22

CLINICAL PROFILE AND OPERATIVE OUTCOME FOR GIANT VESTIBULAR SCHWANNOMAS

SREE CHIRA TIRUNAL INSTITUTE FOR MEDICAL

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PROJECT REPORT

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Programme : **M.Ch. NEUROSURGERY.**

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CERTIFICATE

I, Dr. J.Rajesh kumar hereby declare that I have actually performed all the procedures listed / carried out in the project under report.

Place : Thiruvananthapuram Signature

Date: 2 November 20003 (J. RAJESH KUMAR)

Forwarded. He has carried out the minimum requirement of

procedures/etc.

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CONTENTS PAGE NUMBERS

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INTRODUCTION:

Giant vestibular schwannomas are tumors with an average diameter of more than 4.5 cm in any dimension (axial, coronal or sagittal). A retrospective analysis of 100 cases of vestibular schwannomas operated in our institute over a period of 4 years was carried out and among which 25 were giant tumors. All patients were evaluated with CT scan, MRI scan or both after proper preoperative clinical assessment to document the deficits. Hearing assessment was done with emphasis on decibel loss, speech discrimination score and speech reception threshold in all of them and those with profound hearing loss (> 50 dB) were identified in the preoperative period. Size of the tumor was measured by taking the largest dimension of the lesion in either sagittal, coronal or axial sections. Tumors with size more than 4.5 cm were included in the giant group, between 2.5 to 4.5 cm were considered as large and those less than 2.5 cm were considered as small tumors. The clinical and radiological features, intra-operative problems, postoperative course, surgical outcome and period of hospital stay were analyzed with respect to the tumor size. All patients underwent surgery by retrosigmoid approach and total excision was done in most of the cases. Statistically significant positive correlation between tumor size and degree of neurological deficits was not

observed as well as there was no significant difference in the outcome based on the size between the two groups of tumors.

AIMS OF STUDY

To evaluate, analyze and compare the clinical spectrum, operative techniques and outcome for Giant Vestibular schwannomas to tumors of smaller size.

REVIEW OF LITERATURE

Treatment of vestibular schwannomas has improved dramatically in the second half of the 20 century, especially since the introduction of microsurgical procedure in 1960s, the revolutionary inventions in neuroradiology in the 1970s and 1980s and further refinements in surgery using technological assistance in the 1990s.Treating benign lesions in such a delicate area as the cerebellopontine angle challenges neurosurgeons. Moreover, in some patients, one stage surgery leads not only to permanent cure but also to completely normal quality of life and life expectancy. Many phenomena encountered in this disease are not yet understood, for example, in general, a positive correlation between the size of the tumor and the degree of hearing loss has been found; however, patients with marked hearing loss or even deafness can have tiny intra-meatal tumors and large tumors can lead moderate hearing loss. It is not well understood how various symptoms, duration, and sequence of occurrence correlate with the size and extension of the tumor or with actual objective cranial nerve damage. An analysis of these epidemiological aspects is necessary before formulating an optimal diagnostic and therapeutic protocol.

Madjid Samii's clasification of Vestibular Schwannomas

Tumor sizes were measured in the axial planes, considering intra and extrameatal tumor extension.

Tumor larger than 30 x 20 mm were defined as large

Tumor less than 30 x 20 mm were defined as small

Tumor extension classes were described as follows-

T1: Purely intrameatal.

T2: Intra-extra meatal.

T3a: Filling the cerebellopontine cistern.

T3b: Reaching the brain stem.

T4a: Compressing the brain stem.

T4b: Severely displacing the brain stem and compressing the fourth Ventricle.

With advancing tumor growth and extension, additional cranial nerves, other than vestibulocochlear nerves, increasingly become more seriously involved; the patient's age is another factor leading to deteriorating clinical function. However if deafness occurs and no additional symptoms develop, the deafness will persist without any tumor detection for long time, whereas the symptom duration will be shorter than the average for the tumor class, if additional symptoms develop and stimulate concern by the patient or doctor.

Moreover, because auditory function is not strictly correlated with tumor size, the cause of auditory nerve impairment must be a multifactorial matter.

Comparison of symptoms and signs of various cranial nerves

Regarding all cranial nerves, a higher incidence of involvement and usually, a higher degree of nerve impairment is present than is noticed by the patient. This corresponds to the observations made by some investigators, who found normal hearing in 4% of patients versus 15% presumed normal hearing by the patients. Partial deficits and multifocal involvement of several cranial nerves are factors that raise suspicion of a lesion worth investigating.

Age distribution and tumor extensions

Vestibular schwannoma generally occurs more in the fifth decade. Mean age group for this tumor in Samii's series was 46.9years. There is an inverse correlation between age and degree of tumor extension. Very large tumors diagnosed at an early age might indicate an especially fast rate of growth. Increased growth rate might lead to an earlier diagnosis than in the average case.

Trigeminal nerve

In the case of complete deafness, additional onset of trigeminal nerve disturbances lead to a shortening of the overall symptom duration. In case of trigeminal neuralgia, however the symptom duration is longer as it is usually considered as an idiopathic entity.

Facial nerve

Signs of vascular compression, such as facial spasm, are tolerated for longer periods than was facial nerve deficit, an observation close to that in with trigeminal nerve. Taste disturbances were reported very rarely and were usually not considered to be serious. The incidence in Sammii's series is 1.8%, found to be much lower than what was reported early. A systematic search for taste disturbances should be included in history taking, when doubt or suspicion is present, electrogustometry can detect can detect the disorder early. Detection of taste disturbance is useful because of the increased risk of facial paresis.

Cochlear Nerve

Although disturbed cochlear nerve function was the most frequently noticed symptom, the limited awareness by the patient of the frequency and extent of

this lesion may be extraordinary. *Samii's* series- Fifteen percent of patients did not remember the start of the deficit and 10% had not noticed any hearing deficit or the ensuing deafness. Only 62% of those presenting with definite deafness were aware of it. Incidence of complete hearing loss in Samii's series was 23 %.*Strekers* et al found a rate of 22% deafness in a series 92 patients. Severe hearing disturbances were reported by *Mathew et al*, who found sudden onset of hypoacusis in 4% and vertigo in 5% of their patients. Samii's series 16% of patients' experienced sudden hearing disturbance in 3% of all patients in this series, sudden hearing disturbances resulted in permanent deafness. Patients with preoperative deafness had become deaf gradually (23%) or suddenly (3%) .Of those became deaf before surgery 12% experienced sudden deafness. Therefore in patients who are known to have neuromas, even when good hearing has been stable, sudden hearing loss can occur. Young patients at presentation usually associated with a large tumor growing at an especially fast rate.

