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Surgical Treatment of Acoustic Neuromas Through Retrosigmoid Approach

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ABSTRACT

Background: Acoustic neuromas are rare, benign tumors arising from Schwann cells of vestibular portion of 8th cranial nerve. Frequent presenting symptoms are unilateral hearing loss, and subsequent tinnitus and vertigo.

Objectives: Surgical treatment of acoustic neuromas is to perk up patient outcomes and quality of life through development of safe, effective, and efficient surgical approaches.

Methods: The study was conducted at Mardan Medical Complex, Mardan, from January 2018 to January 2023, comprising 25 patients diagnosed with acoustic neuromas through CT scan and MRI. The tumor size was categorized based on its maximum diameter, with tumors less than 1.5 cm classified as small, tumors computing 1.5-3 cm classified as medium, and tumors > 3 cm classified as large, removed surgically through middle fossa, retrosigmoid and translabyrinth approach.

Results: We found that 56% of patients had small-sized tumors, 28% had medium and 16% had large-sized tumors. They were surgically managed and preserved hearing in 33.33, 25 and 0% of small, medium and large-sized tumors, respectively. Facial nerve was preserved in 85.71, 71.42 and 50% of patients of the former categories, respectively. No complications were seen in patients with small-sized tumors, 14.28% in medium and 25% in large-sized.

Conclusion: Surgical treatment is one of the mainstays of management, and relies on various factors, and goal of surgery should be to minimize complications and maximize functional outcomes. For instance, translabyrinthine approach involves the removal of large-sized tumors, middle fossa for small and retrosigmoid for medium-sized neuromas.

Keywords: *Facial nerve; Neurofibromatosis; Retrolabyrinthine surgery; Retrosigmoid surgery; Translabyrinthine surgery; Vestibular Schwannomas*

INTRODUCTION

Acoustic neuromas, also known as Vestibular Schwannomas, are non-cancerous tumors extended to nerve connecting inner ear to the brain. These tumors can cause hearing loss, tinnitus, and balance problems, among other symptoms. These account for 5-10% of entire intracranial neoplasms. They arise from Schwann cells of vestibular portion of 8th cranial nerve and render momentous morbidity and mortality if left untreated 1-3.

Several pieces of research have manifested that genetic mutations take part in development of acoustic neuromas. For example, mutations in NF2 gene are linked to developing bilateral vestibular schwannomas. Exposure to certain environmental factors, such as ionizing radiation or chemicals like vinyl chloride, augments hazards of developing acoustic neuromas. Neurofibromatosis type 2 is a rare genetic disorder that increases risk of developing multiple tumors, including vestibular schwannomas. These are more common in middle-aged and older adults. Once an acoustic neuroma develops, it grows slowly and gradually compresses the vestibular nerve. This compression can cause symptoms associated with acoustic neuromas, such as hearing loss and balance problems. Progressive tumors also compress other adjacent organizations, such as facial nerve and brainstem 4-6.

The clinical manifestations of acoustic neuroma fluctuate depending on size and location of tumors. The most prevailing symptom is plodding auditory loss in one ear. This auditory loss may be escorted by tinnitus or ringing in affected ear. It can cause dizziness, vertigo, and balance problems. These symptoms may be worsened by head movements or changes in position. In various instances, the patients experience numbness in face, tingling or repeated headaches 7.

Due to the swift advancements and widespread accessibility of both neuroradiological imaging (such as CT and MRI) and electrophysiological techniques (like a recording of brainstem auditory evoked potentials), huge number of acoustic neuromas are now being detected in their early stages. Consequently, the percentage of patients who experience inconsequential functional mutilation of facial nerve and hearing is rising progressively, presenting a challenge for physicians, who must meet the patient's expectations regarding preservation of nerve function 8-9.

Therapeutic options for acoustic neuromas are based on size and location of tumor, along with the severity of signs. In some cases, the tumor may be small and slow-growing, and no treatment may be necessary. In other cases, the tumor may be large or growing rapidly, and surgery or radiation therapy may be necessary. Surgery is a common treatment option for larger or faster-growing acoustic neuromas. During surgery, the tumor is removed from the nerve. Kind of surgery used is based on location of tumor and severity of the symptoms. Usual surgical approaches include translabyrinthine, retrosigmoid, and middle fossa approaches. Radiation therapy may also be used to treat small or slow-growing neuromas. This involves the use of high-energy radiation to shrink or destroy the tumor and is typically done over several weeks 10-12.

