



CLINICAL PRESENTATION OF ACOUSTIC SCHWANNOMA: EXPERIENCE AT A TERTIARY CARE INSTITUTION

Neurosurgery

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ABSTRACT

Background: Acoustic neuroma is a benign tumour and it is the most common tumor of the cerebellopontine angle. Descriptions of the signs and symptoms of acoustic neuroma within the literature are largely from older studies, which may not be applicable to contemporary practice. It presents with hearing loss, tinnitus, disequilibrium, headache, fifth, seventh and lower cranial nerve symptoms, depending upon size.

Aims and Objectives: To study symptoms of acoustic schwannoma with respect to age, sex, clinical presentation, and correlation between size of tumor and various manifestations as well as association with hydrocephalus

Materials and Methods: A retrospective study carried out in the department of neurosurgery, Institute of Medical Sciences, BHU. It includes 42 patients of acoustic schwannoma who were admitted from January 2015 to December 2017.

Results: Out of all 45.24% were male and 54.76% female. Patients were mostly more than 20 years age. Hearing loss (95.4%) and tinnitus (78.57%) were commonest symptoms. 71.43% tumors were more than 3cm size. Symptoms like disequilibrium, facial numbness lower cranial nerve symptoms were significantly more common in tumors larger than 3cm. Hydrocephalus is also seen in larger tumors only.

Conclusion: Acoustic schwannoma usually present in young and middle aged persons. These tumors usually present when becomes large. Facial numbness, disequilibrium, facial weakness, diplopia and lower cranial nerve symptoms and hydrocephalus are present only in large tumors.

KEYWORDS

Acoustic schwannoma, cerebellopontine angle, vestibular schwannoma

Introduction

Acoustic neuroma or vestibular schwannoma is a benign tumour of the Schwann cell that most commonly arises from the vestibular nerve [1]. It is one of the most common intracranial tumors, comprising 8-10% of tumors in most series[2]. It is the most common tumor of the cerebellopontine angle (CPA) (80–90%). These tumors may remain dormant without any symptom or can present with various symptoms which could be due to tumor mass and involvement of the adjacent cranial nerves, cerebellum and brainstem. There could be associated signs and symptoms due to raised intracranial pressure (ICP) due to regional brainstem compression and hydrocephalus. Descriptions of the signs and symptoms of acoustic neuroma within the literature are largely from older studies, which may not be applicable to contemporary practice [3,4]. Furthermore, adequate statistical analysis of these variables with multivariable regression has yet to be performed. Acoustic schwannoma typically become symptomatic after 30 year age. The most common symptoms of are unilateral sensorineural hearing loss (98%), dysequilibrium (67%), tinnitus (70%), headache (32%), facial numbness (29%), facial weakness (10%), nausea and vomiting (9%), otalgia (9%) and diplopia (10%) and change of taste (6%) [2]. Hearing loss, tinnitus and disequilibrium are related to pressure on the eighth nerve complex in the internal acoustic canal. Otagia, facial numbness, weakness and taste change occurs as tumor enlarges and compresses the fifth and seventh nerves. These symptoms occur usually tumors grow beyond 2 cm[5]. Larger tumors cause brainstem compression (with ataxia, headache, nausea vomiting, cerebellar signs, and if unchecked, coma, respiratory depression and death) and lower cranial nerve palsies (hoarseness, dysphagia). Obstruction of CSF circulation by larger tumors may produce hydrocephalus[6].

Aims and objectives

To study symptoms of acoustic schwannoma with respect to age, sex, clinical presentation, and correlation between size of tumor and various manifestations as well as association with hydrocephalus. The study of clinical and historical features within each patient's presentation and the identification of the features associated with larger tumours will allow for an accurate diagnosis to be made quickly and, therefore, the appropriate investigations can be targeted towards those patients with the greatest clinical need. Hence, a secondary objective of this study was to identify those clinical features associated with larger tumour size at initial diagnosis.

Material and methods

This was a retrospective study carried out in the department of

neurosurgery, Institute of Medical Sciences, BHU in patients with diagnosis of acoustic schwannoma. It includes 42 patients who were admitted from January 2015 to December 2017. Detailed record of age, sex and various symptoms was made. Every patient's MRI and/or CT scan record was made and based upon it tumor size and presence or absence of hydrocephalus was recorded.

Results

Total 42 patients were included in the study. 19 (45.24%) were male and 23 (54.76%) 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 2 and both were male. Patients between 21 and 40 years age were 22 (52.38%) and between 41 and 60 years were 18 (42.86%). Average age of presentation was 39.24 and standard deviation was 12.79.

Table 1: Age distribution in different age groups

Age group	No. of patient	Percentage
0-20 years	2	4.76
21-40 years	22	52.38
41-60 years	18	42.86

Symptom in patients varied, hearing loss was the commonest symptom present in 40 (95.24%) patients. Other symptoms like tinnitus, disequilibrium and headache were also common, their frequency is shown in table 2. Symptoms like facial numbness, facial weakness, hoarseness of voice and dysphagia were somewhat lesser in frequency. We had four patients with complaints of diplopia also.

