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Dural arteriovenous fistula of the torcular herophili presenting with hydrocephalus and venous congestion in an 8-month-old child: A case report

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Abstract:

Dural arteriovenous fistulas (DAVFs) are direct communication between the dural arterial and venous systems. They are more common in adults. In children, they are relatively rare. Hydrocephalus is a common problem in pediatrics with a variety of causes. However, very few cases of hydrocephalus as a complication of DAVF have been reported in the literature. This case describes an 8-month-old male child with a large DAVF at the torcular herophili who presented with regression of milestones and hydrocephalus. Magnetic resonance imaging (MRI) on admission showed triventricular hydrocephalus and a massively dilated torcular with a compressed fourth ventricle. Angiography confirmed the presence of a DAVF at the torcula with arterial feeders from the posterior circulation. Endovascular embolization was performed with >80% embolization of the fistula with no complications. Control MRI immediately postoperative was acceptable. No cerebrospinal fluid (CSF) diversion was performed. At a 3-month follow-up, the child had attained all developmental milestones for age. MRI showed normal CSF dynamics and a further reduction in the size of the torcula. Despite being rare, DAVFs should be considered as a possible cause of pediatric hydrocephalus, and treating them can lead to a resolution of the mechanisms inducing hydrocephalus. CSF shunting should be reserved for those cases with persistent hydrocephalus and raised intracranial pressure despite endovascular treatment.

Keywords:

Cerebral vascular malformations, confluence of sinuses, dura arteriovenous fistula, dural arteriovenous fistulas, embolization, hydrocephalus

Introduction

Dural arteriovenous fistulas (DAVFs) are direct communication between the dural arterial and venous systems, the meningeal veins, or the cortical veins. DAVFs are more frequent in adults and can occur in any part of the dura mater. They occur more frequently in the transverse, sigmoid, and cavernous sinuses. Although the etiology in some cases is idiopathic, its appearance is related to trauma, surgery, venous stenosis, or thrombosis of the dural sinuses. They represent 10%–15% of intracranial vascular malformations.[1-3] The appearance of DAVF in childhood is rare. There are several classification systems for DAVF, the most used are the Borden classification and the Cognard classification, both based on the flow pattern in the sinus.[3] Depending on the location of the fistula and the interruption of normal venous drainage, there will be changes in the blood and cerebrospinal fluid (CSF) flow dynamics, giving rise to the symptoms

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presented by the patient. Among the symptoms that may occur include pulsatile tinnitus, murmurs, macrocrania, headache, vision changes, alterations in mental status, bleeding or ischemic disorders, cranial nerve injury, and motor or sensory defects. However, hydrocephalus is not common.[1,2,4] Endovascular treatment is currently the most accepted option, although it can be complemented with surgery or radiosurgery in some cases. In all cases, the objective is to alleviate the symptoms.[4-6] This report describes a rare case of DAVF at the confluence of the sinuses associated with hydrocephalus due to compression of the fourth ventricle by the gigantic fistula, as well as alterations in CSF absorption in the venous sinus, present in an 8-month-old patient.

Case Report

An 8-month-old boy was admitted to the pediatric neurosurgery department due to abnormal and disproportional head enlargement and an inability to support his head and sit. According to his mother, they noticed the problem from birth but thought it would eventually normalize. When there was no improvement, she consulted a neurologist. The occipital frontal circumference (OFC) of the head was 47 cm (>97th percentile) with the chest measuring 44 cm (slightly below the average for the age). On neurosonography (NSG), hydrocephalus was suspected, and the patient was referred to the pediatric neurosurgeons the next day.

On admission, he was conscious, with increased motor activity, and a large, level, mildly tense fontanel measuring 2 cm × 1.5 cm, with diastasis of the metopic sutures. Head circumference was 47.4 cm, with the rigidity of occipital muscles and bilateral Babinski reflex. NSG on admission showed signs of ventriculomegaly.

The electrocardiogram showed sinus arrhythmia, and episodic bradycardia, with features of atrial overload and increased electrical activity in the myocardium of the left ventricle.

