Childs Nerv Syst
DOI 10.1007/s00381-010-1253-4

SPECIAL ANNUAL ISSUE

Cerebral developmental venous anomalies

Diego San Millán Ruíz · Philippe Gailloud

Received: 20 July 2010 / Accepted: 23 July 2010

Abstract
Introduction Cerebral developmental venous anomalies (DVAs) are the most frequently encountered cerebral vascular malformation. As such, they are often observed incidentally during routine CT and MRI studies. Yet, what DVAs represent from a clinical perspective is frequently not common knowledge and DVAs, therefore, still generate uncertainty and concern amongst physicians. This article reviews our current understanding of developmental venous anomalies.
Results In the majority of cases, DVAs follow a benign clinical course. On rare occasions, DVAs become symptomatic generally due to an underlying associated vascular malformation such as cavernous malformations or thrombosis of the collecting vein. Rare forms of DVAs include arterialized DVAs and DVAs involved in the drainage of sinus pericranii, which warrant additional investigation by digital subtraction angiography. Cerebral abnormalities such as atrophy, white matter lesions and calcifications within the drainage territory of asymptomatic DVAs, are often identified on CT or MR imaging studies and likely represent secondary changes due to venous hypertension. There is increasing evidence that DVAs have a propensity for developing venous hypertension, which is thought to be the cause of associated cavernous malformations and parenchymal abnormalities.
Conclusions DVAs represent variations of the normal cerebral venous angioarchitecture and by enlargement follow an uneventful clinical course. Complications can, however, occur and their management requires a thorough understanding of the nature of DVAs, including their frequent coexistence with other types of vascular malformation, and the existence of more complex but rare forms of presentation, such as the arterialized DVAs.

Keywords Developmental venous anomaly · Imaging · Venous hypertension · Regional parenchymal brain abnormalities · Clinical implications · Arterialized developmental venous anomaly · Morphology

D. San Millán Ruíz (✉)
Neuroradiology Unit, Department of Diagnostic and Interventional Radiology, Hospital of Sion,
Avenue du Grand-Champsec 80,
1951 Sion, Valais, Switzerland
e-mail: diego.san-millan-ruiz@rsv-gnw.ch

P. Gailloud
Division of Interventional Neuroradiology, The Johns Hopkins Hospital,
Baltimore, MD, USA

Introduction

Cerebral vascular malformations are classified into capillary telangiectesias, cavernous malformations (CM), arteriovenous malformations (AVM), and developmental venous anomalies (DVAs) [46]. DVAs, CMs, and capillary telangiectesias involve the venous circulation, with DVAs being the most frequent of all cerebral vascular malformations. The term cerebral "developmental venous anomaly", proposed by Lasjaunias et al. [38], is now widely used as a synonym for venous angioma, cerebral venous malformation, or cerebral venous medullary malformation. This appellation relates to the nature of the DVA, which is currently viewed as a normal variation of the cerebral parenchymal venous angioarchitecture rather than a true malformation. Though most DVAs are discovered fortuitously and bear no clinical significance, their diagnosis often generate concern among physicians less familiar with the field of intracranial vascular malformations. Complications can, however, occur and their management requires a thorough understanding of the nature of DVAs, including their frequent coexistence with other types of vascular malformation, and the existence of more complex but rare forms of presentation, such as the arterialized DVAs.

This article reviews our current understanding of DVAs, focusing in particular on their morphological and radiolog-

Published online: 12 August 2010

ical aspects, their development, and their clinical implications and rare complications. To the best of our knowledge, there are no existing reports specifically studying DVAs in the pediatric population. Therefore, much of what is currently known about DVAs derives from studies mostly including adult patients. It is likely, however, that differences exist between children and adults in terms of type and frequency of associated findings.

Epidemiology, developmental considerations, and morphological characteristics

DVAs are the most frequently encountered cerebral vascular malformations, with an incidence of up to 2.6% reported in a series of 4,069 brain autopsies [65]. They are often discovered fortuitously during routine brain imaging using computed tomography (CT) or magnetic resonance imaging (MRI). DVAs are seen both in the pediatric and adult populations, with a slight male predominance [32, 62].

DVAs form during intrauterine life. Though there is no consensus as to the pathogenetic mechanism underlying their development, DVAs likely result from the recruitment of locoregional parenchymal veins compensating for the absence or loss of part of the cerebral venous system (e.g., arrested development or thrombosis) [38, 61, 80]. Therefore, they may be better described as being the result of a fetal pathological event rather than true anatomic variations. As DVAs usually provide sufficient collateral outflow for the brain parenchyma, they are in the vast majority of cases not associated with congenital cerebral lesions. Rare association between anomalous parenchymal veins and migrational brain disorders exist, but the venous anomaly is thought to be a manifestation of the underlying abnormal neuronal migration and not its cause [8, 18, 74].

DVAs represent an anomalous venous disposition due to the absence of normal pial or subependymal veins. They may be understood as a reconfiguration of the venous angioarchitecture, in which a deep venous territory drains centrifugally towards either the pial veins of the cerebral surface or directly into a dural venous sinus, or, alternatively, a cortical and subcortical venous territory drains centripetally towards deep subependymal veins.

The morphological hallmark of a DVA is a cluster of venous radicles that converge into a larger collecting vein, giving the DVA its typical *caput medusae* appearance. The collecting vein crosses a variable length of brain parenchyma to either join the superficial or deep venous system. Occasionally, DVAs may drain into both the superficial and deep venous systems, one of these drainage pathways being predominant [64]. The essential role played by DVAs in the normal cerebral venous drainage is illustrated by cases of catastrophic venous ischemic and hemorrhagic complications resulting from the surgical removal of a DVA [2, 69]. In order to prevent such complications, care must be taken to preserve the collecting vein of a DVA during the surgical evacuation of a cerebral hematoma or the resection of a CM located in its vicinity.

Histopathological descriptions of DVAs are scarce. Our observations [63] of hyalinated collecting veins with parietal fibrous thickening, absence of elastic lamina, and loosely arranged smooth muscle layers are consistent with the few ultrastructural studies currently available in the literature [17, 45]. The cerebral tissue within the drainage territory of a DVA has been classically described as normal. Noran [51], however, found evidence of "demyelination, degenerative alterations of nerve cells, gliosis and leukomalacia…" around DVAs, which is in keeping with the documentation of white matter abnormalities on CT and MRI studies (see below) [62].

A stenosis of the collecting vein of DVAs is commonly observed [19, 62, 63, 74], typically at the point where the vein crosses the dura to drain into a dural venous sinus. Another frequently encountered abnormality is an ampullary dilatation of the proximal segment of the collecting vein, which can be seen in up to 27.3% of cases [62]. The cause of this dilatation is not clear in the absence of histological correlation, but the proximal widening of the collecting vein suggests venous stasis and outflow obstruction perhaps caused by focal thickening of the collecting vein wall.

DVAs can affect a variable volume of brain parenchyma, ranging from a few sulci or a circumscribed wedge of periventricular white matter to an entire hemisphere, in which case their venous architecture may be very complex (Fig. 1). They are more frequently located at the supratentorial level, with a frontal predominance [39, 62]. Several collecting veins may be observed in about 6% of DVAs [62] (Fig. 5), most frequently with posterior fossa and large DVAs. In addition, two or more DVAs coexisting in separate regions of the brain have been observed in 1.2% to 16% of cases [39, 62, 75].

