CASE REPORT

Isolated intraventricular hemorrhage secondary to dural arteriovenous fistula

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SUMMARY

Isolated intraventricular hemorrhage due to dural arteriovenous fistula (dAVF) is extremely rare and has been reported only a few times in the literature. The pathophysiological cause of isolated intraventricular hemorrhage in these cases was hypothetically attributed to retrograde venous flow into subependymal vessels, although none of these cases demonstrated radiographic evidence of such reflux. We present the first case of a dAVF with radiographic evidence of severe cortical venous reflux into the subependymal venous network causing isolated intraventricular hemorrhage, thus lending proof for the underlying pathophysiology. Furthermore, ours is the first case of dAVF with isolated intraventricular hemorrhage that was successfully treated using multimodality transvenous and transarterial embolization techniques. In cases of high-grade dAVF, multimodality treatment may offer the greatest chance for success, and should be strongly considered for management.

BACKGROUND

Dural arteriovenous fistulas (dAVFs) may have a wide range of clinical manifestations including benign pulsatile tinnitus, transient ischemic attacks, seizures, cranial nerve palsies, and intracranial hemorrhage.¹ Intracranial hemorrhage associated with dAVFs is usually described in varying combinations of subarachnoid, parenchymal, and intraventricular components.² Intraventricular hemorrhage (IVH) usually results from extension of an adjacent intraparenchymal hematoma.¹ Isolated IVH is exceedingly rare in cases of dAVF, and has been reported in only a few instances.^{3–8} The pathophysiological cause of isolated IVH in these cases was hypothetically attributed to venous reflux into subependymal vessels, although none of these cases demonstrated radiographic evidence of such reflux.

We present the first case of a dAVF causing isolated IVH with radiographic evidence of retrograde dilatation of the subependymal venous network, thus lending pathophysiological proof for the underlying etiology. Furthermore, ours is the first case of dAVF with isolated IVH that was successfully treated using both transvenous and transarterial embolization techniques. In addition, we discuss the previous literature and treatment options for such high-grade lesions.

CASE PRESENTATION

An 84-year-old woman with hypertension, diabetes, and a modified Rankin Scale score of 0 presented

to the emergency room with 4 days of headache, nausea, vomiting, and worsening confusion with episodes of excessive drowsiness. CT scan of the head showed IVH in both lateral ventricles, without associated hydrocephalus (figure 1A). CT angiogram of the brain showed multiple dilated meningeal and pial vessels overlying and within the right parietal and temporal lobes (figure 1B). The differential diagnosis after non-invasive imaging included dAVF with venous engorgement versus an arteriovenous malformation (AVM) with fistulous component. Given the patient's favorable baseline condition, a diagnostic cerebral angiogram was necessary to further investigate the etiology of the IVH and numerous dilated vessels. Furthermore, angiography was necessary to guide management considering the disparity of treatment options between dAVFs and AVMs. Cerebral angiography showed a right transverse-sigmoid sinus Cognard type IIa+b dAVF with arterial supply from the right middle meningeal artery, the right occipital artery, and the right posterior auricular artery (figure 1C,D). (figure 1B,C). Venous outflow was through reflux of the temporo-occipital cortical veins draining into deep medullary veins, subependymal veins and into the internal cerebral veins and vein of Galen, as well as through an ectatic venous pouch of the right transverse/sigmoid sinus with a more distal high-grade stenosis (figure 1E,F). (figure 1D). The patient's somnolence and confusion worsened. Serial CT scans of the head showed unchanged ventricular size, continuous electroencephalography was negative for seizure activity, and the patient did not show improvement after lumbar puncture with removal of a large volume of cerebrospinal fluid. Her deterioration was thus attributed to worsening venous hypertension. On hospital day 5 she underwent transvenous coil embolization of the venous pouch. Transfemoral catheterization of the ectatic venous pouch was achieved with a Marksman 3.2 Fr microcatheter (Covidien, Irvine, California, USA) through a 6 Fr Envoy-DAXB guide catheter (Codman, Raynham, Massachusetts, USA), which was positioned in the right internal jugular vein. The ectatic pouch was embolized using a Penumbra Occlusion Device-5 (POD5) and a total of eight Penumbra-400 microcoils (Penumbra, Alameda, California, USA), ensuring microcoil occlusion across the multiple arterial feeder entry points, with resolution of fistula and venous reflux (figure 1 G,H). (figure 1E,F). The patient's condition improved and she was discharged in her baseline condition. A follow-up angiogram 4 weeks

