 doi: 10.2176/jns-nmc.2024-0292

NMC Case Report Journal 12, 197-201, 2025

Hydrocephalus after Gamma Knife Surgery for Vestibular Schwannoma Resolved by Tumor Removal without Cerebrospinal Fluid Diversion: Report of Two Cases

Miyuki OGANE,[1] Iwao YAMAKAMI,[2] Shunsuke KUBOTA,[3] Toru SERIZAWA,[4] and Yoshinori HIGUCHI[1]

[1]*Department of Neurosurgery, Chiba University Hospital, Chiba, Chiba, Japan*
[2]*Department of Neurosurgery, Sosen Hospital, Chiba, Chiba, Japan*
[3]*Department of Neurosurgery, Chiba Medical Center, Chiba, Chiba, Japan*
[4]*Tokyo Gamma Unit Center, Tsukiji Neurological Clinic, Tokyo, Japan*

Abstract

Hydrocephalus following Gamma Knife surgery for vestibular schwannoma is typically treated with cerebrospinal fluid diversion. However, additional cerebrospinal fluid diversion (shunt placement) causes a lifelong risk of shunt malfunction and infection. We report two cases of vestibular schwannoma in which the hydrocephalus with progressive tumor growth after Gamma Knife surgery was treated by an initial tumor removal, resolving hydrocephalus without cerebrospinal fluid diversion and causing long-term tumor control.

Patient 1 underwent Gamma Knife surgery for a 22-mm tumor vestibular schwannoma of Koos grade III and developed symptomatic hydrocephalus with progressive tumor growth. Tumor removal at 17 months after Gamma Knife surgery resolved the hydrocephalus without tumor recurrence 8 years after surgery. Patient 2 underwent Gamma Knife surgery for an 18-mm tumor vestibular schwannoma of Koos grade IV and developed rapid tumor growth and symptomatic hydrocephalus 2 years after Gamma Knife surgery. Patient 2 underwent subtotal tumor removal at 40 months after Gamma Knife surgery resolving hydrocephalus without residual tumor progression at 14 years after Gamma Knife surgery.

Subtotal tumor removal may be a primary treatment option in patients with vestibular schwannoma treated with Gamma Knife surgery and developing hydrocephalus with tumor progression. This might help avoid cerebrospinal fluid diversion with long-term tumor control.

Keywords: hydrocephalus, gamma knife surgery, vestibular schwannoma

Introduction

Gamma Knife surgery (GKS) is widely used as a minimally invasive treatment for vestibular schwannomas (VS), particularly for small to medium-sized tumors.[1)] Hydrocephalus occurs in the incidence of 4%-6% after GKS for VS, and hydrocephalus following GKS is managed by cerebrospinal fluid (CSF) diversion.[2,3)] Hydrocephalus occurs without GKS in 3.7%-42% of VS patients.[4)] Since 87% of hydrocephalus coexisting with treatment-naive VS was resolved after tumor removal (TR) alone,[4)] TR was the initial treatment for VS with hydrocephalus. We report two VS patients who developed hydrocephalus along with tumor growth after GKS and who were treated only by TR, resulting in hydrocephalus resolution without CSF diversion and long-term tumor control.

