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A Retrospective Study of Clinical Profile and Surgical Outcomes of Vestibular Schwannomas

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Abstract:
Introduction: *Vestibular schwannomas manifest with very serious deficits of hearing, facial expression, lower cranial nerve, cerebellar and brain stem functions either preoperatively or as a consequence of surgical intervention. Our objective was to analyze the symptomatology and clinical signs and try to correlate with postoperative outcome and complications in the setting of limited infrastructure, therapeutic and monitoring aids and multi-surgeon involvement with varied spectrum of surgical expertise in a single large tertiary teaching institute.*
Materials and Methods: *A retrospective analysis was done using available discharge summary database. Clinical assessment was done using House Brackmann grading for facial nerve function,puretone audiometry for hearing and MRI Brain imaging [Figures 1]. Size of the tumour was taken as the maximum transverse diameter demonstrated on contrast enhanced MRI brain. All patients underwent retromastoid retrosigmoid suboccipital craniectomy. Patients were placed in supine position with head turned to contralateral side and fixed with Mayo's head frame [Figure 3]. Retromastoid C shaped incision was taken to reflect suboccipital muscles and overlying fascia and subcutaneous tissue as a single layer. Suboccipital craniectomy was done to expose transverse sinus, sigmoid sinus and their junction [figure 4]. Foramen magnum rim was opened electively. Mastoid air cells were packed with muscle/abdominal fat and bone wax. Initial small dural opening was done in the inferior part to expose and drain CSF from cerebello medullary cistern.*
Results: *A total of 52 patients who were entered in the discharge summary database between January 2012 to April 2019 were included in the study. Headache was the most common presenting symptom [65.4 %] [Table 1]. Hydrocephalus was present in 45.4 % of patients out of which 34.5 % required perioperative CSF diversion procedure. Papilloedema was present in 38.2 % of patients. 21 % patients presented with preoperative lower cranial nerve palsy. The mean tumor size was 4.4 cm. Giant tumours [>4 cm] constituted 74 % of patients [Figure 3]. Only 6 % [n=3] tumours were < 3 cm in size. Cystic Vestibular schwannomas [> 50 % tumour having cystic component] constituted 27 % [n=14]. 4 % [n=2] of patients had bilateral vestibular schwannomas. All patients underwent Retrosigmoid retromastoid suboccipital craniectomy.*
Conclusion: *Hearing loss is the main presenting symptom of vestibular schwannoma. The indication and the timing of tumor resection is dependent on the tumor extension and related necessity of brain stem decompression and on the auditory function. The chances of good outcomes [Facial nerve preservation and post op morbidity] are best when surgery is performed early [smaller size tumours] and when there is good preoperative facial and lower cranial nerve function. As an optimal goal, completeness of resection with functional Facial nerve preservation is formulated and as an acceptable compromise, near total microsurgical resection with functional Facial nerve preservation is suggested.*
Key Words: *Vestibular schwannomas , Suboccipital craniectomy*

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I. Introduction

The internal auditory canal (IAC) is 10–17 mm in length and contains the facial, vestibular and cochlear nerves, surrounding blood vessels and a common dural sheath.[1]An acoustic neuroma is a benign tumor involving cells of the myelin sheath that surrounds the vestibulocochlear nerve (eighth cranial nerve). Acoustic neuromas are often called vestibular schwannomas because they are tumors that arise from the myelin sheath that surrounds the vestibular nerve.[1] Acoustic neuromas are considered benign (non-cancerous) tumors since they do not spread to other parts of the body. They can occur anywhere along the vestibular nerve but are most likely to occur where the vestibulocochlear nerve passes through the tiny bony canal that connects the brain and the inner ear. A unilateral tumor is a tumor arising from one nerve and a bilateral tumor arises from both vestibular nerves. Unilateral acoustic neuromas usually occur spontaneously (by chance). Bilateral acoustic neuromas occur as part of a hereditary condition called Neurofibromatosis Type 2 (NF2).[2]

