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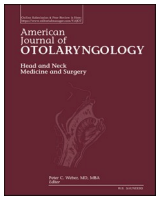

Unique characteristics of pediatric sporadic vestibular schwannoma

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ABSTRACT

Objective: To investigate characteristics of pediatric sporadic (non-NF2) vestibular schwannoma (VS), we compare outcomes of sporadic adult-type vestibular schwannoma which were size- and location-matched cohort.
Study design: Single-institution retrospective matched case–control study.
Methods: Pediatric patients (≤21 years) with sporadic unilateral VS and ≥ 15 months of follow-up were identified and each matched 1:2 to adult patients (>21 years) by tumor size and location. The primary outcome was tumor control (stable vs recurrence/regrowth requiring treatment). Outcomes within the pediatric cohort were further analyzed for age-related differences.
Results: Nine pediatric patients were matched to 18 adult controls. Baseline characteristics and presenting symptoms were similar. Gross total resection was achieved in 44% of children and 22% of adults ($P = 0.071$). Tumor control differed significantly: recurrence/regrowth occurred in 44% of children versus 6% of adults ($P = 0.005$). Within the pediatric group, patients with recurrence were younger (13.8 ± 2.8 vs 17.6 ± 2.6 years; $P = 0.043$). Using a 14-year cutoff, three of four recurrences (75%) occurred in children <14 years versus none in older children ($P = 0.048$).
Conclusions: Children, particularly those under 14 years, experienced significantly poorer tumor control after surgery for sporadic VS compared with adults, despite similar tumor size and location. The higher recurrence and regrowth rates observed in the pediatric cohort suggest that a more aggressive treatment and surveillance approach may be warranted.

1. Introduction

Vestibular schwannoma (VS), also known as acoustic neuroma, is a benign tumor of the vestibulocochlear nerve that typically arises in the internal auditory canal and extends into the cerebellopontine angle [1]. In adults, most cases are sporadic and unilateral, presenting in midlife with hearing loss or tinnitus, with an incidence of 1–2 per 100,000 and earlier detection in recent years due to widespread MRI use [2,3].

By contrast, sporadic VS in the pediatric population is extremely rare, accounting for only 0.8% of intracranial tumors in children, since most pediatric cases occur in the context of neurofibromatosis type 2 (NF2) [4,5]. Case series confirm that truly sporadic pediatric VS (SPVS) without NF2 features are exceptional [6–9]. Children frequently experience diagnostic delays, often presenting with advanced disease, larger tumors, and symptoms of mass effect such as hydrocephalus and brainstem compression [4,6,8,10]. Some studies suggest that pediatric tumors may also demonstrate more rapid growth compared to adult VS [5,9].

Microsurgical resection is the mainstay of treatment for sporadic VS. Pediatric series show that gross total resection (GTR) provides durable tumor control, whereas subtotal resection (STR) almost always leads to regrowth [4,6,7]. Adult studies confirm this pattern, with STR carrying a markedly higher recurrence risk [11]. Current pediatric strategies therefore emphasize maximal safe resection and close MRI follow-up when STR is unavoidable [1,4,5,8,12,13]. Despite this, no study has directly compared pediatric and adult sporadic VS in a matched analysis. The present study addresses this gap by evaluating tumor control and surgical outcomes in pediatric patients and matched adult controls with similar tumor size and location.

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2. Methods

This study was designed as a retrospective cohort analysis performed at Severance Hospital, Seoul, and included patients who underwent surgical resection of vestibular schwannoma (VS) between January 2000 and December 2024. The study protocol was approved by the institutional review board, and the requirement for informed consent was waived due to the retrospective nature of the work (IRB no. 4-2020-1352).

Eligible pediatric patients were defined as those aged 21 years or younger at the time of surgery with sporadic unilateral VS, consistent with prior pediatric VS series [2,4,8,9,13]. To ensure adequate observation, only patients with a minimum of 15 months of clinical and radiological follow-up were included. Patients with NF2, those with a history of prior surgery or radiosurgery at another hospital, and those lost to follow-up or followed for fewer than 15 months were excluded. Nine pediatric patients fulfilled these criteria. For comparison, we identified adult patients (aged over 21 years) with sporadic unilateral VS who underwent surgery during the same period. Each pediatric case was matched with two adult patients based on tumor size and location as determined by preoperative MRI and operative records, yielding a 1:2 matched cohort design.