Received November 14, 2024; Accepted February 27, 2025

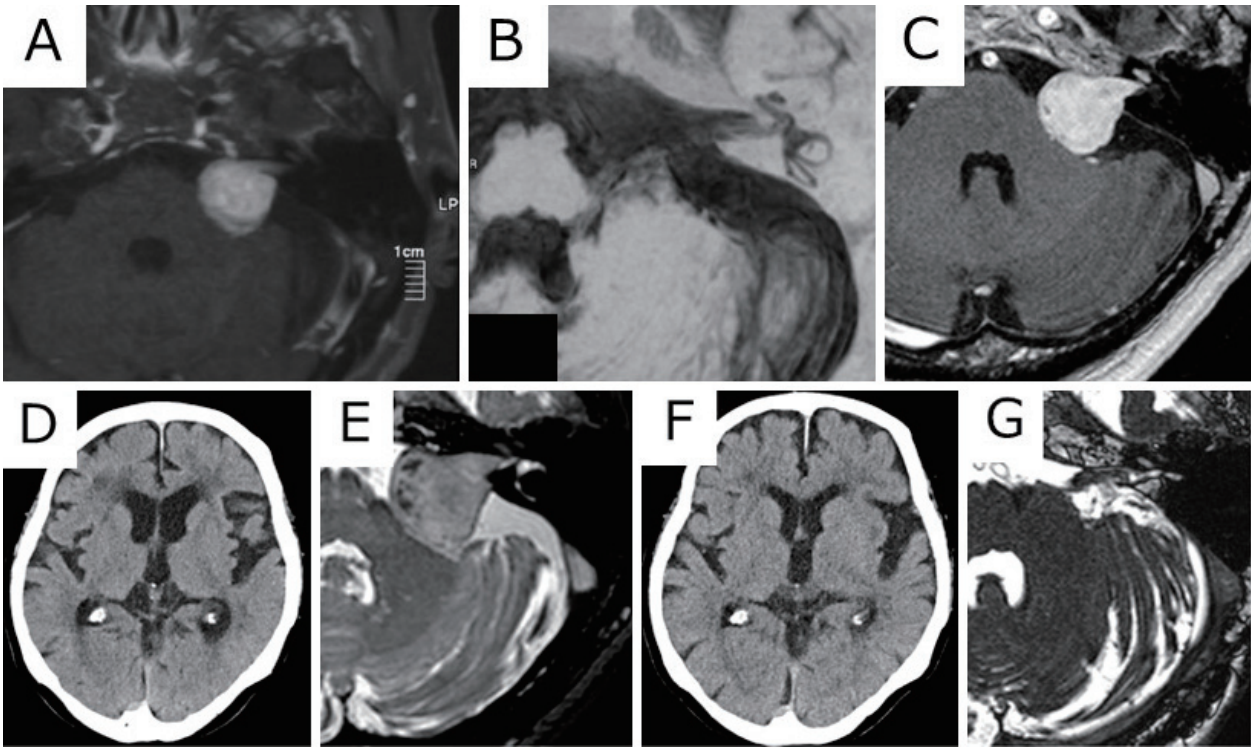

Fig. 1 Case 1.
MRI at 9 years before diagnosis of vestibular schwannoma (B) showed no tumor in the left cerebellopontine angle (CPA). At the time of diagnosis, MRI (A) revealed a 21-mm solid mass in the left CPA. The tumor measured 22 mm on MRI at the time of GKS (C). A CT scan 1 year after GKS (D) showed ventricular enlargement. MRI at the time of surgery (E) showed that the tumor grew to 26 mm. Eight years after diagnosis, there was no tumor growth nor the recurrence of hydrocephalus (F: CT, G: MRI).
CT: computed tomography; GKS: Gamma Knife surgery; MRI: magnetic resonance imaging

