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Case Report

Obstructive hydrocephalus due to aqueductal stenosis from developmental venous anomaly draining bilateral medial thalami: a case report^{☆,☆☆}

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ARTICLE INFO

Article history:

Received 20 January 2020

Revised 19 February 2020

Accepted 20 February 2020

Keywords:

Aqueductal stenosis

Obstructive hydrocephalus

Developmental venous anomaly

ABSTRACT

Hydrocephalus is a pathological buildup of cerebrospinal fluid within the ventricles leading to ventricular enlargement out of proportion to sulci and subarachnoid spaces. Developmental venous anomaly is a common benign and usually asymptomatic congenital cerebrovascular malformation. Hydrocephalus caused by aqueductal developmental venous anomaly is extremely rare. We describe a case of a 47-year-old man who presents with short-term memory impairment who was found to have a developmental venous anomaly draining bilateral medial thalami through a common collector vein that causes aqueductal stenosis and obstructive hydrocephalus.

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Case report

A 47-year-old African-American man presented with slowly progressive short-term memory impairment for the past 5 years. This manifested primarily by forgetfulness, misplacing items, and repeating self in conversations. Symptoms were exacerbated by stress and fatigue. His medical history was only remarkable for a fall in his early 20s resulting in concussion with brief loss of consciousness and cerebrospinal fluid (CSF) otorrhea, which was treated conservatively. He was re-

portedly diagnosed with hydrocephalus at age of 31 years but he was lost to follow-up.

Neurologic exam was non-focal with intact praxis and executive functions. Montreal Cognitive Assessment score was 18/30, with loss of points for delayed recall (5 points), visuospatial function (2), language (2), abstraction (2), and attention (1). Basic laboratory testing was only significant for borderline elevated hemoglobin A1c (6.0%) and homocysteine (11.9 μmol/L).

Magnetic resonance imaging (MRI) of the brain with and without intravenous gadolinium contrast demonstrated hy-

☆ Declaration of competing interest: None.

☆☆ Support and financial disclosures: None.

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<https://doi.org/10.1016/j.radcr.2020.02.014>

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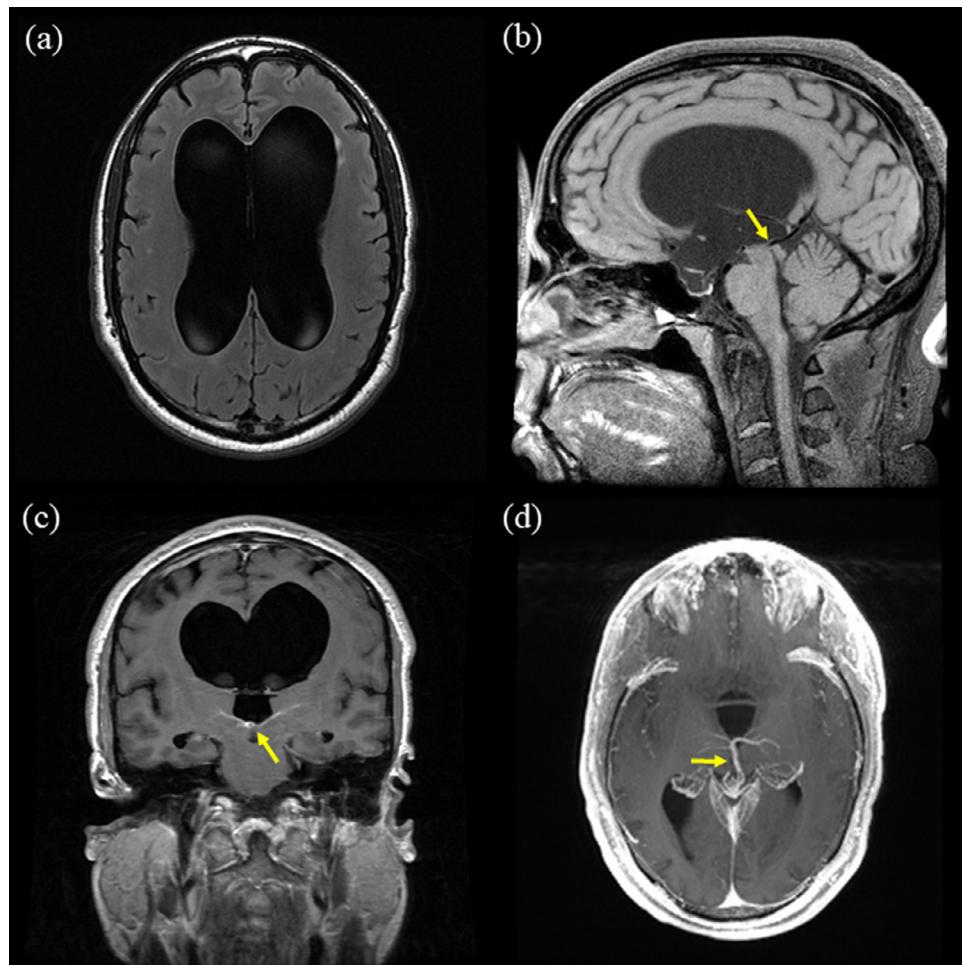