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Figure 1 (A) Non-contrast CT of the head showing isolated intraventricular hemorrhage. (B) CT angiogram, maximum intensity projection, showing multiple abnormal dilated meningeal and pial vessels overlying and within the right parietal and temporal lobes. (C) Frontal and (D) lateral projections of the right external carotid artery digital subtraction angiogram (DSA) showing a dural arteriovenous fistula (dAVF) with arterial supply via the petrosquamosal and temporal branches of the right middle meningeal artery and the right occipital artery. Note the prominent ectatic venous pouch of the right transverse/sigmoid sinus. Late venous phase in (E) frontal and (F) lateral projections showing extensive reflux of the temporo-occipital cortical veins into the transcerebral veins (arrows), subependymal veins (arrowhead), and internal cerebral veins (open arrow) to the vein of Galen (asterisk). (G) Unsubtracted lateral view showing coil mass within the ectatic venous pouch. (H) Immediate follow-up DSA showing resolution of the dAVF without further venous reflux.

later showed recurrent low-grade dAVF with arterial supply via the right middle meningeal artery and the right occipital artery (figure 2A,B), for which she underwent successful transarterial Onyx-18 embolization (figure 2C). A 6-week follow-up angiogram showed complete resolution of dAVF (figure 2D). Clinically, her symptoms had resolved and her mental status, neurologic examination (including cranial nerve testing), and functional status were normal.

INVESTIGATIONS

The investigations performed were non-contrast CT scan of the head, CT angiogram of the brain, and digital subtraction cerebral angiogram.

DIFFERENTIAL DIAGNOSIS

Worsening somnolence in a patient with IVH due to dAVF may be due to obstructive hydrocephalus, venous hypertension, or seizure.

TREATMENT

1. Transvenous coil embolization of the right transverse/sigmoid sinus using POD5 and Penumbra-400 coils.
2. Transarterial Onyx-18 embolization of the right middle meningeal artery.

OUTCOME AND FOLLOW-UP

- 4-week and 6-week follow-up diagnostic angiograms
 - 6-week follow-up clinical examination
- The 6-week follow-up angiogram showed complete resolution of the dAVF. Six-week clinical follow-up showed the patient's

headache had resolved and her mental status had returned to baseline.

DISCUSSION

dAVFs account for approximately 10–15% of all intracranial vascular malformations.³ Clinically, dAVFs may present with pulsatile tinnitus, headache, cranial nerve deficits, and spontaneous intracranial hemorrhage. Intracranial hemorrhage has been reported in up to 35–42% of dAVFs and is thought to be secondary to the rupture of congested cortical veins from retrograde venous drainage.¹

Intracranial hemorrhage due to dAVFs is usually intraparenchymal or within the subarachnoid/subdural space. Isolated IVH due to dAVF is extremely rare and has only been reported six times in the literature.^{3–8} In all the previously reported cases the hypothetical etiology of IVH was retrograde cortical venous reflux into the deep medullary veins, then subsequently into the subependymal venous network with ensuing congestion and rupture. While this hypothesis makes intuitive sense from an anatomic perspective, none of the aforementioned cases demonstrated actual radiographic evidence of such reflux. Our patient demonstrated radiographic reflux through the temporo-occipital veins into the superficial and then the deep medullary veins via visualized transcerebral veins. Additionally, there was radiographic reflux to the level of the internal cerebral veins, and even to the level of the vein of Galen, lending radiographic proof for the underlying pathophysiology.

Extensive venous reflux was not the only factor contributing to our patient's deleterious presentation. In a recent report on the angioarchitectural features of high-grade intracranial dAVFs,

Figure 2 (A) Frontal and (B) lateral view of 4-week follow-up digital subtraction angiogram showing recurrent low-grade dural arteriovenous fistula (dAVF) with residual arterial supply via the petrosquamosal and temporal branches of the right middle meningeal artery (arrows) and transosseous right occipital artery branches (asterisks). (C) Unsubtracted lateral view after Onyx-18 embolization showing Onyx casting of the middle meningeal and occipital arterial feeders. (D) Six-week follow-up angiogram showing resolution of the dAVF.

venous outflow restriction, a pseudophlebitic appearance of cortical vessels, venous aneurysms, and ‘double thrombosis’ of a dural sinus (isolated sinus) were all shown to be associated with either intracranial hemorrhage or an aggressive clinical presentation.¹ In addition to severe reflux, our patient demonstrated outflow restriction in the form of a highly stenotic ipsilateral sigmoid sinus as well as a pseudophlebitic appearance of the cortical vessels, each of which confers an increased odds ratio risk of aggressive presentation of 350% and 420%, respectively.¹ We hypothesize that the combination of extensive reflux, outflow restriction, and the pseudophlebitic venous pattern of our patient all contributed to the severity of venous hypertension and her rapid clinical deterioration—a clinical consequence which was not explicitly reported in previously reported cases of isolated IVH. Prompt recognition of these high-risk angioarchitectural features of dAVFs is imperative for prognostication and effective management.