Case Report

Case 1: A 74-year-old woman at diagnosis

The patient experienced sudden sensorineural hearing loss 23 years before and noticed progressive hearing decline thereafter. She consulted a local otologist; however, imaging studies were not performed. The hearing loss persisted and gradually progressed to deafness. Magnetic resonance imaging (MRI) after a minor head injury demonstrated the left 21-mm solid VS of Koos grade III (Fig. 1A). MRI 9 years before the diagnosis showed no tumor in the left cerebellopontine angle (Fig. 1B), suggesting a relatively rapid tumor growth rate and no association between the VS and the sudden sensorineural hearing loss 23 years earlier. Although TR was proposed as a primary treatment, GKS (12 Gy at periphery) was performed two months after the diagnosis under the patient's preference, when the tumor diameter was 22 mm (Fig. 1C). The tumor enlarged progressively after GKS, increasing to 23 mm and 24 mm at 3 and 6 months after GKS, respectively. The patient developed gait disturbance and had difficulty in daily activity 12 months after GKS, and progressive ventricular enlargement led to a diagnosis of hydrocephalus following GKS (Fig. 1D). Repeated CSF tap tests resulted in temporary improvement of hydrocephalus symptoms and revealed an elevated CSF protein level of 150 mg/dL. MRI showed progressive brainstem compression by the tumor with 26 mm in diameter, 16 months after GKS (Fig. 1E). The patient underwent subtotal TR (90% in resection rate) preserving the facial nerve function 17 months after GKS. The histopathological examination of the resected tumor revealed a schwannoma with a Ki-67 index of less than 5% in the highest area and fibrosis probably due to the effects of GKS. No mitotic and malignant figures led to a diagnosis of benign schwannoma. The symptoms were resolved after surgery. MRI showed no residual tumor progression or hydrocephalus (Fig. 1F and G) 8 years after surgery.

Case 2: A 56-year-old woman at diagnosis

The patient presented with left facial sensory disturbance, leading to a diagnosis of left 28-mm solid VS of Koos grade IV (Fig. 2A). She had lost her left hearing. She underwent an intentional subtotal TR to preserve the facial nerve function. Because of the rapid residual tumor growth (Fig. 2B), a second TR was performed 15 months after surgery. The histopathological diagnosis of the second surgery was a schwannoma with a Ki-67 index of approximately 2%-3% in the highest area and partial hemorrhage.

