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Improvement of Isolated Abducens Nerve Palsy with Hydrocephalus after CSF Diversion: A Possible Evaluative Role of Retroclival-pontomedullary Distance

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Abstract

Isolated abducens nerve palsy (IANP), caused by secondary communicating hydrocephalus, has been rarely documented; in addition, its mechanism and appropriate treatment are not understood well. This study presents a case of bilateral IANP with hydrocephalus in a 62-year-old man who was successfully treated with cerebrospinal fluid (CSF) diversion to correct an enlarged retroclival space during the follow-up of recurrent brain tumor in the right parieto-occipital lobe. The patient was treated with three resections, temozolomide, and irradiation before developing IANP. Magnetic resonance imaging (MRI) revealed a recurrent tumor and ventriculomegaly with an expanded retroclival cisternal space. The patient underwent subtotal tumor resection and external ventricular drain placement in the anterior horn of the lateral ventricle. His bilateral IANP persisted for 4 days after surgery but gradually improved and disappeared by Day 7. Four weeks later, the patient underwent ventriculoperitoneal (VP) shunt surgery to establish a permanent CSF diversion that continued to control the symptoms. Retrospective MRI review revealed the distance between the clivus and pontomedullary junction on the sagittal section (retroclival-pontomedullary distance; RPD) of 9.0, 12.8, 10.7, and 10.6 mm before IANP, on IANP onset, on postoperative Day 4, and post VP shunt surgery, respectively. In conclusion, VP shunt surgery was an appropriate approach for IANP with communicating hydrocephalus to correct the enlarged retroclival cisternal space. RPD thus may be used as one of possible evaluation methods for IANP with hydrocephalus, which can be caused by various factors.

Keywords: isolated abducens nerve palsy, hydrocephalus, VP shunt

Introduction

Isolated abducens nerve palsy (IANP) is a major cranial neuropathy resulting in decreased everyday activities due to diplopia.[1)] Various etiologies affecting any portion of the abducens nerve can cause IANP, including trauma, demyelinating diseases, inflammatory diseases, infections, tumors, subarachnoid hemorrhage, neurovascular compression, increased intracranial pressure (ICP), and communicating hydrocephalus.[2-6)] Hydrocephalus with an increased ICP is a common cause of IANP.[4)] However, the mechanism and appropriate treatment have not been understood well. Additionally, to the best of our knowledge, no method has been proposed to evaluate IANP quantitatively.

This study presents a case of bilateral IANP with communicating hydrocephalus and a brain tumor successfully treated by ventriculoperitoneal (VP) shunt surgery, while correcting the retroclival-pontomedullary distance (RPD). Here, we discuss the surgical approach for IANP in patients with hydrocephalus based on a literature review and the mechanism inferred from the quantitative evaluation we used.

Case Report

A 62-year-old man initially presented with headache,

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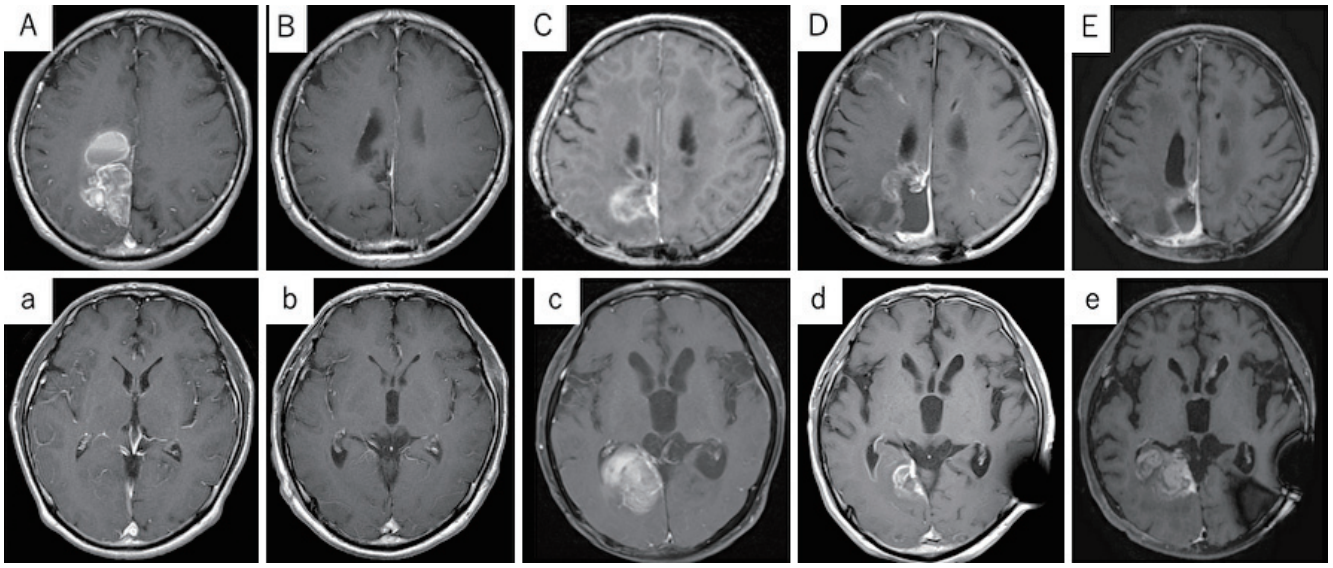

