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Original Article

Outcome of Surgery of Vestibular Schwannomas in a Tertiary Care Hospital in Bangladesh

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Abstract

Background: *Presenting symptoms, treatment considerations, and outcome are strongly related to the extension of vestibular schwannomas (VS). The aim of the current retrospective study was to analyze the clinical features, microsurgical treatment, and outcome of VS with brainstem compression.*

Methods: *Seventy patients presented with VS who had undergone operative procedures performed in our unit from 2017 to 2019. The facial nerve function and hearing assessment was done according to House-Brackmann [HB] grading and pure tone audiometry (PTA) respectively. All patients were operated by retro-mastoid sub-occipital approach.*

Results: *Most patients had large tumors and had no useful hearing (85%), had disabling cerebellar ataxia (92.86%) and presented with features of raised intracranial pressure (48.57%). Large sized tumors were in 32.86% and giant sized tumors were in 57.14% cases. Complete tumor excision was carried out 92.86% and anatomical preservation of facial nerve was achieved in 73.85% cases. Hearing preservation was achieved in 4 patients. Cerebrospinal fluid leak with or without meningitis and transient lower cranial nerve paresis were common complications. The mortality rate was 7.14%.*

Conclusions: *Complete tumor excision with good facial nerve preservation can be achieved in large vestibular schwannomas. Hearing preservation is difficult in larger tumors. Primary microsurgical resection is an appropriate management option for large VS. In our experience, this goal can be achieved safely and successfully by using the retrosigmoid approach.*

Key Words*: Vestibular schwannomas , cerebeo-pontine angel, retrosigmoid approach, facial nerve preservation.*

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Introduction:

Vestibular schwannoma (VS), previously also called acoustic neuroma, is a benign, slow-growing tumor originating from the Schwann cells of the vestibular branch of the vestibulocochlear nerve[1]. The annual incidence of VS is 1–2:100,000 making it the third most common benign intracranial tumor. In its location, the cerebellopontine angle (CPA), VS is the most common type of tumor[2,3,4]. The typical symptoms of VSs are caused by compression on the adjacent

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cranial nerves and may present as hearing loss, tinnitus, dizziness, facial numbness or weakness. Large VSs may even cause hydrocephalus or brainstem compression[5,6] . Sporadic VSs are almost exclusively unilateral, whereas bilateral VSs are typically associated with neurofibromatosis type 2 (NF2)[7]. Since VSs are benign of nature, their treatment options are dependent on the symptoms caused by the tumor. Small VSs with mild symptoms may be followed-up by repeated MR-imaging, whereas larger tumors with pronounced symptoms or tumors with a rapid growth rate may warrant aggressive treatment. Treatment indications and modalities for VSs vary between different centers. The aim of VS surgery is maximum safe resection without causing additional neurological defects in the function of adjacent cranial nerves. In this perspective, the preservation of facial nerve function is crucial due to its location in the immediate proximity of the tumor. However, surgical treatment of VSs results in permanent facial weakness in 10–40% of patients [8,9,10].

The objective of this article is to evaluate the signs and symptoms exhibited in 70 cases of vestibular schwannoma treated surgically between 2017 and 2019, describing the relevant aspects of clinical, audiometric, and imaging diagnosis; the operative techniques used in this series; and perioperative and postoperative complications, thereby proposing a standardized methodology for diagnosis and treatment.

Materials and methods:

We performed a retrospective study of the patients with VS who had undergone operative procedures performed in our unit from 2017 to 2019. Among seventy three patients with VS, three elderly patients were excluded from the study and only a venticulo-peritoneal (VP) shunt was performed.

Clinical evaluation:

All patients underwent complete perioperative neurological evaluation. Functional outcomes at final follow-up were assessed by a phone survey using the Karnofsky performance scale. Facial nerve function was assessed before and after surgery and at each follow-up using the House–Brackmann scale and categorized as good (HB I–II), fair (HB III–IV), and poor (HB V and VI). Pure tone audiograms were performed prior to surgery to assess the option of hearing preservation, as well as postoperatively in patients with serviceable hearing. Criteria for useful hearing was taken as hearing loss <60 decibel (Norstadat Classification for audiometric hearing[11]. The speech discrimination test was not done.

