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Case Report

Aqueductal developmental venous anomaly causing obstructive hydrocephalus: A case report and review of the literature ☆

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ABSTRACT

A developmental venous anomaly (DVA) is a venous drainage of the associated parenchyma that is normally asymptomatic. However, a DVA located adjacent to the aqueduct can cause obstructive hydrocephalus by blocking the flow of cerebrospinal fluid. We describe a rare case of obstructive hydrocephalus due to aqueductal stenosis secondary to a DVA. A 43-year-old man presented with sudden bilateral temporal pain during weight training. Using a 3-Tesla scanner, cranial magnetic resonance imaging (MRI) was performed, and hydrocephalus was found with mild enlargement of the lateral and third ventricles. Susceptibility-weighted imaging and postcontrast MRI revealed that the DVA from the bilateral thalami narrowed the orifice of the aqueduct on its drainage route towards the vein of Galen. We assumed that force exerted during weight training may have caused dilation of the anomalous veins, leading to his symptom. A review of the relevant English-language literature yielded only 19 cases of aqueductal stenosis due to DVA. In comparison to these cases, the duration of symptom in our case was extremely short because the patient had a history of ventriculomegaly detected on plain computed tomography and was diagnosed quickly based on the characteristic finding of DVA: the caput medusae appearance.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Introduction

Developmental venous anomalies (DVAs) are the most common cerebrovascular malformations, occurring in 2%-3% of the population [1,2]. This malformation was previously known as venous angioma, venous malformation, or medullary venous malformation [3]. Currently, DVA is the most commonly used term. Normally, a DVA involves venous drainage of its associated parenchyma without any symptoms. However, in rare situations, it can cause symptoms. In some cases, a DVA can be located adjacent to the aqueduct and block the flow of cerebrospinal fluid (CSF), leading to obstructive hydrocephalus [1]. We present a rare case of obstructive hydrocephalus caused by stenosis of the aqueduct due to a DVA.

Case report

A 43-year-old man with a medical history of mild asymptomatic ventriculomegaly, which was detected using plain computed tomography 17 years previously, presented with sudden bilateral temporal pain during weight training. The patient underwent cranial magnetic resonance imaging (MRI) with and without a gadolinium-based contrast agent on the day following the onset using a 3-Tesla scanner (Magnetom Skyra; Siemens Healthineers, Erlangen, Germany).

T2-weighted imaging (T2WI) revealed hydrocephalus with mild enlargement of the lateral and third ventricles and a linear flow void midline along the floor of the third ventricle, while the fourth ventricle was not dilated (Fig. 1). Susceptibility-weighted imaging (SWI) and postcontrast T1-weighted imaging (T1WI) showed a large abnormal vein running posteriorly upward along the floor of the third ventricle ultimately drained into the vein of Galen (Figs. 2 and 3). This prominent vein was formed by the aggregation of medullary veins from the bilateral thalami and basal ganglia, similar to the caput medusae, and bridged across the orifice of the aqueduct just before draining into the Galenic system. Thus, these abnormal veins were identified as a DVA and considered to be the underlying cause of obstructive hydrocephalus. SWI also depicted an old hemorrhage at the right globus pallidus adjacent to the medullary veins of the DVA (Fig. 2). We hypothesized that the dilation of the anomalous veins caused by exerting force during weight training worsened the stenosis of the aqueduct. This exacerbation of stenosis would temporally lead to further ventricular enlargement and increased intracranial pressure, which could result in headache. The headache was relieved within a few days of the onset, without the use of medication. Surgical procedures such as endoscopic third ventriculostomy (ETV) were not performed because of the patient's mild and transient symptom. The patient's clinical course was uneventful.

Discussion

Generally, hydrocephalus is characterized by the abnormal enlargement of the cerebral ventricles due to an imbalance between CSF production and absorption. There are two types of hydrocephalus: communicating hydrocephalus and noncommunicating hydrocephalus. The former type includes malabsorptive hydrocephalus, hypersecretory hydrocephalus, and idiopathic normal pressure hydrocephalus, whereas the latter type includes only obstructive hydrocephalus. Regarding obstructive hydrocephalus, obstruction of the CSF pathways enlarges the ventricles upstream of the flow according to the classic CSF circulation model, known as the "bulk flow model" [4,5].

