Vestibular schwannoma appears to be very rare in a region of Sub-Saharan Africa

S. Ohaegbulam¹, O. Okwunodulu¹, C. Ndubuisi¹,³, W. Mezue¹,², M. Chikani¹,², S. Nkwerem¹, M. Ekuma¹

¹Neurosurgery Department, Memfys Hospital for Neurosurgery, ²Neurosurgery Department, University of Nigeria Teaching Hospital, ³Enugu State University Teaching Hospital, Enugu, Nigeria

E-mail: *S. Ohaegbulam - samcohaegbulam@hotmail.com; O. Okwunodulu - okwuife@gmail.com; C. Ndubuisi - chikandu@yahoo.com; W. Mezue - mezuec@hotmail.com; M. Chikani - markochikani@yahoo.com; S. Nkwerem - nkwerempatrick@yahoo.com; M. Ekuma - mekumarock@gmail.com

*Corresponding author

Received: 09 March 17  Accepted: 12 April 17  Published: 01 August 17

Abstract

Background: Vestibular schwannoma (VS) is a significant neurosurgical problem hence it enjoys a special attention at conferences and workshops. It accounts for about 8–10% of all intracranial tumors with an annual incidence of about 11–14 per million per year. Most VS are sporadic with 5–10% attributed to neurofibromatosis type 2 (NF2). However, VS is alleged to be rare in Africans and uncommon in African Americans, connoting a racial bias. To our knowledge, no study from sub-Saharan Africa has addressed this subject. The aim of this study is to determine the frequency of VS in a tertiary neurosurgical hospital in sub-Saharan Africa.

Methods: This is a retrospective study of all intracranial neoplasms and VS managed in a major tertiary hospital in sub-Saharan Africa from January 2003 to December 2015. Patients’ records and neuroimaging studies were reviewed retrospectively. Additionally, database of all cranial computed tomography (CT) and magnetic resonance imaging (MRI) done for various indications within the same period was searched retrospectively for “incidental” VS cases.

Results: Over the study period of 13 years, out of 612 cases of intracranial neoplasms, only three (0.49%) were VSs (two sporadic, one bilateral VS in NF2). A search for “incidental” cases of VS from a pool of 7475 cranial scans (CT: 5290; MRI: 2185), yielded none.

Conclusions: The findings strongly suggest that VS is very rare in the study population. It is hoped that other centers in Africa and beyond would perform similar studies.

Key Words: Intracranial tumors, geographical neurosurgery, neurofibromatosis, vestibular schwannoma
INTRODUCTION

Vestibular schwannoma (VS), commonly termed acoustic neuroma, arises on the eighth cranial nerve and accounts for about 8–10% of all intracranial tumors, with an annual incidence of about 11.8–14 per million per year. The designation “acoustic” is actually a misnomer as most are schwannomas that arise on either branch of the vestibular nerve. They are typically encapsulated benign slow growing tumors that negatively impact quality of life. VSs constitute approximately 60% of all schwannomas and roughly 90% of all intracranial schwannomas.

Hearing loss is the most common presenting symptom due to compression of adjacent cochlear nerve. Most VS are sporadic, but 5-13% are associated with NF2, which is an autosomal dominant disorder characterized by bilateral VS, other CNS tumors, and characteristic growth rate of VS. The etiology of the sporadic form is largely unknown and although several factors have been suspected, none has been decisively incriminated.

The incidence rates of VS vary worldwide from 1 to 20 cases per million inhabitants per year, and is reported to be on the increase globally. Although African studies are few, they suggest that VS is rare in Africans, connoting a racial bias. In an important study that captured the entire population of Denmark, the incidence of VS was 17.4/1 million inhabitants per year. A study in the USA found that the incidence of VS was 0.8 per 100,000 person-years. The incidence was similar among males and females, higher in whites than in nonwhites, lowest among the 0–19 year age group, and highest among the 45–64 year age group. VS is a significant health problem in many parts of the world for which no preventive measures are known. The tumor is often diagnosed too late to save hearing or even life. It is therefore no surprise that it is a subject of much clinical and pathological research producing voluminous publications. Neurosurgery and Otorhinolaryngology conferences allocate considerable time to VS. The American Hearing Research Foundation (AHRF) and other organizations have funded research on VS, particularly those leading to early detection before the onset of hearing loss.

The aim of this study is to determine the frequency of VS in a major neurosurgical hospital in south east Nigeria.

MATERIALS AND METHODS

This is a retrospective study of all cases of VS diagnosed at the study tertiary hospital over a 13-year period (1 January 2003 to 31 December 2015). The medical records, CT, MRI, and histopathology records were used. Additionally, database of all cranial CT and MRI done for various indications within the same period was searched for “incidental” cases of VS. The hospital is the main referral neurosurgical hospital serving the city (population 722,664) as well as all the neighboring cities and states with a catchment population of over 20 million. The hospital also offers CT, MRI, neurology, and neurosurgery services.