Sequence of symptoms

Most longstanding symptoms affect the cochlear nerve, with a mean duration of 3.6 to 5.1 years for hypacusis and deafness. Caudal cranial nerves disturbances as long as 2.7 years until diagnosis, but because of low incidence, they are not diagnosed earlier. Vestibular disturbances are the

next most commonly occurring they occur far more frequently than is generally reported. Close follow up of patients presenting with the triad of hearing disturbance, unilateral tinnitus ,and vertigo should be the rule, and vestibular tests should be performed subsequently. Trigeminal nerve disturbances are the third most commonly occurring symptom (16.5%) when considering incidence and sequence of onset. They usually occur after cochlear symptoms have persisted for longer than 2 years and vestibular symptoms for longer than 1 year. In 1980, Tarlov claimed that " clinical suspicion is essential for early diagnosis of *Acoustic neurinomas.*No absolutely characteristic pattern of hearing loss occurs".

Radiological features

Regarding the anatomic structures in general, one would expect larger dimensions in male than in female patients. The basic anatomic parameters evaluated by Samii's series also noticed significant larger measurements in males when compared to female patients except one, which was the size of the internal auditory meatus. The length of the posterior IAC wall on healthy side, contralateral to the tumor was slightly larger in female patients. The mean patient age at presentation with acoustic neurinoma is significantly higher in female patients because of relatively more space for seventh and eighth nerve bundle for the growing tumor inside the auditory meatus.

Pneumatization is especially important in evaluating the region of the petrous apex near the IAC because of the danger of postsurgical CSF fistula; petrous apical cells larger than 1.5 cm in diameter were encountered in up to 0.9 % of cases (Samii's series).

Tumor induced bony changes

Whereas the destruction and widening of IAC by acoustic neurinomas is well-accepted phenomenon, a reliability of this experience for the diagnosis of a suspected tumor is not established. There are reports of only 50% interear differences of IAC despite proven tumors. Definite differences of the IAC could be identified between the tumor free side and side with the tumor. The length of the posterior wall of auditory meatus and the interear difference of maximal IAC diameters showed statistically reliable relation with preoperative hearing function. The more the posterior auditory meatus was destroyed by the tumor, the shorter the posterior wall was or the larger the interear difference of maximal diameters. Severe bony destruction is correlated with more severe acoustic nerve involvement before surgery and with resulting reduced chance of hearing preservation. It is hypothesized that severe bony destruction might be a sign of serious tumor aggressiveness.

Tumor extension and acoustic function

The extension of the tumors correlated with preoperative acoustic nerve function. With regard to the chances of hearing preservation, two growth directions of tumor extension were especially critical, caudal and anterior to the auditory meatus. These tumor extensions showed significant limits with regard to deafness in large tumors, but did not play an important role in small tumors of a mean diameter of 25mm. In such small tumors, the tendency for caudal or anterior growth is small and other factors, such as adherence with the tumor, are more important in causing the deafness. In large tumors anterior and caudal extensions are very critical. These factors emphasize that large tumor size does not necessarily exclude any chance of hearing preservation but that tumor extension plays a major role. At surgery, the acoustic nerve is usually found inferior to the tumor and main tumor extends upwards. When the tumor grows upward or backward from IAC it does not affect the cochlear nerve so severely. If tumor grows in an especially caudal or anterior direction it may surround and infiltrate or stretch and elongate the nerve, the cochlear nerve is extremely vulnerable to stretching, as is known from intraoperative brain stem auditory evoked potential. Hearing conserving results are impaired by tumors with such

predominant growth directions. The extent of caudal and anterior tumor growth is of prognostic value for hearing preservation.

Tumor extension and Facial nerve function.

The same criteria as for the cochlear nerve are applicable to facial nerve to a milder degree. The incidence of impaired facial nerve function is increased in case of tumor growth anteriorly and caudally, but larger extensions are better tolerated than in case of cochlear nerve. This evaluation is based on the experience that deteriorated preoperative function is more often associated with necessity of nerve reconstruction. Moreover, exact analysis of the tumor extension may help to identify those patients who are at increased risk of nerve damage despite normal preoperative function. Despite increasing numbers of reports on neuroradiological delineation of the tumor- nerve relationship by MRI, it is a misunderstanding to think that the demonstration of neighborhood between the facial, cochlear, trigeminal nerves and the tumor really informs about their relations. Cochlear nerve symptoms occur in 95% and facial nerve symptoms only in maximally 17% demonstrates that biological relation is the important one and is very different from the anatomico-radiological hints. Factors like tumor shape or cystic changes or inclusions might provide information on the tumor biology and correlate with biological relation and function. Mulkens et al reported

the problem of inhomogeneous contrast uptake on MR images, which limit the delineation of tumor extensions. Samii et al reported that contrast uptake is variable inside the IAC, limiting preoperative prediction of the extent of intrameatal tumor extension.

The labyrinthine system and hearing function

If the preoperatively calculated minimal length of the posterior IAC wall is not respected, fenestration or complete opening of labyrinthine system is most likely; even in case of cochlear nerve preservation, the rate of functional acoustic preservation is reduced by at least 50 to 66%, mostly to a minimum in such cases. As has been shown in previous study, fenestration and even complete opening of labyrinthine structures do not necessarily cause complete hearing loss but they reduce the rate of hearing preservation and reduced the quality of preserved hearing. The danger of labyrinthine opening can be calculated preoperatively by the *fundus-sinus line.* If this indicative of a danger of fenestration (Grade B), real fenestration or opening occurred in 10 to 16%; if this was indicative of destructive opening (grade C), this 30 to 33% (Samii's series).

These datas speak in favor of ascertaining these calculations before surgery to establish the danger and to decide in which cases intraoperstive

microendoscopy should be used to reduce the necessity of bony opening or when a middle fossa approach might be preferable.

Limitations of Radiological investigations

The four aspects that remain obscure despite all radiological refinements and analyses, includes the tightness of the tumor nerve relationship, the consistency of the tumor, the vascularisation of the tumor and the extent of tumor within the meatus. The tightness of the tumour nerve relationship might depend on the involvement of arachnoid sheath; this biological aspect is unforeseeable, although it is highly important avoiding surgical difficulties and achieving functional nerve preservation. Tumor consistency or vascularisation cannot be predicted. The degree of contrast uptake; sometimes cystic tumor formation is visible and usually predicts a more critical involvement of neural tissue and higher tendency towards bleeding. The intrameatal tumor extension is not completely reliably as determined by MRI because intrameatal contrast uptake is inhomogeneous.

