Occurrence of Lower Cranial Nerve Impairment in Large CPA tumors

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Abstract:

Introduction: *CPA tumors are customarily divided into small (<3cm) and large (>3cm). CP Angle region has been divided into 3 different compartments-upper, middle, and lower based on their association with neurovascular structures and the Subarachnoid cisterns. The lower cranial nerves (7-12) are grouped into 3 groups-group A,(7,8), B(9-11) & C(12) based on their commonality in origin, pathway and similarity in functions, dysfunction and affections. Lower cranial nerve involvement means clinical, &/or radiological &/or microsurgical affections of the lower cranial nerves (7-12).*

Methods: *In this purposive retro-prospective study 66 CPA tumors were included of which 65 were large in size. Out of 65 patients with large CPA tumors (>3cm), 57 tumors were analyzed for the location and the lower cranial nerves. Anova, was implied for the test of significance.*

Findings and results: *Two cases in upper, 15 in middle compartments were encountered whereas rest 40 cases were multicompartmental. There was not even single case, which was solely located in the lower compartment. Of the 40 multicompartmental tumors 22 cases were in upper and middle (UM) compartment, 4 in middle and lower (ML) compartment and 14 in all three (UML) compartments. On the whole there were 37 tumors located in upper, 55 in middle and 19 in lower compartment. Of 57 tumors analyzed for the location, only 2 cases were situated in the upper compartment alone, 15 cases in the middle compartment only. Rest of the tumors (14 cases) was multicompartmental. Twenty-two patients had tumors in the upper and middle compartment (UM), 4 in middle and lower (ML) compartment and 14 were in all three (UML) compartments. On the whole, 37(33.3%) tumors occupied upper, 55(49.5%) middle, and 19(17.1%) occupied the lower compartment. Analysis of location revealed that if all these tumors were located in lower compartment there would be 2/3rd chance that group B/C LCN would be involved-the local/neighborhood effect. Similarly if all of them were in upper compartment, nearly 1/3rd chance of group B/C LCN involvement-the distant effect.*

Conclusion*: With the present definition, group A lower cranial nerve involvement was found to be 92%, group B- 46%, and group C in 7%.*

Key words: *Occurrence of LCNI, Large CPA tumors, CPA compartments*

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Introduction:

There has been a lot of discussion and the work in the field of the CPA tumor, their behavior and characteristics. Most of the studies either involved or characteristically determined the role of the sophisticated equipment in the diagnosis and in the prognostic anticipation of CPA tumors.

In the developing countries like ours these are not only not affordable but also not available. In this context, if there could be some clinical work to determine the state of the impact of the tumor; and have a prognostic guideline, to save time of the surgeon and resources of the patients and their families; which are not covered by any insurance companies here, would be a good work. The patients usually present here at late stages and in fact we do get mostly the large and giant CPA tumors only. Tumours exclusively located in cistern called “cisternal tumors” [1] are almost not seen.

The tumors in CPA include a heterogeneous group and are classified into small (1.5cm), medium (1.5-3cm) and large (>3 cm) depending on their size [2]. But the tumors more than 4 cm can be classified Giant or huge tumors as the challenge is different in the management of these tumors[3].

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The controversy of the CPA tumor starts from the nomenclature. The so-called acoustic neuroma which constitutes almost about 80% of tumors, is in fact inferior vestibular nerve in origin in about 90% of the times, hardly if ever, from the cochlear branch, and from the Schwann cells not from the axon to qualify it's name, even though it can arise from any cranial nerves (CN) except CN I and II. There are three common theories about the origin of this tumor thus their names. The Nerve fiber theory-neuroma; the Fibroblastic theory- Fibroblastoma and the Schwann cell theory-the Schwannoma the latter is mostly accepted in the recent literatures. There was a belief that this tumor arises from near the brain stem and spreads laterally into the IA Canal[4]. In fact these tumors arise from the Obersteiner's zone, which lies near about the internal acoustic meatus most of the time. Intra-canalicular intra-neuronal proliferation of the tumor is called the Schwannosis and multiple schwannomas at a time is often called Schwannomatosis, in the recent literatures. The Consensus Development Conference of 1991 has resolved the controversy about the origin of CPA Shcwannomas[5].