Over the years, surgical techniques for acoustic neuroma have evolved, resulting in decreased intraoperative and perioperative morbidity and better preservation of facial nerve and hearing functions. Ultimate goal of surgery is to completely remove it without causing any further morbidity 13. The suboccipital approach is carried out with patient in semi-sitting position. However, this position is still a topic of debate among neurosurgeons and anesthesiologists 14-15.

Such surgical interventions are classified into two categories: non-hearing and hearing preservation surgery 16. Non-hearing preservation surgeries are typically performed using translabyrinthine approach, while transpromontorial approaches are also implied in various instants. Other surgical methods concluded retrosigmoid, middle cranial fossa, and retrolabyrinthine approaches, whereby hearing conservation is preferred, and assortment of surgical loom depends on factors like degree of residual hearing, tumor size, location, and patient inclination. If hearing preservation surgery is chosen, the main focus of the procedure should be on preserving hearing. Furthermore, regardless of the type of surgery performed, measures to prevent facial nerve palsy must be ensured, especially in cases where no preoperative deficit is evident 10, 17.

The objectives of research on surgical treatment of acoustic neuromas included evaluation of safety and efficacy of different surgical approaches to remove acoustic neuromas, including translabyrinthine, retrosigmoid and

middle fossa approaches. Assessment of long-term outcomes of surgical treatment, including preservation of facial nerve function and hearing, Identification of risk factors for postoperative complications and strategies to minimize these risks and investigation of patient satisfaction and quality of life following surgical treatment of acoustic neuromas. Overall, main goal of research on surgical management of acoustic neuromas is to improve patient outcomes and quality of life through the development of safe, effective, and efficient surgical approaches.

MATERIAL AND METHODS

Study location

The study was conducted at the Department of Neurosurgery, Mardan Medical Complex, Mardan, Khyber Pakhtunkhwa, Pakistan from January 2018 to January 2023 (05 years).

Study design

A cross-sectional study was performed on 25 patients diagnosed with acoustic neuromas through CT scan and MRI, belonging to different age and sex groups. The tumor size was categorized based on its maximum diameter, with tumors less than 1.5 cm classified as small, tumors measuring 1.5-3 cm classified as medium, and tumors > 3 cm classified as large 18.

Preoperative Evaluation

Before surgery, all patients underwent preoperative audiometry and Brainstem Auditory Evoked Potential (BAEP) recording. Imaging was conducted using routine multiplanar MRI to accurately outline the tumor, as well as a high-resolution skull base CT for evaluating the degree of pneumatization and excavation of petrous bone of internal auditory canal and to establish degree of jugular bulb concerning internal auditory canal. Plain radiography of the cervical spine was also conducted to rule out extensive spondylosis, which could potentially result in nerve root or spinal cord compression due to head rotation and semi-sitting position desired during surgery. The preoperative evaluation included detailed history and physical examination, audiometric testing, imaging and neurologic evaluation. Magnetic resonance imaging is most preferred modality for diagnosis and staging of acoustic neuromas 9.

Procedure of surgery

To perform the surgical approach, a 2.5-3.5 cm incision was made behind mastoid, with inferior part of incision curved slightly laterally to access cerebellomedullary cistern and lower part of CP angle. The initial burr hole was made 2-2.5 cm below superior nuchal line, with 2/3rd behind and 1/3rd in front of occipitomastoid suture. A craniectomy was then performed to expose borders of the transverse and sigmoid sinuses, with a smaller bone opening preferred to avoid potential sinus laceration or desiccation and later thrombosis. The bone opening extended inferior to horizontal plane of posterior fossa. A curvilinear dural incision was made 1.5-2 mm medial to sigmoid and inferior to transverse sinus 19.

Surgical Techniques

Surgical management of acoustic neuromas can be exigent, and option of surgical approach depends on several elements, including size and location of tumors, patient's age and health status, surgeon's experience and inclination. Most commonly implied surgical approaches are translabyrinthine, retrosigmoid and middle fossa.

Translabyrinthine approach involves removal of inner ear and the destruction of hearing and balance organs. It provides direct entrance to tumor, allowing for complete removal, but at the expense of hearing loss and the potential for balance problems. The approach is typically used for larger tumors or those in which hearing preservation is not possible.