Dysphagia and vocal cord function were evaluated to assess lower cranial nerve involvement. Evaluation of trigeminal nerve included motor and sensory function. Motor examination, including strength of all extremities, coordination, and gait, were examined routinely to assess brainstem and cerebellar affection. In addition, level of consciousness, orientation, memory, and other signs of elevated intracranial pressure (ICP) were evaluated. In selected cases (visual impairment, diplopia, nystagmus), ophthalmological examination was performed before and after surgery.

Radiologic evaluation:

High-resolution bone window computed tomography (CT) studies, essential for visualizing the anatomy of the posterior semicircular canal and its vicinity to the posterior wall of the internal auditory canal and a high jugular bulb, were obtained before surgery. In addition, MRI was performed among all patients to assess tumor extension and maximal extrameatal diameter. The size of the tumor was determined based on linear planimetric measurements, and only the largest extrameatal diameter was used. The tumor size was measured (in CT and/or MRI scan) in three axis that is diameter parallel to petrous ridge, perpendicular to petrous ridge or the vertical diameter in the coronal slices. Giant tumors were defined as those larger than 40 mm; Large tumors measured up to 25 to 40 mm and small tumors measured up to 10 to 25 mm[12].T-2 weighted MR-imaging studies were used to assess hydrocephalus and tumor consistency (solid or cystic). Follow-up MRI was performed in all patients three months after the surgery to exclude residual tumor, and then every year to exclude recurrence. Residual contrast enhancement along facial nerve or brainstem with a diameter exceeding 5 mm were indicative of subtotal resection (STR), whereas residual contrast-enhanced tissue measuring less than 5 mm was assumed to represent near-total resection (NTR). However, in the latter case, differentiation between residual tumor and scar tissue remains difficult.

Operative procedure:

All the patients were operated via the retro-mastoid sub-occipital approach, with the patient positioned in the park bench position. Cavintron ultrasonic aspirator was used in few cases. The facial nerve stimulator was not utilized. The intrameatal component of tumor was removed and the lateral aspect of the intrameatal facial nerve defined, after drilling the roof internal acoustic meatus (IAM). A piece of muscle was used to seal the drilled IAM in all patients. Mean operation time was 6.25 hours (range: 2.5–10 hours).

Follow up:

All the patients were followed up at 6 weeks, at 3 months, at 6months and yearly thereafter.

Results:

There were a total of 70 patients in this study. The male to female ratio was fairly equal at 7:13. Regarding laterality, 28 (40%) tumors were right-sided and 42 (60%) were left-sided.

There was no useful hearing in approximately 85% patients. Varying degree of facial nerve paresis was observed in 67% patients. The other symptoms were of raised intracranial pressure, trigeminal dysfunction and cerebellar ataxia (Table I).

Regarding the duration of the chief complaint at diagnosis, 8 (11.43%) patients had the symptom for less than six months, 14 (20%) for six months to one year, and 48 (68.57%) for more than one year.

The majority, 32.86% were between the age from 31-45 years. The youngest patient was 17 years and the oldest was 66 years. The mean age was 40 years. Patient ages at diagnosis were distributed as follows:

15–30 years: 22 (31.43%)

31–45 years: 23 (32.86%)

46–60 years: 21 (30%)

61–75 years: 4 (5.71%)

Forty patients (57.14%) had giant, twenty three patients (32.86%) had large and seven patients (10%) had medium size tumors. (Figure I)

Twelve patients (17.14%) underwent ventriculo-peritoneal shunt procedure prior to definitive surgery.

Five patients (7.14%) required EVD (External Ventricular Drainage) surgery in the post-operative period.