Imaging of developmental venous anomalies

Both CT and MRI can demonstrate the typical *caput medusae* draining into a collecting vein, allowing confident diagnosis of DVAs without the need to obtain digital subtracted angiography (DSA) (Figs. 1, 2, 3, 4, and 5) [58, 74, 79]. However, because of its higher temporal resolution, DSA remains the best imaging modality to study the hemodynamic behavior of DVAs. For this reason, DSA is performed in patients presenting with ischemic or hemorrhagic complications within the drainage territory of a DVA, or whenever an associated vascular malformation is suspected on CT or MRI. DVAs typically opacify during the venous phase of the angiogram, concomitantly to the

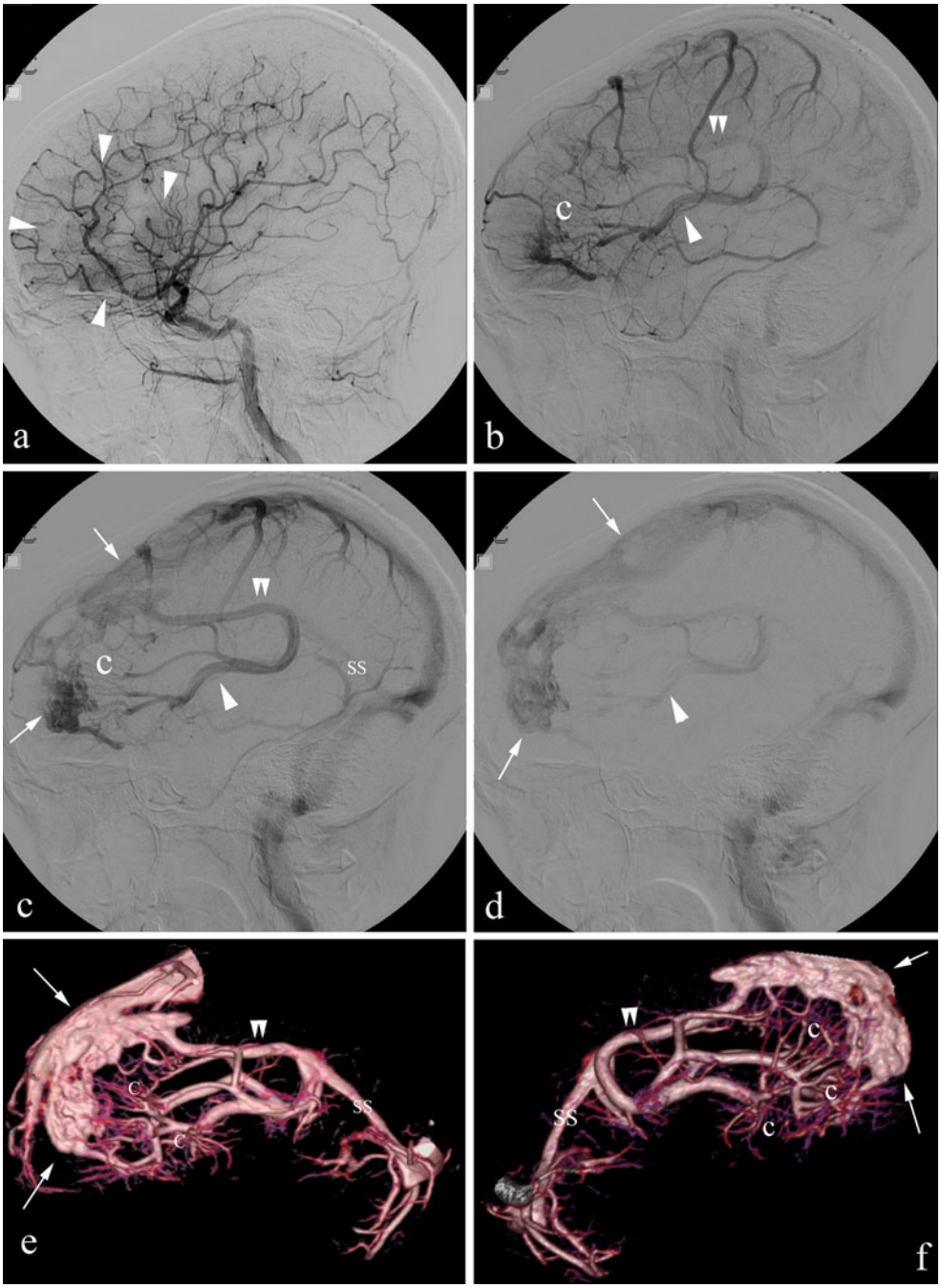


Fig. 1 A 20-year-old woman investigated for craniopharyngioma, in whom MRI demonstrated a large complex deep developmental venous anomaly. **a**–**d** DSA, left common carotid artery injection, arterial to late venous phases. A parenchymal blush appears in the left frontal lobe (*arrowheads*) during the arterial phase (**a**). The early, intermediate, and late venous phases (**b**–**d**) demonstrate a complex DVA of the deep venous system. The DVA appears early in the venous phase and there is delayed washout. The classical anatomy of the internal cerebral veins is not identifiable. Instead anomalous collecting veins (*arrowhead*) drain a network of caput medusae (caput medusae-c) located in the parenchyma surrounding the frontal horns and body of the lateral ventricles on both sides. The principal drainage of the DVA is through a dysplastic and dilated superior sagittal sinus (*arrow*) by way of a tubular venous channel corresponding to the inferior sagittal sinus (*double arrowhead*). Secondary minor drainage also occurs towards the straight sinus (*SS*) by way of the vein of Galen. **e**–**f** Volume rendered reconstructions of the venous phase of a subtracted dynamic CT angiography obtained with a 320-multidetector row CT, left lateral (**e**) and right lateral (**f**) views, demonstrating the anatomy of the DVA (the surrounding structures and the distal two thirds of the superior sagittal sinus have been removed). Figure legend as in **a**–**d**

normal cerebral veins. Delayed outflow of a DVA suggesting drainage impairment may sometimes be observed, even in the absence of a demonstrable collecting vein stenosis.

On non-contrast CT, the collecting vein of the DVA may appear to be isodense or slightly hyperdense to the cortex if patent, or markedly hyperdense if acutely thrombosed (Fig. 2). In addition, non-contrast CT may disclose an associated hemorrhage, parenchymal calcifications, and atrophy or white matter lesions in the drainage territory of the DVA [62]. Both the collecting vein and the *caput medusae* enhance after administration of contrast material; the anatomy of the DVA is best demonstrated by thin-section CT venography.

Non-contrast T2- and T1-weighted MRI may demonstrate flow voids and phase-shift artifact produced by the collecting vein of a DVA and by the larger venous radicles of the *caput medusae.* [39]. The DVA is best appreciated after gadolinium administration, particularly with 3D-contrast-enhanced echo-gradient T1-weighted images. MRI is superior to CT in demonstrating associated parenchymal abnormalities, such as white matter lesions, locoregional cerebral atrophy, and CMs

