

PRESENTATION OF SYMPTOMS AND AUDIOLOGICAL FINDINGS IN SUBJECTS WITH ACOUSTIC SCHWANNOMAS - A RETROSPECTIVE STUDY

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Abstract : Several symptoms such as Hearing loss, head ache, tinnitus etc was associated with patients with acoustic schwannoma. A high-frequency sensory-neural hearing loss is the most common type, occurring in approximately two-thirds of patients. However, in Indian population the presentation of symptoms, the order of symptoms is not known. The present retrospective study was attempted to account for the frequency of symptoms onset, degree of hearing impairment as well as the measures of distortion product otoacoustic emissions, auditory brainstem responses and acoustic stapedius reflex. The analysis was done on 100 confirmed patients (44 females and 56 Males) of acoustic schwannoma. The patients were in the range of 45-60 years with a mean age of 52 years (SD = 2.5). The present study has revealed hearing loss as the most common symptom in 90% of the patients which is in agreement with other studies. The cochlear pattern was observed in 63.4% of patients while 36.6% was non-cochlear type.

Index Terms – *Acoustic schwannoma, Vestibular schwannoma, Acoustic neuroma, Cerebello pontine angle tumor, Tympanometry, Acoustic stapedial reflex, Otoacoustic emission, ABR*

INTRODUCTION

Acoustic neuromas better referred to as vestibular schwannomas¹ is a benign, slow growing, well encapsulated tumour that arises from 8th cranial nerve and usually expands the internal acoustic meatus and spills into the cerebello pontine angle. They occur equally often in the inferior vestibular nerve, and some indicate that they are most common in the superior vestibular nerve. A retrospective study conducted at Japan found that 84.8 per cent of tumours originated from the inferior vestibular nerve, 8.9 percent from superior vestibular nerve, 0.7 percent originated from facial nerve. Remaining 5.6 per cent of tumours could not be determined the origin from superior or inferior.² Acoustic neuroma has an incidence of approximately 12 per million in the United States of America and approximately 20 per million in a contemporary European cohort.^{3,4}

Hearing loss has been reported to be the most frequent symptom, occurring in more than 95 percent of patients. About 90 percent present with a one-sided, slowly progressive hearing impairment. A high frequency sensorineural is the most common type, occurring in approximately two-thirds of patients.⁵ A large multi-national survey-based study identified unilateral hearing loss in 86% of patients, with unsteadiness in 61%, tinnitus in 57% and headache in 36%.⁶ But in this study the main presenting symptom and severity of symptom was not identified. As there is a variation in epidemiology of acoustic neuroma in different parts of the world, the

presentation of symptoms of patients with this disease will also be changing. As the presentation of symptoms and the audiological findings of this disease is not known in Indian population, there exists a need to find out the presentation of symptoms in Indian population. Thus it will help the health care providers in India to be aware about presentation of symptoms, order of symptoms and any new emerging pattern of this disease be it in primary care hospital or a surgical specialty hospital. Descriptions of the signs and symptoms of acoustic neuroma within the literature are largely from older studies, which may not be applicable to contemporary practice. Therefore, the objectives of this study were to describe the most common clinical features associated with subjects with acoustic neuroma. The other objectives are to find out the hearing loss severity by pure tone audiometry, speech discrimination, audiological findings such as DP otoacoustic emission test (OAE), auditory brain stem response (ABR) and Acoustic stapedius reflex (ASR)