Fig. 1 Pre- and postoperative MRI images.
Images denoted by capital letters show the tumor progression, and those denoted by lowercase letters show the progression of ventricular enlargement.
A, a: A preoperative contrast-enhanced T1-weighted MRI (T1W-CE MRI) at initial presentation showing a neoplastic lesion with a maximum diameter of 2 cm in the right parietal lobe. The fluid-fluid level in the tumor indicates hemorrhagic nature. In addition, ventricles are of normal size.
B, b: A T1W-CE MRI 3 months after surgery shows no obvious contrast-enhanced lesions, and ventricular enlargement is not observed.
C, c: A T1W-CE MRI at the onset of isolated abducens nerve palsy shows a recurrent lesion and ventriculomegaly not observed in prior imaging study.
D, d: A postoperative T1W-CE MRI following VP shunting shows slightly improved ventricular enlargement.
E, e: A T1W-CE MRI 2 months after shunting shows no tumor growth and no ventricular re-enlargement.

numbness, and weakness on his left side at the age of 56. Magnetic resonance imaging (MRI) revealed a hemorrhagic lesion in the right parietal lobe (Fig. 1A). Since the hemorrhage was considered to result from a benign tumor, the patient was given a wait-and-scan plan at another hospital. However, due to the progression of numbness, motor weakness on his left side, and tumor growth, the patient was eventually referred to our hospital and underwent gross total tumor resection at the age of 57 years. MRI after the initial surgery revealed no remaining contrast-enhanced lesions (Fig. 1B). Histopathological examination revealed pilocytic astrocytoma. Two years later, a second resection was performed for tumor recurrence; the pathology showed malignant transformation, consistent with a diagnosis of pilocytic astrocytoma with anaplasia (not as a World Health Organization classification type; diagnosed descriptively). Thus, the patient underwent chemotherapy (temozolomide) and irradiation with 60 Gy/30 Fr. Adjuvant chemotherapy with temozolomide was continued for 2 years.

A year later, the patient developed diplopia and his daily activity decreased owing to impaired mobility. Extraocular muscle function tests revealed bilateral IANP (Fig. 2A). MRI revealed a recurrent lesion and communicating ventriculomegaly that had not been observed during his prior visit (Fig. 1C). A lumber puncture was performed before surgery, and we found that the protein level in cerebrospinal fluid (CSF) was 65 mg/dL (institutional normal range: 8-43 mg/dL), although the opening pressure was not documented. Therefore, tumor resection was performed via occipitoparietal craniotomy and external ventricular drainage (EVD) of the left anterior horn of the lateral ventricle. After the surgery, his consciousness improved slightly. The opening pressure of the EVD was 15 cm H_2O under general anesthesia. However, it eventually stabilized at 20 cm H_2O following EVD placement surgery, with a daily drainage volume of 300 mL. The pre- and postoperative CSF properties suggest the presence of secondary communicating hydrocephalus, with relatively higher CSF pressure within the normal range. His bilateral IANP persisted for 4 days following surgery but gradually improved and disappeared by postoperative Day 7 (Fig. 2B). One month after surgery, given a relatively higher EVD pressure of 20 cm H_2O, the daily drainage of CSF of 300 mL, the prolonged enlargement of the ventricles, and the improvement on alertness following EVD placement, we decided to perform ventriculoperitoneal shunt (VP shunt). The patient underwent VP shunt surgery to establish permanent CSF diversion (Fig. 1