They are typically benign and slow growing, but can cause symptoms through mass effect and pressure on local structures, eventually becoming life-threatening. Considering the possibility can enable earlier diagnosis, increase management options and may decrease morbidity. Acoustic Neuroma represents 6-10%of primary intracranial tumours but is the most common form of cerebellopontine angle (CPA) tumour. Sporadic tumours are unilateral and account for 95% of cases, whilst those associated with neurofibromatosis are bilateral and account for the other 5%.[3]

II. Materials And Methods

A retrospective analysis was done using available Discharge summary database. Clinical assessment was done using House Brackmann grading for facial nerve function,puretone audiometry for hearing and MRI Brain imaging [Figures 1]. Size of the tumour was taken as the maximum transverse diameter demonstrated on contrast enhanced MRI brain.All patients underwent retromastoid retrosigmoid suboccipital craniectomy. Patients were placed in supine position with head turned to contralateral side and fixed with Mayo's head frame [Figure 2]. Retromastoid C shaped incision was taken to reflect suboccipital muscles and overlying fascia and subcutaneous tissue as a single layer. Suboccipital craniectomy was done to expose transverse sinus,sigmoid sinus and their junction. Foramen magnum rim was opened electively. Mastoid air cells were packed with muscle/abdominal fat and bone wax. Initial small dural opening was done in the inferior part to expose and drain CSF from cerebello medullary cistern.[4]

The dura was opened based on Transverse and Sigmoid sinuses. Self-retaining retractors were intermittently applied to optimally retract the cerebellar hemispheres. After adequate CSF drainage from the CP angle and adjoining cisterns, intratumoural decompression was done strictly staying within the arachnoid planes, using suction apparatus, sharp scissors, ring curette and tumour knife. After achieving significant tumour decompression, tumour capsule became lax and collapsible which was separated from the surrounding neurovascular structures under magnification of operating microscope. Intrameatal component of tumour was delivered after drilling the internal auditory meatus using high speed diamond burr. Finally, the tumour adjacent to the brain stem was delivered after ensuring minimal handling or traction. Patient was closely monitored for fluctuations in Heart rate and B.P;Ultrasonic aspirator, intraoperative facial and lower cranial nerve monitoring was not available. Hemostasis was confirmed by achieving normotensionand valsalva manouevre. Water tight dural closure was achieved by using pericranial graft or temporalis fascia graft for closure. The craniectomy defect was sealed by replacing bone dust and bone chips. Postoperative CT scan was done either immediate postop or electively after 48 hours depending on the patient's neurological status.[5]

III. Results

A total of 52 patients who were entered in the discharge summary database between January 2012 to April 2019 were included in the study. Headache was the most common presenting symptom [65.4 %] [Table 1]. Hydrocephalus was present in 45.4 % of patients out of which 34.5 % required perioperative CSF diversion procedure. Papilloedema was present in 38.2 % of patients. 21 % patients presented with preoperative lower cranial nerve palsy. The mean tumor size was 4.4 cm. Giant tumours [>4 cm] constituted 74 % of patients [Figure 5]. Only 6 % [n=3] tumours were < 3 cm in size. Cystic Vestibular schwannomas [> 50 % tumour having cystic component] constituted 27 % [n=14]. 4 % [n=2] of patients had bilateral vestibular schwannomas. All patients [except 1 case of underwent Retrosigmoid retromastoid suboccipital craniectomy.

Neurosurgeon's familiarity as well as the advantage of being able to treat virtually any size tumour by this approach was the reasons for choosing this approach. In 80 % of cases, gross total tumor resection was achieved. In 20 % of cases, deliberate subtotal resections were performed [Figure 6]. These were due to adhesions to brain stem in 4 % cases and for facial nerve preservation in 5% cases. Small residues were left attached to lower cranial nerves in 8%[n=4] and trigeminal nerve in 3% [n=2] cases. 6.3% [n=3] were recurrent vestibular schwannomas with a mean recurrence duration of 42.1 months.

