RESEARCH ARTICLE

Demographic variables of Vestibular schwannoma's patients presented in Radio Surgical out- patient department of Cyberknife Robotic Radiosurgery, JPMC Karachi

Shazia Kadri, Naveed Ahmed, Aneeta Ghulam Muhammad, Kamran Saeed, Syed Furqan Ahmad Hashmi, Tariq Mahmood

Abstract

Objective: To determine the age, gender and sites of vestibular schwannoma cases using contrast-enhanced magnetic resonance imaging.

Method: The retrospective descriptive study was conducted at the Department of Cyberknife Robotic Radiosurgery, Jinnah Postgraduate Medical Centre, Karachi, and comprised data of patients with vestibular schwannomas from January 2016 to September 2018. Some of them were histologically proven and rest were radiologically proven. Cases were reviewed on contrast-enhanced magnetic resonance imaging of the brain. Statistical Package for the Social Sciences version 20 (SPSS) was applied.

Results: Of the 500 cases of vestibular schwannomas identified with 515 tumours, 300(60%) were males and 200(40%) were female. The overall mean age was 42.7±14.4 years (range: 17-85 years). Out of 515 tumours, the commonest site was the right cerebellopontine angle in 340(66%) cases. There were 15(3%) cases of radiologically-proven neurofibromatosis type 2. Overall, 490(98%) patients had main clinical complaint of progressive unilateral hearing loss, 5(1%) had vertigo and 5(1%) had facial palsy.

Conclusion: Vestibular schwannomas were found to be more common among adults, with male preponderance and right cerebellopontine angle being the common site.

Keywords: Vestibular schwannoma, VS, Cerebellopontine angle, CPA, MRI, Magnetic resonance imaging, Neurofibromatosis type II, NF II. (JPMA 72: 62; 2022) **DOI: https://doi.org/10.47391/JPMA.1198**

Introduction

Vestibular schwannoma (VS) are the most common cerebellopontine angle (CPA) mass arising from the Schwann cells of the vestibulocochlear nerve. It is also called acoustic neuroma. Unilateral lesion is seen in more than 90% cases with median age 50 years having complaints of unilateral hearing problem and tinnitus.[1] These patients can be planned for surgery, radiotherapy or conservative management, according to age, symptoms, size of tumour and its growth.[2]

The VS incidence is increasing with advancement of imaging techniques. There was three-fold increase in annual VS number seen over a period of 23-years.[3] VS has slight female predominance in the United States.[4] There is higher incidence of 2.66/100,000 person-years in Taiwan and 1.37/100,000 person-years in Asia-Pacific islanders, but the rate is 0.45 per 100,000 persons in Hispanic populations.[5,6]

Increasing incidence and changing trends in VS presentation make it vital for surgeons and physicians to have adequate knowledge of the emerging patterns of disease presentation. Significant research has been carried out on this topic in international settings, but unique population demographics and variable access to care in local settings may render the existing data insufficient or not fully applicable to Pakistani population. The current study was planned to evaluate the age, gender and VS sites on contrast-enhanced magnetic resonance imaging (MRI) in local tertiary care settings.

Materials and Methods

The retrospective descriptive study was conducted at the Cyberknife Robotic Radiosurgery Department of the Jinnah Postgraduate Medical Centre (JPMC), Karachi and comprised data of VS patients from January 2016 to September 2018. After approval from the institutional ethics review board, the sample size was calculated on the basis of literature. Written consent had been taken from all the patients at the time of the actual treatment. Data retrieved through medical records related to VS patients who had been diagnosed on the basis of either characteristic radiological findings on MRI of the brain contrast, or proven on histology report following surgery.

Patients had been referred from different centres of Pakistan and abroad. Those included in the current study were patients with hearing problem, facial palsy and vertigo having been diagnosed VS radiologically on

Department of Cyberknife Robotic Radiosurgery, Jinnah Post Graduate Medical Centre, Karachi, Pakistan.

Correspondence: Shazia Kadri. Email: shaziaum_wk@yahoo.com

imaging or histopathologically after surgery. CPA tumours other than VS and radiologically doubtful CPA VS with suspicious radiological appearance and without histology report were excluded. Neurofibromatosis type II (NF II) cases were also diagnosed on the basis of imaging as bilateral VS. Patients of either gender aged 17-85 years presented with complaints, like hearing loss, facial palsy, tinnitus, dizziness, etc. Even those having had schwannoma surgery showed symptoms of hearing loss, headaches and facial muscle weakness. The cases were reviewed on contrast-enhanced MRI of the brain to confirm the radiological diagnosis. Two senior radiologists reviewed the scans in cases where histopathology was not available. Additional balanced turbo field echo (BTFE) sequence of MRI brain had been done in a few cases to evaluate the lesion in detail. Analysis was done using Statistical Package for the Social Sciences version 20 (SPSS). Descriptive statistics was used.