Approaches outcome and complications

Approaches

The prevalence of certain problems and complications is generally correlated with the surgical approach that is translabyrinthine, middle fossa, and retrosigmoid approaches. By the extended translabyrinthine-transotic

approach, as reported by Chen and Fisch in 147 patients, very good results with regard to mortality (1 of 147 patients) and morbidity and anatomic facial nerve preservation (95%) are achieved. Brackmann and Green advocate the translabyrinthine approach in patients who are deaf or who have poor chances of hearing preservation, they state that this approach has the lowest morbidity with regard to spinal fluid leaks and postoperative headaches and provides the advantage of minimum cerebellar retraction, identification of facial nerve proximally and medially and ability to repair immediately the facial nerve when severed during acoustic tumor removal. Because the value of hearing preservation is now appreciated, a categorical decision for the translabyrinthine approach has been given up. Brieggs et al found a higher incidence of CSF fistulas (11.6%) and infection in large tumors (4.6%) with hydrocephalus. Hoffman found no significant difference in CSF fistula incidence between translabyrinthine group (21%) and the retrosigmoid transmeatal group (16%). Pellet et al reported on 228 patients, 85% were operated on using the translabyrinthine approach and 15% using the middle fossa approach. The mortality rate was 1.75%. Anatomic facial nerve preservation was achieved in 94% of the patients. Hypoglossal facial anastomosis was necessary in 10% and radical excision was achieved in 99%. Five percent were cured by lumbar punctures, and 2.5% were cured by

surgical revision. Meningitis occurred in 0.4%, postoperative hematomas occurred in 1.75%, and balance troubles occurred in 67 %. Haid and Wigand reported on 263 patients who were operated on using the middle fossa approach for tumors that had up to 35mm extrameatal extension with radical excision in 96%, excellent facial function in 78%, persistent paralysis in 6%, and good rate of hearing preservation. Mazzoni et al reported on selected series 90 of 300 patients operated on using the suboccipital approach for a trial of hearing preservation. The facial nerve was preserved in 99%, with completely normal function in 78%. The cochlear nerve was anatomically preserved in 96% and functionally in 44%. CSF leak occurred in 6.6% of the cases, meningitis in 2.2%, paresis of lower cranial nerves in 3% and ataxia in 2%. Glasscock et al reported on the suboccipital and middle fossa approaches in 161 selected patients. They found a lower incidence of temporary facial nerve paresis using the suboccipital approach but also a higher incidence of postoperative headaches using this approach. The literature provides proof of the statement that results are best achieved by the technique with which the surgical team is most familiar.

Completeness of resection

The feasibility of total tumor removal and hearing preservation is under suspicion by some colleagues because of histological finding of tumorous

nerve infiltrations. In samii's series, minor incidences of recurrences (6\880) were reported by total microsurgical removal where relevant tumor rests evidently were not left behind for the sake of hearing preservation. The attitude of removing the tumor immunohistologicaly complete would imply the occasional resection of a macroscopically intact cochlear nerve. That nerve according to many surgeons never shows any tumor recurrences and would function well for decades, as is evident in increasing numbers. Regarding CSF leak (Samii's series), only 1.6 % of patients needed surgical revision because of a real internal fistula, paradoxical rhinorrhea was observed in 7.6% of patients. The incidence of bacterial meningitis in Samii's series was 1.3% Current mortality rate is approximately 1 to 2%. Careful selection of patients and decision against surgery for patients who are at high risk, such as patients with preexisting nerves deficits, hemiparesis, or cardiopulmonary compromise, would, as in all surgical disciplines improves the outcome.

Patients having major risk for surgery

1. Premobid patients with previous surgery or with severe brain stem compression.
2. Tumors with caudal cranial nerve involvement and complete palsy after Surgery.
3. Cystic tumors where the chance of postoperative hemorrhage is higher.

Complication avoidance

Brain stem safety:

Regarding the safety of brain stem, the technique of dissection under continuous irrigation is especially helpful because cauterization may be reduced to minimum. Identification and control of vascular supply to the brain stem are most reliably provided by the suboccipital approach, whereas overview as well as access is limited in the middle fossa and the translabyrinthine/ transotic approaches.

Facial nerve

The argument that the middle fossa or posterior approaches, the only routes enabling hearing preservation, are more dangerous to the patient or especially to the facial nerve than the translabyrinthine route was discarded some time ago. Regarding the facial nerve, the translabyrinthine and the suboccipital approaches offer the best opportunities for preservation as well as for reconstruction of the facial nerve. Tos et al found suction to be a damaging process during nerve preparation. They emphasis that the most dangerous part for facial nerve dissection is just medial to the porus.

Cochlear nerve

Cohen was one of the first and few who reported on the indications for using the middle fossa and suboccipital approaches for hearing preservation when

the majority of his colleagues still insisted on the translabyrinthine approach. Even his early results were very favorable, with only 8% facial nerve paralysis and no mortality. Many authors now report functional cochlear nerve preservation rates from 25% to higher than 50%. Additional experience with both approaches enabled Cohen to formulate the advantages of the suboccipital approach as providing visualization, access and control of all relevant structures and the chance of functional nerve preservation. During the analysis of intraoperative auditory brain stem responses three changes can be observed, latency increase, amplitude decrease and loss of an ABR component. The latency increase and amplitude decrease usually indicate and precede the loss of a wave. The assumptions and criteria for successful ABR preservation, with resultant hearing preservation, have been quite controversial.

- *Possibility of early recognition of significant ABR changes.* One of the most important aspects criticized in monitoring is the phenomenon of the point of no return, i.e., in the worst situations; the neurophysiologist reports wave losses equivalent to deafness when the situation is already permanent. There should be hints for recognition of impending danger of permanent loss while the loss is still temporary.

