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DEVELOPMENTAL VENOUS ANOMALY: MR AND ANGIOGRAPHIC FEATURES

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Developmental venous anomaly (DVA) is probably the most common anomaly of the intracranial vasculature. DVAs consist of multiple, radially oriented dilated medullary veins that converge into a transcerebral vein. We describe the imaging findings of this vascular anomaly in different patients and the role of different imaging modalities.

Key-words: Cerebral blood vessels, abnormalities – Cerebral blood vessels, MR – Cerebral angiography.

Developmental venous anomaly (DVA) was first considered a rare vascular malformation (1, 2). Nowadays, with the advent of Computed Tomography (CT) and especially Magnetic Resonance Imaging (MRI), DVAs are seen every week to month by radiologists (3, 4). Most DVAs are solitary, asymptomatic lesions and are discovered incidentally. They have a relatively benign nature with a low incidence of hemorrhage. When they do bleed, this is thought to be due to associated vascular malformations, like cavernous angiomas. The typical angiographic appearance of a DVA is a caput medusae appearance in the venous phase. MRI combined with MR angiography (MRA) replaces angiography in most uncomplicated cases as a non-invasive alternative (3, 5).

Case reports

Case 1

A 32-year-old woman presented with headache, with no particular location and no neurological deficit. MRI of the brain was made in another hospital that showed a flow void running transcerebral, suggestive for a vascular malformation (Fig. 1A,B). Initially, there was no gadolinium contrast given and an arterial feeder could thus not be excluded with MRI. Patient was referred to our hospital to perform a cerebral angiography to further characterize the vascular malformation and to exclude an arteriovenous malformation (AVM). The angiography gives the definitive diagnosis of a DVA in the left parietal lobe by showing typical dilated medullary veins – caput medusae appearance –, without arterial feeders, that drain into a transcerebral vein. This vein drains into the superior sagittal sinus (Fig. 1C, D).

