Uncommon Symptomatic Cerebral Vascular Malformations

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Summary: We describe three cases of unusual vascular malformations in which the most relevant angiographic findings were the presence of a pathologic arteriovenous shunt through multiple small direct arteriovenous fistulas and the lack of a clearly identifiable nidus. All malformations were symptomatic. Such lesions are relatively rare, but they should be taken into consideration in the differential diagnosis of cerebrovascular malformations.

Index terms: Angioma; Veins, abnormalities and anomalies; Fistula, arteriovenous

The classification of cerebrovascular malformations generally includes four types (1–3): arteriovenous malformation; developmental venous anomaly (DVA), also known as venous angioma; cavernous angioma; and telangiectasia, or capillary angioma. The clinical features and natural course of the high-flow vascular malformations (ie, arteriovenous malformations) have been described extensively and their pathologic role assessed definitively. On the other hand, the poor visibility of cavernous angiomas and telangiectasias on routine angiograms and the usual lack of symptoms of DVAs have resulted in a less clear understanding of their pathology. In particular, DVAs are commonly considered asymptomatic anatomic variations (4–11), even if some contrast exists (12–16).

In more recent studies, symptomatic DVAs have been linked to the frequently associated cavernous angiomas (7, 8, 17, 18). However, this classification does not consider the other, more rare, vascular lesions that have been described, albeit infrequently (19–23). A correct approach, therefore, must consider all the above differential diagnoses for correct management. We report three symptomatic cases of these rarer malformations.

Case Reports

Case 1

A previously healthy 35-year-old woman had a 1-month history of partial seizures. On admission no neurologic deficits were found; electroencephalography revealed a left temporal focus. Computed tomography (CT) and magnetic resonance (MR) imaging showed small anomalous vessels deep in the left hemisphere, without signs of previous hemorrhage (Fig 1A). A diagnosis of DVA was suggested. Angiography revealed two vascular malformations in the left basal ganglia and parietal lobe. In the arterial phase, two abnormal blushes were recognizable and, probably because of the presence of an arteriovenous shunt, the deep venous system was opacified early in the arterial phase (Fig 1B and C).

A superselective angiographic study showed that the malformations were fed by perforating vessels from the middle cerebral and anterior choroidal arteries, respectively (Fig 1D), and by terminal branches of the middle cerebral artery (Fig 1E). Flow was shunted through multiple small fistulas connecting small dilated arteries with small dilated veins; normal brain parenchyma was apparently interposed between the vessels, a separate nidus was not clearly identifiable (Fig 1D and E). Findings on the right carotid angiogram were normal.

The location of the malformations, their extent, and the lack of an identifiable nidus suggested a conservative approach, and medical treatment was initiated (carbamazepine 1200 mg/d), resulting in complete control of seizures.

Case 2

A 35-year-old woman was studied with MR imaging and angiography 3 years after the sudden onset of diplopia and hemiparesis. At that time, a CT scan obtained at another hospital was reported to show a brain stem hematoma. The patient recovered nearly completely in a few months; an MR study showed some small linear signal void structures in the dorsal mesencephalon that seemed to converge into a large vessel lying on the floor of the fourth ventricle (Fig 2A). In the arterial phase of the angiogram, several tiny vessels were seen projecting on the mesen-
cephalon and upper pons, along with a large venous collector corresponding to the vessel seen on the MR image; final drainage was primarily into the precentral cerebellar vein. A nidus was not clearly seen (Fig 2B and C). We interpreted this as a vascular malformation characterized by minute shunts between branches of the posterior cerebral and superior cerebellar artery into veins of the mesencephalon and pons. Because of the location of the malformation and the lack of signs of recent hemorrhage, the patient was discharged without therapy.

Case 3

A 39-year-old man was admitted because of a sudden right-sided motor weakness, aphasia, and headache. A CT scan showed a large left temporoinsular hematoma. The patient made a partial recovery without surgical intervention. Two months later, the hematoma had completely resolved. An angiogram at this time showed a high-flow malformation in the temporoinsular region, which was most likely responsible for the hematoma. Multiple tiny vessels from the middle cerebral and anterior choroidal arteries filled short dilated vessels, probably veins, converging in the basal vein, which filled early in the arterial phase (Fig 3A). In the venous phase, several medullary veins of the temporal lobe converged into the basal vein in a pattern typical of DVA (Fig 3B and C). Superficial venous drainage was poor. Moreover, two aneurysms of the left and one of the right carotid artery were detected. While waiting for stereotactic radiosurgery of the malformation, one aneurysm was occluded with platinum coils; similar treatment is planned for the others.
Discussion

In addition to the four types of vascular malformations that have well-defined radiologic and pathologic characteristics (1–4), combinations of different types of lesions, in particular, DVAs and cavernous angiomas, are also well known. Furthermore, other, more rare, malformations are known that do not fit any of these types. Rather, they have features of more than one single type and appear to be mixed forms in which several variations are present (19–23). The cases reported here did not have pathologic confirmation, but their MR and angiographic features are similar to some previously described cases that were confirmed histologically.

In two of our patients (cases 1 and 2), multiple tiny slightly dilated arterial feeders drained...
From a practical point of view, such malformations were symptomatic, in contrast to the usual lack of symptoms reported with DVAs (12). In our third case, a DVA was present, but we could not determine whether it was part of a more complex malformation or simply associated with an arteriovenous malformation. The malformation of the third case was different. It was characterized by a single large venous collector opacified in the early arterial phase by arteriovenous shunting. The same vein, however, also served as drainage for several tiny medullary veins seen in the late venous phase with the typical appearance of a DVA. It is not clear whether we were dealing with an arteriovenous malformation with a very small nidus, possibly partially destroyed by the hemorrhage, associated with a DVA or with a more complex malformation in which a small portion of a DVA was involved with an arteriovenous shunt. A similar combination of arteriovenous malformation and DVA was described by Meyer et al (23).

The most relevant feature of all these cases was the multiple arteriovenous fistulas draining into a large collector. Unlike with an arteriovenous malformation, there was no nidus visible either on angiograms or MR images. Normal-appearing parenchyma was interposed between the fistulas, similar to those that had pathologic confirmation (19–21). The presence of several veins converging through the normal parenchyma into a large collector is a feature of a DVA in which there is no arteriovenous shunt, such as occurred in our first two cases and in similar cases reported in the literature (19, 21). In our third case, a DVA was present, but we could not determine whether it was part of a more complex malformation or simply associated with an arteriovenous malformation.

From a clinical point of view, it is worthwhile to note that all the above-mentioned malformations were symptomatic, in contrast to the usual lack of symptoms reported with DVAs (12). From a practical point of view, such malformations present a diagnostic and therapeutic problem, since they may be similar to DVAs on CT and MR imaging. Only an angiographic study, possibly superselective, can give precise information on flow velocity and arteriovenous shunting. In accord with Awad et al (21), we recommend an angiographic study when a symptomatic venous anomaly is suspected.

References

18. Wilms G, Bleus E, Demaerel P, et al. Simultaneous occurrence of