Impact of Vestibular Schwannomas on Cerebrospinal Fluid (CSF) Pathway

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Abstract

Background: *Vestibular schwannoma is a slowly growing non-cancerous tumor of the vestibular components of eighth cranial nerve. This tumours are located in cerebellopontine angle region of the posterior fossa. As the tumor enlarges in size it may cause obstruction of cerebrospinal fluid pathway at the level of fourth ventricle.*

Objective: *The purpose of this study was to evaluate the size of Vestibular schwannoma with their impact on Cerebrospinal Fluid (CSF) pathway.*

Methodology: *This study was designed as cross sectional study of 50 patients of 6-60 years (27 males and 23 females). Data were collected from patients with unilateral non-recurrent vestibular schwannomas over two and a half years period from January 2003 to June 2005 in Neurosurgery Department of Bangabandhu Sheikh Mujib Medical University. The tumor size was measured in three axes that is the diameter parallel to the petrous ridge, perpendicular to the petrous ridge or the vertical diameter in coronal slices. The size of the vestibular schwannoma was taken as the largest diameter of extrameatal tumor in any one of these three axes. The analysis of the demographic parameter and factor affecting the circulation of CSF were carried out.*

Results: *Twenty eight patients (56%) had giant, 36% had large and 6% of patients had medium sized tumors. There was no useful hearing in 96% patients. Features of raised ICP (headache, papilledema/ optic atrophy) was observed in 82% cases, cerebellar dysfunction were in 78%. Other features were of trigeminal dysfunction (54%), facial nerve paresis (18%) and planter extensor response (22%). Forty one patients (82%) had hydrocephalus and among them approximately one-quarter were blind. Size of the vestibular schwannoma was important factor for hydrocephalus. Those who had tumors > 3 cm, more than two-third of them had hydrocephalus while those who had tumors 3 or below 3 cm only 2.4% of them demonstrated hydrocephalus suggesting that greater the size of the tumor the higher is the chance of occurrence of hydrocephalus.*

Conclusion: *It was revealed that Vestibular schwannomas with larger than 3 cm are strongly associated with obstruction of CSF circulation.*

Keywords: *Vestibular Schwannoma, Cerebrospinal Fluid (CSF) Pathway, Hydrocephalus, Tumor Size.*

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Introduction:

Vestibular schwannoma (VS)s are benign neoplasm of the vestibular components of eighth cranial nerve that never turn into malignancy[1]. These tumors are the most common cranial nerve schwannoma representing more than 90 percent. Though these tumors account for approximately 6-8 percent of all primary intracranial tumors, vestibuiar schwannomas are the most common mass of the cerebellopontine angle (CPA), representing between 80-90 percent [2]. Vestibular schwannomas affect about 10 people in one million per year[3]. These tumors are usually unilateral. Roughly 5% of patients with vestibular schwannomas are bilateral. The presence of bilateral vestibular schwannoma is considered pathognomic for neurofibromatosis type II[5]. The symptoms of vestibular schwannomas arise from compression of local structures within or at boundaries of the cerebellopontine angle. The symptom complexes follow the size and growth of the tumor. Patients usually present with hearing loss, tinnitus, dysequilibrium, headache, and facial numbness. Other more rare symptoms may also manifest such as facial weakness, nausea and vomiting, otalgia,

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and taste abnormalities. The progression of these symptoms is usually slow and insidious, and often patients will not present to medical attention until their tumors have become quite large[6]. The radiological features of vestibular schwannoma have been well documented. The anatomic location of the tumor is usually centered about porus acousticus. The tumors appear on MR studies is slightly hypo-to-isointense to brain on T1 - weighted imaging and the signal intensity is decreased on T2 - Weighted imaging. Patients with large VS may have secondary obstructive hydrocephalus and occasionally significant neurologic deficit develops[7]. The aim of this study was to investigate the incidence of hydrocephalus and clinical presentations as well as their relation to size of VS.

Methodology:

This Cross-sectional study based research was performed on 50 unilateral non-recurrent vestibular schwannomas patients since January 2003 to June 2005 at Department of Neurosurgery, BSM Medical University. Sample collection of this study was done by both Inclusion and Exclusion Criteria. The inclusion criteria incorporate patients with clinical and radiological evidence of CPA tumor in all ages and sexes. Otherwise, Patients with CPA tumor but not proven as vestibular schwannoma histopathologically were in exclusion Criteria. The evaluation of all patients were performed on the basis of detailed history of illness, meticulous physical examinations, rational neuro-imaging (CT scan/ MRI of Head) and finally were confirmed by histopathological report. The tumor size was measured (CT Scan/ MRI of Head) in three axes that was diameter parallel to the petrous ridge, perpendicular to the petrous ridge or the vertical diameter in coronal slices. The size of the vestibular schwannoma was taken as the largest diameter of extrameatal tumour in any one of these three axes. Tumor size was classified as small (<2 cm), medium (2-3 cm), large (3-4 cm) and giant (>4 cm) (Jackler et al, 1990). The images proven ventriculomegaly associated with tumor mass in CPA region are the way to determine the hydrocephalus. The variables such as age, sex, clinical presentations, clinical findings, hydrocephalus, location of tumor, and size of tumor were entered into a SPSS (Statistical Package for Social Sciences) version-11.5. Data were analyzed using descriptive statistics as well as Fisher's exact probability test and presented in descriptive, tabular and graphical form.