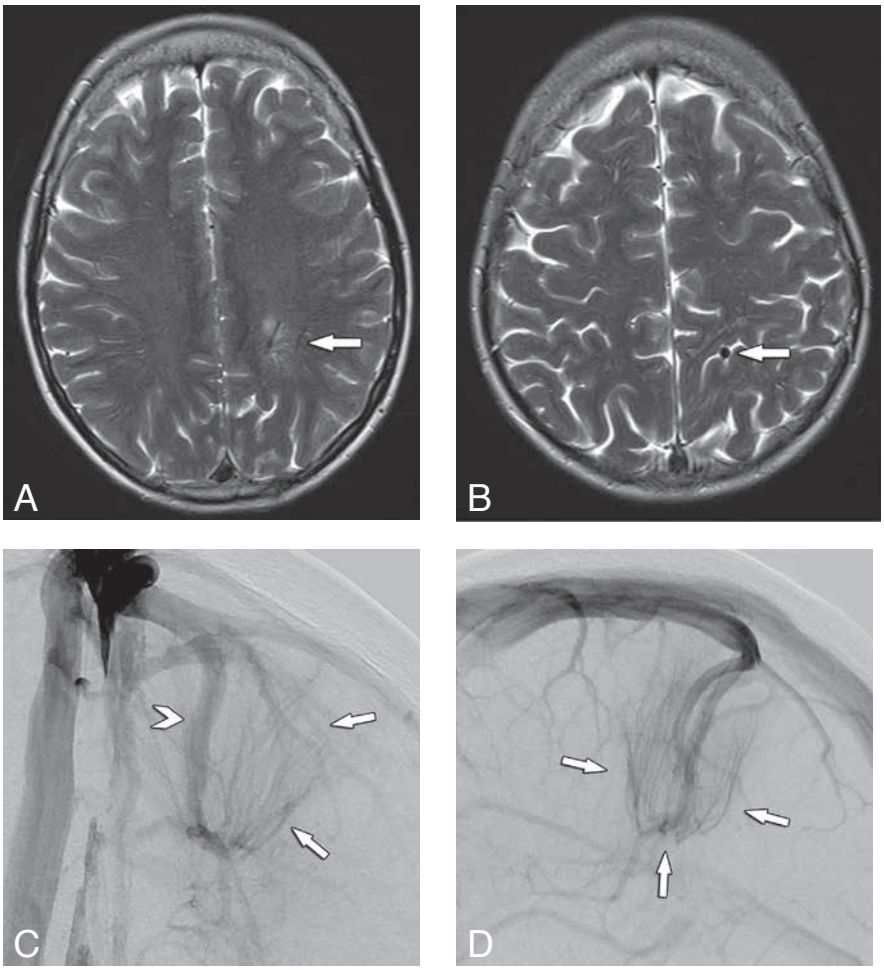

Fig. 1. — Axial T2-weighted MR-images showing a focus of abnormal signal in the left parietal lobe (A) with a flow void running transcerebral, suggestive for a vascular malformation (B). Anteroposterior (C) and lateral (D) cerebral angiography, venous phase, show the typical appearance of a DVA in the left parietal lobe with the *caput medusae* of dilated medullary veins (arrows), accompanied by a single, large draining vein that drains in the superior sagittal sinus (arrowhead).

Case 2

A 53-year-old man presented with acute headache in another hospital. MRI of the brain showed a lesion with heterogeneous signal intensity, hypointense rim in the left frontal white matter and is in close relationship with the frontal horn of the left lateral ventricle. This lesion represents a cavernous malformation. There are some additional vascular structures adjacent to the cavernoma. Cerebral angiography was performed to further characterize these structures and to exclude an AVM. The angiography showed the presence

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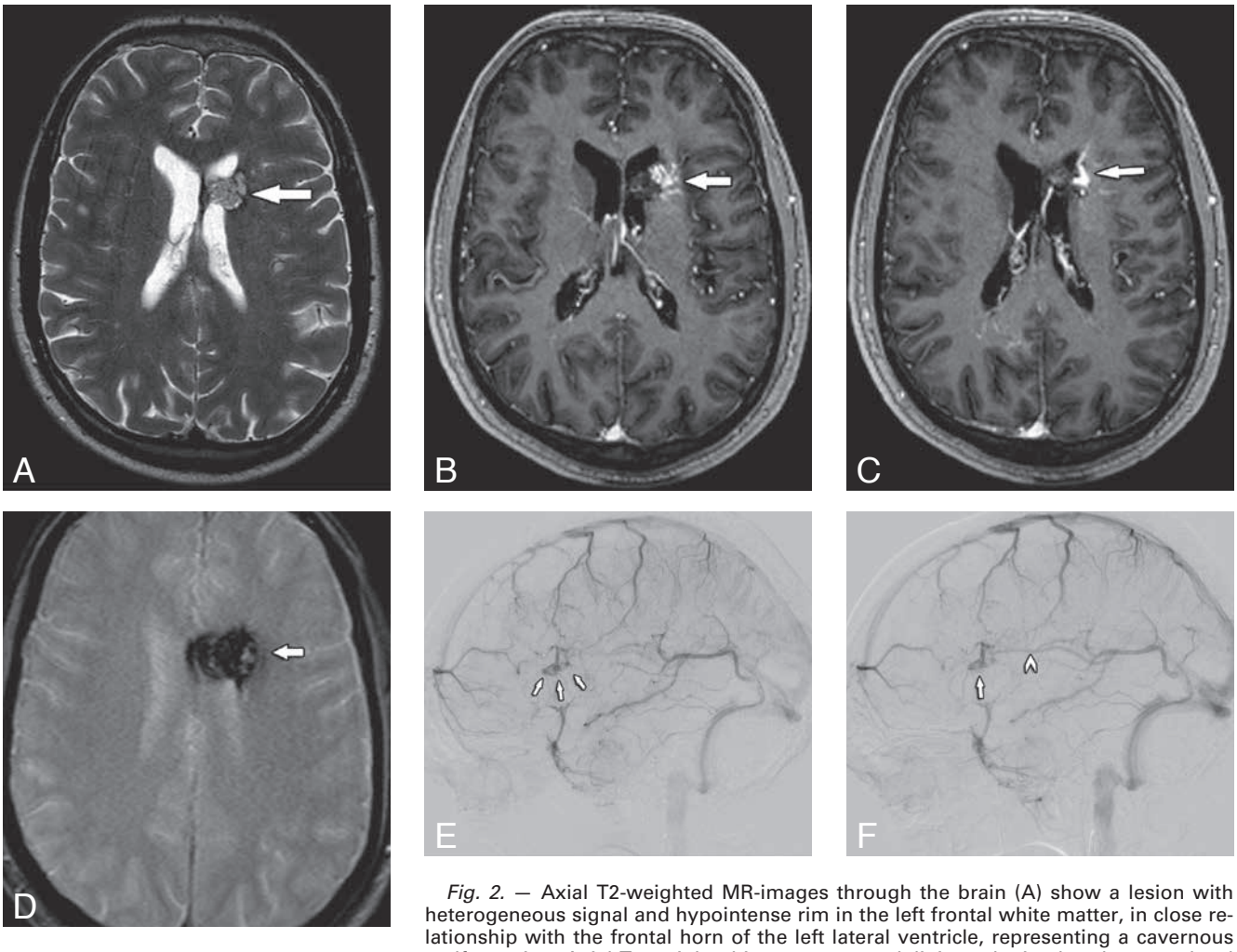