Results

Of the 900 patients with hearing problem and vertigo, 500(55.6%) were found to have VS radiologically on imaging or histopathologically after surgery. Of them, 300(60%) were males and 200(40%) were female. The overall mean age was 42.7±14.4 years (range: 17-85 years). Out of 515 tumours, the commonest site was the right CPA VS in 340(66%) cases (Table). Overall, 490(98%) patients had main clinical complaint of progressive unilateral hearing loss, 5(1%) had vertigo and 5(1%) had facial palsy.

Table: Demographics data (n=500).

Number of Tumours	515
Location of Tumour [n (%)]	
Right Cerebellopontine angle	340 (66)
Left Cerebellopontine angle	175 (34)
Gender [n (%)]	
Male	300 (60)
Female	200 (40)
Mean Age (years)	42.7±17-85

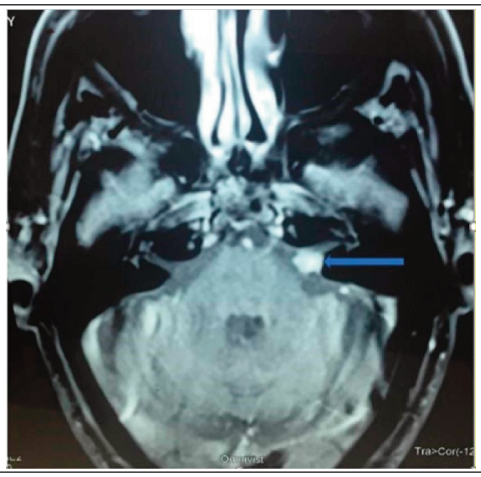

Figure-2: Left vestibular schwannoma. Magnetic resonance imaging (MRI) brain T1-weighted image (T1WI) showing small left vestibular schwannoma.

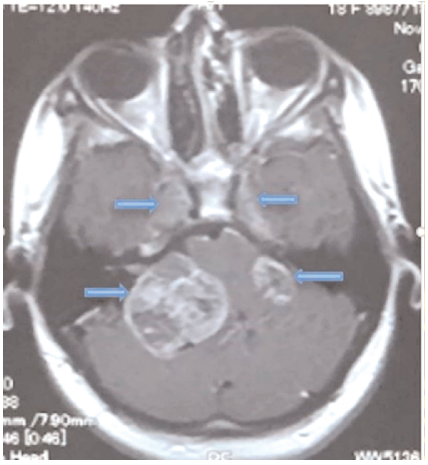

Figure-3: Neurofibromatosis Type II. T1 axial contrast image of a 35-year-old female showing bilateral vestibular schwannoma and para cavernous meningiomas.

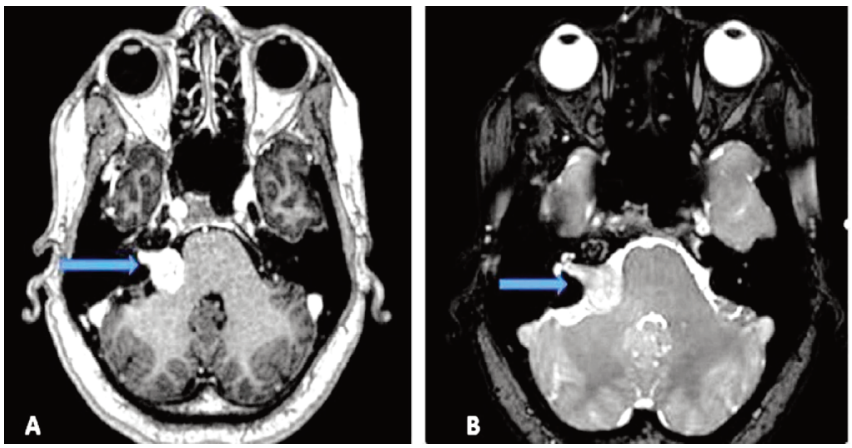

Figure-1: Right vestibular schwannoma. Magnetic resonance imaging (MRI) brain T1-weighted image (T1WI) contrast axial image (A) and Balanced Turbo Field Echo (BTFE) image (B) of a 45-year-old male showing right vestibular schwannoma with intracanalicular extension.

CPA VS were detected on both the right (Figure-1) and left (Figures-2) sides. Out of the 500 cases, 15(3%) had bilateral VS(Figure-3). A case of NF II presented with radiological hallmark of disease as bilateral VS on imaging (Figure-3).

Discussion

VS are mostly sporadic benign CPA tumours of myelin-forming cells of the vestibulocochlear nerve. Its incidence is increasing likely due to improved quality

and techniques of diagnostic imaging. Out of 500 VS cases in the current study group, the male: female ratio was 1.5:1 with median age of 46 years, and with the most common site being right side, and there were 15 cases of radiologically-proven NF II.