In distinction, middle fossa approach involves accessing the tumor through middle cranial fossa, which offers access to internal auditory canal while preserving hearing and balance organs. It is typically implied for smaller tumors that are located in internal auditory canal and that are causing hearing loss. However, this approach is more technically challenging than the translabyrinthine approach and is correlated with higher chance of complications.

Retrosigmoid is another surgical technique utilized for removal of neuromas. It pertains to removing a skull bone segment behind ears, providing access to cerebellopontine angle where tumor is retained. This approach is less invasive than the translabyrinthine approach and allows for preservation of facial nerve and possibility of hearing preservation. During the procedure, the surgeon first makes an incision behind the ear and removes a section of the skull bone to expose cerebellopontine angle. Tumor is then removed using microsurgical techniques, with care taken to preserve the facial nerve and other nearby structures. Following the removal of the tumor, the skull bone is replaced and secured with plates and screws. Retrosigmoid approach is typically utilized for medium-sized tumors that are located in cerebellopontine angle and that may be causing signs like hearing loss, facial numbness or weakness.

The choice among these surgical approaches depends on several factors, including size and location of tumor, patient's preoperative hearing and facial nerve function, and surgeon's option.

Postoperative Care

After surgery, the patients were monitored in ICU. Brain CT, audiometry, and BAEP recordings (for those having preserved hearing) were conducted, and a neurological examination was performed before the patients were discharged from the hospital. Follow-up appointments were scheduled for 6 weeks, 6 months, and yearly intervals thereafter at outpatient department, where the patients underwent neurological assessments, audiometry, and MRI scans to evaluate their current status. The postoperative care of patients with acoustic neuromas focused on minimizing complications and maximizing functional outcomes. Rehabilitation was performed for patients with hearing loss or balance disturbances.

Ethical consideration

The study was commenced after a grant of ethical approval from the Mardan Medical Complex Ethics Review Board and informed patients' consent and willingness were ensured.

Statistical analysis

The statistical analysis was conducted using IBM SPSS for Windows, version 24.0. Group differences in distribution of clinical attributes were assessed using Chi-square tests and Student t-Test, while significance was defined as a probability of a two-sided type 1 error is less than 5% ($p < 0.05$).

RESULTS

Table 1 presented the demographic data and corresponding statistical analysis of study population, intending to identify any significant differences or associations between different demographic factors and study outcome. Average age of participants was 59.82+11.90 years. The gender distribution of the study population revealed that out of 25 patients, 17 were male and 08 were female, with a chi-square value of 3.567 and a corresponding p-value of 0.0589. This indicated that the gender distribution was not significantly different but the p-value is relatively close to the significance threshold of 0.05. Location distribution of the study population showed that 14 lived in urban areas and 11 in rural areas. Out of 25 patients, 04 were working and 21 were not working, with a chi-square value of 10.496 and a corresponding p-value of 0.0011*. This indicated that the employment status distribution was significantly different from what would be expected by chance alone, with a p-value less than the significance threshold of 0.05 (Table 1).

TABLE 1: Demographic data of patients with acoustic neuromas

S. No	Demographic data	No. of patients (n)	Frequency (%)	$\chi 2$	p-value
1	Age (Mean+SD) years	59.82+11.90	----	----	----
2	Gender Male Female	 17 08	 68 32	3.567	0.0589
3	Location Urban Rural	 14 11	 56 44	0.1043	0.7467
4	Employment status Working Not working	 4 21	 16 84	10.496	0.0011*