- *Correlation between temporary or permanent wave losses and the risk of deafness:* A loss of wave 5 is the most definite sign most often associated with deafness, whether this loss is temporary or permanent, and the permanence cannot be foreseen when the wave first disappears. Although it is fairly reliable in indicating definite hearing loss, this sign is the least helpful because it is the consequence of earlier warning signs, i.e. the disappearance of wave 1 or 3. Because wave 3 has the highest incidence of actions with associated loss, it is the most sensitive warning sign, even more sensitive than wave 1 loss. In cases with some hearing preservation, ABRs without wave 3 generally indicate poor quality hearing, as was also shown by correlation of ABR quality with speech discrimination. Complete loss of wave should be prevented by early recognition of its deterioration. Any deterioration of wave 3 must attract special attention, and the surgeon must be warned. The intraoperatively recorded ABR is a snapshot like documentation of the function of the auditory pathway; in the case of loss of single component or several components during surgery, the initiated degenerative processes may continue after the end of surgery and monitoring and may lead to total wave loss, associated with deafness. In conclusion, isolated wave loss during

surgery bears some risk of deafness, early warnings and information during monitoring is helpful. Waves 1 and 3 must be monitored especially carefully during drilling, intrameatal preparation, pulling medially or laterally, hemostasis, and implantation of muscle on the opened mastoid. As soon as soon as one wave is not visible twice, this is reported and a break in surgical dissection is suggested. If the neurophysiologist finds deterioration and asks for a short break, the surgeon immediately stops the ongoing action and awaits recovery or changes the site and type of surgical action. Thus, those dangerous actions are performed stepwise on very sensitive nerves and are tolerated far better.

- *Action dependent wave changes :* Wave 5 changes especially after cerebellar retraction and during mobilization of AICA during acoustic neurinoma surgery (Soulier et al). Intraoperative ABRs predictive of hearing outcome in selected cases"; specifically, when wave 5 was unchanged at the end of the operation, even if it may have been transiently lost during surgery, useful hearing was invariably preserved.
- *Specificity of wave changes for surgical manipulations:* Pulling of the tumor nerve bundle in the medial direction, toward the brain stem and

away from the cochlea, is associated with frequent amplitude reduction and even loss wave 1, which represents the cochlear activity. Wave 1 is mostly stable with pulling in other directions. If wave 1 disappears on medial pulling, then wave 3 may also be disturbed, as a consequence of either wave 1 deterioration, some compression of tumor nerve bundle onto brain stem, or distortion of the nerve. Direct dissection at the auditory nerve is also followed by deterioration of waves 1 and 3.

Hearing function (Samii's experience with 1000 operated cases)

Is completeness of resection and hearing preservation possible or contradictory ? and what are there indications for subtotal resections?. Some question the feasibility of complete excision and reject the principle of total tumor removal and simultaneous hearing preservation because of histological or immunohistochemical findings of tumorous infiltrations of cochlear nerve. To date the minor incidence of recurrences (1.4% in the case of hearing preservation in patients without NF-2) and stable long-term hearing function in Samii's series illustrate the safety of the presented way of total microsurgical removal. In the majority of cases the so-called acoustic neurinoma is a vestibular schwannoma originating from the cochlear nerve. In patients with good hearing, the intraoperative situation is

usually characterized by an easy separation between the cochlear nerve and the tumor. Deliberate subtotal tumor removal should be performed only in rare cases of life saving decompressive surgery or in the case of a special agreement with patients in whom a tumorous cochlear nerve is anticipated. Several successful cases of partial tumor removal or of decompressive surgery performed by opening the IAC have been reported; these procedures should be performed only by someone with special experience in hearing preserving surgery, because otherwise, as a result of the extreme vulnerability of the cochlear nerve, hearing would most likely be seriously impaired or lost despite incomplete tumor removal. Moreover such patients need especially close radiological and clinical follow-up, to detect tumor regrowth and to establish whether additional surgery is necessary.

Radiotherapy alone or in combination before acoustic neuromas surgery

The prolonged goal in radiotherapy / radiosurgery is tumor control, the radiological proof of no tumor growth, which is reported in more than 90% of patients with 1 to 2 years of follow-up whereas tumor shrinkage occurs on approximately 20% of patients. However, spontaneous growth cessation and secondary re-growth or slow and variable growth of 1 to 4mm per year have been reported and put the actual therapeutic impact of radiation under dispute. Hearing preservation rates show a time dependent decrease initially

above or near 50% to approximately 30% within 1 year. Although experience is still limited, radiosurgery of any kind probably does not provide a long-term solution for most patients and does not offer better hearing results than does surgery with an experienced surgical team. Radiosurgery may be a good alternative in elected rare cases that are inoperable because of other medical reasons or for multiple tumors in NF –2, it is apparently no alternative if stable hearing preservation is required; presently, it cannot be recommended for the remaining ears of patients with NF-2, because delayed hearing deterioration is too frequent and too severe after radiotherapy. Surgery after previous radiotherapy is more difficult, is more dangerous, and provides less favorable results. The neural structures are paler and appear less vascularized, and the tumor is more adherent because of scarring of the arachnoid plane. Chances for hearing preservation are much better in the case of short symptom durations. Some surgeons advocate the pre-microsurgical philosophy of observation rather than removal, to avoid disturbing intact functions. Early surgical removal advocated by Shelton and Hitselberger, who reported 67% hearing preservation and nearly normal facial function in 97% of cases at 1 year postoperatively for tumors up to 5mm in size, with the middle fossa approach.

- *Criteria of useful hearing quality:*

  Hinton et al mentioned "the important difference between hearing preservation which pleases the surgeon and that which will be appreciated by the patient", and they think that only 1 to 10% of patients might meet the criteria of useful hearing, such as an SDS of 50% or more, a PTA hearing loss of up to 30 dB and a tumor size of less than 2 cm.

Gardener Robertson modification of Silverstein Norrell system

Class	Decibel loss	Speech discrimination score
Good (class 1)	0-30	70-100%
Serviceable (class 2)	31-50	50-59%
Non serviceable (class 3)	51-90	5-49
Poor (class 4)	> 90	1-4 %
No hearing	0	0

Rowed et al classified serviceable hearing as a speech reception threshold of less than 50 dB and a SDS of more than 60%, similar to Post et al suggesting PTA hearing level of 50dB and an SDS of more than 50% as good / useful hearing. Nadol et al defined useful hearing as a speech reception threshold of at least 70 dB or an SDS of at least 15%.