A brain magnetic resonance imaging (MRI) done on day 3 of admission showed a Type 1 Chiari anomaly with minimal compression at the medulla oblongata, a compressed fourth ventricle, and triventricular ventriculomegaly. The superior sagittal sinus was massively dilated. There was a large cystic flow void involving the whole confluence of sinuses and proximal parts of both transverse sinuses measuring 47 mm × 30 mm × 51 mm [Figure 1].

Computer tomography (CT) angiography performed on day 6 showed a pronounced expansion of the sagittal sinus lumen and the confluence of sinuses (60 mm × 30 mm × 45 mm) and transverse sinuses dilatation up to 26 mm [Figure 1].

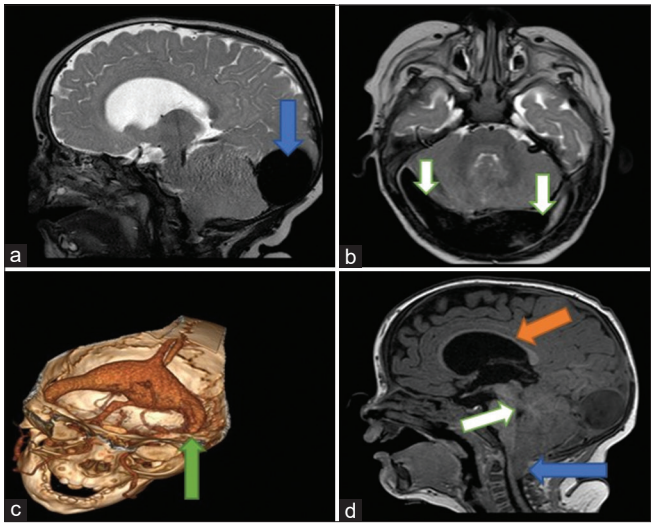

Figure 1: The preoperative imaging. (a and b) Sagittal and axial MRI. A massively dilated torcular herophili (blue arrow) and transverse sinuses are visualized (white arrows). (c) A 3D reconstruction of the CTA with arterial feeders from the posterior circulation (green arrow). (d) A sagittal T1 MRI showing the thinned corpus callosum (orange arrow), the compressed aqueduct of Sylvius and fourth ventricle (white arrow) and herniation of the tonsils through the foramen magnum (blue arrow). MRI: Magnetic resonance imaging, 3D: Three-dimensional, CTA: Computer tomography angiography,

On day 10, microcatheter-aided super-selective endovascular embolization of the arteriovenous fistula afferents using n-Butyl cyanoacrylate (an adhesive embolizing substance) was performed through the left vertebral artery and left external carotid artery in a standard fashion. Preembolization angiogram showed the multiple feeders of the DAVF and the dilated torcular [Figure 2].

Control angiography at the end of the procedure showed near-total (>80%) arteriovenous fistula embolization [Figure 3].

A series of MRI controls performed 2 days to 3 months after embolization showed significant regression in the size of the DAVF and torcular herophili. The CSF dynamics were greatly improved with a significant reduction in the size of the lateral and third ventricles and re-expansion of the fourth ventricle [Figure 4]. On examination at 3 months after discharge, head circumference was 49 cm (>97th percentile). No focal neurologic signs or signs of increased intracranial pressure (ICP) were observed. The child had attained all developmental milestones for age.