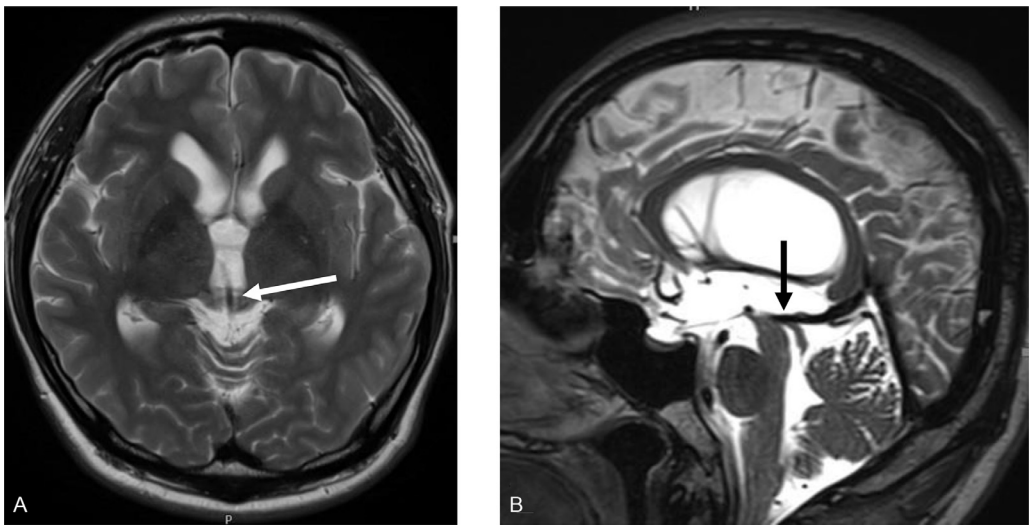

Fig. 1 – T2-weighted imaging obtained on the day following the onset shows mild enlargement of the lateral and third ventricles. The fourth ventricle is not dilated. A linear flow void is depicted midline along the floor of the third ventricle (*arrows*).

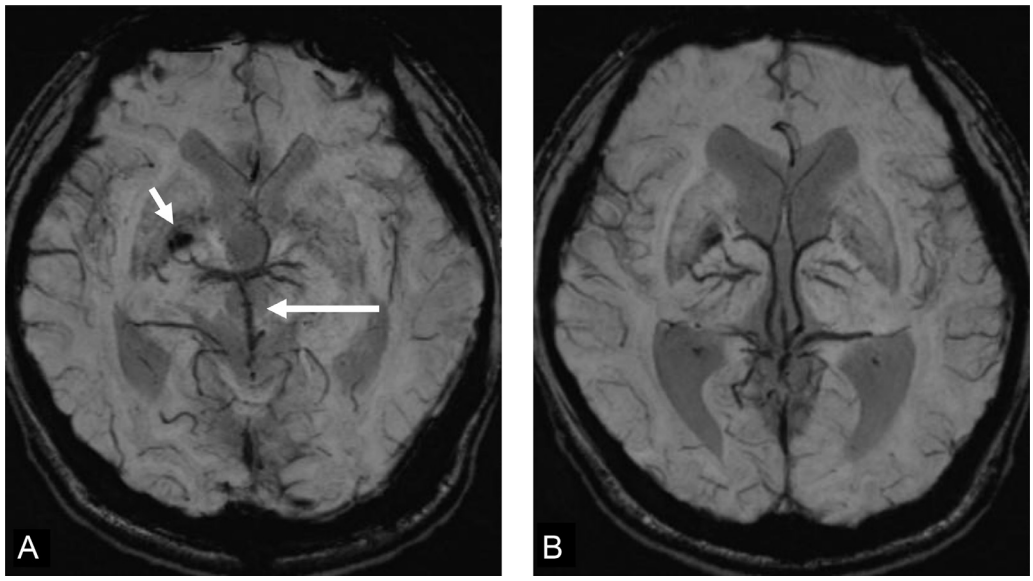

Fig. 2 – Susceptibility-weighted imaging obtained on the day following the onset shows a developmental venous anomaly (DVA) starting from the bilateral thalami and basal ganglia (*long arrow*). Old hemorrhage is depicted at the right basal ganglia (*short arrow*).