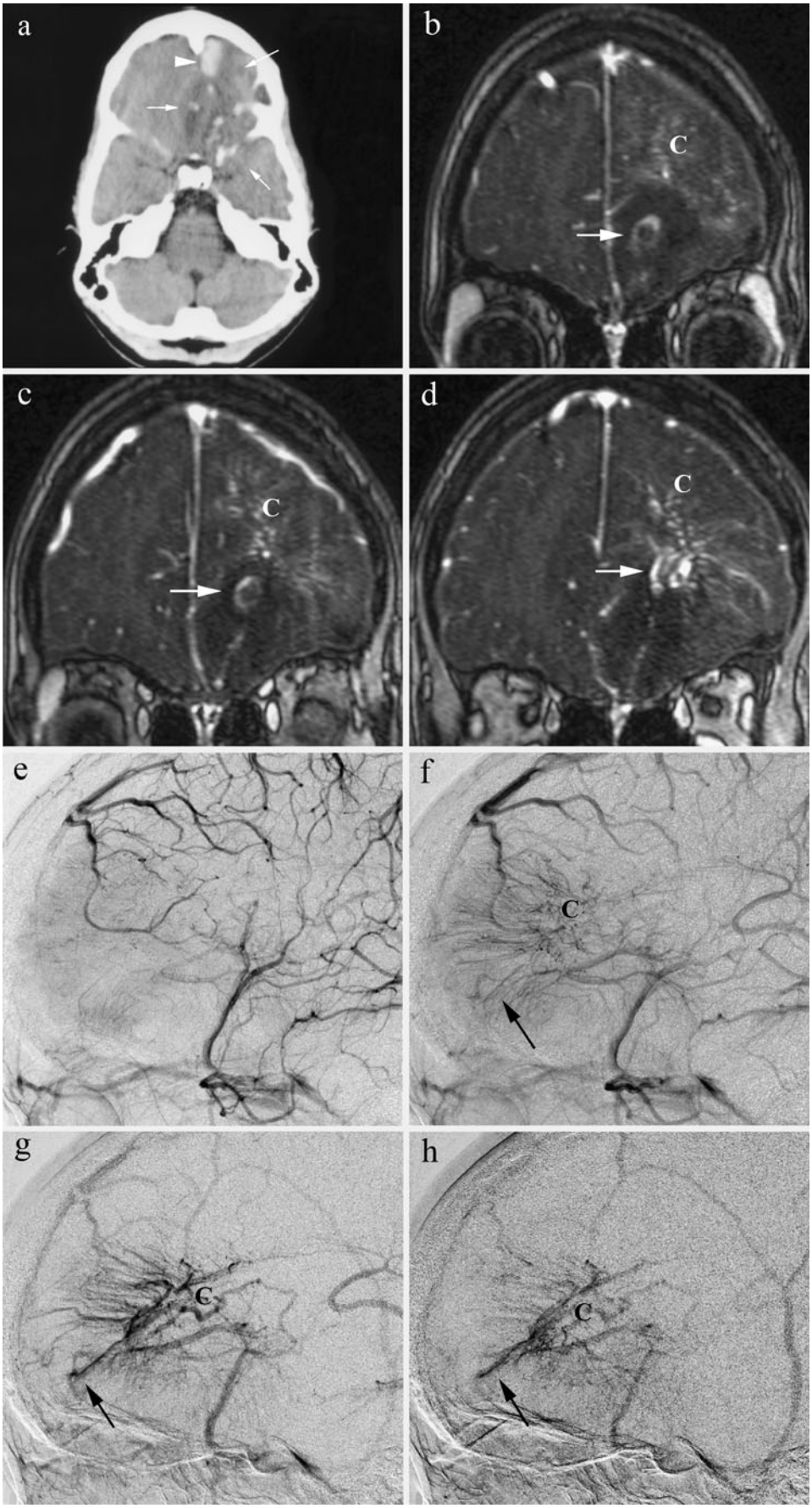


Fig. 2 A 17-year-old girl with left fronto-orbital venous infarction secondary to a thrombosed DVA. **a** Non-contrast CT demonstrating an area of venous infarction (*arrows*) and the hyperdense, acutely thrombosed collecting vein of a DVA (*arrowhead*). **b**–**d** Coronal gadolinium-enhanced 3D T1-weighted images showing a filling defect within the collecting vein consistent with a thrombus (*arrow*) and the enhancing caput medusae (**c**). **e**–**h** DSA, early to late venous phases of a left common carotid artery injection, lateral projection. A parenchymal blush is observed during the early venous phase (**e**). The caput medusae (**c**) shows delayed filling and washout consistent with the collecting vein thrombosis. The collecting vein is only partially filled (*arrow*)

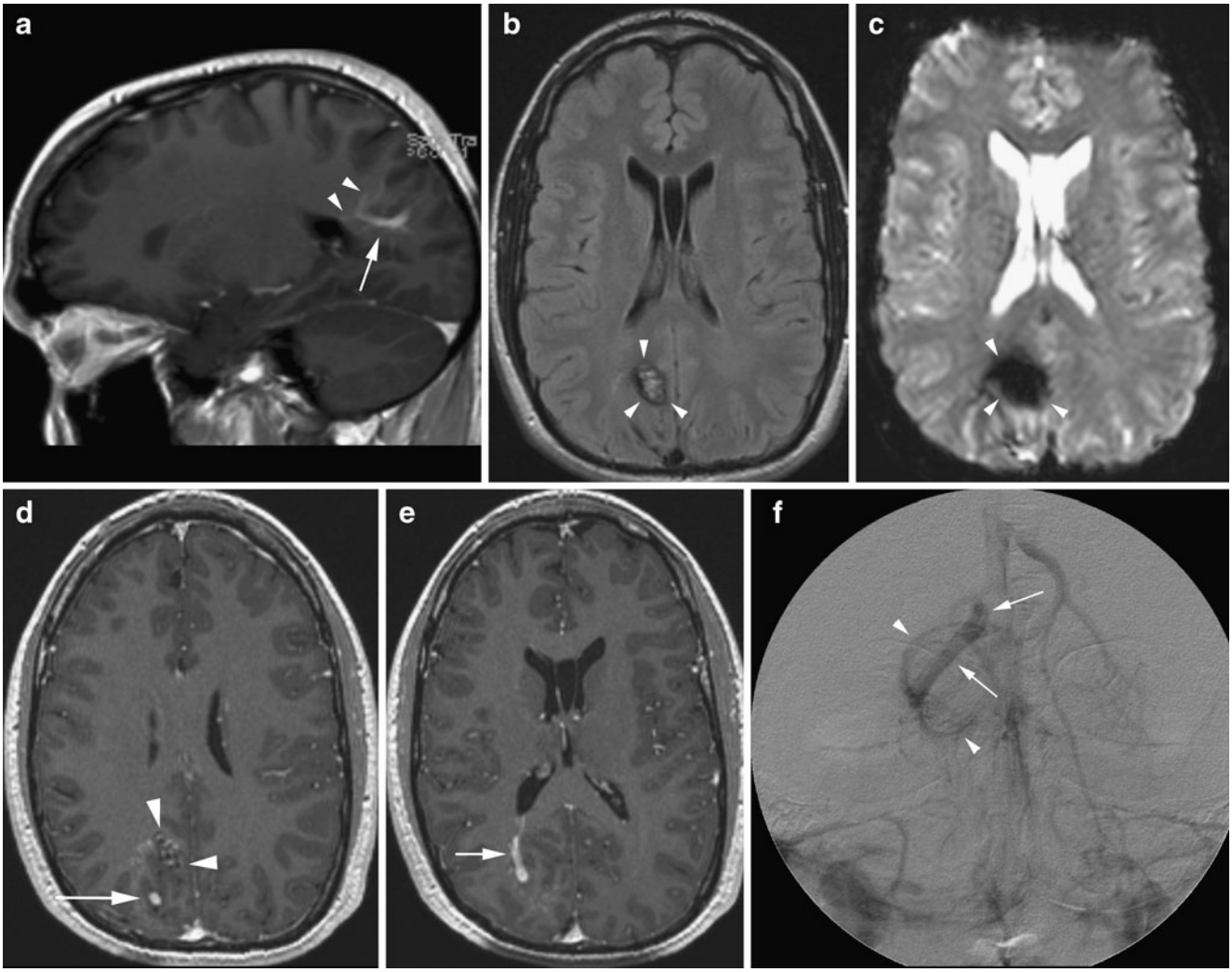


Fig. 3 A 34-year-old man presenting with seizures. **a** Sagittal Gadolinium-enhanced spin-echo T1-weighted image showing a typical DVA with its collecting vein (*arrow*) and small venous radicles corresponding to the caput medusae (*arrowheads*). **b** Axial FLAIR, documenting an associated cavernous malformation (*arrowheads*) consisting of a heterogeneous, well demarcated lesion surrounded by a hypointense rim consistent with hemosiderin deposition. **c** Axial B0 diffusion-weighted image showing susceptibility artifact of the cavernous malformation (blooming effect). **d**, **e** Axial gadolinium-enhanced 3D T1-weighted image demonstrating the heterogeneous enhancement of the cavernous malformation (*arrowheads*) within the drainage territory of the DVA. The collecting vein is opacified as well (*arrow*). **f** DSA, venous phase of a left vertebral artery injection, anteroposterior projection, showing the DVA with its caput medusae (*arrowheads*) and collecting vein (*arrows*) draining into the posterior third of the superior sagittal sinus. Note the change in caliber of the collecting vein immediately prior its junction with the sagittal sinus indicating a stenosis

[62]. Because CMs are frequently associated with DVAs (see below), they should be specifically sought for by using hemosiderin or deoxyhemoglobin sensitive sequences such as echo-gradient T2-weighted images or susceptibility-weighted images (SWI) [67].