The site of origin is the main factor in making a preoperative diagnosis for an unusual lesion of the CPA [6]. In addition, it is essential to analyze attenuation at computed tomography (CT), signal intensity at magnetic resonance (MR) imaging, enhancement, shape and margins, extent, mass effect, and adjacent bone reaction[6]. CPA masses can primarily arise in the cerebellopontine cistern and other CPA structures (arachnoid cyst, nonacoustic schwannoma, aneurysm, melanoma, miscellaneous meningeal lesions) or from embryologic remnants (epidermoid cyst, dermoid cyst, lipoma[7]. Tumors can also invade the CPA by extension from the petrous bone or skull base. Root entry zone primary CPA glioma is on record[8] as are bilateral arachnoid cysts [9].

Tumors of the cerebellopontine angle (CPA) are frequent; neuromas and meningiomas represent the great majority of such tumors. However, a large variety of unusual lesions also encounter in the CPA (Cholesterol granuloma, paraganglioma, chondromatous tumors, chordoma, endolymphatic sac tumor, pituitary adenoma, apex petrositis). Finally, CPA lesions can be secondary to an exophytic brainstem tumors[10] or ventricular tumor (choroid plexus papilloma[11], lymphoma, hemangioblastoma, ependymoma, medulloblastoma, dysembryoplastic neuroepithelial tumor)[12]. A close association between CT and MR imaging findings is very helpful in establishing the preoperative diagnosis of unusual lesions in CPA[4]. While an acoustic neuroma is the most common cause of a cerebellopontine angle (CPA) mass, it accounts for only 1-10% of cases of sensorineural hearing loss (SNHL)[13]. Gradually progressive unilateral SNHL is not a *sine qua none* of CPA tumors, as some common and other very unusual lesions like metastatic brochogenic carcinoma can present with SNHL, at the other end sudden hearing loss can be a feature of CPA tumors in about 3% of cases[14,15].

The other common tumors in this region are meningioma (10%) and epidermoid (7%). Meningiomas rarely can involve the endolymphatic sac[16]. Unlike a schwannoma Meningioma on the other hand involves the VIIth nerve earlier than the VIIIth, bony hypertrophy and calcification, and both meningioma and epidermoid may present with the tic douloureux like symptoms more often than acoustic [17]. The most common presenting symptoms of CPA meningiomas are hearing loss, tinnitus, dizziness and dysequilibrium[18]. It cannot be easily distinquished from an AN only on the history and physical examination. Even with an audiogram, evoked response audiometry (AER) and vestibular function tests, it still cannot be distinquished. CT and MRI are helpful in differentiating these two tumors radiographically[19] more so when neuromas (4th & 6th cranial nerves) with meningiomas occur simultaneusly[20].

In general these tumors even after the treatment in case of large sizes may leave permanent sequelae particularly the ataxia from the brain stem or the cerebellar compression. Again the involvement of the IX, X, XI cranial nerves collectively causing swallowing difficulties may tax the life of an individual by aspiration. Similar damage may persist after the iatrogenic damage to the stem during the surgery.

The 8th cranial nerve is disproportionately frequent followed by 5th nerve at distance; others being rare cranial nerves to be involved by these tumors. There are at least two cases of Intra-labyrinthine Schwannomas reported.

Similarly the controversy lies in naming the lower cranial nerves. Some people believe that the lowermost cranial nerves namely the IX, X, XI and XII are the lower cranial nerves. But recently Professor H. Ric Harnsberger in his book called Handbook of head and neck imaging states that because of the similarity the lower six cranial nerves should be divided in to three groups[21]. Group A, the 7-8complex, for their similar path, common affections and better understanding, taken together. Group B- IX, X, XI complex again having similar path and common affection and similar approach of visualizing them and group C, the isolated XII; should resolve the issue reasonably. For this study this is the definition based on.

The anteriorly positioned cranial nerves like CN XII are preserved even with compartmental tumors regardless of the size and situation of tumors. But the sensory nerves like Vth Cranial nerve dysfunction occurred even when tumor remained away and untouched (surgical confirmation). It may be due to the dragging effect of large tumors falling from IAM or pressure on the trigeminal tract or nucleus on the stem. Whereas motor nerves like Facial function is relatively preserved even when the tumor is large and compressing the nerve directly, and may be due to the presence of the thick layer of myelin.

Involvement of the upper cranial nerve (I-6) has been correlated with the raised ICP or retraction effect due to gravitational fall of the tumor with consequent descent or distortion of the stem.