Discussion

There are few reported cases of DAVF associated with hydrocephalus in the literature. Our literature review revealed nine patients with DAVF and hydrocephalus. It should be noted that in most cases, treatment of the DAVF led to the resolution of hydrocephalus, and shunting was

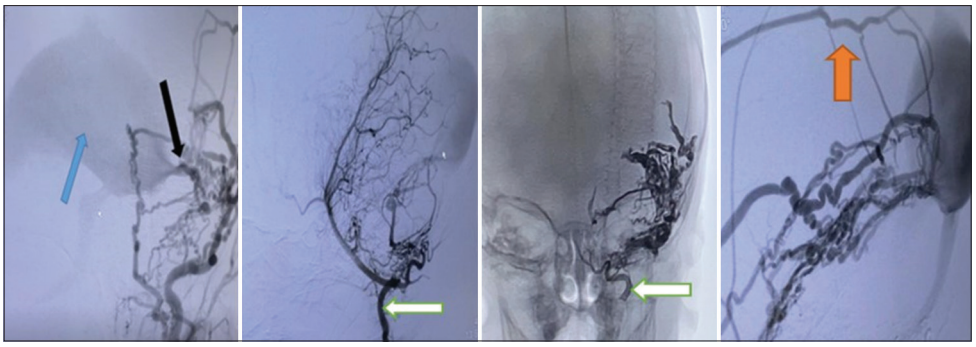

Figure 2: Angiography performed during embolization showing the fistula with multiple feeding arteries (black arrow) and the venous drainage into the torcular (blue arrow). It is noted that the major afferents of the DAVF came from the vertebral system (white arrows) with other minor feeders from the carotid system through the choroidal arteries (orange arrow) and the meningeal branches of the external carotid artery. DAVF: Dural arteriovenous fistulas

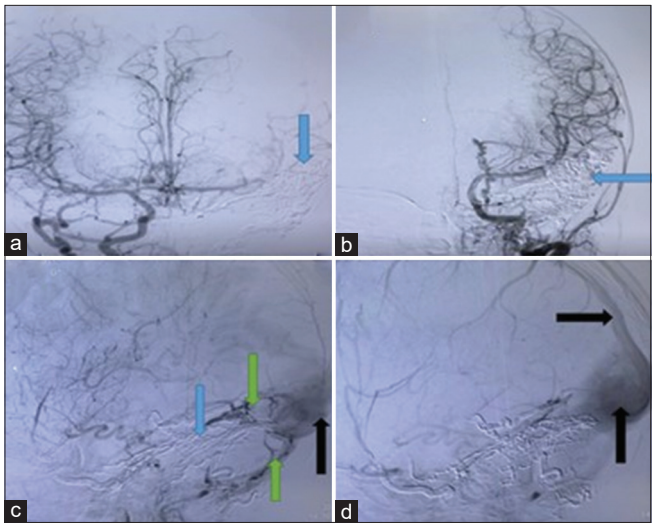

Figure 3: (a and b) Following microcatheters superselective catheterization and embolization with n-Butyl cyanoacrylate (blue arrows). (c and d) The embolization was >80% with partial filling of the DAVF and sinuses (black arrows) from the branches of the choroidal arteries and small meningeal arteries (green arrows) seen on the postembolization angiogram. DAVF: Dural arteriovenous fistulas

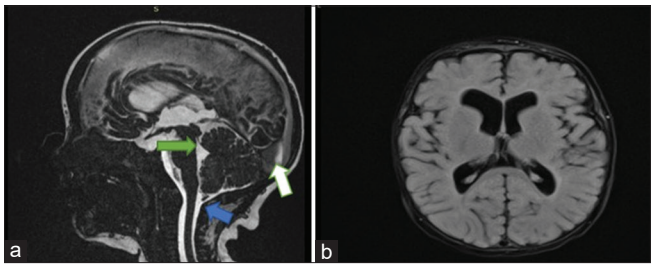

Figure 4: (a) Postoperative image at 3 months compared to the preoperative imaging, the tonsilar herniation has almost completely reversed on the sagittal view (blue arrow). The torcula diameter had significantly reduced (white arrow) and the fourth ventricle appears normal with no signs of compression (green arrow). (b) No features of increased ICP were seen on the axial scan. ICP: Intracranial pressure

rarely required (only two patients) [Table 1]. Depending on the location of the DAVF the patients may present with different clinical features. Some of the described causes of hydrocephalus in DAVF include obstruction of the third ventricle and quadrigeminal cisterns by multiple tortuous vascular malformations, engorgement of the vein of Galen leading to obstruction of the cerebral aqueduct, engorgement of the straight sinus resulting in compression at the junction between the cerebral aqueduct and the fourth ventricle or associated hemorrhage in the adjacent ventricles.[2]

