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Rate of Recurrent Vestibular Schwannoma After Total Removal Via Different Surgical Approaches

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Objectives: The objective of this study was to assess the differences in the recurrence rates of vestibular schwannoma (VS) after total tumor removal through enlarged translabyrinthine (ETL), retrosigmoid (RS), and middle cranial fossa (MCF) approaches. Our results were compared with previously published data, and literature reviews were done to identify the possible causes for the recurrence of VS.

Methods: We performed a retrospective analysis of 2,400 cases of VS that underwent removal at the Gruppo Otologico, Piacenza, Italy, from 1983 until 2010. The minimum postoperative follow-up was 12 months. We also reviewed the previously published data on recurrence rates of VS after ETL, RS, and MCF approaches.

Results: Total tumor removal was achieved in 2,252 cases (93.8%). The recurrence rate was 0.05% for the ETL approach, 0.7% for the RS approach, and 1.8% for the MCF approach. Literature reviews of 3 previously published case series utilizing the translabyrinthine approach showed that none of the primary tumors were less than 2.0 cm in size. Recurrences were seen between 1 and 13 years after the initial surgery.

Conclusions: The rate of VS recurrence after total removal is exceptionally low in experienced hands. Undetected microscopic deposits left on crucial points such as the facial nerve, the preserved cochlea nerve, or the fundus of the internal auditory canal could be possible causes for the recurrence. A definite advantage of an ETL approach is the excellent internal auditory canal exposure, resulting in an extremely low rate of VS recurrence. The patients should be followed up to 15 years with gadolinium-enhanced magnetic resonance imaging (with fat suppression sequence in ETL approach cases). Recurrent VS may exhibit a faster growth rate than primary VS.

Key Words: middle cranial fossa approach, recurrence, retrosigmoid approach, translabyrinthine approach, vestibular schwannoma.

INTRODUCTION

Complete removal is often the main objective in the surgical management of vestibular schwannoma (VS). This is possible with refinements in surgical techniques, microneurosurgical technology, and an experienced surgical team. Excellent surgical outcomes have been reported regarding preservation of the facial nerve.[1-4] The benign, well-encapsulated nature of VS facilitates tumor resection from the surrounding structures. There are 3 principal surgical approaches for VS resection: the enlarged translabyrinthine (ETL) approach, the retrosigmoid (RS) approach, and the middle cranial fossa (MCF) approach. Among these approaches, only the ETL gives adequate access to the whole length of the internal auditory canal (IAC) and the cerebellopontine angle (CPA), allowing a more extensive exposure for total tumor removal. In contrast, the RS approach has limitations in regard to the fundus of the IAC, and the MCF approach has limitations in regard to the CPA. Total tumor removal is achieved when no remnants are visible under the surgical microscope at the end of the surgery. Tumor that regrew after incomplete removal at the initial surgery was not considered in this study as tumor recurrence.

There are a number of published reports concerning the recurrence of VS removed through various approaches.[1,3,5-12] Most of these reports only briefly evaluate residual or recurrent VS. Only a few mention the recurrence rate. In this study, we identified 2,252 cases (93.8%) that underwent total resection, of which 4 cases developed tumor recurrence. By thoroughly analyzing these cases, we attempted to identify the possible causes for the recurrence. The aims of this study were to determine the recurrence rate with each surgical approach, identify the possible causes of recurrence, and recommend a follow-up period.