Result:

Patients were categorized in five groups based on their age from 6 to 60 years. Among them males were a higher (54%) than the female (46%).

This study, showed the age distribution of the patients. Peak age incidence of VS (36%) was found to lay between 25 - 35 years, followed by 26% between 35 - 45 years, 20% between 45 - 55 years. Some 10% were 55 years or above and 6% between 15-25 years. Very few 1(2%) was below 15 years (Table 1).

Table-I

Distribution of patients (n=50)

Age (yrs)	Frequency	Percentage
5-15	01	2.0
15-25	03	6.0
25-35	18	36.0
35-45	13	26.0
45-55	10	20.0
e"	05	10.0
Total	50	100

*Mean ±SD=36.0 ± 1.60 (Range 6 to 60 years)

In this study, majority (96%) of the patients exhibited hearing loss and the rest (4%) did not have such experience. Ninety percent of the cases had vertigo or imbalance, while the rest (10%) did not have such problem. Majority (82%) of the patients did not have facial weakness. Only 18% of the patients had such problem. Sixty two percent of the patients developed tinnitus, while the rest (38%) did not have such complaints. Only 8% of patients complained of either nasal regurgitation or dysphagia. Majority (92%) did not experience so. More than 80% of the cases complained of headache, while the rest (18%) did not have such complaints. Nearly half (49%) of the patients had hypoesthesia, while the rest (51%) did not have the same problem. More than half (56%) of the patients experienced occasional vomiting, and the rest (44%) did not have such experience. (Figure 1).

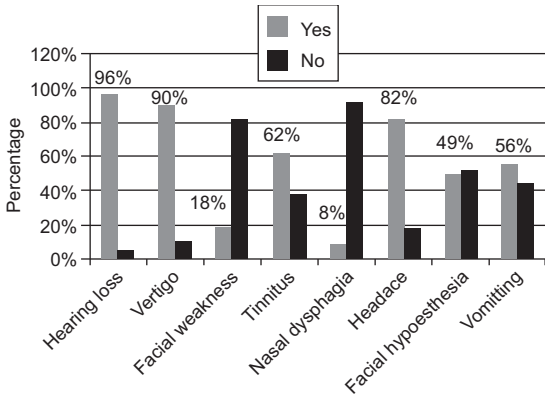

Fig.-1: *Symptoms of the Patients*

Table-II

Distribution of patients by tumor size (n=50)

Group	Tumor Size	Percentage
Giant	>4 cm	56
Large	3-4 cm	36
Medium	2-3 cm	6
Small	<2 cm	2
Total	100	100

Fifty six percent of the patients had tumors of >4 cm size, followed by 36% 3-4 cm, 6% 2 - 3 cm and 2% <2 cm (table: 2).

Fisher's Exact test was done to analyze the data and p-value <0.05 was considered significant. Table 3 describes the association between size of tumor and hydrocephalus. Those who had tumors > 3 cm, more than two-third (97.6%) of them had hydrocephalus while those who had tumors 3 or below 3 cm only 1(2.4%) of them demonstrated hydrocephalus suggesting that greater the size of the tumor the higher is the incidence of hydrocephalus ($p < 0.001$) (Table III).

Table-III

Association between size of tumor and frequency of occurrence of hydrocephalus (n=50)

Size of Tumor (cm)	Hydrocephalus				P-value
	Present		Absent		
	Frequency	Percentage	Frequency	Percentage	
<3 cm	1	2.4	6	66.7	<0.001
>3 cm	40	97.6	3	33.3	
Total	41	100	9	100	

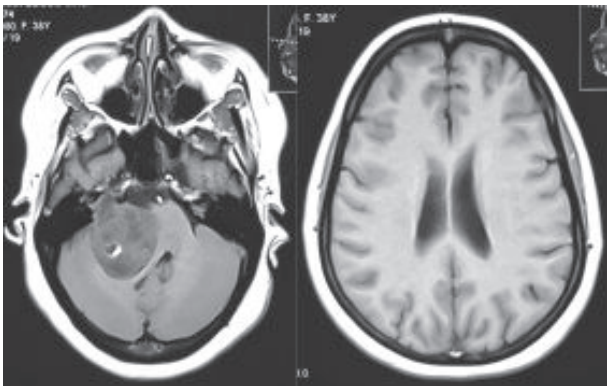

Fig.-2: *Vestibular schwannoma (Right CPA) without CSF pathway obstruction.*

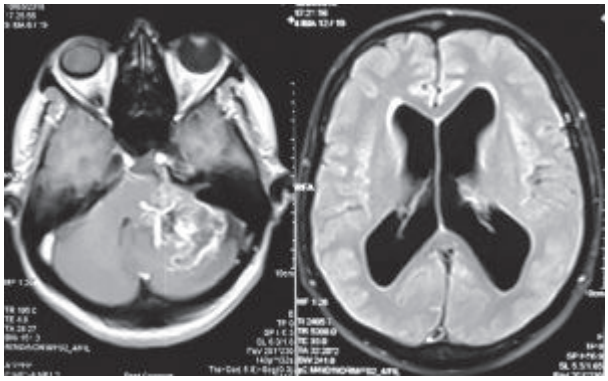

Fig.-3: *Vestibular schwannoma (Left CPA) with CSF pathway obstruction.*

Discussion:

