Epidemiology and Clinical Features of Vestibular Schwannoma in Manitoba, Canada

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ABSTRACT: The incidence of vestibular schwannoma (acoustic neuroma) in Manitoba, Canada was reviewed. From 1987 through 1991, 71 tumors were diagnosed in 69 patients. The overall annual incidence rate for both sexes was 1.27/100,000 with male and female annual incidences of 1.31/100,000 and 1.24/100,000 respectively. For males, there was an early peak in the age group 30 - 39 years (2.1/100,000). Following the fifth decade, the incidence for males plateaued (2.7 - 3.6/100,000). For females, the incidence increased with age up to age 60 - 69 years (4.1/100,000). This was followed by a progressive decline in the incidence. Although the incidence of vestibular schwannoma was relatively high, the clinical features were not unlike those previously reported in the literature.

RÉSUMÉ: Épidémiologie et aspects cliniques du schwannome vestibulaire au Manitoba, Canada. Nous avons revu l'incidence du schwannome vestibulaire (neurinome acoustique) au Manitoba, Canada. De 1987 à 1991, 71 tumeurs ont été diagnostiquées chez 69 patients. L'incidence annuelle pour les deux sexes était de 1.27/100,000, l'incidence annuelle chez l'homme et la femme étant de 1.31/100,000 et 1.24/100,000 respectivement. Chez l'homme, nous avons noté un pic précoce entre 30 et 39 ans (2.1/100,000) et un plateau après la cinquième décade (2.7-3.6/100,000). Chez la femme, l'incidence augmentait avec l'âge jusqu'à 60-69 ans (4.1/100,000), puis diminuait progressivement. Bien que l'incidence du schwannome vestibulaire soit relativement élevée, ses manifestations cliniques n'étaient pas différentes de celles rapportées antérieurement dans la littérature.

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The epidemiology of vestibular schwannoma (acoustic neuroma) has been evaluated in two ways, either as frequency or incidence data. Previous studies report a frequency of vestibular schwannoma relative to other primary intracranial neoplasms of 2 - 8%.[1-11] Autopsy series report a frequency of occult vestibular schwannoma of 0 - 2.4%.[12-15] In general, the tumors detected at autopsy were small, measuring only 0.8 - 5 mm in size and restricted to the internal auditory canal.

The incidence of vestibular schwannoma derived from population-based studies shows considerable variation.[6,7,10,11,16,17] A Denmark study (1976 - 1983) found the incidence of clinically symptomatic vestibular schwannoma to be 0.78/100,000.[16] A study from the Wessex region of England (1965 - 1974) found an incidence of 0.48/100,000.[7] An Israeli study (1960 - 1964) found an incidence of 0.40/100,000[11] while a Connecticut survey (1935 - 1964) reported an incidence of only 0.08/100,000.[10] The Manitoba Primary Intracranial Tumor Study (1980 - 1985) reported an incidence of 0.51/100,000 for vestibular schwannoma.[6] We are unaware of any population based study that examined the incidence of vestibular schwannoma following the widespread introduction of computerized tomographic (CT) and magnetic resonance (MR) imaging. In the present study, therefore, we examined the incidence of vestibular schwannoma in the population of Manitoba, Canada over a 5-year interval, 1987 through 1991, during which time both CT and MR imaging were readily available. In Manitoba, the population has remained constant at approximately one million. The number of people emigrating from or immigrating to Manitoba was minimal, therefore providing an ideal population to examine the incidence of vestibular schwannoma. In addition, all individuals in Manitoba with suspected vestibular schwannoma are referred to one of two teaching hospitals in Winnipeg for their investigation and treatment. Furthermore, the clinical features of the patients with vestibular schwannoma were analyzed.

Data Sources and Methods

Case Material

In this retrospective study, vestibular schwannoma operated on or radiologically diagnosed were reviewed. Radiological

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diagnosis was based on an enlarged porus acousticus plus adjacent enhancing neoplasm. In many cases, computed tomography (CT) was supplemented with magnetic resonance (MR) imaging. Residency in Manitoba at the time of diagnosis was necessary for inclusion in the study. The neurosurgical operating room log books for both teaching hospitals were reviewed. To complete case ascertainment, radiological files were examined in which a possible diagnosis of vestibular schwannoma was entertained. Medical charts were reviewed, extracting the clinical, surgical, histological and audiological data.

