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Case Reports & Case Series

Developmental venous anomaly presenting as obstructive hydrocephalus

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ABSTRACT

Background: Vascular malformations can present with hydrocephalus following external compression of ventricular outlet. This can be managed by a CSF diversion.

Case presentation: We report a case of hydrocephalus secondary to a congenital venous anomaly treated successfully with cerebrospinal fluid diversion. Here we present a case to literature of a 57 years old male patient who had presented with hydrocephalus secondary to aqueductal obstruction by developmental venous anomaly. Patient underwent right ventriculoperitoneal shunt placement and post operative Computed Tomography showed reduction in size of bilateral lateral ventricles and third ventricle.

Conclusion: Although developmental venous anomaly as a cause for hydrocephalus have been reported, we present one more case of hydrocephalus secondary to a congenital venous anomaly who was treated successfully with cerebrospinal fluid diversion.

1. Background

Aqueduct of Sylvius, a channel connecting third and fourth ventricle situated in the dorsal midbrain, surrounded by periaqueductal gray matter is the narrowest part of CSF pathway and the commonest site of CSF flow obstruction [1]. Commonest etiology for aqueduct cerebrospinal fluid flow obstruction include congenital stenosis and gliosis secondary to inflammatory conditions, tumors [2]. While vascular malformations form an infrequent cause of aqueductal obstruction, developmental venous anomalies are extremely rare with only about 10 cases reported so far [3]. Here we report a case of developmental venous anomaly in a 57 years old male causing obstruction of aqueduct of sylvius thereby causing hyrodephalus.

2. Case representation

A 57-year-old male presented with three months history of giddiness, associated with progressive difficulty in walking, memory disturbances. No complaints of headache, nausea, vomiting, blurring of vision. Patient had two episodes of urinary incontinence. Patient was conscious, attentive, having tremors in both upper limbs and spasticity of both lower limbs.

Magnetic resonance imaging of the brain (Fig. 1a–c) showed dilatation of bilateral lateral ventricles, third ventricle with periventricular seepage and a suspicious isointense focus measuring 2.0 x 1.6 cm posterior to midbrain with post contrast enhancement causing obstruction of aqueduct of Sylvius. The fourth ventricle appeared normal in size.

Cerebral 4 vessel digital subtraction angiogram (Figs. 2, 3) showed developmental venous anomaly in bilateral cerebellum. After obtaining informed consent and anesthetic fitness, patient underwent right ventriculoperitoneal shunt placement under general anesthesia. Post operative Computed Tomography (Fig. 4) was done which showed reduction in size of bilateral lateral ventricles and third ventricle. Clinically, patient and attender reported improvement in gait, absent involuntary movements of upper limbs. Spasticity was reduced as well. Patient is currently still under follow up for over 1 year.

3. Conclusion

Aqueductal stenosis from vascular lesions is a rare entity. Vascular anomalies including developmental venous anomaly, vein of Galen aneurysm, ecstatic arteries, dilated abonromal veins and arteriovenous malformations can cause mechanical compression and thus obstructive hydrocephalus requiring CSF diversion procedures [4]. Developmental Venous Anomalies (DVA) earlier known as venous malformation, medullary venous malformation or venous angioma, and medullary venous malformation is one of the commonest vascular malformations within the brain [5]. They contain dilated centripetally draining dilated medullary veins that drain into a collecting transcerebral vein which further drains into either superficial subcortical veins or subependymal veins, thereby forming the a caput medusa [6]. They are considered to result from medullary vein developmental arrest between the Padget's

