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Parenchymal abnormalities associated with developmental venous anomalies

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Abstract
Introduction To report a retrospective series of 84 cerebral developmental venous anomalies (DVAs), focusing on associated parenchymal abnormalities within the drainage territory of the DVA.
Methods DVAs were identified during routine diagnostic radiological work-up based on magnetic resonance imaging (MRI) (60 cases), computed tomography (CT) (62 cases) or both (36 cases). Regional parenchymal modifications within the drainage territory of the DVA, such as cortical or subcortical atrophy, white matter density or signal alterations, dystrophic calcifications, presence of haemorrhage or a cavernous-like vascular malformation (CVM), were noted. A stenosis of the collecting vein of the DVA was also sought for.
Results Brain abnormalities within the drainage territory of a DVA were encountered in 65.4% of the cases. Locoregional brain atrophy occurred in 29.7% of the cases, followed by white matter lesions in 28.3% of MRI investigations and 19.3% of CT investigations, CVMs in 13.3% of MRI investigations and dystrophic calcification in 9.6% of CT investigations. An intracranial haemorrhage possibly related to a DVA occured in 2.4% cases, and a stenosis on the collecting vein was documented in 13.1% of cases. Parenchymal abnormalities were identified for all DVA sizes.
Conclusion Brain parenchymal abnormalities were associated with DVAs in close to two thirds of the cases evaluated. These abnormalities are thought to occur secondarily, likely during post-natal life, as a result of chronic venous hypertension. Outflow obstruction, progressive thickening of the walls of the DVA and their morphological organization into a venous convergence zone are thought to contribute to the development of venous hypertension in DVA.

Keywords Developmental venous anomaly · Regional parenchymal brain abnormalities · Venous hypertension

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Introduction

Cerebral developmental venous anomalies (DVAs), also referred to as cerebral venous angiomas or cerebral medullary venous malformations, are the most frequent vascular malformation of the brain [1–3]. Some specialists interpret them as extreme anatomical variants of the medullary venous system [4], whilst others view them as an abnormality resulting from disturbed angiogenesis and

regression [5–7]. Whatever their origin, it is generally accepted that they are congenital, generally follow a benign and clinically asymptomatic course [2, 8–10] and are typically diagnosed incidentally during imaging investigations for symptoms unrelated to the DVA. DVAs are a purely venous entity with no arteriovenous shunt and drain normally functioning brain parenchyma. Their essential role in normal cerebral venous drainage is illustrated by cases of catastrophic venous ischemic and haemorrhagic complications resulting from the surgical removal of a DVA [11].

Although the parenchyma drained by a DVA is generally reported as normal brain tissue, there have been reports of parenchymal abnormalities in the vicinity of a DVA. The most frequently encountered association is that of a sporadic cavernous venous malformation (CVM). This association is found in up to 40% of DVAs depending on the published series [12, 13] and generally explains presenting neurological symptoms if these concord with the location of a DVA and the CVM [6, 14, 15]. Other reports have mentioned the existence of parenchymal abnormalities, including signal modifications of the white matter on magnetic resonance imaging (MRI) scans, dystrophic calcification and locoregional brain atrophy [6, 12, 16], although no clear information as to the frequency or significance of these findings was forwarded.

The purpose of this study was to evaluate the type and frequency of brain abnormalities found within the drainage territory of the DVA based on computed tomography (CT) and MRI imaging and to discuss their possible pathogenic mechanism.

Methods

This study is based on the retrospective analysis of 84 consecutive DVAs in 83 patients, diagnosed over a 3-year period by routine CT and MRI diagnostic procedures. This study complied with the requirements of the Ethics Committee of our Institution. Sixty patients underwent MRI, 62 patients underwent CT, 36 underwent both imaging techniques.

CT was performed on 16-channel multirow-detector scanner (model MX 8000 IDT; Philips Medical Systems, Best, The Netherlands). Non-enhanced 3-mm-thick images were obtained from C1 to the vertex (index: 0.7 mm; EB filter) on all patients. Some patients also underwent high-resolution CT angiography (CTA) (12 out of 62) or CT venography (CTV) (4 out of 62). CTA was obtained at the arterial phase with bolus tracking after systemic bolus injection of 120 ml of contrast product (4 ml/s), with 1.5-mm-thick slices (index: 0.7 mm; pitch: 0.663; UB filter) acquired from the ascending aorta to the vertex. CTV was performed without bolus tracking, with acquisition starting 50 s after the bolus injection at the vertex and covering the head and cervical region up the level of C4. A late venous phase acquisition following the non-contrast acquisition protocol was obtained 2–3 min after contrast injection in all patients, whether they underwent CTA/CTV or not.

