

Aqueductal Developmental Venous Anomaly Presenting with Mimic Symptoms of Idiopathic Normal Pressure Hydrocephalus in an Elderly Patient: A Case Report

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Developmental venous anomalies (DVAs) are generally asymptomatic; however, they can sometimes cause central nervous disorders. Aqueductal stenosis caused by DVAs is so rare that only 14 cases have been reported to date. Moreover, most patients are children or young adults, presenting with headaches or consciousness disturbances, associated with raised intracranial pressure. Here, we report on an 83-year-old man presenting with mimic symptoms of idiopathic normal pressure hydrocephalus (cognitive disorder, gait disturbance, and urinary urgency: Hakim's triad) because of obstructive hydrocephalus caused by a DVA located in the aqueduct. Endoscopic third ventriculostomy (ETV) was performed to relieve his symptoms, and the opening pressure of the lateral ventricle was recorded to be 10 cm-H₂O. Endoscopic examination of the intraventricular system clearly revealed a vein within the aqueduct converging with the adjacent subependymal vessels. These findings were compatible with the characteristics of DVAs. His symptoms improved after the ETV. This case suggested that DVAs within the aqueduct, despite of their congenital nature, could give rise to decompensated obstructive hydrocephalus even in elderly patients, resulting in Hakim's triad.

Keywords: developmental venous anomaly, aqueductal stenosis, obstructive hydrocephalus, endoscopic third ventriculostomy

Introduction

Developmental venous anomalies (DVAs), previously termed venous malformations, are currently considered congenital non-pathologic anatomical variants of normal deep parenchymal veins, which are composed of mature venous vessels that lack malformed or neoplastic elements.^{1,2)} Although they are benign entities in nature, they occasionally cause headaches, seizures,

and some focal neurological deficits.^{3,4)} Obstructive hydrocephalus as a complication of DVAs is so rare that only 14 cases of aqueductal stenosis (AS) have been reported so far.^{5,6)} The previously reported cases of AS induced by DVAs were mainly in children and young adults, manifesting as chronic or intermittent headaches or consciousness disturbances resulting from elevated intracranial pressure (ICP).^{5,7)}

Here, we present a case of AS caused by a DVA in an 83-year-old man, manifesting symptoms of Hakim's triad,⁸⁾ with a normal ICP; this is the oldest patient reported upon so far. We present the radiological and endoscopic findings related to this uncommon entity, and then discuss the characteristics of clinical manifestations.

Case Presentation

An 83-year-old man, with Hakim's triad, cognitive disorder, gait disturbance, and urinary urgency over the past 4 months, was referred to our hospital. His general condition was good, with neither nausea nor headaches.

Magnetic resonance imaging (MRI) assessment showed supratentorial ventricular dilatation with ballooning of the third ventricle floor, which was consistent with radiological findings observed in obstructive hydrocephalus caused by AS (Figs. 1A–1C). A faint gadolinium-enhanced lesion was found at the dorsal part of the aqueduct, although it was difficult to determine whether the lesion contributed to the hydrocephalus (Fig. 1B, magnified image).

To resolve the symptoms associated with hydrocephalus, as well as to investigate the causative lesion, endoscopic third ventriculostomy (ETV) to inspect the ventricular system was carried out using a flexible neuro-fiber endoscope (VEF-V, Olympus Corporation, Tokyo, Japan). The right-side anterior horn of the lateral ventricle was punctured with least possible leakage of cerebrospinal fluid (CSF), and an initial opening pressure of 10 cm-H₂O was recorded. During endoscopy, we identified a vein accompanying the subependymal veins within the aqueduct (Fig. 1D). The appearance of this vascular lesion was compatible with the characteristics of a DVA, with an enlarged transcortical or subependymal collector vein draining radially arranged medullary veins. ETV was then performed in the conventional manner; his postoperative course was uneventful. His clinical symptoms remained good for 6 months after the operation, as summarized in Table 1. Follow-up MRI showed the flow void sign at the third ventricle floor, revealing patency of the stoma (Fig. 1E).

