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Original Research Article

Study of Clinico-Radiological Features and Surgical Outcome in Acoustic Schwannoma

Pawan Choudhary[1], Vivek Yadav[2], Amit Dagar[3], K B Shankar[4], Puneet Garg[5], Dipanker Singh Mankotia[6]

[1]MCH Resident, Department of Neurosurgery, VMMC and Safdarjung Hospital, New Delhi
[2]Associate Professor, Department of Neurosurgery, VMMC and Safdarjang Hospital, New Delhi
[3]Associate Professor, Department of Neurosurgery, VMMC and Safdarjang Hospital, New Delhi
[4]HOD and Professor, Department of Neurosurgery, VMMC and Safdarjang Hospital, New Delhi
[5]Associate Professor, Department of Radiology, VMMC and Safdarjang Hospital, New Delhi
[6]Associate Professor, Department of Neurosurgery, VMMC and Safdarjang Hospital, New Delhi

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Corresponding Author: Dr. Vivek Yadav
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Abstract

Introduction: CP Angle tumors account for 5-10% of intracranial tumors. Most CPA tumors are benign, with over 85% being vestibular schwannomas, lipomas, vascular malformations and hemangiomas. The most frequent nonacoustic CPA tumors are meningiomas, epidermoids. Primary malignancies or metastatic lesions accounting less than 2% of neoplasm in the CPA.This study attempts to present the clinico-radiological profile as well as results of surgical management in acoustic schwannoma

Material and Methods: This study was conducted in the Department of Neurosurgery, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi. Patients having Contrast enhanced MRI brain with features suggestive of acoustic schwannoma were included in the study. We included 50 patients in this study.

Observation and Results: In our study 12% of patients were of the age group 0-20 yr, 28% were 21-30 yr, 26% were 31-40 yr, 16% were 41-50 yr, 8% were 51-60 yr, 10% were 61-70 yr. Females in our study were 44% whereas the males were 56%. In our study sensorineural hearing loss was observed in 84% cases, vertigo was present in 36% cases, headache was present in 90%, facial nerve dysfunction was present in 24%, tinnitus was present in 56%, motor weakness was present in 8%. Hydrocephalus was present in 28% cases. Our study depicted that majority tumours were large size i.e. 64% were Koos grade 4 and rest 36% were Koos grade 3. As the size increases, possibility of facial nerve palsy also increases post-operatively but the association was statistically significant (P value 0.03).

Conclusion: Post-operatively Facial nerve palsy along with lower cranial palsies were the most common complications. As the size of the lesion increased, possibility of these complications also increased. Thus, once diagnosed these tumours must be managed by multi-disciplinary team consisting of a Neurosurgeon, ENT specialist, Hearing and rehabilitation specialist and Radiation oncologist.

Introduction

CP Angle tumors account for 5-10% of intracranial tumors. Most CPA tumors are benign, with over 85% being vestibular schwannomas, lipomas, vascular malformations and hemangiomas. The most frequent nonacoustic CPA tumors are meningiomas, epidermoids. Primary malignancies or metastatic lesions accounting less than 2% of neoplasm in the CPA.

CPA S.O.L can be divided into extra-axial tumors, intra-axial tumors, extradural tumors and petrous apex lesions. Extra-axial tumors can be divided into those common and rare. Vestibular schwannomas are by far the most common extra-axial tumors. Other common extra-axial tumors include meningiomas and epidermoid cyst, arachnoid cyst. Rare extra-axial tumors include other cranial nerve neuromas (V, VII, IX, X, XI, XII) and vascular malformations (aneurysms, malformations). Intra-axial tumors include parenchymal lesions such as astrocytoma, ependymomas, papillomas, hemangioblastomas and metastases.

Extradural tumors include glomus tumors and bone lesions. Petrous apex lesions include cholesterol granulomas, epidermoid cysts, mucoceles and aneurysms of the carotid artery. These tumors may be asymptomatic or may present with a constellation of symptoms which could be due to tumor mass and involvement of the adjacent cranial

nerves, cerebellum and brainstem. Furthermore there could be associated signs and symptoms due to raised intracranial pressure (ICP) due to regional brainstem compression and hydrocephalus. The most common symptoms of are unilateral sensorineural hearing loss (96%), unsteadiness (77%), tinnitus (71%), headache (29%), mastoid pain or otalgia (28%), facial numbness (7%) and diplopia (7%).[1]

CT and MRI are widely used radiological methods for cerebellopontine angle imaging. The main radiological diagnostic goal is the description of the relation of the tumour to IAM, the brain stem and cerebellar hemispheres. The second line basic information is if the lesion is extra- or intracerebral.

