What the Neurosurgeon needs to know about Cerebral Developmental Venous anomalies

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Abstract

Venous Angiomas or Developmental venous anomalies (DVA) are extreme variations of normal transmedullary veins that are necessary for the drainage of white and gray matter, also are one type of cerebrovascular malformation (CVM), sharing category with capillary telangiectasias, cavernous malformations (CM), and arteriovenous malformations (AVM), each of which may also be associated with a DVA. DVA are the most commonly encountered CVM, accounting for up to 60% of all CVM. We present a review of the literatura

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Introduction

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transmedullary veins that are necessary for the drainage of white and gray matter, also are one type of cerebrovascular malformation (CVM), (1) sharing category with capillary telangiectasias, cavernous malformations (CM), and arteriovenous malformations (AVM), each of which may also be associated with a DVA. (2) DVA are the most commonly encountered CVM, accounting for up to 60% of all CVM. Their prevalence is around 2.5–9%, and they are usually solitary. (3)

Huang et al., in 1984 (4) analyzed the angiographic findings of DVA and proposed terming them “medullary venous malformations,” and Russel and Rubinstein (5) defined cerebrovascular malformations as “cerebral vascular hamartomas” as intention to embrace malformations with a pathological process, however, the most accurate term, DVA, was first suggested by Lasjaunias and Burrows in 1986, (6), because venous

malformations originate from persisting embryologic medullary veins related to alterations in the embryonic period; thus it has been used to describe what is also known as venous angioma, cerebral venous malformation, and cerebral venous medullary malformation. Abandonment of the other designations reflects the nature of the DVA, although the exact etiology remains unknown, does not involve endothelial proliferation, has no proliferative potential, no direct arteriovenous shunts, and generally normal brain parenchyma between the dilated veins. (6, 7)

The use of current neuroimaging techniques has led to great advances in the diagnosis of CVM, and particularly the DVA diagnosis (1), the aim of this work is to review relevant issues regard DVA, which can help a better understanding of the natural course of this type of cerebrovascular malformation.

DVA architecture

An appropriate knowledge of the cerebral venous anatomy is necessary to understand how DVA is constituted and which can be the meaning of its architecture, thus next will be discussed briefly the cerebral venous anatomy.

Cerebral venous anatomy

The superficial venous system:

This system drains the outer 2 centimeters of white matter and cortex in centrifugal direction through subcortical and intracortical veins toward pial veins, cortical veins and then dural sinuses.

Subcortical veins are composed of three segments: medullary, arcuate and

intracortical, in their traveling to the brain surfaces. White matter have also a superficial drainage provided by superficial medullary veins. The cortex's superficial drainage is predominately from intracortical veins, which travel to join either pial or superficial cortical veins. Pial veins, which lays in the brain surface, deep to the pia matter, collect blood drained from intracortical, subcortical and superficial medullary veins. Pial veins coalesce into cortical veins and then toward a dural venous sinus.

The deep venous system

Also known as Galenic venous system, drains blood in centripetal fashion from the more central white matter and paraventricular nuclei through tributaries of the great cerebral vein to the straight sinus. The deep medullary veins perform the deep drainage of white matter, they connect with subependymal vein, and as they traverse in their travel converge into 4 zones: include (1) a superficial zone within the white matter just below the corticomedullary junction; (2) a candelabra zone formed by horizontal connections between deep medullary veins; (3) a palmate zone consisting of connections between very small deep medullary veins just superficial to the lateral ventricles; and (4) a subependymal zone that represents the convergence of the venous trunks that drain from the palmate zone. The internal cerebral veins (ICV) or the vein of Galen are the major collectors of the subependymal zone. The bilateral ICV converge at or near the midline along with the bilateral basal veins of Rosenthal and medial and lateral atrial veins to form a singular midline vein: the great vein of Galen.

The drained blood runs into the major dural sinuses: superior sagittal sinus (SSS), inferior sagittal sinus (ISS), lateral sinus (LS), cavernous sinus and straight sinus, and then to the internal jugular vein (IJV). Superficial venous system predominantly drains the SSS and the LS. Deep white matter and basal ganglia are drained by the deep venous system toward the vein of Galen. Between these two cerebral vein system can be found many anastomoses.