Clinical presentation

DVAs are associated with one or more regional CMs in 13% to 40% of the cases [30, 62]. These CMs are now thought to be responsible for the vast majority of symptoms attributed to DVAs in the pre-CT/MRI era [47, 57]. Garner et al. [24] retrospectively evaluated the risk of hemorrhage associated with DVAs to be of 0.22% per year, while McLaughlin et al. [47] prospectively found that risk to be of 0.68% per year, though only half of their patients were symptomatic. It was initially proposed that DVAs located in the posterior cranial fossa had a higher propensity to bleed than supratentorial DVAs [60], but recent evidence suggests that this is not the case [47].

Hemorrhagic and/or ischemic infarction, or reversible cerebral edema around a DVA may result from acute thrombosis of the collecting vein [12, 22, 25, 26, 33, 34, 37, 41, 48, 54, 68, 71, 76, 77, 82] (Fig. 2), although these changes can also be occasionally observed in the presence of a patent venous collector [5, 27, 55]. At the time of this

review, there were 21 cases of symptomatic thrombosed DVAs documented in the literature, presenting with venous ischemic infarction (53%), parenchymal hemorrhage (37%), or subarachnoid and intraventricular hemorrhage (5%); in 5% of cases, there was no intra- or extra-axial lesions [1, 12, 22, 25, 26, 33, 34, 37, 41, 48, 54, 68, 71, 76, 77, 82]. Management was conservative in nine cases (43%), involved anticoagulation in seven (33%) or surgical decompression for a cerebral hematoma or for mass effect secondary to venous infarction in five cases (24%). Clinical outcome was favorable in eighteen out of 21 patients (86%), with complete recovery or persistence of mild neurological symptoms, though Rankin scores were usually not made available. One of the reported cases described a 9-year-old boy with clinical improvement after heparin administration [76]. There are no large or controlled studies to support systemic anticoagulation over conservative management in cases of thrombosed DVAs. However, the experience gained from the treatment of cortical or dural venous sinus thrombosis suggests that systemic and prolonged anticoagulation should be beneficial in preventing clot propagation, in favoring recanalization, and in promoting reversibility of the symptoms in cases involving DVAs as well [20, 66]. Screening for prothrombotic conditions should be carried out, as it would with a cortical or dural venous sinus thrombosis. Symptomatic cerebral hematomas and/or edema may warrant decompression craniectomy, taking particular care to preserve the drainage pathway of the DVA during clot aspiration.

Anecdotal publications have reported the association of DVAs with choreoballismus [14], obstructive hydrocephalus through compression of the cerebral aqueduct [11], ophthalmoplegic migraine [9], or nerve root compression [43, 56]. There seems to be no demonstrable relation between uncomplicated DVAs and epilepsy or headaches [50, 70, 73].

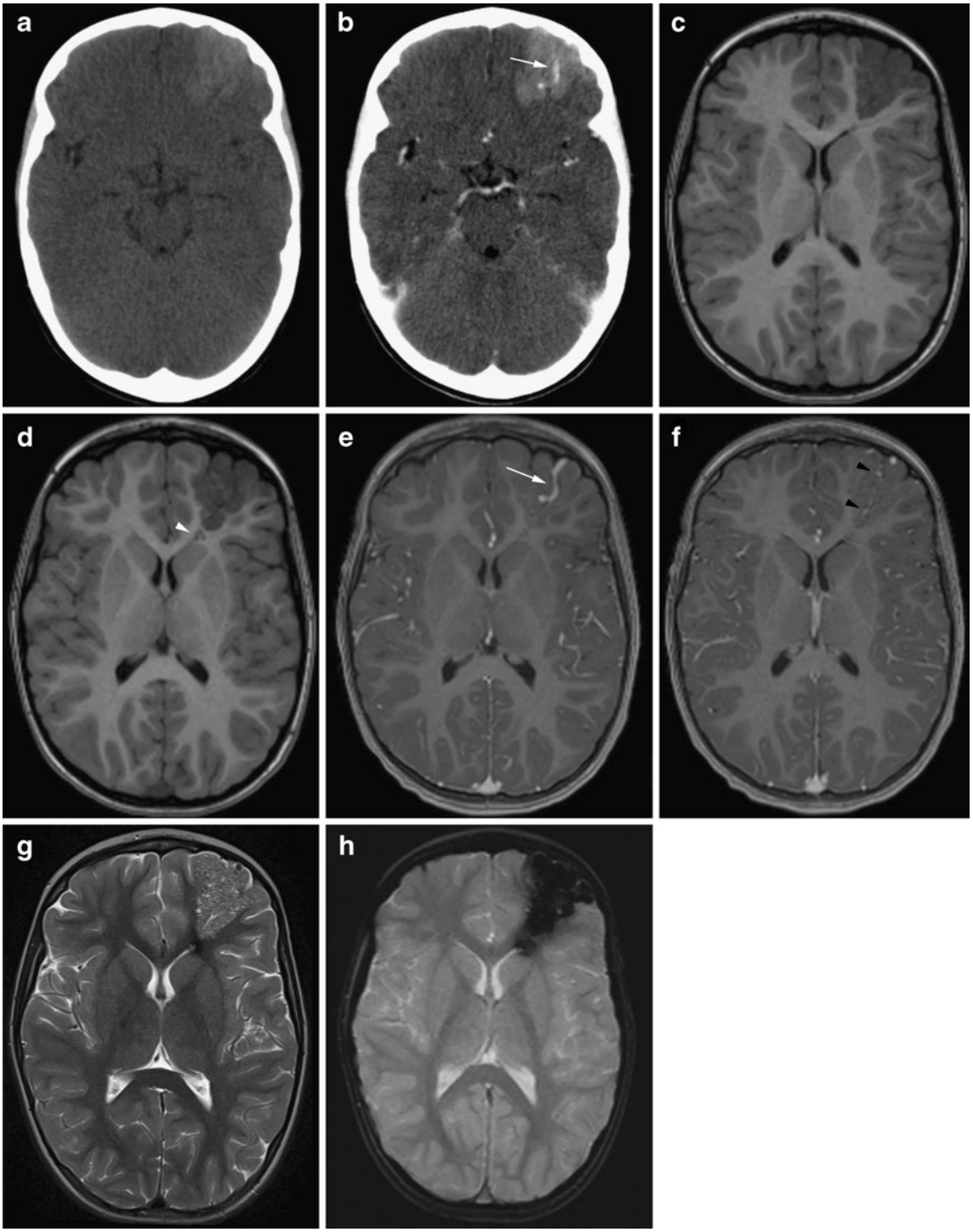
Association with other vascular malformations

DVAs are associated with sporadic CMs in 13% to 40% of cases [30, 62]. The CM is typically located in the region of the *caput medusae* (Fig. 3). This close topographic relation between CMs and DVAs, as well as reports describing the de novo formation of CMs or CM-like lesions in the proximity of a DVA [15, 16, 19, 42, 63], suggest a causative link between CMs and DVAs. Evidence of subclinical microhemorrhages in the form of hemosiderin-laden macrophages surrounding a DVA has been demonstrated by San Millan Ruiz et al. [63], supporting the hypothesis that blood diapedesis through the walls of the venous radicles or rupture of one of the radicles may occur [19, 80]. Repeated microhemorrhages around DVAs are thought to induce the formation of CM-like lesions by activating angiogenic growth factors such as the vascular endothelial growth factor (VEGF), and lead to reactive angiogenesis with vessel formation and coalescence, a process that has been referred to as "hemorrhagic angiogenic proliferation" [7, 59]. The hemorrhagic risk of CMs associated with DVAs may be higher [81] than the 2.6% to 3.1% per patient–year reported for isolated CMs [35, 49].

