Developmental venous anomaly with an arteriovenous shunt and a thrombotic complication

Case report

SIVIERO AGAZZI, M.D., LUCA REGLI, M.D., ANTOINE USKE, M.D., PHILIPPE MAEDER, M.D., AND NICOLAS DE TRIBOLET, M.D.

Departments of Neurosurgery and Neuroradiology, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland

Developmental venous anomalies (DVAs) are common congenital variations of normal venous drainage that are known for their benign natural history. Isolated cases of symptomatic DVAs with associated arteriovenous (AV) shunts have recently been reported. The present case, in which thrombosis occurred in a DVA involving an AV shunt, raises intriguing questions regarding the clinical characteristics of these lesions and can be used to argue in favor of considering such lesions to be arteriovenous malformations (AVMs).

A 39-year-old man presented with acute thrombosis in a complex system of anomalous hemispheric venous drainage, which included two distinct DVAs, one of which involved an AV shunt. The hemodynamic turbulences induced by a communication between shunted and normal venous outflows were the possible predisposing factor of the thrombosis. Follow-up angiographic and magnetic resonance images revealed complete recanalization of the thrombosed vessel and provided a thorough visualization of the particular angioarchitecture of the DVA.

Acute thrombosis within a DVA with an AV shunt has not been reported previously and, thus, this case can be added to other reports of complications that arise in this particular type of DVA. The authors hypothesize that the presence of an AV shunt in a DVA is a risk factor for aggressive clinical behavior of the anomaly, rendering those lesions prone to complications similar to AVMs.

Although no treatment can be offered, the presence of an AV shunt in a DVA warrants close follow-up observation because such lesions may represent a particular subtype of AVM and, therefore, may exhibit an aggressive clinical behavior.

KEY WORDS • developmental venous anomaly • venous angioma • arteriovenous shunt • cerebral vein thrombosis • arteriovenous malformation

Case Report

This 39-year-old man was found to harbor a complex DVA when, at the age of 28 years (1988), he underwent cerebral angiography at another hospital for investigation of head and neck pain. No treatment was recommended at that time and the patient was well until May 1995, when he visited our institution following three episodes of generalized tonic–clonic seizures. On admission, the patient was sedated, intubated, and given ventilation. A head CT scan revealed blood in the patient’s right basal cisterns and an acute thrombosis of a venous channel that was located on the surface of the right temporal lobe. Four-vessel angiographic and cerebral MR images demonstrated the presence of thrombosis in one of the venous channels that participated in the drainage of the DVA. After extubation on the following day the patient was found to have a mild left hemiparesis and homonymous left hemianopsia. These neurological deficits resolved over a period of 10 days and the patient was discharged home on a regimen of antiaggregation (acetylsalicylic acid) and antiepilepsy...
systems communicate abnormal. A caput medusae During the venous phase, the superficial venous system appears a dilated draining vein toward a midline tentorial sinus. Lower: (arrow) are visible in the basal ganglia region as they drain through caput medusae During the late arterial phase, dilated medullary veins forming a caput medusae (Fig. 1 upper right) of the basal ganglionic DVA as it drained toward a midline tentorial sinus. During the normal venous phase (Fig. 2 lower left), the second caput medusae was visible as it drained near the communication site between the superficial and deep venous drainage systems. Compared with the angiograms obtained in 1988, there were marked changes in the superficial venous drainage of the right hemisphere. There was a lack of opacification of the main draining vein, which extended from its origin at the converging point of the cortical veins to the point at which it connected with the deep venous drainage system. The deep venous outflow was dilated and partly drained through the distal portion of the superficial system by the veins’ connection on the basal surface of the temporal lobe, as demonstrated by the filling of the distal portion of the superficial venous channel despite the proximal thrombosis. Gadolinium-enhanced T1-weighted MR images demonstrated a lack of opacification of the superficial effluent, which could be observed up to the communication site between the two systems (Fig. 2 lower right). No other associated vascular anomalies were visible.

In 1996, four-vessel angiograms revealed complete recanalization of the superficial venous drainage system and unchanged hemodynamic characteristics of the DVA. Lateral views (data not shown) were identical to the ones displayed in Fig. 1. Cerebral MR images (Fig. 3 left) confirmed good canalization of the two draining systems. The dilated basal ganglionic medullary veins and the abnormal main superficial draining vein are demonstrated in Fig. 3 center. An angiogram (Fig. 3 right) obtained in 1999 confirmed the patency and superficial location of the thrombosed venous channel.