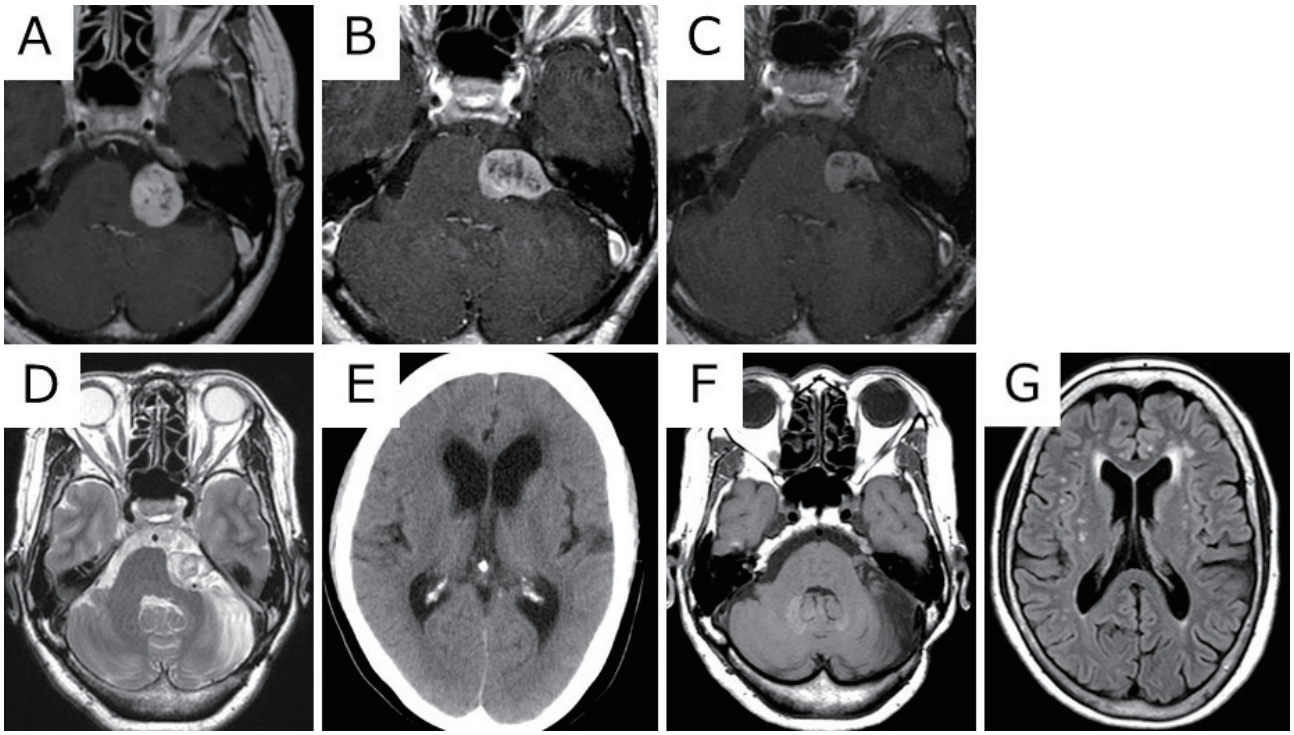

Fig. 2 Case 2.
MRI at diagnosis of vestibular schwannoma (A) showed a tumor measuring 28 mm. At the time of the second surgery (B), the tumor size was 28 mm. By the time of GKS (C), the tumor size was 18 mm. MRI at 5 years after the diagnosis (D) showed the tumor regrew to 27 mm, and CT scan (E) revealed the ventricular enlargement, leading to the final tumor removal. CT scan (F) and MRI (G) confirmed no recurrence of the residual tumor nor hydrocephalus 12 years after the third surgery.
CT: computed tomography; GKS: Gamma Knife surgery; MRI: magnetic resonance imaging

No karyorrhexis or malignant findings were present. Planed GKS (12 Gy in the periphery) was applied 6 months after the second surgery when the tumor diameter was 18 mm (Fig. 2C). The residual tumor showed rapid growth, and the patient developed gait disturbance 2 years after GKS. Computed tomography scans demonstrated progressive ventricular enlargement with progressive tumor growth 3 years after GKS (27-mm tumor, Fig. 2D and E). The patient underwent subtotal TR (resection rate 80%) preserving facial nerve function. The histopathology of the third surgery revealed a benign schwannoma with a Ki-67 index of less than 2% in the highest area. The proliferation of collagen fibers and degenerated hyaline fibers, presumed to be tissue reactions following GKS, as well as neutrophil infiltration around blood vessels, were observed. No karyorrhexis or malignant findings were present. Both hydrocephalus and the symptoms resolved after the third surgery. MRI showed no tumor recurrence or hydrocephalus (Fig. 2F and G) 12 years after the third surgery.

Discussion

Hydrocephalus after GKS for VS occurs because of the elevated CSF protein due to tumor necrosis and impaired CSF absorption.[5] Additionally, in cases of large tumors, compression of the fourth ventricle may lead to impaired CSF circulation, contributing to hydrocephalus.[6] CSF diversion, i.e. shunt placement, is a standard treatment. However, unnecessary shunt placement should be avoided because the shunt placement causes a lifelong risk of shunt malfunction and infection. In some cases, hydrocephalus can be resolved after TR alone, making shunt surgery unnecessary. We presented two VS cases with hydrocephalus after GKS and TR alone resolved hydrocephalus with long-term tumor control.

VS treated with GKS involves transient tumor progression; tumor volume increases by an average of 27% at 6 months and typically shrinks to its original size at 12 months after GKS.[7] Some of the patients had persistent tumor growth more than 1 year after GKS and required TR in approximately 2%-3% of the patients.[8,9] In the present two cases with tumors growing for over a year after GKS and developing the symptomatic hydrocephalus, we performed TR to resolve the hydrocephalus and to expect long-term tumor control.

The hydrocephalus in treatment-naive VS patients is mostly resolved by TR without CSF diversion.[10] Both intracranial pressure and CSF protein reduce after TR.[11] In the present cases, TR might be associated with decreased CSF protein, improved CSF absorption, and resolving hy-

drocephalus.