RESULTS

During the study period, 612 patients with intracranial tumors were diagnosed out of which there were two cases of sporadic VS and a case of bilateral VS or NF2 [Table 1].

Search for “incidental” VS

This was accomplished through the search of the database of all cranial CTs and MRIs performed at the hospital between 2003 and 2015. From the 7475 cranial scans (CT: 5290; MRI: 2185), no “incidental” cases of VS were found [Table 2].

DISCUSSION

Spanning over a period of 13 years, out of 612 cases of intracranial neoplasms, only three (0.49%) were VS. This was undoubtedly a rather small proportion of all intracranial neoplasms managed at the institution during the study period. The patient population consisted

| Table 1: Characteristics of vestibular schwannoma (VS) cases |
|--------------------|-----------------|-----------------|-----------------|
| Patient Characteristics | Patient #1 | Patient #2 | Patient #3 |
| Age in years | 64 | 28 | 24 |
| Sex | Fem | Male | Fem |
| Duration of symptoms | 10 year | 2 year | 3 year |
| Seizures | No | Yes | No |
| Headaches | Yes | Yes | Yes |
| GCS at presentation | 14 | 15 | 10 |
| Vision | NPL | NPL | HM |
| Hearing loss (deaf) | Left | Bilateral | Left |
| Facial nerve palsy | Left | None | Left |
| Other cranial nerve palsies | V | None | IX, X |
| Ataxia | Yes | No | Yes |
| Hemiparesis | Lt | None | Lt |
| MRI/CT findings CPA tumor | Large Lt | Bil. Lt> Rt | Large Lt |
| Histology/Diagnosis | VS | VS/NF2 | VS |

| Table 2: Search for “incidental” vestibular schwannoma (VS) |
|----------------|------------------|------------------|------------------|
| Investigation type | All scans | Cranial scans | No of VS |
| CT (2003-2015) | 8760 | 5290 | 00 |
| MRI (2009-2015) | 8029 | 2185 | 00 |
| Total | 16789 | 7475 | 00 |

Radiology records (2003-2015)
of black Africans of predominantly single ethnicity, suggesting that VS is rare in this part of Africa and may have a racial bias. An earlier study of intracranial tumors from the same area also reported no cases of VS.[17] However, the total number of tumors was relatively small and the non-availability, at that time, of CTs and MRIs in the country raised some doubts on the findings. The situation is very different now as those facilities have become available.

Schwannomas actually represent 8–10% of all intracranial neoplasms, but this frequency appears to have a racial bias.[13,20,26] In a review of the clinical characteristics of 4886 adult patients who underwent acoustic neuroma excision in the USA, it was reported that 85.4% were Caucasians while only 3.7% were African Americans.[13] This racial bias has been demonstrated in a study from South Africa that showed a low incidence of VS of approximately 0.3 per 100,000 population per year.[26] Simpson et al. (1990), in a study of 163 cases of primary intracranial neoplasms in black South Africans, reported that acoustic schwannomas accounted for 3.7%. Although this figure is low by world standards, it represented a far higher rate that had been previously reported in Africa.[21]

It is possible that asymptomatic lesions may account for the relative rarity of VS in this study and amongst black people in general. However, the zero “incidental” VS in this study despite the relatively large sample size suggests the rarity of VS in this study black population. This is in sharp contrast to the findings in the retrospective analysis of MRI scans by Lin et al.[12] in the University of California that yielded an incidence of 0.02% of incidental VS in their study population. Also, an MRI based study found cases of incidental VS to be 12% of the patients with VS,[19] while autopsy studies showed even higher incidence rates of VS.[29]

The question then arises as to why VS should be rare in this study population when the incidence is rising in other parts of the world. In the search for an answer, it is appropriate to examine some of the etiological factors that have been incriminated in the pathogenesis of VS which include loud noise and use of mobile phones.[4,5,19] It is well known that in Africa, loud noise is a major nuisance to the extent that laws have been enacted to confront and hopefully reduce it.[18,21] It is therefore reasonable to assume that there is abundance of this factor which then cannot be credited with the reduction in the incidence of VS. Some studies have reported weak association between VS and the use of mobile phones.[24] Mobile phones are in widespread use, the density which is the highest in Africa is quite close to Western figures. Other factors like smoking were shown to have inverse association while multiparous women have increased risk.[25] These factors if real, should have probably led to increased incidence of VS in the study population. However, the only proven etiology factor, just like other types of tumor, is exposure to radiation but no information is available on this in this locality.

