Stenotic and occlusive disease of the venous drainage system of deep brain AVM's

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Angiograms obtained prior to treatment in 53 cases of deep-seated cerebral arteriovenous malformations (AVM's) were retrospectively analyzed with particular attention to the topography of the AVM nidus and the venous drainage. The location of the lesion was determined by a combination of angiography and computerized tomography. Twenty-seven AVM's involved the basal ganglia and thalamus, 12 were located in the corpus callosum, six were intraventricular, and eight involved the mesencephalon and brain stem. Forty-one patients (77.3%) presented with intracranial hemorrhage. Vessel wall irregularities and/or stenosis of the system of the vein of Galen were observed in 14 cases, and occlusion of the deep venous system was present in seven cases. These AVM's showed numerous collateral venous pathways through enlarged medullary and cortical regional veins. There was dominant participation of the basal vein of Rosenthal in all cases. Unique local hemodynamic factors produced by the convergence of the draining veins of the AVM's into the vein of Galen and straight sinus may lead to a higher incidence of stenosis and/or occlusion of the venous drainage. The relatively high incidence of intracranial hemorrhage in these deep-seated AVM's may suggest a relationship between an increased incidence of intracranial bleeding and impaired venous outlets.

Key Words • cerebral arteriovenous malformation • vein of Galen • venous occlusion • collateral pathway

Cerebral arteriovenous malformations (AVM's) are developmental abnormalities of the vascular bed of the brain. The etiology and pathogenesis of these anomalies are not well known or understood. Cerebral AVM's are classified as dural, pial, or mixed, depending on topography and blood supply. Pial AVM's are fed by intracranial branches of the internal carotid and/or vertebral arteries and are drained by cortical veins and/or the Galenic venous system. Examination of the venous outlets of cerebral AVM's reveals a predictable pattern depending on topography. When veins draining primary brain territory are overloaded or occluded, redistribution and shunting toward regional veins is seen.

The cerebral venous territories reveal a marked degree of overlapping since connecting links trespass across their boundaries. The Galenic system is no exception to the general principle of the plexiform arrangement of venous systems. Schlesinger described two types of anastomotic veins related to the vein of Galen: intracerebral (transverse caudate veins, trans-cerebral anastomotic channels, and septal veins), and extracerebral (internal occipital vein, basilar vein of Rosenthal, and cerebellar veins).

Abnormalities of the Galenic system are rare, and they are particularly unusual in the adult population. However, when they do occur they are manifested by a florid clinical syndrome related to venous hypertension and infarction of deep brain structures. This is not true in patients having large deep-seated cerebral AVM's. In these cases, morphological changes in the venous walls are sometimes demonstrated and the vein of Galen and straight sinus may not be visible angiographically, despite a lack of clinical or pathological evidence of deep venous infarction. This report describes our findings in a series of cases with deep-seated AVM's.

Clinical Material and Methods

Fifty-three patients with deep-seated cerebral AVM's underwent angiographic analysis of the venous drainage
Occlusive disease of veins draining deep brain AVM’s

TABLE 1

Clinical presentation in 53 cases of deep brain arteriovenous malformations

<table>
<thead>
<tr>
<th>Clinical Presentation</th>
<th>Cases</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>hydrocephalus</td>
<td>2</td>
<td>3.7</td>
</tr>
<tr>
<td>seizures</td>
<td>4</td>
<td>7.5</td>
</tr>
<tr>
<td>intracranial hemorrhage</td>
<td>41</td>
<td>77.3</td>
</tr>
<tr>
<td>progressive deficit</td>
<td>5</td>
<td>9.4</td>
</tr>
<tr>
<td>severe headaches</td>
<td>1</td>
<td>2.1</td>
</tr>
</tbody>
</table>

TABLE 2

Location of deep brain arteriovenous malformations (AVM’s) in the 53 cases in this series

<table>
<thead>
<tr>
<th>Location of AVM’s</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>thalamus</td>
<td>9</td>
</tr>
<tr>
<td>basal ganglia</td>
<td>8</td>
</tr>
<tr>
<td>thalamus &amp; basal ganglia</td>
<td>10</td>
</tr>
<tr>
<td>corpus callosum</td>
<td>12</td>
</tr>
<tr>
<td>intraventricular</td>
<td>6</td>
</tr>
<tr>
<td>brain stem</td>
<td>6</td>
</tr>
<tr>
<td>brain stem &amp; thalamus</td>
<td>2</td>
</tr>
</tbody>
</table>

Results

In 32 cases no evidence of morphological changes in the system of the vein of Galen was noted. In 11 cases there was evidence of irregularities with or without stenosis of the vein of Galen (Fig. 1), and in 10 cases the vein of Galen and straight sinus were not visualized (Fig. 2 left). The deep AVM’s showing occlusion of the vein of Galen and straight sinus typically exhibited redistribution of blood flow through deep medullary veins toward the superficial venous system. The great basal vein of Rosenthal was particularly well developed in all these cases, carrying retrograde flow toward the cavernous sinus and superficial and deep Sylvian veins (Fig. 2 right). In some cases the draining veins were straight and very dilated (Fig. 3 left); in other cases they were very tortuous, serpiginous, and less dilated (Fig. 3 right).