MRI was performed on a 1.5-Tesla scanner (Intera, Philips Medical Systems). For all patients, we obtained the following images: standard fast-spin echo T2-weighted [T2-W; TR 4450, TE 100, Flip 90°, thickness: 5 mm, gap: 1 mm, Field of view (FOV): 220 mm, matrix: 384/512]; pre- and post-contrast (Magnevist, Shering, Switzerland) spin-echo (Gadovist) T1-weighted (T1-W and Gd T1-W; TR 420, TE 10, Flip 90°, thickness: 5 mm, gap: 1 mm, FOV: 240 mm, matrix: 304/512), diffusion-weighted (DWI; TR 3562, TE 95, thickness: 6 mm, gap: 1 mm, FOV: 240 mm, matrix: 128/256); fluid-attenuated inversion recovery (FLAIR; TR 11000, TE 140/IR2800, Flip 90°, thickness: 5 mm, gap: 0.5 mm, FOV: 230 mm, matrix 256/432). Post-contrast (Gadovist) injected three-dimensional (3D) T1-weighted (Gd 3D T1-W; TR 25, TE 4.7, Flip 30°, thickness: 1 mm, gap: 0 mm, FOV: 240 mm, matrix: 240/256) and 3D time-of-flight angiography (Gd 3D TOF; 3D acquisition mode, TR 22, TE 9.2, Flip 25°, thickness: 0.6 mm, gap: 0 mm, 1 chunk, FOV: 170 mm, matrix: 352/512) images were obtained for 12 and nine patients out of 60 respectively.

On post-contrast CT, a DVA was identified on source or reconstructed images as a superficial or deeply seated typical caput medusa draining into the deep or superficial venous system by way of an enlarged collecting vein. On MRI, recognition of a DVA was based on GD-T1 sequences demonstrating the same morphological characteristics of DVAs as CT.

CT and MRI images were analysed for regional parenchymal abnormalities within the drainage territory of the DVA that were thought to be related with the DVA, such as cortical or subcortical atrophy, white matter density or signal alterations, dystrophic calcifications, presence of haemorrhage or a CVM. It was not the purpose of this study to evaluate parenchymal lesions outside of the DVA's drainage territory.

Stenosis of the collecting vein of the DVA was sought in images that allowed appreciation of the collecting vein, such as CTA/CTV, Gd 3D T1-W and Gd 3D TOF sequences. The locations of the DVAs were noted. A stenosis was identified on the basis of a dilatation of the collecting vein proximal to a zone of marked narrowing, usually at or close to the point of dural passage towards a dural venous sinus. DVAs were classified comparatively as small, medium, and large sized in relation to the smallest and largest DVA in the series; a quantitative analysis of the size was not undertaken due to the inherent difficulty in calculating the volume of a DVA and the inhomogeneity of the imaging protocols. Analysis of the data was performed conjointly by two trained neuroradiologists (DSMR, JD).

Results

The average age (±SD) of the patients was 50±19.9 years (range: 11 months–89 years) and 52.4% of the study cohort were males. Of the 84 DVAs that were found, 80 (95.2%) were fortuitous findings unrelated to presenting neurological symptoms. DVA location distribution is reported in Table 1. One patient harboured both a right cerebellar DVA and a left vermian DVA, which drained into the right superior petrosal sinus and the straight sinus, respectively.

DVA size

Of the 84 DVAs identified, 40 (47.6%) were assessed as medium sized, 32 (38.1%) as small and 12 (14.3%) as large. Five of the 12 large DVAs were complex with two or more large confluent collecting veins.

Parenchymal abnormalities

A total of 55 of the 84 DVAs (65.4%) harboured a coexisting locoregional parenchymal abnormality in the form of atrophy, density or signal abnormality, calcification, a CVM or a combination of these findings.

Loco-regional atrophy

Loco-regional brain atrophy was the most frequent parenchymal abnormality encountered within the drainage territory of a DVA, being observed in 29.7% of the cases. Brain atrophy was encountered for all DVA locations, with the exception of the mesencephalon, and for all DVA sizes. Atrophy was found to be either cortical, subcortical or a combination of both (Fig. 1). The extension of the atrophy was variable, ranging from focal enlargement of a sulcus or a ventricle to atrophy involving the entire drainage territory of the DVA.