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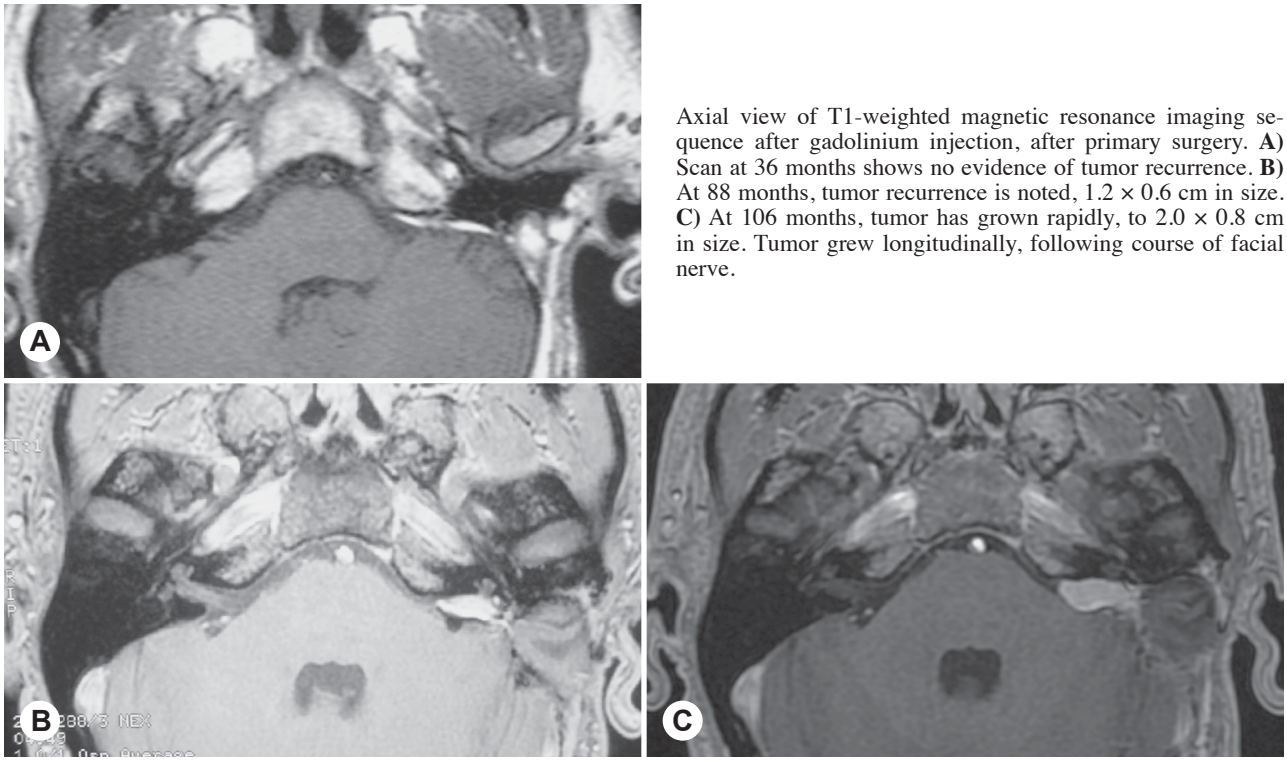

Axial view of T1-weighted magnetic resonance imaging sequence after gadolinium injection, after primary surgery. **A)** Scan at 36 months shows no evidence of tumor recurrence. **B)** At 88 months, tumor recurrence is noted, 1.2 × 0.6 cm in size. **C)** At 106 months, tumor has grown rapidly, to 2.0 × 0.8 cm in size. Tumor grew longitudinally, following course of facial nerve.

MATERIALS AND METHODS

A retrospective case review was performed of the medical records of 2,400 patients with VS who underwent removal at the Gruppo Otologico from 1983 to 2010. The minimum postoperative follow-up was 12 months. Patients with neurofibromatosis type 2 (NF2) and patients who had undergone previous treatment elsewhere for their tumors were excluded. The details of the surgical techniques in the various approaches have been described elsewhere.[13,14] The radiologic and pathological features of the recurrent cases were reviewed to assess the tumor behavior and ascertain any cause for the recurrence. Facial nerve function was reported according to the House-Brackmann (HB) classification.[15] A detailed search of the PubMed database was performed with a review of the English-language literature published by December 2010 by use of the following key words: recurrence in vestibular schwannoma, translabyrinthine approach, retrosigmoid approach, and middle cranial fossa approach.

RESULTS

There were 2,400 cases (47% male and 53% female patients). The patients' ages ranged from 10 to 83 years (mean, 50.4 years). The follow-up periods ranged from 12 to 262 months (mean, 90 months). The tumors ranged from 0 cm (purely intrameatal) to 5.5 cm (mean, 1.8 ± 0.97 cm). The follow-up radiologic evaluations were performed at 2 months and 1, 3, 5, and 7 years. Total tumor removal was achieved in 2,252 cases (93.8%). Of these, 2,011 patients underwent surgery via the ETL approach, 131 via the RS approach, and 110 via the MCF approach. Of the 2,252 cases with total tumor removal, 4 recurrences of VS were subsequently noted. Two of the cases were resected through an MCF approach, and 1 each were resected through the ETL and RS approaches. The facial nerve was preserved anatomically in 2,058 of these 2,252 cases (91.4%). Among these 2,058 cases, 1,255 (61%) retained a postoperative HB grade I or II status, and 601 (29.2%) retained HB grade III status; the remaining 202 cases (9.8%) had HB grade IV-VI status.

