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Original Article

Cerebellopontine Angle Tumors, Presentation, Treatment and Outcome: Experience at a Tertiary Care Institution

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***ABSTRACT*:**
Aims and objectives: To study tumors of cerebellopontine angle with respect to age, sex, clinical presentation, radiological and pathological appearance, management and complications related to tumor excision. **Material and methods:** This was a prospective study carried out in the department of neurosurgery, Institute of Medical Sciences, BHU with clinic radiologic diagnosis of SOL in cerebellopontine angle on 38 patients. Various parameters were analyzed. **Results:** 15 (39.5%) were male and 23 (60.5%) were female. Average age of presentation was 37.95 and standard deviation was 14.22.Eight patients were having tumors less than or equal to 3cm size while 30 patients were having large tumors that is more than 3cm size.Hearing loss was the commonest symptom present in 78.95% patients followed by tinnitus and disequilibrium in 65.8%. Disequilibrium, headache, facial symptoms and lower cranial nerve symptoms were seen only in larger tumors. Acoustic schwannoma was the commonest (76.3%), followed by meningioma (13.16%) and epidermoid (7.9%).Based on MRI we found solid lesion in 8 cases, 26 patients had mixed echogenic tumors and cystic appearance was seen in 4 patients. In ten patients ventriculoperitoneal shunt was placed first then excision of tumor was done and 28 patients underwent excision directly. Facial nerve dysfunction was the commonest postoperative complication followed by trigeminal nerve dysfunction and lower cranial nerve involvement. Eight patients had residual tumor and 2 patients expired. **Conclusion:** These tumors usually present when becomes large and there is no significant sex predominance. Smaller tumors are usually solid and larger are mixed echogenic.
Key words: Cerebellopontine Angle Tumors, schwannoma.

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INTRODUCTION

The cerebellopontine (CP) angle is bound anterolaterally by the posterior aspect of the petrous temporal bone and posteromedially by the cerebellum and pons. It contains important vascular structures and cranial nerves and is subject to a certain gamut of lesions, notably tumors with interesting radiological manifestations. Lesions of the CP angles usually are divided into those native to the angle (vestibular schwannoma, meningioma, epidermoid, arachnoid cyst, metastases, lipoma, etc.) and those extending to the angle from adjacent structures (gliomas, ependymomas, choroid plexus papillomas, vascular malformations)(1).

The Acoustic schwannoma is the most common tumor ofthe cerebellopontine angle (CPA) (80–90%) followed by Meningioma (5–10%), Ectodermal inclusion tumours, Epidermoid and Dermoid (5–7%). Other tumors of the CPA include primary cholesteatoma, and other cranial nerve schwannoma (V, VII, IX, X), arachnoid cysts etc. These tumors may be asymptomatic or may present with a constellation ofsymptoms 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%) (2)The options available for management includes

Observation, Surgery, Stereotactic Radiosurgery, Fractionated radiotherapy. Some patients might also be candidates for a combination ofthese therapies (3). The ideal treatment is total excision of tumor. Surgery is indicated for small, medium and large size tumors.(4) The Suboccipital retrosigmoid approach has become the most popular method ofexcising CPA tumors.(5)

AIMS AND OBJECTIVES

To study tumors of cp angle with respect to age, sex, clinical presentation, radiological and pathological appearance, management and complications related to tumor excision.

MATERIAL AND METHODS

This was a prospective study carried out in the department of neurosurgery, Institute of Medical Sciences, BHU with clinicoradiologic diagnosis of SOL in cerebellopontine angle. It includes 38patients who were admitted from May 2016 to January 2018. Every patient underwent MRI and CT scan and then were operated.

The various parameters that were used in the study were size oftumor, preoperative neurological deficits, extent of cranial nerve involvements, functional status of patients in preoperative and postoperative periods, presence ofsigns ofraised ICP etc. and postoperative complications were recorded in immediate and delayed post-operative periods. The management was done either by surgical excision directly by RMSO craniotomy or ventriculoperitoneal shunting followed by excision by RMSO craniotomy.

Facial nerves functioning in post-operative periods were measured by House Brackmann Facial nerve Grading system (6). A post-operative follow up of the patient was done minimum for 1 month. Post-operative lower cranial nerve paresis was assessed by presence or absence of Gag reflex and deglutination difficulty. The data ofrelevant information was collected and tabulated and systematically analysed.

