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Original Article

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

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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

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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.[6] 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[30] and roughly 90% of all intracranial schwannomas.[22]

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.[1,6] 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.[7,8,11] Although African studies are few, they suggest that VS is rare in Africans, connoting a racial bias.[17,26,27] In an important study that captured the entire population of Denmark, the incidence of VS was 17.4/1 million inhabitants per year.[29] 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.[13] 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	Patent #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.[27]

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,[9] 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.[23] 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.[25] 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.[3] 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 30% 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*.[16]	2011	Japan	5448	9.9
Cordera *et al*.[2]	2002	Italy	285	8.0
Mehrazin *et al*.[14]	2006	Iran	3437	8.1
Surawicz *et al*.[28]	1999	USA	20765	6.5
Kuipers *et al*.[10]	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 [Table 3].[2,10,14,16,28]

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.

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Conflicts of interest
There are no conflicts of interest.

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