*indicated that the value is significant

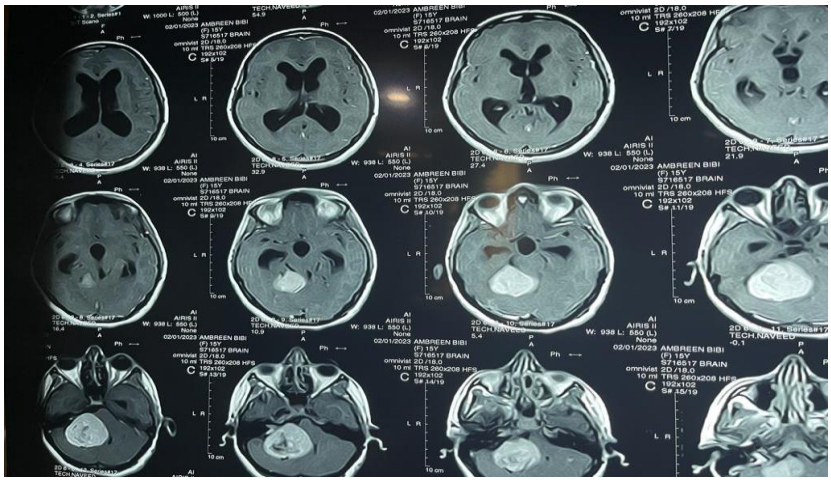
Different categories of tumor sizes were observed in patients (Figure 1 a,b,c). Specifically, tumors were categorized as small if they were less than 1.5 cm, medium if they were between 1.5 and 3.0 cm, and large if they were greater than 3.0 cm. Out of a total of 25 patients, 14 had tumors of small sized category, with a frequency of 56%. The p-value for this group is <0.00001*, indicating a highly significant association between tumor size and the study outcome. Out of a total of 25 patients, 07 had tumors of the medium size category, with a frequency of 28%. Large tumors with a size greater than 3.0 cm were found in 04 patients, with a frequency of 16%.

Overall, the table suggests that tumor size is an important factor in analysis of this patient population, with significant differences observed in the distribution of tumors across different size categories. Specifically, majority of the patients had small tumors < 1.5 cm in size (Table 2).

TABLE 2: Categorization of participants based on the tumor size

S. No	Size of tumor	Category of tumor	Number of patients (n)	Frequency (%)	p-value
1	< 1.5 cm	Small	14	56	0.00001*
2	1.5-3.0 cm	Medium	07	28	
3	> 3.0 cm	Large	04	16	

*indicated that the value is significant



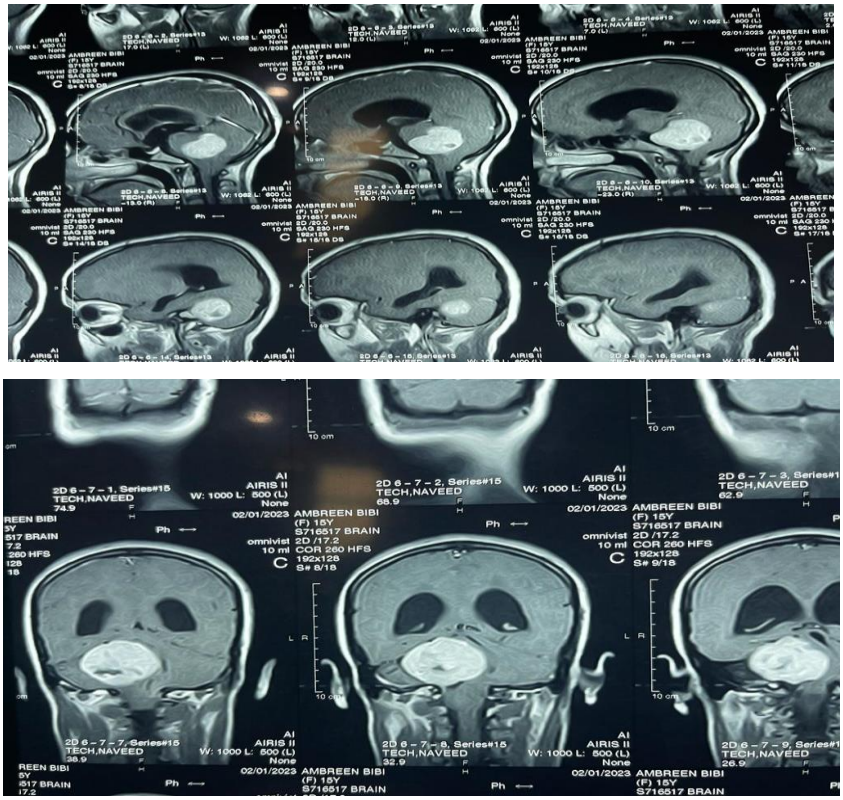


FIGURE 1 (A,B,C): Small, medium and large sizes of intracanlicular acoustic neuromas, respectively

The graphical presentation of the clinical manifestations of the patients with acoustic neuromas revealed that most of the patients had hearing problems (85%), followed by tinnitus (70%), dizziness (55%), vertigo (47.5%), gait problems (47.5%), cephalalgia or headache (42.5%), disequilibrium (25%) and facial numbness (20%) (Figure 2).