Fig. 2. — Axial T2-weighted MR-images through the brain (A) show a lesion with heterogeneous signal and hypointense rim in the left frontal white matter, in close relationship with the frontal horn of the left lateral ventricle, representing a cavernous malformation. Axial T1-weighted images post gadolinium obtained at the same level (B, C) reveal contrast-enhancing vascular structures adjacent to the cavernoma, representing an associated DVA. Susceptibility-weighted imaging (SWI) (D) shows striking hypointense 'blooming' of the cavernous malformation. Cerebral angiography, venous phase, sagittal views (E, F) confirm the presence of a DVA in the right frontal white matter (arrows), draining into the thalamostriate vein (arrowhead), in turn draining to the great cerebral vein.

of a DVA in the right frontal white matter, draining into the thalamostriate vein, in turn draining to the great cerebral vein (Fig. 2).

Discussion

DVA is probably the most common anomaly of the intracranial vasculature. Usually, they are discovered as incidental lesion on imaging studies. DVA is about 3 to 4 times more prevalent than AVM. They represent 60% of the cranial vascular malformations found at autopsy and occur in up to 4% of the population (3, 6). Most DVAs are asymptomatic and solitary lesions.

DVAs consist of multiple, radially oriented dilated medullary veins that drain normal cerebral nervous tissue and are located in the white matter. These medullar veins converge into a trancortical or subependymal draining vein. This vein drains into a superficial cortical vein or dural sinus or to a deep ependymal vein. The enlarged veins provide the main venous drainage for intervening normal brain.

DVAs were previously known as venous angiomas, venous malformations or medullary venous malformations. This terminology is replaced by *DVA* because it represents an anatomical variant of venous drainage of the white matter rather than a true vascular malformation (3, 4, 7). The etiology is unclear. DVAs are mostly found in the frontal and cerebellar white matter, respectively in 50 and 25% of cases (3, 8)

DVAs are thought to be benign entities that are unlikely to become symptomatic, although headache and epilepsy are reported. Most DVAs are discovered incidentally at contrast-enhanced CT or MRI (9, 10). There is controversy about the hemorrhagic risk of a DVA. It remains unclear whether the hemorrhage is always caused by an associated occult cavernous malformation, or that it may be caused by the DVA itself.

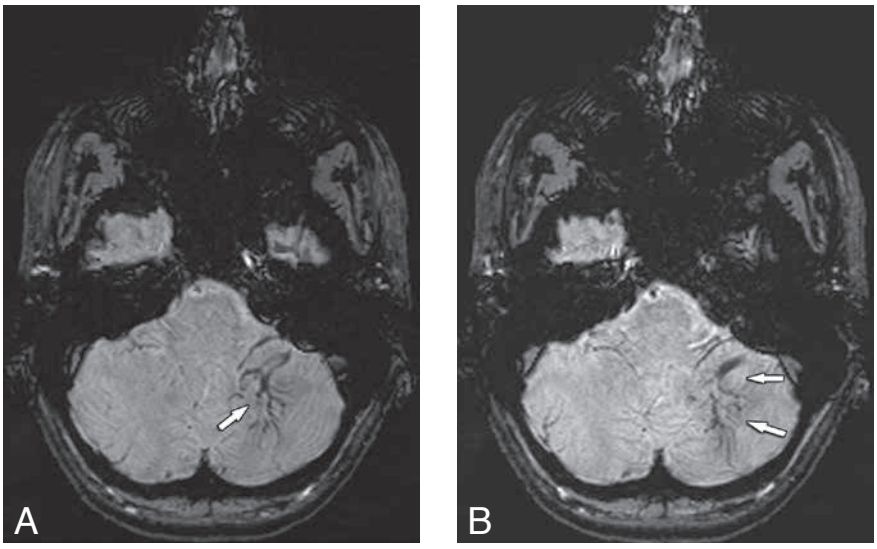

Fig. 3. — Susceptibility-weighted imaging (SWI) is very sensitive to venous blood and show the DVA as a hypo-intense lesion.