A large nationwide cohort study in Denmark revealed that high level of education and high disposable income were associated with the occurrence of VS in both sexes.[22] This might be due to increased awareness which is usually aided by higher education and high disposable income that will facilitate seeking medical attention and the ability to pay for it. But then, Denmark has free Medicare to which all citizens have free access suggesting that awareness and delays in seeking help may be the problem and not high income. Interestingly, a similar study in the USA also arrived at similar conclusions that the largest social gradient in all brain tumor types was seen in VS with increased risk.[15] These factors may be relevant in our locality as well as other parts of Africa where poverty and illiteracy rates are still high.

VS causes impaired hearing and one would expect that anybody who experiences progressive deafness would seek medical help at the earliest opportunity. Educated people are more likely to recognize early deafness because of increased communication needs. Paradoxically, it is known that tumor size is not always directly proportional to the degree of hearing impairment since large VS may cause mild hearing loss while small tumors may cause marked deafness.

Late presentation is a common phenomenon in Africa hence intracranial tumors reach giant size before diagnosis. In a recent analysis of pituitary tumors, about 50% of the patients were already giant tumors by the time of presentation.[15] However, VS by virtue of its anatomical location will, in addition to hearing loss, produce other signs and symptoms of cerebellopontine angle (CPA) mass lesion such as ataxia, facial nerve paresis, vertigo, tinnitus, other cranial nerve palsies, and even obstructive hydrocephalus as the tumor increases in size. These signs and symptoms should ultimately force the patient to seek help, albeit at an advanced stage of the disease. This has not been the case with VS unlike other intracranial tumors, thereby adding credence to the belief that VS is truly rare in the study population.

Are there genetic factors to be considered as possible reasons for the rarity of VS in the study population? It is known that VS is due to loss of tumor suppressor gene on the long arm of chromosome 22. This is a somatic mutation in sporadic cases of VS, but in NF2 this is an inherited abnormality or a new mutation that is transmitted to the offspring.

Given the increase in the availability of neuroimaging and neurosurgical services in our locality as in other parts of Africa, the prevalence of VS has not displayed rising trends. A study of the global frequency of
Table 3: Global frequency of VS

| Author             | Year | Country | ICT   | VS as % of ICT |
|--------------------|------|---------|-------|----------------|
| Nakamura et al.    | 2011 | Japan   | 5448  | 9.9            |
| Cordera et al.     | 2002 | Italy   | 285   | 8.0            |
| Mehrzian et al.    | 2006 | Iran    | 3437  | 8.1            |
| Surawicz et al.    | 1999 | USA     | 20765 | 6.5            |
| Kuipers et al.     | 2012 | Suriname| 251   | 4.7            |
| Current study      | 2016 | Nigeria | 612   | 0.5            |

VS: Vestibular schwannoma, ICT: Intracranial tumors

VS in intracranial tumor series demonstrates this difference [12,10,14,16,28]

CONCLUSION

Vestibular schwannoma seems to be rare in the study population. This finding strengthens the belief that VS exhibits racial bias with relative rarity in blacks. If this is confirmed from other researchers, the quest for the factors that protect some people from VS should be intensified as this could ultimately lead to prevention and even cure.