Clinical information was obtained retrospectively from hospital charts, operative notes, and radiological records. Demographic variables included age, sex, and tumor side. Presenting features were extracted, with particular attention to hearing loss, tinnitus, and imbalance or vertigo, as well as the interval from the first radiologic diagnosis to surgery. Tumor characteristics were documented according to preoperative MRI, including the largest tumor diameter and anatomical location, together with the descriptive "type" recorded in the charts (vestibule, facial nerve, or acoustic schwannoma). Operative details included the surgical approach and the extent of resection, classified as gross total or subtotal according to the operative report and/or early postoperative imaging. Follow-up data encompassed both clinical evaluations and serial MRI studies, with the total duration of postoperative surveillance calculated from the date of surgery.

Outcomes of interest were tumor control, defined as radiographic stability without need for further intervention, and tumor progression, defined as radiographic regrowth or and the requirement for additional treatment. Secondary outcomes included preoperative hearing level graded according to the American Academy of Otolaryngology–Head and Neck Surgery (AAO-HNS) classification, facial nerve function assessed by the House–Brackmann (HB) grading system before and after surgery, the need for second treatment (revision surgery or radiotherapy), and any treatment-related complications when recorded [14–16].

All statistical analyses were performed to compare pediatric and adult cohorts. Continuous variables were summarized as means with standard deviations or medians with interquartile ranges, as appropriate, and compared using parametric or nonparametric tests. Categorical variables were expressed as counts and percentages and compared using Fisher's exact or chi-square tests. A two-sided p value <0.05 was considered statistically significant. Analyses were conducted using SPSS 23.

3. Results

3.1. Pediatric vs. adult cohort comparison

Nine pediatric patients were matched with 18 adult controls by tumor size and location. The mean age was 14.8 ± 2.4 years in children and 54.9 ± 13.3 years in adults. Sex distribution (M:F 3:6 vs 6:12) and tumor laterality (R:L 5:4 vs 9:9) were comparable. Presenting symptoms were similar, with hearing loss present in all children and in 17 of 18 adults, tinnitus in all children and 15 adults, and imbalance in 3 children versus 10 adults (all $P > 0.05$). The anatomical nerve of origin did not differ between groups (Table 1).

Extent of resection was gross total in 4 children (44.4%) and 4 adults (22.2%), with the remainder undergoing subtotal resection ($P = 0.071$). Time from first radiologic diagnosis to surgery was shorter in children (9.3 ± 5.7 months) than adults (16.3 ± 4.1 months), though not significant ($P = 0.32$). Mean follow-up was 41.0 ± 12.2 months in children and 54.4 ± 12.9 months in adults ($P = 0.13$) (Table 1).

Preoperative serviceable hearing (AAO-HNS A-B) was present in 2 of 9 children (22.2%) compared to 11 of 18 adults (61.1%), a difference that did not reach statistical significance ($P = 0.084$). Facial nerve function at presentation was good (HB I-II) in 8 children and 15 adults, with one severe palsy in each group ($P = 0.65$). Postoperatively, good facial function (HB I-II) was maintained in 6 children (66.7%) and 11 adults (61.1%), with the remainder demonstrating moderate dysfunction (HB III-IV); no patient in either group had HB V-VI at follow-up ($P = 0.756$) (Table 1).

Tumor control differed significantly between groups. Stable disease was achieved in 5 children (55.6%) compared with 15 adults (94.1%), whereas progression or recurrence occurred in 4 children (44.4%) versus 1 adult (5.9%) ($P = 0.005$). Among children with recurrence, 2 underwent repeat surgery and 2 received salvage radiotherapy. The single adult recurrence was managed with radiation (Table 2, Fig. 1a, b).

3.2. Pediatric subgroup analysis

Within the pediatric cohort, four patients developed tumor recurrence or regrowth and five remained stable during follow-up. Children with recurrence were significantly younger at surgery compared with those with stable disease (13.8 ± 2.8 vs 17.6 ± 2.6 years; $P = 0.043$). To further explore this finding, patients were stratified by age 14 years, a threshold corresponding to the approximate median of the cohort and the transition into mid-adolescence. Three of the four children with recurrence (75%) were younger than 14 years, whereas none of the five children with stable disease fell into this age group ($P = 0.048$). Tumor size at presentation was larger in the recurrence group (23.7 ± 14.4 vs 14.0 ± 10.4 mm), although this difference was not significant ($P = 0.3$). Follow-up duration was also similar between groups (39.2 ± 15.1 vs 42.4 ± 11.0 months; $P = 0.5$) (Table 3).