It is found that the invasion of the nerves, which varies with histological nature, is also responsible for the LCN involvement in addition to the direct sudden uncompensated pressure. So far as the consistency of the tumor is concerned; softer the tumor more likely that the nerves is involved making it difficult to remove in toto. Early symptomatic tumors seem to be fast growing and quickly decompensate or are of soft variety and vice versa.

Method:

The study was designed as correlational retro-prospective and the study period was 3.5 years(Jan 1998-June 2001). A total of 66 (42 prospective and 24 retrospective) consecutive patients (47 in Neurosurgery Department of BSMMU and 19 in addition diagnosed in Popular Diagnostic Centre) were included in the study with their consent. All of these prospective cases 41, by definition, were large or giant tumors .

Patients who deny or are with recurrent tumors were excluded.

(Two recurrent cases and one Giant basilar aneurysm presenting with CPA syndrome were excluded.)

Data Analysis:

The Analysis was made using multivariate analysis (Anova Test) and the conclusion drawn based on the F value at the 5% level of significance. The composite picture of the multicompartmental involvement is settled using modified Leikert's principle.

Results:

The demographic profile of the study group revealed 2/3rd male and 1/3 female patient, mostly of, both socially and economically productive age group (30-50years), suffered from CPA tumors. They were 12.69 % left handed and the tumor was located on the right side most of the time (53.12%). The earlier occurrence of CPA tumors by one decade in SAARC region has been verified in this study also.

Clinical presentation is one of the ways gauzing of involvement of LCN [table 4] and size of tumor is another [table 3].. One in six patients had associated medical conditions. Most common clinical presentations were headache (94.54%), hearing loss (85.45%), vertigo or imbalance (67.27%) and visual troubles (67.27%). Signs of raised Intracranial pressure was found in about half of patients and evidence of lowermost cranial nerves (9-12) involvement in 1/3rd . Only less than one third of patients (27%) presented with typical CPA syndrome.

With the present definition, group A. lower cranial nerve involvement was found to be 92%, group B- 46%, and group C in 7%.

About the size of the tumor, diameter and volume both were not significantly different in LCN involvement than those occurring by chance ($p>0.05$) (tables 1 and 2).

The most common presenting (average duration is 1.3years) symptoms (n=55) were Headache (94.54%) and Hearing loss of varying grade (85.45%). Vertigo or imbalance was present in 67.27% cases but in most instances were the responses rather than the complaint. Strikingly visual symptoms were the reason for consultation in most of the instances (67.27%). Vomiting was found in 54.54% of the times and difficulties in deglutition or voice change were

complained of in 29.09% cases. Tinnitus was found only in 27.27% cases and it was the complaint mostly in lower diameter tumors.

Many patients were blind at the time of admission and nealy 3/4th patients had visual problems and 60% had papilledema at presentation .

73.58% had dysfunction of the trigeminal nerve though there were only 16.36% complaints and 69.09% cases had clinical evidence of Facial Nerve dysfunction of which only 1 in 4 complained off .

Unlike other studies the dysfunction of group B of the lower cranial nerve (IX, X, XI) were clinically found impaired in 34.54% cases which is a bit higher than Ramamurthi's series [22]. The more anterriorly located nerves in the CPA, 6th and 12th cranial nerves, were least impaired in this series (9% and 7% respectively).

The commonest tumor in CPA remained the Schwannoma (76%) followed by Meningioma (13.3%) and Epidermoid (4.44%). Two unsual forms Ependymoma one-case and Hemangiopericytoma 2 cases and 2 cases of Neurofibroma (type 2) were reported histologically. On the whole (n=65) Schwannoma/NF represent 70.76% and Meningioma 15.38%.

The tumors were analyzed on the basis of their location on the upper, middle or lower CPA compartments respectively. Naturally most often the tumor remained in the middle compartment as most of the tumor originates in this compartment in relation to the lower cranial nerves. The following tables will illuminate the observations.

