In the course of embryological development, four successive cerebral venous patterns may be recognized. As each one becomes occluded, a new one forms. In the final stage there is extensive development and reabsorption of superficial veins. Should this final pattern fail to mature in a particular region, a preexisting pattern would persist to produce effective venous drainage. The cerebral venous malformation (CVM) is thought to represent such a failure. This malformation is characterized by a triad consisting of an absence of superficial veins, a “star-cluster” system of deep collecting veins, and a deep draining vein. In previous communications, it has been suggested that the CVM is the template of the arteriovenous malformation (AVM) and that the AVM is, in fact, a fistulized CVM. This implies that there may be transition forms in which features of both the CVM and AVM could be recognizable. The present communication illustrates the radiological features of eight cases. Three of these cases are used to document the essential triad that constitutes a CVM. Four cases include AVMs that display some elements of a CVM. One demonstrates an AVM that contains a CVM inside it. The clinical features of these cases are irrelevant to the main theme of this article and are not included.

Illustrative Cases

Three Classic Cerebral Venous Malformations

On routine cerebral angiography the middle cerebral vein may appear to be subject to much variability. It may be absent or may fail to connect to the cavernous sinus. It is routinely absent in perisylvian CVMs. In Case 1 a deeply seated CVM in the region of the genu drains though a wide collecting vein into the anterior sagittal sinus (Fig. 1 left). A more common type of “star cluster” can be seen in Case 2 (Fig. 1 center). Here, too, the middle cerebral vein is absent. The combination of a posterior fossa star cluster, a draining vein, and a defective lateral sinus is evident in Case 3 (Fig. 1 right).

Four Arteriovenous Malformations With Some Cerebral Venous Malformation Features

The anteroposterior and lateral views of the anomaly presented in Case 4 (Fig. 2 upper and lower left) are reminiscent of the classic CVM with its venous star cluster and draining vein. However, the lateral view of the venous phase reveals some unusual opacifications in relation to the venous dendrites (Fig. 2 lower left). The middle cerebral vein and the cavernous sinus are not visible. The sagittal sinus exhibits a peculiar appearance around its midpoint, and the vein of Rosenthal is unusually prominent. In the early-to-intermediate arterial phase on lateral projection, some abnormal small vessels may be identified (Fig. 2 upper right). In the late arterial phase, the malformation can be seen to have filled fully and it is clear that it is a late-filling AVM (Fig. 2 lower right). There is an absence of surface cortical veins.

In Case 5 the computerized tomography (CT) scan displays an anomaly (Fig. 3A) reminiscent of a CVM, which can also be seen on the venous-phase cerebral angiogram.
However, the very earliest lateral vertebral angiogram (Fig. 3C) reveals an abnormal perforating vessel, and a later view shows a very clearly defined fistulous anomaly (Fig. 3D). An overlay of Figs. 3B and 3D indicates that this fistulization occurs around the terminal of the main venous sac or on one of its major dendrites located immediately anterior to it. Despite an appropriate course of radiosurgery (accelerated helium nucleus), the malformation, although reduced in size, was seen to persist 2 years later. The size reduction provided a better view of the fistulization linkage (Fig. 3E) and an especially clear view of the core vein (Fig. 3F). In this case also there were venous and sinus abnormalities. The sagittal sinus was deficient anteriorly (Fig. 3G). It was compensated for by the presence of two longitudinally running surface veins (Fig. 3H). This is a classic appearance of the failure of the two independently formed sagittal plexuses to fuse. The middle cerebral vein drained toward the posterior sagittal sinus. The cavernous sinus was not visualized. A persistent filling defect in the region of the vein of Galen (Fig. 3B and E–G) might be interpreted as due to washout or to further organic defect.

Two magnetic resonance (MR) images (Fig. 4A and B) in Case 6 suggest a CVM, as does a venous-phase arteriogram (Fig. 4C); however, a late arterial-phase arteriogram displays filling of the malformation, claiming the anomaly to be yet another AVM (Fig. 4D).

Case 7 provides an example of a straightforward AVM filling in the early arterial phase. Unlike most AVMs, in which the image of the convoluted arterial supply and the convoluted venous drainage overlie the area of fistulization, thus obscuring its anatomical detail, this AVM is stretched out, revealing more clearly its basic anatomy (Fig. 5). The draining vein system closely resembles the CVM star cluster exhibited in Figs. 1 right, 3F, and 4D. There is an associated paucity of surface cortical veins (not illustrated). The pattern is one of direct fistulization of the venous structures without the formation of an extensive intervening capillary-like rete.

**Cerebral Venous Malformation Within an Arteriovenous Malformation**

Case 8 represents the rare combination of a CVM within an AVM. The early arterial phase shows a wedge-
shaped or triangular AVM, within which there lies a smaller triangular filling defect (Fig. 6A). In a later phase, after all of the early arterial filling has totally washed out, a venous structure appears and persists in the exact position previously occupied by the filling defect (Fig. 6B). Its vessels demonstrate the linear converging pattern that is typical of a CVM. In an intermediate-to-late phase, after the arterial phase of the AVM has washed out but before the CVM has begun to fill, the veins of the AVM are apparent (Fig. 6C). These veins are identical in appearance to those of the CVM and, in fact, are in continuity with the defective anterior sagittal sinus.

