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Case Report

Refractory communicating hydrocephalus after radiation for small vestibular schwannoma with asymptomatic ventriculomegaly: A case report ☆,☆☆,★,★★

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ABSTRACT

Communicating hydrocephalus is a known tumor-related syndrome associated with vestibular schwannoma, which can occur even in small tumor. Radiation has become a popular primary treatment option for small schwannoma; however, little is known about the efficacy and risk accompanying asymptomatic ventriculomegaly on images. We report a case of a 59-year-old woman who suffer from refractory communicating hydrocephalus after radiation for small vestibular schwannoma with asymptomatic ventriculomegaly. After the surgical removal of the tumor, hydrocephalus was gradually improved due to intermittent lumbar puncture and finally resolved without shunt placement. Surgical removal should be considered as the first option for the treatment, even if the patient is asymptomatic and the images revealed a small vestibular schwannoma with only slight ventricular enlargement.

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Abbreviations list: ADC, apparent diffusion coefficient; CSF, cerebrospinal fluid; GKRS, gamma knife radiosurgery; ICP, intracranial pressure; MR, magnetic resonance.

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Introduction

Hydrocephalus associated with vestibular schwannoma is well recognized condition, which is usually noncommunicating (obstructive) hydrocephalus related to tumor size but occasionally communicating (nonobstructive) hydrocephalus unrelated to tumor size [1]. Communicating hydrocephalus is considered to be a tumor-related syndrome due to elevated cerebrospinal fluid (CSF) protein concentration secreted by the tumor [2–4]. In some cases, it could be found as asymptomatic ventriculomegaly associated with small vestibular schwannoma. For small vestibular schwannoma, radiation has increasingly become a popular primary treatment option; however, little is known about the efficacy and risk of radiation for the small vestibular schwannoma accompanying ventriculomegaly on the images.

Here, we report a case of communicating hydrocephalus exacerbated and become refractory after radiation for vestibular schwannoma with ventriculomegaly on the preoperative images, which was gradually improved and finally resolved on the both clinically and radiographically after surgical removal of the tumor without shunt placement.

Case report

A 59-year-old woman initially presented with decreased hearing on the right side in another hospital. A magnetic resonance (MR) scan revealed a 20-mm tumor at the right cerebellopontine angle accompanying mild ventriculomegaly (Fig. 1A–C). The diagnosis of the tumor was vestibular schwannoma because the MR scan showed the tumor was extended into the internal auditory canal, which was strong homogeneous enhancement and created an acute angle with petrous bone. Given the patient preference for radio surgery, the patient received gamma knife radiosurgery (GKRS; 8-Gy single fraction).

The patient developed headache and gait disturbance 5 months after GKRS. An MR scan demonstrated moderate tumor growth with central necrosis, and with slightly more ventricular enlargement (Fig. 2A–C), while the periventricular hy-

perintensity area on fluid-attenuated inversion recovery MR images was not revealed (Fig. 2D and E). A lumbar puncture showed a high intracranial pressure (ICP) of 25 cm H₂O and an increased CSF protein concentration of 101 mg/dL. A ventriculoperitoneal shunt (CERTAS; Codman, Raynham, MA) was placed to treat the communicating hydrocephalus, and the symptoms improved. However, the symptoms recurred 1 month after the procedure, with lumbar puncture revealing the ICP of 30 cm H₂O and the CSF protein concentration of 168 mg/dL. A shunt revision was performed but also failed to manage the hydrocephalus because of the shunt dysfunction. Then the patient was referred to our hospital for the treatment of the enlarged tumor and concomitant refractory communicating hydrocephalus.