Discussion

The great vein of Galen is a very thin-walled structure that lies in a ventrodorsal direction as it curves around the splenium of the corpus callosum. Below the splenium, the vein of Galen receives several venous tributaries. Among these, the most important are the internal cerebral veins, the basal veins of Rosenthal, the internal occipital veins, and the posterior pericallosal and superior cerebellar veins. The Galenic system is responsible for venous drainage of the hemispheric deep white matter, basal ganglia, corpus callosum, lateral and third ventricles, septum pellucidum, pineal region, and parts of the limbic system and cerebellum.

The position and course of the deep veins are more constant than those of the cortical cerebral veins. Many intracerebral anastomoses exist between the terminal tributaries of the central and superficial veins, thus providing a margin of safety in conditions that may alter the drainage by one system or the other. Hammock, et al., have described the angiographic and pathological changes observed in primates after surgical ligation of the vein of Galen. In their study, 16 Rhesus monkeys and six mongrel dogs underwent this surgical procedure. The animals were followed clinically for 6 months, and showed good clinical tolerance and no evidence of hydrocephalus or venous infarction. Hammock, et al., demonstrated that the venous collateral routes of drainage are readily available and that circulatory changes are very well tolerated following acute
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FIG. 2. Left carotid angiograms, venous phase, showing a basal ganglia arteriovenous malformation and corresponding draining veins. *Left:* Dominant participation of the internal cerebral vein (large arrow), basal vein of Rosenthal (curved arrow), and mesiotemporal and occipital cortical veins (small arrows) can be seen. The vein of Galen and straight sinus are not visualized. *Right:* The late venous phase shows the basal vein of Rosenthal draining in a retrograde direction into the deep Sylvian veins (curved arrows) and frontal cortical veins (open arrows).

occlusion of the vein of Galen if these collateral pathways are not involved. The angiographic patterns they described are similar to the ones that we found in our cases of deep-seated cerebral AVM’s with concomitant occlusion of the Galenic system.

The clinical outcome is entirely different if occlusion of the Galenic system includes significant compromise of the venous collateral pathways. Thrombosis of the Galenic system in children has been associated with prematurity, birth injury, and pyogenic infections, and Heinz, et al., have described its association with an AVM of the vein of Galen.

Thrombosis of the Galenic system in adults is rare. Bots has reported pathology of three adult cases in which the patients died following thrombosis of the vein of Galen. He described hemorrhagic infarction involving thalami, fornices, basal ganglia, and the ventral aspect of the corpus callosum. He also found thrombosis of the cerebellar veins, hippocampal veins, basal vein of Rosenthal, lateral ventricular veins, and internal occipital veins. He postulated that extensive venous thrombosis and hemorrhagic infarction are explained by the unique anatomical features of the Galenic system in which an extensive region of the brain drains into a single large vein. Garcia, et al., described spontaneous thrombosis of deep cerebral veins associated with an anterior hypothalamic AVM. At autopsy, they found thrombosis of the Galenic system and hemorrhagic encephalomalacia involving the draining territories of the internal cerebral veins and basal vein of Rosenthal. The venous hemorrhage was confluent and profuse and involved gray and white matter. They postulated that thrombosis of the vein of Galen is related to the development of venous stagnation secondary to a sudden local increase in pressure produced by intraventricular hemorrhage.

In 10 of our 53 cases of deep-seated AVM’s showing obliteration of the Galenic system, there was no clinical evidence of dysfunction of the deep-seated cerebral structures. At angiography, very well developed collateral pathways were seen in all the cases. As expected, the most important collateral vessels were the basal vein of Rosenthal, the mesio-occipital and parietal cortical veins, and the deep and superficial Sylvian veins (Fig. 2 right). Not infrequently, the same contralateral veins were also involved when the ipsilateral venous pathways had been “overloaded.” In 11 of 53 cases, there was evidence of vessel wall irregularity with or without stenosis of the vein of Galen and its tributaries (Fig. 1). These angiographic changes are probably related to endothelial damage secondary to turbulent, unstable, and irregular flow. Nornes and Grip documented the direction and pattern of flow in brain AVM’s, using a directional Doppler technique in the operating room. They described turbulent and highly irregular flow in the draining veins proximal to the AVM nidus and a more regular parabolic pattern in keeping with stable laminar flow in the distal portions of draining veins.

