angle is required to evaluate or rule out retrocochlear pathology. Unilateral retro-cochlear pathologies (ex. acoustic neuroma) may be associated with sudden asymmetrical SNHL. Acoustic neuroma is a benign tumor arising from abnormally proliferative Schwann cells, which envelope the lateral portion of the vestibular nerve in the internal acoustic meatus. To identify the incidence of acoustic neuroma on MRI among cases of sudden asymmetrical SNHL. A cross-sectional study was carried out. A total of 21 cases of sudden asymmetrical SNHL that reported to our tertiary care institution during the period Sep 2017 and June 2020 were selected for this study. All patients underwent MRI Brain to rule out acoustic neuroma. In our study, 02 (9.52%) patients with sudden asymmetrical SNHL, were found to have acoustic neuroma on MRI brain. Both the patient had more than 15 dB asymmetrical hearing loss on pure tone audiometry. Hearing loss was unilateral and sudden in onset without any predisposing factors and had associated tinnitus. Patients with sudden ASNHL, inter-aural asymmetry  $\geq 15$  dB HL at any two frequencies between 2,000 Hz and 8,000 Hz or  $\geq 20$  dB HL at two contiguous frequencies are usually associated with an abnormal finding on MRI e.g. acoustic neuroma or retro-cochlear lesions.

**Keywords:** Asymmetric SNHL (ASNHL); Pure tone audiometry; MRI Brain; Acoustic Neuroma; CP angle (Cerebello pontine angle); unilateral SNHL.

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## INTRODUCTION

Asymmetric Sensorineural Hearing Loss often presents a diagnostic challenge. But occasionally it may be indicative of retrocochlear disease [1]. Various etiological factors such as Viral or bacterial infection causing labyrinthitis, CNF infections, head injuries, loud noise exposure, tumors of cerebellopontine angle, meningioma's, ototoxic drugs and idiopathic etiology are known to cause sudden Asymmetric sensorineural hearing loss. The cause is often multi factorial in nature [1, 2]. Pure tone audiometry is the audiological screening as well as a diagnostic test. Various studies have defined asymmetric sensorineural hearing loss as an inter-aural asymmetry of  $\geq 20$  dB at two contiguous frequencies or an inter-aural asymmetry of  $\geq 15$  dB at any two frequencies between 2,000 Hz and 8,000 Hz on PTA [3].

Incidence of unilateral SNHL varies in different studies and range from 3.24% to 19.3%. In India unilateral SNHL affects 7.9% - 13.3% of general population [17]. Occasionally Asymmetric Sensorineural Hearing Loss (ASNHL) may be indicative of an underlying retrocochlear disease [1]. In all these cases magnetic resonance imaging (MRI) with cerebellopontine angle is required to evaluate or rule out retrocochlear pathology. Over 95% of American neuro-otologists reported ordering MRI for patients with suspected ASNHL [2].

Since there are many cases of SNHL with intra cranial tumour documented, MRI has become a routine investigation in evaluation of SNHL to rule out intracranial tumor [2]. Vestibular schwannoma (VS) / acoustic neuroma is the most common tumor of the cerebellopontine angle (CPA) and accounts for 5% to 10% of all intracranial tumors in adults [5, 6]. However, VS is rare, with an overall prevalence of 1 per 100 000, and is found only in 2% to 8% of patients with ASNHL [1, 2]. It is a benign tumour arising from abnormally proliferative Schwann cells, which envelopes the lateral portion of the vestibular nerve in the internal acoustic meatus. While patients with VS often present with the classic symptoms of unilateral hearing impairment, tinnitus, and/or imbalance, up to 45% are asymptomatic [2, 6].

The purpose of this present study is to evaluate the association between clinical and audiometric factors to determine the criteria which can be used to increase the diagnostic yield of the MRI examination in patients presenting with ASNHL.