CMs within the drainage territory of a DVA may be unique or multiple and of variable sizes. MRI, and to a lesser extent CT, is helpful in demonstrating acute hemorrhage within the CMs, mostly by demonstrating perilesional vasogenic edema and mass effect. On rare occasions, diffuse cavernous transformation of the entire drainage territory of the DVA may be observed (Fig. 4).

The association between DVAs and superficial venous malformations of the head and neck is well established. DVAs may be encountered in up to 20% of patients with a large superficial venous malformation, a much higher incidence than in the general population [13]. DVAs are also associated with lymphatic or lymphaticovenous malformations of the orbit or periorbital region, with an incidence as high as 60.6% in a recent series of 33 patients [10]. In addition, these patients presented other concomitant intracranial vascular abnormalities, such as CMs (6.1%), dural arteriovenous fistulas (12.1%), pial arteriovenous malformations (3%), and sinus pericranii (3%). Finally, DVAs may be part of the manifestations of some neurocutaneous disorders, such as in the blue-rubber-bleb syndrome [21].

Fig. 4 A 6-year-old girl presenting with partial complex seizures. **a** ▶ Non-contrast CT demonstrating a spontaneous hyperdense wedge-shaped lesion in the left frontal lobe, involving the cortex and superficial white matter, extending to the deep white matter around the frontal horn of the left lateral ventricle on other slices (not shown). **b** Contrast-enhanced CT documenting mild enhancement of the lesion and a superficially draining DVA within the lesion (*arrow*). The DVA showed no evidence arterialization on DSA and CTA (not shown), but several segmental narrowings were demonstrated on the collecting vein on its course towards the superior sagittal sinus. **c**–**f** Axial 3D T1-weighted images before and after administration of Gadolinium. The hyperdense lesion shown on CT is spontaneously hypointense (**c**, **d**), with slight enhancement observed after contrast administration (**e**, **f**). The collecting vein of the DVA (*arrow*) and the venous radicles of the caput medusae (*black arrowheads*) enhance markedly. A small focal area of signal abnormality is also observed in the periventricular white matter around the frontal horn of the lateral ventricle (*white arrowhead*). **g** Axial fast-spin-echo T2-weighted image showing the heterogeneous appearance of the cortical and subcortical lesion, which contains multiple hyperintense foci. The deep white matter around the left frontal horn is hypointense suggesting the presence of deoxyhemoglobin or hemosiderin. **h** Axial echo-gradient T2 reveals a marked susceptibility effect with "blooming" of the lesion in keeping with hemosiderin deposition. These findings are consistent with a diffuse cavernous transformation of the parenchyma drained by the DVA. The focal lesion adjacent to the left frontal horn is typical for a cavernous malformation as well. In this case, outflow obstruction in the form of segmental stenoses was demonstrated by DSA



The concomitant occurrence of a DVA with a sinus pericranii (SPi) is well established, suggesting a common developmental origin for both venous malformations. Sinus pericranii consist of an anomalous extracranial drainage of the intracranial circulation occurring through a diploic emissary veins into an enlarged superficial venous pouch, itself connected to the subgaleal and scalp venous systems. DVAs can occasionally drain partially or completely into a sinus pericranii. In a recently published series, a DVA was found in seven out of 15 patients with a SPi [23], and in at least two cases, the SPi was the major drainage pathway of the DVA. This finding emphasizes the need for a precise exploration of the cerebral venous anatomy when planning the treatment of a sinus pericranii.

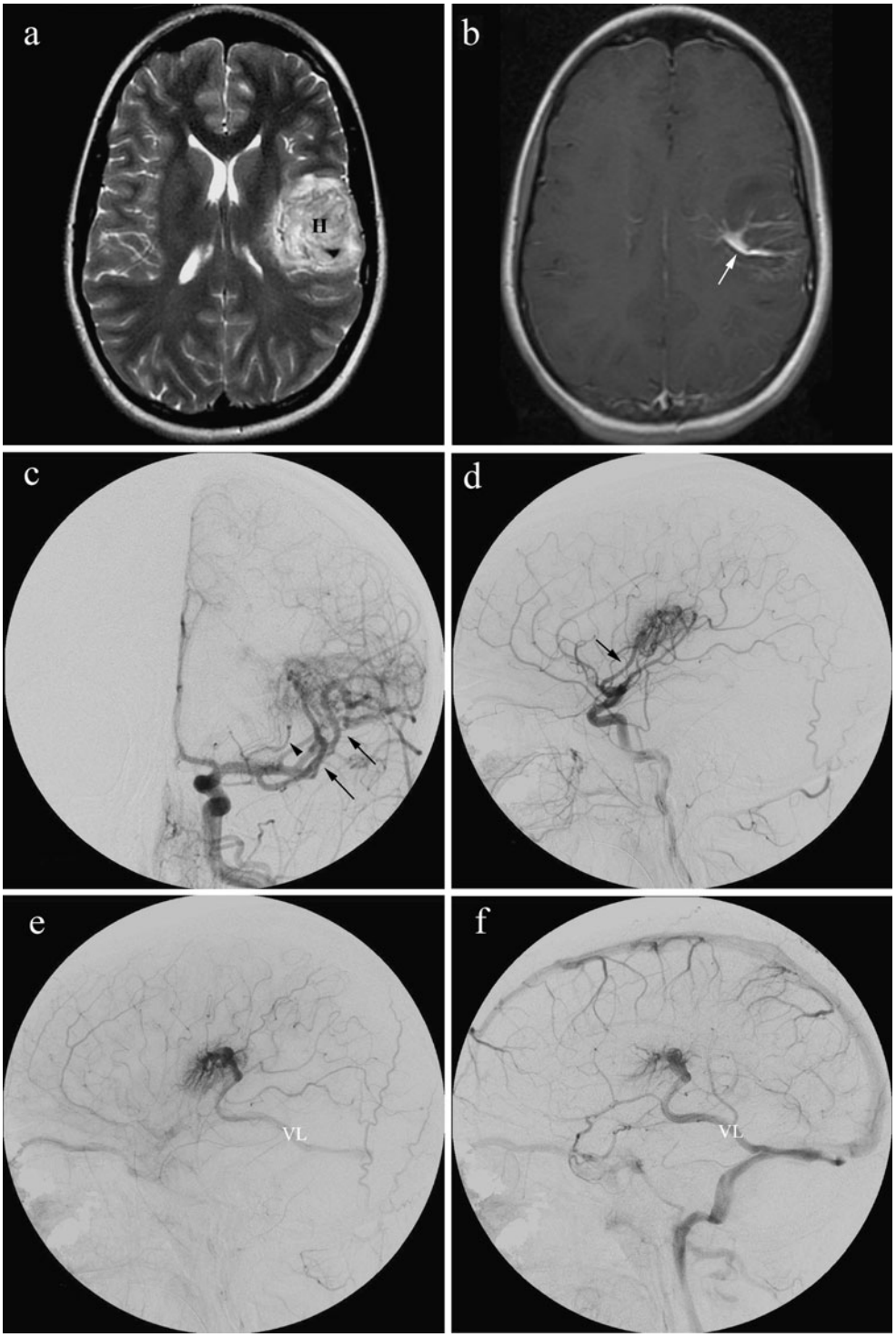


Fig. 5 A 16-year-old girl with a left fronto-opercular hemorrhage. **a** Axial T2-weighted image demonstrating the acute left fronto-opercular acute hemorrhage (H). **b** Gadolinium-enhanced T1-weighted image showing the typical appearance of a DVA with its collecting vein and caput medusae. The collecting vein drains into the left lesser anastomotic vein of Labbé and is patent. No distal stenosis of the collecting vein was visible, however, an ampullary dilatation of the proximal collecting vein can be seen (*arrow*). **c–f** DSA was performed to rule out the presence of an associated arteriovenous shunt. A left common carotid artery injection, arterial phase, anteroposterior projection demonstrates a parenchymal blush projecting over the left fronto-opercular region (**c**). There is enlargement of the insular branches of the left middle cerebral artery (*arrows*) and of a lateral lenticulostriate artery (*arrowhead*). These findings are also demonstrated on the lateral projection, in the intermediate (**d**) and late (**e**) arterial phases. The collecting vein drains into the lesser anastomotic vein of Labbé (VL), which appears early during the late arterial phase (**e**). The dilated venous radicles of the caput medusae of the DVA are well delineated in the venous phase (**f**). No collecting vein stenosis is observed, but DSA confirms the ampullary dilatation visualized by MRI. In the absence of an AVM nidus, these findings are consistent with a Type 2 arterialized DVA

When surgical or endovascular closure of a sinus pericranii that drains a DVA is contemplated, particular care must be taken in demonstrating an alternative drainage pathway of the DVA [63]. If no such pathway exists, closure of the sinus pericranii is precluded by the risk of cerebral venous infarction secondary to interrupting the outflow of the DVA.

