The Laryngoscope
Lippincott Williams & Wilkins, Inc.

Sublocalization and Volumetric Growth Pattern of Intracanalicular Vestibular Schwannomas

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Objective: **None of several previous reports on the growth pattern of vestibular schwannomas (VS) have dealt with the sublocalization and volumetric growth pattern of intracanalicular tumors. This paper reports such data from 196 patients.** ***Study Design:*** **All VS patients have been registered prospectively at one center in Denmark since 1975. Data on intracanalicular tumors were drawn from the database, yielding 196 patients with a diagnostic and at least one control magnetic resonance imaging scan. All images were retrieved and the tumor sublocalization, size, and growth rate determined.** ***Results:*** **The majority (50%) of the tumors was located centrally in the internal auditory canal (IAC), whereas 31% were porus-near and 19% fundus-near. Of the 196 tumors, 88 (45%) displayed growth, 20 (10%) shrinkage, and 88 (45%) remained unchanged. Thirty-eight (19%) tumors grew to extrameatal extension. Growth occurred only within 5 years after diagnosis. In the 88 growing tumors, the mean absolute growth rate was 111mm^3/year and the relative rate 114%/volume/year. The occurrence of IAC expansion at diagnosis was higher for tumors displaying subsequent shrinkage. Growth occurrence and rate, IAC expansion, and progression to extrameatal extension were not related to tumor sublocalization.** ***Conclusion:*** **Most intracanalicular VS are located centrally in a nonexpanded IAC at diagnosis. Growth occurs within 5 years after diagnosis in up to 45% of the tumors, although only 19% extend into the cerebellopontine angle. IAC expansion, growth occurrence, and rate are not related to tumor sublocalization. These findings justify primary observation of all purely intracanalicular tumors, unless realistic hearing preservation is intended.** ***Key Words:*** **Acoustic neuroma, intrameatal, natural history.**

Laryngoscope, **116:1131–1135, 2006**

INTRODUCTION

The literature contains several reports on the growth pattern of vestibular schwannomas (VS).[1–11] These reports document that some tumors grow continuously, whereas other tumors grow to a certain size followed by stagnation or even shrinkage. Most studies have shown a percentage of growing tumors from 30% to 55%.

The incidence of diagnosed VS is increasing, caused at least in part by an increased number of magnetic resonance imaging (MRI) scanners.[12,13] Because MRI allows visualization of smaller tumors than computed tomography (CT), the increased incidence is primarily a question of an increased detection of small and often exclusively intracanalicular tumors.

Only a few of the aforementioned growth studies have touched on the growth of these intracanalicular VS,[2,9,11] and in one of these reports only by somewhat rough measures (e.g., growth including extrameatal expansion into the cerebellopontine angle).[11] Thus, specific details of tumor sublocalization and the occurrence and rate of growing intracanalicular tumors in an unselected patient material are not available in the literature.

This paper reports data on the sublocalization and growth of intracanalicular VS registered in 196 patients primarily allocated to observation by repetitive MRI. The patient group represents all patients diagnosed and prospectively registered with a sporadic, unilateral, and exclusively intracanalicular VS in Denmark during the period 1976 to 2004 and with at least two sufficient MRI scans available at data analysis.

MATERIALS AND METHODS

All patients diagnosed with a cerebellopontine angle tumor resembling a VS have been registered prospectively in a database at one center in Denmark (5.2 million inhabitants) since 1975. In November 2004, data on all sporadic, unilateral, and purely in-

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Editor's Note: This Manuscript was accepted for publication February 28, 2006.

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DOI: 10.1097/01.MLG.0000217528.37106.2D

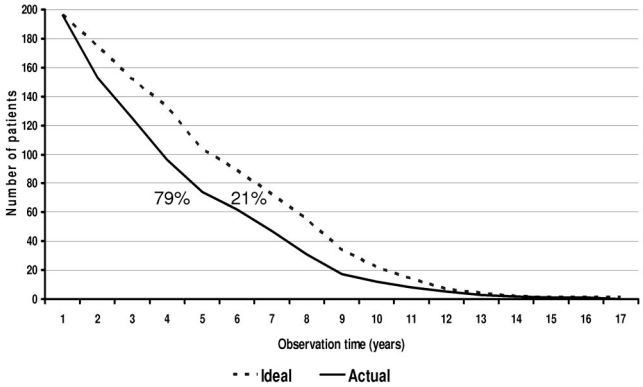

Fig. 1. Actual and ideal observation in 196 patients with an intrameatal vestibular schwannoma.

