SHORT REPORT

Clinical significance of intracranial developmental venous anomalies

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Abstract

Objectives—Venous angiomas, or developmental venous anomalies (DVAs), represent the most often occurring cerebral vascular malformation. The clinical significance of a DVA is, however, at present unclear.

Methods—A retrospective analysis was carried out on two series of consecutive cranial MRIs performed between January 1990 and August 1996 in a university department of neuroradiology and in a large radiological private practice. The medical records of all patients in whom a DVA was diagnosed were screened to identify the specific complaint which necessitated the imaging procedure.

Results—A total of 67 patients with DVA could be identified. In 12 patients an associated cavernoma was found. The main reason for performing the MRI was the evaluation of seizures or of headaches. In all patients with DVA in whom an intracerebral haemorrhage was diagnosed an associated cavernoma was present at the site of the haemorrhage. None of the 67 patients showed an association between the complaints that led to the MRI and the location of the DVA.

Conclusions—DVAs do not seem to be associated with a specific clinical presentation. In a significant percentage of cases, however, coexisting cavernomas are found which have a defined bleeding potential and should be treated independently of the DVA. This study supports the hypothesis that DVAs are a congenital abnormality of venous drainage without clinical significance.

(J Neurol Neurosurg Psychiatry 1999;67:234-238)

Keywords: developmental venous anomaly; venous angioma

leangiectasias, cavernomas, and venous angiomas.¹ All four types of cerebral malformations can also be differentiated by distinct neuroradiological features that enable the radiologist to make an exact diagnosis without knowing the patient's history.

Venous angiomas are characterised histologically by a composition of sometimes thickened and hyalinised veins with interspersed normal neural parenchyma.² On angiography they present typically as deep, medullary veins during the early or middle venous phase accompanied by a single large draining vein. On CT they are rarely detectable without contrast enhancement. After application of contrast medium the draining veins appear as a linear focus of enhancement.³ On MRI venous angiomas have a characteristic flow void on both T1 and T2 weighted images.⁴ Administration of MR contrast material considerably improves demonstration of the draining vein and often allows visualisation of the medullary veins.⁴ Although once considered rare, they are now thought to be the most frequent cerebral vascular abnormality. A postmortem study in 1978 including 4069 serial cases reported an incidence of 2.5%.⁵ A review of the literature in 1982, however, disclosed only 45 clinical cases reported up to that time.⁶

With the advances in non-invasive imaging technology, visualisation of DVAs became easier. As a consequence larger series of patients with DVAs were published including many patients in whom the DVA was apparently an incidental finding.⁷⁻⁸ A high coincidence with other intracranial vascular malformations was noted.⁸⁻⁹ Coexisting cavernomas were reported in up to 33% of patients with DVAs.⁸⁻¹² The cause of cerebral haemorrhage in these patients was usually attributed to the cavernoma. These studies resulted in a change in the therapeutic recommendations for patients presenting with a DVA. Whereas previously surgical removal of the DVA was considered as a therapeutic option by some authors,¹³ there is general agreement nowadays that a surgical approach should be reserved for associated cavernous angiomas which have a bleeding risk of about 1% a year.¹⁴⁻¹⁵ The change in the assessment has been reflected in the names which have been applied to this type of cerebral venous malformation. Whereas in the older literature it is

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Received 21 July 1998 and in
revised form
7 January 1999
Accepted 20 January 1999

With the wider availability of MR imaging, neurologists are often faced with radiological reports of findings, the clinical importance of which is uncertain. Such examples are venous angiomas or developmental venous anomalies (DVAs). According to the widely acknowledged classification of cerebral vascular malformations it is normal to differentiate between arteriovenous malformations, capillary te-

Table 1 Patient characteristics

Mean age (range)	46.5 (19–77)
Sex (M/F)	30/37
Location of the DVA:	
Supratentorial	41
Frontal	23
Temporal	6
Parietal	6
Occipital	4
Basal ganglia	2
Infratentorial	26
Cerebellum	23
Brain stem	3
Associated lesions	13
Cavernoma	12
Brain tumour	1

commonly known as venous angioma, Lasjaunias *et al* subsequently suggested the term DVA to underline its character as an embryological variant of venous drainage.¹⁶

To investigate whether DVAs are associated with a particular clinical presentation we performed a retrospective study which included all patients in whom a DVA was diagnosed during the period from January 1990 to August 1996 in a neuroradiological university department and in a radiological private practice.