Case 1. A 42-year-old man presented with left sensorineural hearing loss in December 2001. Gadolinium-enhanced magnetic resonance imaging (Gd-MRI) showed a 2.0-cm VS in the left CPA region, extending into the IAC. The tumor was completely resected through an ETL approach in January 2002. During the operation, the fundus of the IAC was found to be clear, and a good plane of cleavage was found between the tumor and the facial nerve. Afterward, the patient had HB grade V facial palsy, but recovered to HB grade III 6 months later and remained there until 2009. A 1.2 × 0.6-cm recurrence was noted in April 2009 after a routine Gd-MRI scan. The patient was asymptomatic, apart from left-sided grade III HB facial weakness. A repeat Gd-MRI scan in October 2010 showed an increase in size to 2.0 × 0.8 cm (see Figure). In January 2011,

the patient underwent tumor removal again through an ETL approach. In the CPA region, the facial nerve was found to be lying anterior to the tumor with a good plane of cleavage, and the tumor was totally removed. Afterward, the patient's facial nerve status was still HB grade III, and histopathologic analysis confirmed a benign schwannoma.

Case 2. A 42-year-old woman presented with a 1-year history of sudden progressive right sensorineural hearing loss. A Gd-MRI scan showed a 0.8-cm intrameatal VS located close to the fundus. The tumor was totally removed through the MCF approach in June 2005. A good plane of cleavage was noted between the tumor and the surrounding structures during the operation. The postoperative period was unremarkable, and the patient retained her normal facial function; the histopathologic study confirmed a benign schwannoma. Three years after the primary surgery, a 0.5-cm recurrent tumor located within the right IAC was noted during routine Gd-MRI investigation. The patient is still under follow-up, and the size of the recurrent tumor has remained stable.

Case 3. A 51-year-old woman presented with tinnitus in the right ear. An audiogram showed mild high-frequency sensorineural hearing loss in the right ear. Right intrameatal VS was confirmed later through Gd-MRI. The size of the tumor was about 1.0 × 0.6 cm, and the fundus was free of tumor extension. The tumor was totally removed through an MCF approach in July 1993. The postoperative course was unremarkable. Five years after her initial surgery, the patient developed right-sided facial palsy of HB grade III and reduction of her hearing level. A routine Gd-MRI scan revealed a 0.6-cm recurrent intrameatal tumor. Revision surgical removal was proposed, but the patient declined and opted for a watch-and-rescan policy. The tumor was stable on subsequent routine Gd-MRI investigation.

Case 4. A 48-year-old woman was referred to the Gruppo Otologico in June 2005 with a 2-year history of left tinnitus and poor speech discrimination on the telephone. The audiogram revealed only mild high-frequency sensorineural hearing loss. A Gd-MRI investigation showed VS in the left IAC with minimal extension toward the CPA area. The size of the tumor was 1.0 × 0.8 cm, and the fundus was free of tumor extension. The patient underwent total removal of her tumor through the RS approach in December 2005. Her cochlea and facial nerve were preserved in the surgery. A recurrent tumor was noted during routine Gd-MRI investigation in June 2010. The tumor was 0.5 cm and was located within the left IAC. The recurrent tumor has been managed conservatively with a watch-and-rescan policy.

DISCUSSION

Complete resection of VS requires an experienced surgical team. The recurrence rate is lowest with the ETL approach, followed by the RS and MCF approaches. The recurrence rates after the ETL approach range from 0.05% to 0.99%, according to the published literature[1,5,6] and the present series (see Table[1,3,5-11]). Because of the previous sacrifice of the vestibular and cochlear nerves, often the recurrent tumor is asymptomatic until it reaches a large size. The symptoms include headache, trigeminal nerve disturbances, ataxia, and, in some cases, disturbance of the 9th and 10th cranial nerves.[1,16-19]