RESULTS

Total 38 patients were included in the study. 15 (39.5%) were male and 23 (60.5%) were female. Youngest was 5 years and eldest was 60 years old. Age group wise distribution of patients is shown in table 1. Patients less than 20 years of age were only 4 and all were male. Young patients and middle aged patients were almost equal in numbers but most of them were females. Average age of presentation was 37.95 and standard deviation was 14.22.

Table 1: Sex distribution of CP angle tumors in different age groups

Age group	Males	Females
0-20 years	4	0
21-40 years	4	12
41-60 years	7	11

Sixteen patients had tumor in right CP angle and 22 in left CP angle. Side wise distribution is shown in Table 2. Younger patients had tumors mostly of left side.

Table 2: Side wise distribution of CP angle tumors in different age groups

Age group	Right side	Left side
≤20 years	2	2
21-40 years	5	11
40-60 years	9	9

On the basis of MRI tumors were grouped in two categories (table 3). Group A included tumors less than and equal to 3cm size and group B with more than 3cm. Eight patients were having tumors less than or equal to 3cm size while 30 patients were having large tumors that is more than 3cm size.

Table 3: Sex and size wise distribution of CP angle tumors

Size of tumor	Male	Female
≤3cm	3	5
>3cm	12	18

Patients presented with various symptoms (table 4). Hearing loss was the commonest symptom present in 78.95% patients followed by tinnitus and disequilibrium in 65.8%. Disequilibrium was seen only in larger tumors. Similarly headache was asymptom of larger tumor and was present in 52.6% patients. Other symptoms like facial numbness, facial weakness, diplopia, lower cranial nerve symptoms were seen only in larger tumors.

Table 4: Various clinical presentations of CP angle tumors

Clinical presentation	≤3cm size tumor (N=8)	>3cm size tumor (N=30)	p-value
Hearing loss	5	25	0.1994
Tinnitus	5	20	0.8523
Disequilibrium	0	25	0.001
Headache	2	18	0.0781
Facial numbness	0	10	0.1271
Facial weakness	0	10	0.1271
Hoarseness of voice	0	6	0.4302
Dysphagia	0	6	0.4302
Diplopia	0	5	0.5678

Every patient underwent MRI brain as well as CT scan brain to see for exact location of tumor, size, radiological appearance, hydrocephalus and acoustic canal width. Based on radiology we found four types of tumor which was further confirmed in histopathology after excision. This is shown in table 5. Acoustic schwannoma was the commonest (76.3%), followed by meningioma (13.16%) and epidermoid (7.9%). There was a single case of large arachnoid cyst also.

Table 5: Histological diagnosis of CP angle tumors

Diagnosis	≤3cm size tumor(N=8)	>3cm size tumor (N=30)	p-value
Acoustic schwannoma	7	22	0.4024
Meningioma	1	4	0.9506
Epidermoid cyst	0	3	0.9625
Arachnoid cyst	0	1	0.999

Based on MRI we found solid lesion in 8 cases out of which 4 were meningioma and 4 were acoustic schwannoma. 26 patients had mixed echogenic tumors, mostly larger tumors. Cystic appearance was seen in 1 patient of arachnoid cyst and 3 of epidermoid cyst. All cystic lesions were large that is more than 3cm size.This is shown in table 6

Table 6: Radiological appearance of CP angle tumor based on MRI

Radiological appearance	≤3cm size tumor (N=8)	>3cm size tumor (N=30)	p-value
Solid	4	4	0.023
Mixed	4	22	0.2075
Cystic	0	4	0.7425

In ten patients ventriculoperitoneal shunt was placed first then excision of tumor was done. These were cases which included all the large tumors with associated hydrocephalus. 28 patients underwent excision directly. This is shown in table 7. Excision of tumor was done by RMSO craniotomy in all the cases.

Table 7: Surgical management of CP angle tumors

Surgical management	≤3cm size tumor	>3cm size tumor
VP shunt followed by RMSO and excision	0	10
RMSO and excision	8	20

We encountered several complications following surgery as shown in table 8. The commonest being facial nerve dysfunction which was seen in 14(36.8%) patients, mostly larger tumors. Transient trigeminal nerve dysfunction was seen in 5(13.2%) patients and lower cranial nerve symptom developed in 6(15.8%) patients. However these symptoms recovered up to an extent after 2-3 weeks in some patients. In 8 patients we had to leave part of tumor unresected due to various reasons like proximity with brain stem and lower cranial nerves, all were large tumors. Our 2 patients expired due to respiratory complications in postoperative period.