## MATERIALS AND METHODS

A cross-sectional study was carried out at ENT department of 7 Air Force hospital Kanpur. All patients with unilateral hearing loss that reported to ENT department Sep 2017 to May 2020 were considered as our study population. All these individuals were evaluated clinically to rule out any active middle ear disease, followed by which Oto-neurological examinations were carried out. A detailed history was taken and individuals who are contraindicated for MRI owing to the presence of metallic implants (e.g., pacemakers, aneurysm clips) and claustrophobia were excluded from the study. Audiological evaluation was done for these individuals which consisted of Pure Tone Audiometry (PTA), tympanometry and free field hearing. Of these with unilateral sensorineural hearing loss further underwent MRI brain to rule out cerebellopontine angle lesions. MRI was done on a 1.5-T or 3.0-T machine using either a conventional or acoustic protocol. The conventional protocol included sagittal T1, axial T2 fast spin-echo (FSE), and axial T2 fluid attenuated inversion recovery (FLAIR), axial T1 pre-contrast, axial T1 post-contrast, and coronal T1 post-contrast sequences using 5-mm slice thickness. The acoustic protocol included all sequences from the conventional brain protocol plus high-resolution axial T2, axial T1 pre-contrast, axial T1 post-contrast, and coronal T1 post-contrast sequences of the internal auditory canal and posterior fossa with 3-mm slice thickness. One-millimeter reconstructed axial slices from the 3-dimensional sequences were reviewed. Individuals with CP angle lesions or inner ear anomalies further required contrast enhanced MRI brain study. After attaining normal renal function test these patients underwent conventional post-contrast imaging. Intravenous administration of a standard dose (0.1 mmol/kg body wt) of Gadolinium contrast medium was used for this study. Finally, a total of 21 patients of unilateral SNHL on Pure tone audiometry were included in the study. Other associated clinical symptoms of retrocochlear disease such as tinnitus, vertigo and duration of onset of these symptoms were also recorded.

**RESULTS**

The age of our study population ranged between 18 years to 50 years. The mean age was calculated to be 34.76 years. Among the 21 cases, 12 of them were female and 09 were male. ASNHL was seen almost equally distributed among both the genders with no statistical significance. Of these 21 cases, 06 individuals had associated systemic co-morbidities. 04 individuals had Hypertension and two patients had type 2 Diabetes Mellitus. In our study right sided SNHL was found to be more common, accounting to 57.1% (12 individuals) when compared to left side with only 42.8% (9 individuals). 33% of cases had tinnitus as an associated symptom and 9.5% of patients had giddiness as an associated symptom.

On MRI scan, 9.52% that is 02 patients were found to have acoustic neuroma. Figure 1: MRI showed a well defined homogeneously enhanced extra axial mass lesion in right CP (cerebellopontine) angle with intracanalicular extension along VIII nerve. Minimal widening of internal auditory canal on ipsilateral side was present. Both these patients had asymmetrical hearing loss and tinnitus. Neither of these individuals had any giddiness. Of the 90.5% of non-acoustic neuroma cases, about 26% of patients had tinnitus and only 10% had an associated vertigo as symptom.

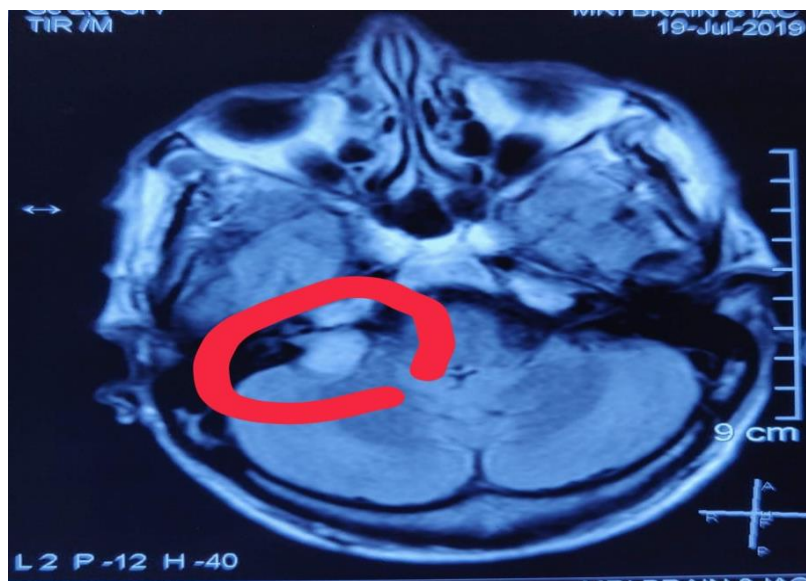
Table 1 shows different variables which can help in deciding for MRI in case of sudden asymmetrical sensorineural hearing loss. Only one patient (4.76%) had Mondini’s dysplasia as an incidental finding on MRI.

**Table 1: Variables for CPA mass prediction**

Indices	No CPA mass (19)	CPA mass (02)
Male	08(42.10%)	01(50%)
Female	11(57.89%)	01(50%)
Right ear	11(57.89%)	01(50%)
Left ear	08(42.10%)	01(50%)
IA asymmetry of $\geq 20$ dB HL at two contiguous frequencies	12((63.15%)	02(100%)
IA asymmetry $\geq 15$ dB HL at any two frequencies between 2,000 Hz and 8,000 Hz	07(36.84%)	02(100%)
Tinnitus	05(26.31%)	02(100%)
Giddiness	02(10.52%)	00