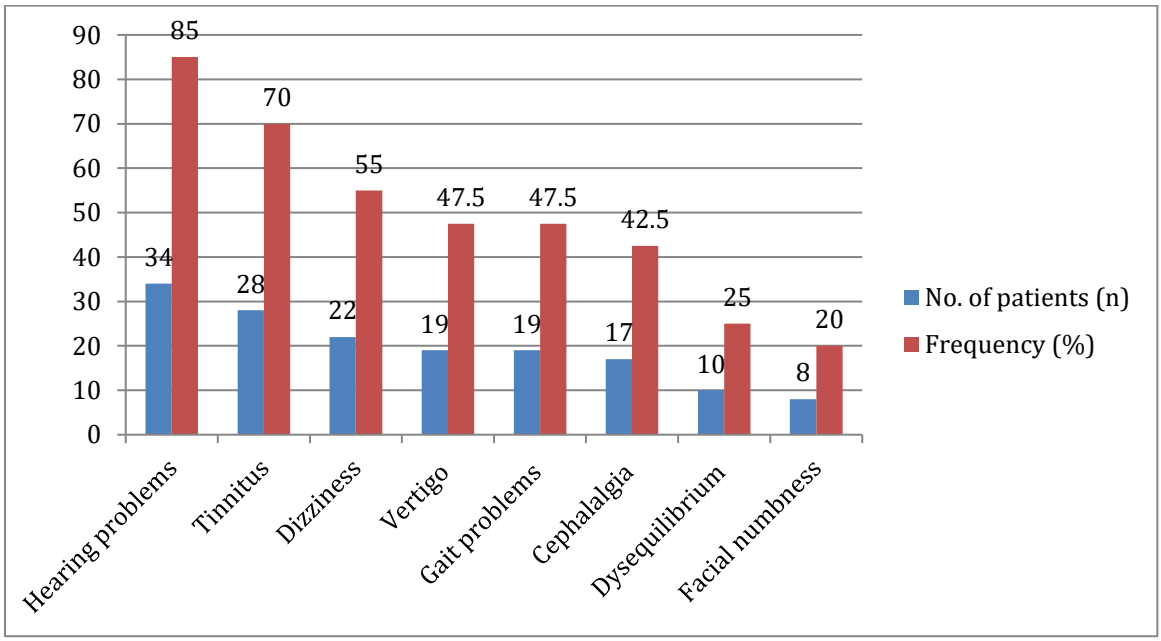


FIGURE 2: Graphical presentation of the clinical manifestations of the patients with acoustic neuromas

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This table presented data related to the pre-and post-operative hearing of patients in different categories of tumor size, along with the percentage of patients who had hearing preservation after surgery, and the corresponding chi-square ($\chi 2$) values and p-values. The objective of this table was to determine whether there was a significant association between tumor size and hearing preservation after surgery. Category of small tumors was with a total of 14 patients. Out of these, 12 patients had a pre-operative hearing, and 04 of these patients had hearing preservation after surgery, which corresponds to a percentage of 33.33%, with a non-significant relationship. Category of medium tumors had a total of 07 patients. Out of these, 04 patients had a pre-operative hearing, and 01 of these patients had hearing preservation after surgery, which corresponds to a percentage of 25%. Large tumors had a total of 04 patients. Out of these, 02 patients had pre-operative hearing, but none of them had hearing preservation after surgery. Therefore, percentage of hearing preservation in this group is 0% (Table 3).

Overall, this table suggested that there was no significant association between tumor size and hearing preservation after surgery in patients with small or medium-sized tumors. However, small sample size for the large tumor category limited the conclusions that can be drawn about this group.

TABLE 3: Data of patients presenting the preservation of hearing after acoustic surgery

Category	No. of patients	Pre-operative hearing	Post-operative hearing	Hearing preservation n(%)	$\chi 2$	p-value
Small	14	12	04	33.33	1.0777	0.2992
Medium	07	04	01	25	0.2187	0.6399
Large	04	02	00	00	2.666	0.1411

The data of patients regarding facial palsy in patients after acoustic surgery demonstrated that facial nerve was preserved in 12 out of 14 (85.71%) patients with small-sized tumors after acoustic surgery, 5 out of 7 (71.42%) in medium-sized and 2 out of 4 (50%) patients had facial nerve palsy in the category of large-sized tumors (Figure 3).