According to a study,[7] the incidence of VS was 1.1 per 100 000 (range 1.03-1.21) for patients diagnosed between 2004 and 2011. The fact that VS incidence seems to be increasing can be attributed in part to incidental diagnosis with the growing use of MRI and computed tomography (CT). The rise in incidence and subsequent prevalence of disease calls for continuous monitoring of VS presentation in patients reporting to local care facilities, so that a snapshot of disease characteristics in our population may be formed.

Also, the figures established in VS epidemiological studies do not account for the undiagnosed and asymptomatic tumours which may be forming a sizeable chunk of the total disease load. A comprehensive study on VS over a 20-year period from 1995 to 2015 with a total of 1,296 patients established the median age of disease presentation as 59.2 years and a female preponderance of 53.5%.[8] Another study in a developing country showed mean age of the patients as 46±3 years and a female preponderance of 1.2:1.[9] A nationwide study based on the Swedish Brain Tumour Registry (SBTR) for all diagnosed with VS between 2009 and 2015 showed 348 patients among whom 165(47.4%) were female and all underwent surgery with a mean age of 50.6±14.5 years.[10] In the current study, the median presenting age was 46 years and the male:female ratio was 1.5:1, showing male preponderance.

The VS incidence with age has been studied in USA and Nordic countries and it was found that the yearly average surge was mainly in 65 years or elder patients.[11,12] A study showed an interesting increase in age of diagnosis, from an average of 52 years to 62 years.[13] Most of the VS patients are usually of advanced age, revealing a high-risk population due to high post-surgical complication rates, including age and comorbidities.[14] These findings possibly are a much more specific and important predictor of the groups, are likelier to present with disease. An important aspect is the increasing number of elderly patients in whom open surgery and tumour removal revealed worse outcome in contrast to the young population. Quality of life (QOL) in the management of small VS studied with management options include observation, radiation or hearing-preserving microsurgery.[15] But recently with advancement of radiosurgery, the growth of tumour could be stopped with minimal side effects and preservation of nerve function.

This may be explained by general perceptions of life expectancy as well as cost-versus-outcome approach in case of older patients. Nevertheless, it is an important factor to consider by the medical team when undertaking decision-making regarding the choice to treat the disease and the preferred mode of treatment. Data regarding median age of presentation may also be helpful in counselling patients regarding the prevalence of their condition among the local population of similar age and helping them decide on the treatment option chosen by most patients of their group. These findings shift the focus of future research towards age as a very important predictor of VS diagnosis, treatment and outcome.

Out of 500 patients with acoustic neuroma enrolled in the current study, a total of 485 patients presented with unilateral lesions. It has been studied that approximately 1,000 cases of unilateral VS were noted annually in the United Kingdom from 2013 to 2016.[16] In cases of unilateral VS, which make up majority of the current study group, there was a clear tendency seen for tumour occurrence on right side. Bilateral VS were found in 15 patients which is in line with previous studies as patients with NF II are prone to develop multiple intracranial neoplasms, such as schwannomas of the cranial nerves and meningiomas.[17] They have to be differentiated from VS with regard to their biological and clinical characteristics as they show a highly variable growth behaviour and are associated with other tumours.

The hearing outcomes in NF II-related VS are predictable by considering age at diagnosis of NF II. However, between 2008 and 2015, the number of certified NF clinics grew from 32 to 50, and annual patient volume rose from 6,776 to 10,245 patients (13% of the total estimated NF patient population in the United States).[18] The characteristic feature of NF II is bilateral VS, usually by age 30 years. Even though radiosurgery shows promising results in NF II-related VS, it is still a matter of argument.[15] A study suggested that the age of onset in patients with NF II is more important than the multiplicity of tumours for forecasting the clinical course. VSs are tumours with relatively slow growth rate of approximately 1-3 mm/year and are, therefore, expected to become symptomatic late and are usually diagnosed much later in life.[19] However, another study indicated that with the rise in number, there is a the need for fresh studies to be conducted on incidence of neurofibromatosis and associated VS in the future.[20]

The current study has its limitations, as it was done at a single centre for a relatively short period and with a relatively small sample size. All these elements mean the current study and its findings may not account for the overall disease load in the whole population. However, the study does provide a current picture of VS epidemiology in

the community.

Conclusions

Vestibular Schwannomas were found to be more common among adults, with a male preponderance and the right CPA being the most common site. Most individuals presented with clinical complaints of hearing loss, vertigo and facial palsy. With advancement of imaging techniques, there are better chances of early diagnosis of this benign lesion.

Disclaimer: None.

Conflict of Interest: None.

Source of Funding: None.

