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Uncommon Symptomatic Cerebral Vascular Malformations

Mauro Bergui and Gianni Boris Bradac

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-

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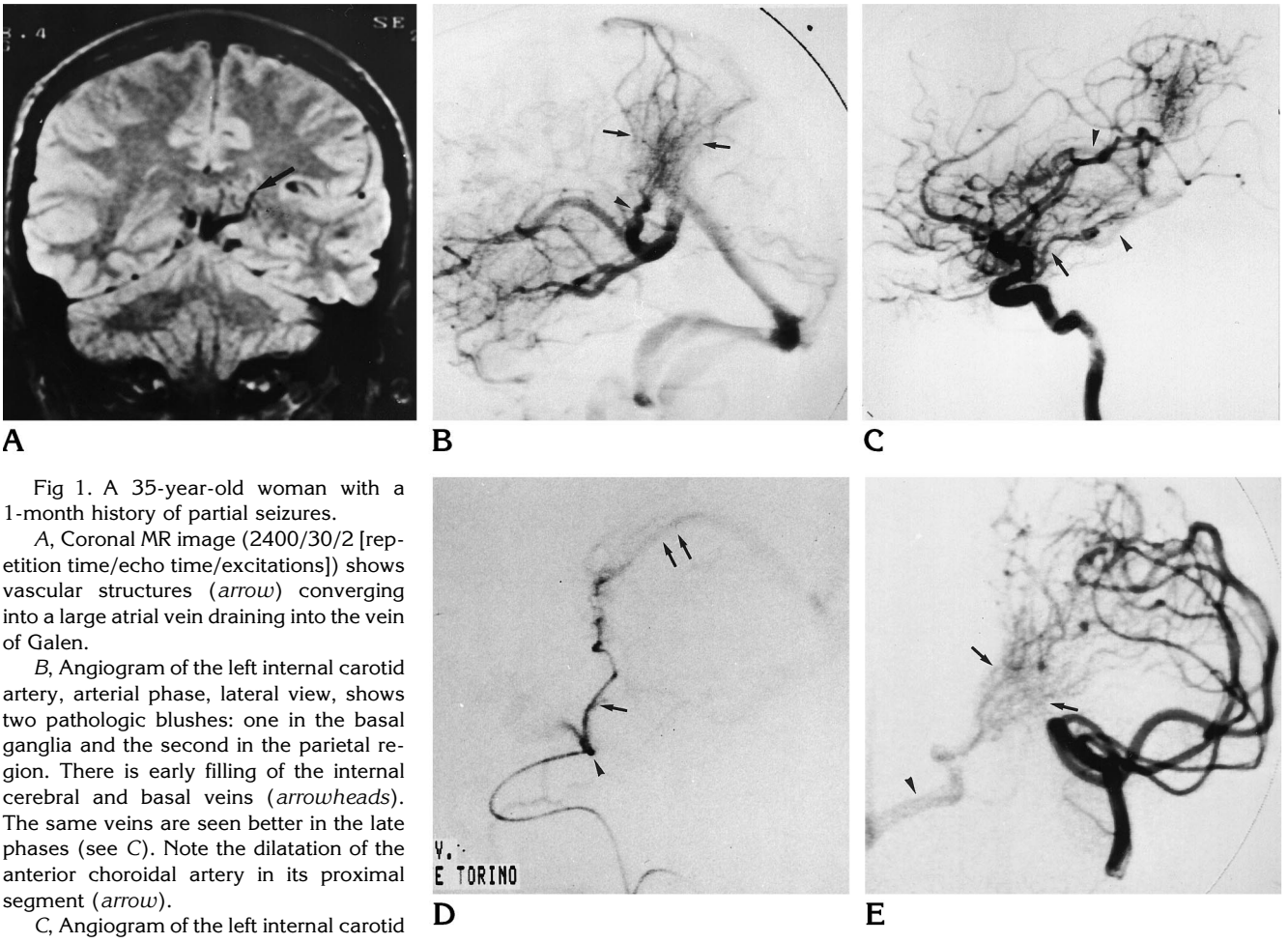

Fig 1. A 35-year-old woman with a 1-month history of partial seizures.

A, Coronal MR image (2400/30/2 [repetition time/echo time/excitations]) shows vascular structures (*arrow*) converging into a large atrial vein draining into the vein of Galen.

B, Angiogram of the left internal carotid artery, arterial phase, lateral view, shows two pathologic blushes: one in the basal ganglia and the second in the parietal region. There is early filling of the internal cerebral and basal veins (*arrowheads*). The same veins are seen better in the late phases (see *C*). Note the dilatation of the anterior choroidal artery in its proximal segment (*arrow*).

C, Angiogram of the left internal carotid artery, venous phase, lateral view, shows multiple deep medullary veins (*arrow*) draining into a dilated atrial vein (*arrowhead*). Note the enlarged basal and internal cerebral veins and the vein of Galen.

D, Selective injection of a perforating vessel of the left anterior choroidal artery (lateral view) shows well the arteriovenous shunt (*arrowhead* indicates catheter tip; *single arrow*, the perforating vessel; *double arrows*, the internal cerebral vein).

E, Selective injection of the left middle cerebral artery, anterior view, shows early filling of deep medullary veins (*arrows*) converging into a single atrial vein (*arrowhead*) and draining into the internal cerebral vein.

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 resorbed. 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.