METHODOLOGY

The study was conducted in department of audiology & speech pathology, NIMHANS hospital Bangalore. Sample was taken from the patients who were referred from department of neuro surgery to the department of audiology & speech pathology for the pre-operative detailed audiological evaluation. 100 consecutive patients referred to department were taken into the study. Study design was a retrospective study. Subjects with acoustic neuroma who were diagnosed by neurosurgeons with relevant investigations were included in the study. Then Tympanometry test was done to confirm the middle ear clearance.⁷ As middle ear pathology affects the Audiological examination results, those who failed the test were excluded from the study. Subjects were interviewed by the first author. Subjects were asked to recollect the initial symptom during the course of the disease. They were also asked to recollect what the other symptoms were. Detailed medical history and history of symptoms have been documented from these patients. The details of previous audiological examination such as pure tone audiological evaluation DP otoacoustic emission test (OAE), auditory brain stem response (ABR) and Acoustic stapedius reflex (ASR) have been collected from the medical records as well as from the patient's side. Data was also collected were onset of each symptom, order of occurrence of symptoms and previous audiological results.

The degree of hearing loss was classified on the basis of all frequency average with the assumption that it would depict the hearing acuity close to the audiometric configuration. For classification in terms of the degree of hearing loss modified Goodman's classification by Clark (1981) was used as reference.⁸ The speech discrimination score (SOS)⁹ wherever permissible were also classified as normal (>90%), mild (80-89%), moderate (70-79%), severe (50-69%) and profound (<49%). The inter-peak latency of the ABR was analyzed wherever feasible on the lesion side as well as on the non-lesion side as either normal, delayed or absent.¹⁰ The ASR measurements were analyzed based on Jerger classification^{11, 12} and the DPOAE were also analyzed wherever available in terms of either present or absent.¹³ Descriptive statistics was used to summarize the data in percentages.

RESULTS AND DISCUSSION

Results

This is a retrospective study to account for the frequency of symptoms, onset, and degree of hearing impairment as well as the measures of distortion product otoacoustic emissions (DPOAE), auditory brain-stem responses (ABR) and acoustic stapedius reflex (ASR). The analysis was done on 100 patients (44 Females and 56 Males). The patients were in the range of 45-60 years with a mean age of 39.26 years (SD=12.2).

Symptom analysis: Symptom analysis done on 100 AN patients revealed the occurrence of 14 prominent symptoms. The

percentages of occurrence of each of these symptoms were analyzed. Hearing loss appeared to be the most prominent symptom occurring in 90% of patients, followed by headache in 77%, swaying in 72%, blurring of vision in 50%, vertigo, tinnitus, vomiting and hypesthesia of face in the 30% range, followed by slurring of speech and weakness of extremities in the 20% range. Swallowing difficulty, angle of deviation of mouth and nasal regurgitation was 18%, 14% and 12% respectively. Bowel and bladder incontinence was the rare symptom appearing in 5% of patients (Table 1). The individual symptoms were classified into six symptom groups based upon the order of occurrence. Hearing loss appeared as first symptom in 64% of patients, followed by headache in 36%, tinnitus in 19% and vomiting in 10% of patients. Thus the analyses revealed the general order of occurrence of symptoms as hearing loss, headache, tinnitus, vomiting, swaying, vertigo, weakness of extremities, swallowing difficulty, bowel and bladder incontinence, nasal regurgitation, blurring of vision, hypesthesia of face, slurring of speech, and deviation of angle of mouth.

Table-1 Percentage of symptoms observed in the present study

Sl. No	Symptoms	Percentage
1	Hearing loss	90%
2	Headache	77%
3	Swaying	72%
4	Blurring Of Vision	50%
5	Vertigo	39%
6	Tinnitus	39%
7	Vomiting	31%
8	Hypesthesia Of Face	30%
9	Slurring Of Speech	29%
10	Weakness Of Extremities	20%
11	Swallowing Difficulty	18%
12	Deviation Of Angle Of Mouth	14%
13	Nasal Regurgitation	12%
14	Bowel And Bladder Incontinence	5%

Puretone audiometry: The conventional audiometry in 93 patients (out of the total 100 patients) with unilateral acoustic schwannoma revealed profound degree of hearing loss, greater than 90 dB, in 63.4% of the patients, greater than 90dB. In the remaining the distribution of degree of hearing acuity was in the order of; severe in 15.1%, moderately severe in 3.2%, moderate in 6.5%, mild in 4.3% and normal in 7.5%. In the remaining seven patients out of 100, pure tone audiometry could not be carried out in four and in three who had bilateral acoustic schwannoma had unilaterally profound hearing loss and mild hearing loss contralaterally in one patient, bilaterally profound degree in another patient and in the third patient unilaterally severe degree with contralateral ear having mild SN hearing loss (Table 2).