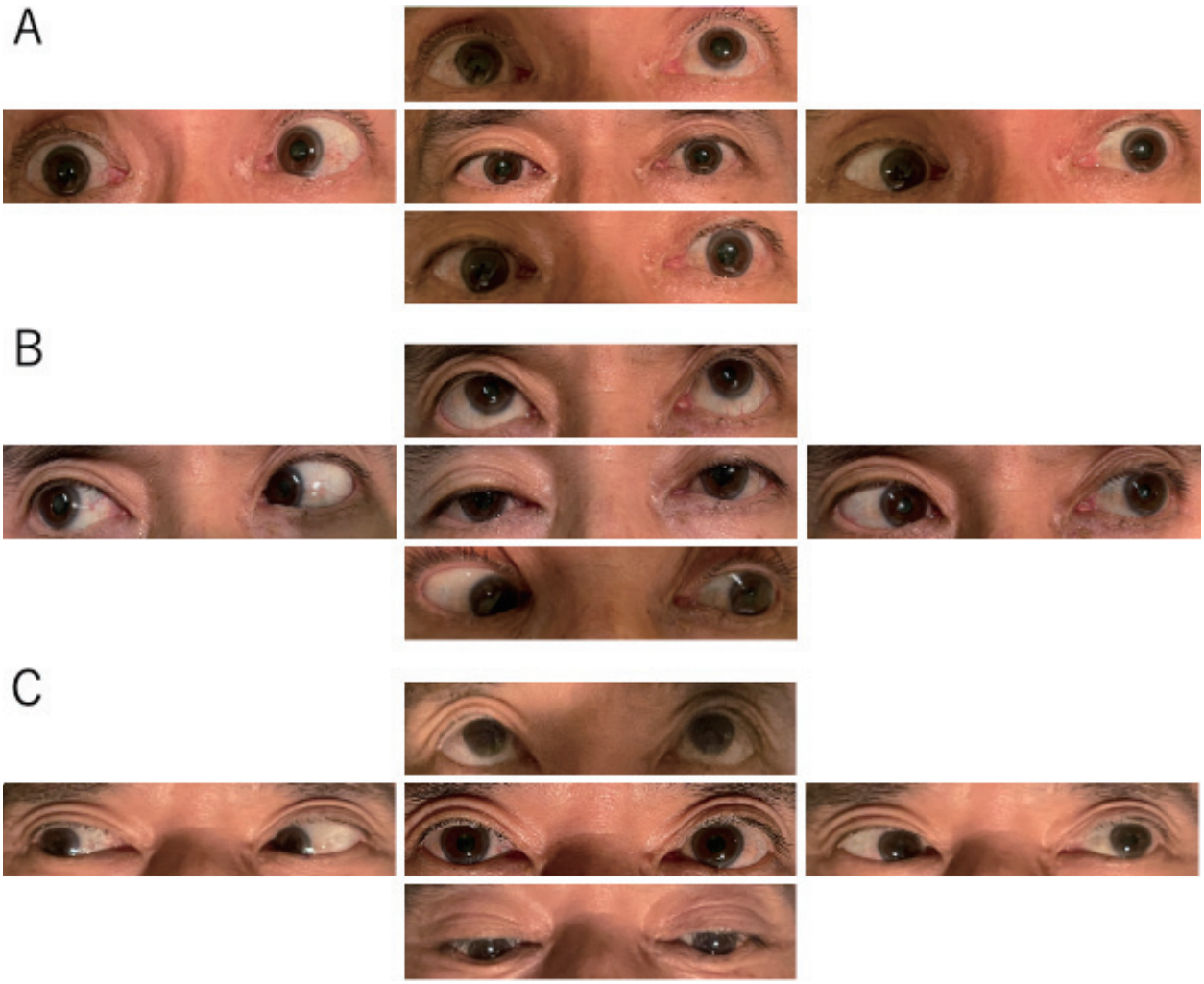

Fig. 2 Extraocular muscle function tests at the onset, postoperatively, and immediately before discharge.
Extraocular muscle function test at onset (A) shows that the sclera of the outer pupil of the right eye is visible when looking to the right, and vice versa when looking to the left, indicating that the abduction of both eyes is impaired. The test at postoperative Day 7 (B) shows the virtually unrestricted range of motion for abduction in both eyes. The improvement was retained following VP shunting at the time of discharge, 2 months postoperatively (C).

D). A retrospective MRI review revealed that the distances between the clivus and the pontomedullary junction on the sagittal section (RPD) were 9.0, 12.8, 10.7, and 10.6 mm (4 months before surgery: baseline; 1 month before surgery: onset of IANP; postoperative Day 4: during EVD; and 1.5 months following tumor surgery: after VP shunt, respectively; Fig. 3), consistent with the onset and improvement in ocular motor abnormality. Due to reduced daily activities, he was moved to a rehabilitation hospital 2 months after tumor surgery, with no progression of hydrocephalus (Fig. 1E), and the bilateral IANP disappeared at discharge (Fig. 2C). Eventually, the patient passed away due to tumor progression 3 months after discharge.