All patients had significant preoperative hearing loss [Class C & D of American Academy of Otolaryngology-Head and Neck Surgery Foundation hearing classification system].[2]57.3% [n=30] had significant [House Brackmann grade 3-6] preoperative facial nerve palsy out of which 61 patients had > 4 cm tumour [Table 2]. Anatomic facial nerve preservation was achieved in 84 % in tumours < 3 cm size. The overall anatomic facial nerve preservation rate was 33.6 %. The rate of functional facial nerve preservation [H-B Grade 1-2] was 67.2 % in cases of small tumors (< 30 mm). The overall functional facial nerve preservation [H-B Grade 1-2] was 28.2 %. Only 52.7 % [n=27] were available for follow-up [at 6-12 months]. The Facial nerve functional status at follow-up was almost comparable to the immediate postoperative status. However the veracity of this finding may not be tenable due to the significant follow-up drop-out rate.

Postoperatively 71.9% patients had Facial Nerve palsy [House Brackmann Grade 3-6] out of which 57.3% preoperative facial nerve palsy and 14.6% had new onset facial nerve palsy. Postoperative lower cranial nerve palsy was present in 33.6% out of which 23.6% had preoperative palsy and 10% had new onset palsy. Postoperative CSF leak was present in 11.8% patients. Overall mortality was 5.4% [2 cases] out of which 4.5% [2 cases] were due to operative site haematoma.

Fig.1 Preoperative MRI imaging sequences showing tumour characteristics of Vestibular schwannoma

