Spontaneous Intracerebral Hemorrhage Caused by an Unusual Association of Developmental Venous Anomaly and Arteriovenous Malformation

K.F. FOK, S. HOLMIN, H. ALVAREZ, A. OZANNE, T. KRINGS, P.L. LASJAUNIAS

From the Service de Neuroradiologie Diagnostique et Thérapeutique, CHU Le Kremlin Bicêtre, Paris, France

Key words: developmental venous anomaly, brain arteriovenous malformation, hemorrhage

Summary

We describe three patients who presented with spontaneous intracerebral hemorrhage resulting from the close association of developmental venous anomaly (DVA) and arteriovenous malformation (AVM). Angioarchitecturally, either the DVA formed the draining pathway for the AVM or they shared a common venous channel. The AVMs were treated by targeted embolization and the DVAs were carefully preserved. It is suggested that the unusual association of an AVM with the less flexible DVA was the cause of hemorrhage.

Introduction

Developmental venous anomalies, previously known as venous angiomas, are congenital venous anomalies of normal venous drainage. Once thought to be a rare lesion with a high propensity for hemorrhage, DVAs are now recognized as the most frequent cerebral vascular anomaly and are rarely symptomatic. A conservative approach is usually recommended. However, DVAs constitute an extreme deviation of venous drainage with reduced flexibility to venous haemodynamic changes. Any additional stress on the drainage system may upset the delicate haemodynamic balance and present clinically. We report three unusual cases showing an association of a DVA with an arteriove-

nous malformation that presented with intracerebral hemorrhage as a result of reduced adaptability of the DVA.

Case Reports

Case 1

A six-year-old boy with hereditary haemorrhagic telangiectasia (HHT) presented in December 2004 with severe occipital headache and vomiting after an emotional outburst. He had malaise but was conscious and except for right dysmetria, the physical examination was normal. Emergency CT scan of the brain showed a right cerebellar hematoma with compression of the fourth ventricle but no CSF retention. Thus, he was transferred to us for further management.

Magnetic resonance imaging (MRI) and cerebral angiogram showed a sizable DVA in the right cerebellum, which drained through the lateral mesencephalic vein and then the precentral cerebellar vein without outlet obstruction. Interestingly, at the top of the cerebellum there was an arteriovenous fistula (AVF) fed by the superior cerebellar artery draining into the DVA as the venous outlet (figure 1). The AVF was thought to be responsible for the hemorrhage and subsequent venous congestion. The fistula was successfully embolized by superselective glue injection, which led to total occlusion of the fistula with

preservation of the DVA. The child recovered favorably after the intervention and his dysmetria resolved. Follow-up angiogram four months later confirmed a stable obliteration of the arteriovenous fistula and a patent DVA. (figure 2).

Case 2

A 51-year-old man who was seen in November 2003 for right-sided headache presented with nausea and vomiting for one week. He had a history of suspected cerebral hemorrhage many years ago which resulted in right hemiparesis and epilepsy, and since then he has been treated with anticonvulsants. Physical examination showed that the right upper limb power was grade 4/5 and the right lower limb power was grade 3/5 with hypoaesthesia in the right lower limb.

A contrast-enhanced CT scan showed a left posterior frontal hemorrhagic lesion with surrounding encephalomalacia and, interestingly, a concomitant right temporal hemorrhage with surrounding vessels. A cerebral angiogram was performed which showed a left frontal arteriovenous malformation (AVM) with several intranidal aneurysms fed by frontal branches of the anterior cerebral artery. A concomitant developmental venous anomaly (DVA) was seen subependymally in the right temporal lobe, draining through a transcerebral vein and a temporal vein and finally emptying into the lateral sinus. Magnetic resonance imaging was performed which ruled out any cavernous malformation in the brain (figure 3).

Staged embolization of the left frontal AVM was performed and follow up MRI five months later confirmed the absence of any cavernous malformation in the vicinity of the DVA. After three sessions of glue embolization of the AVM, the flow, size and venous congestion was significantly reduced, since the remaining nidus did not allow further safe endovascular intervention, gamma knife radiosurgery was chosen to tackle the small residual AVM nidus.

Case 3

A 25-year-old previously healthy man without familial vascular diseases presented in December 2005 with a sudden onset of left-sided hemiparesis including left facial paresis and loss of consciousness. The CT scan showed a right-sided frontoparietal hematoma. MRI raised the suspicion of an arteriovenous mal-

formation adjacent to the hematoma (figure 4). Angiography was performed in the beginning of March 2006, i.e. two months after the hemorrhage. The patient had then recovered well from the hemorrhage with only a mild persisting paresis in the left leg.