The options available for management includes Observation, Surgery, Stereotactic Radiosurgery, Fractionated radiotherapy. Some patients might also be candidates for a combination of these therapies. The ideal treatment is total excision of tumor. Surgery is indicated for small, medium and large size tumors. Several approaches and techniques are available for the resection of CPA tumors, including translabyrinthine, retrosigmoid, suboccipital, retrolabyrinthine, transcochlear, transotic and middle fossa approaches. Surgery improves quality of life in patients but each approach is associated with post operative complications like mortality, haematoma, pneumocephalus, cranial neuropathies, cerebellar dysfunction, brainstem complications, infections, CSF leaks etc.

The Suboccipital retrosigmoid approach is a popular method of excising CPA tumors. Advantages of this approach include a low complication rate, particularly with regard to facial nerve function, and total tumor removal in the vast majority of cases. Moreover, the technique is safe and effective, even with the largest of tumors.[2] Technical advances in surgery and anaesthesiahas revolutionized the results of surgery in cerebellopontine angle lesions with reduced mortality and morbidity. Use of intra-operative nerve stimulators and intraoperative evoked potentials have made the resection of the lesions possible along with preservation of cranial nerves.

This study attempts to present the clinico-radiological profile as well as results of surgical management in acoustic schwannoma.

Material and Methods

This study was conducted in the Department of Neurosurgery, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi. All patients having Contrast enhanced MRI brain with features suggestive of acoustic schwannoma were included in the study. Except previously operated cases with recurrence.

Sample size

At 95% confidence level and taking the new post operative deficit rate as 35% in patients of acoustic schwannoma (Bartek J et al[3]) and with a relative error of 20%, the sample size estimated was 198 using the formula,

$n = Z\alpha 2\ p\ q\ /\ L2,$

Where n = sample size

$Z\alpha$ = 1.96 value of the standard normal variate corresponding to level of significance alpha 5%

p = new post operative deficit in 35% of patient = 0.35

q = 1 – p = 0.65

L = Relative error = 35*20%

We get only 50 patients, so using finite population formula,

$$= \frac{\text{Sample size}}{1+\frac{\text{Sample Size-1}}{\text{Population}}} = \frac{198}{1+\frac{198-1}{50}} = 40$$

So we included 50 patients in the study with acoustic schwannoma which was above the estimated minimum sample size.

Patients included in the study were evaluated and classified as per Koos [4] Classification of tumour extension.

Approval of the study was obtained from Ethical committee (medical).

Post-operative: patients were assessed for various surgical outcomes like

1. Development of new post operative deficit, if any
2. Cranial nerve palsy
3. Motor weakness
4. Post operative hematoma
5. Hydrocephalus
6. Prolonged ICU stay
7. Mortality

Radiological follow-up studies were performed by means of CT and/or MR imaging. Facial nerve function status was evaluated immediately post-operatively and at 6 months with the help of House-Brackmann scale.

Observation and Results

Table 1: Distribution of Cases according to Sex

Sex	Total Patients	Percentage
Male	28	56
Female	22	44

Above table shows that incidence is more common in males (56%) than females (44%). The Highest Percentage of cases are of the age group 21-30 yr (28%), followed by 31-40 yr (26%), 41-50 yr (16%), 0-20 yr (12%), 61-70 yr (10%) and 51-60 yr (8%).

Table 2: Distribution of cases according to Pre operative clinical presentation

Clinical findings	Total Patients	Percentage
Sensorineural hearing loss	42	84
Tinnitus	28	56
Vertigo	18	36
Headache	45	90
Facial nerve palsy	12	24
Motor weakness	4	8

Above table shows that incidence of headache was maximum ie. in 45 (90%) patients followed by sensorineural hearing loss in 42 (84%) patients, then tinnitus in 28 (56%), vertigo in 18 (36%), facial nerve palsy in 12 (24%) and motor weakness in 4 (8%) patients.