It is well established in children that vascular dysfunction in the form of increased venous pressure can cause communicating hydrocephalus.[1] This can be seen in cases of venous sinus thrombosis, outlet obstruction in craniosynostosis, etc.[7] When the pressure within the superior sagittal sinus approximates or exceeds the pressure in the subarachnoid space, there is a resultant failure of CSF resorption, thus producing an imbalance in brain water or volume and an increase in ventricular size.[1]

Our patient presented with factors favoring both etiological mechanisms of hydrocephalus. The patient had triventricular enlargement due to the direct mass effect of the DAVF on the aqueduct of Sylvius, causing an obstruction. Furthermore, evident in this patient were the massively dilated venous sinuses and torcular indicating an increased venous pressure. We concluded that our patient had mixed communicating and noncommunicating hydrocephalus.

DAVF is not usually suspected clinically and is only revealed through imaging done for the evaluation of cranial symptoms.[8] Usually, the CT or MRI is useful to exclude other possible intracranial pathologies. The gold standard investigation for prognostication and management planning is digital subtraction angiography.[9] It outlines the vascular anatomy, i.e., main arterial feeders and venous drainage of the DAVF, and allows for embolism where possible.[8,10]

If left untreated, these vascular abnormalities have an unfavorable natural history. The mainstay of management is endovascular treatment although open surgical treatment can be performed in highly selected cases.[8,10] Whether to use the transarterial or transvenous

Table 1: Summarizes the cases reviewed in the literature review

Authors and year	Sex/age	Fistula location	Hydrocephalus	Treatment	Outcome
Sirakov *et al*. 2022	Female/9 years	Transverse sinus and the confluence of sinuses	Obstructive	Staged embolization in multiple sessions, microsurgical extirpation, shunt implantation	Hydrocephalus resolved spontaneously
Agrawal *et al*. 2021	Female/15 years	Occipital sinus DAVF	Obstructive	Embolization with Onyx	Hydrocephalus resolved spontaneously
Zhang *et al*. 2019	Female/55 years	Vein of Galen DAVF	Obstructive	Onyx transarterial endovascular embolization	Hydrocephalus resolved spontaneously
Morales *et al*. 2017	Male/4 years	Pial AVF	Communicating	Embolization of the venous pouch with coils and glue (n-BCA)	Hydrocephalus resolved spontaneously
Walcott *et al*. 2013	Male/69 years	Vertebro-vertebral fistula	Communicating	Embolization with combined Onyx and coils followed by VPS	Hydrocephalus resolved with VPS
Nakahara *et al*. 2011	Female/76 years	DAVF in the transverse-sigmoid sinuses	Normal-pressure	Transvenous embolization	Hydrocephalus resolved spontaneously
Johnson *et al*. 2009	Male/8 months	The confluence of sinuses and the transverse sinuses	Communicating	Surgical clipping of the middle meningeal artery near the fistula at birth, embolization at 8 months	Resolution of hydrocephalus, global developmental delay
Ernst *et al*. 2006	Female/77 years	Transverse sinus DAVF	Obstructive	Onyx transarterial embolization	Hydrocephalus resolved spontaneously
Monges *et al*. 2005	Female/50 days	Torcular herophili DAVF	Communicating	Posterior fossa durotomy VPS	A complete recovery Hydrocephalus resolved with VPS

VP: Ventriculoperitoneal, AVF: Arteriovenous fistulas, DAVF: Dural AVF, n-BCA: N-Butyl-2-cyanoacrylate, VPS: VP shunt

approaches depends on the angioarchitecture of the DAVF, the location, and the direction of venous flow.[5] Our patient underwent endovascular management of the fistula with no complications. The hydrocephalus resolved over time, and the patients had no features of increased ICP despite OFC remaining in the >97th percentile range. This emphasizes that these patients rarely require shunting procedures to manage the hydrocephalus as it resolves with normalization of venous pressure and relief of compression.[2] Some authors have performed CSF diversion procedures in cases of severely increased ICP or persisting hydrocephalus after embolization.[6,11]