Acknowledgement

The authors gratefully acknowledge the contributions of the radiology department, the pathologists for reporting on the excised tumors and the ethics committee of the hospital.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Abaza MM, Makariou E, Armstrong M, Lalwani AK. Growth rate characteristics of acoustic neuromas associated with Neurofibromatosis type 2. Laryngoscope 1996;106:674-9.
2. Cordera S, Bottachi E, D’Alessandro G, Machado D, Gonda F, Corso G. Epidemiology of primary intracranial tumours in NW Italy, a population based study: Stable incidence in the last two decades. J Neurol 2002;249:281-4.
3. Curry WT Jr, Barker FG 2nd. Racial, ethnic and socioeconomic disparities in the treatment of brain tumours. J Neurooncol 2009;93:25-39.
4. Edwards CG, Schwartzbaum JA, Lönn S, Ahlbom A, Feychting M. Exposure to loud noise and risk of acoustic neuroma. Am J Epidemiol 2006;163:327-33.
5. Edwards CG, Schwartzbaum JA, Nise G, Forsén UM, Ahlbom A, Lönn S, et al. Occupational noise exposure and risk of vestibular schwannoma. Am J Epidemiol 2007;166:1252-8.
6. Evans DG, Moran A, King A, Saeed S, Gurusinghe N, Ramsden R. Incidence of vestibular schwannoma and Neurofibromatosis 2 in the North West of England over a 10-year period; higher incidence than previously thought. Otol Neurotol 2005;26:93-7.
7. Hoffman S, Propp JM, McCarthy BJ. Temporal trends in incidence of primary brain tumours in the United States, 1985–1999. Neuro Oncol 2006;8:27-37.
8. Howitz MF, Johansen C, Tso M, Charabi S, Olsen JH. Incidence of vestibular schwannoma in Denmark, 1977-1995. Am J Otol 2000;21:690-4.
9. Jeyakuma A, Seth R, Brickman TM, Dutcher P. The prevalence and clinical course of patients with ‘incidental’ acoustic neuromas. Acta Otolaryngol 2007;127:1051-7.
10. Kuipers SE, Kaffluddin E, Joe RT, Peerdeman S. Incidence and treatment of Central Nervous System Tumours in Suriname. World Neurosurg 2013;80:e79-83.
11. Lanser MJ, Sussman SA, Frazer K. Epidemiology, pathogenesis, and genetics of acoustic tumours. Otolaryngol Clin North Am 1999;22:499-520.
12. Lin D, Hagerty JL, Fischbein NJ, Jackler RK. The prevalence of ‘Incidental’ Acoustic Neurroma. Arch Otolaryngol Head Neck Surg 2005;131:241-4.
13. McClelland S III, Guo H, Okuyemi KS, Morbidity and mortality following acoustic neuroma excision in the United States: Analysis of racial disparities during a decade in the radiosurgery era. Neurooncol 2011;13:1252-9.
14. Mehrzian M, Rahmat H, Paviar P. Epidemiology of primary intracranial tumours in Iran, 1978-2003. Asian Pac J Cancer Prev 2006;7:284-8.
15. Mezeu WC, Ohaegbulam SC, Chikani MC, Achebe DN. Management of Giant Pituitary Tumours Affecting Vision in Nigeria. World Neurosurg 2012;77:606-9.
16. Nakamura H, Makino K, Yano S, Kuratsu J. Epidemiological study of primary intracranial tumours: A regional survey in Kumamoto Prefecture in southern Japan-20-year study. Int J Clin Oncol 2011;16:314-21.
17. Ohaegbulam SC, Sagdeo N, Ikerionwu S. Intracranial Tumours in Enugu, Nigeria. Cancer 1980;46:2322-4.
18. Olayinka OS. Effective noise control measures and sustainable development in Nigeria. World J Environmental Engineering 2013;3:5-15.
19. Preston-Martin S, Thomas DC, Wright WE, Henderson BE. Noise trauma in the aetiology of vestibular schwannomas in men in Los Angeles County, 1978–1985. Br J Cancer 1989;59:783-6.
20. Propp JM, McCarthy BJ, Preston-Martin S. Descriptive epidemiology of vestibular schwannomas. Neuro Oncol 2006;8:11.
21. Saadu AA, Onyeonwu RO, Ayorinde EO, Ogisi FO. Road traffic noise survey and analysis in some major Urban centres in Nigeria. Noise Control Eng J 1998;46:146-58.
22. Sarma S, Sekhar LN, Schessel DA. Nonvestibular schwannomas of the brain, a 7-year experience. Neurosurgery 2002;50:437-49.
23. Schoemaker MJ, Swerdlow AJ, Auvinen A, Christensen HC, Feychting M, Johansen C, et al. Medical history, cigarette smoking and risk of vestibular schwannoma: An international case–control study. Int J Cancer 2007;120:103-10.
24. Schüz J, Jacobsen R, Olsen JH, Boice JD Jr, McLaughlin JK, Johansen C. Cellular telephone use and cancer risk: Update of a nationwide Danish cohort. J Natl Cancer Inst 2006;98:1707-13.
25. Schüz J, Marianne S, Søren H, Sven-Eric S, Per CT, Christoffer J. Sociodemographic factors and vestibular schwannoma: A Danish nationwide cohort study. Neuro Oncol 2010;12:1291-9.
26. Seedat RY, Claassen AJ, Mol DA. Incidence and management of acoustic neuromas in South Africa. Otol Neurotol 2002;23:947-49.
27. Simpson RH, Sparrow OC, Duffield MS. Cerебеллопонтине angle tumours in black South Africans-how rare are acoustic schwannomas? S Afr Med J 1990;78:11-4.
28. Surawicz TS, McCarthy BJ, Kupelian V, Kujich PJ, Bruner JM, Davis FG, et al. Descriptive epidemiology of primary brain and CNS tumours: Results from the Central Brain Tumour Registry of the United States, 1990-1994. Neuro Oncol 1999;1:14-25.
29. Tos M, Stangerup SE, Cayé-Thomasen P, Tos T, Thomsen J. What is the real incidence of vestibular schwannoma? Arch Otolaryngol Head Neck Surg 2004;130:216-20.
30. Weller RO, Cervos-Navarro J. Pathology of peripheral nerves. In: Gowing NFC, editor. Tumour Histopathology. London: Butterworths; 1977.