Table 3: Distribution of cases according to Pre operative tumour size as per Koos grading

Koos grade	Total Patients	Percentage
Grade 1	0	0
Grade 2	0	0
Grade 3	18	36
Grade 4	32	64

Above table shows the incidence of acoustic schwannoma as per size according to Koos grade. Highest incidence was of grade 4 size i.e. 32 (64 %) patients, followed by grade 3 with 18 (36%). There was no patient of grade 1 or 2. Hydrocephalus was present in 14 patients out of 50. Rest 36 patients did not develop hydrocephalus.

Table 4: Distribution of cases as per Pre operative Facial nerve palsy

Pre operative Facial palsy	Total Patients	Percentage
Grade 1	44	88
Grade 2	6	12
Grade 3	0	0
Grade 4	0	0
Grade 5	0	0
Grade 6	0	0

Above table shows the incidence of Pre operative Facial nerve palsy as per the House and Brackmann grading. Majority patients were grade 1 with no facial nerve involvement ie. 44 (88%) out of 50 patients followed by grade 2 palsy in 6 (12%). No patient in our study had Pre operative grade 3,4, 5 or 6 palsy. 26 (52%) out of 50 patients who had Koos grade 4 tumour had normal facial nerve status. 18 (36%) out of 50 patients who had Koos grade 3 tumour also had normal facial nerve status. There were 6 (12%) patients with Koos grade 4 tumour who had Grade 2 Facial nerve palsy.

Table 5: Distribution of cases according to size of tumour in Post operative Facial nerve involvement

Post op Facial palsy	Koos grade 1	Koosgrade 2	Koos grade 3	Koos grade 4
Grade 1	0	0	5	8
Grade 2	0	0	10	8
Grade 3	0	0	0	3
Grade 4	0	0	3	9
Grade 5	0	0	0	4
Grade 6	0	0	0	0

Above table shows distribution of cases with Post operative facial nerve involvement according to size of tumour. 8 (16%) out of 50 patients who had Koos grade 4 tumour had normal facial nerve status. 5 (10%) out of 50 patients who had Koos grade 3 tumour also had normal facial nerve status. There were 8 (16%) patients with Koos grade 4 tumour who had Grade 2 Facial nerve palsy. There were 10 (20%) patients with Koos grade 3 tumour who had Grade 2 Facial nerve palsy. There were 3 (6%) patients with Koos grade 4 who had Grade 3 Facial nerve palsy. There were 9 (18%) patients with Koos grade 4 who had Grade 4 Facial nerve palsy. There were 3 (6%) patients with Koos grade 3 who had Grade 4 Facial nerve palsy. There were 4 (8%) patients with Koos grade 4 who had Grade 5 Facial palsy.13 (26%) patients had normal post operative facial nerve. 18 (36%) patients had Grade 2 facial nerve palsy. 3 (6%) patients had grade 3 facial nerve palsy. 12 (24%) patients had grade 4 facial palsy. 4 (8%) patients had grade 5 facial nerve palsy.

Table 6: Distribution of cases as per Post operative complications

Post operative complication	Total Patients	Percentage
Hydrocephalus	3	6
Hematoma	8	16
Third cranial nerve palsy	1	2
Lower cranial nerve palsy	25	50
Prolonged ICU stay	13	26
Deaths	2	4

The above table shows distribution of cases as per post operative complications. 3 (6%) patients who did not have hydrocephalus Pre operatively developed hydrocephalus in the post operative period. 8 (16%) patients had hematoma in post operative period. Only 1 (2%) patient developed 3rd nerve palsy. 25 (50%) patients developed lower cranial nerve palsies. 13 (26%) patients had prolonged post operative ICU stay.

There was death of 2 (4%) patients in the post operative. There was 1 (2%) patient with Hydrocephalus, 4 (8%) patients with post operative hematoma, 2 (4%) patients with Lower cranial nerve palsy, 6 (12%) patients with prolonged ICU stay and 1 (2%) patient with post operative mortality who had Koos grade 3 tumour. Whereas there were 2 (4%) patients with Hydrocephalus, 4 (8%) patients with post operative hematoma, 23 (46%) patients with Lower cranial nerve palsy, 7 (14%) patients with prolonged ICU stay and 1 (2%) patient with post operative mortality who had Koos grade 4 tumour.