Chances of hearing preservation

In 1988, Gardner and Robertson reviewed the current literature and reevaluated cases with a classification system similar to that presented by Silverstein et al. The total number of cases under review was 621, with 221 reported successes. Cases limited to those having unilateral acoustic neuromas, with valid supporting audiometric data, numbered 394, with 131 successful outcomes (33%). there were only 5 cases of hearing preservation with unilateral acoustic neuromas of 3cm or larger. Almost all authors agree that preservation is more likely with smaller tumors with good preoperative hearing, especially with good SDS and in early operations, although Shelton et al did not find any influence of preoperative hearing on the chances of hearing preservation, and McKenna et al did not find any strong correlation between preoperative tumor size or hearing and the chances of hearing preservation. The majority of authors report attempts for hearing preservation with tumors up to 20 mm, equivalent to Samii's classes T1 and T2. Because there is growing agreement about the necessity for training, there are more colleagues advocating attempts for hearing preservation in all cases and there are some using neurophysiological monitoring, with variable success. There are reports of postoperative SDS improvement for some patients with poor preoperative SDS.

Additional factors of predictive importance

Tumor extension plays an important role, advance age as a disadvantageous factor by some authors. There is strong correlation with the preoperative acoustic quality. Male gender seems to be favorable for hearing conservation (samii;s and Nadol et al). Duration of the patient's symptoms may be important, because it correlates with the chances of postoperative hearing preservation; this is valid for the duration of hypoacusis and of vestibular disturbances.

Trends in vestibular schwannoma management

Approximately 2000 patients with vestibular schwannomas are being operated in USA. The number of patients undergoing gamma knife radiosurgery has been steadily increasing each year. Approximately 25% of the patients undergoing radiosurgery had recurrent tumors after previous microsurgical resection failed. Several factors are essential for the continued exponential growth of vestibular schwannoma radiosurgery. First, and most important, it must be documented in peer-reviewed publications that long-term tumor growth control has been achieved in large groups of patients with vestibular schwannomas whose tumor marginal dose was equal to or less than 16Gy. All patients newly diagnosed with small to medium sized

Vestibular schwannomas must be informed of the option of radiosurgery and should have access to this.

Less radical surgery and adjuvant therapy with Radiosurgery

Lownie et al reported intracapsular removal of large acoustic neuromas in 11 patients with long follow-up for functional preservation during the era of microsurgery. The facial nerve was preserved in 9 patients (82%), but in 2 patients (18%) the residual tumor increased in size 2 and 3 years after surgery. *Kameyama et al* followed 19 patients with residual tumor after intracapsular removal and 10 patients (53%) had regrowth. *Ohta et al* reported that the tumor leaving the site of IAC had increased size, and the tumor exiting the brainstem had not increased during the follow-up period of to 8 years. So to reduce the operative morbidity and preserve the functional outcome, intracapsular removal may be one operative approach in the era of microsurgery. But from the standpoint of tumor growth control, some adjuvant therapy may be needed for long - term tumor growth control. Kondziolka et al reported the 5 years follow up results in 165 patients and achieved 98% clinical tumor growth control with radiosurgery. Flicklinger et al reported tumor control rate 91.0%, no patients who received dose less than 13 Gy for tumor margin suffered facial palsy. Prasad and Steiner treated 159 patients with acoustic neurinoma with gamma knife radiosurgery; they

reported 89% tumor growth control in patients that had undergone previous operations and 94% tumor growth in the nonoperated patients along with 40% hearing preservation, facial palsy occurred in only 2% and new trigeminal neuropathy occurred in 3 %. These experiences revealed that radiosurgery could achieve long-term tumor growth control with very low morbidity, including facial palsy with hearing preservation. In cases of large acoustic neuromas, subtotal removal and subsequent radiosurgery is a useful treatment strategy for maintaining cranial nerve function and long- term tumor growth control.

MATERIALS AND METHODS

This is a retrospective study of hundred operated cases of vestibular schwannomas over a period of three and a half years in the department of Neurosurgery in Sree Chithira Thirunal Institute for medical sciences and Technology, Thiruvananthapuram. Chi- square test was applied for Statistical evaluation.

PERIOD OF STUDY

All cases admitted electively from November 1999 to June 2003 were evaluated clinically for documentation of the preoperative deficits with routine investigations and imaging for the lesion done by CT scan, MRI or both. Quantification of the hearing loss was done with pure tone audiometry. Size of the lesion was measured from the available imaging modality by taking the largest diameter among the sagittal, coronal or axial planes. Lesions were classified according to the size as below.

SIZE OF TUMOR	TERMINOLOGY
Less than 1 cm	Small
1 cm to 2.4 cm.	Medium
2.4 cm to 4.5 cm	Large
More than 4.5 cm	Giant

According to this clinical symptoms, signs, operative findings, postoperative deficits, period of hospital stay and operative outcome of giant tumors were

compared with that of tumors of smaller size. All patients were followed up till this time and their recovery from the postoperative deficits were also recorded and compared between two groups.

RESULTS AND ANALYSIS

NUMBER OF CASES

TOTAL NO. OF CASES.	MALES	FEMALES
100	44	56

$X^2 = 1.44$ (Chi- square test, not significant)

SIDE OF LESION

Total no. of cases	Right	Left	Bilateral (NF-2)
100	45	50	5

NATURE OF TUMOR

SOLID TUMOR	CYSTIC
94	6

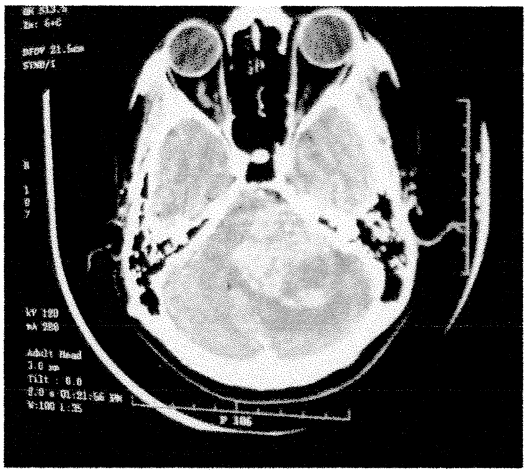
AGE INCIDENCE

AGE	No. of cases	Percentage
0 -10 yrs	0	
11-20 yrs	0	
21-30 yrs	7	7%
31- 40 yrs	38	38%
41-50 yrs	43	43%
> 50 yrs	12	12%

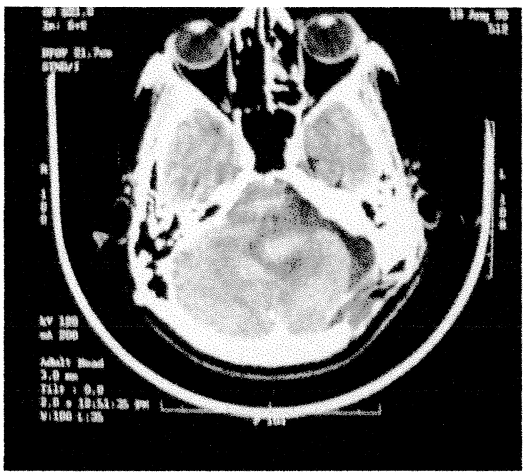
Median age is 42.162 years.