Neurological examination revealed mild consciousness disturbance, Bruns nystagmus, right facial sensory disturbance, right facial palsy (House–Brackmann grade II), right ear deafness, and gait disturbance. An MR scan showed further exacerbated ventriculomegaly and enlarged tumor with the development of necrosis (Fig. 3A–C). The tumor was compressing the brain stem, and the fourth ventricle was shifted but was not fully obstructed (Koos grade IV). To manage the hydrocephalus, we urgently performed external ventricular drainage. The ICP was 25 cm H₂O, and the CSF protein level collected from the drainage tube was 322 mg/dL. (Fig. 4)

Two weeks after, we performed surgical removal of the tumor via retrosigmoid approach. The arachnoid was thickened and partly yellow (Fig. 5A), and the tumor was grayish, soft, and hemorrhagic (Fig. 5B). Subtotal tumor resection was performed except for anchoring of central nerve VII (Fig. 5C), and the external ventricular drainage system was removed. Histopathological analysis revealed a necrotic region, microvascular proliferations, and hemosiderosis in the tumor tissues (Fig. 5D–G).

On day 1 postoperatively, the symptoms of hydrocephalus did not improve immediately and computed tomography showed ventricular size was almost no change (Fig. 6A); the ICP was 45 cm H₂O and the CSF protein level was 155 mg/dL by lumbar puncture (Fig. 4). However, the ICP and CSF protein level gradually decreased by repeated lumbar puncture, and her symptoms gradually improved (Fig. 4). Her consciousness became clear and she walked steadily. She was discharged 24 days after the surgery without a shunt procedure. At the 5-

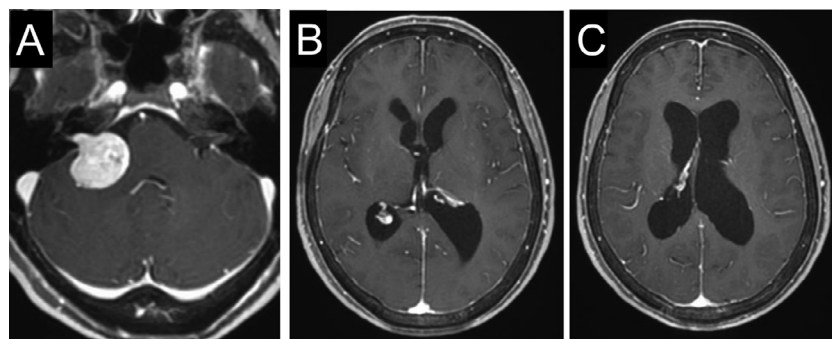


Fig. 1 – (A–C) An axial gadolinium-enhanced T1-weighted magnetic resonance images. At the time of the first admission, it showed a 20-mm (5.5 cm³) enhanced mass extending into the right internal auditory canal accompanying mild ventriculomegaly. The appearance was consisted with a vestibular schwannoma.

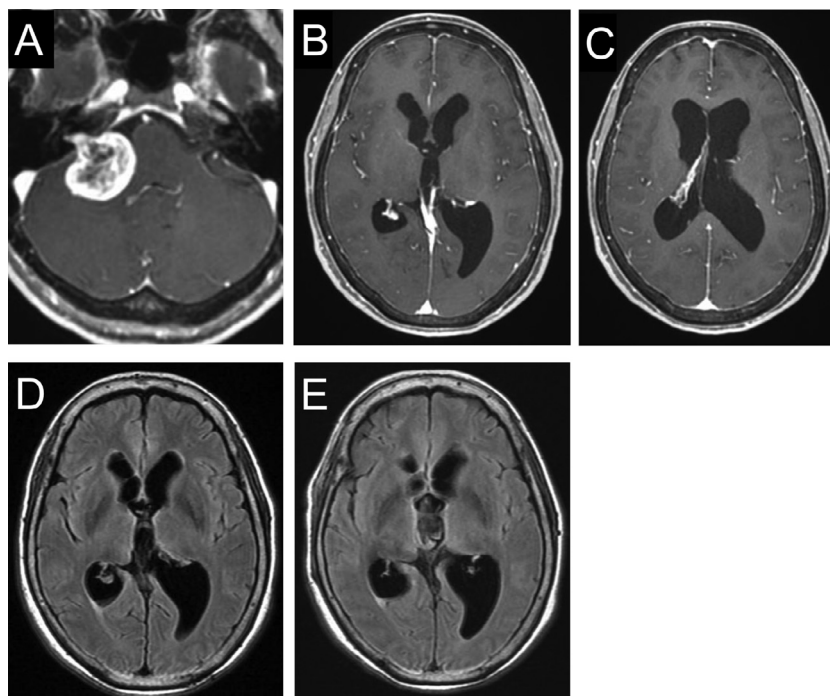


Fig. 2 – (A–C) At the 5 months after radiation, axial gadolinium-enhanced T1-weighted magnetic resonance images demonstrated moderate tumor growth about 30 mm with central necrosis, and with slightly more ventricular enlargement. **(D and E)** At the 5 months after radiation, axial fluid-attenuated inversion recovery magnetic resonance imaging confirmed that there was no periventricular hyperintensity area.