4. Discussion

VS are exceedingly rare in children, constituting only about 2% of

Table 1
Demography and symptoms, and tumor parameters.

	Children	Adults	
N	9	18	
Age, y (sd)	15.9 (3.2)	54.9 (13.3)	
Gender M:F	3: 6	6:12	
Side R:L	5:4	9:9	
Symptoms			
Hearing loss (%)	9 (100)	17 (87.5)	
Tinnitus (%)	9 (100)	15 (94.4)	
Imbalance (%)	3 (33.3)	10 (55.5)	
Tumor parameters			
Location\type			
Labyrinth	1	2	
FN	1	2	
Vestibular schwannoma	7	14	
Size, mm (SD)	16.67 (13.35)	16.67 (13.35)	
AAO-NHS hearing scale			
Serviceable hearing (A–B) before surgery, (%)	2 (28.5)	11 (61.1)	p = 0.084
HB facial nerve grading			
Before surgery, good-moderate function (I–III), (%)	8 (0.88)	17 (94.4)	P = 0.61
After surgery, good-moderate function (I–III), (%)	7 (0.77)	12 (66.6)	P = 0.55

Table 2
Extent of resection, time to surgery, follow-up duration, and tumor control.

	Children	Adults	P-value
Extent of resection			
Sub-total	5 (55.6%)	14 (77.8%)	
Total	4 (44.4%)	4 (22.2%)	0.071
Time to surgery, months (SD)	9.3 (5.7)	16.3 (4.1)	0.32
Follow-up after, months (SD)	41 (37.2)	54.4 (12.9)	0.13
Tumor control rate - Stable	5 (55.6%)	15 (94.1)	**p = 0.005**

Note: Values in bold indicate statistically significant differences between groups ($p < 0.05$).

posterior fossa tumors but representing the majority (40–86%) of cerebellopontine angle lesions, a distribution pattern comparable to that seen in adults [2,4,5,8,10]. The vast majority of pediatric VS are associated with NF2 [4,5,7,9,11], whereas SPVS account for only a very small minority. Mazzoni et al. reported that SPVS represent approximately 2% of all VS across age groups, underscoring the exceptional rarity of this entity [6].

In this size- and location-matched comparison of SPVS versus adults, the principal finding in the pediatric group is a markedly lower rate of postoperative tumor control in the pediatric cohort. Despite similar baseline characteristics, recurrence or regrowth occurred in 44.4% of children compared to 5.9% of adults ($P = 0.005$), highlighting a clinically meaningful difference that is not explained solely by tumor size or location (Fig. 2).

Within the pediatric group, younger age emerged as an adverse marker. Children with recurrence were significantly younger at surgery, and when stratified by a 14-year cutoff, three of four children with recurrence (75%) were under 14 years, whereas none in the stable group fell below this threshold. Tumor size and follow-up duration did not differ significantly between outcomes, suggesting that age itself may influence tumor behavior. Similar signals have been observed in pediatric case series and meta-analyses, which have noted that younger children may present with larger tumors, faster growth, and more aggressive clinical courses [1,4,5,8,9,14].

Our findings align with prior literature demonstrating that subtotal resection (STR) in pediatric VS is associated with high rates of regrowth. Several series reported regrowth in nearly all children following STR, whereas gross total resection (GTR) provided durable tumor control [1,4,6,7,13]. Malina et al., in their systematic review, emphasized the importance of complete resection whenever feasible, given the vulnerability of pediatric residual tumors [5]. Moreover, pediatric data showing higher recurrence and regrowth rates compared with adults suggest that a more aggressive treatment approach may be warranted in this population [5]. This trend may also have a biological basis beyond surgical factors. Zhang et al. reported that pediatric patients demonstrated significantly higher Ki-67 proliferation indices than adults [17]. Such elevated Ki-67 levels have not typically been observed in adult cohorts [18]. Although molecular analysis was not performed in the present study, these clinical outcomes imply that pediatric VS may harbor distinct biological characteristics. Potential factors could include aggressive genetic variants, undetected genetic mosaicism, or an upregulated expression of growth factors that drive rapid turnover, unlike the indolent course often seen in adults. These findings collectively support the concept that age-related biological factors contribute to the more aggressive course observed in pediatric VS and highlight the need for future molecular studies [18–22].

