Original Articles

Acoustic neuroma : a clinical study of 45 cases

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Abstract:
Acoustic Neuroma is the commonest CPA tumor. A cross-sectional study of 45 cases was carried out in one year period. Sex incidence was just opposite to western countries with male to female ratio 3:2. The patients were younger than those found in available literature with mean age 39 years. Three bilateral tumor were found with no peripheral lesion and 2 had exhibited positive family history. In this series most of the tumors were fairly large. Common presentations were hearing loss, tinnitus, raised intracranial pressure and multiple cranial nerve palsy. A large number of patients had features of brain stem and cerebellar compression.

Key word: *Accoustic Neuroma, Clinical presentation.*

Introduction

By the early 1900s, the diagnosis of brain tumors had progressed to the points at which patients with particular symptoms were definitely found to have brain tumors.

Cushing emphasized that unilateral hearing loss is the first symptom of an acoustic tumor and that this symptom must be specifically sought by the examiner. As predicted by him early diagnosis has resulted in decreased morbidity and mortality rates from this tumor.

Acoustic neuroma accounts for nearly 90% of internal auditory canal and CPA tumours. Te second most common is meningioma followed by congenital cholesteatoma. CPA tumours particularly acoustic neuroma though is histologically benign, pose as a source of serious morbidity and high mortality. Removal of these tumours has not only been a changed life saving decompressive procedure to a functional preservation of not only facial motion but also the hearing with complete removal of the tumour.

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The aim of the surgical advancement is life saving decompression and functional preservation with complete clearance of lesion.[2]

In almost every pathologic entity of the IAC, hearing loss is the most common feature, although it is most prevalent in acoustic neuromas. The hearing loss can be and usually is gradual in onset, probably due to gradual compression of the cochlear nerve. However, a significant number (10-20%) of patients with intracanalicular lesions will experience sudden SNHL[3]. This is believed to be caused by occlusion of the IAC artery by the lesion. The sudden onset may be attributable vascular occlusion secondary to new onset tumor edema, intratumoral hemorrhage, etc. This sudden SNHL is frequently reversible, especially in response to steroid treatment and no doubt may be dismissed as idiopathic in such cases. This is a problem because most of these occurrences are thought o occur with small intracanalicular tumors, and early diagnostic opportunities are potentially lost. Lack of early diagnosis bears a negative impact on hearing preservation [2,3]

Routine evaluation in most institutions includes fully audiometric testing including retroeochlear tests, ABR & sometimes ENG. If suspicion persists after these evaluations, an MRI i performed. This creates a dilemma for the clinician. The MRI is beloved to be the most sensitive test, especially for very small lesions, but the expenses of routine MRI evaluation of every patient presenting with a symptom associated with ANs is prohibitive. Management of ANs is a controversial topic. The reason for this is the issue of hearing preservation. Studies of post operative results show variability, but overall, preservation attempts have been statistically disappointing. Most series reveal a minority of attempts to be successful.[3]

Aims and Objectives

General: To review the clinical presentations of acoustic neuroma.

Specific:

- To find out the hearing status of patients of acoustic neuroma.
- To find out the frequency of involvement of different cranial nerves.
- Occurrence of acoustic neuroma in different age group and sex.

Materials and Methods

Time of study: July 2004 to June 2005

Type of study: Cross sectional

Place of study: Bangabandhu Sheikh Mujib Medical University Hospital, Dhaka

Sampling technique: Purposive sampling from consecutive cases diagnosed on the basis of clinical, radiological and histopathological findings.

Sample size (N) : 45 (Forty five)

Patients: All consecutive cases of acoustic neuroma.

Evidences: Images, histopathology reports and pictures, int4ra-opertive notes, questionnaires, examination notes.

Selection of patients Inclusion criteria:

- All patients with radiologicaly suggested and then histologically proved cases of acoustic neuromas.
- Cases with clinical evidence of acoustic neuroma and then histologically diagnosed tumor.

Exclusion criteria

- Cases which are clinically/apparent but histologically not proven.
- Recurrent cases.
- Patients who denied operation
- Patients with inadequate information

Limitations of study:

Sample size is small as it is not a common tumor.

- Some audio logical investigations (i.e. ABR) could not be done because of lack of facility.

Observation and Results.