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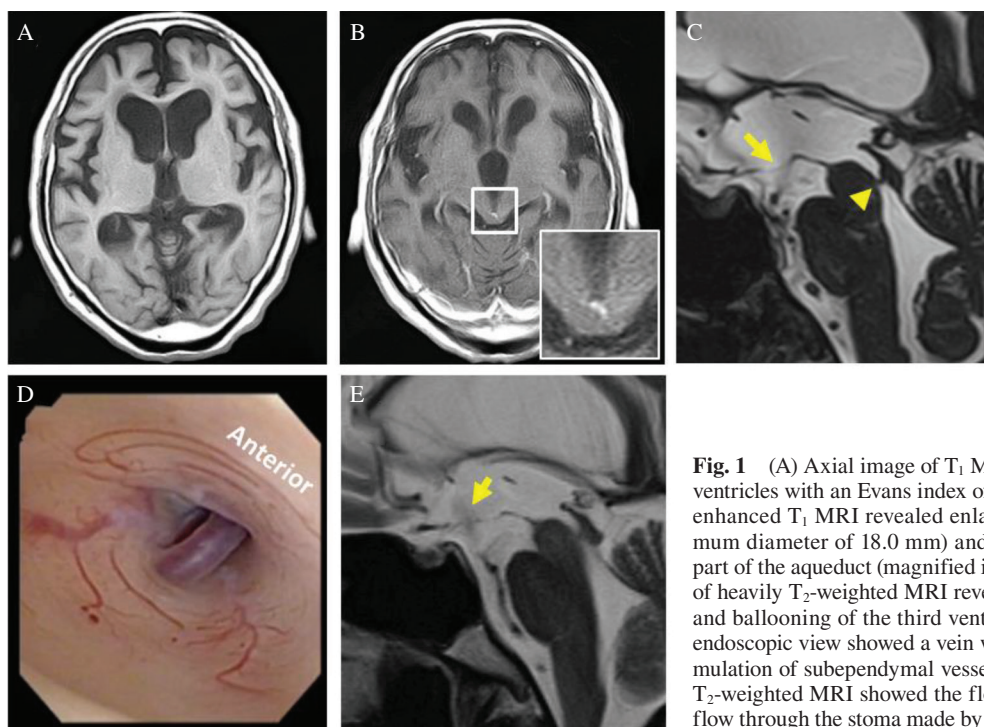


Fig. 1 (A) Axial image of T₁ MRI revealed enlargement of the lateral ventricles with an Evans index of 0.39. (B) Axial image of gadolinium-enhanced T₁ MRI revealed enlargement of the third ventricle (maximum diameter of 18.0 mm) and a faint enhancement at the posterior part of the aqueduct (magnified image). (C) Preoperative sagittal image of heavily T₂-weighted MRI revealed aqueductal stenosis (arrow head) and ballooning of the third ventricle floor (arrow). (D) Intraoperative endoscopic view showed a vein within the aqueduct and adjacent accumulation of subependymal vessels. (E) Postoperative sagittal image of T₂-weighted MRI showed the flow artifact (arrow), revealing the CSF flow through the stoma made by endoscopic third ventriculostomy.

Table 1 Clinical evaluation in pre- and post-ETV

Evaluations	Pre-ETV	Post-ETV
MMSE (full score; 30)	15	20
3 m TUG (s)	25	14
INPHGS total score	6	3
Gait	2	0
Cognition	3	2
Incontinence	1	0
Modified Rankin Scale	2	1

ETV: endoscopic third ventriculostomy, INPHGS: idiopathic normal pressure hydrocephalus grading scale, TUG: timed up and go.

Discussion

Aqueductal stenosis may be induced by various central nervous system disorders, such as infection, hemorrhage, head trauma, tumors, cysts, or vascular lesions.^{9,10} AS caused by DVAs is so rare that only 14 cases have been reported previously, all of which involved infants and relatively young individuals (0–58 years) (Table 2).^{5–7,11–20} Apart from mental deterioration in the case of a 58-year-old woman¹¹ and seizure in the case of a 43-year-old woman,¹⁴ the main symptoms in the remaining 12 cases (0–42 years) were severe or intermittent headaches and consciousness disturbances, which were closely associated with raised ICP.

Taking into consideration the congenital nature of DVAs, this case is quite unique, in that the intra-aqueductal DVA had been asymptomatic until he manifested with Hakim’s triad at the age of 83. Although the underlying mechanisms of very late onset obstructive hydrocephalus in this patient is

unclear, the clinical characteristics in this case are consistent with a previous study on idiopathic AS. Younger patients tend to have headaches with raised ICP, whereas older patients tend to show partial or complete Hakim’s triad with normal ICP.²¹ Long-standing intra-aqueductal DVAs, together with senile changes in the craniospinal environment, such as decrease in brain compliance, might induce decompensated obstructive hydrocephalus.^{22,23}

In cases of obstructive hydrocephalus caused by AS, ventriculoperitoneal shunting (VPS) with programmable pressure valves is another treatment choice. On the other hand, it was reported that the failure rate of initial VPS was much higher than ETV for adult patients with AS.²⁴ Regarding the first-line treatment option for patients with adult AS whose CSF absorbable capacity is expected to be preserved, ETV may be recommended rather than VPS, because ETV can prevent various problems related to VPS, such as over-drainage, shunt malfunction, or infection.