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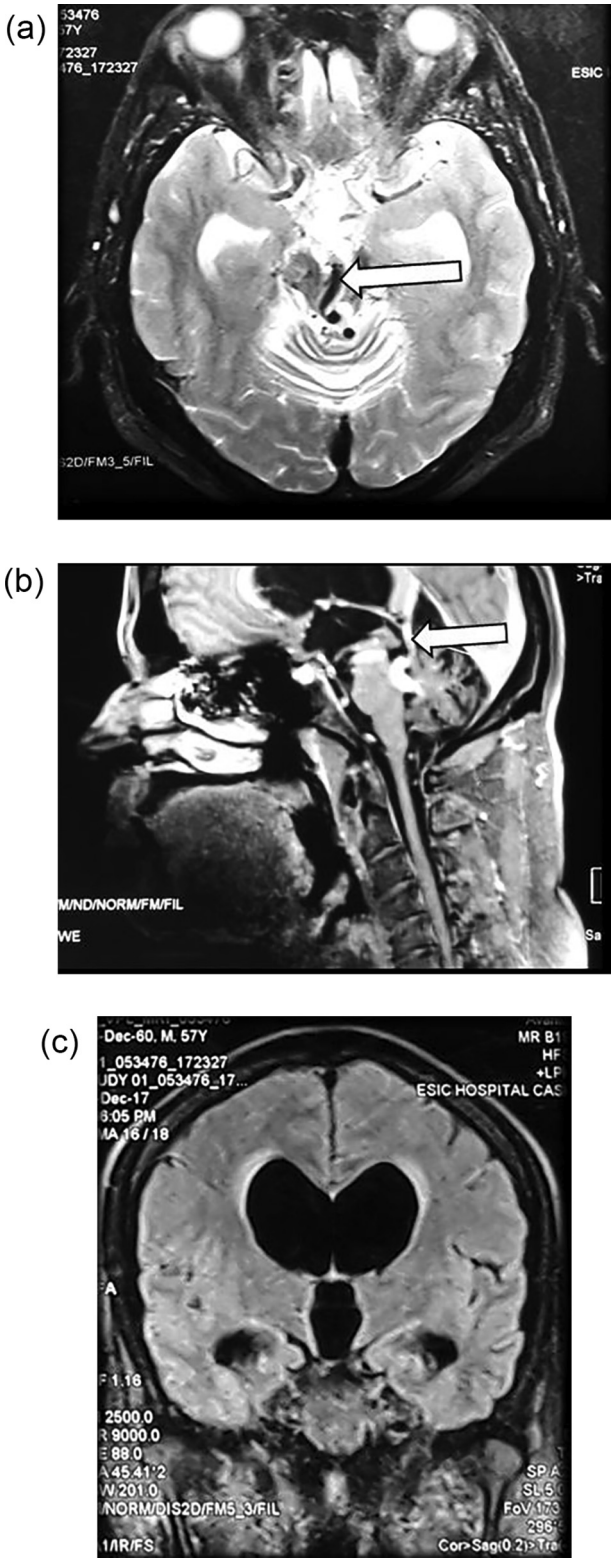

Fig. 1. a–c. Magnetic resonance imaging- Dilatation of bilateral lateral ventricles, third ventricle with periventricular seepage and a suspicious isointense focus.

fourth and seventh stages [2]. The incidence according to autopsy studies of Developmental venous anomalies (DVAs) is about 2.6%. Although most of these lesions are benign; these lesions may however sometimes clinically present with either mechanical or flow related complications [3]. In very few incidences these anomalies have been

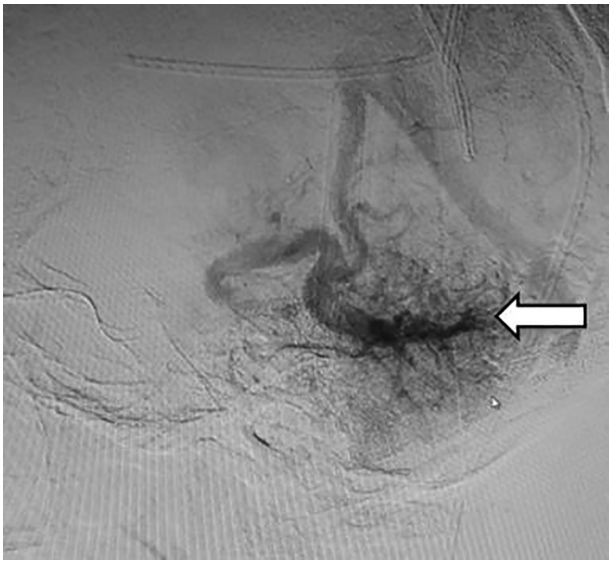

Fig. 2. Cerebral 4 vessel digital subtraction angiogram showed developmental venous anomaly in bilateral cerebellum.

reported to cause ventricular outflow obstruction causing hydrocephalus [7]. Vein of Galen malformations, one of the differential diagnosis is actually a misnomer. During development, the median prosencephalic vein fails to regress and this becomes aneurysmal. Hydrocephalus is associated with venous hypertension or mechanical obstruction [4].