Some of the venous wall irregularities observed in our cases are highly suspicious of local damage. This local endothelial damage may be produced by turbulent flow as described by Nornes and Grip. Fry studied acute endothelial vascular changes associated with different flow patterns. He was able to reproduce different flow patterns and analyze the interaction of laminar and turbulent flows with endothelial surfaces. He theorized that intimal damage observed in turbulent flow is produced by a combination of the mechanical inter-
Action of pressure and shearing stress on the endothelial surface and the change in the electrochemical characteristics of the endothelial membrane produced by convective properties of flow. It is conceivable that this primary endothelial damage and subsequent thrombosis in situ could end in complete obliteration of the Galenic system, as observed in 10 of our cases.

Patients with AVM's of similar size and location displayed marked differences in the anatomical characteristics of their draining veins. There was a spectrum of variation in the caliber and tortuosity of the draining veins, most probably related to different predominant hemodynamic factors, such as lateral wall pressure, turbulence, and blood velocity.

The diameter of a vein depends on the elastic properties of its walls and the pressure inside it. Because the distensibility curve is nonlinear, the biggest difference in diameter occurs at a relatively low pressure. Veins, in contrast to arteries, elongate in response to pressure. This phenomenon gives rise to the tortuosity of varicose veins in the legs. Thus, one could predict that if a vein draining an AVM is subjected to a high lateral pressure it will become long, serpiginous, and slightly wider (Fig. 3 right). On the other hand, the phenomenon of post-stenotic dilatation observed in arteries shows that an artery becomes wider if it is subjected to turbulent flow. Assuming that the same phenomenon occurs in veins, the presence of a highly turbulent flow in the draining vein of an AVM could produce marked dilatation. These premises allow speculation that, if a vein draining an AVM is very long and tortuous, and slightly wide, the increased lateral pressure is the dominant hemodynamic factor; or, if the vein is very wide and relatively straight, turbulence probably plays the dominant role.

Turbulence will decrease the pressure, and therefore the vein will not be very tortuous.

In 41 of the 53 cases of deep-seated AVM's, the patients presented with an intracranial hemorrhage. This figure appears to be high when compared with AVM's involving other areas of the brain. Since our hospital specializes in treating patients with hemorrhage, we recognize that this factor may modify our relative figures of bleeding AVM's compared to non-bleeding AVM's, but we also postulate that the morphological changes in the Galenic system herein described may play a role in this increased incidence of hemorrhage. It is conceivable that an impairment of the venous outlet (either partial thrombosis or occlusion) may yield regional increased pressure in the AVM nidus, with resultant damage of the AVM capillary bed. One may also speculate that the hemorrhage may be produced on the venous side of the malformation, a phenomenon well established in dural AVM's presenting with intracranial hemorrhage secondary to a bleed from arterialized pial draining veins. Albert has related the frequency of intracranial hemorrhage in cerebral AVM's to the number of veins draining these lesions. He hypothesized that deep-seated AVM's with one or two draining veins are more prone to bleed than brain AVM's with three or more draining veins. This experience was not reproduced in our cases, and we did not find a direct relationship between the rate of bleeding and the number of veins draining an AVM. The great majority of the deep AVM's in our series had numerous venous pathways.

Dobbelaere, et al., have stressed the importance of performing a complete angiographic evaluation of the venous drainage of an AVM. They emphasized the
importance of: 1) identifying agenesis of primary venous pathways for a specific brain territory; 2) determining, if possible, the drainage of the normal brain around the AVM; and 3) identifying what they call "the phenomenon of saturation of the AVM venous drainage." They postulated that nonvisualization of the sinus rectus, accompanied by a major development in collateral pathways that also participate in the drainage of the normal brain, is associated with profound hemodynamic changes with a very precarious equilibrium. They stated that those cases are prone to develop postsurgical paroxysmal brain swelling when the venous pedicles are ligated during surgery. These observations, as well as the numerous morphological changes described in this present investigation, point to the importance of performing a thorough analysis of the venous drainage of all brain AVM's. It is possible that some correlation may exist between morphological and dynamic characteristics of the venous drainage of an AVM and the clinical evolution of these lesions. This information may also be valuable in predicting the severity of vasogenic edema after embolization and/or resection, and in differentiating those veins that, by participating in venous drainage of the normal parenchyma, should not be ligated or accidentally occluded by embolization.19

References


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