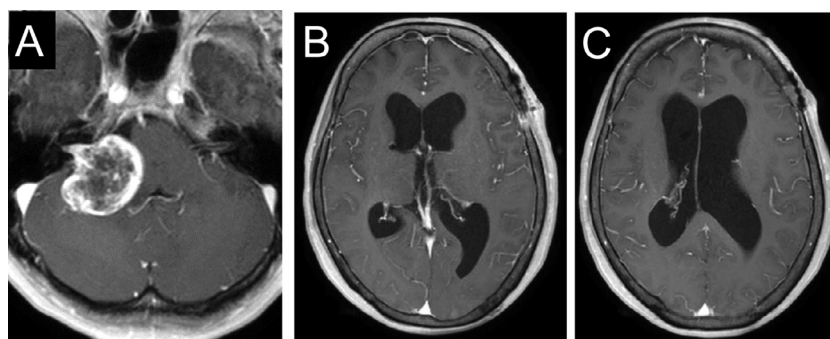


Fig. 3 – (A–C) Axial gadolinium-enhanced T1-weighted magnetic resonance images. When the patient was referred to our hospital, it revealed further exacerbated ventriculomegaly and tumor expansion to about 35 mm with the development of necrosis. The tumor was compressing the brain stem and shifting the fourth ventricle.

month follow-up, she had no symptoms, and the ventricle size decreased throughout the follow-up period on computed tomography findings. (Fig. 6B)

Discussion

We have reported a case of refractory communicating hydrocephalus after GKRS for small vestibular schwannoma with asymptomatic ventriculomegaly on the preoperative images.

For the enlarged tumor and refractory hydrocephalus after GKRS, we performed surgical removal of the tumor as the definitive treatment. Hydrocephalus was gradually improved and finally resolved on the both clinically and radiographically after tumor removal without shunt placement.

The incidence of hydrocephalus for vestibular schwannoma ranges from 3.7% to 18.0% of cases [5–7], and 3.0% to 14.0% of cases after GKRS [2,3,8–10]. GKRS has become a popular primary treatment option for small vestibular schwannoma up to approximately 25 mm in diameter. However the risk of developing hydrocephalus after GKRS might be high.

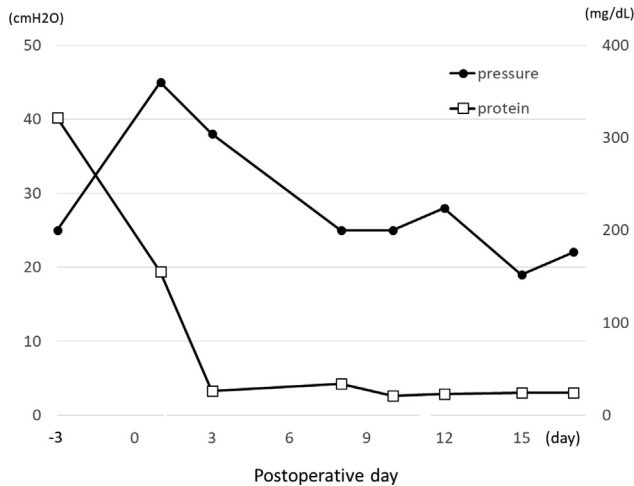


Fig. 4 – This diagram shows the perioperative intracranial pressure (ICP; black dot) and cerebrospinal fluid protein level (white square). First, the protein level gradually decreased; the ICP slightly decreased thereafter.