Associated cerebral regional abnormalities and physiopathologal considerations

Anecdotal literature reports mention abnormalities of the brain surrounding DVAs [6, 19, 30, 51, 61, 74, 75]. However, in a recent series of 84 consecutive DVAs explored by MRI and CT, parenchymal abnormalities (other

than CMs) within the drainage territory of the DVA were found in 65% of the cases [62]. These included locoregional cerebral atrophy found in close to a third of the cases (29.7%), dystrophic calcification (9.6% by CT), and non-specific white matter lesions reminiscent of the lesions observed in patients with vascular leukoencephalopathy and likely identical to the lesions described by Noran [51] (28.3% by MRI, 19.3% by CT). Parenchymal abnormalities were not related to the size or the location of the DVA. The incidence of these abnormalities in the pediatric population is unknown, although they are likely less frequently encountered than in adults as parenchymal changes are thought to be secondary to long-standing venous hypertension (VHT) within the DVA system.

There is a substantial body of evidence from the literature to support that VHT is the underlying mechanism leading to the spectrum of brain lesions associated with DVAs. Impaired brain perfusion attributed to venous congestion was documented by several authors in areas drained by either small or large DVAs [44, 72]. Intraoperative documentation of increased venous pressure within a DVA was published by Dillon [19]. The venous outflow impairment due to a stenosis of the collecting vein may account for VHT in a substantial number of cases. As previously mentioned, parietal thickening of the veins forming the DVA may also contribute to the development of VHT, even in the absence of a demonstrable stenosis, by reducing the size and compliance of the vessel lumen, increasing the resistance to flow, and diminishing the vessel's capacity to adapt to pressure modifications [62, 63]. In addition, DVAs represent a point of venous confluence, where a single collecting vessel drains an abnormally large venous territory. The resulting volume overload may participate in the development of VHT [62]. A recent study suggests that angioarchitectural factors, such as tortuosity of the medullary veins (venous radicles) and collecting vein, may predispose to the development of CMs within the drainage territory of a DVA [29].

Atypical forms of developmental venous anomalies: arterialized DVAs

Early angiographic opacification of a DVA during the mid or late arterial phases may exceptionally be observed on DSA studies, typically in large supratentorial DVAs. There is no accepted term designating this subtype of DVAs, which are variably referred to as arterialized DVAs, mixed-type vascular malformations, arteriovenous malformations with venous predominance, or intracerebral venous angiomas with arterial blood supply [4, 7, 19, 28, 31, 52, 53, 61, 63, 78]. Uncertainty remains in regard to the nature and pathogenesis of this subtype of DVA, which could represent a spectrum between a simple DVA and a "classic" AVM. Some authors have suggested that the arteriovenous shunt develops secondarily from a pre-existing DVA. In this hypothesis, thrombosis of some venous radicles leads to venous hypertension and ischemia, triggering the expression of VEGF, which in turn leads to neoangiogenesis and, ultimately, to the formation of the arteriovenous shunt [3]. A similar mechanism has been discussed above in regard to the formation of CMs associated with DVAs. Though appealing, these theories do not exclude the possibility of an underlying infra-radiological vascular abnormality, such as a capillary telangiectasia or a micro arteriovenous shunt, acting as precursors for the CM-to-be or the arterialized portion of the DVA.

DSA remains necessary to adequately characterize arterialized DVAs. A recent publication has proposed that three types of "arterialized DVAs" may exist [63]:

- Type 1 typical DVAs with a *caput medusae* blush occurring during the mid or late arterial phases, without demonstrable arterial feeders or AVM nidus (Fig. 1);
- Type 2 "arterialized DVAs" with enlarged arterial feeders to the *caput medusae*, without an angiographically demonstrable AVM nidus (Fig. 5). This type of DVA seems to correspond to the lesions described by Im et al. [31], in which histological examination revealed dilated arterialized veins with hyalinized walls containing elastic laminae interspaced with normal brain tissue;
- Type 3 DVAs draining an angiographically demonstrable AVM [4, 36, 40, 52, 53].

To the best of our knowledge, there are no reports of Type 1 arterialized DVAs presenting with hemorrhage. On the other hand, Types 2 and 3 seem to have a propensity to bleed [4, 36, 40, 52, 53], and their clinical behavior could be similar to a typical AVM [31]. The management principles of arterialized DVAs are not well established, and there is not sufficient data in the literature to delineate a standardized treatment strategy [63]. In the few case reports or small series published so far, Types 2 and 3 arterialized DVAs presenting with hemorrhage have been treated using various combinations of endovascular embolization, in particular when an AVM nidus was demonstrated, surgical resection, and Gamma knife surgery [4, 36, 40, 52, 53]. Some authors recommend conservative management.

Conclusions

Cerebral DVAs are frequently encountered during routine imaging studies of the brain. They are currently

considered extreme variations of the cerebral venous angioarchitecture that puts them at risk of developing venous hypertension. In the vast majority of cases, however, DVAs follow a benign clinical course and do not require follow-up imaging studies or specific medical management. Their frequent association with other vascular malformations, in particular with CMs, usually accounts for cases presenting with cerebral hemorrhage or seizure activity. On rare occasions, a DVA itself may be responsible for neurological complications secondary to thrombosis of its collecting system, or to mass effect exerted by a dilated collecting vein. Thrombosed DVAs should be managed as cortical or dural venous sinus thrombosis. In cases with cerebral hematoma or edema requiring surgical management, particular care must be taken to preserve the collecting vein of the DVA in order to avoid catastrophic venous infarction. There is growing awareness of rare and atypical forms of arterialized DVAs that generally follow an aggressive clinical course and likely carry a hemorrhagic risk similar to typical AVMs. Three types of arterialized DVAs have been defined based on the angiographic demonstration of abnormal arterial supply to the DVA or an associated AVM that drains into the DVA system. DSA remains required in all cases of DVAs presenting with a cerebral hemorrhage in which CT or MRI has failed to demonstrate a collecting vein thrombosis or a cavernous malformation. The management of arterialized DVAs is not yet clearly established in the literature due to the small number of reported cases.