Table 8: Outcome/Complication following surgery

Outcome /Complication following surgery	≤3cm size tumor	>3cm size tumor
New facial nerve dysfunction	2	12
New trigeminal nerve dysfunction	0	5
New lower cranial nerve involvement	0	6
Residual tumor	0	8
Mortality	0	2

DISCUSSION

We had a total of 38 patients out of which 15 were male and 23 female. Patients less than 20 years of age were only 4 and all were male while more than 20 year aged patients were 34 and the female population in this age outnumbered the male. In a similar study done on acoustic schwannoma by Sourabh et al had maximum number ofcases i.e. 38.46% in31 to 40 years ofage.(7) The average age ofpresentation in our study was 37.95 with standard deviation 14.22. In Saurabh et al study mean age was 37.23 years and standard deviation was 11.41. Memari F et al had mean age of49 years in their series.(2)There was a female predominance in our study similar to Saurabh et al.(7) But in series ofMemari F et al(2) there was male predominance.

We had 16 patients of tumor in right CP angle and 22 of left CP angle. P-value regarding side of tumor was not significant and so far no study is available in literature regarding side predominance as well.

We had tumors larger than 3cm more common in our study that is total 30 (78.95%) and tumors smaller than 3cm were only 8 (21.05%). Saurabh et al(7) had maximum number of case between 2 cm-3.9 cm i.e 53.84% with average size 3.21 cm. However Memari F et al(2) observed the mean tumor size 24 mm, ranging from <15 mm to >35 mm. We encounter larger tumors more because of ignorance of health and illiteracy in this part of world. Mostly patients reach to us only after their symptoms start affecting their routine life. The most common symptom in our patients was ipsilateral sensorineural hearing loss (SNHL) which was present in 30 (78.95%) cases including both small and large tumors. Tinnitus was the second most common symptom with 25 (65.79%) patients whether small or large tumors. According to saurabh et al(7) SNHL and tinnitus was present in all patients (100%). The difference is because we included all types of tumors in CP angle and most of our cases of meningioma, epidermoid and arachnoid cyst did not had hearing loss or tinnitus as their complaint. Disequilibrium was present in 25 (65.79%) patients and all were larger tumors. In no patient with less than 3cm tumor disequilibrium was present and this difference was statistically significant. Disequilibrium is a cerebellar sign so is present only once significant cerebellar compression occurs so it is present in larger tumors only. Epidermoids are slow growing tumors and also they compress the adjacent structures very late so patients with epidermoids remain free from cerebellar or brain stem signs. Cerebellar signs were present in 76.92%patients. Headache was present in 20 (52.63%) patients of which mostly were males. In the study by Saurabh et al(7) headache was present in all cases. The difference is because we have included cases of epidermoid and arachnoid cyst which usually are painless. Facial numbness and facial weakness was there in 10(26.3%) patients all of which were more than 3cm size. Facial symptoms were seen in all epidermoid patients. Saurabh et al(7) had some different observation, they hadfacial paresis in 76.92% trigeminalinvolvement in 53.84% patients. Lower cranial nerve involvement was present in 38.46% patients in saurabh et al(7) study while in ours 6 (15.79%) patients had lower cranial nerve symptoms. Symptoms like facial numbness, facial weakness, diplopia, hoarseness of voice and dysphagia was present only in tumors more than 3cm size.

Memari F et al(2) had observed that Forty-seven patients (94%) presented with tinnitus, and vertigo was present in 30 patients (60%). M. Javad Mirzayan et al(8) had also obtained similar findings with Hypacusis/Deafness 70% (12) Tinnitus 30% (6)Vertigo 15% (3) Cerebellar symptoms 10% (2) Facial palsy 10% (2)Trigeminal hypesthesia 5% (1) Symptoms ofincreased ICP 15% (3).

Based on MRI we had 8 (21.05%)solid tumors, 26 (68.42%) mixed echogenic tumor and 4 cases (10.53%) cystic tumors. Percentage of smaller tumors being solid was significantly higher in number than larger tumors.

In total we had 29 (76.31%) patients of acoustic schwannoma, 5(13.16%) of cp angle meningioma, 3(7.9%) epidermoid and single case of arachnoid cyst of cp angle confirmed histopathologically. Acoustic neuromas remain by far the most prevalent lesion to be found in the cerebellopontine angle.(9) Meningiomas are the second most common tumour to be found in this site, accounting for exactly 6.5% of all cases in two separate British series.(10,11) In the present series in 10 (26.31%) patients ventriculoperitoneal shunt was placed first then excision of tumor was done. These were cases where tumor wasmore than 3cm in size and were associated with hydrocephalus. 28 (73.68%) patients underwent excision directly that is no preoperative shunting was done. Most of these patients had not hydrocephalus. In few even if associated with hydrocephalus ventricular tapping through safety burr hole was done during the time of excision. In all the cases excision of tumor was done by RMSO craniotomy.