Gadolinium-enhanced MRI is the ideal tool for evaluation of postoperative tumor recurrence. With the suppression techniques recommended by Battista et al,[20] is possible to eliminate the signal of the abdominal fat used to obliterate the surgical defect in ETL cases. For RS and MCF approaches, long-term postoperative evaluation by Gd-MRI is necessary to distinguish a suspected recurrent tumor from muscle tissue or fascia used to fill the surgical defect in the IAC. According to Mazzoni et al,[10] the enhancement from the substitute tissue may disappear after 1 year. There may be confusion between dural enhancements and tumor recurrence on MRI immediately after surgery. Elster and DiPersio[21] noted that dural enhancement could be present on MRI within the first 2 years after surgery. There are certain criteria on MRI to suggest tumor recurrence. An enhancing image with a rounded profile and a nodular mass located at the fundus or filling the meatotomy cavity at the IAC is diagnostic for tumor.[10] It has been our policy to follow up VS cases up to 10 years after the primary surgery. According to our data and previously published articles (see Table), recurrence may occur up to 13 years after operation. Therefore, it is reasonable that postoperative follow-up should be lengthened to at least 15 years, with Gd-MRI performed during follow-up at 2 months and at 1, 3, 5, 7, 10, and 15 years after surgery. Documented growing recurrent tumors should be surgically resected, depending on the patient's general condition and life expectancy. In recurrent cases, we believe that the ETL approach is preferable.[12] Radiotherapy may be indicated for patients in poor general condition, for elderly patients, or in the case of refusal of surgical intervention and small lesions.

Postulations for the recurrence involve the anatomic aspect of the fundus of the IAC, the surgical approach, and tumor biological aspects. The vestibular and cochlear components of the eighth cranial

PUBLISHED CASE SERIES WITH RECURRENCE RATES REPORTED ACCORDING TO TRANSLABYRINTHINE, RETROSIGMOID, OR MIDDLE CRANIAL FOSSA APPROACH

Authors	*No. of Cases*	*Recurrence Rate*	*Time to Recurrence After Surgery*	*Size of Primary Tumor*	*Size of Recurrent Tumor*	*Treatment for Recurrence*
Translabyrinthine approach						
Shelton[1]	1,668	0.29% (5 cases)	9 y	2.0 cm	3.2 cm	NS
			10 y	3.5 cm	5.0 cm	TL
			11 y	4.5 cm	3.0 cm	TL + TC + TS
			11 y	4.0 cm	2.5 cm	TL
			13 y	4.5 cm	1.0 cm	TL*
			(10.8-y median follow-up)			
Glasscock et al[5]	568	0.17% (1 case)	5 y	5.0 cm	2.0 cm	NS
Freeman et al[6]	707	0.99% (7 cases)	NS; 9-y median follow-up	NS	NS	NS
Present series	2,011	0.05% (1 case)	7 y	2.0 cm	2.0 cm	ETL
Retrosigmoid approach						
Ebersold et al[3]	255	1.2% (3 cases)	NS	NS	NS	NS
Freeman et al[6]	122	0.8% (1 case)	NS; 9-y median follow-up	NS	NS	NS
Mazzoni et al[10]	104	7.7% (8 cases)	7 y	NS	0.3 cm	ETL
			7 y		1.5 cm	ETL
			7 y		1.5 cm	None
			6 y		0.3 cm	None
			5 y		0.9 cm	ETL
			3 y		0.5 cm	ETL
			3 y		0.1 cm	None
			3 y		0.1 cm	None
Samii and Matthies[11]	880	0.7% (6 cases)	1 y	NS	NS	See below†
			4 y		2.5 cm	
			Others NS		Others NS	
Present series	131	0.7% (1 case)	5 y	1.0 cm	0.5 cm	Watch and rescan
Middle cranial fossa approach						
Gjurić et al[7]	735	0.3% (2 cases)	NS	NS	NS	Both had revision surgery, but approach NS
Meyer et al[8]	162	0.6% (1 case)	NS	NS	NS	NS
Kanzaki et al[9]	160	3.7% (6 cases)	3.5-y median follow-up	NS	NS	3 cases had revision surgery, but approach NS
Present series	110	1.8% (2 cases)	3 y	Intrameatal (0.8 cm)	Intrameatal (0.5 cm)	Watch and rescan
			5 y	Intrameatal (1.0 cm)	Intrameatal (0.6 cm)	Watch and rescan

NS — not specified; TL — translabyrinthine approach; TC — transcochlear approach; TS — transsigmoid approach; ETL — enlarged translabyrinthine approach.

*In this case, patient chose removal of recurrent tumor.