The findings of the study derived from the data analysis are furnished below:

Table I
Age distribution of the patients (N=45):

*Age of the patients (yrs)	Number	%
<20	05	11.1
20-30	05	11.1
30-40	12	26.7
40-50	14	31.1
>50	09	20.0

Median age = (40.0 $\pm$ 1.90) years; range; (6-70) years

Table II
Hearing loss of the patients (N=45):

*Hearing loss	Number	%
No loss	07	15.6
Mild	08	17.8
Moderate	08	17.8
Severe	05	11.1
Profound	17	37.8

Median hearing loss = (60.0$\pm$5.40) dB; range: (0-110) dB.

TableIII
Optic atrophy of the patients (N=45):

*Hearing loss	Number	%
Yes	18	40.0
No	27	60.0

Table IV
Rise of ICP of the patients (N=45*):

Rise of ICP	Number	%
Yes	40	88.9
No	05	11.1

Table V

Distribution of the cases by Cranial Nerve (CN) palsy (N=45*):

CN Palsy	Number	%
V Nerve	26	57.8
VI Nerve	03	6.7
VII Nerve	13	28.9
VIII Nerve	38	84.4
IX Nerve	14	31.1
X Nerve	14	31.1
XI Nerve	10	22.2
XI Nerve	01	2.2

*Total will not correspond to 100%, because of multiple response.

Table VI

Different combinations of Cranial Nerve (CN) palsy (N=45*):

CN Palsy	Number	%
V, VIII	26	57.8
V,VII, VII	08	17.78
V,VII, VIII, IX,X,XI	05	11.11
V,VIII,IX,X,XI,XII	01	2.22
IX,X,XI	06	13.33
V,III,IX,X	12	26.67
V,VI,VII,VIII, IX,X,XI	02	4.44

*Total will not correspond to 100%, because of multiple response.

Table VII

Distribution of the cases by clinical features (N=45*):

CN Palsy	Number	%
Hearing loss	38	
Vertigo/Unsteadiness	21	84.4
Tinnitus	33	46.7
Recruitment	02	73.3
Papilloedema	38	4.4
Optic atrophy	18	84.4
Rise of ICP	40	40.0
Cerebellar dysfunction	28	88.9
Hydrocephalous	36	80.0
Brain stem compression	12	26.7

*Total will not correspond to 100%, because of multiple response.

Table VIII

Distribution of the patients by Tumor size (N=45):

*Tumor size (cms)	Number	%
<2	01	2.2
2-3	03	6.7
3-4	22	48.9
4-5	13	28.3
$\geq$5	06	13.3

*Median size = (3.80 $\pm$ 0.12) cms; range: (1.99-5.60) cms

Table IX

Distribution of the patients by hearing loss and Tumor size (N=45):

Tumor size (cms)	Hearing loss					P-value
	No loss	Mild	Moderate	Profound	Severe	
<3	2(50.0)*	00	00	2(50%)	00	
3-4	3(13.0)	5(21.7)	7(30.4)	6(26.1)	2(8.7)	0.182
>4	2(11.1)	3(16.7)	1(5.6)	9(50.0)	3(16.7	

Figures in the parentheses indicate corresponding %.
p<0.05 was considered significant.

Discussion

Tumor of the Cerebellopontine angle would typically be expected to present with a predictable series of symptoms and signs. The natural history of neoplasm's of the internal auditory canal and subsequently CPA was elucidated by Cushing in 1917. Only history and acoustic neuroma does not vary, much from series to clinical examinations were reliability available at that time, although Cushing did perform caloric test on many of the patients suspected of having CPA lesions. The incidence of series it is 6% in all-intracranial tumors according to Hain (2004), Vrabec (1994) estimated it 8-10%.

Prevalence of incidental acoustic neuroma is only 0.02 percent 5,6. Sex distribution of this tumor varies between different centres. Female predominates than male in western studies. According to Vogel (1996) and Verabec (1994), female to male ratio is 3:2. But in our country, male outnumbers female, Barua KK (2004) estimated it just reverse (3:2), in the present series the observaiton is same.[7]

It is generally agreed that acoustic neuroma is a disease of middle age. This is commonest in the fourth to sixth decade8. But here it is commoner in younger age group. 26.7% of patients of this series had presented in 3rd and 4th decade and 31.1% were between 4th and 5th, so about 80% were below 50. The lowest age of the patient was 6 years and the highest was 70, and 9 were above 50 years old. Median age of the patients were 40.0±1.90 years. In this study it was observed that incidence of acoustic neuroma in Bangladesh is earlier. It may be due to higher life expectancy of people in other country or due to improper age recording in our country.