The larger the VS at the time of GKS, the higher the incidence of hydrocephalus after GKS.[12] The larger the tumor at the time of hydrocephalus onset, the greater the effect of TR on reducing CSF protein and improving CSF absorption; it may increase the hydrocephalus resolution rate. A higher TR rate correlates with a higher hydrocephalus cure rate.[11]

In a report on cases of VS larger than 25 mm treated with GKS, 20.4% (11 out of 54 cases) developed hydrocephalus; however, none of these cases exhibited tumor enlargement.[8] Similarly, in other studies, approximately 5.0% (11 out of 211 cases) developed hydrocephalus, but the tumor control rate was 100%.[13] These two reports suggest that hydrocephalus following GKS is not necessarily associated with tumor growth after GKS. However, we believe that not only the presence or absence of final tumor growth but also transient expansion after GKS could influence the development of hydrocephalus. In cases of transient tumor expansion, elevated CSF protein levels during the expansion phase may persist and potentially contribute to hydrocephalus.

In cases where hydrocephalus develops after GKS for VS, previous reports and the present cases suggest that such tumors are often relatively large at the time of GKS (approximately 20-25 mm or larger). Even in the absence of tumor growth, TR may improve CSF protein levels, offering potential benefits for hydrocephalus management. This effect is expected to be particularly pronounced in cases with tumor growth. However, physicians performing GKS tend to avoid invasive treatments, which may lead to hesitation in proceeding with TR for hydrocephalus in the absence of tumor growth. Further investigation is necessary to address this issue and develop appropriate solutions.

Shunt placement is the safe and effective treatment of idiopathic normal pressure hydrocephalus with a success rate of 82%.[14] However, complications such as mortality (0.2%), and morbidity (8.2%), including subdural hematoma (4.5%), intracerebral hemorrhage (0.2%), and infection (3.5%), are reported, with a shunt revision rate of 13%.[14] In a study comparing ventriculoperitoneal (VP) shunts and lumboperitoneal (LP) shunts, there was no difference in effectiveness or severe adverse events between the two groups; however, shunt revisions were more frequent in the LP shunt group (7%) compared to the VP shunt group (1%).[15] Shunt malfunction and infection pose lifelong risks. In the present two cases, TR resolved the hydrocephalus after GKS without CSF diversion and thereby eliminated the lifelong risks indigenous to shunt placement.

GKS increases the risk of postoperative facial palsy and hearing impairment in the TR because GKS causes fibrosis and adhesion between the tumor and surrounding structures.[16] The residual tumors after salvage surgery for GKS-treated VS commonly demonstrated tumor shrinkage, and subtotal TR should be considered to avoid the additional deficit. In the present two cases, TR preserved facial nerve function without any new neurological deficit. Although TR was subtotal (achieving 80-90% TR), hydrocephalus was successfully resolved, and the residual tumors did not show regrowth during the 8 to 12-year follow-up period. These findings suggest that achieving a substantial extent of TR, such as 80-90%, might serve as a critical guideline for treatment, even without pursuing total resection.

Conclusion

Considering the chance of resolving the hydrocephalus without CSF diversion, TR may offer an effective initial treatment for VS patients with hydrocephalus and progressive tumor progression after GKS.

Informed Consent

Informed consent was obtained from all patients included in this case report.

Conflicts of Interest Disclosure

The authors declare no conflicts of interest.