While a general consensus does not exist on the optimal treatment paradigm for transverse/sigmoid sinus dAVFs, treatment is largely dictated by clinical and anatomic circumstances.^{9,10} Transarterial or transvenous embolization techniques alone have been successful in treating low-grade lesions or those that demonstrate non-aggressive neurologic deficits.^{2,9,10} While monotherapy with either transarterial or transvenous embolization may achieve cure in low-grade lesions, a combined transvenous and arterial embolization is considered by many as the most efficient strategy for treatment of high-grade dAVFs, although a consensus does not exist on the optimal treatment paradigm.^{9,10} Accordingly, our patient was successfully treated with transvenous embolization, followed by transarterial Onyx embolization of the middle

Table 1 All reported cases of isolated intraventricular hemorrhage due to dural arteriovenous fistula

Case	Study, Year	Age (year)/sex	Clinical signs/symptoms	Location of IVH	Location of dAVF	Feeders	Grade	Treatment	Outcome
1	Halbach, 1987 ⁶	56/M	NS	NS	NS	NS	NS	IBCA and surgery	Cure
2	Aihara, 1997 ⁸	72/M	NS	Left lateral ventricle	Transverse sigmoid	NS	NS	Transarterial embolization, balloon angioplasty of stenotic right transverse sinus	Unfavorable
3	Kawaguchi, 1999 ⁷	83/F	Headache, vomiting, venous congestion	Left lateral ventricle	Transverse sigmoid	Left occipital, left middle meningeal	Lalwani IV	Transarterial liquid embolization, surgical packing of sinus	Angiographic cure
4	Irie, 2003 ⁵	67/M	Headache, nausea, vomiting	Left lateral ventricle	Transverse sigmoid	Left occipital, left vertebral, tentorial branches of left ICA	Cognard III	Transvenous coil embolization	Complete occlusion, resolved headache
5	Padmanabhan, 2008 ³	52/F	Headache	Left lateral ventricle, third ventricle	Transverse sigmoid	Left occipital, left middle meningeal, tentorial branches of left ICA	Cognard IIa+b	Transvenous coil embolization	Cessation of reflux
6	Srivastava, 2017 ⁴	17/M	Headache, left-sided paresis, slurred speech, ataxia	Bilateral occipital horns, fourth ventricle	Transverse sigmoid	Right middle meningeal, tentorial branches of right ICA	Cognard IIa+b	Transarterial NBCA embolization	Cure
7	Present case	84/F	Headache, confusion, venous hypertension	Bilateral lateral ventricles	Transverse sigmoid	Right middle meningeal, occipital, right posterior auricular	Cognard IIa+b	Transvenous coil embolization, transarterial Onyx-18 embolization	Cure

dAVF, dural arteriovenous fistula; IBCA, isobutyl-2-cyanoacrylate embolization; ICA, internal carotid artery; IVH, intraventricular hemorrhage; NBCA, n-butyl cyanoacrylate; NS, not specified.

meningeal arterial feeders. Of the previously reported cases of dAVFs with isolated IVH, multimodality therapy was employed in only two of the cases, both of which involved surgical management (table 1). We hypothesize that, in similar cases with extensive retrograde venous flow, the use of single-modality therapy with transarterial or transvenous technique alone may lead to partial/incomplete treatment, and persistent risk of intracranial hemorrhage.

CONCLUSION

We present a unique case of a dAVF causing isolated IVH with radiographic evidence of reflux to the subependymal veins, thus lending pathophysiological proof for the underlying etiology. Although isolated IVH may have a variety of etiologies, dAVFs should be considered in the differential and external carotid artery angiography may be warranted in challenging cases where an etiology does not readily present itself. In such cases of high-grade dAVF, we hypothesize that multimodality treatment may

offer the greatest chance for success, and should be strongly considered for management.

Contributors KA, CR: conception, design, analysis and interpretation of data. KA: drafting of the manuscript. KA, FAM, CR: critical revisions for important intellectual content. All authors read and approved the final manuscript.

Competing interests None declared.

Patient consent Obtained.

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Learning points

- ▶ Intracranial hemorrhagic risk of dural arteriovenous fistulas is associated with the degree of venous reflux.
- ▶ Isolated intraventricular hemorrhage due to dural arteriovenous fistula is extremely rare.
- ▶ Intraventricular hemorrhage in these cases is due to cortical venous reflux through the superficial, then the deep medullary veins via the transcerebral veins, and finally into the subependymal venous network.
- ▶ While monotherapy with either transarterial or transvenous embolization may achieve cure in low-grade dural arteriovenous fistulas, a combined transvenous and arterial embolization is considered by many as the most efficient strategy for treatment of high-grade lesions.

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