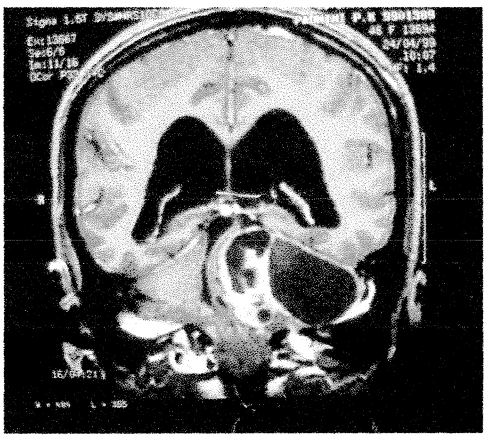
GIANT TUMORS

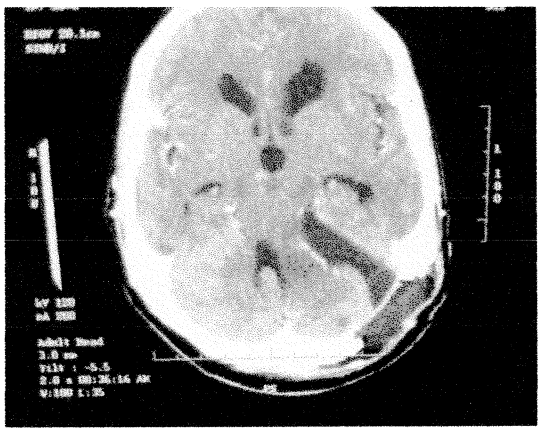
Preoperative



Post-operative

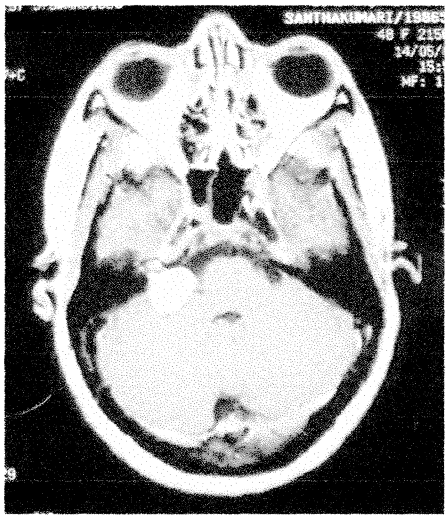




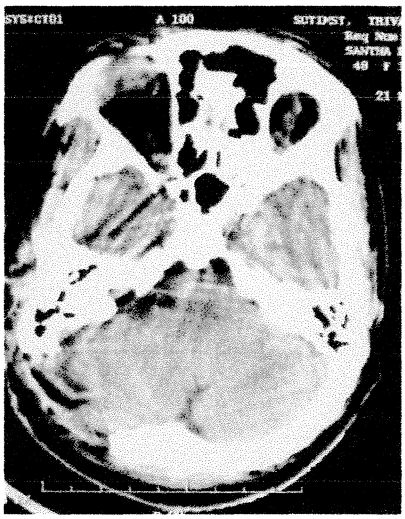


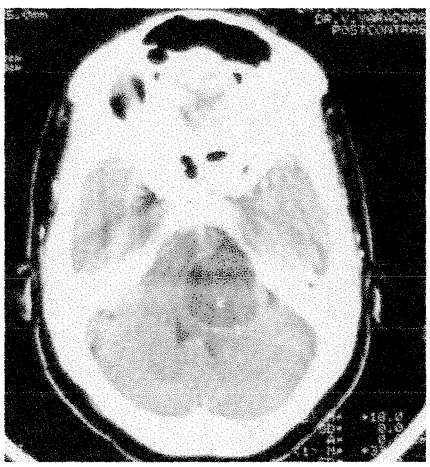
SMALL AND LARGE TUMORS

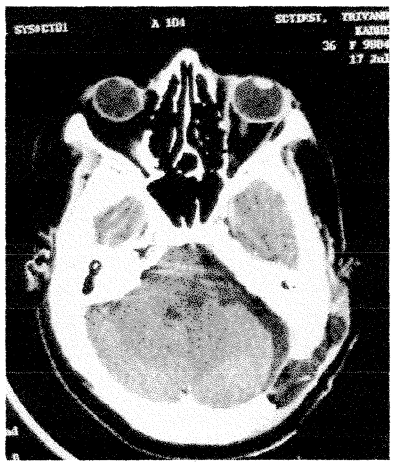
Preoperative scan



Post-operative scan







SIZE OF THE LESION

<1 CM	0
1.0- 2.4 cm	3
2.5 - 4.5 cm	72
> 4.5 cm	25
Total No. of cases	100

PREOPERATIVE SIGNS

RAISED ICP (Papilloedema)

	< 4.5 cm	**4.5 cm**
No. of cases	51 (68 %)	19 (76%)

X^2 = 0. 5714 (not significant)

SENSORY FIFTH NERVE INVOLVEMENT

	< 4.5 cm	**> 4.5 cm**
No. of cases	63 (84 %)	24 (96 %)

X^2 = 2.3813 (not significant)

FACIAL PALSY

	< 4.5 cm	> 4.5 cm
No. of cases	41 (54 %)	14 (56 %)

X^2 =0.0135 (not significant)

PRE-OPERATIVE HOUSE- BRACKMANN GRADING OF FACIAL PALSY

H B GRADES	< 4.5 CM (41)	> 4.5 CM (No.14)
1	0	0
2	25 (60.9%)	0
3	8 (19.5 %)	1 (7.14 %)
4	5 (12. 19%)	1 (7.14 %)
5	3 (7.31 %)	10 (71.4%)
6	0	2 (14.28 %)

PRE-OPERATIVE HEARING STATUS

Decibel loss	< 4.5cm(No: 75)	>4.5cm(No: 25)
> 50dB	55 (73.3%)	20 (80%)

X^2 = 0.444 (not significant)

PRE- OPERATIVE LOWER CRANIAL NERVE PALSY

	< 4.5 cm	> 4.5 cm
No. of cases	6 (8 %)	5 (20%)