Fig. 3. Case 5. Computerized tomography scan (A) and angiograms (B–H) illustrating an anomaly with a venous pattern that is typical of a cerebral venous malformation (CVM). This also is arterialized in the arterial phase and is an arteriovenous malformation (AVM). Two years subsequent to radiosurgery, fistulization continues, but the reduced arterial input permits a better visualization of the fistulization process. Additional features include a defective sagittal sinus and nonvisualization of the cavernous sinus. A: Computerized tomography scan revealing the anomaly’s appearance to be compatible with that of a CVM or an AVM. B: Carotid angiogram, early venous phase, displaying the anomaly’s appearance to be compatible with that of a CVM. C: Early arterial-phase angiogram showing an abnormal perforating artery (arrow). D: Late arterial-phase angiogram showing the main cluster of fistulization at the end of the vein (open arrow) and a more distal anterior cluster (solid arrow). E: Postradiosurgery angiogram revealing that the fistulous vein still fills in the arterial phase. F: Postradiosurgery angiogram showing that in the late venous phase the anomaly looks like a typical CVM. G: Carotid angiogram failing to show the anterior sagittal sinus or the normal vein of Labbé. The middle cerebral vein blood flow is reversed. The cavernous sinus and the vein of Galen–straight sinus junction are not visualized. H: Carotid angiogram, anteroposterior view, revealing that the bilateral collateral longitudinal veins compensate for the defective anterior sagittal sinus.

Fig. 4. Case 6. Magnetic resonance (MR) images (A and B) and angiograms (C and D) displaying a brainstem arteriovenous malformation (AVM) with the typical venous pattern of a cerebral venous malformation (CVM). The lateral sinus on the opposite side does not fill. A: Sagittal MR image of a pontine vascular anomaly. The appearance is compatible with that of a CVM. B: Horizontal MR image showing a very abnormal vein that seems compatible with a CVM. C: Anteroposterior venous-phase vertebral angiogram revealing a star cluster (asterisk) and draining vein (small arrow) compatible with characteristics of a CVM. The opposite lateral sinus is poorly visualized and there is an irregularity of the confluence of the sinuses (large arrow). D: Anteroposterior arterial-phase vertebral angiogram showing fistulization of the abnormal vein. The basilar artery is indicated by a large arrow; the draining vein by a small arrow.
drain through a single common tortuous mediooccipital vein (Fig. 6C–E), which shows a two-phase drainage pattern related in time to the outflow of the two malformations.

Discussion
Throughout this series there have been two recurrent themes: an abnormality of surface veins and a star cluster of collecting veins. In general, the venous anomaly appeared as a paucity or absence of surface veins or a reversal in the direction of venous blood flow in the area overlying the star cluster. The most common venous anomaly we encountered was an absence of filling of the middle cerebral vein and of its associated cavernous sinus. Abnormalities of the sagittal and lateral sinuses were al-
so observed. Not only was the star cluster seen in all of the CVMs, but a structure of virtually identical appearance was recognized in all of the AVMs. Thus arises the suggestion that the two anomalies have a common origin, related to a defective formation of the venous mantle (perhaps an exaggeration of the normal spontaneous occlusion that occurs), and that they differ only in that one is fistulized and one is not. It is impossible to escape the conclusion that both the CVM and the AVM in Case 8 are part of the same structure in which only the periphery has become fistulized. This intimate association further enhances the speculation that the two lesions are variants of a common developmental failure.

During the embryological stages, if the surface veins of a territory fail to develop, the blood must drain deeply into an unblocked escape channel. Thus a number of collecting veins must coalesce deeply to form a draining vein. This is simply a restatement of the classic triad of defective surface drainage, star cluster of collecting veins, and deeply draining vein of the CVM. Although a general failure in the development of the superficial veins is postulated as an etiological or contributory factor in the development of these lesions, no specific role is attributed to individual abnormalities in the sagittal or lateral sinus. Their aberrations, like the combined AVM–CVM in Fig. 6A and the two separate AVMs in Fig. 2 lower right, simply indicate that multiple abnormalities of venous structures may coexist. The later development of the middle cerebral vein and its still later attachment to the cavernous sinus make it unlikely that its recorded variations play a causative role in the development of a CVM or an AVM, although a derivative role could be contemplated. Moreover, it is known that thrombosis of a dural sinus in the adult may be followed by the development of a dural venous fistula. It is reasonable to suspect that should a thrombosis occur in the whole or part of the star cluster then that thrombosis might similarly induce fistulization and thus create an AVM. Figure 1 right might represent the basic template of both malformations. Figures 2 upper left, 3E, and 4D seem to represent the simplest forms of fistulization. The emphasis, seen in Fig. 5, seems to lie in a direct communication between artery and vein, rather than in an elaborate intervening rete. In all cases, the large vein rather than a large rete appears to be the dominating factor.

These speculations are not advanced with any measure of proof but are offered as a “best possible fit” explanation on the basis of the available material.

References


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