†Three cases had RS approach; other 3 cases NS.

nerve usually enter the IAC as a single unit. They divide into separate nerves within the IAC. The point of separation may vary among individuals. Only in the lateral-most 3 to 4 mm of the IAC do the vestibular and cochlear nerves usually divide into distinct, identifiable structures. The inferior vestibular nerve further branches into the singular nerve. The latter has its own canal posteroinferior to the inferior vestibular nerve. Thus, these nerves are delineated individually in the lateralmost aspect of the IAC. At the fundus of the IAC, the vestibular nerve is separated into superior and inferior divisions by the crista falciformis. Therefore, the tumor may microscopically follow these nerves into their own canal in the fundus of the IAC. Visualization of this area requires removal of part of the bony partition between the vestibule and the lateral IAC.[1] The exposure of the fundus of the IAC will be insufficient unless the

bony labyrinth is violated through an RS or MCF approach.[22] Therefore, it is very critical to address this area during the surgery in order to achieve complete tumor resection. Mazzoni et al[23] have described a modified RS approach in which the perimeatal bone is removed up to the blue line of the labyrinth to permit the direct exposure of the fundus.

The main aims in the surgical management of VS are usually complete tumor removal and facial nerve preservation. We believe that regardless of the surgical approach adopted, exposure of the IAC is the key factor in avoiding tumor recurrence. Adequate tumor exposure up into the fundus of the IAC has been always a major concern in determining the surgical approach, and the ETL approach is the one that allows the surgeon to directly visualize the entire tumor, decreasing the likelihood of tumor remnants.[12] For the MCF approach, we agree with Haberkamp et al[22] that there will be some limitation by the falciform crest blocking the visualization of the inferior half of the fundus. With the posterior fossa approach, the exposure of the fundus of the IAC will be inadequate without violating the bony labyrinth.[22] The rates of tumor recurrence after translabyrinthine procedures have ranged from 0.17% to 0.99%.[1,5,6] In the present series, the incidence of recurrence was 0.05%. To the best of our knowledge, this is the lowest reported recurrence rate in the literature. Including the present series, the recurrence rates after total removal of VS have ranged from 0.3% to 3.7% for the MCF approach[7-9] and from 0.7% to 7.7% for the RS approach.[3,6,10,11]

According to Gamache and Patterson,[24] recurrent VS tumors behave similarly to the primary tumors, with an average annual growth rate of 0.2 cm. In 1 of the cases in our present series, the recurrent tumor grew at more than twice the average rate (see Figure). Vascularity is an important factor that could promote the growth of recurrent tumors. Cayé-Thomasen et al[25] determined that vascular endothelial growth factor is a potent mediator for angiogenesis and has been shown to correlate with the tumor growth in VS.

Luetje et al[26] and Neely[27] postulated that the eighth nerve stump may be a likely source for recurrent tumors, in view of possible multicentric tumors and microscopic intraneural tumor invasion. Neely[27] found that in some cases of solitary VS, the vestibulocochlear nerve is involved beyond the gross tumor with intraneural invasive skip lesions. Neurofibromatosis type 2 should be taken into account in VS recurrence, since microinvasion is more common in NF2 tumors.[19,27,28] Eckermeier et al[29] studied 30 temporal bones containing unoperated acoustic neuroma and found that both the cochlear and vestibular fibers were involved in 28 of the 30 cases.

According to the data from reported series on the translabyrinthine approach (see Table), the sizes of the primary tumors were 2 cm or more. The size of the tumor and prolonged physical compression on the facial nerve may cause attenuation of the nerve function. We believe that there could be microscopic deposits left on the facial nerve. It is still unclear how often microscopic deposits are left on the facial nerve, because this has not been reported in the literature. The ETL approach has been shown to be advantageous for identification of the facial nerve via a consistent anatomic landmark[13] and yields an increased likelihood of preservation of the nerve, but microscopic deposits may not be noticed with the surgical microscope.

CONCLUSIONS

Vestibular schwannoma can be completely resected, with an exceptionally low recurrence rate, by an experienced surgical team. A definite advantage of the ETL approach is the excellent IAC exposure, which results in an extremely low VS recurrence rate. Past recurrences after use of the ETL approach were possibly unrelated to the approach. Rather, we believe they occurred because VS can be invasive and microscopic deposits on the preserved facial or cochlear nerve may not be identified with the surgical microscope. Recurrence after the RS and MCF approaches may be due to inadequate access along the whole length of the IAC. A large size of the primary tumor may also play a role in recurrence. Further study and more clinical data are needed to evaluate statistically the relationship between tumor size and recurrence of VS after surgery. Recurrent VS may grow at a faster rate than primary tumors and can be asymptomatic. Therefore, Gd-MRI scans at 2 months and at 1, 3, 5, 7, 10, and 15 years after surgery are essential.

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