Recanalization in embolized DAVF is uncommon and not often described in the literature. Adamczyk, *et al*. in 2012 described an 18% risk of recanalization with Onyx,[12] significantly higher than the 1.3% recanalization rate with N-butyl-2-cyanoacrylate described by Guillevin, *et al*.[13] N-butyl-2-cyanoacrylate has been shown to have longer vascular occlusion and slow resorption properties. However, vessel remodeling can take place, leading to glue migration in the blood vessels and recanalization.[14]

Apart from endovascular embolization of DAVF, open surgery and stereotactic radiosurgery (SRS) remain useful alternatives.[5,9] Open surgery requires skeletonization and coagulation of the arterial feeders and arterialized cortical veins. In cases with nonfunctional DAVFs, they can be safely resected.[5,15] One of the most concerning complications is hemorrhage which can be reduced by preoperative selective embolization.[16] The main indication for open surgical management is failed or unfeasible endovascular embolization.[17] Anterior cranial fossa location is a relative indication for surgery when embolization is at high risk for ophthalmic artery involvement.[5,17,18]

Open surgery is associated with 13% long-term morbidity and mortality. Complications include postoperative infection, CSF leaks, stroke, cranial nerve palsy, etc.[16,17]

SRS, on the other hand, is the method of last resort used when both endovascular and surgical methods both failed or are too dangerous.[19,20] Complete obliteration (seen in 50%–93%) may take months, and during this period, the patient remains at risk of hemorrhage.[5,21] However, in low-grade DAVF, i.e., Cognard or Borden Grade 1, it has shown good outcomes with minimal hemorrhage risk.[16] The associated complications include cranial nerve palsies, brain edema, and radiation necrosis.[19,22]