Table -2 Classification of hearing loss by pure tone audiometry

Degree of loss	Percentage of patients' (lesioned side)	Percentage of patients' (non lesioned side)
Profound	63.4%	2.2%
Severe	15.1%	----
Moderately severe	3.2%	2.2%

Moderate	6.5%	5.4%
Mild	4.3%	18.3%
Normal	7.5%	72%

Similarly, in the non lesioned ear (ear contralateral to the tumour side) in the 93 patients, the distribution of hearing acuity was, normal in 72%, mild in 18.3%, moderate in 5.4%, moderately severe in 2.2% and profound in 2.2% patients.

Speech discrimination: Profound discrimination loss compatible with the degree of hearing loss was found in 58 patients (62.4%) out of 93 patients. In four patients the test was not administered. In the remaining 35 patients, seven demonstrated normal discrimination, and 26 patients showed varied discrimination score not in agreement with the degree of hearing level. Similarly, in the non-lesioned ear 78 patients (80.4%) had normal discrimination score with 13 patients having profound loss of speech discrimination and of them nine patients were not compatible with the degree of hearing loss. In the remaining six, varied discrimination patterns were observed disproportionate to the degree of loss. Bilateral acoustic schwannoma patients' showed profound discrimination loss bilaterally in two and one patient showed mild discrimination loss on one side and profound loss on other side.

DP Otoacoustic emission: The analysis of the distribution of DPOAE for its presence or absence in the lesioned ear side which could be obtained on 71 patients revealed absence of DPOAE in 43 patients (60.6%) who demonstrated moderate to profound degree of hearing impairment. On the contrary, 26 patients (36.6%) had presence of DPOAE with degree of hearing level varying from mild to profound. Amongst them, 15 patients (57.7%) had severe to profound degree of hearing impairment. In two patients the DPOAE was abnormal where puretone thresholds could not be ascertained. In 26 (26.8%) patients the testing was not carried out. Similarly, in the non-lesioned ear side, of the 97 patients (excepting 3 with bilateral acoustic schwannoma), 48 (66.7%) patients showed the normal patterns of emission as against 24 (33.3%) patients who demonstrated the absence of DPOAE. In the remaining 25 patients the test was not carried out (Table 3).

Table-3 Dpoae, lesioned side vs non-lesioned side comparison

DPOAE		Lesion side	Non-lesion side
	Present		36.6%
Absent		60.6%	33.3%

Auditory brainstem response: The ABR was not done in 77 patients on the lesioned side and was abnormal in the remaining 20 patients. With respect to the degree of hearing loss, the ABR was abnormal in three patients with normal hearing, four with moderate hearing loss, five with severe hearing loss and eight with profound degree of hearing impairment.

Similarly in the non-lesioned ear the ABR was not done in 72 patients. In the remaining it was normal in six patients while 19 patients demonstrated abnormality (Table 4). The two bilateral acoustic schwannoma cases showed absent ABRs bilaterally and in one patient the test was not carried out.

Table-4 ABR, lesioned side vs non-lesioned side comparison

ABR		Lesion side	Non-lesion side
	Normal		0%
Abnormal		100%	76%

Acoustic stapedius reflex (ASR): The testing could not be carried out in 61 patients. In the remaining 39 patients, bilateral absence was found in 35%, a diagonal pattern was observed in 38% and inverted 'L' in 23%. In bilateral acoustic schwannoma patients', the testing was not carried out.