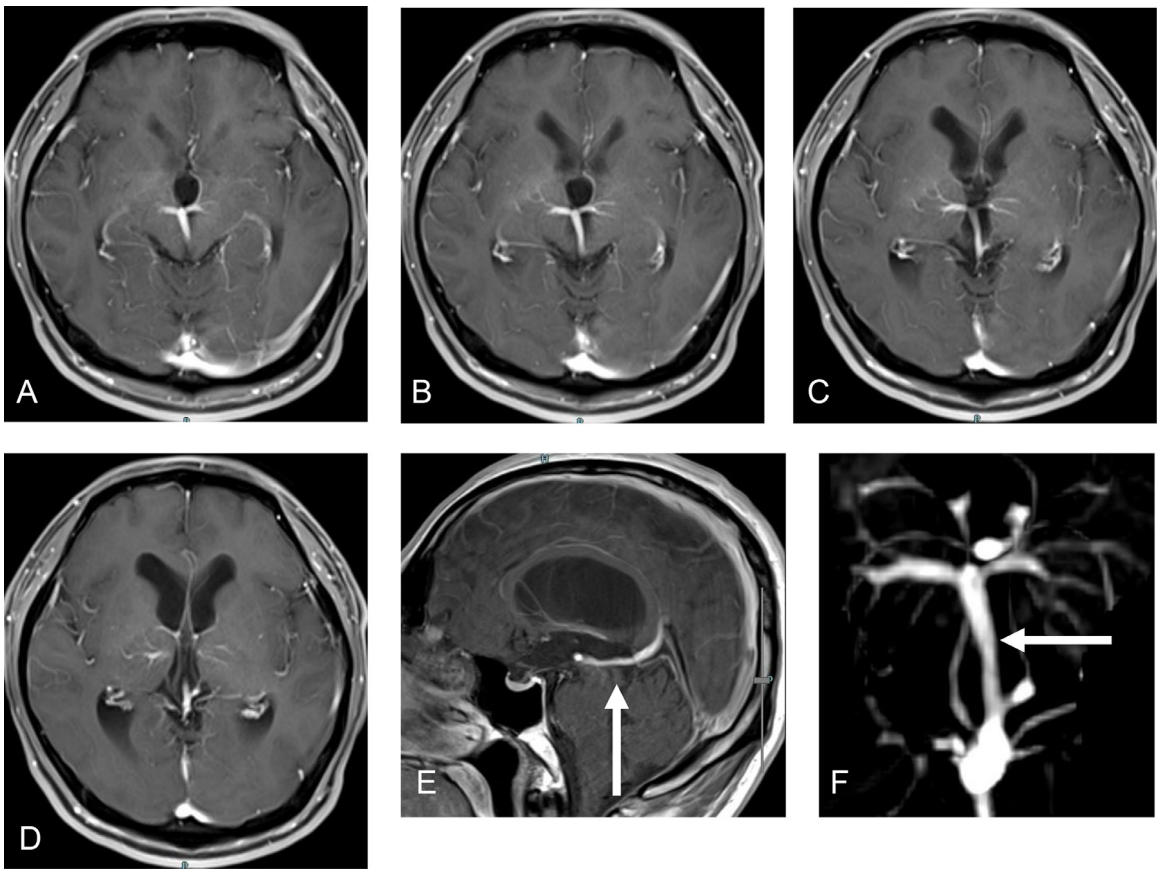

Fig. 3 – Postcontrast magnetic resonance imaging (MRI) (A-E) obtained on the day following the onset shows a DVA (*arrows*), coursing along the floor of the third ventricle before draining into the vein of Galen. This DVA narrows the orifice of the aqueduct on its drainage route and blocks the flow of cerebrospinal fluid. A partial maximum intensity projection image based on subtraction imaging of postcontrast MRI (F) clearly depicts the DVA.

Table 1 – Reported cases of aqueductal stenosis due to DVA (English-language literature).