Discussion

Pathogenesis of DVAs

The congenital origin of DVAs has long been accepted and their previous designation, “venous angioma,” has gradually been abandoned to avoid confusion with a proliferative process. It has been postulated that DVAs represent a venous dysmorphism that involves a whole region of the brain and is caused by an erroneous genetic signal that occurs between the 45th and the 90th day of gestation. In a recent review of the literature on the formation of cerebral veins, four successive cerebral venous patterns have been described. As each one becomes occluded, a new one forms. In the final stage, extensive reabsorption of superficial veins takes place with persistence of a few major effluents. If a failure occurs in this compensatory surface-vein occlusion process or if a superficial venous channel thromboses, the deep coalescing network dilates, forming a caput medusae, and induces the

medications. At the 6-month and 4-year follow-up examination, the patient was found to be neurologically intact and results of control angiography revealed complete recanalization of the thrombosed venous drainage system. The patient was instructed to continue his regimen of anti-aggregation and antiepilepsy medications.

Review of Radiographic Findings

The patient first underwent angiography in 1988. On injection of the right CA, the angiogram revealed during the arterial phase an early appearing vein draining dilated medullary veins that formed a caput medusae (Fig. 1 upper). The image was diagnostic of a DVA in the basal ganglia that was associated with an AV shunt. During the venous phase several anomalies of the superficial venous system were evident (Fig. 1 lower). The superficial middle cerebral vein was absent and the cortical veins converged into a dilated temporal venous channel that drained into the right transverse sinus. Moreover, in the posterior temporal lobe region, dilated medullary veins formed a second caput medusae, corresponding to a second DVA.

In 1995, a nonenhanced CT scan revealed a round hyperdense area on the surface of the right temporal lobe, which was indicative of intraluminal thrombus (Fig. 2 upper left). Diffuse subarachnoid blood was present in the right basal cisterns (data not shown). On injection of the right CA, results of four-vessel angiography revealed the persistence during the late arterial phase (Fig. 2 upper right) of the basal ganglionic DVA as it drained toward a midline tentorial sinus. During the normal venous phase (Fig. 2 lower left), the second caput medusae was visible as it drained near the communication site between the superficial and deep venous drainage systems. Compared with the angiograms obtained in 1988, there were marked changes in the superficial venous drainage of the right hemisphere. There was a lack of opacification of the main draining vein, which extended from its origin at the converging point of the cortical veins to the point at which it connected with the deep venous drainage system. The deep venous outflow was dilated and partly drained through the distal portion of the superficial system by the veins’ connection on the basal surface of the temporal lobe, as demonstrated by the filling of the distal portion of the superficial venous channel despite the proximal thrombosis. Gadolinium-enhanced T1-weighted MR images demonstrated a lack of opacification of the superficial effluent, which could be observed up to the communication site between the two systems (Fig. 2 lower right). No other associated vascular anomalies were visible.

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**Fig. 2.** Neuroimages and angiograms obtained in 1995. *Upper Left:* Nonenhanced CT scan revealing a spontaneous round area of hyperdensity on the surface of the right temporal lobe (arrow). *Upper Right:* Angiogram of the right CA obtained during the arterial phase. In the early appearing basal ganglia DVA (arrowheads), the medullary veins are more dilated. The deep venous drainage opacifies the distal portion of the superficial drainage (small arrows) from the point at which both drainage systems communicate (large arrow). *Lower Left:* Angiogram demonstrating the lack of opacification of the main superficial draining vein, which is clearly visible from its origin (small arrow) to its communication with the deep drainage system (large arrow). *Lower Right:* Gadolinium-enhanced T₁-weighted MR image revealing the thrombosed main superficial draining vein (large arrow) close to the patent and contrast-enhanced deep venous drainage system (small arrow).

**Fig. 3.** Gadolinium-enhanced T₁-weighted images (left and center) obtained in 1996 and a control angiogram obtained in 1999 (right). *Left:* Good opacification of the superficial (large arrow) and deep (small arrow) draining systems is visible. *Center:* Opacification of the dilated transmedullary veins (arrowhead) and the main superficial draining vein (arrow). *Right:* Right anterior oblique CA angiogram obtained during the venous phase demonstrating the patency and superficial location of the main superficial draining vein (arrow).
formation of an abnormal draining vein that arises from the depths of the white matter.

**Thrombosis Within a DVA**

Only recently has thrombosis been reported as a potential complication of a DVA. Although it is well known that DVAs drain normal brain and that surgical occlusion of the abnormal vein may result in massive venous infarction of the drained parenchyma, it was only in 1986 that spontaneous thrombosis of a draining vein was first suspected and documented. Since then, 10 cases, including the present one, have been reported in the literature (Table 1). All reported cases of thrombosis within a DVA have been symptomatic, with no incidental finding and no coagulation disorder discovered during the workup. The age of patients with this complication, including that of the patient in our case, has ranged between 13 and 56 years old; outcome was good in eight cases, fatal in one case, and persistent disability in another. In none of the previous cases was an associated AVM reported.