Methods

The records of all cerebral MRIs performed between January 1990 and August 1996 by the Department of neuroradiology of the Aachen University Hospital and by a large private radiological practice situated in Aachen were reviewed retrospectively to identify patients with a DVA. Brain MRI of all records indicative of a DVA were then re-evaluated by an experienced neuroradiologist to decide whether the described lesion fulfilled the typical neuroradiological criteria for a DVA.

All MR scans at the Department of Neuroradiology were obtained on a 1.5 T Siemens MR imaging system, whereas MR scans in the private practice were obtained with a Philipps 0.5 T unit. In most patients MR imaging was carried out before and after intravenous administration of gadolinium-DTPA.

Medical charts and additional radiographic studies from all patients in whom a DVA was diagnosed were reviewed. In each patient the specific complaint that necessitated the imaging study was determined from the available medical records. Assessment of the relation of the symptoms or signs to the DVA was based on the location of the DVA.

Figure 1 (A) CT, (B) digital subtraction angiography (DSA), and (C, D) MRI/MRA in a 29 year old woman who presented with a history of two unexplained episodes of syncope. The venous anomaly is located in the frontal lobe and drains into the superior sagittal sinus. There is the typical caput medusa-like appearance of the small parenchymal veins (arrowheads) converging into one large medullary draining vein (arrow). (B) The caput medusa can be seen both on DSA, and (C) on the T1 weighted contrast enhanced MRI.

Figure 2 Reason for obtaining the MRI that led to the diagnosis of a DVA in the 67 patients.

Results

A total of 67 patients with DVA were found. In the series of 7266 MR scans obtained at the Department of Neuroradiology 51 patients were identified (group 1). Out of 11192 consecutive brain MR scans obtained in the private practice 16 patients with a DVA were found (group 2). In all but four of the patients in group 1 MRI was performed with and without intravenous application of gadolinium, whereas only eight of the 16 patients in group 2 received gadolinium. In 21 of the 51 patients in group 1 MRI diagnosis was confirmed by an intra-arterial angiography, which was mainly performed to exclude the possibility of a small arteriovenous malformation. None of the patients in group 2 underwent angiography. Patient characteristics and the anatomical location of the DVA are summarised in the table. A typical example of the radiological appearance of a DVA using different imaging techniques is shown in figure 1.

Significant intracranial lesions other than the DVA were identified in 14 patients (21%). In 12 patients (18%) cavernous angiomas were found. In 10 of these patients the cavernous angioma was located in close vicinity to the DVA. In three patients the DVA with the associated cavernoma was located in the cerebellum. Multiple cavernomas were discovered in two of the 12 patients; one patient had two and another patient three cavernous angiomas. No other associated vascular malformations were found. One patient had a grade III astrocytoma, another patient had multiple sclerosis with typical signs of demyelination on MRI.

The patients' complaints that led to the diagnostic neuroradiological procedure are summarised in figure 2.

Four out of 15 patients presenting with seizures had a first ever single generalised tonic-clonic seizure. One patient with simple partial seizures had advanced multiple sclerosis and another patient with complex partial seizures had a left temporal grade III astrocytoma. The remaining patients had generalised tonic-clonic seizures (five patients) or complex partial seizures (four patients) with no pathol-