Authors (Year)	Age/Sex	Complaint	Duration of chief complaint	Location of DVA	Pathway of DVA around the aqueduct	Drainage of DVA	Diagnostic procedure	Treatment	Outcome
Rosenheck et al. (1937) [9]	58/F	Mental deterioration	5 years	Midbrain	NA	NA	Autopsy	No	Dead
Avman et al. (1980) [10]	35/F	Vertex headache, indistinct vision	7 years	Midbrain	Along (oa∼the left lateral recess)	NA	X ray, angiography, direct exploration	Stenting	Improvement
Watanabe et al. (1991) [11]	39/M	Headache	1 year	Midbrain	Across (c2)	→Precentral cerebellar vein→Galen	CT, MRI	Shunting	Improvement in images
Oka et al. (1993) [12]	43/F	Seizure	2 months	Midbrain	Along c1∼pp)	→Precentral cerebellar vein→Galen	CT, MRI, angiography	ETV	Improvement in images
Blackmore et al. (1996) [13]	16/F	Occipital Headache	2 months	Midbrain and thalamus ∼ the floor of the third ventricle	Surrounding (c1)	NA	MRI	No	No treatment
Bannur et al. (2002) [14]	11/M	Occipital headache	5 months	Midbrain ∼ the floor of the third ventricle	Along (oa∼pp)	→Subependymal vein →Galen	CT, MRI	Shunting	Improvement
Yagmurlu et al. (2005) [15]	7/F	Headache	1 month	Thalamus ∼ midbrain	Along (oa∼c1)	→A collector vein of other DVA in the right perimesencephalic sistern→Galen	MRI	No	No treatment
Giannetti et al. (2008) [16]	42/M	Headache, behavior abnormalities	1 year	The floor of the third ventricle ∼ midbrain	Along (proximal part*)	NA	CT, MRI	ETV	Improvement in symptoms
	18/M	Headache, sleepiness, inattention at school and antisocial behavior	6 years	Midbrain ∼ the pineal region	Hairpin curve (oa)	NA	CT, MRI	ETV	Improvement in symptoms
Guhl et al. (2011) [17]	0/M	Delayed psychomotor development	10 months	Midbrain ∼ the orifice of the aqueduct∼ the floor of the third ventricle	Across (oa)	→Perimesencephalic vein→Galen	CUS, MRI	ETV	Improvement in symptoms and images

(*continued on next page*)

Table 1 (*continued*)

Authors (Year)	Age/Sex	Complaint	Duration of chief complaint	Location of DVA	Pathway of DVA around the aqueduct	Drainage of DVA	Diagnostic procedure	Treatment	Outcome
Paulson et al. (2012) [18]	0/F	Asymptomatic macrocephaly	3 days	Thalamus ~ the floor of the third ventricle ~ the orifice of the aqueduct	Across (oa)	NA	CUS, MRI	VP shunt	Improvement in images
Inoue et al. (2013) [19]	10/M	Headache, nausea, vomiting	2 weeks	Cerebellar hemisphere ~ midbrain ~ the orifice of the aqueduct	Along (oa~the fourth ventricle)→Across (oa)	→The vein of the cerebellomesen-cephalic fissure →Galen	CT, MRI	ETV	Improvement in symptoms and images
Kita et al. (2019) [20]	83/M	Hakim's triad	4 months	Midbrain	Across (A)	NA	MRI, endoscopy	ETV	Improvement in symptoms and images
Cavallo et al. (2019) [21]	37/M	Headache, lipothymia	1 year	Pons & cerebellar hemisphere ~ midbrain	Along (oa~pp)	NA	MRI, an-giography	ETV	Improvement in symptoms and images
Higa et al. (2020) [22]	1/M	Head circumference enlargement	11 months	Cerebellar hemisphere ~ midbrain	Along (oa~the fourth ventricle)	→Internal cerebral vein →Galen	MRI	ETV	Improvement in images
Low & Seow (2020) [23]	13/F	Headache	1 year	Midbrain ~ the orifice of the aqueduct	Across (oa)	NA	MRI	ETV	Improvement in symptoms and images
Xian et al. (2020) [24]	47/M	Memory impairment	5 years	Thalamus ~ the floor of the third ventricle ~ **the orifice of the aqueduct**	**Across (oa)**	**→Galen**	MRI	ETV	Improvement in symptoms
Isikbay et al. (2022) [25]	50s*/F	Headache, bilateral leg weakness	1 week	Thalamus ~ the floor of the third ventricle ~ **the orifice of the aqueduct**	**Across (oa)**	**→Galen**	CT, MRI, an-giography	VP shunt	Internal hemorrhage 15 years later
Hiraga et al. (2023) [26]	48/F	Depression	NA	The floor of the third ventricle ~ midbrain	Across (oa)	NA	CT, MRI	ETV	Improvement in symptoms and images
Present case	43/M	Headache	2 days	Thalamus and basal ganglia ~ the floor of the third ventricle ~ **the orifice of the aqueduct**	**Across (oa)**	**→Galen**	MRI	No	No treatment