The study exhibited no side predominance of the tumor. They occurred almost equally distributed in both sides and carried no clinical significance. Three patients had bilateral acoustic neuroma with no peripheral lesion. Two of them had family history of same disease. They were diagnosed as neurofibromatosis type 2. One patient had both acoustic neuroma and meningioma in sameside,[8,9].

Tumor size contributes to clinical presentations of the lesion. Authors defined the size in different ways. Koos etal (1978) had described grades I-IV. Long DM (2000) had studied 1022 patients between July 1969 and January 1998 and divided the tumor in 3 groups; group I tumors (61%) were smaller than 2.5 cm, group II tumors (24%) from 2.5-4.0 cm and group III tumors (15%) larger than 4.0 cm. In his study, majority of patients were smaller tumors. Contrary to this, in the present series, half (48.9%) was medium sized, 42.2% were large tumor (>4 cm), only one was intrameatal small tumors were only 8.9% and rest of the tumors were bigger. Median size of the tumor was (3.80±0.12) cm. This figure indicates that the majority of patients were presented in advanced stage.

Acoustic neuroma is a slow growing tumor. It grows 2 mm per year even less growth rate also reported 0.2-2.0 mm[5]. Duration of symptoms are so longer than other tumors. In the present series, average duration of symptoms is 18 months.

Regarding clinical presentations, there are variations in available data. The earliest symptom reported by the patients in the series of Ojmann et al (1972) was loss of hearing (33 of 46 patients), headache (4 of 46), balance disturbance (3 of 46), facial pain, tinnitus and facial weakness each in single cases. But in all other series hearing loss is the most frequent symptom, occurring in more than 95 percent of patients. About 90 percent present with a one sided, slowly progressive hearing impairment. A high frequency sensorineural pattern is the most common type, occurring in approximately two thirds of patients. In the remaining third the next most common observation is hearing loss at low frequency (which would be more typical of Meniere's disease). Even less commonly, some have the "cookie bite" pattern (suggestive of congenital hearing loss)[10].

A sudden hearing loss occurs in about 25 percent of patients with acoustic neuroma. However, because acoustic neuroma is a rare condition, sudden hearing loss attributable to an acoustic tumor occurs in only 1-5 percent of patients with sudden hearing loss as there are many more common causes. Even a sudden hearing loss with complete recovery can be caused by an acoutic. In our opinion, this percentage is high, but nevertheless certainly acoustic neuroma's can be found in persons with symmetrical hearing. Tinnitus is very common in acoustic neuroma, is usually unilateral and confined to the affected ear. Brackman estimated it 85% and it varies from 77-95% in some other studies. The hearing loss can be and usually is gradual in onset due to gradual compression of cochlear nerve. However a significant number (10-20%) of patients with intracanalicular lesions experience sudden SNHL. McElveen estimated 26% patients with sudden hearing loss, and surprisingly they had been improved with steroid therapy. Tinnitus was the second most common symptom (56%) in his series. Poor SDS and rollover phenomenon were his another findings[10,12].

In this series, 8 (17.8%) patients had mild, same had moderate, 5 (11.1%) had severe, 17 (37.8%) had profound hearing loss. But 7 (15.6%) patients had no hearing loss. Normal hearing is not surprising. Morrison and Sterkers (1996) shown about 11% patients of their study had normal hearing. Thapa BK (2000) had described 16% patients with no hearing loss. Tumors above 4 cm had variable hearing, 2 (11.1%) had no loss, majority (66.7%) had severe to profound hearing loss. Hearing loss of other patients were not proportional to increased size of tumor. In the present series the difference between the groups based on the size of the tumors did not reach the level of significance with respect to hearing loss.

In study of Barer KK (2004), only 2% patients were exhibited normal hearing. He also had shown tinnitus in 88% of his respondents. The degree of hearing loss was not proportionate with the size of the tumor. A2 cm tumor shown profound hearing loss but 4 cm tumor exhibited mild loss. Vertigo and disequilibrium are uncommon presenting symptoms. Rotational vertigo is much more common in smaller tumors. In Samii's series of 16 patients with intracanalicular tumors, 75% of patients presented with vertigo. But

unsteadiness or imbalance, on the other hand, appears to be more common in larger tumors. In the present series, 40% of larger (>3 cm) reported balance disturbance, but these were not their predominant complain. It was explored after careful questioning. In the series of Samii, 10% patients exhibited vertigo or imbalance as presenting feature. According to Hain (2004), 70% of patients with large tumors had unsteadiness [5,13,14].