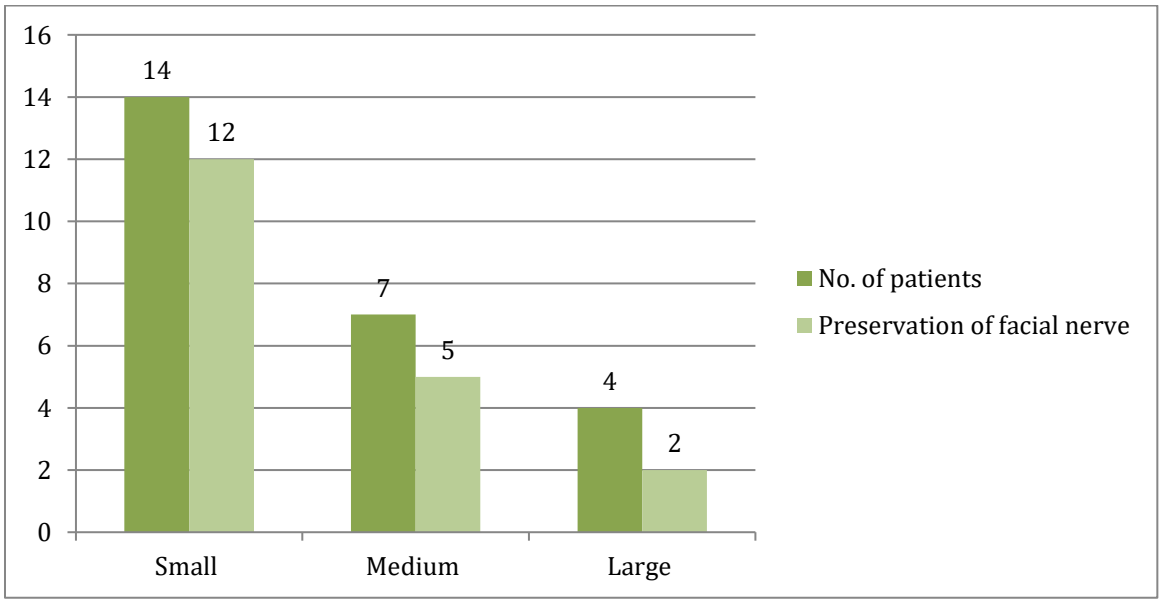

FIGURE 3: Data of patients revealing facial palsy in patients after acoustic surgery

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The number of patients in different categories of tumor size, along with several patients who underwent total tumor resection, partial tumor resection, and slight tumor resection, as well as the number of patients who experienced complications and mortality was shown in Table 4. Patients affected with small tumors were 14, operated on through middle fossa approach and out of these 10 patients underwent total tumor resection, 03 patients underwent partial tumor resection, and 1 patient underwent slight tumor resection. None of the patients in this group experienced any complications or mortality.

Similarly, the category of medium tumors was operated through retrosigmoid approach and had a total of 07 patients. Out of these, 03 patients underwent total tumor resection, 02 patients underwent partial tumor resection, and 01 patient underwent slight tumor resection. One patient in this group experienced complications, but there were no deaths reported. While, category of large tumors was operated through translabyrinth approach and had a total of 04 patients, out of these, 01 patients underwent total tumor resection, 01 patient underwent partial tumor resection, and no patients underwent slight tumor resection. One patient in this group experienced complications, and there was 01 death reported.

Overall, this table suggested that patients with small tumors had the best surgical outcomes, with a high percentage of total tumor resection and no complications or deaths reported. Patients with medium-sized tumors also had good surgical outcomes, with a majority undergoing total tumor resection and a low incidence of complications and mortality. However, patients with large tumors had poorer surgical outcomes, with only half undergoing total tumor resection, and a higher incidence of complications and mortality.