Fig. 1 – MRI brain demonstrates obstructive hydrocephalus secondary to aqueductal stenosis from a developmental venous anomaly draining bilateral medial thalami (arrows). (a) Axial T2-weighted-fluid-attenuated inversion recovery, (b) sagittal T1-weighted image, (c) coronal T1-weighted image postcontrast, and (d) axial T1-weighted image postcontrast maximum intensity projection.

drocephalus with marked dilation of the lateral and third ventricles (Fig. 1). The fourth ventricle was not enlarged, suggesting obstructive hydrocephalus at the level of the cerebral aqueduct. Postcontrast imaging showed a developmental venous anomaly (DVA) draining bilateral medial thalami converging to a prominent anomalous central vein that ran midline along the floor of the third ventricle and ultimately drained posteriorly into the vein of Galen. The central draining vein bridged across the proximal orifice of the cerebral aqueduct, resulting in aqueductal stenosis and severe obstructive hydrocephalus. He was referred to neurosurgery and underwent endoscopic third ventriculostomy (ETV). Although ventriculomegaly was not changed significantly on immediate post-operative imaging, the patient did improve clinically.

Discussion

Hydrocephalus is a hydrodynamic CSF disorder characterized by excessive accumulation of CSF within the ventricu-

lar system of the brain leading to disproportionate ventriculomegaly to any sulcal enlargement that may coexist. Based on its pathophysiology, it is classified as either obstructive (from structural blockage of CSF flow within the ventricular system) or communicating (usually from impaired CSF absorption) [1]. Common causes of obstructive hydrocephalus include aqueductal stenosis from congenital narrowing, septum, web, forking, postinflammatory gliosis, midbrain tumors, and Arnold-Chiari or Dandy-Walker malformations [1–3].

DVA is the most common intracranial vascular malformation, occurring in up to 5% of the general population and they are usually asymptomatic. In very rare circumstances, however, DVA can impede CSF flow through the cerebral aqueduct and become symptomatic [4].

Our review of the literature finds 16 cases of aqueductal stenosis secondary to DVA [4–18]. The mean age of diagnosis in these reported cases was 28 years (range prenatal, defined as 0 days, to 83 years). Fifty-six percent of cases ($n = 9$) were male and 44% ($n = 7$) were female. The most common presenting symptom was headache (63%, $n = 10$). MRI was performed in 88% of cases ($n = 14$), although computed tomography and/or

angiography were also often obtained. ETV was performed in 56% of cases ($n = 9$), while the remaining patients underwent shunting (25%, $n = 4$) or conservative management (19%, $n = 3$). Our patient is older than the typical age of diagnosis, and his presenting symptom is short-term memory dysfunction. Only 2 cases in the literature were diagnosed at a later age (58 and 83 years), and both were described to have cognitive impairment [5,18].

As DVA per se is benign, management of symptomatic DVA is targeted at alleviation of symptoms. Our patient underwent ETV, which is currently considered the treatment of choice for obstructive hydrocephalus [19]. ETV creates an alternative CSF flow pathway that bypasses the obstruction, resulting in improved CSF drainage and ventricle size. All reported patients who have undergone this procedure, including ours, responded well [4,8,11,13–14,16–18].

Despite the frequency with which hydrocephalus and DVA present independently, it is unusual for DVA to cause obstructive hydrocephalus. Clinicians should be aware of this unique combination and its presentation to guide appropriate diagnosis and treatment.