Discussion

Here, we present a case of bilateral IANP with communicating hydrocephalus and a recurrent brain tumor that was successfully treated with a VP shunt, while correcting the enlarged retroclival space. The clinical course of this patient suggested that bilateral IANP associated with communicating hydrocephalus could be improved by a VP shunt and that the expanding retroclival cisternal space may be a pathological IANP mechanism.

The causes of IANP include abducens nerve kink at the entry of Dorello's canal, water hammer effect, tumors, inflammation, trauma, demyelinating diseases, infections, subarachnoid hemorrhage, neurovascular compression, increased ICP, and communicating hydrocephalus.[2-6] Increased ICP is a possible mechanism underlying IANP in

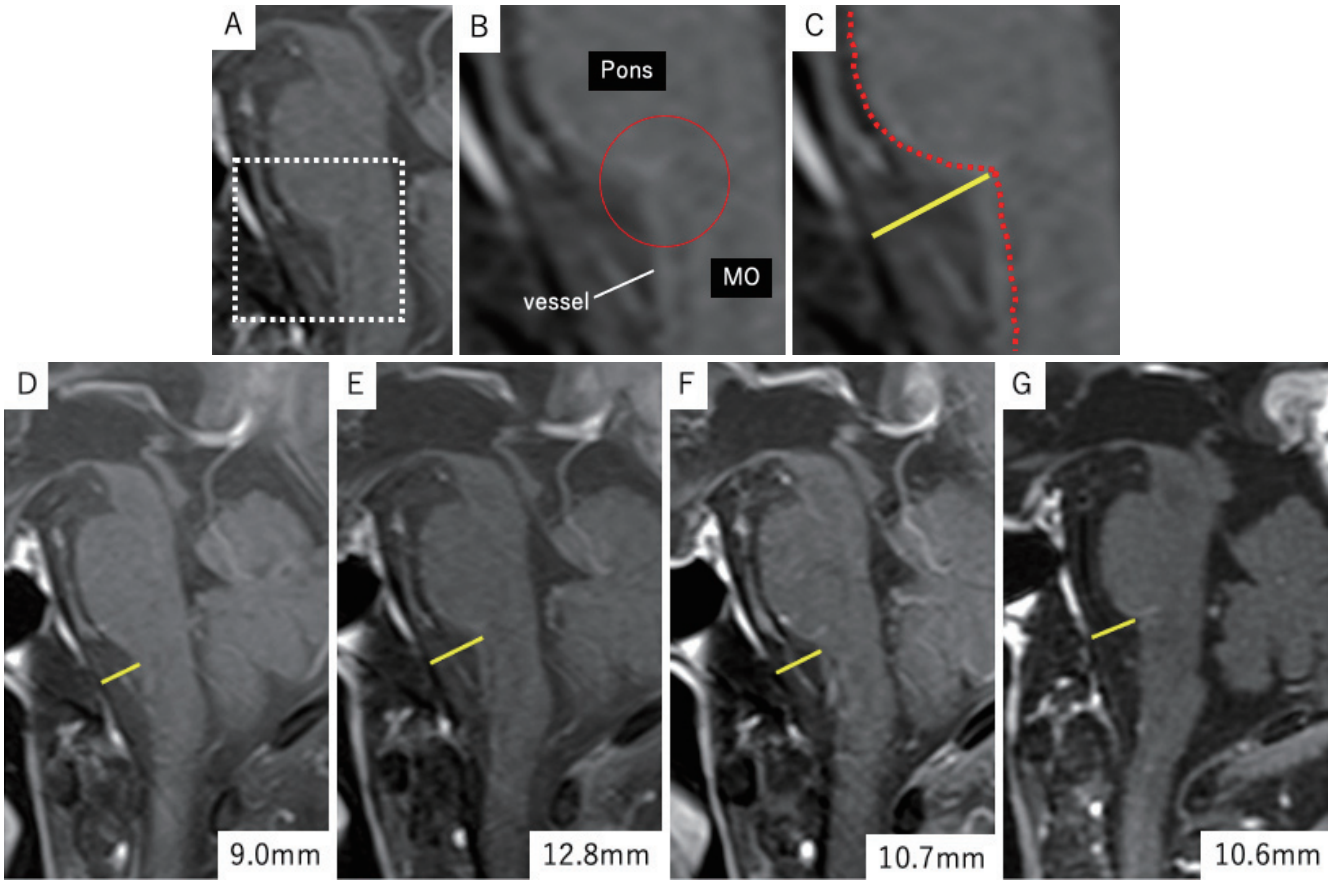

Fig. 3 The distance measurement between the clivus and the pontomedullary junction on the sagittal section: Retroclival-pontomedullary distance (RPD).
Panel A shows the plain sagittal MRI. Panel B (maginified images of A) shows that the basilar or vertebral artery runs in front of the medulla oblongata. Panel C shows the anterior border of the brainstem (red dot line), and the distance measurement between the clivus and the pontomedullary junction on the sagittal section (RPD; yellow line) by drawing a perpendicular line from the pontomedullary junction to the clivus. The measurement shows changes in the length of 9.0, 12.8, 10.7, and 10.6 mm at 4 months before (D: baseline); 1 month before (E: onset of symptom); 4 days after (F: during external ventricular drainage); and 2 months after surgery (G: after VP shunt), respectively.

hydrocephalus.[4] In contrast, spinal overdrainage, which leads to decreased ICP, can also cause IANP, suggesting another mechanism for IANP in disorders of CSF dynamics.[7] The expanded retroclival space can worsen IANP by shifting the brainstem posteriorly and caudally, which stretches the abducens nerve.[8] This is because the abducens nerve is anchored between Dorello's canal in the retroclival region and the pontomedullary junction of the brainstem. This presumed mechanism has been previously demonstrated in several cases. The first is the case of IANP resulting from shunt overdrainage from the fourth ventricle in an isolated fourth ventricle, which stretched the abducens nerve by the backward shift of the brainstem.[9] The second is the retroclival pneumocephalus after trauma compressed the brainstem and stretched the abducens nerve, resulting in the IANP.[10] In the third case, the direction of aneurysmal rupture toward Dorello's canal and the thick cisternal hematoma between Dorello's canal and the brainstem were associated with IANP.[11,12] Finally, in case of Chiari malformation, correction of craniocaudal deviation by foramen magnum decompression improved IANP. In line with these, our case revealed a direct correlation between the enlargement of the retroclival space and the onset of IANP. In this context, an objective evaluation of the shift can assist in identifying a contributor of IANP. To capture the anterior-posterior shift quantitatively, we measured the distance between the petroclival and pontomedullary junctions on sagittal head section images and named it RPD. The RPD enlargement may have been one of the contributors to IANP in this case.