Statistical Analysis

Association of tumour size as per Koos grade with post operative Facial nerve palsy

			7 CN PALSY POST OP					
			0	2	3	4	5	Total
KOOS GRADE	3	Count	5	10	0	3	0	18
		% within KOOS GRADE	27.8%	55.6%	0.0%	16.7%	0.0%	100.0%
	4	Count	8	8	3	9	4	32
		% within KOOS GRADE	25.0%	25.0%	9.4%	28.1%	12.5%	100.0%
Total		Count	13	18	3	12	4	50
		% within KOOS GRADE	26.0%	36.0%	6.0%	24.0%	8.0%	100.0%

Figure 1 :

Fishers exact Test was done, P value was 0.03 which signifies there is a statistically significant association between size of tumour and post-operative facial nerve palsy. Association of tumour size as per Koos grading with post-operative lower cranial nerve paralysis

			LOWER CRANIAL NERVE PARALYSIS		
			0	1	Total
KOOS GRADE	3	Count	16	2	18
		% within KOOS GRADE	88.9%	11.1%	100.0%
	4	Count	9	23	32
		% within KOOS GRADE	28.1%	71.9%	100.0%
Total		Count	25	25	50
		% within KOOS GRADE	50.0%	50.0%	100.0%

Figure 2 :

Fishers exact Test was done, P value was less than 0.01 which signifies there is a statistically significant association between size of tumour and post-operative lower cranial nerve palsy.

CT/MRI Imaging of Cases

Case 1: 50 year old female with Koos grade 4 size vestibular schwannoma on right side. Patient underwent left side VP shunting pre-operatively followed by gross total excision of the lesion via right side retro-mastoid sub occipital approach.

Pre-Operative Scan

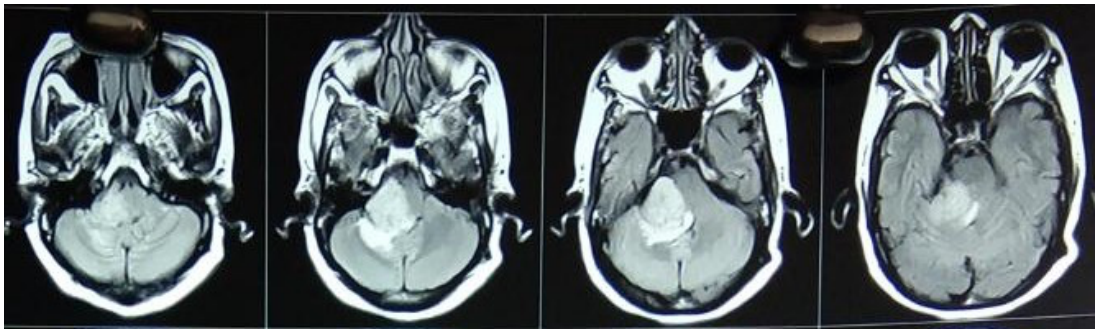

Figure 3 :

Post-Operative Scan

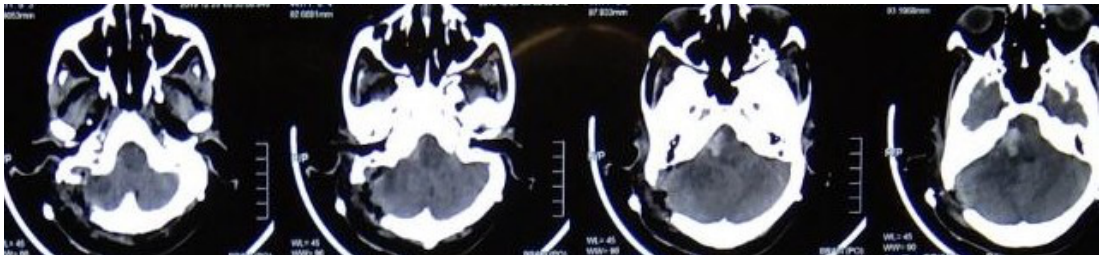

Figure 4:

Case 2: 50 year old female with Koos grade 4 size vestibular schwannoma on left side. Patient underwent gross excision of the lesion via left side retro-mastoid sub occipital approach.