The pathway of DVA is bolded and underlined in cases similar to the present one.
A, ampulla of the aqueduct; oa, the orifice of the aqueduct; CT, computed tomography; CUS, cranial ultrasound; c1, first constriction of the aqueduct; c2, second construction of the aqueduct; ETV, endoscopic third ventriculostomy; F, female; M, male; MRI, magnetic resonance imaging; NA, not applicable; pp, posterior part of the aqueduct; VP, ventriculoperitoneal.
* Details unknown.

Obstructive hydrocephalus with enlargement of the lateral and third ventricles, as in the present case, is caused primarily by stenosis or obstruction of the aqueduct. In adults, common causes include inflammatory septa and membranes in the aqueduct, neurocysticercosis, tectal glioma, pineal gland tumor, and pineal gland cysts [4]. In children, congenital aqueduct stenosis, neural tube defects such as myelomeningocele and Chiari II malformation, and developmental cysts should be included in the differential diagnosis [5]. In contrast, DVA is a rare cause of aqueductal stenoses.

Reflecting the current hypothesis that DVA is formed primarily as compensatory drainage veins due to congenital hypoplasia or occlusion of the original medullary veins [6], DVA is composed of medullary veins and a large collector vein that assembles these veins radially. This finding is referred to as the caput medusae appearance [1] or umbrella-like appearance [2], which is beneficial to differentiating DVA from other causes of aqueductal stenosis. The performance of SWI and postcontrast MRI would be a practical approach to detecting this appearance because both sequences are suitable for visualizing the venous system due to their high contrast. Furthermore, SWI is useful for detecting hemorrhage, which is a complication of DVA, and cavernous hemangioma, which is a comorbidity of DVA [1]. In our case, these sequences could clearly detect DVA blocking the aqueduct and an old hemorrhage adjacent to the medullary veins. Furthermore, considering both its drainage route and the review of the development of the cerebral vein [7,8], we speculated that the DVA of our patient was probably derived from the bilateral absence of the basal vein of Rosenthal.

Our review of relevant English-language literature identified only 19 cases of obstructive hydrocephalus due to aqueductal stenosis secondary to DVA (Table 1) [9–26]. In our study, the mean age at diagnosis in these cases was 30 years (range, 0-83 years), with 55% of the cases (n = 11) being male and 45% (n = 9) being female. The most common symptom was headaches (n = 12). The duration of the chief complaint ranged from 2 days to 7 years. MRI was performed in all cases after the first clinical MRI was performed in the early 1980s [27]. The collector vein of the DVA finally drained into the Galenic system in all cases where the drainage route of the DVA was described (n = 10). The aqueduct was obstructed by an anomalous vein coursing across the orifice of the aqueduct in 8 cases and coursing along in 8 cases. ETV was performed in 11 of the cases (55%), while shunting was performed in 4 cases, stenting was performed in 1 case and conservative management was performed in 3 cases. The outcome was uneventful, with the exception of 1 case that resulted in symptomatic internal hemorrhage 15 years later. In our case, SWI depicted an old hemorrhage in the right basal ganglia, presumably due to the DVA, but it was fortunately asymptomatic. Similar to the cases reported by Xian et al. [24] and Isikbay et al. [25], the DVA of our patient coursed ventrodorsally across the orifice of the aqueduct before draining into the vein of Galen. The duration of the symptom in our case was extremely short because the previous detection of ventriculomegaly led to a quick diagnosis based on MRI, including SWI and postcontrast T1WI.

Conclusions

We present a rare case of obstructive hydrocephalus caused by aqueductal stenosis due to a DVA. Although DVAs are mostly benign, on rare occasions a DVA can cause obstructive hydrocephalus by narrowing the CSF pathway along its route. We detected the caput medusae appearance using MRI with SWI and postcontrast T1WI.

Patient consent

Informed consent was obtained from the patient for publication of the case report.

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