General Population Information

Manitoba has a population of slightly more than one million. This is comprised mainly of ethnic groups from European, Asiatic, American Indian and Inuit stock. The people of Manitoba have equal access to health care facilities through a government-sponsored universal health care system. The age distribution of the population of Manitoba is not unlike that of Canada as a whole for the years 1987 - 1991.

Methods

The average annual crude incident rate as well as age and sex specific incidence rates for vestibular schwannoma were computed utilizing the average population over the 5-year period $[1{,}100 \pm 19] \times 10^3$. Radiographic examinations were re-evaluated to substantiate and expand the original reports. Tumor size was determined by measuring maximum diameter from either CT or MR images. The histopathological criteria used to define vestibular schwannoma were derived according to the World Health Organization classification.[18]

Results

Incidence and Age and Sex Distribution

During the 5-year study interval, the diagnosis of vestibular schwannoma was made in 69 patients. There were 34 females (49%) and 35 males (51%). Bilateral vestibular schwannoma was found in two patients giving a total of 71 tumors. The overall average annual incidence rate for both sexes was 1.27/100,000 with male and female average annual incidence rates of 1.31/100,000 and 1.24/100,000 respectively. The average annual incidence rate for vestibular schwannoma by age and sex is shown in Figure 1. The most noteworthy aspect of the incidence curves was the identification of an early peak for males in the 30 - 39 age group (2.1/100,000). Following the fifth decade, the incidence for males plateaued (2.7 - 03.6/100,000). For females, the incidence increased with age up to the 60 - 69 age group (4.1/100,000) followed by a progressive decline in the incidence.

Clinical Features

The history and physical findings of patients with vestibular schwannoma as a function of tumor size are presented in Table 1. The most common symptom was hearing loss (97%). Fifty-nine patients had unilateral hearing loss and eight had bilateral asymmetrical hearing loss. There were four cases of sudden hearing loss (6%). Other common symptoms include: tinnitus (54%), ataxia/unsteady gait (44%), headache (28%), facial paresthesia (23%), and vertigo/dizziness (20%). Hearing loss and tinnitus are common symptoms for all tumor sizes. Patients with tumors less than or equal to 1 cm in size had the least number of symptoms.

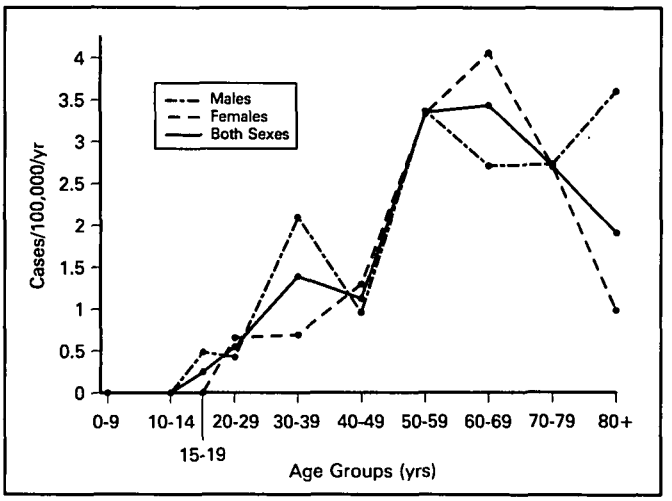

Figure 1 — Average annual incidence rates for vestibular schwannoma by age and sex. Males show an early peak in the 30 - 39 year age group following which the incidence increases with age up to the 50 - 59 year age group. For females, the incidence increases with age up to the 60 - 69 year age group followed by a progressive decline in the incidence.

Unilateral or asymmetric sensorineural hearing loss was found in 97% of the patients, ataxia in 35%, facial sensory impairment in 26%, and nystagmus in 23%. None of the patients with tumor size less than or equal to 1 cm had cranial nerve V or VII deficits or cerebellar signs. Unilateral sensorineural hearing loss and nystagmus were common physical findings and were independent of tumor size.

The size distribution of 71 vestibular schwannomas based on CT or MR imaging observations are presented in Table 2. Ten tumors were ≤ 1.0 cm (14%), 40 were 1.1 - 2.5 cm (56%), 17 were 2.6 - 4.0 cm (24%), and 4 were > 4.0 cm (6%). Homogeneous vs. non-homogeneous enhancement was not a differentiating feature. Fourth ventricular compression with or without obstructive hydrocephalus was dependent on tumor size. Cystic degeneration was observed in six of the tumors larger than 2.6 cm.