Pre-Operative Scan

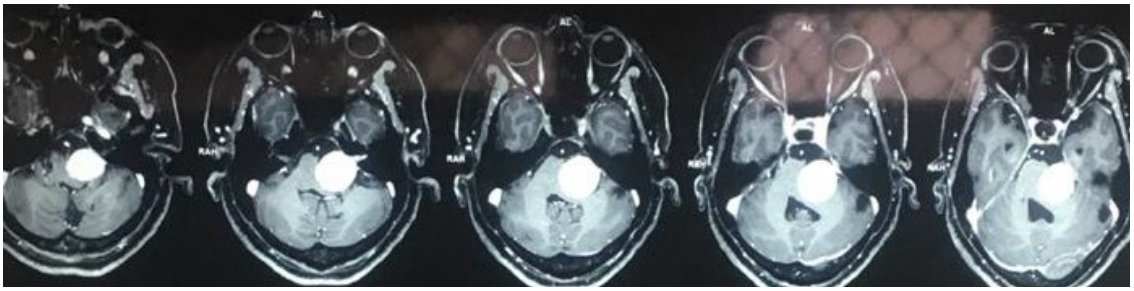

Figure 5:

Post-Operative Scan

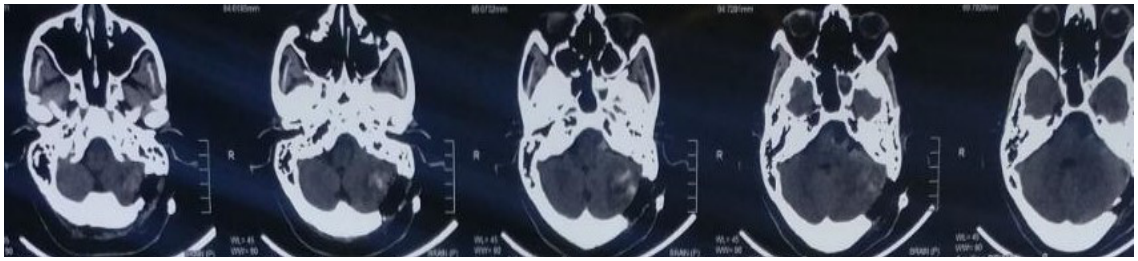

Figure 6:

Case 3: A 46y female with Right acoustic schwannoma underwent surgical excision of tumour and developed new onset grade 4 facial nerve palsy post operatively.

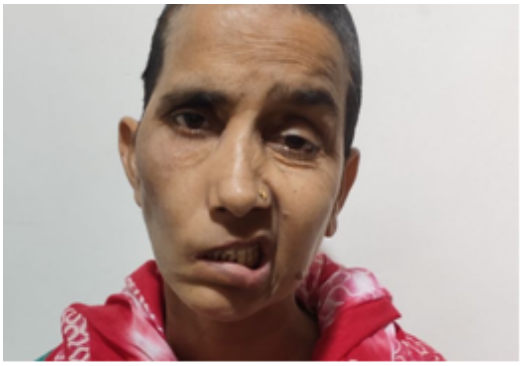

Figure 7:

Case 4: A 32 y female with Left Acoustic schwannoma was operated and surgical excision of the tumour was done. Patient developed new onset Third cranial nerve palsy in the post operative period.

Figure 8:

Case 5: A 49 y male with Left acoustic schwannoma underwent surgical excision of tumour and his facial nerve palsy grade deteriorated from grade 2 (Pre op) to grade 5 (post op)

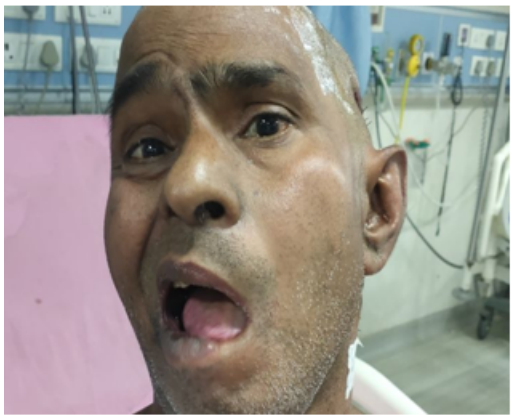

Figure 9:

Discussion

Tumors of the Cerebellopontine have always been difficult for neurosurgeons to manage optimally.