White matter abnormalities on MRI and CT

Subcortical or deep white matter abnormalities within the drainage territory of a DVA were more frequently observed on MR images (28.3%) than on CT (19.3%) images. On CT, white matter abnormalities consisted of well-circumscribed, non-enhancing hypodensities centred by the caput medusa of the DVA (Figs. 2 and 3). On MR imaging, white matter abnormalities consisted of a non-enhancing hyperintense signal on T2-W and FLAIR images and a hypointense signal on T1-W images, with no diffusion restriction on the DWI and no mass effect exertion on the surrounding parenchyma (Fig. 3). The extension of the signal modifications ranged from a small, well circumscribed hyperintense area on T2-W and FLAIR images around the caput medusa to a large volume of brain parenchyma corresponding to the entire drainage territory of the DVA, visible both on T2-W and T1-W images. White matter abnormalities occurred with DVAs of all sizes but were only found in the supratentorial white matter. The lesions were stable over a period of 3 years in one patient investigated by CT and for over 6 months in one patient undergoing both CT and MR imaging. The remaining patients did not have any followup imaging performed.

Cavernous venous malformation

Lesions compatible with a CVM on MR imaging were found abutting the DVA in eight out of 60 cases (13.3%). In one case, a CVM was suspected on the CT image, but MR confirmation could not be obtained because the patient carried a pacemaker. CVMs were solitary lesions, with the exception of one patient harboring five CVMs in the right cingular gyrus and corpus callosum. CVMs were located in the frontal lobe (two cases), in the parietal lobe (one case), in the lateral temporal lobe (one case), in the basal ganglia (one case), in the cerebellum (one case) and in the mesencephalon (two cases). CVMs were usually asymptomatic, being discovered incidentally in all but two patients. The first patient, a 40-year-old man, had seizures from a right frontal CVM associated with a small DVA. In the second case, a 67-year-old woman presented with transient loss of consciousness attributed to a right basal ganglia CVM adjacent to a medium-sized DVA. This CVM was surrounded by vasogenic edema, suggestive of acute thrombosis associated with haemorrhage within the CVM. The patient had an uneventful recovery following this acute episode.

Acute intracranial hemorrhage

Two cases presented with intracranial haemorrhage possibly related to an underlying DVA. In the first case, a 40-year-old woman suffered from a perimesencephalic subarachnoid haemorrhage following a Valsalva manoever. The CTA disclosed a large, complex left cerebellar and vermian DVA consisting of two major collecting veins. Drainage

Table 1 Developmental venous anomalies (DVA) location distribution

Frontal	Cerebellar hemisphere	Temporal	Parietal	Insular	Basal ganglia	Vermian	Mesencephallon	Temporo-parietal
39	12	11	5	5	5	3	2	2

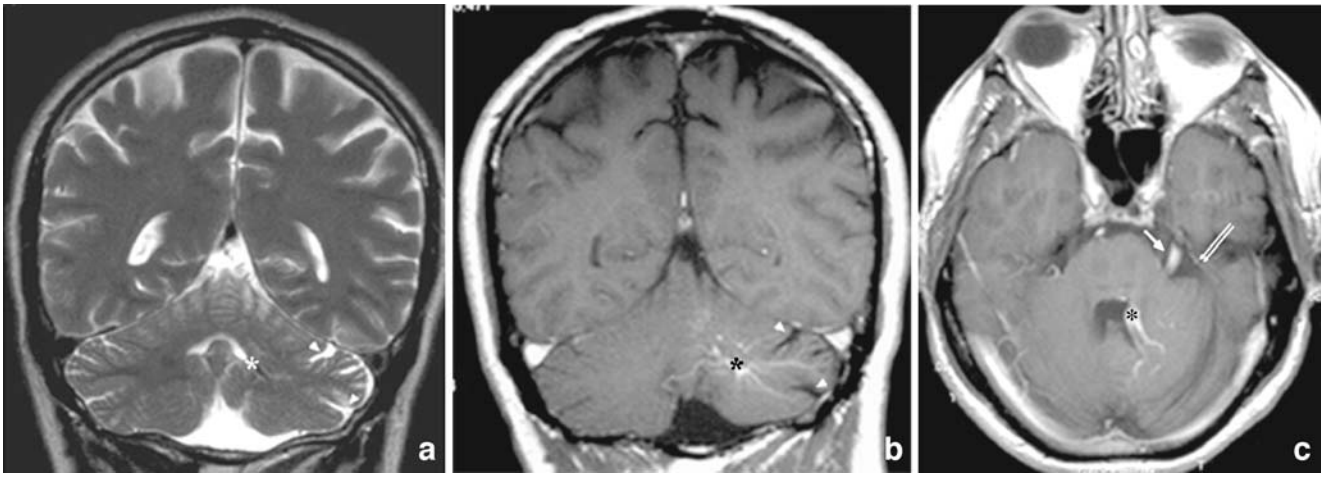

Fig. 1 Thirty-year-old male with multiple sclerosis. **a**, **b** Standard fast-spin echo T2-weighted (T2-W) and post-contrast Gadovist injected three-dimensional T1-weighted (Gd 3D T1-W) coronal views showing a large, left vermian and hemispheric cerebellar DVA (*asterisk*) with cortical cerebellar atrophy (*arrowheads*) within the drainage territory of the developmental venous anomaly (DVA). The DVA collecting vein appears as a flow void on the T2-W image. The left cerebral hemisphere was devoid of any demyelinating disease on the magnetic resonance imaging (MRI) scan and was the only region showing atrophy. **c** Gd 3D T1-W axial view. This DVA ultimately collected into the left superior petrosal vein (of Dandy) (*single arrow*). Note the difference in caliber between the superior petrosal vein and the superior petrosal sinus (*double arrows*) compatible with a stenosis at the point of dural passage of the vein into the sinus