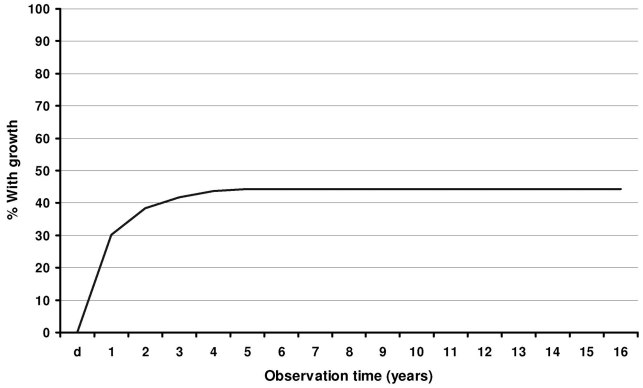

Fig. 2. Nelson-Aalen plot depicting risk of growth with increasing observation time in 196 intrameatal vestibular schwannomas.

tracanalicular tumors were drawn from the base, yielding 325 patients. Fifty-two patients diagnosed by a CT scan were excluded, leaving 273 patients diagnosed by MRI. All diagnostic and control images on these patients were retrieved and the sublocalization and three-dimensional size of the tumor determined by measurement. In addition, the images were analyzed for tumor-induced expansion of the internal auditory canal (IAC) (tumor filling the radial aspect of an enlarged canal compared with the contralateral side). A control MRI was not available in 61 patients (22 patients died before the first follow-up MRI, a control MRI had not been performed in 20 patients, and the images from 19 patients could not be retrieved), and one of the three tumor dimensions was not present on the MRI in 16 patients (the coronal projection was missing), yielding 196 patients with at least two sufficient MRI scans for volumetric determination of tumor size, growth frequency, and rate.

Most tumors had an ellipsoid shape, and the volume was calculated as a product of the three dimensions, using the formula for an ellipsoid: $volume = 1/6 \times 3{,}14 \times d1 \times d2 \times d3$. An increase of more than 2 mm in any diameter was defined as growth and a decrease of more than 2 mm as shrinkage.

Tumor sublocalization categories were defined as either fundus-near (no liquor between the tumor and the fundus and liquor between the tumor and the porus on T2-weighted images), central (liquor, or no liquor in tumors filling the entire canal, between the tumor and the fundus and between the tumor and the porus), or porus-near (no liquor between the tumor and the porus and liquor between the tumor and the fundus). Median age of the 196 patients was 58 (range 15–77) years at diagnosis, and the male to female ratio was 1.31. The mean observation period between first and last MRI was 4.4 (range 1–17) years. The cumulated actual observation was 79% of the ideal observation (Fig. 1).

The nonparametric $\chi 2$, Fisher's exact, Mann-Whitney, and Kruskal-Wallis tests were used for statistical analyses, and $P < .05$ was chosen as the level of significance. A Nelson-Aalen plot was used for calculation of the cumulated risk of growth because of the variable length of the observation period (high number of censored data) (Fig. 2).

RESULTS

Tumor Size and Growth

Most tumors were of a medium size for a purely intracanalicular lesion (χ^2, $P < .0001$), and most tumors did not lead to expansion of the osseous IAC at diagnosis (χ^2, $P < .0001$) (Table I). The overall mean tumor size was 118 (range 14–461) mm^3 at diagnosis and 415 (range 25–2,114) mm^3 at the last MRI (Table II). Of the 196 tumors, 88 (45%) displayed growth, 20 (10%) displayed shrinkage, and 88 (45%) remained unchanged during the observation period (Fig. 2) (Table I). Thirty-eight (19%) tumors grew to extrameatal extension, whereas 158 (81%) remained purely intrameatal (Table I). No tumor growth occurred after the fifth year of observation (Fig. 2).

In the 88 growing tumors, the mean absolute growth rate was 111 (range 11–666) mm^3/year, and the mean relative growth rate was (range 2–660) 114%/volume/year (Table II).

