ORIGINAL ARTICLE

Frequency of Acoustic Neuroma / CP Angle Tumor in Unilateral SNHL

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ABSTRACT

Aim: To determine the frequency of acoustic neuroma /CP angle tumor in patients with unilateral SNHL in 100 patients in 15 yearsin a teaching hospital in Lahore

Methods: This study was conducted from 2000 to 2014 involving 105 patients presenting with unilateral SNHL but without any history of head injury, surgical trauma, mumps and congenital cause of unilateral SNHL.

Patient's demographic data, age and gender were recorded.The SNHL was detected through clinical examination, tuning fork tests and confirmed through PTA and tympanometry. MRI Brain of all patients withunilateral SNHL was done to rule out acoustic Neuroma / CP angle tumor.

Results: Among 100 patients 90 were males and 10 were females with female to male ratio 1:6 and age ranging from 20 to 60 year.AcousticNeuroma and CPangle tumor was found in 4 patients (4%) as compared to 96(96%) in which no cause was found. Out of these 4 patients, 2were male (age 40 and 45 years) and two were females (age 20 and 22 years). One patient had acoustic neuromaand three had CP angle tumor.

Conclusion: Considering the acoustic neuroma and CP angle tumoras a possible cause of SNHL, it is suggested that all patients presenting with unilateral SNHL, should have MRI brain and internal acoustic meatus done to rule out acoustic neuroma and CP angle tumor. Tests like CT Scan brain is of no valuebecause it fails to pick up acousticneuromaspecially if it is less than 5mm of size. Other tests like BERA can be suggestive but not conclusive. Further BERA if normal does not 100% rule out acoustic neuroma.In one of our case, BERA of a patient (doctor and paediatrician) was done and was reported to have cochlear dysfunction, as a cause of unilateral SNHL,and was advised hearing aid by an audiologist of a teaching hospital , which later turned out to be acoustic Neuroma on MRI brain.Therefore BERA cannot be entirely relied upon to differentiate between cochlear & retro cochlear hearing loss.

Keyword: SNHL (Sensorineural hearing loss), Acoustic Neuroma, CP (Cerebello-pontine) angle tumor

INTRODUCTION

Unilateral Vestibularschwanoma is a benign tumor arising from abnormally proliferative schwann cells which envelope lateral portion of vestibular nerve in internal acoustic meatus. The term acoustic neuroma is replaced by the term Vestibular schwanoma(Eldridge and Parry 1992). Acoustic Neuroma represent 80% of CP angle tumours. They arise from Schwann cells and virtually always originate within internal acoustic meatus. The commonest nerve of origin is superior vestibular nerve (Nager 1969).

Histopathological examination of vestibular Schwanoma reveals two morphological pattern. The Antoni A pattern in which there are closely packed cells with small spindle shaped and densely stained cells. A whirlad appearance of Antoni type A cells is calledverocay body. This contrast with Antoni B pattern in which there is looser cellular aggregation of vacuolated pleomorphic cells. In any particular vestibular Schwanoma, one type of cellular pattern may predominate or both types can be completely admixed.

Etiology of vestibular schwanoma is not known. Defect of chromosome 22q may be responsible.As the tumor grows it fills the IAM and eventually protudes out ofporus. Extra metal expansion results in displacement and stretching of seventh and eighth nerve on the anterior aspect of tumor and on the (AICA) on the inferior aspect.After further growth the tumorexpands to touch& compress cerebellum and trigeminal nerve.Further compression and displacement of brain stem and the 4th ventricle which leads to hydrocephalus. AICA and lower cranial nerves are also displaced and become adherent to inferior surface of tumour, overtime 5th and 6th nerve become stretched over the surface of the tumour.

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Symptoms and signs: Symptoms are unilateral deafness or tinnitus or both.Discomfort in the ear and mastoid is common. Hearing loss is usually gradual and slowly progressive over a period of time varying from two months to twenty years (King and Gipson) and is more in higher tones. In 5-10% cases hearing loss is sudden and may be profound. Other non acoustic symptoms such as vertigo, trigeminal or cerebellar symptoms can occur (Thomsom, Tos and Moller 1992).

Later mid facial and corneal hypoesthesia, headache, ataxia and lower cranial nerves symptoms appear. ystagmus either vestibular or cerebellar, signs of fifth and seventh nerve involvement can occur. Long tract signs, weakness of limbs as well as visual loss and severe general headache can also occur. Diagnosis is usually made by first getting PTA which shows SNHLin affected ear. Canal paresis can be detected by caloric test and also by clinical test called head impulse test. This test can detect severe canal paresis but not mild or moderate canal paresis and will not replace caloric test but it is useful adjunct to it (Beyon, Jani or Baguley 1998). Further MRI with gadolinium enhancement is needed.

DISCUSSION

Patient presenting in ENT OPD with gradual onset unilateral hearing loss,detected as SNHL on Tuning forktests and PTA,can be due to head injury, trauma or mumps,But there are many cases without such history. In our casemajority of patientscame with no history ofmumps, ear surgery, or head injury.Majority of them presented with gradual and progressive hearing loss and tinnitus in one ear.However some patient presented with tinnitus in one ear lasting for few month.