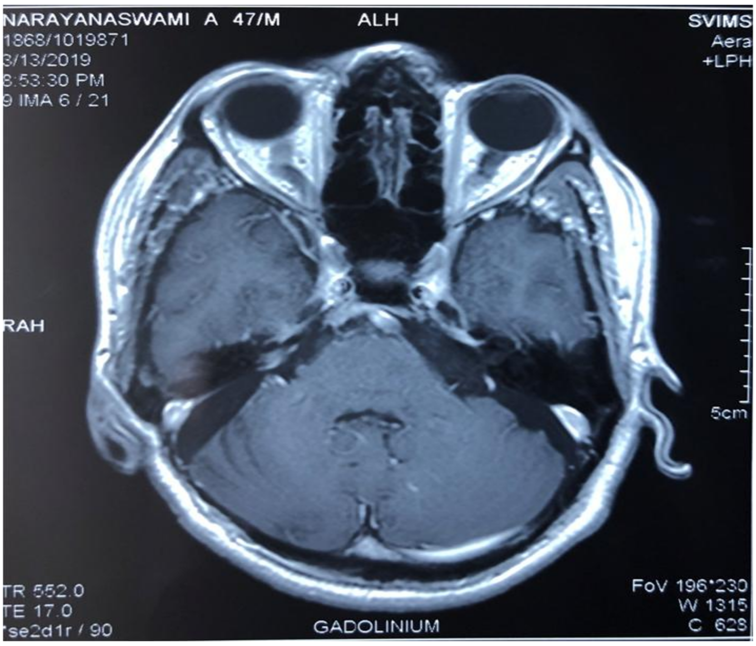

Fig.2 Postoperative MRI imaging sequences showing total removal of tumour

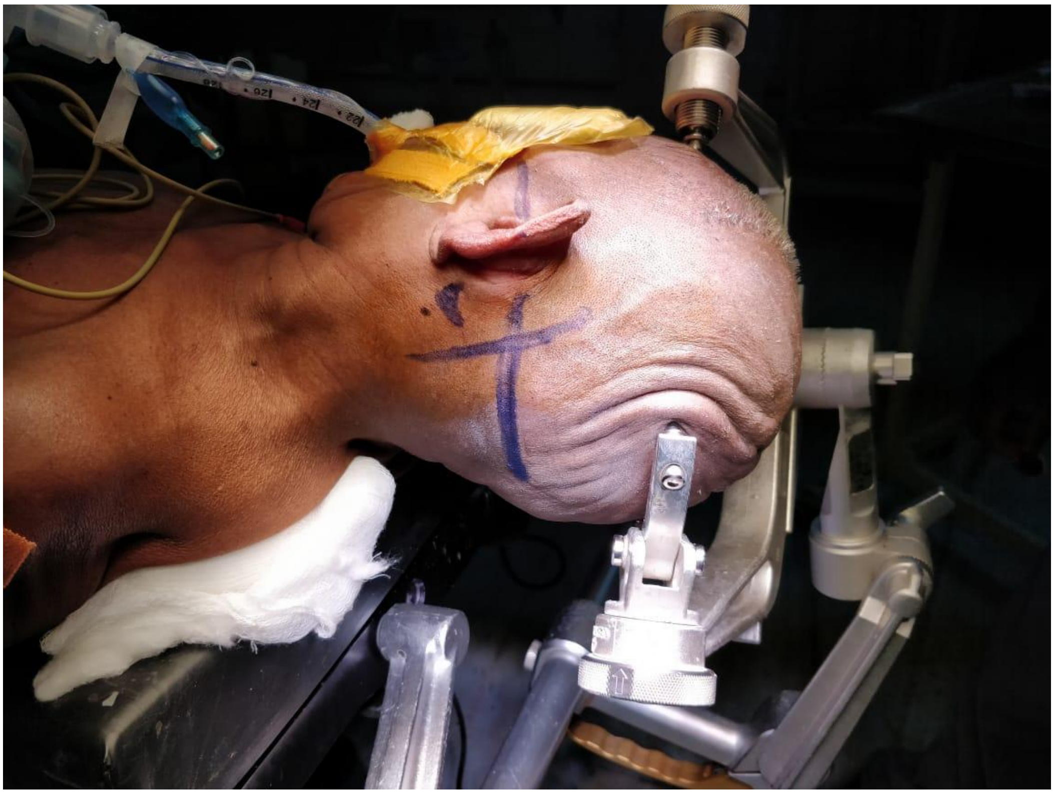

Fig. 3 showing positioning and incision in a patient undergoing Retromastoid suboccipital craniectomy for Vestibular schwannoma