From a clinical perspective, these findings underscore several points. First, counseling for pediatric families particularly when the patient is younger than 14 years should emphasize the elevated risk of post-surgical progression and the potential need for salvage therapy [16]. Second, maximal safe resection should be the surgical goal in children, as facial nerve outcomes were similar between groups, indicating that aggressive resection need not compromise function when performed in experienced centers [15]. Finally, younger pediatric patients may warrant closer MRI surveillance in the early years after surgery, given their higher recurrence risk.

Despite its strengths, this study also carries some limitations. It is a single-institution, retrospective study, and although SPVS are

Table 3
Subgroup analysis of pediatric patients with sporadic vestibular schwannoma by tumor control outcome.

Children	Rec\growth (n = 4)	Stable (n = 5)	P-value
Age, y (SD)	13.8 (2.8)	17.6 (2.6)	**0.043**
Age < 14y	3 (75%)	1 (20%)	**0.048**
Tumor size	19.2 (18.3)	14 (10.4)	0.5
Follow-up duration	39.2 (15.1)	42.4 (11.0)	0.5

Note: Values in bold indicate statistically significant differences between groups ($p < 0.05$).

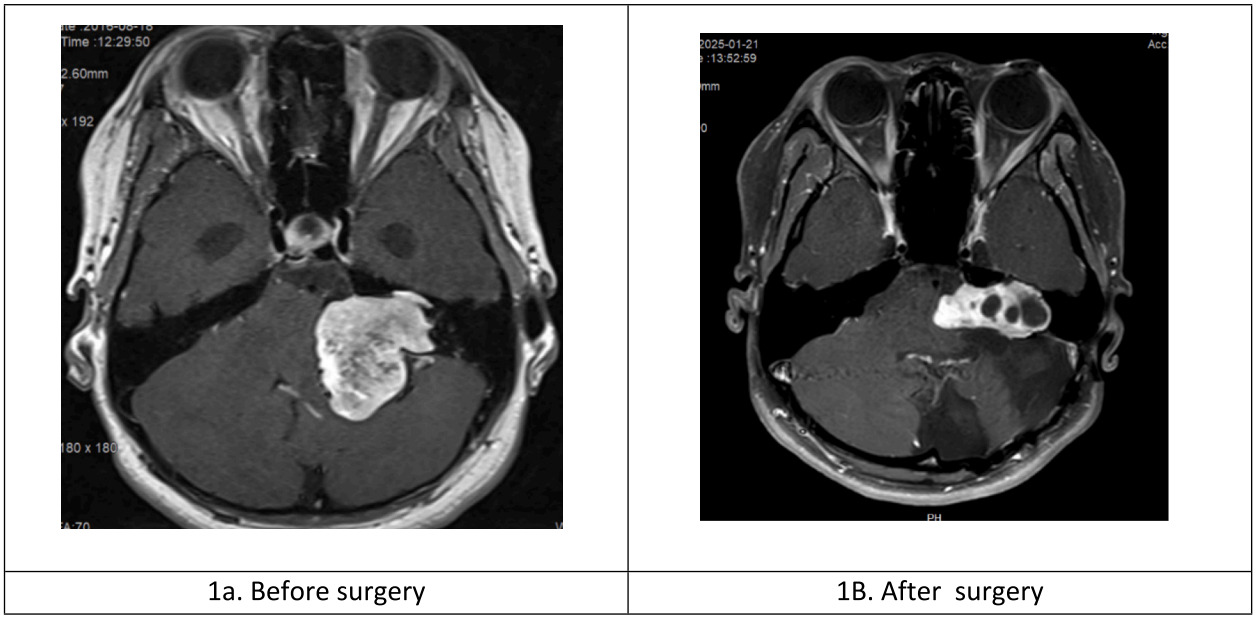

Fig. 1. Pre- and postoperative MRI of an 18-year-old female with a 45-mm vestibular schwannoma, demonstrating tumor regrowth.