The angiogram confirmed the presence of a large DVA located at the site of the previous hematoma in the right frontal lobe. In addition, it demonstrated an associated micro-AVM with a fistula emptying into the upper, posterior part of the DVA (figure 5). The micro-fistula was fed by a small feeder originating from the precentral gyrus branch of the right pericallosal artery. The upper and lower parts of the DVA shared a common large venous collector emptying into the superior sagittal sinus (figure 6). The fistula was embolized with glue and the result was satisfactory (figure 7). A delayed post-embolization control angiography is not yet available since it is scheduled one year after the embolization.

Discussion

According to the widely acknowledged classification of cerebral arteriovenous malformation, four different types can be differentiated both on histological sections and neuroimaging studies: arteriovenous malformations, capillary telangiectasias, cavernomas and venous angiomas¹.

Recently, the term "venous angioma" has been largely replaced by "Developmental venous anomaly" (DVA) as no proliferation of the vascular lumen has been found in this condition. The new term reiterates the fact that these "lesions" in fact only constitute an extreme anatomical variation. However DVAs may demonstrate less flexibility than "normal" venous drainage².

Figure 1 Case 1: Frames A and B demonstrate the T2- and T1-weighted non contrast axial MR scans showing a hemorrhage following suspected AVM bleeding. After left vertebral artery angiography the late arterial phase (Frames C and D) shows the arteriovenous shunt (arrow) supplied by the right superior cerebellar artery. During the venous phase of this injection (Frame E) the DVA is demonstrated. Frame F shows the 3D reconstruction of the DVA. The fistula of the arteriovenous shunt empties into the anterior, superior part of the DVA.

Figure 2 Case 1: Frame A shows the superselective angiogram demonstrating the angioarchitecture of the arteriovenous shunt before glue injection, frame B shows the glue cast following embolization. Four months after embolization, a left vertebral angiogram shows on late arterial phases (Frame C) a stable exclusion of the arteriovenous shunt and on late venous phases (Frame D) the persistence of the DVA.

DVAs were once considered to be rare vascular malformations³ but with the advent of CT and MRI, they are in fact now known to be the most common of the cerebral vascular malformations. In one series of 4069 consecutive brain autopsy studies, 105 DVAs were found, leading to a frequency of 2.5%. In the same series, only 24 AVMs and 16 cavernous malformations were found. Thus, DVAs were more than four times as common as AVMs, and represented 63% of all vascular malformations found⁴. In the clinical setting, a review of over 8200 cran-

iospinal MRI performed at one institution, revealed 50 DVAs, 33 cavernous malformations, and 17 arteriovenous malformations⁵. It is likely that the incidence of DVAs will continue to increase concomitantly with increased use of CT and MRI, higher resolution and more frequent use of contrast enhancement.

McCormick¹ described the classic anatomical features of a DVA: a composition entirely of veins with interspersed neural parenchyma and commonly thickened and hyalinized veins, having minimal smooth muscle and elastic tis-

Figure 3 Case 2: T2-weighted coronal MR scan (Frame A) shows simultaneous hemorrhage at the left frontal lobe and right temporal lobe. On right internal carotid artery angiography the solitary AVM supplied by the anterior cerebral artery can be seen in the mid-arterial phase (Frame B), while, in the venous phase (Frames C and D) the right temporal DVA draining into the right lateral sinus can be appreciated.

sue. On CT, they are rarely detectable without contrast administration. After contrast administration, they appear as linear areas of enhancement radiating either towards the ependymal surface of the ventricles or toward the venous sinuses⁶. On MRI, they may show a characteristic flow void appearance on T1- and T2-weighted images, with normal non-gliotic surrounding parenchyma. Administration of gadolinium further demonstrates the medullary veins and draining collector vein. The classical fan-shaped caput medusae is not always ap-

parent in each case, with the multiple vessels appearing as linear, curvilinear, or dot-like structures, depending on the orientation of the vessel with respect to the plane of imaging⁷.