The occurrence of IAC expansion at the diagnostic MRI was significantly higher for tumors displaying subsequent shrinkage during the following observation period (Fisher´s exact test, $P = .00143$) (Table III).

There were no significant differences in tumor size or growth pattern between male and female patients or between different age groups. Growth occurrence and rate did not correlate with diagnostic tumor size.

Tumor Sublocalization

The majority (50%) of the 196 tumors were located centrally in the IAC (χ^2, $P < .0001$), whereas 61 (31%) were porus-near and 37 (19%) fundus-near (Table I). The diagnostic size of the fundus-near tumors was significantly smaller than the central tumors (Mann-Whitney, $P < .0451$) (Table II). There were no significant differences in sublocalization between male and female patients or between different age groups.

Sublocalization and Growth

Growth occurrence and rate, IAC expansion, and progression to extrameatal extension were not related significantly to tumor sublocalization (Tables II and III). Although the numbers point toward a higher occurrence of growth, IAC expansion, and growth to extrameatal extension for the central tumors (Tables II and III), these apparent differences were without statistical significance.

DISCUSSION AND CONCLUSION

From this first detailed study on the sublocalization and growth pattern of intracanalicular VS, we can conclude that most tumors are of a medium size for a purely intracanalicular lesion, and most tumors are located cen-

TABLE I.
Various Characteristics of 196 Intrameatal Vestibular Schwannomas at Diagnosis and at Last Evaluation.

		Diagnostic MRI		Last MRI	
		n	%	n	%
Largest intrameatal diameter (mm)	1–5	35*	18	29	
	6–10	134*	68	99	
	>10	27*	14	29	
Largest extrameatal diameter (mm)	1–5	–	–	20	51
	6–10	–	–	13	33
	>10	–	–	6	15
IAC expansion	Yes	36¤	18	71	36
	No	160¤	82	125	64
Tumor sublocalization at diagnosis	Fundus	37#	19	–	–
	Central	98#	50	–	–
	Porus	61#	31	–	–
Extrameatal extension	Yes	0	0	38	19
	No	196	100	158	81
Volume	Decreased	–	–	20	10
	Unchanged	–	–	88	45
	Increased	–	–	88	45
Mean volume		112 mm^3		242 mm^3	

¤*# χ^2, $p < .0001$
MRI = magnetic resonance imaging; IAC = internal auditory canal.

trally in the canal. The diagnostic volume of these central tumors is larger than the tumors located in the fundus. The cumulated risk of growth is 45% overall, which is higher than the 34% found in our material on tumors with an extrameatal component at diagnosis.[11] However, only 19% of the intracanalicular tumors grow into the cerebellopontine angle. Tumor growth occurs predominantly within the first few years after diagnosis, which is equivalent to tumors with an extrameatal component.[2,11] Our data showed that no intracanalicular tumors grow after 5 years of observation with an unchanged size. The diagnostic size is not related to sex or age, and the occurrence and rate of growth are not related to tumor sublocalization, diagnostic size, sex, or age. The latter is in agreement with a recent publication addressing potentially predictive parameters for tumor growth.[14]

Bias of Growth Data

The main strength of the present data are the prospective and consecutive one-center registration of all patients diagnosed with a VS during the period 1976 to 2004 in Denmark, with a population of 5.2 million inhabitants. The data are thus without patient referral bias. Because all the present patients diagnosed with a purely intracanalicular VS by MRI have been allocated primarily to observation, the material also is without patient selection bias. The referral and patient selection bias is a problem in a vast majority of previously published papers on VS growth, and this may be the main explanation for the surprising variability of reported growth occurrences (see below).[1–11]

Our sublocalization and growth data are biased by the fact that 52 patients diagnosed by a CT scan were excluded because of the inaccuracy of this type of imaging. Furthermore, a control MRI was not available in 61 patients (22 patients died before the first follow-up MRI, a control MRI had not been performed in 20 patients, and the images from 19 patients could not be retrieved). Last, one of the three tumor dimensions was not accessible in 16 patients because the coronal MRI projection was missing.