Possibly thrombosis or stenosis of the draining vein can cause a hemorrhagic event (9, 11, 12).

There is a coexistence between DVAs and vascular malformations, especially cavernous malformations. Abe et al found a coexistence of 33% between DVA and cavernous malformations with a relative high prevalence of coexisting lesions in the posterior fossa (5). Although less commonly reported, DVAs may be associated with neuronal migration anomalies, such as cortical dysplasias (4). Multiple DVAs can be found in the blue rubber bleb nevus syndrome or in sinus pericranii.

In contrast to other vascular malformations in the brain, DVA resection is not recommended. Because they drain normal brain tissue, resection of these venous pathways is associated with high morbidity and mortality and major neurologic complications (7, 12).

Imaging

DVAs were thought to be rare before the advent of CT and MRI imaging. Nowadays they are seen with high frequency as incidental finding at CT or MRI. Invasive imaging is nowadays mostly reserved for complicated cases or to exclude associated vascular lesions (4, 10, 13).

At cerebral angiography the pathognomic appearance appears during the venous phase of the cerebral angiogram. This includes the caput medusae of deep medullary veins, accompanied by a single, large draining vein that drains in a superficial cortical vein or dural sinus. The arterial phase of the cerebral angiogram is normal in most cases. Angiography is still the best technique to image DVA and to differentiate it from AVMs which have a different clinical significance. Cavernous malformations and capillary telangiectasias are angiographically occult.

Unenhanced imaging studies are normal in approximately 50%. On unenhanced MR images DVAs typically show a transhemispheric flow void on the T1- and T2-weighted images with normal non-gliotic surrounding parenchyma. Enhanced CT and MRI have a sensitivity of over 85% for detection of DVAs. Enhanced CT and MRI can show the draining vein of the DVA as a linear area of enhancement, that courses from the deep white matter to a superficial or deep vein or to a dural sinus. 2D MRI (time-of-flight) angiography can show the pathognomic features of a DVA in more than 85% of cases (10, 13). 2D-MRA is superior to 3D methods for the imaging of slow flow vessels, like veins, because they have less saturation effects. The imaging plane is best chosen perpendicular to the direction of flow. CT and MRI have the advantage of showing hemorrhage, angiographically occult malformations and other pathology, that cannot be demonstrated with angiography.

Contrast enhanced MR angiography is performed to exclude an arterial component – an indication for treatment- and to exclude focal stenosis of the draining vein – which is thought to be associated with hemorrhagic events. However, small AVMs or micro AVMs cannot be shown by MRI/MRA. In some cases there is variable enhancement of the brain parenchyma at enhanced MRI. It is suggested that this enhancement indicates venous restrictive disease in association of the draining vein and thus possibly also associated with a higher risk of bleeding (4, 14). Susceptibility-weighted imaging (SWI) can also be used to image venous structures. SWI appears to have a better detectability of venous structures than conventional T2*-weighted imaging (15). Therefore, SWI can be useful to detect DVA. They typically have a low signal intensity on SWI due to the blood oxygen level-dependent (BOLD) effect in the dilated medullary and draining veins (Fig. 3).

Conclusion

Developmental vascular anomaly (DVA, formerly known as venous angioma, venous malformation or medullary venous malformation) is probably the most common anomaly of the intracranial vasculature and represents an anatomical variant of venous drainage rather than a true malformation. They have a relatively benign nature with low incidence of hemorrhage. When bleeding is found, this is thought to be due to an associated vascular malformation, like a cavernous angioma. Alternatively, thrombosis or stenosis of the draining vein of a DVA is thought to give a hemorrhagic event. DVAs are often seen incidentally at CT or MRI. Although angiography still is the best technique to image DVA, with the typical caput medusae appearance in the venous phase, invasive imaging is nowadays mostly reserved for complicated cases and in case of acute hemorrhage to differentiate it from AVM. MR angiography can be an adequate non-invasive alternative to diagnose a DVA.

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