Pure tone audiogram results as a function of tumor size for 48 patients are presented in Table 3. Hearing loss was the average response in decibels at 500, 1,000 and 2,000 Hz (American National Standards Institute 1969).[19] Mild hearing loss [26 - 40 dB] (31%) was most common followed by moderate [41 - 55 dB] (23%), moderate - severe [56 - 70 dB] (23%), severe [71 - 90 dB] (11%) and profound [> 90 dB] (9%). One patient (3%) had normal hearing. Hearing loss was independent of tumor size. Speech discrimination scores were distributed from 0 to 100%, with the results as a function of tumor size are presented in Table 4. Similar to the hearing loss results, the scores were independent of tumor size.

Sixty of the patients with vestibular schwannoma underwent surgical excision of their neoplasms. All of these tumors were histologically diagnosed as benign schwannomas.

Discussion

Limitations of the Data

It is estimated that more than 95% of clinically symptomatic vestibular schwannomas diagnosed pre-mortem in Manitoba

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Table 1A. Presenting Symptoms Relative to Tumor Size in 69 Patients with Vestibular Schwannoma

Symptoms	≤ 1 cm (n = 9) Per cent	1.1 - 2.5 cm (n = 39) Per cent	2.6 - 4.0 cm (n = 17) Per cent	> 4 cm (n = 4) Per cent
Hearing Loss	100	100	94	75
Tinnitus	56	54	70	100
Ataxia/Unsteady Gait	11	36	52	50
Headache		26	29	75
Facial Paresthesia		18	47	25
Vertigo/Dizziness	11	15	35	25
Retro-auricular Fullness		13	18	25
Visual/Impairment		10	18	50
Nausea/Vomiting	11	5	5	25
Facial Pain		5	18	
Difficulty Swallowing		5		
Syncope			6	
Diplopia		3	12	
Personality Change		3		
Facial Paresis			6	
Extremity Paresis				25
Otalgia		3		

Table 1B. Physical Findings Relative to Tumor Size in 69 Patients with Vestibular Schwannoma

Physical Findings	≤ 1 cm (n = 9) Per cent	1.1 - 2.5 cm (n = 39) Per cent	2.6 - 4.0 cm (n = 17) Per cent	> 4 cm (n = 4) Per cent
Sensorineural Hearing Loss	100	100	94	75
Ataxia		23	65	100
Cranial Nerve V Deficit		26	41	25
Nystagmus	11	18	29	75
Cranial Nerve VII Deficit		5	24	
Cranial Nerves IX, X Deficit		8	12	
Papilledema			18	25
Cranial Nerve VI Deficit			12	
Memory Impairment		3	6	
Cranial Nerve XII Deficit		3		
Visual Field Deficit		3		
Cranial Nerve IV Deficit			6	

during the years 1987 through 1991 have been identified in this study. It is possible that some tumors may have been diagnosed and/or treated outside of Manitoba and thus are not included in the study. This number is likely very small since the Government sponsored health care system requires that such patients be initially diagnosed in Winnipeg where the provincial CT and MR imaging facilities are located and would thus be available to the study. Peripheral Winnipeg hospitals and the City of Brandon did not acquire CT imaging capability until after the study interval.

Table 2. CT and MR Findings in 69 Patients with 71 Vestibular Schwannomas

Findings	Number of Patients	Per cent
Enlarged Porus	50	70
Enhancement		
Homogeneous	46	65
Non-Homogeneous	25	35
Fourth Ventricle		
Compression/Shift	17	24
Hydrocephalus	7	10
Cystic Component	6	8
Tumor Size		
< 1.0 cm	10	14
1.1 - 2.5 cm	40	56
2.6 - 4.0 cm	17	24
> 4.0 cm	4	6

Table 3. Hearing Loss in 48 Patients as a Function of Tumor Size

Hearing Loss (Average Response at 500, 1000, 2000 Hz)	≤ 1 cm (n = 9) Per cent	1.1 - 2.5 cm (n = 30) Per cent	2.6 - 4.0 cm (n = 9) Per cent
0 - 25	-	5	-
26 - 40	38	25	43
41 - 55	-	30	29
56 - 70	38	15	29
71 - 90	25	10	-
91+	-	15	-