Complete excision of the VS along with intra-canicular (intra-metal) portion was achieved in 92.86% (65/70) of patients. Out of the five patients in whom complete excision could not achieved; Two patients had tumor adhere to brainstem and in two patients the tumor was adhere to facial nerve (part of the tumor was left to avoid injury). In one patient, only a subtotal removal was possible due to massive intra-operative hemorrhage.

Facial nerve anatomically preserved in 73.85% (48/65) patients with complete tumor excision. Four patients died in post-operative period, and sixty six patients were analyzed for facial nerve function at time of discharge. The facial nerve preservation rates were 58.82% for giant size VS and increased rates were observed with tumors of smaller sizes (88% for large and 100% for medium size) (Table II). Further the functional status of the facial nerve at follow up were better in patients with relatively smaller tumors who had mild grade facial paresis (H&B Grade 1and 2) pre-operatively (Table III).

Seven patients had useful hearing pre-operatively. Useful hearing could be retained in four patients (5.71%) post-operatively. Amongst these seven patients, one had giant size tumor, four had large tumors and three had medium sized tumor.

Table-I

Pre-operative cranial nerve and neurological deficits in patients with vestibular schwannoma.

Neurological deficits	Number	(%)
Hearing loss (no useful hearing)	60	85.71
Cerebellar signs	65	92.86
Trigeminal dysfunction	45	64.29
Facial nerve paresis	47	67.14
Papilledema	34	48.57
Secondary optic atrophy	05	7.14
Lower cranial nerve paresis	25	35.71

Table-II

Anatomical preservation of facial nerve in patients with complete excision in different tumor sizes

Tumor category	Patients with complete removal	Anatomical preservation of 7th nerve	(%)
Medium	6	6	100
Large	25	22	88
Giant	34	20	58.82
Total	65	48	73.85

Table III

Post-operative complications in patients with complete excision and anatomical preservation of facial nerve

Complications	Number of patients	(%)
CSF leak	4	5.71
Hematoma	3	4.28
Meningitis	3	4.28
Lower cranial nerve paresis	5	7.14

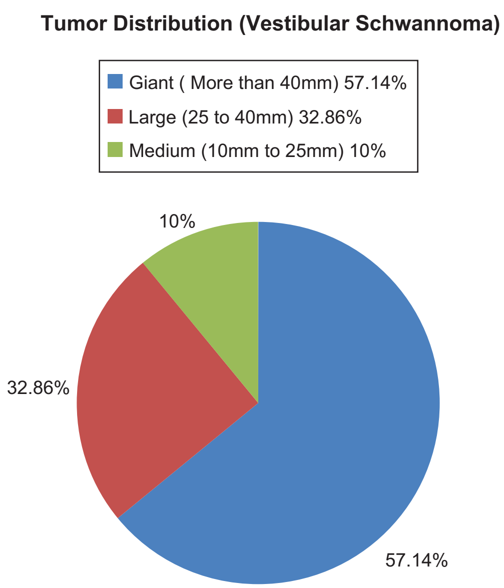

Fig.-1: *Tumour distribution according to size of vestibular schwannoma*

Early postoperative complications (occurring within one month of surgery) included cerebrospinal fluid (CSF) leak occurred in 5.71% (4 patients).CSF otorrhea occurred in two patients. Most cases were managed conservatively with or without lumber puncture and CSF drainage.

Total three patients (4.28%) developed post-operative hematoma. Among two patients developed operative site hematoma and one patient had an extradural hematoma and all of them were managed conservatively.

Total three patients (4.28%) developed post-operative meningitis and responded to anti-meningitic treatment.

Post-operative lower cranial nerve paresis developed in five (7.14%) patients (Table III). These patients required nasogastric tube feeding. However the lower cranial nerve paresis improved functionally during 4-6 weeks follow up.

The mortality rate was in 7.14% patients (5/70). Massive cerebellar edema leading to brainstem failure occurred in one patient. Massive intraoperative haemorrhage, operative site haematoma, post-operative myocardial infraction caused mortality in one patient each. No definite cause of death could be established in one patient who could not be revived following surgery.