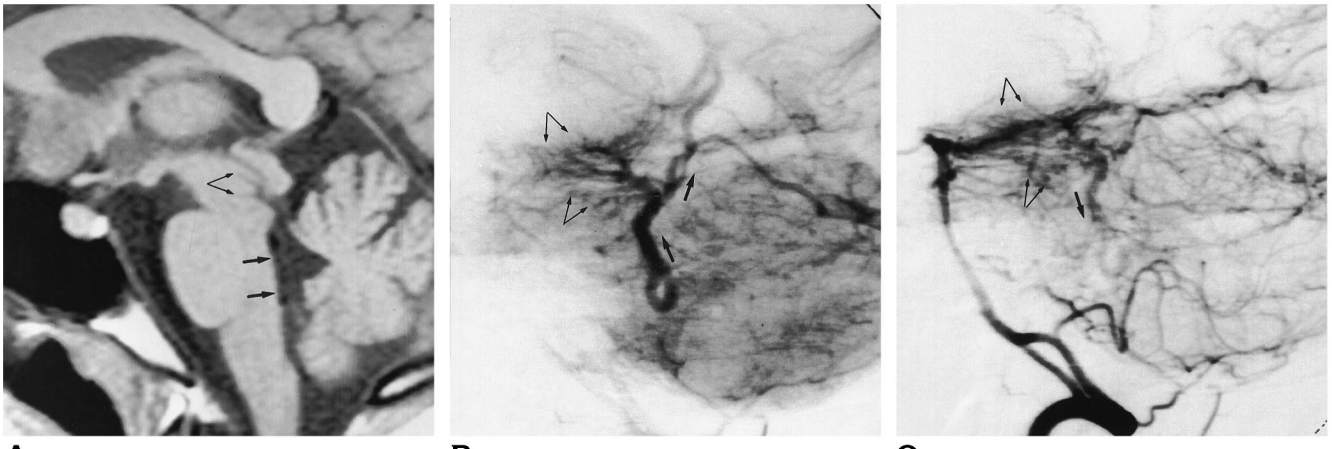

Fig 2. A 35-year-old woman 3 years after sudden onset of diplopia and hemiparesis.

A, Sagittal MR image (600/30/2) shows small vascular structures (*small arrows*) in the dorsal mesencephalon. *Large arrows* mark the venous collector on the floor of the fourth ventricle.

B and *C*, Angiograms of the left vertebral artery, lateral view, early (*B*) and late (*C*) arterial phases. Several tiny vessels (*small arrows*) already visible in the arterial phase fill a large collector located on the floor of the fourth ventricle; this vessel is initially directed downward and then upward to drain mainly into the precentral vein (*large arrows* mark the vessel and the direction of flow) and partially into a cerebellar vein.

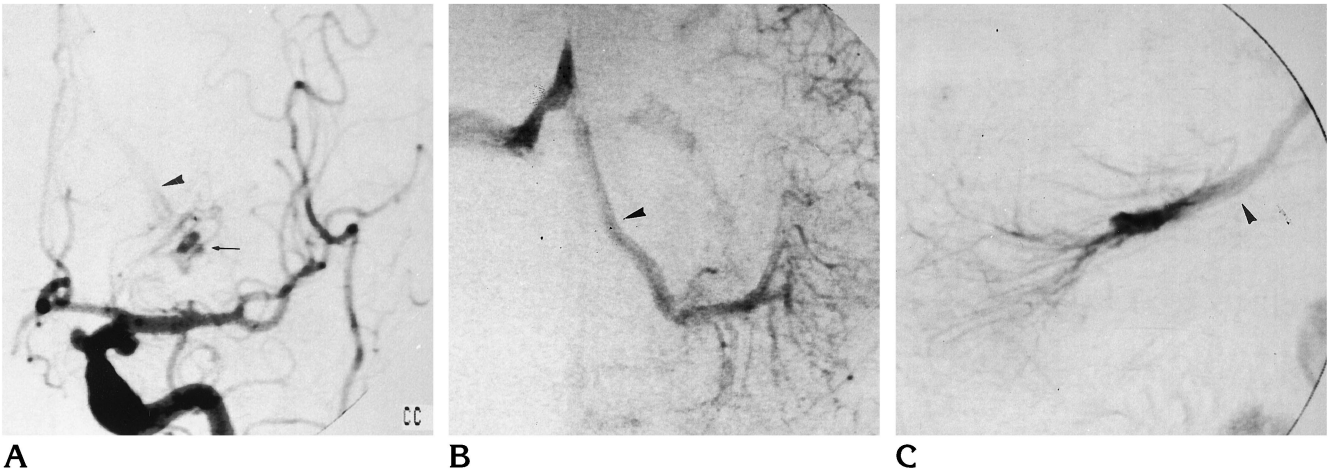

Fig 3. A 39-year-old man with sudden right-sided motor weakness, aphasia, and headache.

A, Angiogram of the left internal carotid artery, arterial phase, anterior view. Perforating vessels produce an early filling of short vessels, probably dilated veins (*arrow*) draining into the basal vein (*arrowhead*). Note also the aneurysms of the carotid siphon.

B and *C*, Angiograms of the left internal carotid artery, venous phase, anterior (*B*) and lateral (*C*) views. A caput medusae of medullary veins, with the typical angiographic aspect of the DVA, converge onto the basal vein (*arrowhead*).

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

directly into veins, with each vessel acting as a single arteriovenous fistula. Normal brain parenchyma was presumably interposed between single vessels, and a nidus was not identifiable. On the venous side, the vessels were organized in a caput medusae, with features similar to that of DVA, but differing in that, because of the arteriovenous shunt, these veins appeared in the early arterial phase. We found five similar cases reported (19–21) in which histologic examination showed a network of little arteries and veins with thickened walls. A common characteristic of all these lesions was the lack of a nidus; the malformation being formed by multiple fistulous vessels within normal brain parenchyma.

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.

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