occurred primarily into the left transverse sinus. The cranial component of the DVA was also drained by a precentral vein into the left basal vein and subsequently connected with the perimesencephalic veins by way of a lateral mesencephalic vein. There was a tight stenosis of the major collecting veins at the point of dural passage into the left transverse sinus. All imaging modalities failed to disclose other vascular lesions that could account for the subarachnoid haemorrhage. A CVM was also disclosed within the drainage territory of the DVA in the white matter of the left cerebellar hemisphere.

In the second case, a large temporal DVA draining into a lateral temporal vein was considered to have caused a temporal lobar haemorrhage in an 82-year-old man, in the absence of other identifiable underlying lesions at the 3- and 6-month follow-up with MR imaging. No thrombosis of the collecting vein of the DVA was disclosed in either case.

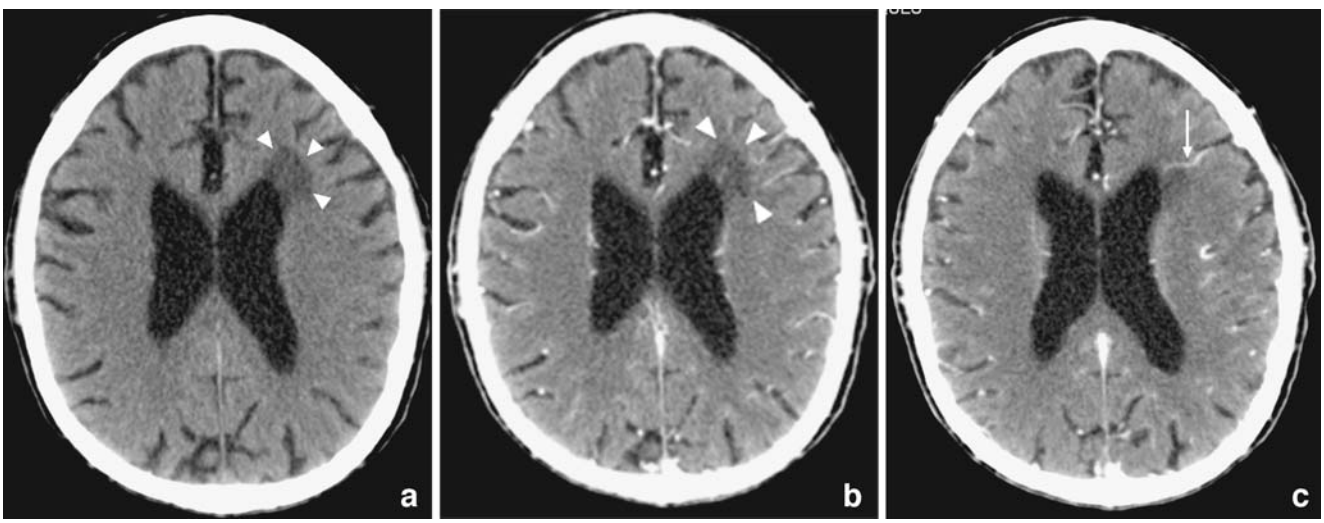

Fig. 2 A 40-year-old alcoholic male presenting with seizure. **a** Non-contrast computed tomography (CT) showing a left frontal horn, periventricular white matter hypodensity (*arrowheads*) with no mass effect. **b**, **c** Post-contrast CT showing a small underlying frontal DVA with superficial drainage (*white arrow*) whose medullary veins are found within the hypodense area demarcated by the *arrowheads* in **a** and **b**. This lesion was unchanged compared to that observed on a CT image performed 3 years prior. The MRI correlate of the white matter lesion (not shown) consisted of both T2 and T1 shortening, absence of diffusion restriction on the diffusion-weighted (DWI) scan, mass effect or enhancement. Furthermore, MRI did not disclose any other zones of white matter abnormalities