The pathogenesis of the development of hydrocephalus after treatment is not yet fully understood. Three potential mechanisms have been suggested, namely, compression of the fourth ventricle, protein shedding by a tumor necrosis and plugging of arachnoid granulation resulting in malabsorption, and alterations of CSF flow dynamics in basilar cisterns [2,5,7,11,12]. In our case, communicating hydrocephalus was exacerbated and become refractory after radiation by increasing the CSF protein. Therefore, GKRS may exacerbate these events.

Previous reports have suggested that the risk factors for developing hydrocephalus are the tumor volume [3,10], Koos grade [10], female sex [2,3], and the apparent diffusion coefficient (ADC) value of the tumor [1]. A large tumor with a high Koos grade causes not only obstructive hydrocephalus but also communicating hydrocephalus. Frischer et al reported the risk of hydrocephalus was 0.6% for patients with Koos grade II neuromas, 2.8% for those with Koos grade III, and 9.1% for those

with Koos grade IV [10]. The CSF protein concentration is correlated with the development of hydrocephalus, as is the tumor size [4]. Also, the mean ADC value of the tumor were related to the association of hydrocephalus [1]. They described that the cut-off for the tumor ADC value according to the receiver operating characteristic curve analysis was $1.35 \times 10^{-3} \text{ mm}^2/\text{s}$. In our case, the tumor rapidly increased from 20 to 35 mm in 5 months, the tumor deformed the brain stem and shifted the fourth ventricle (Koos grade IV), and the patient was female and the mean ADC value of the tumor was $1.52 \times 10^{-3} \text{ mm}^2/\text{s}$; therefore, the risk of hydrocephalus was high. We propose that tumor removal, not GKRS, should be considered in patients with a vestibular schwannoma accompanied by risk factors for developing hydrocephalus, even if the tumor is relatively small and the patient is asymptomatic.

Most case reports have indicated that a shunt procedure is the preferred treatment for communicating hydrocephalus due to a vestibular schwannoma [5]. In another study, patients with communicating hydrocephalus after GKRS for a vestibular schwannoma who underwent a shunt procedure generally had a good clinical course [3,13]. The incidence of hydrocephalus due to a vestibular schwannoma after tumor removal reportedly ranges from 0.40% to 0.68% of cases [8,14]. That is, tumor removal is known to not always resolve hydrocephalus, and shunt procedure may be required [15]. However, our patient developed shunt dysfunction twice and tumor enlargement, so we performed tumor removal. In previous reports, patients with preoperative hydrocephalus usually underwent a shunt procedure within about 1 week after tumor removal [16]. The use of repeated lumbar puncture with the intention to avoid the need for a shunt, although is not a widespread practice, it is a long-known strategy. In the present case, we could avoid a shunt procedure by repeated lumbar puncture in the early period after tumor removal. The CSF protein concentration decreased and the ICP improved within several days. Her symptoms gradually improved. Therefore, we propose that surgical removal of a vestibular schwannoma should be considered as an optimal strategy for refractory shunt dysfunction. Hydrocephalus after tumor removal can be managed by repeated lumbar puncture in the early postoperative days before considering shunt placement.