Table-I

Location of tumor and the cranial nerve relationship (n=111 combinations)

Compartment	Group A Involved	Group B Involved	Group C Involved
Upper (n=57)	36	18	3
Middle(n=78)	52	22	4
Lower (n=31)	19	11	1
Total (n=111)	107	51	8
Percent	92%	46%	7%

Table-II

Histology of large CPA tumor and Cranial Nerve involvement (n=65)

Histo		Group A		Group B	Group C
		VII	VIII	IX X XI	XII
Schwannoma	(n=50)	35	46	13	4
Meningioma	(n= 10)	6	9	7	0
Hemaangiopericytoma	(n=2)	2	2	2	0
Epidermoid	(n=2)	2	2	2	0
Ependymoma	(n=1)	1	1	0	0
Total	(n=65)	46	60	24	4
Percent		71%	92%	37%	6%

Table-III

Tumor Diameter and LCN Involvement (n=40)

Diameter	Group A	Group B	Group C
<3 cm	1	0	0
3-4 cm	23	8	1
>4 cm	13	7	1
Total	37	15	2
Percent	93%	38%	5%

Table-IV

Clinical findings: (n=55)

Dysfunction of Optic nerve:	37 (67.27%)	visual diminution alone in 6 (11%) and rest with papilledema
Dysfunction of the third nerve:	5(9.09%)	
Dysfunction of Trigeminal (Sensory)	39 (73.58%)	of these 2(3.6%) had motor findings as well
Dysfunction of VI		9(16.36%)
Dysfunction of VII		38(69.09%)
Dysfunction of VIII		51(92.72%)
Dysfunction of IX/X/XI	19(35.18%	
XII dysfunction		4(7.2%)
Ataxia / Cerebellar signs		35(63.6%)
Long tract Motor signs		4(7.27%)
Hydrocephalus (n=64)	44(68.75%)	

Pre op hearing *(n=55)* both useful and minimum perception were present in 32/55 (58.18%) patients, therefore less chance of hearing preservation in large tumors.

Discussion:

This clinical study is unique therefore it is natural to have a lot of controversies and complacencies more so because of its purposive nature. Since we get mainly large CPA tumors presenting at their very late stage this golden opportunity was grabbed for this study.

Lower cranial nerve involvement (i.e. IX, X, XI, XII) in most of the previous studies with large Schwannomas is reported about 20% [23]. In this study, the definition includes the VII and VIII cranial nerves as well. This study showed the occurrence of lower cranial nerves in about 80% for the group A, 35% for the group B and 10% for group C.

The causes for lower cranial neuropathy besides CPA tumor are varied. This small area can be considered a panorama box as it can harbor a large number of unusual lesions [12,]. In our study a case of ependymoma and two cases of hemangiopericytoma were among unusual tumors.

Lower cranial nerve involvement is found to be higher in Hydrocephalic patients that without.

Group B lower cranial nerves involvement is more observed than complained of (35% vs 29%), once they are complained of, there is usually risk of complications - Aspiration or choking which often taxes the patient's life.

Similarly, the patients take trigeminal dysfunctions so lightly that only 16% complained whereas 73% presented with it. This is a reflection of the late presentation due to the ignorance of the patients. The average duration of symptoms is 1.3 years. Physicians are also responsible to an extent for this delay.

The more anteriorly placed cranial nerves are less affected than the laterally placed ones- VI (9%) and XII (7%). This is a reflection of the local / compartmental effect of the large tumors.

The occurrence of the common forms of CPA tumors is nearly the same in this study as in most of the literatures. The earlier presentation by a decade in SAARC countries is supported by this study as 50% cases were in their third and fourth decades of life. Contrary to the reports from west, the higher incidence in females (M:F as 1:2), is not found in our series. There is a tendency of increasing number of female patients presenting with CPA tumor with them getting older from 20 years to 60 years, but after sixty this does not hold true. This may possibly be related to hormonal factors.

Maximum tumor diameter was found to be noncontributory in involving the lower cranial nerves($F=3.8$ and $P>0.05$) .

Similarly, the volumetric size of the tumor did not affect the lower cranial nerves,($F=2.65$ & $P>0.05$).

Similarly, as a by product of the study, the consistency of Schwannoma correlated with the distant Cranial nerve involvement as soft variety of the Schwannoma more frequently produced the distant effects than their counterpart firm variety z for group A=3 i.e. $P<0.05$ and for Gr B and C z= " i.e $p<0.001$.

Conclusion:

This study showed the occurrence of lower cranial nerve involvement in about 80% for the group A, 35% for the group B and 10% for group C.

It seems the Involvement of the cranial nerve does not give you the idea about the size of the tumor rather the consistency, which in fact precludes the complete removal most of the times and also advocates the early intervention.