Discussion

As mentioned by Cushing (1917),¹⁴ the hallmark of AN is auditory dysfunction with hearing loss or tinnitus. The present study has revealed hearing loss as the most common symptom in 90% of the patients, which is in agreement with other studies (Jackler and Pfister, 2005).⁵ This has been found to be the first clinical symptom in majority of our patient population. In an attempt to explain the different types of hearing losses seen in AN, Lehnhardt (1990)¹⁵ suggested a theory involving both myelin and axon compression damage. He theorized that "early in the course of compression, the myelin damage might be the only lesion, allowing for tone decay and acoustic reflex decay without recruitment. Auditory brainstem evoked responses would also be delayed. This would be the typical picture of AN. Later on, as myelin and axon compression both become involved, the ABR would be delayed, still without recruitment. If the tumour compression changes, for instance, in cessation of tumour growth or recovery from intra tumour hemorrhage, enough axons may remain to allow for adequate nerve conduction and remyelination may occur. Therefore recruitment may be positive, the ABR may be positive, but tone decay may not occur."^{16, 17}

Number of other mechanisms^{18, 19} that have been implicated for the cause of hearing loss in tumours of the eighth nerve as well as cerebello pontine angle (CPA) are direct compression of the eighth nerve, stretching of the nerve, vascular compression or occlusion of blood supply to the eighth nerve to the cochlea, damage to cochlear efferents, biochemical changes within the inner ear and hemorrhage within the nerve or into the tumour; all may have place in the etiology of hearing loss. The rate and amount of hearing loss vary depending upon the rate of tumour growth, the plasticity of the nerve, the consistency of the tumour, the location within the internal auditory canal (IAC) and the amount of early expansion into the CPA.

In the present study, 63.4% of patients have demonstrated profound degree of hearing loss greater than 90 dB in the tumour ear and in contrast seven patients had normal hearing. The patients who have demonstrated profound degree might have reported to us when other neurological symptoms started bothering them and may be rendering them incapacitated. In the remaining patients varied degree of hearing loss have been demonstrated including the seven patients having normal hearing. In these patients, except the patients with normal hearing, all have demonstrated high frequency slope. As Schuknecht and Woellner (1955)²⁰ put it, if the organ of corti is intact, 75% of the auditory nerve fibers need to be destroyed' before pure tone hearing is affected. This mechanism explains slowly progressive hearing loss of CPA tumours and also partially explains the middle and high frequency hearing losses associated with eighth nerve tumours.

Similarly, in the non-lesioned ear (ear contralateral to the tumour side) in the 93 patients, the distribution of hearing acuity was normal in 72%, mild in 18.3%, moderate in 5.4%, moderately severe in 2.2% and profound in 2.2% patients.

In the lesioned side profound discrimination loss compatible with the degree of hearing loss was found in 62.4% patients out of 93 patients who demonstrated a loss more than 90dB. In 4 patients the test was not administered. In 28 patients the discrimination score was not compatible to the degree of hearing level thus suggesting the neural involvement.

Similarly, in the non-lesioned ear 80.4% patients had normal discrimination score with 13.4% patients having profound loss of speech discrimination. In the remaining 6.2%, varied discrimination patterns were observed.

Historically no correlation has been found between the degree of hearing loss (HL) or speech discrimination (SD) deficit and tumour size. The very poor SDS generally supports a hearing mechanism of neural compression. Thomson et al (1983)²¹ discussed the concept that discrimination at high presentation levels is primarily an inner hair cell function, and 95% of fibers in the auditory nerve originate in the inner hair cells, therefore the neural compression would inhibit hair cell function with the

resultant poor SD, especially at increasing presentation levels.