According to Rammos et al, surgical management is to be directed towards the complications of DVA's mentioned above rather than the DVA lesion itself. Intracranial hemorrhage may require decompression and obstructive hydrocephalus will require CSF diversion [5,8]. Since, they are mostly benign lesions and attempts to removing them can cause venous infarction or edema of normal brain tissue, surgery is generally not advised for excision or occlusion [8]

So far very fewer cases have been reported with obstructive hydrocephalus secondary to ventricular outflow obstruction by DVA with patient's ages ranging from newborn to 58yrs [8]. Hence we have reported our case of developmental venous anomaly in a 57 years old male causing compression of the aqueduct of sylvius thereby causing hydrocephalus. Patient had presented with features suggestive of obstructive hydrocephalus, evaluated with MRI brain which had showed a DVA obstructing the aqueduct of sylvius. As per literature, patient underwent CSF diversion with improvement in clinical condition.

Several cases of developmental venous anomaly as a cause for hydrocephalus have been reported. With this report, we add one more case to the literature of a patient with hydrocephalus secondary to a congenital venous anomaly who was treated successfully with cerebrospinal fluid diversion.

4. Ethics approval and consent to participate section

The study was approved and designed by institutional Ethical Committee, Sri ramachandra institute of higher education and research, and informed consent was obtained from the participant.

5. Consent for publication

The ethical review board allowed the publication of case and it was with patients consent.

Availability of data and material

All case related data are provided in this article and not available elsewhere.

Contribution Details *Dr. Bhaskar Naidu, Dr. Sudha Ram*

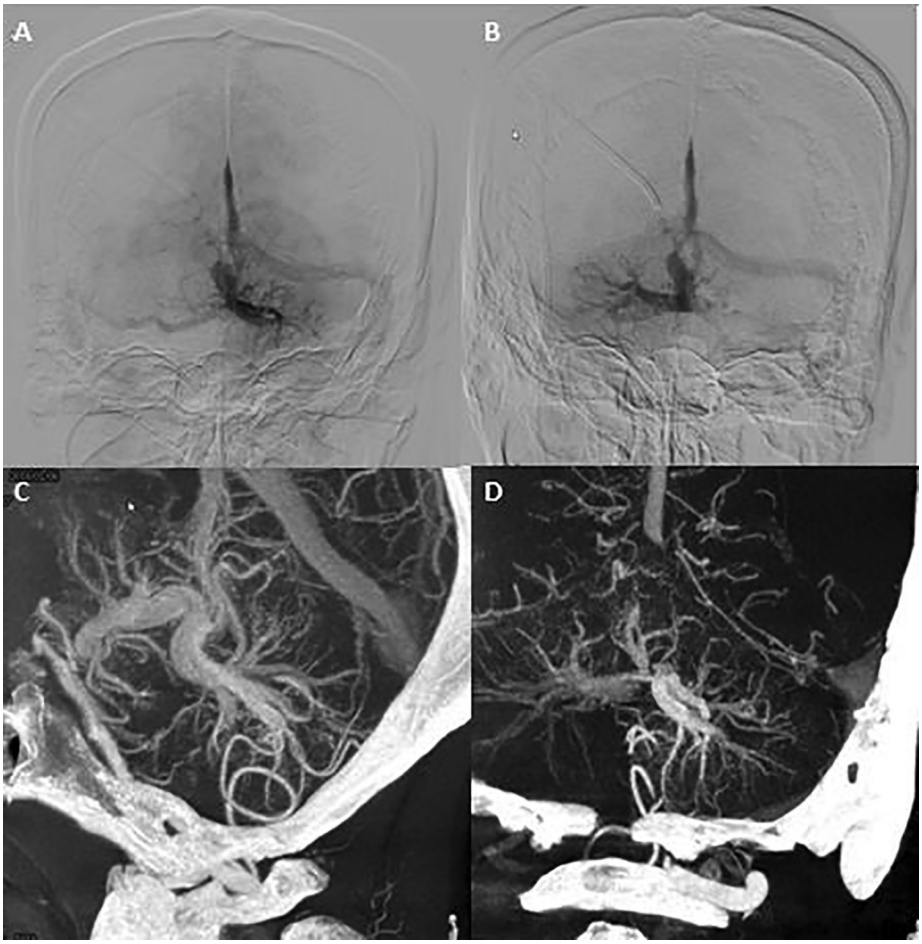

Fig. 3. Reduction in size of bilateral lateral ventricles and third ventricle.