ogy on MRI except the presence of the DVA. Interictal EEG recordings, which had been obtained using the Standard 10/20 System, were reviewed in all patients. The EEG was pathological in nine patients: six patients showed generalised spike wave activity or bilateral rhythmic slow waves; only three patients showed focal EEG abnormalities. In one patient with complex partial seizures who underwent a left hippocampectomy after electroencephalographic identification of a left temporal seizure focus the localisation of the DVA was in the left temporal lobe. After selective amygdalohippocampectomy without removal of the DVA the frequency of seizures declined considerably and the left temporal spike wave activity recorded by EEG had disappeared. In the remaining two patients there was no association between the focal electroencephalographic abnormalities and the location of the DVA. One patient with a normal interictal EEG presented with simple partial motor seizures involving the right arm and the right side of the face. A DVA associated with a cavernoma was located in third frontal gyrus. In the patients presenting with seizures a supratentorial location of the DVA was more frequent (82%) than in the remaining patients (58%).

The headaches in the group of patients with this complaint were classified as tension-type headache in four patients, as migraine with aura in four patients, and as migraine without aura in three patients. The remaining patients were classified as having atypical facial pain (two patients), paroxysmal hemicrania (one patient), and headache associated with a flu-like illness (one patient). There was no correlation between the site of the headaches and the location of the DVA in this group of patients.

Five patients presented with signs and symptoms of an intracerebral haemorrhage. In all five patients an associated cavernous angioma could be detected. In four of these a cerebral angiography was included in the patient's investigation to exclude the possibility of an arteriovenous malformation. In three

patients the site of the haemorrhage was the cerebellum, and in the remaining patients the haemorrhage was located in the thalamus and in the midbrain, close to the superior colliculi. In three of these patients (two with cerebellar haemorrhage and the patient with the thalamic haemorrhage) a cavernous angioma associated with a DVA was found in close vicinity to the site of haemorrhage. In the two remaining patients, a cavernous angioma was discovered close to the site of the bleeding whereas the DVA was located in the right occipital lobe (midbrain haemorrhage) or in the contralateral cerebellar hemisphere (cerebellar haemorrhage). The malformation was neurosurgically resected in only one patient, in whom the diagnosis of a cavernous angioma could be verified histologically.

In 28 patients various neurological signs and symptoms such as vertigo (four patients), syncope, tinnitus, dementia, or asymmetric reflexes were the reason for obtaining a cranial MRI. No correlation between clinical presentation and the location of the DVA could be made in these patients.

In three patients the MRI was ordered for non-neurological reasons (for example, screening for intracranial disease in a patient undergoing cardiac transplantation).

Discussion

Among 7266 patients studied consecutively with brain MRI at the university neuroradiology department a DVA was discovered in 0.7%. By contrast the percentage of detected DVAs in the neuroradiology practice was only 0.14%. In addition to the reliance on an automated computer search to screen the records of all MRI performed in the radiology practice, the main reason for the much lower percentage was the fact that in the radiology practice contrast enhanced MRI, which increases the sensitivity for detecting a DVA, was performed less often. Although no absolute numbers for the percentage of gadolinium enhanced MRI examinations were available, the fact that 50% of patients with the diagnosis of a DVA in the private practice had not received gadolinium compared with only 8% of patients in the university neuroradiology department, strongly supports this explanation. In addition, a 0.5T MRI scanner was used compared with the 1.5 T MRI scanner at the university, which might also be expected to further decrease the diagnostic sensitivity.

The 0.7% found among patients scanned with a 1.5T machine is consistent with a previously published large study of DVAs diagnosed in a series of consecutive MRI studies. A review of 15 000 craniospinal MRI yielded 72 DVAs (0.48%).⁸ This is by contrast, however, with the only consecutive postmortem series comprising 4069 necropsies, in which 104 DVAs were diagnosed (2.56%).⁵ The discrepancy may be due to a limited sensitivity of MRI in detecting small DVAs, but the possibility that normal veins were mistaken for a DVA on necropsy cannot be excluded completely.

The anatomical distribution of the DVAs was consistent with the literature, with about one

third of the DVAs located in the cerebellum and in the brain stem, and the remaining two thirds in a supratentorial location. Also in common with previous investigations a high incidence of associated venous cavernomas was detected in this study.^{8 17} By contrast with earlier studies we did not find an increased incidence of associated brain tumours.^{8 18} Among 67 patients with DVA there was only one patient with a grade III astrocytoma, which most likely represents a chance coincidence.