TABLE 4: Tumor resection status of each category of patients after acoustic surgery

Category	No. of patients	Surgical approach	Total tumor resection	Partial tumor resection	Slight tumor resection	Complications	Mortality
Small	14	Middle fossa	10	03	01	00	00
Medium	07	Retrosigmoid	03	02	01	01	00
Large	04	Translabyrinth	01	01	00	01	01

DISCUSSION

Our findings suggested that patients with small tumors operated through middle fossa approach had the best surgical outcomes, with a high percentage of total tumor resection and no complications or deaths reported. Patients with medium-sized tumors operated through retrosigmoid approach also had good surgical outcomes, with a majority undergoing total tumor resection and a low incidence of complications and mortality. However, patients with large tumors were operated on through translabyrinth approach and had poorer surgical outcomes, with only half undergoing total tumor resection, and a higher incidence of complications and mortality. There is no significant difference in hearing preservation among patients with small or medium-sized tumors. However, patients with small-sized tumors had better outcomes of hearing preservation than medium and large-sized tumors, respectively. Facial palsy was observed only in patients having large-sized tumors, while patients with small and medium-sized tumors were operated and their facial nerves were also preserved with satisfactory outcomes.

When managing patients with acoustic neuromas, several options needed consideration for determining most optimal approach. Decisions regarding the appropriate management approach are typically based on several factors, like patient's age, tumor size, hearing preservation, and any coexisting conditions. Three common surgical approaches used for acoustic neuromas are middle cranial fossa, translabyrinthine, and retrosigmoid approaches. The choice of approach largely depends on the tumor size and whether hearing preservation is a priority. For larger acoustic neuromas, hearing

preservation is often not a significant consideration in selection of surgical approach since post-operative hearing is typically not serviceable. It was reported a lack of successful hearing preservation in patients with tumors that were 25mm or larger via the retrosigmoid approach 20-21.

Our findings are comparable to the research revealing that patients with cerebellopontine angle (CPA) tumors smaller than 1 cm typically have a facial nerve function rate of over 96% (HB grades I and II) after surgery. In contrast, tumors up to 1.5 cm in size have a facial nerve preservation rate of 83% (HB grades I and II), which drops to 70% for tumors over 2.5 cm and to 50% for tumors over 3.5 cm. Although the use of intraoperative monitoring and advancements in surgical tools have improved surgical outcomes over time, functional outcomes have only slightly improved in the last two decades. Therefore, for tumors up to 1.5 cm, observation alone may be the best option. If the tumors grow, low-morbidity surgery may be feasible, provided that it is done promptly before the tumor reaches the critical size of 1.5 cm. This strategy maximized the likelihood of preserving good facial nerve function, as evidenced by the surgical outcomes reported in analyzed series 13, 22. Surgical principle of "removing bone to spare brain" that guided trans petrous approaches to cerebellopontine angle substantially reduced the associated morbidity and mortality rates 13.

According to a study, the retrosigmoid approach resulted in an overall hearing preservation rate of 51%. Long-term serviceable hearing preservation rates range between 83% and 87% five years after tumor surgery using this approach. However, minor hearing loss worsens the prognosis, with 54% of patients losing class A hearing after ten years 19, 23. In another study involving 982 patients with a follow-up of between 26 and 52 months, the overall hearing preservation rate was found to be 54%. An average tumor growth rate of ≤ 2.5 mm/year was identified as a statistically significant factor for hearing preservation 24.

Despite significant advancements in radiation treatment for acoustic neuromas, surgery remains the primary solution in achieving a cure for the disease, long-term control of the tumor, and preservation of cranial nerve function over the long run 23.

CONCLUSION

Acoustic neuromas are rare, benign tumors that require a multidisciplinary approach to management. Surgical treatment is one of the mainstays of management, and several surgical approaches have been developed over the years. Preferred surgical approach relies on several elements, and goal of the surgery should be to minimize complications and maximize functional outcomes. The translabyrinthine approach involves the removal of large-sized tumors but with destruction of hearing and balance organs and severe complications. Middle fossa approach is used to manage small-sized tumors, with good functional capacity and outcomes, preserving the hearing and balance organs. Retrosigmoid approach is another surgical technique used for removal of acoustic neuromas of medium size and is less invasive than translabyrinthine approach and allows for preservation of facial nerve, as well as possibility of hearing preservation. Based on patient's age, morbidity associated with different surgical approaches, tumor size, and hearing function, some guiding principles can inform surgical decision-making. However, when the hearing function is lost or severely impaired, it cannot be restored.

CONFLICT OF INTEREST

None.

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