X^2 = 4.4569 (not significant)

PREOPERATIVE CEREBELLAR SIGNS

	< 4.5 cm	> 4.5 cm
No. of cases	57 (76 %)	23 (92 %)

$X^2 = 3.0$ (not significant)

PREOPERATIVE EVIDENCE OF HYDROCEPHALUS

	< 4.5 cm	> 4.5 cm
No. of cases	44 (58.6 %)	23 (92 %)

$X^2 = 0.015$ (not significant)

PREOPERATIVE CSF DIVERSION

	< 4.5 cm	> 4.5 cm
External Ventriculostomy	1	2
VP shunt	1	0
Endoscopic third ventriculostomy	1	0
Total	3	2

SURGERY

All patients underwent retrosigmoid approach through retromastoid craniectomy in lateral position with head fixed on Mayfield kees clamp, Adequate cerebellar retraction achieved by releasing CSF from Cisterna magna. Technique of surgery involves initial intra-tumoral decompression of tumor followed by separation of wall by dissecting in the proper arachnoid plane. Cavitron ultrasonic surgical aspirator was used in almost all cases for tumor excision. All neural structures in the tumor bed were identified as possible including fifth nerve, lower cranial nerves, seventh – eighth complex as well as vascular structures including superior petrosal vein and anterior inferior cerebellar arteries. Total tumor excision was possible in almost all cases except few were near total removal was done. In almost 60 % of cases in giant as well as large tumors anatomical preservation of facial nerve was done. Fascia lata graft was used for watertight dural closure in all cases. Twenty four to forty eight hours' ventillatory support was given to indicated cases and used to be weaned off afterwards. Patients were allowed to take oral fluids after forty eight hours if there is no features of lower cranial nerve palsy.

SUGERY

SURGERY	< 4.5 cm No. 75	>4.5 cm No. 25
Total excision	75 (100%)	22 (88%)
Near total excision	0	3 (12%)

ANATOMICAL PRESEVATION OF SEVENTH NERVE

	< 4.5 cm	> 4.5 cm
Anatomical preservation of seventh nerve	65 86.6 %)	15 (60%)

$X^2 = 8.335$ (significant)

OPERATIVE CONPLICATIONS

One patient in large tumor group had evidence of thin acute subdural hematoma in the right fronto-parietal region, which was managed conservatively as patient was neurologically intact and was discharged. Later patient presented with deterioration of sensorium and headache after three weeks. On evaluation found to have sub acute subdural hematoma with mass. Patient underwent evacuation of the same and had an uneventful recovery. Whereas in the giant series one patient had an acute extradural hematoma which was detected in the third postoperative day and evacuation of the same done as an emergency procedure and improved well. One more patient in the same group had evidence of posterior fossa hematoma, which was managed conservatively as patient had no deficits and he improved without any deficits. No operative mortality in this series. One patient in the giant as well as one in the larger tumor group underwent Fascio- hypoglossal anastomosis for grade 6 fascial palsy without much rewarding results and still under followup for assessing the outcome of the procedure.

POSTOPERATIVE FRESH SEVENTH NERVE PALSY

	< 4.5 cm (Out of remaining 31 cases)	**>4.5cm (Out of remaining 11cases)**
Seventh nerve palsy	17	11

HOUSE- BRACKMANN GRADES

H-B GRADES	**< 4.5 cm**	**> 4.5 cm**
1	0	0
2	0	0
3	4	0
4	3	3
5	10	8
6	0	0
TOTAL	17	11

POSOPERATIVE LOWER CRANIAL NERVE PALSY

	< 4.5 cm (No. 69)	**>4.5 cm (No. 20)**
Lower cranial nerve palsy	4 (5.79 %)	5 (25 %)

$X^2 = 6.29$ (significant)

POSTOPERATIVE CSF DIVERSION

CSF diversion	**< 4.5 cm**	**> 4.5 cm**
Endoscopic third ventriculostomy	2	1
External ventricular drainage	1	0
VP shunt	3	0
TP shunt	5	0
Total	11	1

MEAN HOSPITAL STAY

	< 4.5 cm	**> 4.5 cm**
Mean hospital stay	15.3 days	18.2 days

DISCUSSION

In our series of hundred *Acoustic neurinomas,* which were operated in our Hospital (S C T I M S T, *department of Neurosugery)* during a period of 4-Years were retrospectively analyzed with respect to the size of the tumor, the extent of clinical profile and operative outcome. Reviewing the literature *Acoustic neurinomas* are classified as giant tumors (L.N.SEKHAR) when it measures more than 4.5 cm in any dimension, as large tumors once the size lies between 2.5 to 4.5 cm and medium and small tumors having the dimension 1.0 to 2.4 cm and < 1.0 cm respectively. We have analyzed all cases by taking the largest measurements of all tumors in axial, coronal and sagittal plane in MRI or CT scan. Out of the total 100 cases operated 25 were giant, 72 cases were large and three cases were below 2.5 cm in diameter. All patients had preoperative assessment of hearing status with pure tone audiometry and those with profound hearing loss were identified. Majority of cases (97%) operated were more than 2.5 cm in diameter with profound hearing loss, so preservation of hearing was out of question and was not attempted also. Samii' series shows show higher incidence of acute hearing loss with a frequency of single attack of 10%, several attacks of 6.1% and acute hearing loss with permanent deafness around 3%. All patients were admitted electively and was properly worked up with specific

emphasis on preoperative deficits (quantification of hearing - loss). All patients were imaged preoperatively with C T scan, MRI scan or both. Of the total 100 cases operated during 4-year period 44 were males and remaining 56 % were females with no definite statistical significance. Among these 43% were in the age group between 41 to 50 years and next Majority were in the 31 to 40 group, youngest patient was of 17 years (N F-2) of age and 63 year old patient who was operated was the eldest in this series. Reviewing the literature in Samii's series age group ranged from 11.1 to 87.6 years, patients with the largest tumors were symptomatic at the earliest age, 44.4 years, where as those with only intrameatal tumor extension had a mean age of 48.9 years at surgery. The average age of the female patients was higher than that of the male patients (47.6 versus 45.2). Samii's series inference is that if age is correlated with hearing loss, but inversely correlated with the tumor size, many large tumors must be found in young patients with good hearing. Out of the total cases, 50% of lesions were on the left side and 5 % of patients had bilateral lesions (N F-2). In 1000 cases of vestibular schwannomas by Samii, he operated 120 tumors in total 82 patients, in 38 patients bilateral tumors were operated and in 44 patients only unilateral resections were done. Todd H Lanman has reported an incidence of 14.2% for NF-2 in his series of operated 190 cases of large

