Venous-predominant parenchymal arteriovenous malformation: a rare subtype with a venous drainage pattern mimicking developmental venous anomaly

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Object. Considerable confusion exists in the literature regarding the classification of cerebrovascular malformations and their clinical significance. One example is provided by the atypical developmental venous anomaly (DVA) with arteriovenous shunt, because it remains controversial whether these lesions should be classified as DVAs or as atypical cases of other subtypes of cerebrovascular malformations. The purpose of this study was to clarify the classification of these challenging vascular lesions in an effort to suggest an appropriate diagnosis and management strategy.

Methods. The authors present a series of 15 patients with intracranial vascular malformations that were angiographically classified as atypical DVAs with arteriovenous shunts. This type of vascular malformation shows a fine arterial blush without a distinct nidus and early filling of dilated medullary veins that drain these arterial components during the arterial phase on angiography. Those prominent medullary veins converge toward an enlarged main draining vein, which together form the caput medusae appearance of a typical DVA.

Results. Based on clinical, angiographic, surgical, and histological findings, the authors propose classifying these vascular malformations as a subtype of an arteriovenous malformation (AVM), rather than as a variant of DVA or as a combined vascular malformation.

Conclusions. Correct recognition of this AVM subtype is required for its proper management, and its clinical behavior appears to follow that of a typical AVM. Gamma Knife radiosurgery appears to be a good alternative to resection, although long-term follow-up results require verification. (DOI: 10.3171/JNS/2008/108/6/1142)

Key Words: angiography • arteriovenous malformation • arteriovenous shunt • developmental venous anomaly • histological study

Cerebrovascular malformations have been divided into 4 categories: that is, AVMs, venous malformations, cavernous angiomas, and capillary telangiectasia. Nevertheless, some vascular malformations lie outside this classification scheme and their angiographic diagnoses are uncertain and questionable. An example is presented by atypical DVAs with arterial components (corresponding to arteriovenous shunts), which lack a typical AVM nidus. The literature on this type of vascular malformation is confusing, although many authors have recognized its existence. In 1967, Wolf et al. presented the first case of an intracerebral venous angioma with angiographic arterial involvement, and isolated cases of “atypical DVAs with arteriovenous shunts” have been reported sporadically. These cases are angiographically characterized by early-appearing dilated medullary veins that drain into a large main draining vein without typical nidus formation. Whether these lesions should be classified as DVAs or as atypical forms of another subtype of cerebrovascular malformation remains controversial. In most cases, the lesions have been angiographically classified as DVAs because of their prominent venous drainage patterns, which show marked similarity to the caput medusae appearance of DVAs and the absence of a typical AVM nidus. Some investigators have interpreted these lesions as mixed malformations in which a DVA shares venous drainage with an adjoining AVM. Some authors have viewed these vascular malformations as...
Venous-predominant parenchymal arteriovenous malformation

AVMs, although their suggestion has not been widely accepted.1,29

Angiographic findings, clinical implications, and treatment strategies for this subtype of vascular malformation are discussed based on our experiences. The present series is the largest study of angiographically and pathologically confirmed cases of this rare subtype of vascular malformation.

**Clinical Materials and Methods**

A retrospective analysis was conducted in a series of consecutive vascular malformations that were angiographically classified as “atypical DVAs,” “unclassified vascular malformations,” or “slow flow type AVMs” at initial angiographic evaluations. Fourteen cases were treated at Seoul National University Hospital between 1991 and 2005, and 1 case (Case 4) was treated at another institution.

Clinical information was obtained from hospital charts, from attending physicians, or by telephone interviews with patients or family members. Medical records were reviewed for demographic information, clinical presentations, neuroradiological findings, treatments, outcomes, and follow-up data.

In all cases cerebral angiography and MR imaging had been performed, and images were comprehensively reviewed. Angiographic examinations were performed using a commercially available biplanar angiographic unit (Integris Allura; Philips Medical Systems) with an image intensifier matrix of 1024 × 1024. Rotational angiography and 3D reconstruction of the lesions were performed in 4 cases. The C-arm rotated over a 240° range at a rate of 55°/second for ~ 4.4 seconds. The contrast medium was injected at a flow rate of 4–5 ml/second, resulting in a total of 16–20 ml for each rotational angiography acquisition. The image data were transferred to a workstation, where 3D image reconstruction by volume rendering was performed using the Integris 3D-RA (release 3.2) software package (Philips Medical Systems). For patients presenting before 2002, angiographic examinations were performed using a monoplanar angiographic unit (Angiocat; Siemens Medical Systems).

Original slides of the histological specimens were reviewed and the diagnoses were confirmed by a neuropathologist in 3 cases. Surgical specimens of the vascular malformations were fixed in formalin, embedded in paraffin, and stained with H & E and elastin for light microscopy. Multiple sequential sections were also cut and prepared for immunostaining of smooth muscle actin and glial fibrillary acidic protein to detect the presence of smooth muscle and gliotic brain tissue, respectively.