The retrosigmoid approach has been accepted as a popular approach for excision of small, medium and large size CP angle tumors. Merari F et al(2)in their series had operated 28% cases by retrosigmiod approach and had low complication rates. Jain Vk et al(12) had all the cases operated by retro-sigmoid approach and concluded that the retro-sigmoid approach in experienced hands is a good option; with good results compared to other series irrespective ofthe tumor size.

In the study by Saurabh et al(7) all patients underwent preoperative ventriculoperitoneal shunting to relieve the raised ICP Followed by excision of tumour through retrosigmoid approach. Jain VK et al(12) also advocated preoperative shunting in patients presenting with features ofraised ICP. The incidence ofpreoperative shunting was as high as 66% in the series reported by Ramamurthy (13).

According to Pirouzmand F et al(14) excision of cp angle tumors can be performed without permanent CSF shunting even if hydrocephalus is present.

The commonest complication after tumor excisionis facial nerve dysfunction which was seen in 14(36.8%) patients, mostly larger tumors. Merari F et al(2)had reported Thirty-two patients (64%) had a Grade 1 or 2 score at 1 year, while 26% had a score of3 or 4, and 8% had a score of5 or 6. They also found that there was a significant correlation between tumor size and facial nerve outcome, with larger tumors yielding worse outcomes. Gormley and Sekhar et al(15)showed House-Brackmann evaluation of postoperative facial nerve function revealed excellent results (Grade I or II) in 96% ofsmall tumors, 74% of medium tumors, and 38% oflarge tumors. A fair postoperative function (Grade III or IV) was achieved in 4% of small tumors, 26% ofmedium tumors, and 58% of large tumors. Samii et al(4)intheir series of1000 cases showed that the facial nerve was anatomically preserved in 929 cases (93%). In 200 cases oftumor resections using the CPA pproach, preservation rates rose to 94%, independent oftumor size. Facial nerve function graded according to the House-Brackmann scale within 2 weeks 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% ofthe patients.Jain V k et al(12)in there series showed Facial nerve was anatomically preserved in 79.2% (198/250) patients with complete tumor excision.

Transient trigeminal nerve dysfunction was seen in 5(13.2%) patients and lower cranial nerve symptom developed in 6(15.8%) patients. However these symptoms recovered upto an extent after 2-3 weeks in some patients.Saurabh et al(7) had transient lower cranial nerve paresis in 46.15% which gradually improved. Jain V k et al(12)had the incidence oflower cranial nerve paresis6.8% in their study. In 8 patients we had to leave part of tumor unresected due to various reasons like proximity with brain stem and lower cranial nerves, all were large tumors. Residual tumour was present in15.38% patients in saurabh et al(7) series. Merari F et al(2) hadresidual tumor in 7% for retrosigmoid approach Gormley and Sekhar et al(15)reported Complete tumor resection was accomplished in 99% of the patients, and there was no evidence of recurrence in this group. Only 1 of the 179 patients underwent incomplete tumor resection; he required subsequent surgery for symptomatic tumor regrowth. Jain V k et al(12) had achieved complete tumor excision in 96.5%.

Our 2 patients expired due to respiratory complications in postoperative period. Merari F et al(2)had mortality of2 % for retrosigmoid approach. Gormley and Sekhar et al(15) had 1% mortality in their case series. Jain V k et al(12)have observed mortality 6% in their series. Samii M et al(16) reported no mortality in their series.

CONCLUSION

Acoustic schwannoma is the commonest tumor of cerebellopontine angle. Tumors of CP angle usually present in young and middle aged persons. These tumors usually present when becomes large and there is no significant sex predominance. Hearing loss, tinnitus, disequilibrium and headache are common symptoms. Facial numbness, weakness, diplopia and lower cranial nerve symptomsare present only in large tumors. Smaller tumors are usually solid and larger are mixed echogenic. VP shunt can be done preoperatively if associated with hydrocephalus or otherwise directly tumor excision can be done by RMSO craniotomy. Facial nerve dysfunction is the commonest complication of surgery.

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