TABLE II.
Tumor Size and Growth Rates in Relation to Tumor Localization within the Internal Auditory Canal.

	n	Mean Volume First MRI, mm^3	Mean Volume Last MRI, mm^3	Mean Absolute Growth, mm^3/yr	Mean Relative Growth, %/yr	IAC Expansion First MRI (%)	IAC Expansion Last MRI, (%)	Growth to Extrameatal Extension (%)
Fundus	13	77 (24–131)*	294 (84–1295)	90 (14–579)	133 (4–579)	15	46	23
Central	50	126 (19–461)*	468 (25–2114)	122 (11–666)	106 (3–571)	20	62	54
Porus	25	123 (14–330)	372 (79–1573)	101 (14–502)	119 (2–660)	4	44	32
All	88	118 (14–461)	415 (25–2114)	111 (11–666)	114 (2–660)	15	55	43

*Mann-Whitney, $P < .0451$.

TABLE III.
Occurrence of Volume Changes and Internal Auditory Canal Expansion (IAC) in Relation to Tumor Sublocalization.

		Decreased		Unchanged		Increased	
	n	n	%	n	%	n	%
Fundus	37	4	11	20	54	13	35
Central	98	11	11	37	38	50	51
Porus	61	5	8	31	51	25	41
IAC expansion	Yes	10	50*#	13	15*	13	15#
	No	10	50	75	85	75	85
Total	196	20	10	88	45	88	45

*# Fisher´s exact test, $P = .00143$.

Comparison with Previously Published Data

Three previous studies have touched on growth of purely intracanalicular VS. Glasscock et al.[2] surveyed 10 patients over a mean period of 38 months and demonstrated growth in 2 (20%) patients and an overall one-dimensional growth rate of 0.06 cm/year. Raut et al.[9] found no growing tumors over a mean observation period of 81 months in 18 patients. A meaningful interpretation and comparison of these data with ours are impossible because of the very limited number of patients included in these studies, which, in addition, are subject to patient referral and selection bias. Stangerup et al.[11] did not examine tumor sublocalization or growth within the canal but demonstrated growth to extrameatal extension in 35 of 228 (18%) patients, which is comparable with the present finding of 19%.

The 45% occurrence of growth for the purely intracanalicular VS is fairly on average with respect to data on tumors with an extrameatal component, reported by other centers to be from 18% to 69%,[1–10] in selected patient materials with referral bias. Our data on extrameatal tumors showed a somewhat lower growth occurrence of 34%.[11] It is difficult to assess the comparability of the growth rate because almost all studies reports growth rates based on one- or two-dimensional measurements only.[1,2,4–7,9–11] Two previous studies using three-dimensional measurement for volume determinations have shown a growth occurrence of, respectively, 32% (12 of 38 patients)[8] and 36% (17 of 47 patients).[3] Unfortunately, none of these studies included calculation of the absolute or relative volumetric growth rate.

Growth and Tumor-Bone Interface

Forty-five percent of the purely intracanalicular tumors grow to some extent, whereas only 19% grow into the cerebellopontine angle. In other words, the growth appears to be arrested within the canal in a large proportion of the tumors displaying growth after diagnosis. Most tumors do not involve expansion of the osseous IAC. However, the occurrence of IAC expansion at the diagnostic MRI is significantly higher for tumors displaying subsequent shrinkage. These observations are very interesting when considering factors with potential influence on tumor growth and could indicate that an intimate tumor-bone interface may induce growth arrest or even tumor shrinkage, although this is purely speculative.

Treatment Strategy

Because the present data show that less than a fifth of the purely intracanalicular tumors expand into the cerebellopontine angle, the only reasonable treatment is primary observation by repeated MRI, unless realistic hearing preservation is intended.[15] We propose the following observational strategy for all sporadic, unilateral, and purely intracanalicular VS: yearly MRI for 5 years, followed by MRI every other year for 4 years, followed by MRI every 5 years for 10 years, after which the observation is terminated. A rigid data interpretation indicates no reason to follow patients for more than 5 years because tumor growth only occurred within the first 5 years after diagnosis. We have, however, chosen the cautious treatment policy above because only a limited number of tumors have been followed for more than a decade (Fig. 1). If significant growth occurs into the cerebellopontine angle, active treatment is recommended (surgery or radiotherapy). Naturally, special considerations may indicate a deviation from this management policy (e.g., continued observation of diseased or elderly patients).

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