REFERENCES

- [1] Langner S, Fleck S, Baldauf J, Mensel B, Kühn JP, Kirsch M. Diagnosis and differential diagnosis of hydrocephalus in adults. *Rofo* 2017;189(8):728–39.
- [2] Jellinger G. Anatomopathology of non-tumoral aqueductal stenosis. *J Neurosurg Sci* 1986;30(1–2):1–16.
- [3] Partington MD. Congenital hydrocephalus. *Neurosurg Clin N Am* 2001;12(4):737–42 ix.
- [4] Pereira VM, Geibprasert S, Krings T, Auroboonyawat T, Ozanne A, Toulgoat F, Pongpech S, Lasjaunias PL. Pathomechanisms of symptomatic developmental venous anomalies. *Stroke* 2008;39(12):3201–15.
- [5] Rosenheck C. Venous angioma of the sylvian aqueduct and the fourth ventricle associated with internal hydrocephalus and mental deterioration. *Arch Neurol Psychiatry* 1937;38:428–38.
- [6] Avman N, Dinçer C. Venous malformation of the aqueduct of Sylvius treated by interventriculostomy: 15 years follow-up. *Acta Neurochir (Wien)* 1980;52(3–4):219–24.
- [7] Watanabe A, Ishii R, Kamada M, Suzuki Y, Hirano K, Okamura H. Obstructive hydrocephalus caused by an abnormal vein in the aqueduct. Case Report. *J Neurosurg* 1991;75(6):960–2.
- [8] Oka K, Kumate S, Kibe M, Tomonaga M, Maehara F, Higashi Y. Aqueductal stenosis due to mesencephalic venous malformation: case report. *Surg Neurol* 1993;40(3):230–5.
- [9] Blackmore CC, Mamourian AC. Aqueduct compression from venous angioma: MR findings. *AJNR Am J Neuroradiol* 1996;17(3):458–60.
- [10] Bannur U, Korah I, Chandy MJ. Midbrain venous angioma with obstructive hydrocephalus. *Neurol India* 2002;50(2):207–9.
- [11] Sato S, Sonoda Y, Kuroki R, Kayama T. [A rare case of aqueductal stenosis due to venous angioma]. *No To Shinkei* 2004;56(12):1042–6.
- [12] Yagmurlu B, Fitoz S, Atasoy C, Erden I, Deda G, Unal O. An unusual cause of hydrocephalus: aqueductal developmental venous anomaly. *Eur Radiol* 2005;15(6):1159–62.
- [13] Giannetti AV, Rodrigues RB, Trivelato FP. Venous lesions as a cause of Sylvain aqueductal obstruction: case report. *Neurosurgery* 2008;62(5):E1167–8.
- [14] Guhl S, Kirsch M, Lauffer H, Fritsch M, Schroeder HW, et al. Unusual mesencephalic developmental venous anomaly causing obstructive hydrocephalus due to aqueductal stenosis. *J Neurosurg Pediatr* 2011;8(4):407–10.
- [15] Paulson D, Hwang SW, Whitehead WE, Curry DJ, Luerßen TG, Jea A. Aqueductal developmental venous anomaly as an unusual cause of congenital hydrocephalus: a case report and review of the literature. *J Med Case Rep* 2012;6:7.
- [16] Inoue K, Yoshioka F, Nakahara Y, Kawashima M, Matsushima T. Obstructive hydrocephalus following aqueductal stenosis caused by supra- and infratentorial developmental venous anomaly: case report. *Childs Nerv Syst* 2013;29(2):329–34.
- [17] Cavallo C, Faragò G, Broggi M, Ferroli P, Acerbi F. Developmental venous anomaly as a rare cause of obstructive hydrocephalus. *J Neurosurg Sci* 2019;63(5):600–6.
- [18] Kita D, Park C, Hayashi Y. Aqueductal developmental venous anomaly presenting with mimic symptoms of idiopathic normal pressure hydrocephalus in an elderly patient: a case report. *NMC Case Rep J* 2019;6(3):83–6.
- [19] Yadav YR, Parihar V, Pande S, Namdev H, Agarwal M. Endoscopic third ventriculostomy. *J Neurosci Rural Pract* 2012;3(2):163–73.