One of the patients had,in addition to hearing loss and tinnitus,feeling of pressure in his left ear and slight degree of unsteadiness on walking occasionally but not always.

Another patient had,in addition to tinnitus and deafness,hypoesthesia of right side of face and right sided facial weakness(misdiagnosed as bell'spalsy and treated byphysiotherapyand by electrical stimulation of facial muscles).

Another patient,initially complaining of hearing loss and tinnitus,later developed unsteadiness on walking and visual problem. Initially this patient was treated by ENT specialist with betahistine for several weeks. In this case MRI was advised which showed a large CP angle tumour pressing brain stem and cerebellum causing hydrocephalus. Ophthalmoscopy showed marked papillodema. After VP shunting her clinical condition improved.Regarding CP angle tumor,neurosurgeon decided to go for debulking of tumor rather than complete removal as according to them this case could havehigh mortality.

Our first patient who wasa doctor and consultant paediatrician initially consulted an ENT specialist, for his left ear hearing loss tinnitusand feeling of aural fullness, who performed tuning fork tests and declared him asa case of conductive hearing loss & tubal catarrah and treated him for several months for his suspected tubal catarrah. When this patient consulted us, our tuning fork tests showed hearing loss of SNHL type, which was confirmed as SNHL ofmoderate degree through PTA. We further suggested MRI scan brain to rule out acoustic neuroma. He ignored our suggestion for several weeks. Later he consulted an audiologist who performed BERA and suggested that his hearing loss was due to cochlear rather than retro-cochlear pathology and therefore he just needs hearing aid and there is no acoustic neuroma or cp angle tumor .Later as his symptoms were worsening, he got his MRI scan done on ours and on advise of ENT surgeon in UK, which showed acoustic neuroma of about 1.8cm. He got his surgery done in America, and as complication he initially developed CSF Rhinorrhoea and later meningitis, but recovered from these problems. Now at present he has no problem and follow up MRI scan is clear. However he has no hearing in left ear because of translabrynthine approach for his surgery.

The importance of performingtuningfork test correctly: is being emphasized. Rinne test should be performed with masking of other ear, if negative, to rule out fasle Rinne negative. Secondlyin Weber testtuningfork should be applied over bone withsufficient pressure to cause slight discomfort to the patient to note the lateralization. Just putting the tuning fork over the bone and saying Weber non-conclusive is a mistake.

Role of Proper PTA with masking: It is suggested that in any patient,after assessment of his hearing with tuning fork tests, pure tone audiometry if asked,should be done by a proper qualified audiologist with proper masking, otherwise a case of SNHL will appear as conductive hearing loss in that ear. Example is a case who was detected as having SNHL in left ear as detected by tuning fork test . When this patient audiometry was performed by a non- qualified person, he reported conductive hearing loss in the left ear as shown in audiogram 1.The same patient PTA was then performed by a qualified audiologist with proper masking and report turned out to be SNHL in the left ear as shown in audiogram 2. This shows importance of PTA with proper masking. If PTA is not done with proper masking, then a SNHL may be reported as a conductive hearing loss putting the patient on totally different treatment.

Role of BERA: It is also suggested that tests like normal BERA or BERA showing cochlear dysfunction or tone decay test showing absent tone decay do not rule out acoustic neuroma or CP angle tumor. It is observed that acoustic Neuroma by compressing labyrinthine vessels can produce changes in cochlear cells which will manifest as cochlear pathology rather retro cochlear in early stages, as a cause of unilateral SNHL, when BERA is performed on such patients.

Role of MRI Scan Brain and Internal acoustic meatus: Therefore, any patient with unilateral SNHL on tuning fork tests and pure tone audiometry with proper masking should have an MRI scan brain and internal acoustic meatus to rule out acoustic neuroma.

CONCLUSION

From the study we have drawn the conclusion that any patient presenting with audio-vestibular symptoms such as tinnitus,deafness in one ear or vertigo, one should keep in mind to rule out acoustic neuroma/cp angle tumor as a cause or possibility when there is no history of mumps, acoustic trauma, or surgical trauma to that ear. For all the patients with audio-vestibular symptoms, we should have proper assessment comprising of history, general physical examination, systemic and ENT examination,with properly performed tuning fork test and pure tone audiometry. When examination suggest a unilateral hearing loss of SNHL type or unilateral tinnitus and vertigo with no obvious cause,PTA with proper masking is mandatory, and if SNHL in one ear or asymmetrical SNHL is found on PTA, the MRI scan with gadolinium enhancement is required to pick acoustic Neuroma or CP angle tumour.

Therefore one should always ask for MRI scan brain with gadolinium enhancement to rule out acoustic Neuroma in a patient with unilateral SNHL rather relying on test like tone decay and BERA which may be suggestive but not diagnostic . Also CTscan brain cannot be relied upon as it can miss intra-canalicular tumor unless there is bony expansion. Investigation of choice is MRI scan with gadolinium enhancement.

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