DVA constitution

DVA consist of radially oriented and converging dilated medullary veins, which drain centripetally into a transcerebral collector (enlarged transcortical or subependymal draining vein) that opens either into the superficial subcortical or deep pial veins. They are composed of angiogenically mature elements; indeed they are widely accepted to represent anatomic variants of normal venous drainage. (3) Histologically the veins are enlarged and sometimes hyalinized, but otherwise normal. (3)

Because of the absence of normal or subependymal veins draining outer zone of the white matter and the cortex to the pial veins, it is generated an abnormal venous arrangement or disposition, in which is produced a reversing of the drainage pattern, i.e., instead of superficial venous drainage, the blood is drainage toward the deep subependymal veins, in a centripetally fashion. The deep venous territory, instead of draining blood centripetally from the more central white matter and paraventricular nuclei, drains blood in a centrifugal fashion toward either the pial veins or directly into a dural

venous sinus. (2)

Figure 1 – Angio CT – venous angiomas on right side

Depending on their location, they may feed into the superficial (dural) or the deep venous systems. Posterior fossa DVA tend to direct venous blood to the veins of the lateral recess of the fourth ventricle, the precentral veins, a transpontine vein, or the longitudinal intrategmental vein, as detected by venous phase angiography. (8)

Hypothesis

Their etiology and mechanism of development are unknown, but it is currently accepted that they act like a compensatory system of cerebral parenchyma venous drainage due to early failure, abnormal development, or an intrauterine occlusion of normal capillaries or small transcerebral veins and thrombosis of normal parenchymal veins. (7) These drainage pathways may have developed as a method for maintaining the hemodynamic equilibrium of the transcortical venous drainage. According to Saito and Kobayashi (9), during embryogenesis, some

accident occurs during the formation of the medullary veins or their tributaries, such as occlusion or maldevelopment, and as compensation results CVA, thus, the main suggested etiology for the formation of CVA is an embryologic accident that results in either arrested formation or thrombosis of the developing venous drainage of the specific region (10–12), that is followed by a secondary compensatory mechanism in which embryologic medullary venules persist and cluster locally in a large draining vein (6, 9, 10, 13). Thus, they may be better described as being the result of a fetal pathological event rather than true anatomic variations.

Associated parenchymal abnormalities

Although the parenchyma drained by a CVA is generally reported as normal brain tissue, there have been reports of parenchymal abnormalities in the vicinity, and they are more common than previously thought (2). The essential role of CVA in normal cerebral venous drainage is shown when cases of catastrophic venous ischemic and haemorrhagic complications arise as consequence from their surgical removal (14). Brain abnormalities found within the drainage territory of a CVA are (15): locoregional brain atrophy, which is observed in close to 30% of patients, followed by white matter, lesions (28.3% on MRI, 19.3% on CT), CM (13.3% of MRI investigations) and dystrophic calcification (9.6% of CT investigations).

To take into account, parenchymal abnormalities are equally observed in small-, medium-, and large-sized CVA, indicating that size is not a determining parameter for their development. Regarding the number of CVA, in the San Millán et al series, was

observed that only 1.2% of patients harboured two coexisting CVA, a lower incidence than the 12% reported by Uchino et al (16).

The coexistence of two or more CVM within the same location has been documented (17–19). The prevalence of this specific subtype of mixed vascular malformations has a wide range, 2.1–25.9% (20–22). Amongst these, the co-occurrence of CM and CVA has been reported most commonly (15, 21, 23, 24), almost up to 40% of CVA (15, 25, 26), this fact generally explains presenting neurological symptoms if there is concordance with the location of a CVA and the CM (13, 20, 27).

Other parenchymal abnormalities, including signal modifications of white matter on magnetic resonance imaging (MRI) scans, dystrophic calcification and locoregional brain atrophy (13,25,28), however with no clear information regarding the significance of these findings.

DVA may also be present adjacent to brain tumors, infarcts, demyelinating areas, and moyamoya malformations; also associated congenital anomalies of the cerebral arterial system such as primitive trigeminal artery, fetal origin of the posterior cerebral arterial system, as well as fetal venous anomalies such as retention of the primitive falx, occipital, and marginal tentorial sinuses, (29) lending credence to the theory that embryonic origin of CVS.

The neuropathologic changes underlying the radiographic changes associated with DVA or DVA-associated clinically significant syndromes are very poorly understood. Vaitkevicius et al (8) reported histopathologic evidence of vascular remodeling related to altered hemodynamics in the region of a DVA, including microvascular wall hyalinization

and calcification, which are consistent with chronic regional blood flow alternation and venous hypertension.

Clinical presentation

CVA are usually asymptomatic, and are frequently discovered incidentally during medical imaging studies of the brain. (3, 15), probably because CVA generally provide sufficient venous drainage of the involved territory (2).

Sometimes DVA may present with headache, seizure, focal neurological deficit, dizziness, and ataxia. (23, 30, 31) Symptoms can be produced either by venous congestion related to flow obstruction or mechanical compression (e.g., hydrocephalus or nerve compression). (32) The clinical sequelae of a CVA are likely related to the regional changes that occur near it.

As previously mentioned, CVA can be associated with cavernous venous malformations in up to 40% cases, and are considered now to be responsible for the vast majority of symptoms attributed to CVA in the pre-CT/MRI era (33, 34). CVA have been linked with ischemic or haemorrhagic infarctions, or with focal neurological symptoms (28, 35), probably due to stenosis or thrombosis of the collecting vein. Several authors have documented impaired brain perfusion attributed to venous congestion in areas drained both by small and large CVA (36, 37).