They are often closely involved with CNs and various blood vessels, posing great risk to surgical intervention. Factors such as tumor size and location, extent of resection, pathology and preoperative CN function have been implicated in long-term outcome. Studies investigating the natural history and treatment of vestibular schwannomas have dominated the literature in this regard. Tumors of the CPA have traditionally been associated with high rates of postoperative facial palsy and/or hearing loss because resection poses a particularly challenging endeavour.

CPA tumours, although uniform in location, are diverse pathologically and with regard to the site of tumour origin and displacement of the neurovascular structures. The main factor underscoring the importance of accurate preoperative diagnosis is the different surgical approach for vestibular schwannomas. There is general agreement that completeness of tumour resection and preservation of the facial nerve are the major neurosurgical goals. An appreciation of the vascular and cranial nerve microanatomy and the relationships between neurovascular structures and the tumour are essential for achieving optimal surgical results.

CT is a widely used method for lesions of the CPA, but MRI is also the imaging modality of choice for CPA and internal auditory canal masses. CT is fairly accurate in diagnosis of schwannomas bigger than 10–15 mm and schwannomas with intrameatal fraction because it can demonstrate enlargement of the acoustic canal as well as the mass itself. However, on CT axial images small intracanalicular schwannomas can be missed. CT and MRI should be used complementarily in CPA diagnostics and have a great impact on therapeutic planning.

A characteristic radiological feature of schwannomas is the presence of intrameatal fraction (“ice cream on cone” sign) and IAM widening, as well as contrast enhancement and the sharp angle between tumour and temporal bone pyramid.

Various approaches have been shown to be useful in resecting tumours in the CP angle region and have been well reviewed. Most of cases are exposed through a retrosigmoid approach. This has afforded good visualisation and allowed resection of the tumour without excessive retraction. In the patient with no prospect of useful postoperative hearing, a translabyrinthine approach may be appropriate especially for the tumours involving the internal auditory meatus although it must always be remembered that hearing may occasionally improve or even return to normal after the resection of cerebellopontine angle tumours and destructive approaches must therefore be used selectively.Vestibular schwannomas rarely recur after complete resection.

In our study 12% of patients were of the age group 0-20 yr, 28% were 21-30 yr, 26% were 31-40 yr, 16% were 41-50 yr, 8% were 51-60 yr, 10% were 61-70 yr. Females in our study were 44% whereas the males were 56%. FaramarzMemari, et al. (2015) [5] showed that the mean age was 49 years and there was a slight male predominance of 55%. Joarder MA, et al. (2015) [6] showed the maximum incidence between 30-50 yrs with female predominance of 55%. Maheswararao Y.V.N, et al. (2018) [7] showed that the highest incidence of extra axial cp angle tumours were found in 51-60 years age group with 70% being females.

In our study sensorineural hearing loss was observed in 84% cases, vertigo was present in 36% cases, headache was present in 90%, facial nerve dysfunction was present in 24%, tinnitus was present in 56%, motor weakness was present in 8%. Hydrocephalus was present in 28% cases. FaramarzMemari, et al. (2015) [5] showed that Forty-seven patients (94%) presented with tinnitus,

and vertigo was present in 30 patients (60%). Seven patients had cranial nerve 7 paresis preoperatively (maximum House Brackmann score, 4). Two patients had hemiplegia preoperatively. Joarder MA, et al. (2015) [6] showed that the most common presenting complaint was sensorineural hearing loss, cerebellar dysfunction and headache.

Our study depicted that majority tumours were large size ie. 64% were Koos grade 4 and rest 36% were Koos grade 3. Faramarz Memari, et al. (2015) [5] showed, tumours ranging from <15 mm to >35 mm. Joarder MA, et al. (2015) [6] showed medium size (10-25mm) tumors in 15%, large size (26-40mm) in 58%, giant (>40mm) in 27%.