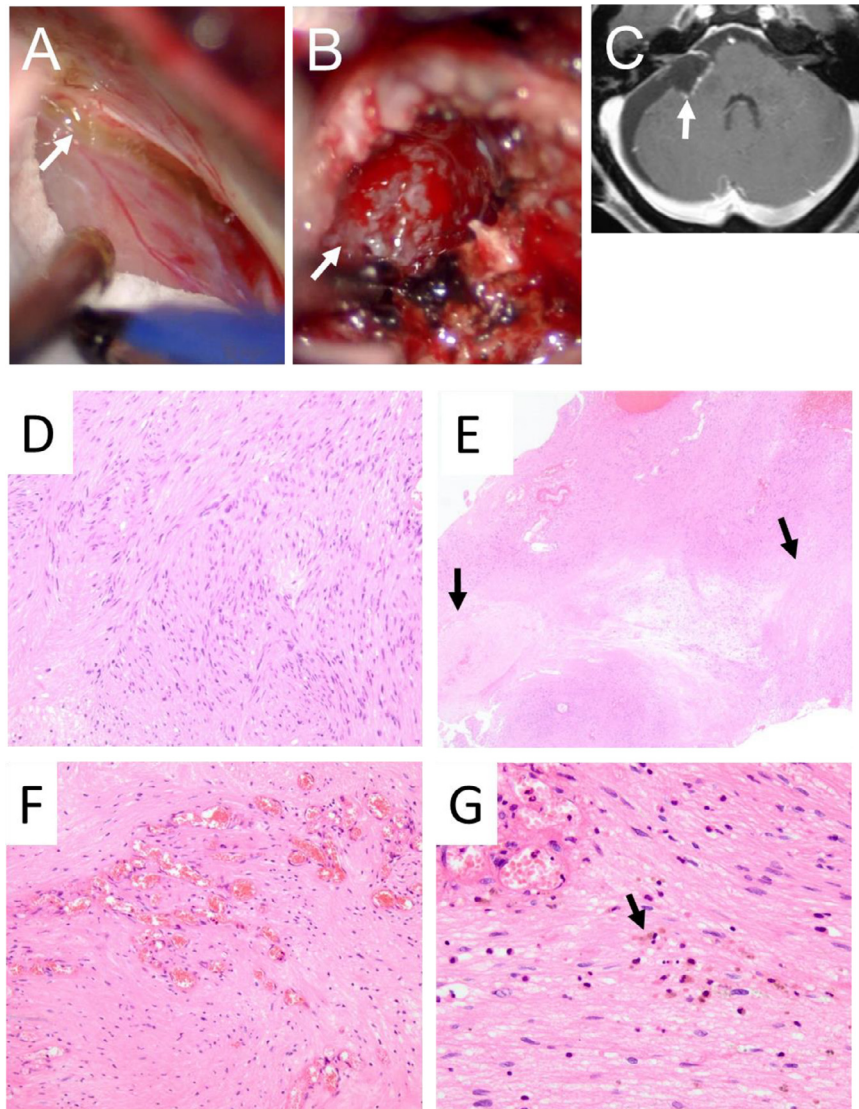


Fig. 5 – An intraoperative finding showed that (A) the arachnoid was thickened and partly yellow (arrow) and (B) the tumor was grayish, soft, and hemorrhagic (arrow). (C) Postoperative axial gadolinium-enhanced T1-weighted magnetic resonance images confirmed subtotal resection of the tumor (arrow head) except for anchoring of central nerve VII. Histological microphotographs showed the typical pattern of a schwannoma with (D) palisading nuclei (hematoxylin and eosin (H&E) stain, $\times 400$), (E) necrosis (arrow, H&E stain, $\times 40$), (F) vascular proliferation (H&E stain, $\times 100$), and (G) hemosiderosis (arrow, H&E stain, $\times 100$).

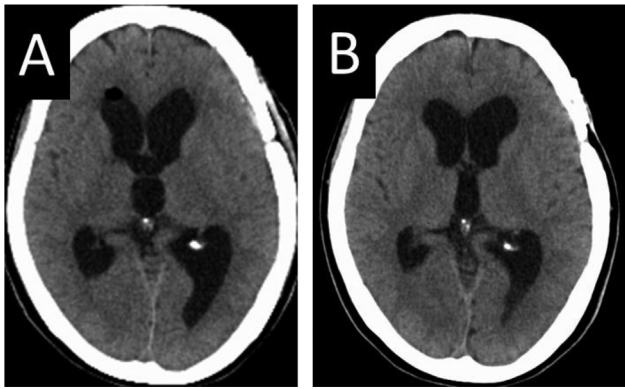


Fig. 6 – Axial computed tomography scan was performed on postoperative days 1 (A) and 155 (B). The ventricle size decreased throughout the follow-up period.

Conclusion

Communicating hydrocephalus can be exacerbated and become refractory after radiation for vestibular schwannoma by increasing the CSF concentration. Surgical removal of the tumor should be considered as the first option for the treatment, even if the patient is asymptomatic and the images revealed a small vestibular schwannoma with only slight ventricular enlargement.

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