Table 2: Symptoms in acoustic schwannoma

Symptom	No. of patient	Percentage
Hearing loss	40	95.24
Tinnitus	33	78.57
Disequilibrium	29	69.05
Headache	20	47.62
Facial numbness	10	23.81
Facial weakness	10	23.81
Hoarseness of voice	12	28.57
Dysphagia	12	28.57
Diplopia	4	9.52

On the basis of MRI tumor size was recorded ranging from 1.5cm to 5cm. Average size was 3.43cm with standard deviation 0.86. Based on tumor size patients were grouped in two categories (table 3). Group A included tumors less than or equal to 3cm size and Group B with more than 3cm.

Table 3: Patients in different size groups

Groups	No. of patients	Percentage
Group A (≤ 3 cm)	12	28.57
Group B (> 3 cm)	30	71.43

Again based on MRI brain, it was seen that 17 (40.48%) patients had hydrocephalus and 25 (59.52%) had not any hydrocephalus

Comparison between both groups i.e, Group A and B was made with regards to sex, age, different symptoms and presence or absence of hydrocephalus in MRI brain. These are shown in table 4, 5, 6 and 7.

Table 4: Sex and size wise distribution

Size of tumor	Male	Female
Group A (≤ 3 cm)	6	6
Group B (> 3 cm)	13	17

p-value: 0.6950

On comparison between Group A and Group B regarding frequency of male and female patients, it was found insignificant

Table 5: correlation between age and size

Age group	Group A (≤ 3 cm) (n=12)	Group B (> 3 cm) (n=30)	p-value
0-20 years	1	1	0.4918
21-40 years	7	15	0.6252
41-60 years	4	14	0.4302

Comparison between three age groups and and Group A and B showed more frequency of larger tumors in higher age groups, however this observation was not found significant.

Table 6: Various clinical presentations vs tumor size

Clinical presentation	Group A (≤ 3 cm) (n=12)	Group B (> 3 cm) (n=30)	p-value
Hearing loss	10	30	0.153
Tinnitus	8	25	0.235
Disequilibrium	4	25	0.001
Headache	2	18	0.011
Facial numbness	0	10	0.040
Facial weakness	0	10	0.040
Hoarseness of voice	0	12	0.015
Dysphagia	0	12	0.015
Diplopia	0	4	0.4897

On comparing size of tumor and different symptoms it was found that Larger tumors presented more frequently with all symptoms. Hearing loss and tinnitus were common symptoms in smaller tumors as well. Headache and disequilibrium were also present in two and four cases respectively, but in larger tumors these symptoms were significantly higher in frequency. Facial symptoms, diplopia and lower cranial nerve symptoms were seen only in larger tumors.

Table 7: correlation between tumor size and hydrocephalus

MRI finding	Group A (≤ 3 cm) (n=12)	Group B (> 3 cm) (n=30)
Hydrocephalus	0	17
No hydrocephalus	12	13

p-value: 0.0009

hydrocephalus was also seen only in larger tumors.

Discussion

Acoustic schwannoma is the most common tumor of the cerebellopontine angle (CPA) (80–90%). We had a total of 42 patients out of which 19(45.24%) were male and 23(54.76%) were female. Patients less than 20 years age were only 2. Patients between 21 and 40 years age were 22(52.38%) and between 41 and 60 years were 18

(42.86%). Average age of presentation was 39.24 and standard deviation was 12.79. In a similar study done on acoustic schwannoma by Sourabh et al had maximum number of cases i.e 38.46% in 31 to 40 years of age[7]. The average age of presentation in our study was 39.24 with standard deviation 12.79. In Saurabh et al study mean age was 37.23 years and standard deviation was 11.41. Memari F et al had mean age of 49 years in their series[8]. There was a female predominance in our study similar to Saurabh et al[7]. But in series of Memari F et al[8] there was male predominance. No significant correlation was found between age and tumor size and tumor size vs sex in our study. Neither any study in literature shows any significant correlation between them. We had tumors larger than 3cm more common in our study that is total 30 (71.43%) and tumors smaller than 3cm were only 12 (28.57%). Average tumor size in our study was 3.43cm. 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[8] 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 40 (95.24%) cases including both small and large tumors. Tinnitus was the second most common symptom with 33 (78.57%) patients including small or large tumors. According to saurabh et al[7] SNHL and tinnitus was present in all patients (100%). In our study disequilibrium was present in 69.05% patients and Headache was present in 20 (47.62%) patients. In the study by Saurabh et al[7] headache was present in all cases. In our study facial numbness and facial weakness was there in 10(23.81%) patients all of which were more than 3cm size. Saurabh et al[7] had some different observation, they had facial paresis in 76.92% trigeminal involvement 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[8] had observed that Forty-seven patients (94%) presented with tinnitus, and vertigo was present in 30 patients (60%). M. Javad Mirzayan et al[9] 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 of increased ICP 15% (3).