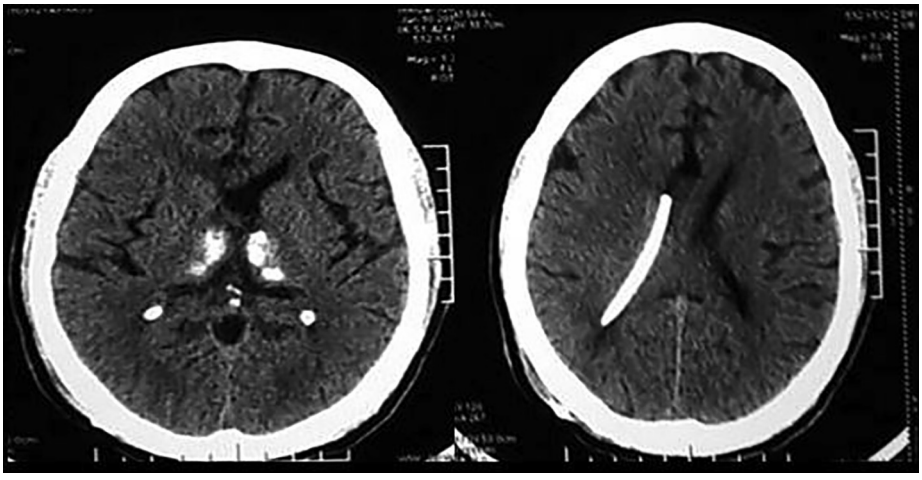

Fig. 4. a–d. Cerebral 4 vessel digital subtraction angiogram showing DVA.

	Contributor 1	Contributor 2
Concepts		Yes
Design		Yes
Definition of intellectual content		Yes
Literature search	Yes	
Clinical studies	Yes	
Experimental studies	Yes	
Data acquisition	Yes	
Data analysis	Yes	
Statistical analysis	Yes	
Manuscript preparation	Yes	
Manuscript editing		Yes
Manuscript review		Yes
Guarantor	Yes	Yes

Declaration of Competing Interest

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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References

[1] David Paulson, Steven W. Hwang, Daniel J. Curry, Thomas G. Luerssen, Andrew Jea, Aqueductal Developmental Venous Anomaly as an Unusual Cause of Congenital Hydrocephalus: Case Report and Review of the Literature, J. Med. Case Rep. 6 (2012) 7.

[2] Keng-Liang Kuo, Feng-Ji Tsai, Yao-Ju Liu, Yu-Kai Huang, Ann-Shung Lieu, Unusual Location of Developmental Venous Anomaly within Fourth Ventricle Causing Obstructive Hydrocephalus – A Case Report, Neurol. Neurochir. Pol. 52 (2018) 112–115.

[3] G.M. Santucci, J.L. Leach, J. Ying, S.D. Leach, T.A. Tomsick, Brain Parenchymal Signal Abnormalities Associated with Developmental Venous Anomalies: Detailed MR Imaging Assessment, Am. J. Neuroradiol. 7 (2008) 1317–1323.

[4] V.M. Pereira, S. Geibprasert, T. Krings, T. Aurboonyawat, A. Ozanne, F. Toulgoat, S. Pongpech, P.L. Lasjaunias, Pathomechanisms of symptomatic developmental venous anomalies, Stroke 39 (2008) 3201–3215.

[5] S.K. Rammos, R. Maina, G. Lanzino, Developmental venous anomalies: current concepts and implications for management, Neurosurgery 65 (2009) 20–29.

[6] C.C. Blackmore, A.C. Mamourian, Aqueduct compression from venous angioma: MR findings, Am. J. Neuroradiol. 17 (1996) 458–460.

[7] M. Sarwar, W.F. McCormick, Intracerebral venous angioma: case report and review, Arch. Neurolol. 35 (1978) 323–325.

[8] U. Bannur, I. Korah, M.J. Chandy, Midbrain venous angioma with obstructive hydrocephalus, Neurol. India 50 (2002) 207–209.