References

1. Abarca-Olivas J, Botella-Asuncion C, Concepcion-Aramendia LA, Cortes-Vela JJ, Gallego-Leon JI, Ballenilla-Marco F (2009) Two cases of brain haemorrhage secondary to developmental venous anomaly thrombosis. Bibliographic review. Neurocirugia (Astur) 20:265–271
2. Abe M, Hagihara N, Tabuchi K, Uchino A, Miyasaka Y (2003) Histologically classified venous angiomas of the brain: a controversy. Neurol Med Chir (Tokyo) 43:1–10, discussion 11
3. Aboian MS, Daniels DJ, Rammos SK, Pozzati E, Lanzino G (2009) The putative role of the venous system in the genesis of vascular malformations. Neurosurg Focus 27:E9
4. Aksoy FG, Gomori JM, Tuchner Z (2000) Association of intracerebral venous angioma and true arteriovenous malformation: a rare, distinct entity. Neuroradiology 42:455–457
5. Amemiya S, Aoki S, Takao H (2008) Venous congestion associated with developmental venous anomaly: findings on susceptibility weighted imaging. J Magn Reson Imaging 28:1506–1509
6. Augustyn GT, Scott JA, Olson E, Gilmor RL, Edwards MK (1985) Cerebral venous angiomas: MR imaging. Radiology 156:391–395
7. Awad IA, Robinson JR Jr, Mohanty S, Estes ML (1993) Mixed vascular malformations of the brain: clinical and pathogenetic considerations. Neurosurgery 33:179–188, discussion 188
8. Barkovich AJ (1988) Abnormal vascular drainage in anomalies of neuronal migration. AJNR Am J Neuroradiol 9:939–942
9. Berbel-Garcia A, Martinez-Salio A, Porta-Etessam J, Saiz-Diaz R, Gonzalez-Leon P, Ramos A, Campollo J (2004) Venous angioma associated with atypical ophthalmoplegic migraine. Headache 44:440–442
10. Bisdorff A, Mulliken JB, Carrico J, Robertson RL, Burrows PE (2007) Intracranial vascular anomalies in patients with periorbital lymphatic and lymphaticovenous malformations. AJNR Am J Neuroradiol 28:335–341
11. Blackmore CC, Mamourian AC (1996) Aqueduct compression from venous angioma: MR findings. AJNR Am J Neuroradiol 17:458–460
12. Bouchacourt E, Carpena JP, Bories J, Koussa A, Chiras J (1986) Ischemic accident caused by thrombosis of a venous angioma. Apropos of a case. J Radiol 67:631–635
13. Boukobza M, Enjolras O, Guichard JP, Gelbert F, Herbreteau D, Reizine D, Merland JJ (1996) Cerebral developmental venous anomalies associated with head and neck venous malformations. AJNR Am J Neuroradiol 17:987–994
14. Burke L, Berenberg RA, Kim KS (1984) Choreoballismus: a nonhemorrhagic complication of venous angiomas. Surg Neurol 21:245–248
15. Cakirer S (2003) De novo formation of a cavernous malformation of the brain in the presence of a developmental venous anomaly. Clin Radiol 58:251–256
16. Campeau NG, Lane JI (2005) De novo development of a lesion with the appearance of a cavernous malformation adjacent to an existing developmental venous anomaly. AJNR Am J Neuroradiol 26:156–159
17. Courville CB (1963) Morphology of small vascular malformations of the brain. With particular reference to the mechanism of their drainage. J Neuropathol Exp Neurol 22:274–284
18. Desai K, Bhayani R, Nadkarni T, Limaye U, Goel A (2002) Developmental deep venous system anomaly associated with congenital malformation of the brain. Pediatr Neurosurg 36:37–39
19. Dillon WP (1997) Cryptic vascular malformations: controversies in terminology, diagnosis, pathophysiology, and treatment. AJNR Am J Neuroradiol 18:1839–1846
20. Ferro JM, Canhao P (2008) Acute treatment of cerebral venous and dural sinus thrombosis. Curr Treat Options Neurol 10:126–137
21. Gabikian P, Clatterbuck RE, Gailloud P, Rigamonti D (2003) Developmental venous anomalies and sinus pericranii in the blue rubber-bleb nevus syndrome. Case report. J Neurosurg 99:409–411
22. Gama RL, Nakayama M, Tavora DG, Bomfim RC, Carneiro TC, Pimentel LH (2008) Thrombosed developmental venous anomaly associated with cerebral venous infarct. Arq Neuropsiquiatr 66:560–562
23. Gandolfo C, Krings T, Alvarez H, Ozanne A, Schaaf M, Baccin CE, Zhao WY, Lasjaunias P (2007) Sinus pericranii: diagnostic and therapeutic considerations in 15 patients. Neuroradiology 49:505–514
24. Garner TB, Del Curling O Jr, Kelly DL Jr, Laster DW (1991) The natural history of intracranial venous angiomas. J Neurosurg 75:715–722
25. Guerrero AL, Blanco A, Arcaya J, Cacho J (1998) Venous infarct as presenting form of venous angioma of the posterior fossa. Rev Clín Esp 198:484–485
26. Hammoud D, Beauchamp N, Wityk R, Yousem D (2002) Ischemic complication of a cerebral developmental venous anomaly: case report and review of the literature. J Comput Assist Tomogr 26:633–636
27. Herbreteau O, Auffray-Calvier E, Desal H, Freund P, De Kersaint-Gilly A (1999) Symptomatic venous angioma. Report of a case. J Neuroradiol 26:126–131