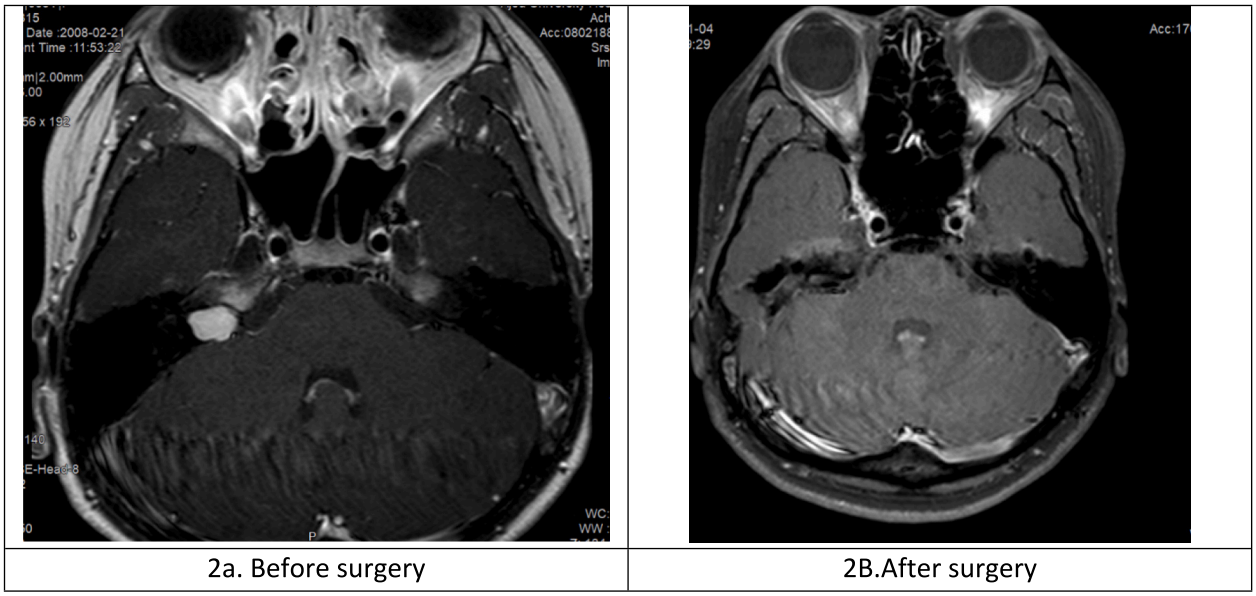

Fig. 2. Pre- and postoperative MRI of an 11-year-old female with a 17-mm vestibular schwannoma, demonstrating complete resection without recurrence.

exceedingly rare and ours is among the largest reported series of sporadic cases [1,4–7] the absolute numbers remain relatively low, which limits statistical power. Furthermore, while all pediatric patients had at least 15 months of follow-up, the adult cohort had longer surveillance on average, which could bias toward detecting more recurrences in adults rather than fewer. Larger multi-institutional cohorts will be necessary to validate the age effect and refine treatment strategies.

5. Conclusion

This matched analysis—the first to directly compare pediatric and adult sporadic vestibular schwannoma of similar size and location—demonstrates that children, particularly those under 14 years, experience significantly poorer tumor control after surgery compared with adults. Taken together with prior pediatric series [1,4–7], our data support maximal safe resection and vigilant postoperative surveillance, especially in younger patients.

CRediT authorship contribution statement

Udi Shapira: Writing – review & editing, Writing – original draft, Visualization, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Riana Kipiani Abdul Halim:** Writing – review & editing, Investigation, Conceptualization. **Jeong Geum Lee:** Writing – review & editing, Methodology, Formal analysis, Data curation. **In Seok Moon:** Writing – review & editing, Writing – original draft, Visualization, Supervision, Investigation, Conceptualization.

Authorship, originality and prior publication

This manuscript is original, has not been published previously in whole or in part, and is not under consideration for publication elsewhere. All authors meet the criteria for authorship as defined by the International Committee of Medical Journal Editors (ICMJE), have approved the final version of the manuscript, and agree to be accountable for all aspects of the work.

Originality and prior publication

This manuscript is original, has not been published previously in whole or in part, and is not under consideration for publication elsewhere.

Institutional review board statement

This study was approved by the Insti-tutional Review Board (IRB) of Yonsei University Health System (IRB no. 4-2020-1352).

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Declaration of competing interest

The authors declare no conflict of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amjoto.2026.104788.