Discussion:

The management of large VS is challenging. Patients usually present with multiple cranial nerve deficits and signs of brainstem compression or intracranial hypertension. There are numerous options and factors to be considered in the optimal management of patients with acoustic neuromas. The options include microsurgical management; stereotactic radiosurgery and conservative 'wait and scan'. The decision is based on a number of factors which include the age of patient, size of tumor, preservation of hearing and the presence of co-morbid factors. With the availability of operating microscope, safe modern anesthesia and refinement in the microsurgical technique the goal of VS surgery has shifted from complete excision to excellent facial nerve function and preservation cochlear nerve function.

In this present study, 90% of patients had either a large or giant sized VS. Eighty five percent of patients had no useful hearing at time of presentation. In contrast to certain western literature[13,14], majority of our patients sought medical attention at a stage when they developed disabling cerebellar ataxia (92.86%) and /or the symptoms of raised intra-cranial pressure.

Pre-operative ventriculo-peritoneal shunt is not required in majority of patients, but some patients report late with signs of raised intra-cranial pressure requiring emergency shunt procedures. In the present study the pre-operative shunt was done in five patients at our center and seven patients had shunt done elsewhere before they were referred for definitive surgery. Gerganov et al recommended placement of external ventricular drainage (EVD) or VP shunt prior to surgery as surgery in patients with hydrocephalus and increased ICP is presumably more challenging, and related to worse outcome or higher complication rates. They found that the general and functional outcome in patients with primary VS removal is independent of the presence of hydrocephalus[14,15,16].

Complete tumor excision was achieved in 92.86% (65/70) of the patients. Yamakani et al [17] reported complete tumor excision in 86% of patients by retrosigmoid approach for large acoustic tumors. Lanman et al reported at higher rate (96.3%) of total removal by trans-labrynthine approach[14]. Ebersold et al[18] achieved total tumor resection in 97.2% (249/256) by retro-mastoid approach for tumors of all sizes. Samii et al[19] have reported complete excision in 97.9% patients by sub-occipital trans-meatal approach. The translabyrinthine and retrosigmoid approaches allow removal of VS of almost any size[20]. Each approach has advantages and disadvantages. The benefits of the translabyrinthine approach are a short distance to the tumor and avoidance of cerebellar retraction with early identification of the facial nerve. The disadvantages of the translabyrinthine approach include inevitable hearing loss and, in cases of large VS, restricted access to the trigeminal nerve, caudal cranial nerves, and anterior aspect of the CPA[21,22]. The retrosigmoid approach is the most popular approach among neurosurgeons[23,24,25,26]. It is fast, straightforward, and offers excellent visualization of the CPA, trigeminal nerve, lower cranial nerves, and majority of the posterior fossa arteries including the upper part of the vertebral artery and superior cerebellar artery [26].

Anatomical preservation of facial nerve was achieved by 73.85% (48/65) patients. In the giant category facial nerve preservation was 58.82% (20/34) and in the large category was 81.3% (22/25) and 100% (7/7) for medium size tumors. In some western literatures the anatomical preservation rate is 80-90% with the removal of large tumors either by trans-lybrinthine approach [27,28]. Samii and Matthias reported preservation rate of 87% with tumor size Ã3 cm until 1988, but in most recent 200 cases preservation rate rise to 94% independent of tumor size [27]. This data confirms that there is a learning curve for surgery of VS. Microsurgical skills and experience of the surgeon influence postoperative facial nerve function[28,29]. According to Whittaker et al a surgeon operating less than twelve cases per year cannot expect to get equal results of large series [21]. Another major factor influencing facial nerve function is the tumor size. The risk of facial nerve palsy may increase by up to six-fold in large VS [29]. Facial nerve, having a reciprocal relationship with tumor size i.e. larger size of tumor lesser the chances of preservations facial nerve [14,15,18] was also observed in this present study.