Table 4. Speech Discrimination in 41 Patients as a Function of Tumor Size

Speech Discrimination Per Cent	≤ 1 cm (n = 9) Per cent	1.1 - 2.5 cm (n = 25) Per cent	2.6 - 4.0 cm (n = 7) Per cent
90 - 100	25	13	17
70 - 89	13	33	-
40 - 69	13	33	50
10 - 39	38	7	-
0 - 9	13	13	33

Incidence of Vestibular Schwannoma

Few studies have examined the clinical incidence of vestibular schwannoma and the majority of these pre-date the widespread use of CT and MR imaging. Post-mortem series have revealed a frequency of occult vestibular schwannoma of 0 - 2.4%. It should be emphasized that autopsy series are biased by population selection. In the autopsy series reported by Leonard and Talbot,[12] many of the patients were derived from the neurosurgical service and had audiological studies. Autopsy series are also biased as they tend to examine an older population and therefore are not representative of the population as a whole. Tumor size within the post-mortem studies is well below that generally expected to manifest clinical symptoms and/or

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signs. Loss of regions of the long arm of chromosome 22 has been observed in large clinically symptomatic vestibular schwannoma.[20] These regions may subtrend a tumor-suppressor gene, inactivation of which results in stimulation of cell growth.[20,21] The increasing frequency of vestibular schwannoma with age implies that the loss of such a suppressor gene is age-dependent. It would be interesting to speculate that small tumors such as those reported in the post-mortem series lack chromosome 22 changes and therefore are not of a similar biological nature to those reported in studies examining clinically symptomatic tumors.

The average annual incidence rate for vestibular schwannoma reported in the present study is higher than that obtained in previous population-based clinical studies.[6,7,10,11,16,17] This higher incidence likely reflects the impact of technological advancement on the diagnosis of vestibular schwannoma. The Manitoba Primary Intracranial Tumor Study (1980 - 1985) reported an average annual vestibular schwannoma incidence rate of 0.29/100,000 and 0.76/100,000 for males and females respectively. Coincident with that publication, the number of CT scanners in Winnipeg increased from two to five. Furthermore, access to MR imaging became available. Although tumor size has been constant over the five-year interval of the present study, the mean tumor size is less than that previously reported by us (2.21 ± .11 vs. 2.91 ± .16 cm).[6] All previous population studies were published prior to the introduction of MR imaging.

A striking feature of this study is the occurrence of an early incidence peak for males. This may reflect more frequent auditory tests in occupations with excessive noise exposure which are more likely to employ males. The trend towards a decreasing incidence in females after the seventh decade implies trophic interaction between vestibular schwannoma and the endocrinological milieu unique to women. Vestibular schwannoma rarely becomes symptomatic before the onset of puberty and the tumors have been shown to have both estrogen and progesterone receptors.[22-24] Furthermore, pregnancy may exacerbate symptoms and increase the growth rate of tumors in women with neurofibromatosis type 2.[25,26]

Clinical Features

Clinical features among the patients with vestibular schwannoma in the present study are similar to those previously reported.[27-30] The high percentage of patients with asymmetrical sensorineural hearing loss emphasizes the importance of excluding the diagnosis of vestibular schwannoma in such a patient population. Sudden hearing impairment likely reflects vascular occlusion.[31] Increasing cerebellar and brainstem compression such as that associated with larger tumors accounts for the increased number of clinical signs and symptoms among this subgroup.

The increasing use of CT and MR imaging has diagnosed a higher percentage of tumors. In the previously reported Manitoba study (1980 - 85), the average size of vestibular schwannoma was 2.91 ± .16 compared to 2.21 ± .11 cm in the present study. Furthermore, patients in the initial Manitoba study were more likely to present with evidence of brain stem compression with or without associated hydrocephalus compared to the present study. Diagnosis of small tumors such as those found in the present study has only been possible through the use of MR imaging.

A relationship between average pure tone hearing loss or speech discrimination with the size of tumor was not demonstrated. This suggests that the intracanalicular component of the neoplasm and its compressive effects on the cochleovestibular nerve, the blood supply to the nerve or the blood supply to the cochlea, is the main determinant of hearing impairment. The potentially much larger intracranial component seems to contribute less to the hearing deficit.

In summary, this study has demonstrated the epidemiology and clinical features of vestibular schwannoma in a stable population that is representative of the population of Canada.

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