The angiographic aspect is typical: multiple medullary veins draining normal cerebral tissue converging toward a central enlarged collector associated with absence of the venous pathways that normally drain the territory. This image is always seen during the usual angiographic venous phase⁸ and reflects the capillary-venous convergence and trans-hemispheric course that

Figure 4 Case 3: CT scan showing the right-sided fronto-parietal hematoma and T1 - weighted contrast-enhanced coronal MRI showing a suspected DVA adjacent to the hematoma. The arrow indicates the large venous collector of the DVA.

constitutes the DVA. DVA can be classified by their location as juxtacortical, subcortical and deep, according to Valavanis et Al⁹. The terminal or draining vein to which the caput medusae joins was classified as either deep or superficial². The etiology of the DVA remains specula-

tive. It is commonly thought that DVAs represent an arrest during the development of the venous system resulting in the retention of primitive embryological medullary veins draining into a single large vein. Thinking in a functional-anatomical way, Lasjaunias et Al² pointed out

Figure 5 Case 3: Right internal carotid artery angiogram. On the late arterial phase (Frame A) the fistulous point and the draining vein (arrows) emptying into the upper part of the DVA can be seen. The inserted image shows the fistula and the DVA in greater magnification. The venous phase (Frame B) demonstrates the DVA.

that the characteristic trans-hemispheric anastomotic pathway was developed in response to a hemodynamic need. Each one of them can, in a given region, be the dominant drainage of an unusual territory. They should be considered a stable functionally important “venous deviation”. Their importance was unfortunately proven in patients who underwent surgical ligation of DVAs or radiosurgical treatment and consequently developed massive venous infarcts in the corresponding territory, or resulted in significant rates of irradiation complications, respectively^{10,11}.

Attempts to ascertain the hemorrhagic risks of DVAs have been hampered by the inability to diagnose and follow the lesion before the development of a significant complication. It remains unclear whether the hemorrhagic event is really caused by the DVA or is always associated with an occult cavernous malformation¹². The issue has been further complicated by the finding that some of the “venous angiomas” identified in earlier literature were later proven to be arteriovenous malformations¹³. Garner et Al⁵ in his retrospective MRI study of DVA concluded the bleeding risk to be 0.22% / year (one hemorrhage per 4498 person-years of follow-up period), while the symptomatic bleeding risk calculated in another prospective MRI study of 80 patients was 0.34% per year¹⁴. Understandably these are groups of patients who were referred to a tertiary centre for neurolog-

Figure 6 Case 3: 3D reconstruction of the venous vascular tree on the right side showing the DVA after image editing with removal of the large right-sided bridging veins. The fistula of the associated microAVM empties into the posterior upper part of the DVA.

Figure 7 Case 3: Targeted embolization of the arteriovenous shunt was performed with glue (Frame A). Post-embolization right internal carotid artery angiogram (Frame B) shows the exclusion of the fistulous point of the microAVM. The inserted image shows the absence of the fistula at higher magnification.

Figure 8 Schematic diagram showing the association of DVAs with different types of venolymphatic defects.

Figure 9 Schematic diagram showing the different types of symptomatology associated with DVAs.

ical manifestation, and the calculated risk of hemorrhage must be viewed as the “maximal possible risk” because of the referral bias.

However, as implied by its name, DVA constitutes a venous deviation and inevitably a decreased flexibility for the territorial drainage is expected. Cases like cerebral infarction and secondary hemorrhage as a result of venous thrombosis of a DVA have been reported¹⁵. Intracranial hemorrhage as a result of DVA associated with AVM was, however, rarely discussed in the literature.

The presenting symptom of an AVM depends mostly on the venous status, and venous outlet obstruction is known to increase the bleeding risk of the lesion. While in our first case, the venous outlet to the deep venous system was not anatomically narrow, as depicted by angiography, the peculiar angioarchitecture

to use the DVA as the drainage pathway to the deep venous system constitutes a functionally restricted outlet for the high flow arteriovenous shunt. Coupled with Valsalva challenge induced by severe emotional distress and crying, we believed that it further reduced the venous outflow and triggered the hemorrhage. A similar case had been described in a paper by Gardner⁵ in which a 15-year-old girl with a solitary DVA presented with right parietal hemorrhage while she was singing, however there was no documentation of any outlet obstruction in her DVA.

In the third case presented in this article, it is also likely that the hemorrhage was caused by added stress to the venous system by the microfistula, but the patient did not describe any events that could be associated with impaired venous outflow. It rather points to the vulnera-

bility of the combination of a fistula emptying into a large DVA.