In the group of patients presenting with seizures there was only one patient in whom the location of the DVA was in the same area as the EEG focus. After hippocampectomy, however, the focus was no longer present. In the remaining patients no association between the location of the DVA and focal EEG abnormalities could be found. It can, therefore, be concluded that the DVA itself is rarely, if ever, the seizure focus. The high incidence of seizures in the DVA group, however, merits some consideration. Another series published on patients with DVAs also noted a high incidence of seizures in this patient group.⁷ It can be argued that this developmental anomaly of the venous vasculature is associated with anomalies of neuronal migration. Although no grey matter heterotopias were detected in association with a DVA in our patients there is still a possibility that DVAs are associated with small scale migration deficits that are not yet detectable with neuro-radiological methods. Such an association of a DVA with an anomaly of neuronal migration has been described occasionally in the literature.^{19 20}

The second most frequent reason for obtaining an MRI examination was headache, but no association could be made between location of headaches and location of the DVA. This lack of association is not surprising, given the fact that even patients with large arteriovenous malformations rarely present with headaches.

An associated cavernous angioma was found in all patients with DVA in whom an intracerebral haemorrhage was diagnosed. In all patients the cavernous angioma was situated close to the site of the bleeding. In three patients the DVA and the cavernous angioma were in such close proximity that it could be argued that the separation in DVA and associated cavernous angioma was artificial, and that it would be more suitable to consider the lesion as a complex malformation consisting of venous and cavernous parts.^{14 21} A different explanation could be that there is a close developmental relation between these types of venous anomaly and venous malformation. In studies presenting many patients with DVAs there was only one patient in whom a left parietal DVA was associated with a left parietal haemorrhage without demonstration of an associated cavernous angioma.⁷ In all other patients with DVA and cerebral haemorrhage the symptoms could be attributed to an associated cavernous angioma located at or near the site of the haemorrhage.^{8 17} The previously reported high incidence of symptomatic haemorrhage in patients with DVA may result from the fact that in the pre-MRI and pre-CT era

the angiographically occult cavernous angiomas were notoriously difficult to diagnose.²² A cerebral haemorrhage was often, therefore, causally linked to an angiographically diagnosed DVA. In addition, some of the apparent symptomatic DVAs could later be diagnosed correctly as arteriovenous malformation during surgery.²³

Case reports linking neurological or psychiatric symptoms with DVA should also be treated with care. In some reports the neuroradiological information given is insufficient to make the diagnosis of a DVA with certainty.²⁴ In others the correlation of symptoms and location of the DVA may well be incidental.²⁵

Although published follow up data up to now are not large enough for a definite conclusion,²⁶ DVA can be considered a benign cerebral anomaly which is very unlikely to become symptomatic. To our knowledge no cases of enlarging DVAs have been described in the literature; DVA can be considered a variation of venous drainage of otherwise normal brain tissue, as the region of the malformation is characterised by a complete absence of normal draining veins.¹⁶ This concept is supported by the catastrophic outcome of patients in whom a DVA was ligated neurosurgically and in whom extensive infarcts caused by venous congestion occurred postoperatively.^{27,28} The aetiology of the DVA remains speculative. It is commonly thought that DVAs represent an arrest during the development of the venous system which results in the retention of primitive embryological medullary veins draining into a single longer vein.¹⁶

It is concluded that in this series of 67 patients with DVA there was no association between the complaints that gave rise to the neuroradiological procedure and the DVA diagnosed on MRI. A patient in whom such a vascular anomaly is diagnosed should be reassured as to the benign nature of the DVA. Because the DVA is, however, a very unlikely explanation for the symptoms that led to the imaging procedure, additional investigations may be necessary to clarify the specific cause of the complaints.

We thank the radiologists of the Röntgenpraxis Dahmengraben, Aachen, for giving us the opportunity to screen their MRI records.

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