Analysis of our patients on DPOAE measurements revealed the absence of DPOAE in 43 patients (60.1 %) (out of 71 patients tested for DPOAE) in whom the degree of hearing loss varied from moderate to profound degree. The absence of DPOAE suggests the involvement of the cochlea. However 26 patients (36.6%) have demonstrated the presence of DPOAE in whom 15 patients had a profound degree of hearing impairment. This implies the normal functioning of the outer hair cells. Thus profound hearing loss may be due to the severe compression of the nerve sparing the vascular supply to the cochlea. Telischi (2000)²² reported on 97 patients with AN who underwent DPOAE testing. He found that 37% to 57% of tumours were of a cochlear loss pattern and 41%-59% was retrocochlear pattern. He concluded that the majority showed evidence of reduced outer hair cell function in at least one frequency. In our study cochlear pattern was observed in 66.6% and non-cochlear in 38.6%.

In our study 33.3% patients in the non-lesioned side also have demonstrated the absence of DPOAE suggesting the contralateral vascular compression resulting in cochlear disorder. This is expected in patients with large tumours and its strategic location, which could cause severe compression on the brainstem distorting the vascular supply to the cochlea in the non-lesioned ear.

Marangos et al (2001)²³ studied 50 AN patients who had normal ABR, found 20% of them having normal hearing (PTA) and 18% having binaurally symmetrical hearing. In our study the ABR was done only on 20 patients. With respect to the degree of hearing loss, the ABR was abnormal in three patients with normal hearing, four with moderate hearing loss, five with severe hearing loss and eight with profound degree of hearing impairment. Excepting the patients with profound degree of hearing impairment where the stimulus was not sufficient to stimulate the auditory nerve, patients have demonstrated abnormal ABRs. This implies the highest sensitivity of the ABR in demonstrating the eighth nerve pathology. Interestingly the abnormality of the ABR pattern was also observed in 19 patients (out of 25) contralateral to the lesion side suggesting the pressure effect of the tumour on the brainstem distorting the auditory synchrony. Grabel et al(1991)²⁴ suggested that the chronic effect of high tumour volume within the infratentorial compartment might also play a role in AN hearing loss. Eggermont et al (1980)²⁵ suggested that the abnormal ABR was probably from abolition of the synchronized firing of nerve fibers rather than a prolongation of the nerve conduction velocities. This was especially true for the high-frequency fibers, which run around the outside of the auditory nerve, versus the middle and apical turn fibers, which are positioned more toward the middle of the nerve and possibly are less disturbed in the early stages of tumour growth.

The limitation of the study was the sample size was small. Another limitation was that many of the patients reported with hearing loss, the audiological findings were not documented in case sheet. ABR test was not done for many of the patients. As the test was time consuming and this facility was not there in many of the hospital. This could be the reason why these reports were not documented in the case sheet. The present study evaluated only initial symptoms and audiological findings. The changes in symptoms and audiological findings over the period of time were not examined. As the tumour progresses, the symptom order and disease severity may change. This could have affected the present study since the study was done retrospectively. During the study period tumour size and location may change. This may result in change in presenting symptoms. The length of time to presentation is an important factor in tumour size, as longer length of symptom duration is associated with larger tumours.⁶ Size and location of the tumour were not documented during data collection. This may not be uniform among subjects in the present study. So these variables would have affected the results. There is a need for correlating the audiological deficits with that of the tumour location and its extension, volume and the nature of the tumour per se. Further study can be done to find out the association of symptom with tumour size.

Conclusion

The objectives of this study were to describe the most common clinical features associated with subjects with acoustic neuroma. The other objectives are to find out the hearing loss severity by pure tone audiometry, speech

discrimination, audiological findings such as DP otoacoustic emission test (OAE), auditory brain stem response (ABR) and Acoustic stapedius reflex (ASR). The present study has revealed hearing loss as the most common symptom in 90% of the patients which is in agreement with other studies. The cochlear pattern was observed in 63.4% of patients while 36.6% was non-cochlear type. The present retrospective study emphasizes the importance of recording the symptom evolution, which could guide in understanding the impact of lesion as well as understanding the underlying pathophysiology of the auditory behavior in patients with acoustic schwannoma.

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