References

1. Rajput MSA, Ahmad AN, Arain AA, Adeel M, Akram S, Awan MS, et al: Preservation of Hearing and Facial Nerve Function with the Microsurgical Excision of Large Vestibular Schwannomas: Experience with the Retrosigmoid Approach. Cureus. 2018;4:3684.
2. Lin EP, Crane BT. The management and imaging of vestibular schwannomas. Am J Neuroradiol. 2017, 38:2034-43.
3. Foley RW, Shirazi S, Maweni RM, Walsh K, McConn Walsh R, Javadpour M, et al. Signs and Symptoms of Acoustic Neuroma at Initial Presentation: An Exploratory Analysis. Cureus 2017;;9:e1846.
4. Early S, Rinnooy KC, Eggink M. Progression of Contralateral Hearing Loss in Patients With Sporadic Vestibular Schwannoma. Front Neurol 2020;11:796.
5. Carlson ML, Marston AP, Glasgow AE, Habermann EB, Sweeney AD, Link MJ, et al. Racial differences in vestibular schwannoma. Laryngoscope. 2016;126: 2128–33.
6. Koo M, Lai JT, Yang EY, Liu TC, Hwang JH. Incidence of Vestibular Schwannoma in Taiwan from 2001 to 2012: A Population-Based National Health Insurance Study. Ann Otol Rhinol Laryngol. 2018;127:694.
7. Carlson ML ,Habermann EB ,Wagie AE, Driscoll CL, Gompel JJV, Jacob JT, et al. The changing landscape of vestibular schwannoma management in the united states—a shift toward conservatism. Otolaryngol Head Neck Surg.2015;153:440-6.
8. Hunter JB, Francis DO, O'Connell BP, Kabagambe EK, Bennett ML, Wanna GB, et al. Single Institutional Experience With Observing 564 Vestibular Schwannomas: Factors Associated With Tumor Growth. Otol Neurotol. 2016, 37:1630-6.
9. Thapa PB, Shahi S, Jha RK, Shrestha D. Vestibular Schwannoma: An Experience in a Developing World. World J Oncol. 2019, 10:118-22.
10. Bartek Jr J, Förander P, Thurin E, Wangeird T, Henriksson R, Hesselager G, et al.: Short-term surgical outcome for vestibular schwannoma in sweden: a nation-wide registry study. Front Neurol . 2019;10:43.
11. Cioffi G, Yeboa D N, Kelly M, Patil N, Manzoor N, Greppin K, et al. Epidemiology of vestibular schwannoma in the United States, 2004–2016, Neuro-Oncology Advances. 2020;2:1.
12. Marinelli JP, Lohse CM, Carlson ML: Incidence of vestibular schwannoma over the past half-century: a population-based study of Olmsted County, Minnesota. Otolaryngol Head Neck Surg 2018;159:223-8.
13. Kshettry VR, Hsieh JK, Ostrom QT., Kruchko C, Barnholtz-Sloan JS., et al.: Incidence of vestibular schwannomas in the United States. J Neurooncol. 2015;124:223-8.
14. Hatch JL, Bauschard MJ, Nguyen SA, Lambert PR, Meyer TA, McRackan TR. National trends in vestibular schwannoma surgery: influence of patient characteristics on outcomes. Otolaryngol Head Neck Surg. 2018; 1:194599818765717.
15. Deberge S, Meyer A, Le Pabic, Peigne L, Morandi X, Godey B, et al. Quality of life in the management of small vestibular schwannomas: observation, radiotherapy and microsurgery.Clin Otolaryngol. 2018;43:1478-86.
16. Evans DG, Wallace AJ, Hartley C, Freeman SR, Lloyd SK, Thomas O, et al.: Familial unilateral vestibular schwannoma is rarely caused by inherited variants in the NF2 gene. . Laryngoscope. 2019;129:967-973.
17. Lawson McLean AC, Rosahl SK. Growth Dynamics of Intracranial Tumors in Patients with Neurofibromatosis Type 2. World Neurosurg. 2017;98:152–61.
18. Vanessa L. Merker, Annie Dai, Heather B. Radtke, Pamela Knight, Justin T. Jordan & Scott R. Plotkin. Increasing access to specialty care for rare diseases: a case study using a foundation sponsored clinic network for patients with neurofibromatosis 1, neurofibromatosis 2, and schwannomatosis. BMC Health Serv Res 2018;18:668.
19. Paldor I, Chen AS, Kaye AH. Growth rate of vestibular schwannoma. J Clin Neurosci. 2016;32:1-8.
20. Merker VL, Dai A, Radtke HB, Knight P, Jordan J T, Plotkin SR, et al. Increasing access to specialty care for rare diseases: a case study using a foundation sponsored clinic network for patients with neurofibromatosis 1, neurofibromatosis 2, and schwannomatosis.. BMC Health Serv Res. 2018;18:668.