Moreover, in this case, use of a flexible endoscope for ETV enabled us to identify the DVA within the aqueduct, which was difficult to detect on preoperative MRI. Together with the previous eight cases involving the use of endoscopes (Table 2),^{5,14,17,19,20} this case suggests that neuro-fiberscopes are not only useful for the treatment of AS but are also helpful in diagnostic exploration for AS with uncommon etiologies.

Conclusion

We presented a rare case of an elderly man with mimic symptoms of idiopathic normal pressure hydrocephalus (iNPH) caused by a DVA within the aqueduct. This case was quite unique for the following reasons: (1) the patient was much older than those in previous reports, (2) he showed

Table 2 Previously reported cases of aqueductal stenosis due to developmental venous anomaly

Age (yr)	Sex	Symptoms	Duration	Location of DVA	Findings of DVAs	Diagnostic procedure	Treatment	References
58	M	Mental deterioration	5 yr	Aqueduct, 4th ventricle	An angioma blocking the aqueduct and the 4th ventricle	Autopsy	No	11
35	F	Headache	1 yr	Aqueduct	A venous element partially blocking the aqueduct	Angiography, direct exploration	Stenting	12
39	M	Headache	1 yr	Lower aqueduct	An abnormal draining vein penetrating the lowest part of the tectum mesencephali	MRI	VP shunt	13
43	F	Seizure	2 mo	Periaqueduct	An enlarged pulsatile vein protruding into the aqueduct on the left side	MRI, angiography, endoscopy	ETV	14
16	F	Headache, behavior abnormalities	2 mo	Periaqueduct	Vascular channels partially surrounding the aqueduct	MRI	No	15
11	M	Headache	5 mo	Aqueduct	A draining vein traversing the aqueduct	MRI	Shunting	16
28	F	Headache, diplopia	n.a.	Ostial aqueduct, 3rd ventricle	An abnormal vein passing through the orifice of the aqueduct	MRI, endoscopy	ETV	17
7	F	None	1 mo	Aqueduct	An abnormal vein coursing through the aqueduct	MRI	No	18
42	M	Headache, behavior abnormalities	1 yr	Ostial aqueduct, 3rd ventricle	Two draining veins passing near the orifice of the aqueduct	MRI, endoscopy	ETV	5
18	M	Headache	6 yr	Aqueduct (venous loop)	A loop of the subependymal DVA protruding into the aqueduct	MRI, endoscopy	ETV	5
0 (10 mo)	M	Delayed psychomotor development	10 mo	Ostial aqueduct	Large veins of the DVA compressing the entry into the aqueduct	MRI, endoscopy	ETV	19
0 (Birth)	F	Increased head circumference	–	Ostial aqueduct, 3rd ventricle	A DVA passing through the orifice of the aqueduct	MRI	VP shunt	20
10	M	Headache, nausea, vomiting	2 wk	Aqueduct	A supra- and infratentorial DVA coursing through the aqueduct	MRI, endoscopy	ETV	21
37	M	Headache	1 yr	Aqueduct	A large collector vein coursing within the aqueduct	MRI, angiography	ETV	6
83	M	Hakim's triad	4 mo	Aqueduct	A DVA coursing within the aqueduct	MRI, endoscopy	ETV	This case

DVA: developmental venous anomaly, ETV: endoscopic third ventriculostomy, mo: month(s), n.a.: not available, VP shunt: ventriculoperitoneal shunt, wk: week(s), yr: year(s).

symptoms of Hakim's triad, which were resolved by ETV, and (3) an intra-aqueductal DVA was clearly shown on endoscopy. Even in elderly patients manifesting Hakim's triad, not only iNPH, but also obstructive AS caused by congenital lesions, such as DVAs shown in this case, should be considered as differential diagnosis.

Conflicts of Interest Disclosure

The authors have no conflict of interest to declare. This work was supported by a grant-in-aid for scientific research from Japanese Ministry of Health, Labor, and Welfare (# H26-NANCHITOU-IPPAN-052, to D.K.).

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