Reference:

1. Yamakami I, Oka N, Yamaura A: Acoustic neurinoma located exclusively in cerebellopontine angle cistern "cisternal" acoustic neurinoma): a case report. No Shinkei Geka 1996 Jul; 24(7): 661-4

5. Korean Brain Tumor study group: Post graduate course in Neurosurgery for Brain Tumors: 1997: pp $_{407-432}$

6. Bonneville F, Sarrazin JL, Marsot-Dupuch K, Iffenecker C, Cordoliani YS, Doyon D, et all: Unusual lesions of the cerebellopontine angle: a segmental approach. Radiographic J 2001 Mar-Apr; 21(2): 419-38

7. Bigelow DC, Eisen MD, Smith PG, Yousem DM, Levine RS, Jackler RK et all; Lipomas of the internal auditory canal and cerebellopontine angle. Laryngoscope 1998 Oct; 108(10): 1459-69

8. Arnautovic KI, Husain MM and Linskey ME: Cranial nerve root entry zone primary cerebellopontine angle gliomas: a rare and poorly recognized subset of extraparenchymal tumors. J Neurooncol 2000 Sep; 49(3): 205-12

9. Ucar T, Akyuz M, Kazan S and Tuncer R: Bilateral cerebellopontine angle arachnoid cysts: case report. Neurosurgery 2000 Oct; 47(4): 966-8

10. Ahn MS and Jackler RK: Exophytic brain tumors mimicking primary lesions of the cerebellopontine angle. Laryngoscope 1997 Apr; 107(4): 466-71

11. Beskonakli E, Cayli S, Bostanci U, Kulacoglu S and Yalcinlar Y: Choroid plexus papillomas of the posterior fossa: extraventricular extension, intraventricular and primary extraventricular location: Report of four cases. J Neurosurg Sci 1998 Mar; 42 (1): 37-40

12. Kohan D, Downey LL, Lim J, Cohen NL and Elowitz E: Uncommon lesions presenting as tumors of the internal auditory canal and cerebellopontine angle. Am J Otol 1997, May; 18(3): 386-92

13. Heier LA, Comunale JP and Lavyne MH: Sensorineural hearing loss and cerebellopontine angle lesions: Not always an acoustic neuroma—a pictorial essay. Clin Imaging 1997 May-Jun;21(3):213-23

14. Chaimoff M, Nageris BI, Sulkes J, Spitzer T and Kalmanowitz M: Sudden hearing loss as a presenting symptom of acoustic neuroma. Am J Otolaryngol 1999 May-Jun; 20(3):157-60

15. Domenech E, Paya LM, Gozalbo JM and Aldasoro J: Metastasis of bronchogenic lung carcinoma in CNS beginning as sensorineural hearing loss. Acta Otorhinolaryngol, Esp 1997: 48: 493-495.

16. Mark S Greenberg: Acoustic Neuroma and CPA tumors: Hand Book of Neurosurgery: Greenberg Graphics, vol 2, Theme Publications, US 1997; 261-271; 945.

17. Friedman RA, Nelson RA and Harris JP: Posterior fossa meningiomas intimately involved with the endolymphatic sac. Am J Otol 1996 Jul;17(4):612-6.

18. Tos M, Charabi S and Thomsen: Clinical experience with vestibular schwannomas: epidemiology, Symptomatology, diagnosis, and surgical results. J. Eur Arch Otorhinolaryngol 1998; 255(1): 1-6.

19. Asawavichiang-inda S, Vaewvichit K and Taecholarn C: Cerebellopontine angle meningiomas with primary otologic symptoms: J Med Assoc Thai 1997.

20. Beppu T, Yoshida Y, Wada T, Arai H, Suzuki M, Kuroda K, et all: Trochlear and abducens nerve neurinomas accompanied by a cerebellopontine angle meningioma—case report. Neurol Med Chir (Tokyo) 1997 May; 37 (5): 416-21.

21. Ric H Harnsberger: Lower Cranial nerves: Hand Book of Head and Neck Imaging:, Mosby, US: 1994; 488-542.

22. MJ Chandy and A Ranjan: Acoustic Schwannomas: Tandon and Ramamurthy's Text Book of Neurosurgery II, , First Edition :BI Publications, India: 1996; 1037-1056.