vestibular schwannomas. According to the criteria, of the total 100 cases operated, 25 patients were having giant lesions with an average diameter more than 4.5cm, 72 patients having large tumors (2.5 to 4.5 cm) and remaining 3 patients were having evidence of medium sized lesions. Contrary to the literature our patient series comprised large lesions as majority and giant tumors formed a significant bulk of the total. Nineteen patients (76%) in the giant series had clinical as well as radiological evidence of hydrocephalus and almost similar incidence was observed in the large tumor group with an incidence of 68%, no statistical significance was observed. Sensory fifth nerve involvement was a predominant clinical sign noted in 96% of patients in the giant group compared to 84 % of the remaining patients, with no statistical significance. Samii's series showed an incidence of 11 to 16% for the same, remarkably low when compared to our series comparing to the literature the incidence of pre –operative facial palsy was higher in our series with no statistical difference between giant as well as large tumors with involvement of 56% and 54% respectively. Out of total 43 patients in the large tumor group 60.9% had grade 2 (mild) palsy and three-fourth of the giant series had profound facial weakness (grade 4 and 5). Samii' s series showed incidence of facial nerve symptoms in 6% of cases and he has reported frank facial paresis in 17% of patients with House-

Brackmann score of 2 to 6 in three-fourth of cases. Except few all patients in the giant as well as large tumor group had presented with progressive hearing loss as the initial and common Symptom with no statistically significant difference, among these 73.3% of patients with large tumors and 80% of patients having giant lesions had evidence of non-serviceable hearing loss. Considering this factor hearing preservation out of surgery was out of question in our series. In Samii's series 95% noticed some degree of hearing deficit for an average 3.7 years. Five percent of patients had no clinical as well as audiometric evidence of hearing deficit. Three percent of patients presented with evidence of acute deafness with permanent disability. Out of the total 46% of patients presented with tinnitus along with deafness. Lower cranial nerves were involved in 6 (8%) in the larger tumor series were as it was seen in 20% of the larger group (significant, X^2 = 4.456), no patients required a pre-operative tracheostomy. Samii's series showed an incidence of 3.5% for the lower cranial nerve palsy, among this 2.7% of patients had symptoms pertaining to that. Cerebellar ataxia was observed as frequent affection in both groups with an incidence of 92% and 76% in giant and large tumor series (no statistical significance) respectively. Samii's series has showed an incidence of 30.3% for the same, mainly gait unsteadiness, and Todd. H. Lanman in his series of 190 patients with large

tumors has shown an incidence of 12.1%, our group of patients noticed remarkably high occurrence of this deficit. Three patients in the large group and 2 in the giant tumor group underwent pre-operative CSF diversion for hydrocephalus. Out of the total 3 patients had external ventriculostomy, one had ventriculoperitonial shunt and for one patient endoscopic third ventriculostomy was done. All patients underwent *retrosigmoid approach* and total excision of lesion was achieved in all cases except 3 cases in giant tumor group where near total excision was done. Anatomical preservation of seventh nerve was possible in 86.6% of large tumor group and 60% of giant group, has shown statistically significant possibility in large tumor series. In Samii's series of 1000 operated cases 979 patients had total removal of the lesion and in 21 cases deliberate subtotal removal was done with the aim of life saving surgery in 10 cases and of hearing saving surgery in 11 patients. In Samii's series out of the total 979 tumors for which radical removal was done anatomical preservation of facial nerve was achieved in 929 cases (93%). The preservation rate ranged from 87 to 94% for large and small tumors respectively. Facial nerve function, graded according to the House-Brackmann scale within 2 weeks in Samii's series after surgery, was grade 1 in 47%, grade 2 in 12%, grade 3 in 14%, grade 4 in 6%, grade 5 in 10%, and grade 6 in 11% of cases. Whereas in our series out of total 17 patients who

had fresh facial palsy in postoperative period with large tumors 58.8% had grade 5 score and 72.2% of patients out of the total 11 patients in the giant group noticed grade 5 weakness. Almost all patients had uneventful postoperative period except one patient in the large tumor series who had evidence of supratentorial acute subdural hematoma which was conservatively managed initially and later underwent burr hole evacuation for the same for symptomatic sub acute *SDH* after 2 weeks, patient had an uneventful recovery. Another patient in the giant had undergone evacuation of posterior fossa *EDH* and one more patient in the same group had evidence of cerebellar hematoma, which was managed conservatively. There was no operative mortality in our series. M.Samii had 11 deaths in series (1.1%) majority where due to co-morbid medical illnesses. Samii's series showed evidence of hemiparesis in total 10 patients and one patient had quadriparesis, in our group of patients we didn't have any motor weakness. Samii has shown an incidence of CSF fistula in 9.2% of patients, whereas in our series 5% of patients had intractable pseudomeningocele for which theco-peritonial shunt was done and had good recovery. Postoperative hemorrhage in Samii's series was 2.2% where surgical revision was needed in 1.5%, hematoma was located in the CP angle in 4 cases, intrapontine in 2, and in the epidural space in one case, and this is comparable to our series.

Postoperative lower cranial involvement was seen in 5.79% and 20% in large and giant groups respectively with appreciable statistical significance between two groups, Samii 'series has shown almost same incidence of 5.5 %. Eleven patients in the large tumor series and one patient in the giant group underwent postoperative CSF diversion procedure; majority in these had thecoperitonial shunt for intractable psuedomeningocele (5/11). Mean hospital stay for large tumor group was 15.3 days where as it was 18.2 days for the giant series. Reviewing the literature recurrence rate noticed in M.Samii's series was 0.8% (7/880), the recurrence was higher for those with hearing preservation (1.4%) than for those without hearing preservation (0.48%). In our series it is quite premature to comment about this as our cases are still under follow-up.

CONCLUSION

Vestibular schwannomas remains as an operative challenge for even experienced neurosurgeons, operative outcome depends mainly on the operative skill and experience of the surgeon rather than size of the lesion and extent of clinical deficits. In our series by analyzing 100 cases of operated vestibular schwannomas of which 25 cases fulfilled the criteria of Giant tumors, we compared the clinical course and operative outcome of these tumors with tumors of smaller size and found to have no significant statistical correlation for the tumor size with clinical spectrum and operative outcome.

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