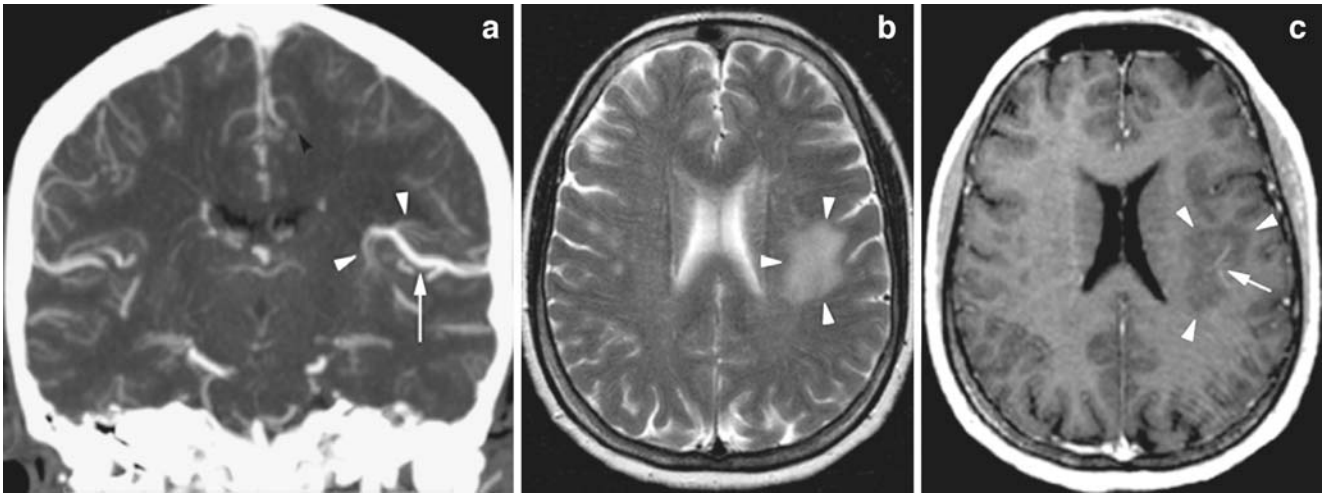

Fig. 3 A 56 year-old female presenting with a Guillain–Baré syndrome with bilateral 6th cranial nerve palsy. Brain MRI disclosed an incidental white mater lesion centred upon a small DVA with superficial drainage. **a** Coronal multiplanar reconstruction (MPR) of an a CT angiography (CTA), showing the left insular DVA with the typical caput medusa (*arrowheads*) centred around a collecting vein (*arrow*). **b** MR imaging, axial T2-W images, showing a hyperintense subcortical white matter lesion around the left insular region (*arrowheads*), without mass effect or regional atrophy. **c** MR imaging, Gd 3D T1-W axial images. There is no contrast uptake of the lesion, which appears hypointense and well-demarcated (*arrowheads*). This zone is centred by the caput medusa of the DVA (*arrow*). There was no diffusion restriction on the DWI image (not shown), and no stenosis is demonstrated on Gd 3D T1-W or Gd 3D TOF images. The lesion was unchanged at the 6-month follow-up

Dystrophic calcifications

DVA were associated to dystrophic parenchymal calcifications in 9.6% of CT cases (six out of 62). Dystrophic calcifications were found in the frontal white matter (three cases), cerebellar hemisphere (one case), caudate nucleus (one case), basal ganglia (one case; Fig. 4). Dystrophic calcifications occurred in DVAs regardless of their size. Dystrophic calcifications were not sought for in MRI as echo gradient T2-W images were not available for all patients.

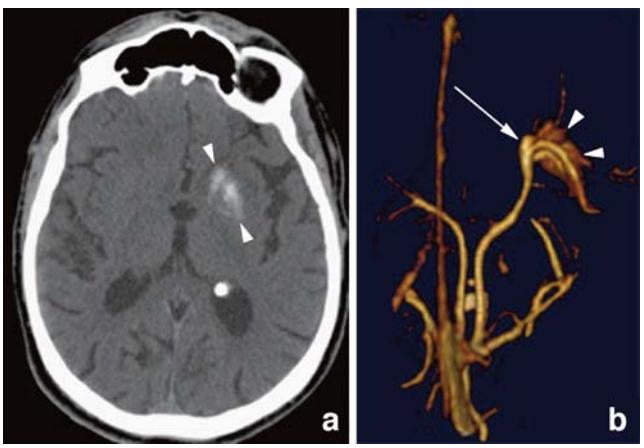

Fig. 4 An 83 year-old man presenting a right middle cerebral artery (MCA) infarction. **a** Non-injected CT showing dystrophic calcifications in the left basal ganglia (*arrowheads*). **b** 3D reconstruction of CTA acquisition (Vitrea workstation, Vital Images, Minnetonkam Minn.) showing a medium-sized DVA draining the left basal ganglia and internal capsule into the internal cerebral vein. Calcifications are visible between the venules forming the caput medusa (*arrowheads*). Note the ampullary dilatation (*arrow*) of the proximal portion of the DVA when compared to the more distal portion of the collecting vein. The left sylvian cistern looks larger than the right one, although this was due to an asymmetric acquisition rather than atrophy in this case