In the present series, most of the patients attended for consultation in advanced stage. Usually they does not seek advice in early, otological stage. Out of 45 patients, only 11 (24.4%) patients were attended initially by otolaryngologists and then referred to neurosurgeon after imaging study. Headache was the earliest symptom for which they seek advice of physician but all of them noticed hearing loss as and for this, they did not attended for consultation. They did not give any significance of this symptom. Headache was present in 94% of patients. Edward and Patterson (1994) found 84% of patients with headache as their initial symptom. He also found a strong correlation between tumor size and headache. In their series, 92% of tumor was large, >2.5 cm. In the present series, 94% of patients had tumor size more than 2.5 cm. So there is a close correlation between size of the tumor and headache. It also indicates rise of intracranial pressure. Other features of intracranial pressure rise are vomiting, pailloedema, failing vision, bradycardia and hypertension. Optic atrophy, diplopia, anosmia are secondary to rise of intracranial pressure. When tumor size exceeds 4 cm, it compresses brain stem and causes motor deficit, obstructs CSF pathway and develops hydrocephalus[9,17]. In the study of McElveen and Saunders (1994), these advanced feature of tumor was presented in 66-76% patients. In this study, these features were as follows; vomiting 80%, papilloedema 70%, optic atrophy 40%, Anosmia 6%, diplopia 12%, features of brain stem compression 20%. Four patients had complete failure of vision. Eighteen had undergone ventriculoperitoneal shunt for getting immediate relief from raised intracranial pressure. Neuroradiological evidence of hydrocephalus was found in 82% of patients. But in other studies, it ragnes from 5-50% (McElveen 1994). Evidence suggests that as tumor size increased, severity of hydrocephalus also increased.

Symptoms related to cerebellar dysfunction are found in patients with large tumors. These have been reported in 46% of patients with acoustic neuroma. In the present series, cerebellar dysfunction was estimated in 62% of patients[17].

Cranial nerves involvement in acoustic neuroma is an important issue. `Involvement' was defined by Long DM (2000) as adherence between neurovascular structures and the tumor (or capsule), for which the surgical resection was required to free the st4ructure. Preoperatively it can be diagnosed by clinical examination. Fifth to twelfth cranial nerves are directly involved by enlarging tumor but upper nerves dysfunction (anosmia, papilloedema, optic atrophy) are secondary to raised intracranial pressure[9,17].

In the series of Prakash and Long (2000), larger tumors (>4 cm) had a higher incidence of involvement of 4th Cranial nerve (41%), 5th CN (100%), 9th to 11th CN (99%) and 12th CN (31%). Involvement of cranial nerves were proportional to the size of tumors. Larger tumor affects nerves more than smaller tumors but it is not true in case of 8th nerve as mentioned before 18. In the early reports of relatively large tumors documented the findings of these nerves involvement in approximately 30% of patients (Rengachery et al 1999). Compression of lower cranial nerves results from extension of tumor inferiorly towards jugular foramen. In the present series. Trigeminal nerve was most commonly involved (57.8%) followed by Glossopharyngeal, vagus along with triggerman nerve (26.7%). All of them had hearing loss. Facial nerve involvement was 28.9% and the tumor > 2.5 cm involved this nerve, smaller tumors are not to cause any facial nerve dysfunction. This nerve is predominantly motor nerve and relatively resistant to tumor invasion. If facial nerve is involved in early stage of tumor, the diagnosis is more likely to be meningioma or Cholesteatoma than acoustic neuroma (Ramsden 1997). In the series of Hain (2004), Facial nerve disturbance was uncommon, found only in 50% of patients of the larger tumors (>2.5 cm), exhibited by facial twitching, hemi facial spasm and facial asymmetry[10,19,20].

It is not surprising that size of the tumor plays major role in producing clinical features of acoustic tumor and huge works had done on this ground in home and abroad especially with lower cranial nerves manifestation 20. In the series of Thapa BK (2002), 73% had St" nerve, 69% 7" nerve and 36% had 9th, 10th and 11th nerve dysfunction; cerebellar signs were present in 68% patients. Hypoglossal nerve involvement was uncommon despite the size of the tumor (2.22%)19. From above discussion, we se a small picture of patients of acoustic neuroma that they presents almost always in advanced stage.

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