**Developmental Venous Anomaly Associated With AV Shunt**

The association between a DVA and an AV shunt has already been reported in the neurosurgical and neuroradiological literature. A particularly detailed analysis of the angiographic characteristics of these lesions has been provided by Mullan and colleagues. In all their patients there was an abnormality of the surface venous drainage system (most frequently, an absence of filling of the middle cerebral vein), the classic star cluster of DVAs, and a fistula. In their description, those lesions were called AVMs, not DVAs. These authors have asserted that AVMs and DVAs have a common origin related to a defective formation of the venous mantle, their sole dif-

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**TABLE 1**

Review of 10 cases of thrombosed DVAs

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>DVA Location</th>
<th>Symptom(s)</th>
<th>Imaging Presentation</th>
<th>Treatment</th>
<th>Outcome</th>
<th>Length of Follow Up</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bouchacourt, et al., 1986</td>
<td>37, F</td>
<td>lt fronto-parietal</td>
<td>hemiplegia</td>
<td>infarction</td>
<td>anticoagulation medication</td>
<td>good</td>
<td>5 mos</td>
<td>recanalization, DNA visible only after recanalization of vein</td>
</tr>
<tr>
<td>Field &amp; Russell, 1994</td>
<td>34, F</td>
<td>rt parieto-temporal</td>
<td>headache</td>
<td>hemorrhage, hemianopsia</td>
<td>anticoagulation medication</td>
<td>good</td>
<td>3 mos</td>
<td></td>
</tr>
<tr>
<td>Kim, et al., 1996</td>
<td>13, M</td>
<td>rt temporoparietal</td>
<td>raised ICP</td>
<td>brain edema</td>
<td>craniotomy, ICP monitoring</td>
<td>fatal</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>Merten, et al., 1998</td>
<td>50, F</td>
<td>lt putamen</td>
<td>aphasia</td>
<td>hemorrhage</td>
<td>anticoagulation medication</td>
<td>good</td>
<td>2 yrs</td>
<td>decreased size of draining vein at 2-yr follow up</td>
</tr>
<tr>
<td>Konan, et al., 1999</td>
<td>31, M</td>
<td>rt cerebellum</td>
<td>raised ICP, obstructive hydrocephalus</td>
<td>infarction</td>
<td>EVD</td>
<td>disabled</td>
<td>1 yr</td>
<td>cavernous angioma close to DVA</td>
</tr>
<tr>
<td>Lai, et al., 1999</td>
<td>56, F</td>
<td>rt parieto-frontal</td>
<td>epilepsy</td>
<td>NP</td>
<td>antiepilepsy medication</td>
<td>good</td>
<td>6 mos</td>
<td>thrombosis on MR image</td>
</tr>
<tr>
<td>Thobois, et al., 1999</td>
<td>25, F</td>
<td>rt temporal</td>
<td>epilepsy</td>
<td>NP</td>
<td>anticoagulation medication</td>
<td>good</td>
<td>4 mos</td>
<td>resolution of thrombosis on follow-up MR image</td>
</tr>
<tr>
<td>Masson, et al., 2000</td>
<td>43, M</td>
<td>lt parietal</td>
<td>epilepsy</td>
<td>NP</td>
<td>anticoagulation medication</td>
<td>good</td>
<td>3 mos</td>
<td></td>
</tr>
<tr>
<td></td>
<td>68, M</td>
<td>lt parietal</td>
<td>epilepsy</td>
<td>NP</td>
<td>anticoagulation medication</td>
<td>good</td>
<td>30 mos</td>
<td>associated thrombosis of SLS</td>
</tr>
<tr>
<td>present study</td>
<td>35, M</td>
<td>rt temporoparietal</td>
<td>epilepsy</td>
<td>NP</td>
<td>acetylsalicylic acid, antiepilepsy medication</td>
<td>good</td>
<td>4 yrs</td>
<td>AV shunt recanalization</td>
</tr>
</tbody>
</table>

* EVD = external ventricular drainage; ICP = intracranial pressure; NP = no presentation (patient had epilepsy); SLS = superior longitudinal sinus; — = not applicable.
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ference being the presence of a fistula in AVMs as opposed to DVAs. The role of thrombosis in a DVA would be to induce fistulization, as occurs in dural sinus thrombosis and development of dural fistulae. Although fistulized DVAs are not always explicitly called AVMs, several authors1,2,9,11,22 believe that such lesions are at greater risk to develop complications than classic DVAs, and that their natural history resembles that of classic AVMs. The implication of DVAs in the development of AVMs has also been indicated by Nussbaum and associates,22 based on an observation of de novo development of several AVMs close to and draining into a large DVA.

Conclusions

This challenging case illustrates the potential complexity of DVAs. Despite the general opinion that DVAs are benign lesions, an increasing number of reports have demonstrated their potential complications. The simultaneous coexistence of a DVA, an AV shunt, and thrombosis has not previously been documented. Findings in our case support the theory that DVAs with AV shunts should not be considered a simple variant of venous angioma, but rather a particular type of AVM.

References


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