Abbreviations: CPA – Cerebellopontine Angle; IA – Inter-aural

**Figure 1: MRI Brain showing mass in CP angle- Acoustic neuroma**



## DISCUSSION

ASNHL, especially in the presence of unilateral tinnitus, vertigo/disequilibrium, or focal neurological deficits involving the 5th or 7th cranial nerve distributions, there is increased concern for retrocochlear disease [8, 9]. Standard Pure tone audiometry fails to identify retrocochlear disease. In addition, abnormal acoustic reflex testing may suggest retro cochlear abnormality but cannot identify the cause. Findings on DPOAE testing may not help in determining if a retrocochlear disease is present. Unilateral vestibular schwannoma (VS) is a benign tumour arising from abnormally proliferative schwann cells, which envelope the lateral portion of the vestibular nerve in the internal acoustic meatus. As the VS grows, it gradually fills into the internal acoustic meatus and eventually protrudes out of the Porus. Patients with ASNHL may be screened by ABR. The ABR test is sensitive for a vestibular schwannoma larger than 1 cm but is limited in the evaluation of smaller tumors or in patients with significant hearing loss [10]. ABR testing may be useful in certain clinical scenarios, such as in older patients in whom the missed diagnosis of a small tumor may be less consequential or if an MRI is contraindicated owing to the presence of metallic implants (e.g pacemakers, aneurysm clips) [10, 11].

An MRI of the brain and internal auditory canals with gadolinium is the most sensitive test for detecting retro cochlear disease [10]. This study aims to find out role of MRI in asymmetrical sensorineural hearing loss. According to, Mahrous AK1, Kalepu Ra [12] retrospective study of 100 consecutive patients, the diagnostic rate was 1% for CPA lesion which was vestibular schwannoma. However, Kesser BW et al; and Jiang ZY et al; in their study found that Vestibular schwannoma was detected in 2% to 8% of patients with ASNHL [1, 2] which concurs with our study. As we detected 9.52% of cases had vestibular Schwannoma on MRI.

According to Stachler et al,[10] the overall rate of MRI abnormalities directly related to sudden SNHL ranged from 7% to 13.75%, which is similar to our study where 9.52% patients with asymmetrical SNHL were detected to have pathological on MRI. MRI showed a well-defined homogeneously enhanced extra axial mass lesion in CP (cerebellopontine) angle with intracanalicular extension along VIII nerve. Minimal widening of internal auditory canal on ipsilateral side was present. This supports that the concept that acute onset of unilateral hearing loss may increase the diagnostic yield of MRIs.

Clinical symptoms of vestibular schwannoma cases were studied by David M. Baguley, et al [12] and he found that about 76% of patients had associated tinnitus symptom. In our study both the patients of acoustic neuroma had tinnitus as an associated symptom. These two patients with acoustic neuroma were referred to higher centre for further management. Usually, patients with sudden sensorineural hearing loss may improve with corticosteroids or other modalities of treatment but it is always advised to use imaging technique to rule out acoustic neuroma or CP angle lesion. As Nageris BI et al [14] have found that complete recovery of hearing loss in patients with sudden sensorineural hearing loss does not exclude acoustic tumor, and these patients therefore require full evaluation. So in present study, we advised MRI sequences in all the the patients who were presented with sudden asymmetrical sensorineural hearing loss. Wilson YL et al [15] mentions that although radiographic imaging with MRI is not as cost effective, its value in detecting for acoustic neuroma is undeniable. MRI has role not only in diagnosing the vestibular schwannoma but also has role in follow up of the disease. As per Kartikesh and S vsrshney et, at; about 75.8% of MRI brain in unilateral SNHL were found to be normal but MRI brain was diagnostic in 6% of cases [17] , similarly in our study MRI brain was diagnostic in 9.52% of cases and rest were normal. But even a normal MRI aids in diagnosing the cause of asymmetrical SNHL which further helps in definitive management of the patient. The current trend in treatment is from traditional microsurgical resection or stereotactic radiotherapy to conservative management (it is also called wait and scan). Detailed non-contrast MRI has the potential to replace contrast MRI in diagnosing and following growth of VSs in the future [16].

## CONCLUSION

Patients with sudden Asymmetrical SNHL with or without tinnitus/dizziness/vertigo are also more likely to have an abnormal MRI finding like acoustic neuroma or retrocochlear lesions considering the other causes of sudden sensorineural hearing loss have been ruled out. Those patients who present with sudden ASNHL with inter-aural asymmetry  $\geq 15$  dB HL at any two frequencies between 2,000 Hz and 8,000 Hz or  $\geq 20$  dB HL at two contiguous frequencies is usually associated with an abnormal finding on MRI and should undergo MRI of the brain. Gadolinium enhanced contrast MRI or special sequence non

contrast serial MRI brain also have added advantage in “wait and scan” conservative treatment of vestibular schwannoma.

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