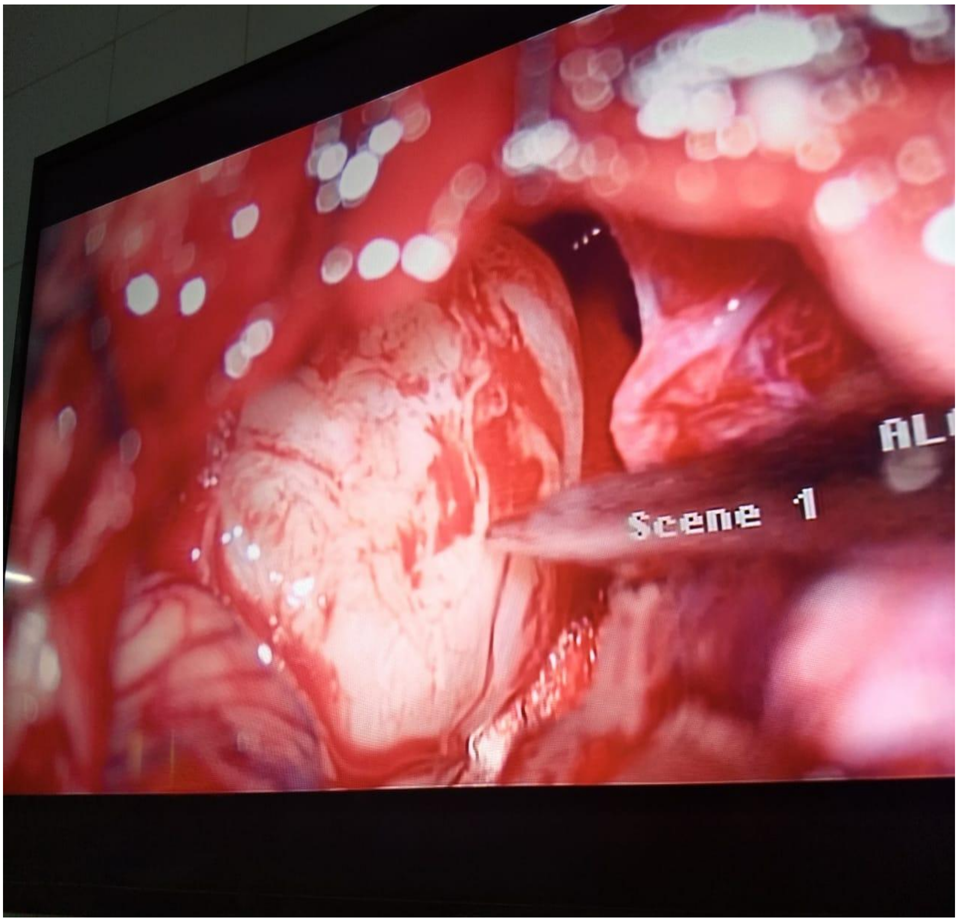

Fig. 4 intraoperativepicture showing Vestibular schwannoma

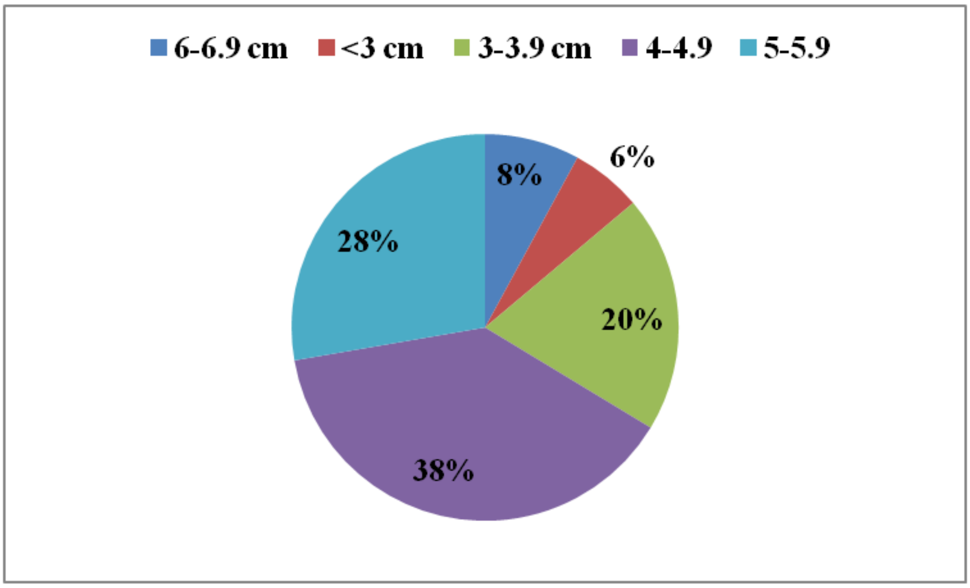

Fig. 5 showing distribution of vestibular schwannomas as per tumour size

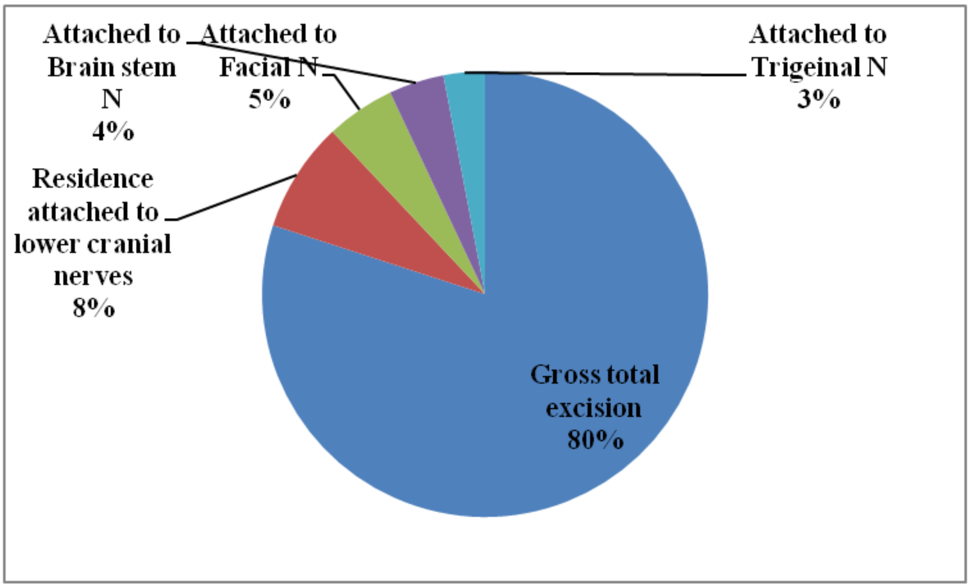

Fig.6 showing extent of tumour resection in vestibular schwannoma

S.No	Clinical Parameter	No of patients	Percentage
1	Hearing Impairment	52	100
2	Headache	34	65.4
3	Facial Nerve weakness	28	53.6
4	Hydrocephalus	23	45.4
5	Papilloedema	20	38.2
6	Lower cranial Nerve Palsy	11	21

Table 1: Showing Clinical profile of presentation in Vestibular schwannoma

Tumour Size	<3 cm	3-3.9 cm	4-4.9 cm	5-5.9 cm	6-6.9 cm	Total
No	6	22	43	31	8	110
Preop Facial N palsy Grade 3-6	0	2	25	29	7	30(57.3%)
Preop Intact Facial N function	6	20	18	2	1	23(43.7%)
Anatomical Facial N preservation	5(84%)	18(90%)	14(77.8%)	0	0	17(33.6%)
Immediate Postop Facial N palsy Gr 1-2	4(67.2%)	16	11	0	0	15(28.2%)

Immediate Postop Facial N palsy Gr 3-4	1	2	3	1	0	3(6.4%)
Immediate Postop Facial N palsy Gr 5-6	1	4	29	30	8	34(65.5%)
Followup Facial N palsy Gr 1-2	3	8	7	0	0	16(31%)
Followup Facial N palsy Gr 3-4	0	1	1	0	0	2(3.4%)
Followup Facial N palsy Gr 5-6	1	2	15	16	4	34(65.5%)

Table 2: Showing corelation of tumour size and postop facial nerve function in vestibular schwannoma

IV. Discussion

The trend was comparable to a similar study from Jain et al.[7]which is from a comparable setup from this part of the world. Jain et al., had 90 % of the patients presenting with significant hearing loss preoperatively. Preoperative facial nerve palsy [H-B Grade 3-6] was seen in 57.3 % of our patients.[6] This might be attributed to the relatively larger size of tumours in our series[74 % cases being > 4 cm in diameter]. Although the overall anatomical facial nerve preservation rate was only 33.6% in our series, in smaller sized tumours [<3 cm: 84%, 3-3.9 cm: 90%] it was comparable to series by Jain et al.,[81.3 %].[7] This might be attributed to many factors like multiple surgeons with various levels of learning curves, larger size of tumours and unavailability of intraoperative cranial nerve monitoring and ultrasonic aspirator.[8]