Regardless of the mechanism, CSF diversion can be used for managing IANP with hydrocephalus by normalizing ICP or correcting the CSF space deformity. However, overdrainage by lumbo-peritoneal shunt may cause downward sagging of the brain, resulting in traction on the abducens nerve and symptom exacerbation.[7] Therefore, a VP

shunt may be a safer option. Regarding the timing of treatment initiation, no clear strategy has been described in the literature in which IANP was improved by a VP shunt. In this case, treatment was initiated within 1 month after the symptoms' onset, indicating that a range of therapeutic time window may exist for IANP with hydrocephalus.

This study also has some limitations. The pathology of the rare combination of pilocytic astrocytoma with anaplastic features, communicating hydrocephalus, and IANP was explored. A possible explanation for developing communicating hydrocephalus in anaplastic pilocytic astrocytoma is the hemorrhagic nature of the tumor. Additionally, neuroinflammation induced by repeated surgeries may decrease tolerance to nerve stretching, potentially contributing to IANP in communicating hydrocephalus.[13] IANP is uncommon in most patients with communicating hydrocephalus. However, in the demonstrated case, neuroinflammation or other factors following multiple surgeries may have affected the abducens nerve, resulting in IANP with a change in RPD of only a few millimeters. In addition, while the RPD measurement method used reflects shifts in the anteroposterior direction, it may not capture shifts in the craniocaudal direction. Furthermore, the cutoff value for RPD could not be determined because this was a single case. Thus, our findings require further investigation in a larger population.

Nonetheless, our case showed that VP shunting is an effective surgical treatment of isolated bilateral abducens nerve palsy with communicating hydrocephalus, suggesting a potential utility of RPD in this clinical condition. The points discussed here should be investigated in future studies.

Conclusion

VP shunting is effective in the treatment of isolated bilateral abducens nerve palsy associated with communicating hydrocephalus. Additionally, an expanded retroclival cisternal space, which could exacerbate nerve stretching, might be a contributing factor to isolated bilateral abducens nerve palsy. Therefore, the RPD measurement could potentially be used in evaluating this clinical condition.

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Conflicts of Interest Disclosure

There are no conflicts of interest to declare.

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