Follow-up data were available for 14 patients; the other patient died of surgical complications. The clinical follow-up duration for these 14 patients ranged between 2 months and 20 years.

**Results**

**Clinical Findings**

Patient characteristics and the anatomical location of the lesions are summarized in Table 1. Eight patients presented with an ICH. Cases 7 and 12 presented with ICH on 2 and 3 occasions, respectively. In 6 patients, an intracranial vascular malformation was found incidentally. Three lesions (Cases 1, 9, and 10) were detected in patients presenting with minimal or vague symptoms after a head trauma. In Case 6, a diffuse hemispheric vascular malformation was found when we investigated a pituitary adenoma by using brain MR imaging. In 1 patient (Case 13), an intracranial multiple vascular malformation was found during a work-up for facial cavernous malformations. One patient (Case 11) presented with epilepsy. In Case 5, a typical AVM had been treated with GKS, and another spatially separate vascular malformation was identified on follow-up angiography performed after GKS. This vascular malformation had originally coexisted with the AVM according to initial preradiosurgery angiography, but had been overlooked because of its faint angiographic staining in the presence of a large flow to the AVM.

**Neuroimaging Findings**

Magnetic resonance imaging showed the usual imaging patterns of typical DVAs in all patients.30 Radiating wheel spoke patterns of contrast-enhancing dilated medullary veins in white matter and an enlarged central draining vein was a common finding on MR imaging (Fig. 1). No tightly packed vascular mass suggesting an AVM nidus was noted in any case.

An example of the typical angiographic appearance of this vascular malformation is shown in Fig. 1. In the arterial phase, networks of tiny “cracklike” vessels and early appearing enlarged medullary veins were seen in all cases. In the early arterial phase, a poorly marginated, homogeneous, abnormal arterial blush was noted. In the midarterial phase, diffusely distributed, fine vascular staining mixed with poorly defined capillary blushlike parenchymal staining composed the vascular malformation. These abnormally staining vascular structures were connected to a network of radially arranged dilated medullary veins that converged toward a larger draining vein that in turn emptied into a superficial cortical or deep venous system in the late arterial and parenchymal phases. This caput medusae appearance of vein collections was a consistent finding in all cases. An angiographically defined arterial feeding vessel was noted in 1 case (Case 9). In this case, a moderately enlarged but otherwise normal-appearing anterior cerebral artery branch was visualized on angiographic studies; preoperative embolization was performed through this channel. The absence of angiographically documented enlarged feeding arteries and the absence of a typical AVM nidus were common angiographic findings in all other cases. The 3D reconstruction angiographic images showed detailed angioarchitecture of this type of vascular malformation (Fig. 2).

The largest lesion (Case 6) involved an entire hemisphere. In this case, angiography demonstrated abnormally dilated medullary, transcerebral, superficial cortical, and deep venous structures involving the entire right hemisphere, with no identifiable nidus.

**Treatment and Outcome**

Resection of the lesions was performed through a supratentorial craniotomy in 3 patients (Cases 4, 7, and 9). In these cases, the vascular malformations were composed of enlarged veins connected to markedly dilated red draining venous channels, and were intermingled with brain pa-
Lesions have been variously described as with "DV As with A V shunts" and "arterialized DV As" in the literature. In Case 8, follow-up angiography performed 14 days after GKS showed partial obliteration of the vascular malformation, which showed reduced lesion size and slower venous flow. This patient still occasionally experiences a mild headache.

There was 1 surgery-related death (Case 7), which occurred days after reoperation, and was due to uncontrolled brain swelling. Another patient (Case 12) died of a massive ICH during a third rebleeding episode. Two surgically treated patients (Cases 4 and 9) remain well and asymptomatic. In Case 5 the vascular lesion remains untreated. The patient in Case 11 still has seizures despite treatment with antiepileptic medication, and 1 patient (Case 10), whose vascular lesion still has not been treated, reports occasional throbbing headaches. Conservative management of asymptomatic vascular malformations was decided on by 3 patients (Cases 5, 6, and 13). Ten patients remained asymptomatic. In treated patients, no episode of rebleeding has been documented during the follow-up period.

**Histological Findings**

In 3 cases, a diagnosis of AVM was inferred based on histological findings. Dilated arterialized veins usually had asymmetrically thickened and hyalinized walls. Some of the blood vessel walls contained elastic laminae, characterizing them as true arteries. In Cases 4 and 9, anomalous vessels of different sizes were diffusely interspersed in normal brain parenchyma (Fig. 3). A compactly arranged smooth muscle layer was observed in most of the vessel walls after smooth muscle actin staining.