In the present study 10% (7/70) patients had pre-operatively useful hearing. Post-operative hearing could be preserved in four of these seven patients (5.71%) in spite of their giant size in one, large in four and medium in three patients. Though the retro-mastoid approach gives the surgeon great opportunity for saving hearing in small sized tumors, but in tumors move than 4 cm, the post-operative hearing is usually very poor as observed by Ebersold et al,[18] who reported no post-operative hearing in any of patients with tumor size more than 4 cm . According to Samii et al, patients with large tumor (30 mm x 20 mm) hearing was preserved in 23.6% (78/330) [11]. Almost all authors agree that hearing preservation is more likely with smaller tumors with good pre-operative hearing.[29,30.]

The reported incidence of cerebrospinal fluid leak ranges between 0% and 30%, with the average approximately 12%, although making comparisons between published series is difficult because of the various methods and reporting criteria used by different authors[31.] In the present study 5.71% (4/70) of patients of cerebrospinal fluid leak .Most cases were managed conservatively with or without lumber puncture and CSF drainage and the incidence is at par with the series published by Yamakani et al,[17]. However the incidence of associated meningitis in our study is 4.28% also similar as compared to other series (between 3.7 to 9.2%).[14,32-34]

Although loss of cochlear and seven nerve function are two of the major cranial nerve injuries that can occur during the surgery, there are risks of injury of lower cranial nerves in large and giant sized tumors, which can complicate the postoperative course. Judicious use of nasogastric tube feeding and planned tracheostomy can avoid major respiratory complications post-operatively. The incidence of lower cranial nerve paresis has been reported to range from 1.5% to 5.5%[14,18,35,19] against 7.14% in the present study. Cerebellar retraction is the event with the greatest influence on the surgical risk, and should be avoided. Appropriate preoperative patient positioning and intraoperative general anesthesia should enable spontaneous retraction[.35]

The mortality rate was in 7.14% patients (5/70). Massive cerebellar edema leading to brainstem failure occurred in one patient. Massive intraoperative haemorrhage, operative site haematoma, post-operative myocardial infraction caused mortality in one patient each. No definite cause of death could be established in one patient who could not be revived following surgery. The mortality rate was similar to study of Jain et al 6% patients (15/250)[12]. As compared to other study the mortality rate in our series was high. Some authors have stated that 20 to 60 operations are necessary to achieve results similar to those of highly experienced surgeons[35-38]. Vestibular schwannoma surgery requires continuing refinement and improvement.

In the present study, all the cases were operated by retro-sigmoid approach with park bench position. This concludes the fact that the retro-sigmoid approach in experienced hands is a good options; with good results compared to other series irrespective of tumor size. This is an extension to the view put forward by Semii et al that from any of the available approaches, such as sub-occipital, the middle fossa, and the trans-labyrinthine; surgeons can develop expertise to high standards, by training and experience, with respect to the optimum patient's safety, morbidity and mortality[19].

Gormley and Sekhar et al[15] used the combined transpetrosal and retrosigmoid approach for tumors greater or more than 4 cm in the cerebello-pontine angel, especially when they extend up to the tentorial notch, because the combination allows good visualization of tumor-brainstem interface and the tentorial notch and better facial nerve outcome for these group of tumors. But in our series, using retro-sigmoid approach alone visualization of tumor-brainstem interface and facial nerve preservation could be possible in giant sized tumors without much difficulty. Although the choice is influenced by surgeons' preferences, the retrosigmoid approach is recommended in surgery for acoustic neuroma whenever hearing preservation surgery is an option, or for tumors of any size irrespective of hearing function[36].

Conclusions:

Vestibular schwannomas tend to be diagnosed late in our local setting, with large tumors and compressive symptoms. The goal in treating VSs should be total removal in one stage and preservation of neurological functions, because these factors determine the quality of life for patients. In our experience, this goal can be achieved safely and successfully by using the retrosigmoid approach.

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