References

[1] Zipfel J, Gorbachuk M, Gugel I, Tatagiba M, Schuhmann MU. Management of sporadic vestibular schwannomas in children—volumetric analysis and clinical outcome assessment. Children (Basel) 2022;9(4):490.
[2] Walcott BP, Sivarajan G, Bashinskaya B, et al. Sporadic unilateral vestibular schwannoma in the pediatric population. J Neurosurg Pediatr 2009;4(2):125–9.
[3] Fernández-Méndez R, Wan Y, Axon P, Joannides A. Incidence and presentation of vestibular schwannoma: a 3-year cohort registry study. Acta Neurochir 2023;165(10):2903–11.
[4] Başar İ, Hanalioğlu Ş, Narin F, Bilginer B. Pediatric vestibular schwannomas: evaluation of clinical features, treatment strategies and long-term results of 10 cases. Turk Neurosurg 2018;28(3):116–25.
[5] Malina GEK, Heiferman DM, Riedy LN, et al. Pediatric vestibular schwannomas: case series and a systematic review with meta-analysis. J Neurosurg Pediatr 2020;26(3):302–10.
[6] Mazzoni A, Dubey SP, Poletti AM, Colombo G. Sporadic acoustic neuroma in pediatric patients. Int J Pediatr Otorhinolaryngol 2007;71(10):1569–72.
[7] Zipfel GJ, et al. Unilateral vestibular schwannomas in childhood without evidence of neurofibromatosis: experience of 10 patients at a single institute. Neurosurgery 2000;46(2):328–36.

[8] Janz TA, et al. Characteristics and outcomes of pediatric vestibular schwannomas. Otol Neurotol 2021;42(4):557–65.
[9] Garg K, et al. Pediatric vestibular schwannoma with or without neurofibromatosis: characteristic features and operative nuances. Childs Nerv Syst 2007;23(4):399–406.
[10] Holman MA, Proops DW, Grant DB. Pediatric cerebellopontine angle and internal auditory canal tumors. Childs Nerv Syst 1999;15(2–3):121–7.
[11] Choi JW, Park HJ, Kim CH, Kim DG, Chung YS, Park SH. Clinical course of vestibular schwannoma in pediatric neurofibromatosis type 2. J Neurosurg Pediatr 2014;13(6):650–7.
[12] Nakatomi H, Jacob JT, Carlson ML, et al. Long-term risk of recurrence and regrowth after gross-total and subtotal resection of sporadic vestibular schwannoma. J Neurosurg 2017;133(4):1052–8.
[13] MacNally SP, Rutherford SA, Ramsden RT, Evans DG, King AT. Outcome from surgery for vestibular schwannomas in children. Br J Neurosurg 2009;23(3):226–31.
[14] Han MS, Kim SY, Kim HJ, et al. Comparison of clinical characteristics and surgical outcomes of cystic and solid vestibular schwannomas. Otol Neurotol 2018;39(7):e381–6.
[15] Kim J, Moon IS, Jeong JH, Lee HR, Lee WS. What really decides the facial function of vestibular schwannoma surgery? Clin Exp Otorhinolaryngol 2011;4(4):168–73.
[16] Lee HJ, Kim MJ, Koh SH, Chang WS, Moon IS. Comparing outcomes following salvage microsurgery in vestibular schwannoma patients failing gamma-knife radiosurgery or microsurgery. Otol Neurotol 2017;38(9):1339–44.
[17] Zhang Z, Zheng X, Zhang D, et al. Vestibular schwannomas in young patients: a 12-year experience in a single center. World Neurosurg 2022;158:e166–78. https://doi.org/10.1016/j.wneu.2021.10.145.
[18] Iannella G, de Vincentiis M, Di Gioia C, Pasquariello B, Greco A, Ciofalo A, et al. Subtotal resection of vestibular schwannoma: evaluation with Ki-67 measurement, magnetic resonance imaging, and long-term observation. J Int Med Res 2017;45(3):1061–73. https://doi.org/10.1177/0300060516686873.
[19] O'Reilly BF, Kishore A, Crowther JA, et al. Correlation of growth factor receptor expression with clinical growth in vestibular schwannomas. Otol Neurotol 2004;25:791–6.
[20] Charabi S, Engel P, Charabi B, et al. Growth of vestibular schwannomas: in situ model employing the monoclonal antibody Ki-67 and DNA flow cytometry. Am J Otolaryngol 1996;17:301–6.
[21] Charabi S, Engel P, Jacobsen GK, et al. Growth rate of acoustic neuroma expressed by Ki-67 nuclear antigen versus symptom duration. Ann Otol Rhinol Laryngol 1993;102:805–9.
[22] Yokoyama M, Matsuda M, Nakasu S, et al. Clinical significance of Ki-67 staining index in acoustic neurinoma. Neurol Med Chir (Tokyo) 1996;36:698–702.