In our series, gross total excision could be achieved only in 80% of cases as safe removal of residue was not found to be possible due to adherence to vital neurovascular structures in the remaining 20% of cases. Lower cranial nerve palsy was seen in 33.6% of patients postoperatively out of which 23.6% of patients had significant lower cranial nerve palsy preoperatively.[9] Effectively there was an increase by 10% of lower cranial nerve involvement postoperatively which was comparable to other series [Table 3]. In our series, the mortality was 5.4%, which was comparable to that of Jain et al. Misra et al., had 39% of cases with>4 cm tumour and 96% anatomical Facial Nerve preservation.[10]

V. Conclusion

Hearing loss is the main presenting symptom of vestibular schwannoma. The indication and the timing of tumor resection is dependent on the tumor extension and related necessity of brain stem decompression and on the auditory function. The chances of good outcomes [Facial nerve preservation and post op morbidity] are best when surgery is performed early [smaller size tumours] and when there is good preoperative facial and lower cranial nerve function. As an optimal goal, completeness of resection with functional Facial nerve preservation is formulated and as an acceptable compromise, near total microsurgical resection with functional Facial nerve preservation is suggested.

In the background of a multisurgeon, resource constrained setup an acceptable level of complications and a reasonable outcome can still be achieved. The necessity of intraoperative cranial nerve monitoring, ultrasonic aspirator and the role of surgeon's learning curve cannot be undermined**.**

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