DVA stenosis

Stenosis of the collecting vein was observed in 11 cases (13.1%). However, this is likely to be an underestimation as only a small proportion of MRI studies included Gd 3D T1-W and 3D Gd TOF images (28.3%), and about one half of the CT studies included angiography sequences (45.1%). Furthermore, a stenosis at the point of dural passage of the collecting vein in cases of superficial DVA was difficult to disclose on CTA due to the partial volume effect with bone and beam-hardening artifacts. The presence of parenchymal abnormalities could, therefore, not be correlated to the presence of a stenosis in this study.

An ampullar dilatation of the proximal portion of the DVA, located at the convergence zone of all the medullary veins forming the caput medusa, was observed in 23 of 84 DVAs (27.3%). Of these 23 DVAs, 15 (65.3%) had an associated parenchymal abnormality, which was comparable to the 65.4% rate of parenchymal abnormalities for the entire DVA population studied. The significance of these ampullar dilatations is unknown in the absence of a histopathological confrontation.

Discussion

Some authors consider DVA to be normal anatomic variants of the cerebral medullary venous system [4, 7, 11], an

opinion that may be reasonable from a clinical perspective, given that the number of reported complications directly attributable to a DVA remains anecdotal. A few case reports have linked DVA with ischemic or haemorrhagic infarctions, or with focal neurological symptoms [2, 16–27]. Stenosis or thrombosis of the collecting vein of a DVA was reported in several of the cases presenting with haemorrhagic or non-haemorrhagic infarctions involving the drainage territory of a DVA. Consequently, increased venous pressure within the DVA territory has been suggested in complicated DVAs [18]. Impaired brain perfusion attributed to venous congestion was documented by several authors in areas drained both by small and large DVAs [28, 29]. Direct proof of increased venous pressure within a DVA was presented by Dillon [30], who measured a 15-mm-Hg pressure gradient across a stenosis of the collecting vein of a DVA in one patient. This patient had presented with recurrent episodes of brain haemorrhage in the drainage territory of the DVA which, although permeable, exhibited a tight stenosis of its collecting vein. It remains unknown, however, whether all DVAs develop venous hypertension (VHT) and whether factors other than draining vein stenosis may be involved.

This study focused on brain abnormalities found within the drainage territory of a DVA, which proved to be very frequent both on CT and MR imaging. Sixty-five percent of all DVAs harbored at least one coexisting parenchymal abnormality, excluding two cases presenting with acute intracranial haemorrhage. The most frequent finding was locoregional brain atrophy, which was observed in close to one third of the cases (29.7%), followed by white matter, lesions (28.3% on MRI, 19.3% on CT), CVMs (13.3% of MRI investigations) and dystrophic calcification (9.6% of CT investigations). Parenchymal abnormalities were equally observed in small-, medium-, and large-sized DVA, suggesting that DVA size is not a determining parameter for their development. In this study, DVAs were not associated with other cutaneous, mucosal or orbital vascular malformations. Only one of the 83 patients (1.2%) harboured two coexisting DVAs, a lower incidence than the 12% reported by Uchino et al. [31].

Outflow obstruction by a collecting vein stenosis was observed in 13.1% of the DVAs. However, this likely represents an underestimation for two reasons: first, not all cases benefitted from imaging protocols that would disclose a stenosis, such as Gd-3D TOF, Gd- 3D T1-W MRI, or CTA or CTV; second, it has been previously demonstrated that the rate of stenosis is underestimated if DSA is not performed [30]. A correlation between the presence of parenchymal abnormalities and a collecting vein stenosis was, therefore, not shown in this study. Such a relation is, however, strongly suspected based on the observation by Dillon [30] of an outflow obstruction in most DVAs with coexisting cryptic vascular malformation.

In all but four cases, DVAs were considered to be incidental findings unrelated to the presenting symptoms. One patient presented with seizures related to a coexisting frontal CVM, a second patient lost consciousness due to an acute haemorrhage within a pre-existing CVM in the basal ganglia and the two other patients presented with perimesencephalic haemorrhage and temporal lobe haemorrhage, speculatively related to an underlying DVA.