Our study showed 12% patients had pre-operative grade 2 facial palsy, all of them were Koos grade 4 size tumours. Post-operatively there were 16% patients with Koos grade 4 tumour who had Grade 2 Facial nerve palsy. There were 20% patients with Koos grade 3 tumour who had Grade 2 Facial nerve palsy. There were 6% patients with Koos grade 4 who had Grade 3 Facial nerve palsy. There were 18% patients with Koos grade 4 who had Grade 4 Facial nerve palsy. There were 6% patients with Koos grade 3 who had Grade 4 Facial nerve palsy. There were 8% patients with Koos grade 4 who had Grade 5 Facial palsy. In our study it is observed that there is a positive relationship between size of tumor and incidence of post-operative facial nerve palsy. Post operatively 26% patients developed facial palsy in Koos grade 3 size tumors and 48% patients developed facialpalsy in Koos grade 4 tumors. Anatomical preservation of facial nerve was achieved for Koos grade 3 size lesions in 28% cases and for Koos grade 4 lesions in 25% cases. As the size increases, possibility of facial nerve palsy also increases post-operatively but the association was statistically significant (P value 0.03). Joarder MA, et al. (2015) [6] and Faramarz Memari, et al. (2015) [5] also depicted a significant correlation between tumor size and facial nerve outcome, with larger tumors yielding worse outcomes (P value 0.001). Gormley and Sekhar, et al. (1997) [8] study showed House-Brackmann evaluation of postoperative facial nerve function revealed excellent results (Grade I or II) in 96% of small tumors, 74% of medium tumors, and 38% of large tumors. Samii M, et al.[9] showed post operative grade 1&2 facial nerve function in 64% cases, grade 3&4 in 21% cases and grade 5&6 in 15% cases.

We observed hematoma in 16% cases and lower cranial nerves palsy was seen in 50% cases out of which 46 % were Koos grade 4 tumours and they were managed with nasogastric tube feeding. This also revealed a higher post operative lower cranial nerve palsy rate in large size tumours (P value less than 0.01). Third cranial nerve palsy was seen in 2% patients. Vijendra K. Jain, et al. (2005) [10] in their study on VS had the incidence of lower cranial nerve paresis of 6.8%. Sourabh Dixit, et al (2017) [11] in their study on VS had transient lower cranial nerve paresis in 46.15% patients which gradually improved. The reported incidence of lower cranial nerve paresis in the rest of literature ranges from 1.5% to 5.5% [12]. In our study 3 patients (6%) developed hydrocephalus post operatively. As quoted by F Pirouzmand, CH Tator, J Rutka et al [13] 3% to 15% patients developed hydrocephalus. In our study overall prolonged ICU stay post operatively was seen in 26% cases and mortality was seen in 4% cases mainly. Faramarz Memari, et al. (2015) [5] in their study on CP angle lesions, had mortality of 2% for retro-sigmoid approach. Gormley and Sekhar, et al. (1997) [8] had 1% mortality in their case series of VS. Vijendra K. Jain, et al. (2005) [10] have observed mortality of 6% in their series of VS. Sami M, et al.[9] reported no mortality in their series of VS.

Conclusion

Acoustic schwannoma was common in middle age group, with incidence in males slightly more than females. Headache and sensorineural hearing loss were the most common complaints. Majority of lesions were of large size (Koos grade 4). Post-operatively Facial nervepalsy along with lower cranial palsies were the most common complications and there was significant association between the size of lesion and increased post operative Facial and Lower cranial nerve palsy. As the size of the lesion increased, possibility of these complications also increased.

Prolonged ICU stay seen in 26% cases was a major concern. Hearing loss and facial paralysis may improve over time for many individuals. Along with facial nerve and other lower cranial palsies it significantly added to the post operative morbidity. These post operative complications along with residual hearing loss also had a significant impact on quality of life. Overall mortality was in 4% cases only. Thus, once diagnosed these tumours must be managed by multi-disciplinary team consisting of a Neurosurgeon, ENT specialist, Hearing and rehabilitation specialist and Radiation oncologist.

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