Garner et al. (31) retrospectively evaluated the risk of hemorrhage associated with DVA to be of 0.22% per year, while McLaughlin et al (33) prospectively found that risk to be of

0.68% per year, though only half of their patients were symptomatic.

It was initially proposed that CVA located in the posterior cranial fossa had a higher propensity to bleed than supratentorial DVA, but that is not currently considered as truth. Only in about 25% of cases are CVA diagnosed because of the occurrence of epileptic seizures (38). Moreover, these lesions are often difficult to relate causally or topographically to the epileptogenic zone (19). More often they are occasionally observed during the diagnostic evaluation of different clinical pictures.

Pereira et al. (7) reviewed the pathomechanisms of symptomatic DVA. Based on imaging findings and clinical symptoms, two major pathomechanisms can be identified: mechanical and flow-related. Although mechanical complications lead either to hydrocephalus or to vessel-nerve conflicts, flow-related mechanisms can be further subdivided into those that are related to an increase in inflow into the DVA or to an obstruction of the outflow. See Tabel 1.

Imaging

The CVA is a large transparenchymal venous structure with many abnormal tributaries that converge into a larger vein, which in turn drains into a venous sinus (39, 40). The morphological hallmark of a CVA is a cluster of venous radicles that converge into a larger collecting vein, giving the CVA typical "caput medusae" appearance, demonstrable through both, CT and MRI, allowing confident diagnosis of CVA without the need to obtain digital subtracted angiography (DSA). (12)

TABEL I
Pathomechanisms of symptomatic DVA

Symptomatic DVA	Mechanical	Hydrocephalus	Interventricular foramen	
			Aqueduct	
		Neurovascular compression	CN V, VII, VIII	
	Flow-related	Increased inflow	Microshunt into DVA	
			AVM draining into DVA	
		Dreased outflow	Anatomical	Obstruction of the venous collector
			Functional	Distant high flow shunt
CN: cranial nerve; DVA: developmental venous anomaly				

As mentioned previously, the collecting vein crosses the brain parenchyma to either join the superficial or deep venous system. (41)

Noncontrast CT may show a collecting vein as isodense or slightly hyperdense to cortex (11). Contrast-enhanced CT readily detects the draining vein of a CVA as a linear or curvilinear focus of enhancement that courses from the deep white matter to a cortical vein or to a dural sinus. (35)

However, because of its higher temporal resolution, DSA remains the best imaging modality to study the hemodynamic behavior of CVA. For this reason, DSA is performed in patients presenting with ischemic or hemorrhagic complications within the drainage territory of a CVA, or whenever an associated vascular malformation is suspected on CT or MRI (2, 12).

DVA have begun to be studied by resonance magnetic techniques, appearing as a hypointense signal globular or linear phase

and hypo or hyperintense T1 - T2 in the same way hyperintensity found surrounding parenchymal injury in 50% of cases. T1- and T2-weighted images demonstrate a characteristic parenchymal flow void corresponding to CVA. The enhancement of both the draining vein and the caput medusae may be seen also on gadolinium-enhanced T1-weighted sequences. MRI is particularly useful and far superior to CT for the evaluation of related parenchymal abnormalities as well as for the detection of highly associated cavernous malformations (15).

Overall, developmental venous anomalies are low-flow vascular malformations, which are usually less conspicuous in conventional non-contrast MR images (42). Susceptibility weighted imaging is ideal for screening patients with a high clinical suspicion of low-flow vascular malformations (43) and limits the use of contrast studies and digital subtraction angiograms in their

demonstration. State-of-the-art 320-multidetector row CT (320-MDCT) is available in a few institutions and can perform dynamic subtracted CT angiography, allowing for morphologic as well as hemodynamic analyses of DVA.

Arterialized DAV (venous predominant parenchymal AVMs or arterialized venous malformations)

These are atypical DVA which are characterized by early angiographic opacification during the mid or later arterial phases, are also known as. (44) Although the pathogenesis of this subtype of DVA remains uncertain, they may represent a spectrum evolving from a simple DVA to a "classic" AVM, (2, 12) in which the DVA may serve as a foundation for future AVM development.

Management

Conservative management is strongly suggested (45). Their management requires a thorough understanding of the nature of CVA, 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.

Indications for surgical removal are reserved for patients with large or recurrent hemorrhage, poorly controlled seizures, secondary trigeminal neuralgia or refractory to medical treatment, hemifacial spasm or hydrocephalus for aqueduct compression. (46)

Despite being the most frequent CVM, its treatment principles have not been established. A number of case series have reported a favorable outcome in DVA

thrombosis after anticoagulation. (30, 47–53) Prevailing this benefit despite hemorrhage. (48, 51)

Because most patients do well, no aggressive therapy, surgical or radiologic, needs to be performed. Failure to recognize this may cause needless anxiety and will result in potentially injurious treatment. It is certainly important to provide careful follow-up monitoring because aging can cause hemodynamic stress to DVA, causing possible occlusion and venous infarction.

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