Locoregional atrophy around a DVA included cortical, subcortical or cortico-subcortical atrophy. Atrophy was found in all DVA locations except the mesencephalon. A few reports have mentioned the association between brain atrophy and DVA. Saito and Kobayashi [7] described one case of six DVAs in which the third ventricle and the right lateral ventricle were slightly enlarged and displaced towards a DVA that drained the right thalamus, caudate nucleus and putamen. Uchino et al. [16] reported a case of cerebral hemiatrophy related to a complex drainage abnormality assimilated to multiple right hemispheric DVAs; no stenosis was visible in any of the collecting veins in this case. Huber et al. [12] described the presence of focal brain atrophy in ten of 17 cases where DVAs and CVMs coexisted. Finally, Noran [32], based on pathology studies, mentioned that "venous angiomas (DVA)...not uncommonly causes definite atrophy of the region...", although he did not report how frequently atrophy was encountered or whether an outflow obstruction had been encountered.

White matter lesions were more frequently observed on MR imaging than CT, in keeping with the better sensitivity of MRI in detecting white matter lesions. White matter lesions were found to involve both the superficial and deep supratentorial white matter, and were stable over time in those cases where follow-up was available. No cerebellar white matter lesions were observed, though the significance of this observation is limited by low numbers of posterior fossa cases studies. These lesions showed no contrast uptake, diffusion restriction, or mass effect, and behaved similarly to leukoaraiosis both on CT and MR imaging. Reports of white matter abnormalities related to DVA are rare. Again, Uchino et al [16] described white matter T2 hyperintensities in the case of cerebral hemiatrophy and hemispheric DVA cited above. Truwit [2], in his pictorial essay on venous angioma, illustrated a case of white matter abnormalities around a frontal DVA similar to the ones presented here. Dillon [30] illustrated a right-sided centrum semiovale lesion with partial calcification around a relatively large DVA. Interestingly, the patient described by Dillon suffered from slowly progressive left-sided hemiparesis. None of the cases in our series with white matter lesions, however, had any neurological signs attributable to these lesions. Augustyn et al. [33] reported white matter abnormalities adjacent to DVA in up to 57% of their cases,

however, only seven DVA were studied in their report, limiting the statistical significance of their findings.

The significance of the white matter lesions is difficult to ascertain in the absence of histopathological correlation. Such lesions may be assimilated to vascular-induced leukoaraiosis, resulting from chronic cerebral ischemia consequent to VHT, rather than from acute venous infarction. Several findings substantiate this assumption. The histopathology correlate of leukoaraiosis includes a spectrum of lesions that range from edema on one end, and demyelination, loss of oligodendrocytes and gliosis on the other [34]. Though the parenchyma drained by DVAs is generally considered to be normal, lesions similar to those of leukoaraiosis were reported on histology by Noran [32], who observed zones of "demyelination, degenerative alterations of nerve cells, gliosis and leukomalacia..." around DVAs. Such histological lesions would behave similarly to leukoaraiosis on CT and MRI, and could well correspond to the lesions that were observed in the present study.

Further support may be found in the outstanding work of Moody et al. [35], who identified a new type of cerebral vascular pathology causing leukoaraiosis and involving the deep venous system, which they named periventricular venous collagenosis (PVC). Moody et al. [35] observed progressive parietal thickening of the periventricular and subependymal veins draining the deep white matter, resulting in luminal narrowing and ultimately leading to occlusion of a large proportion of these vessels. The authors further correlated PVC with the presence and degree of importance of radiologically and pathologically proven leukoaraiosis in elderly individuals devoid of small arterial vessel disease. They concluded that narrowing or occlusion of the veins draining the deep white matter caused an outflow obstruction leading to chronic cerebral edema and ischemia.

Several parallels may be traced between DVAs and deep cerebral veins affected by PVC, suggesting common pathophysiological pathways. On ultrastructural studies, DVA have been shown to have thickened and hyalinized walls [1, 36, 37], similar to PVC. Parietal thickening could reduce vessel lumen size and compliance, increase resistance to flow and diminish the vessel's capacity to adapt to pressure modifications. In addition, DVAs represent a point of venous confluence, where a single collecting vessel drains an abnormally large venous territory, resulting in a relative volume overload. This anatomic configuration is also encountered, to a lesser extent, in the subependymal veins affected by PVC, which are disposed as a venous convergence zone around the lateral and superior aspects of the lateral ventricles [5]. This pattern is not shared with cortical veins which, interestingly enough, do not become significantly thickened with age [35]. Outflow obstruction in the form of stenosis, loss of compliance due to parietal thickening and an anatomic configuration favouring a relative volume overload likely predispose DVAs to developing VHT and the subsequent atrophy and white matter lesions observed in this study. The limited capacities of DVA to adapt to sudden pressure changes in a condition of underlying VHT is emphasized by cases where a sudden increase in intracranial venous pressure following a Valsalva maneuver resulted in haemorrhagic complications, as observed in one of our cases and also reported by other authors [8, 19]. Smaller repeated parenchymal haemorrhages could alternatively trigger the cascade of events leading to the formation of a CVM, as has been suggested by several authors [7, 30].