Conclusion

Hydrocephalus is a common problem in pediatrics with a variety of causes. Despite being rare, DAVF should be suspected as a possible cause of pediatric hydrocephalus. In these patients, treating DAVF can lead to a resolution of the mechanisms inducing hydrocephalus and CSF shunting should be reserved for those cases with persistent hydrocephalus and raised ICP despite endovascular treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for the child's images and other clinical information to be reported in the journal. The guardian understands that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Wen HY, Yang ST, Lo YL. Hydrocephalus with ventricular papillary structure induced by aggressive intracranial Dural arteriovenous fistulas. World Neurosurg 2019;132:208-10.
2. Zhang CW, Fu W, Richard SA, Xie XD. Obstructive hydrocephalus associated with spontaneous Dural arteriovenous fistula: A case report. Medicine (Baltimore) 2019;98:e15026.
3. Gandhi D, Chen J, Pearl M, Huang J, Gemmete JJ, Kathuria S. Intracranial Dural arteriovenous fistulas: Classification, imaging findings, and treatment. AJNR Am J Neuroradiol 2012;33:1007-13.
4. Ernst GL, Carlson AP. Transverse sinus Dural arteriovenous fistula presenting with acute hydrocephalus from cerebellar venous engorgement resolving with endovascular treatment. J Neurol Surg A Cent Eur Neurosurg 2016;77:538-42.
5. Baharvahdat H, Ooi YC, Kim WJ, Mowla A, Coon AL, Colby GP. Updates in the management of cranial Dural arteriovenous fistula. Stroke Vasc Neurol 2020;5:50-8.
6. Walcott BP, Berkhemer OA, Leslie-Mazwi TM, Chandra RV, Ogilvy CS, Yoo AJ. Multimodal endovascular treatment of a vertebrovertebral fistula presenting with subarachnoid hemorrhage and hydrocephalus. J Clin Neurosci 2013;20:1295-8.
7. Morales-Gómez JA, Garza-Oyervides VV, Arenas-Ruiz JA, Mercado-Flores M, Elizondo-Riojas CG, Boop FA, *et al.* Hydrocephalus in a patient with an unruptured pial arteriovenous fistula: Hydrodynamic considerations, endovascular treatment, and clinical course. J Neurosurg Pediatr 2017;19:307-11.
8. Yadav V, Bhatt S, Athaullah, Dangwal S. Rare occurrence of Dural arteriovenous fistula in a child: Multi-modality imaging and literature review. Radiol Case Rep 2021;16:879-83.
9. Kuwayama N. Management of Dural arteriovenous fistulas. In: Kato Y, Ansari A, editors. Cerebrovascular Surgery: Controversies, Standards and Advances. Cham: Springer International Publishing; 2022. p. 251-64.
10. Yu J, Lv X, Li Y, Wu Z. Therapeutic progress in pediatric intracranial Dural arteriovenous shunts: A review. Interv Neuroradiol 2016;22:548-56.
11. Monges JA, Galarza M, Sosa FP, Ceciliano A. Direct surgical approach of a congenital Dural arteriovenous fistula at the torcular herophili in a neonate: Case illustration. J Neurosurg 2005;102:440.
12. Adamczyk P, Amar AP, Mack WJ, Larsen DW. Recurrence of "cured" Dural arteriovenous fistulas after onyx embolization. Neurosurg Focus 2012;32:E12.
13. Guillevin R, Vallee JN, Cormier E, Lo D, Dormont D, Chiras J. N-butyl 2-cyanoacrylate embolization of spinal Dural arteriovenous fistulae: CT evaluation, technical features, and outcome prognosis in 26 cases. AJNR Am J Neuroradiol 2005;26:929-35.
14. Comby PO, Guillen K, Chevallier O, Lenfant M, Pellegrinelli J, Falvo N, *et al.* Endovascular use of cyanoacrylate-lipiodol mixture for peripheral embolization: Properties, techniques, pitfalls, and applications. J Clin Med 2021;10:4320.
15. Collice M, D'Aliberti G, Arena O, Solaini C, Fontana RA, Talamonti G. Surgical treatment of intracranial Dural arteriovenous fistulae: Role of venous drainage. Neurosurgery 2000;47:56-66.
16. Kakarla UK, Deshmukh VR, Zabramski JM, Albuquerque FC, McDougall CG, Spetzler RF. Surgical treatment of high-risk intracranial Dural arteriovenous fistulae: Clinical outcomes and avoidance of complications. Neurosurgery 2007;61:447-57.
17. Oh SH, Choi JH, Kim BS, Lee KS, Shin YS. Treatment outcomes according to various treatment modalities for intracranial Dural arteriovenous fistulas in the onyx era: A 10-year single-center experience. World Neurosurg 2019;126:e825-34.
18. Giannopoulos S, Texakalidis P, Mohammad Alkhataybeh RA, Charisis N, Rangel-Castilla L, Jabbour P, *et al.* Treatment of ethmoidal Dural arteriovenous fistulas: A Meta-analysis comparing endovascular versus surgical treatment. World Neurosurg 2019;128:593-9.e1.
19. Cifarelli CP, Kaptain G, Yen CP, Schlesinger D, Sheehan JP. Gamma knife radiosurgery for Dural arteriovenous fistulas. Neurosurgery 2010;67:1230-5.
20. See AP, Raza S, Tamargo RJ, Lim M. Stereotactic radiosurgery of cranial arteriovenous malformations and Dural arteriovenous fistulas. Neurosurg Clin N Am 2012;23:133-46.
21. Koebbe CJ, Singhal D, Sheehan J, Flickinger JC, Horowitz M, Kondziolka D, *et al.* Radiosurgery for Dural arteriovenous fistulas. Surg Neurol 2005;64:392-8.
22. Söderman M, Edner G, Ericson K, Karlsson B, Rähn T, Ulfarsson E, *et al.* Gamma knife surgery for Dural arteriovenous shunts: 25 years of experience. J Neurosurg 2006;104:867-75.