Patients presenting with simultaneous hemorrhage in an AVM and a DVA, as shown in the second case, are even rarer and, to our knowledge, have never been reported in the literature. It is hard to link the two bleeding lesions together except for their venous drainage. The right posterior temporal DVA drained to the right lateral sinus and apparently with a focal venous stenosis at the point where it entered the lateral sinus. Since the left parasagittal AVM also utilized the sagittal sinus and bilateral transverse sinus as the venous drainage, one might speculate whether enhanced flow or pressure in the drainage pathway of the arteriovenous shunt increase the back pressure for the less flexible DVA and lead to hemorrhage. Although it is difficult to prove in retrospect, similar experiences have been reported by Kunz et Al.¹⁶ who showed a patient with bilateral frontal dural arteriovenous fistula and a frontal DVA that shared the same drainage pathway.

The venous hypertension induced by the arteriovenous shunt impeded the drainage of the DVA and led to frontal venous infarction.

DVAs frequently accompany various types of venolymphatic defects as shown in figure 8. Depending on the association with such defects, DVAs can thus in rare cases contribute to or cause different types of symptomatology (figure 9). These three cases illustrate the fact that DVA must be considered non-pathological, normal venous structures, but at the same time they carry a reduced adaptability to venous hemodynamic stress¹⁷. The rare association of an arteriovenous malformation and a DVA may stretch the limit of this less flexible venous anomaly and lead to spontaneous hemorrhage.

Acknowledgements

This study was supported by Svensk-Franska Stiftelsen in Stockholm and the European Society for Neuroradiology (ESNR).

References

- McCormick WF: The pathology of vascular (arteriovenous) malformations. *J Neurosurg* 24: 807-816, 1966.
- Lasjaunias P, Burrows, Planet C: Developmental venous anomalies (DVA): the so call venous angioma. *Neurosurg Rev* 9: 233-244, 1986.
- Huang YP, Wolf BS: Veins of the white matter of the cerebral hemispheres (the medullary veins): diagnostic importance in carotid angiography. *Am J Roentgenol* 92: 739-755, 1964.
- Sarwar M, McCormick WF: Intracerebral venous angioma. Case report and review. *Arch Neurol* 35: 323-325, 1978.
- Gardner TB, Del Curling O et Al: The natural history of intracranial venous angiomas. *J Neurosurg* 75: 715-722, 1991.
- Hammoud D, Beauchamp N et Al: Ischemic complication of a cerebral developmental venous anomaly: case report and review of the literature. *J Comput Assist Tomogr* 26(4):633-636, 2002.
- Lee C, Pennington MA, Kenney CM: MR evaluation of developmental venous anomalies: Medullary venous anatomy of venous angiomas. *Am J Neuroradiol* 17: 61-70, 1996.
- Goulao A, Alvarez H et Al: Venous anomalies and abnormalities of the posterior fossa. *Neuroradiology* 31: 476-482, 1990.
- Valavanis A, Wallauer J, Yasargil MG: The radiological diagnosis of cerebral venous angioma: Cerebral angiography and computed tomography. *Neuroradiology* 24: 193-199, 1983.
- Tropper R, Jurgens E, Reul J: Clinical significance of intracranial venous anomalies. *J Neurol Neurosurg Psychiatr* 67: 234-238, 1999.
- Lindquist C, Guo WY et Al: Radiosurgery of venous angiomas. *J Neurosurg* 78: 531-536, 1993.
- Rigamonti D, Spetzler RF, Medina M: Cerebral venous malformations. *J Neurosurg* 73:560-564, 1990.
- Cabanes J, Blasco R, Garcia M: Cerebral venous angiomas. *Surg Neurol* 11: 385-389, 1979.
- McLaughlin MK, Kondziolka D, Flickinger JC: The prospective natural history of cerebral venous malformation. *Neurosurg* 43(2): 195-201, 1998.
- Masson C, Godefroy O, Leclerc X: Cerebral venous infarction following thrombosis of the draining vein of a venous angioma (developmental abnormality) *Cerebrovasc Dis* 10: 235-238, 2000.
- Kunz A, Voros E, Varadi P: Venous cerebral infarction due to simultaneous occurrence of dural arteriovenous fistula and developmental venous anomaly. *Acta Neurochir (Wien)* 143: 183-1184, 2001.
- Lasjaunias P, Berenstein A, terBrugge K: *Surgical neuro-angiography*, vol 1. Springer, Berlin Heidelberg, New York, Tokyo, 656-661, 2001.

Pierre L. Lasjaunias, MD, PhD
Hôpital de Bicêtre, Service de Neuroradiologie
Diagnostic et Thérapeutique,
78, Rue du Général Leclerc
94275 Le Kremlin-Bicêtre, France
E-mail: pierre.lasjauniasct.ap-hop-paris.fr