Symptoms like disequilibrium, headache, facial symptoms and lower cranial nerve symptoms were seen significantly more frequently in larger tumors. similarly A large multi-national survey-based study identified unilateral hearing loss in 86% of patients, with unsteadiness in 61%, tinnitus in 57% and headache in 36% [3]. However, this study from Wiegand et al. does not draw comparisons between these symptoms and patient tumour size. Nor does this particular study differentiate between the main presenting symptoms or discuss symptom severity. The contemporary series of patients outlined in this article showed similar levels of hearing loss and tinnitus, but smaller numbers of positive cranial nerve abnormalities and cerebellar signs. The explanation for this is likely because hearing loss and tinnitus are not size-dependent features of acoustic neuroma, whilst symptoms and signs associated with compression of the nearby cranial nerves, brainstem and cerebellum are size dependent. Matthies et al. provide a series of 1,000 patients and explain that 95% of patients undergoing surgery have hearing loss prior to their operation and 63% were suffering from tinnitus [10]. The authors go on to report vestibular disturbances in 61%, headaches in 12% and taste disturbances in 2% Based on MRI brain, we found that 17 (40.48%) patients had hydrocephalus (HCP) and 25 (59.52%) had not any hydrocephalus and all the hydrocephalus patients were larger tumors. This means that hydrocephalus occurs in larger tumors only when due to tumor growth aqueduct is obstructed. The incidence of HCP associated with vestibular schwannoma ranges from 3.7% to 42%[11,12,13,14,15].

CONCLUSION

Acoustic schwannoma 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, disequilibrium, facial weakness, diplopia and lower cranial nerve symptoms are present only in large tumors. Hydrocephalus is associated with large tumors only.

REFERENCES

1. Vestibular schwannomas: Lessons for the neurosurgeon: part II: molecular biology and histology. DeLong M, Kirkpatrick J, Cummings T, Adamson D. *Contemp Neurosurg.* 2011;33:1-4.
2. Harner S G, Laws E R: Clinical findings in patients with acoustic neuromas. *Mayo Clin Proc* 1983; 58:721-8
3. Acoustic neuroma--the patient's perspective: subjective assessment of symptoms, diagnosis, therapy, and outcome in 541 patients. Wiegand DA, Fickel V. *Laryngoscope.* 1989;99:179-187.
4. Moffat DA, Baguley DM, Beynon GJ, Da Cruz M. *Am J Otol.* Vol. 19. Accessed; 1998. Clinical acumen and vestibular schwannoma; pp. 82-87.
5. Tarlov E C. microsurgical vestibular nerve section for intractable Meniere's disease. *Clin Neurosurg* 1985; 33:667-84
6. Greenberg Mark S. *handbook of Neurosurgery.* 7th ed. Thieme publishers, 2010
7. Dixit Saurabh, Banga Manpreet Singh, Saha Suniti Kumar, Roy Kaushik, Gosh Partha, BV Sandeep. A study assessing the post operative outcome in patients of acoustic schwannoma operated through retrosigmoid approach at tertiary care institutions- An experience of one year. *Asian Journal of Medical Sciences* July-Aug 2017(8):44-49
8. Memari F, Hassannia F and Abtahi SHR. Surgical Outcomes of Cerebellopontine angle Tumors in 50 Cases. *Iranian Journal of Otorhinolaryngology* 2015; 27(78):29-34.
9. Mirzayan MJ, Gerganov VM, Lüdemann W, Oi S, Samii M and Samii A. Management of vestibular schwannomas in young patients—comparison of clinical features and outcome with adult patients. *Child's Nervous System* 23:8, 891-895.
10. Samii M and Matthies C. Management of 1000 vestibular schwannomas (acoustic neuromas): Surgical management and results with an emphasis on complications and how to avoid them. *Neurosurgery* 1997; 40:11-23.
11. Briggs RJ, Shelton C, Kwartler JA, Hitselberger W. Management of hydrocephalus resulting from acoustic neuromas. *Otolaryngol Head Neck Surg.* 1993;109:1024-4.
12. Atlas MD, Perez de Tagle JR, Cook JA, Sheehy JP, Fagan PA. Evolution management of hydrocephalus associated with acoustic neuroma. *Laryngoscope.* 1996;106(2 PT 1):204-6
13. Pirouzmand F, Tator CH, Rtko J. Management of hydrocephalus associated with vestibular schwannoma and other cerebellopontine angle tumors. *Neurosurgery* 2001 Jun; 48(6):1246-53
14. Gerganov VM, Pirayesh A, Nouri M, Hore N, Luedemann WO and Oi S. Hydrocephalus associated with vestibular schwannomas: management options and factors predicting the outcome. *J Neurosurg* 2011; 114:1209-1215.
15. Fukuda M, Oishi M, Kawaguchi T, Watanabe M, Takao T, Tanaka R, et al. etiopathological factors related to hydrocephalus associated with vestibular schwannoma. *Neurosurgery.* 2007;61:1186-92.