28. Hirata Y, Matsukado Y, Nagahiro S, Kuratsu J (1986) Intracerebral venous angioma with arterial blood supply: a mixed angioma. Surg Neurol 25:227–232
29. Hong YJ, Chung TS, Suh SH, Park CH, Tomar G, Seo KD, Kim KS, Park IK (2010) The angioarchitectural factors of the cerebral developmental venous anomaly; can they be the causes of concurrent sporadic cavernous malformation? Neuroradiology. doi:10.1007/s00234.009-0640-6
30. Huber G, Henkes H, Hermes M, Felber S, Terstegge K, Piepgras U (1996) Regional association of developmental venous anomalies with angiographically occult vascular malformations. Eur Radiol 6:30–37
31. Im SH, Han MH, Kwon BJ, Ahn JY, Jung C, Park SH, Oh CW, Han DH (2008) Venous-predominant parenchymal arteriovenous malformation: a rare subtype with a venous drainage pattern mimicking developmental venous anomaly. J Neurosurg 108:1142–1147
32. Kapp JP, Schmidek HH (1984) The cerebral venous system and its disorders. Grune & Stratton, Orlando
33. Kim P, Castellani R, Tresser N (1996) Cerebral venous malformation complicated by spontaneous thrombosis. Childs Nerv Syst 12:172–175
34. Konan AV, Raymond J, Bourgouin P, Lesage J, Milot G, Roy D (1999) Cerebellar infarct caused by spontaneous thrombosis of a developmental venous anomaly of the posterior fossa. AJNR Am J Neuroradiol 20:256–258
35. Kondziolka D, Lunsford LD, Kestle JR (1995) The natural history of cerebral cavernous malformations. J Neurosurg 83:820–824
36. Kurita H, Sasaki T, Tago M, Kaneko Y, Kirino T (1999) Successful radiosurgical treatment of arteriovenous malformation accompanied by venous malformation. AJNR Am J Neuroradiol 20:482–485
37. Lai PH, Chen PC, Pan HB, Yang CF (1999) Venous infarction from a venous angioma occurring after thrombosis of a drainage vein. AJR Am J Roentgenol 172:1698–1699
38. Lasjaunias P, Burrows P, Planet C (1986) Developmental venous anomalies (DVA): the so-called venous angioma. Neurosurg Rev 9:233–242
39. Lee C, Pennington MA, Kenney CM 3rd (1996) MR evaluation of developmental venous anomalies: medullary venous anatomy of venous angiomas. AJNR Am J Neuroradiol 17:61–70
40. Lindquist C, Guo WY, Karlsson B, Steiner L (1993) Radiosurgery for venous angiomas. J Neurosurg 78:531–536
41. Lovrencic-Huzjan A, Rumboldt Z, Marotti M, Demarin V (2004) Subarachnoid haemorrhage headache from a developmental venous anomaly. Cephalalgia 24:763–766
42. Maeder P, Gudinchet F, Meuli R, de Tribolet N (1998) Development of a cavernous malformation of the brain. AJNR Am J Neuroradiol 19:1141–1143
43. Malinvaud D, Lecanu JB, Halimi P, Avan P, Bonfils P (2006) Tinnitus and cerebellar developmental venous anomaly. Arch Otolaryngol Head Neck Surg 132:550–553
44. Matsuda H, Terada T, Katoh M, Ishida S, Onuma T, Nakano H, Yagishita A (1994) Brain perfusion SPECT in a patient with a subtle venous angioma. Clin Nucl Med 19:785–788
45. McCormick WF, Hardman JM, Boulter TR (1968) Vascular malformations ("angiomas") of the brain, with special reference to those occurring in the posterior fossa. J Neurosurg 28:241–251
46. McCormick WF, Schochet SS (1976) Atlas of cerebrovascular disease. Saunders, Philadelphia
47. McLaughlin MR, Kondziolka D, Flickinger JC, Lunsford S, Lunsford LD (1998) The prospective natural history of cerebral venous malformations. Neurosurgery 43:195–200, discussion 200-191
48. Merten CL, Knitelius HO, Hedde JP, Assheuer J, Bewermeyer H (1998) Intracerebral haemorrhage from a venous angioma following thrombosis of a draining vein. Neuroradiology 40:15–18
49. Moriarity JL, Wetzel M, Clatterbuck RE, Javedan S, Sheppard JM, Hoenig-Rigamonti K, Crone NE, Breiter SN, Lee RR, Rigamonti D (1999) The natural history of cavernous malformations: a prospective study of 68 patients. Neurosurgery 44:1166–1171, discussion 1172-1163
50. Morioka T, Hashiguchi K, Nagata S, Miyagi Y, Yoshida F, Mihara F, Sakata A, Sasaki T (2006) Epileptogenicity of supratentorial medullary venous malformation. Epilepsia 47:365–370
51. Noran H (1945) Intracranial vascular tumors and malformations. Arch Pathol 39:393–416
52. Nussbaum ES, Heros RC, Madison MT, Awasthi D, Truwit CL (1998) The pathogenesis of arteriovenous malformations: insights provided by a case of multiple arteriovenous malformations developing in relation to a developmental venous anomaly. Neurosurgery 43:347–351, discussion 351-342
53. Oran I, Kiroglu Y, Yurt A, Ozer FD, Acar F, Dalbasti T, Yagci B, Sirikci A, Calli C (2008) Developmental venous anomaly (DVA) with arterial component: a rare cause of intracranial haemorrhage. Neuroradiology 51:25–32
54. Peltier J, Toussaint P, Desenclos C, Le Gars D, Deramond H (2004) Cerebral venous angioma of the pons complicated by nonhemorrhagic infarction. Case report. J Neurosurg 101:690–693
55. GS PVM, Krings T, Aurboonyawat T, Ozanne A, Toulgoat F, Pongpech S, Lausjaunias PL (2008) Pathomechanics of symptomatic developmental venous anomalies. Stroke 39:3201–3215
56. Peterson AM, Williams RL, Fukui MB, Meltzer CC (2002) Venous angioma adjacent to the root entry zone of the trigeminal nerve: implications for management of trigeminal neuralgia. Neuroradiology 44:342–346
57. Rigamonti D, Spetzler RF (1988) The association of venous and cavernous malformations. Report of four cases and discussion of the pathophysiological, diagnostic, and therapeutic implications. Acta Neurochir (Wien) 92:100–105
58. Rigamonti D, Spetzler RF, Drayer BP, Bojanowski WM, Hodak J, Rigamonti KH, Plenge K, Powers M, Rekate H (1988) Appearance of venous malformations on magnetic resonance imaging. J Neurosurg 69:535–539
59. Rothbart D, Awad IA, Lee J, Kim J, Harbaugh R, Criscuolo GR (1996) Expression of angiogenic factors and structural proteins in central nervous system vascular malformations. Neurosurgery 38:915–924, discussion 924-915
60. Rothfus WE, Albright AL, Casey KF, Latchaw RE, Roppolo HM (1984) Cerebellar venous angioma: "benign" entity? AJNR Am J Neuroradiol 5:61–66
61. Saito Y, Kobayashi N (1981) Cerebral venous angiomas: clinical evaluation and possible etiology. Radiology 139:87–94
62. San Millan Ruiz D, Delavelle J, Yilmaz H, Gailloud P, Piovan E, Bertramello A, Pizzini F, Rufenacht DA (2007) Parenchymal abnormalities associated with developmental venous anomalies. Neuroradiology 49:987–995
63. San Millan Ruiz D, Yilmaz H, Gailloud P (2009) Cerebral developmental venous anomalies: current concepts. Ann Neurol 66:271–283
64. San Millan Ruiz D, Gandhi D, Levrier O (2010) Venous anomaly. J Neurosurg 112:213–214, author reply 214
65. Sarwar M, McCormick WF (1978) Intracerebral venous angioma. Case report and review. Arch Neurol 35:323–325
66. Schaller B, Graf R (2004) Cerebral venous infarction: the pathophysiological concept. Cerebrovasc Dis 18:179–188
67. Sehgal V, Delproposto Z, Haacke EM, Tong KA, Wycliffe N, Kido DK, Xu Y, Neelavalli J, Haddar D, Reichenbach JR (2005) Clinical applications of neuroimaging with susceptibility-weighted imaging. J Magn Reson Imaging 22:439–450
68. Seki Y, Sahara Y (2007) Spontaneous thrombosis of a venous malformation leading to intracerebral hemorrhage—case report. Neurol Med Chir (Tokyo) 47:310–313
69. Senegor M, Dohrmann GJ, Wollmann RL (1983) Venous angiomas of the posterior fossa should be considered as anomalous venous drainage. Surg Neurol 19:26–32

70. Striano S, Nocerino C, Striano P, Boccella P, Meo R, Bilo L, Cirillo S (2000) Venous angiomas and epilepsy. Neurol Sci 21:151–155
71. Thobois S, Nighoghossian N, Mazoyer JF, Honnorat J, Derex L, Froment JC, Trouillas P (1999) Cortical thrombophlebitis and developmental venous anomalies. Rev Neurol (Paris) 155:48–50
72. Tomura N, Inugami A, Uemura K, Hadeishi H, Yasui N (1991) Multiple medullary venous malformations decreasing cerebral blood flow: case report. Surg Neurol 35:131–135
73. Topper R, Jurgens E, Reul J, Thron A (1999) Clinical significance of intracranial developmental venous anomalies. J Neurol Neurosurg Psychiatry 67:234–238
74. Truwit CL (1992) Venous angioma of the brain: history, significance, and imaging findings. AJR Am J Roentgenol 159:1299–1307
75. Uchino A, Hasuo K, Matsumoto S, Masuda K (1995) Double cerebral venous angiomas: MRI. Neuroradiology 37:25–28
76. Vieira Santos A, Saraiva P (2006) Spontaneous isolated non-haemorrhagic thrombosis in a child with development venous anomaly: case report and review of the literature. Childs Nerv Syst 22:1631–1633
77. Walsh M, Parmar H, Mukherji SK, Mamourian A (2008) Developmental venous anomaly with symptomatic thrombosis of the draining vein. J Neurosurg 109:1119–1122
78. Wendling LR, Moore JS Jr, Kieffer SA, Goldberg HI, Latchaw RE (1976) Intracerebral venous angioma. Radiology 119:141–147
79. Wilms G, Demaerel P, Marchal G, Baert AL, Plets C (1991) Gadolinium-enhanced MR imaging of cerebral venous angiomas with emphasis on their drainage. J Comput Assist Tomogr 15:199–206
80. Wilson CB (1992) Cryptic vascular malformations. Clin Neurosurg 38:49–84
81. Wurm G, Schnizer M, Fellner FA (2005) Cerebral cavernous malformations associated with venous anomalies: surgical considerations. Neurosurgery 57:42–58, discussion 42–58
82. Yamamoto M, Inagawa T, Kamiya K, Ogasawara H, Monden S, Yano T (1989) Intracerebral hemorrhage due to venous thrombosis in venous angioma—case report. Neurol Med Chir (Tokyo) 29:1044–1046