The extent of brain atrophy and white matter lesions observed in our study was variable. Atrophy could be circumscribed to a sulcus or a portion of a ventricle, or involve the entire parenchyma drained by the DVA. Similarly, white matter lesions could involve a small zone of periventricular white matter, or include all of the white matter drained by the DVA. In all cases, including those with dystrophic calcifications and CVMs, the lesions were confined to the caput medusa, which consists of the smaller venular component of the DVA. This is in accordance with observations made by Ciricillo et al. [38] and Dillon [30], who found CVMs around the venous radicles of the caput medusa. That only parts of the drainage territory of a DVA show parenchymal abnormalities suggests either that venous hypertension does not necessarily involve the whole of the DVA system or that some zones of the DVA, such as the small venous radicles, are more sensitive to VHT.

The significance of the calcifications is also difficult to ascertain in the absence of histological correlation. McCormick, in a histology study, found calcifications in venous malformations consisting of seven arteriovenous malformations, four CVMs and one DVA [1]. Calcifications were deposited within the walls of the abnormal vessels and, at times, in the surrounding "gliotic parenchyma", although he did not specifically refer to whether the latter involved the case of DVA or the other malformations. Although calcifications are frequently reported findings in CVMs, with or without DVAs [16, 39], they are not generally included in the radiological descriptions of DVAs. Dillon [30] illustrated a case, already mentioned above, with coexisting periventricular white matter abnormalities and calcifications surrounding a DVA. Saito and Kobayashi [7] reported the presence of calcifications in the basal ganglia and thalamus of the patient described above, with subcortical atrophy in the drainage territory of a large DVA. In our cases, calcifications were found either in the supratentorial or cerebellar white matter, the basal ganglia or the caudate nucleus. No calcifications were detected in the cortex. Whereas calcifications within a CVM are related to old

haemorrhages, it is not clear whether similar calcifications in isolated DVAs correspond to old haemorrhages or result from long-standing cerebral ischemia.

The mechanisms leading the formation of DVAs in utero is still unclear and, as mentioned earlier, remains controversial. The development of the superficial and deep parenchymal veins is intimately related to the expansion of the cerebral hemispheres [5]. There is supporting evidence that an ischemic insult occurring during the period of neuronal migration, between weeks 8 and 25 of gestation, may arrest neuronal migration and secondarily result in abnormal venous development [40, 41]. Such a vascular insult is an unlikely cause of the development of most DVAs based on the fact that neuronal migration abnormalities are not characteristically associated with DVAs. In the present study, none of the 60 patients that had undergone MRI showed any evidence of migrational abnormalities. It is possible that DVAs represent a collateral venous network, speculatively kept patent or developing in response to the loss of the normal medullar veins, possibly secondary to thrombosis. The absence of migrational abnormalities within the drainage territory of the DVA suggests that collateral drainage through the DVA is likely to be sufficient in most cases.

The parenchymal abnormalities observed in our study, therefore, likely developed after birth and possibly in adult life. In the case of the hemispheric drainage anomaly coexisting with cerebral hemiatrophy reported by Uchino et al [16], the skull was symmetric, which led the authors to consider that atrophy was acquired after childhood. Further evidence is provided by cases reporting de novo formation of a CVM around DVAs [30, 42]. As mentioned earlier, VHT is presumably the common physiopathological mechanism leading to atrophy, white matter lesions, calcifications and CVMs. Considering that a large proportion of DVAs had recognizable parenchymal abnormalities on CT and MR imaging, it may be safely assumed that VHT likewise affects a large proportion of DVAs.

Conclusion

Parenchymal abnormalities in the form of atrophy, white matter lesions, calcifications and CVM within the drainage territory of a DVA were encountered in almost two thirds of the DVAs reported in this series. The close topographic relation between parenchymal abnormalities and the drainage territory of the DVA suggested a cause-to-effect relation. These abnormalities were considered to be acquired after birth and did not generally lead to neurological deficits. The presence of parenchymal abnormalities seemed to be unrelated to DVA sizes. Chronic cerebral ischemia secondary to VHT is thought to be the physiopathological mechanism leading to the development of atrophy and white matter lesions, although VHT may also become manifest acutely in the form intracranial haemorrhage. Morphological considerations, such as parietal thickening of the DVA and DVA arrangement into a venous convergence zone, likely predispose DVA to developing VHT. Further investigation is warranted in order to clarify why some DVA are prone to forming CVMs while others develop white matter lesions or atrophy, what the histopathological correlate of the frequently encountered white matter lesions is, what proportion of DVAs develops thickened walls and what the triggering factor to this thickening is.

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Conflict of interest statement We declare that we have no conflict of interest.

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