The cerebellopontine angle is a small area located in the posterior fossa of cranial cavity. Vestibular schawannomas are the most common lesion of CPA. Hearing loss is the most frequent symptoms, occurring in more than 95% patients with vestibular schwannoma. In this study, out of 50 patients only two patients (4%) had no hearing loss but the majority exhibited hearing loss. Tinnitus is very common in vestibular schwannoma. It is usually unilateral and confined to affected ear. In a western study, it was 70%. In our study, 62% of patients had complained of this symptom. Imbalance & vertigo are common because the tumor origin is nearly always from vestibular nerves. Harners and laws noted that 67% of patients with acoustic neuroma presented with vertigo/imbalance. In our study, only 10% of patients did not have such complains. Headache in patients with VS, usually results from increased intracranial pressure (ICP). But it may also occur in the absence of ICP, due to pressure or traction upon blood vessels.

In their series Edwards & Patterson found that 84% of patients with VS had headache. In our study, 82% of patients had complained of headache and 44% of patients were not associated with vomiting. In their series, Thomsen and Tos found overall Incidence of trigeminal dysfunction in vestibular schwannomas was 67%[8]. In our study more than half of patients (54%) had the signs of trigeminal nerve dysfunction. Facial weakness is a rare or late occurrence, even though the VIIth nerve is almost always distorted early; whereas facial numbness occurs sooner once trigeminal compression occurs (often in the presence of normal facial movement), despite the fact that the Vth nerve is farther away[9]. In our series, VIIth nerve dysfunction was 18% where more than half of patients (54%) had Vth nerve dysfunction. Involvement of lower cranial nerves, especially the glossopharyngeal (IX CN) and vagus (X CN), usually occurred late, and manifest as palatal paresis, hoarseness of voice and dysphagia. Lower cranial nerves involvement was reported in up to 20% of patients with acoustic neuroma[10]. In our study, 16% of patients were presented with lower cranial nerve palsy. In an earlier study, Thomsen & others revealed that with lesions larger than 4cm, about 60% of patient with VS were exhibited with cerebellar dysfunction[4]. In our study, approximately 70% of patients with VS who had cerebellar dysfunction exhibited >4cm size of tumor, where as about 9% of those did not have cerebellar dysfunction had >4cm size of tumor. A large tumor can cause compression of brain stem with resultant contra lateral/bilateral planter extensor response. In a western series, Babinski's signs were found in 5% patients with VS, but in our series, more than one fifth (22%) cases revealed extensor planter response. Vestibular schwannoma compresses the cerebellum and gives rise to signs of cerebellar dysfunction. Jain and others reported that they found cerebellar dysfunction in 88% of patients with acoustic neuroma[11]. In our study, more than three-fourth (78%) cases exhibited cerebellar dysfunction. Larger tumor may cause brain stem compression as with involvement of other neural structure. As the brain stem compression becomes severe, the fourth ventricle collapses and ultimately resulting in obstruction of cerebrospinal fluid (CSF) pathway. Obstruction of CSF circulation by larger tumors may produce hydrocephalus with increased ICP[12]. In our series 82% of patients with VS developed obstructive hydrocephalus. An elevation of intra cranial pressure (ICP) may cause headache, vomiting, diplopia, papilloedema and in late cases irreversible loss of vision (Secondary optic atrophy). In some study it was reported that these advance features were present in 66-76% cases[13]. In our study more than three-fourth (82%) patients had headache, 56% patients had vomiting, 12% patients had diplopia, 62% had papilloedema and 20% patients had blindness (Secondary optic atrophy). Either papilloedema or secondary optic atrophy were in 82% patients. Finally our study has shown strong association between size of tumor and frequency of occurrence of Hydrocephalus. Those who had tumors >3cm, more than two-third of them had hydrocephalus (Table 3) while those who had tumors 3 cm or below only 1 (2.4%) of them demonstrated hydrocephalus suggesting that greater the size of the tumor the higher is the occurrence of Hydrocephalus.

Conclusion:

In this study, strong association between the sizes of vestibular schwannoma and the frequency of occurrence of hydrocephalus are found. Vestibular schwannomas with larger than 3 cm are strongly associated with obstruction of CSF circulation but the possibility of hydrocephalus is rare when the tumor size is 3 cm or less

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