References

1) Tatagiba M, Wang SS, Rizk A, et al. A comparative study of microsurgery and gamma knife radiosurgery in vestibular schwannoma evaluating tumor control and functional outcome. Neurooncol Adv. 2023;5(1):vdad146. doi: 10.1093/noajnl/vdad146
2) Lee S, Seo SW, Hwang J, et al. Analysis of risk factors to predict communicating hydrocephalus following gamma knife radiosurgery for intracranial schwannoma. Cancer Med. 2016;5(12):3615-21. doi: 10.1002/cam4.955
3) Pirouzmand F, Tator CH, Rutka J. Management of hydrocephalus associated with vestibular schwannoma and other cerebellopontine angle tumors. Neurosurgery. 2001;48(6):1246-53; discussion 1253. doi: 10.1097/00006123-200106000-00010
4) Gerganov VM, Pirayesh A, Nouri M, et al. Hydrocephalus associated with vestibular schwannomas: management options and factors predicting the outcome. J Neurosurg. 2011;114(5):1209-15. doi: 10.3171/2010.10.JNS1029
5) Cauley KA, Ratkovits B, Braff SP, et al. Communicating hydrocephalus after gamma knife radiosurgery for vestibular schwannoma: an MR imaging study. AJNR Am J Neuroradiol. 2009;30(5):992-4. doi: 10.3174/ajnr.A1379
6) Fairhead R, Harris L, Shoakazemi A, et al. Hydrocephalus in patients with vestibular schwannoma. Acta Neurochir. 2023;165(12):4169-74. doi: 10.1007/s00701-023-05866-2
7) Nagano O, Higuchi Y, Serizawa T, et al. Transient expansion of vestibular schwannoma following stereotactic radiosurgery. J Neurosurg. 2008;109(5):811-6. doi: 10.3171/JNS/2008/109/11/0811
8) Bailo M, Boari N, Franzin A, et al. Gamma knife radiosurgery as primary treatment for large vestibular schwannomas: clinical results at long-term follow-up in a series of 59 patients. World Neurosurg. 2016;95:487-501. doi: 10.1016/j.wneu.2016.07.117
9) Pollock BE, Lunsford LD, Kondziolka D, et al. Vestibular schwan-

noma management. Part II. Failed radiosurgery and the role of delayed microsurgery. J Neurosurg. 1998;89(6):949-55. doi: 10.3171/jns.1998.89.6.0949
10) Nakajima M, Yamada S, Miyajima M, et al. Guidelines for management of idiopathic normal pressure hydrocephalus (third edition): endorsed by the Japanese Society of Normal Pressure Hydrocephalus. Neurol Med Chir (Tokyo). 2021;61(2):63-97. doi: 10.2176/nmc.st.2020-0292
11) Miyakoshi A, Kohno M, Nagata O, et al. Hydrocephalus associated with vestibular schwannomas: perioperative changes in cerebrospinal fluid. Acta Neurochir. 2013;155(7):1271-6. doi: 10.1007/s00701-013-1742-9
12) Frischer JM, Gruber E, Schöffmann V, et al. Long-term outcome after gamma Knife radiosurgery for acoustic neuroma of all Koos grades: a single-center study. J Neurosurg. 2019;130(2):388-97. doi: 10.3171/2017.8.JNS171281
13) Lee SH, Seol HJ, Kong DS, et al. Risk factors and tumor response associated with hydrocephalus after gamma knife radiosurgery for vestibular schwannoma. Acta Neurochir. 2012;154(9):1679-84. doi: 10.1007/s00701-012-1350-0
14) Toma AK, Papadopoulos MC, Stapleton S, et al. Systematic review of the outcome of shunt surgery in idiopathic normal-pressure hydrocephalus. Acta Neurochir. 2013;155(10):1977-80. doi: 10.1007/s00701-013-1835-5
15) Miyajima M, Kazui H, Mori E, et al. One-year outcome in patients with idiopathic normal-pressure hydrocephalus: comparison of lumboperitoneal shunt to ventriculoperitoneal shunt. J Neurosurg. 2016;125(6):1483-92. doi: 10.3171/2015.10.JNS151894
16) Iwai Y, Ishibashi K, Nakanishi Y, et al. Functional outcomes of salvage surgery for vestibular schwannomas after failed gamma knife radiosurgery. World Neurosurg. 2016;90:385-90. doi: 10.1016/j.wneu.2016.03.014

Corresponding author: Miyuki Ogane, MD
Department of Neurosurgery, Chiba University Hospital, 1-8-1 Inohana, Chuo-ku, Chiba, Chiba 260-8677, Japan.
e-mail: cffa6405@chiba-u.jp