**Discussion**

Considerable debate exists concerning the use of the description “DVAs with AV shunts” and “arterialized DVAs” to describe this type of vascular malformation. The common angiographic finding of this entity is the presence of early-filling prominent medullary veins that converge on a main draining vein with or without an ill-defined arterial blush. Lesions have been variously described as follows: mixed malformations of atypical AVM and

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**TABLE 1**

Clinical features of 15 patients with vascular malformations*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Presenting Symptoms</th>
<th>Lesion Location</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8, M</td>
<td>incidental occipital</td>
<td></td>
<td>GKS</td>
<td>asymptomatic, partial obliteration</td>
</tr>
<tr>
<td>2</td>
<td>9, F</td>
<td>ICH frontal</td>
<td></td>
<td>hematoma evacuation, GKS</td>
<td>asymptomatic</td>
</tr>
<tr>
<td>3</td>
<td>15, F</td>
<td>ICH</td>
<td></td>
<td>GKS</td>
<td>asymptomatic</td>
</tr>
<tr>
<td>4</td>
<td>21, M</td>
<td>ICH temporal</td>
<td></td>
<td>resection</td>
<td>asymptomatic</td>
</tr>
<tr>
<td>5</td>
<td>24, F</td>
<td>incidental frontal</td>
<td></td>
<td>conservative</td>
<td>asymptomatic</td>
</tr>
<tr>
<td>6</td>
<td>27, F</td>
<td>incidental entire hemisphere</td>
<td></td>
<td>conservative</td>
<td>asymptomatic</td>
</tr>
<tr>
<td>7</td>
<td>29, F</td>
<td>ICH frontotemporal</td>
<td></td>
<td>resection</td>
<td>died</td>
</tr>
<tr>
<td>8</td>
<td>34, M</td>
<td>ICH frontal</td>
<td></td>
<td>hematoma evacuation, GKS</td>
<td>headache, partial obliteration</td>
</tr>
<tr>
<td>9</td>
<td>29, M</td>
<td>incidental frontal</td>
<td></td>
<td>resection</td>
<td>asymptomatic</td>
</tr>
<tr>
<td>10</td>
<td>34, M</td>
<td>incidental basal ganglia</td>
<td></td>
<td>conservative</td>
<td>headache</td>
</tr>
<tr>
<td>11</td>
<td>49, M</td>
<td>epilepsy temporal</td>
<td></td>
<td>conservative</td>
<td>seizures, medicated w/ AEDs</td>
</tr>
<tr>
<td>12</td>
<td>71, M</td>
<td>ICH basalganglia</td>
<td></td>
<td>conservative</td>
<td>died</td>
</tr>
<tr>
<td>13</td>
<td>26, F</td>
<td>incidental cerebellum, basal ganglia</td>
<td></td>
<td>conservative</td>
<td>asymptomatic</td>
</tr>
<tr>
<td>14</td>
<td>13, M</td>
<td>ICH cerebellum</td>
<td></td>
<td>GKS</td>
<td>asymptomatic</td>
</tr>
<tr>
<td>15</td>
<td>22, M</td>
<td>ICH cerebellum</td>
<td></td>
<td>GKS</td>
<td>asymptomatic</td>
</tr>
</tbody>
</table>

* AED = antiepileptic drug.
venous angioma; arterIALIZED venous anomaly; atypical AVMs; transitional forms between venous malformations and AVMs; AVMs draining into a large DVA; or small AVMs with venous predominance. 2–7,9–11,13,18–20,26,29

Findings in the present cases support the suggestion that DVAs with arteriovenous shunts should be viewed as an AVM subtype, and differentiated from DVAs. 1,21 Nevertheless, there appears to be considerable overlap between the angiographic patterns of the cases presented here and the patterns of DVAs. The caput medusae configuration of draining veins observed in the presented cases is highly suggestive of DVA. Nevertheless, overall angiographic findings were not specific for a diagnosis of DVA because of the presence of arteriovenous shunts and their frank arterial components. Early venous drainage is a finding that suggests the presence of an arteriovenous shunt in a cerebrovascular malformation, and should raise suspicion of an AVM. 25 A DVA is a venous-based lesion with normal arterial and capillary phases and circulation time, and therefore “DVA with arterial components” and “arterIALIZED DVA” are inconsistent designations. 8,24–27 At initial angiographic evaluations, we classified the presented cases as “atypical DVAs,” due to our preoccupation with the prominent caput medusae–like venous drainage pattern, which is known to be the most characteristic feature of DVAs. Moreover, the angiographic findings in these vascular lesions were not uniformly compatible with a diagnosis of AVM because of the absence of a typical AVM nidus of tightly packed vessels and enlarged feeding arteries. However, based on anatomical variations of subcortical medullary veins, some vascular malformations can adopt draining patterns resembling those of DVAs. 26,27 Thus, the caput medusae appearance may be considered an extreme form of variation in the venous drainage pattern of AVMs.

Although not all cases were histologically examined in the present series, the histological findings of all resected vascular malformations were interpreted as AVMs, and faithfully reflected their angiographic architectures. Histological studies showed numerous venous and arterial channels interspersed in normal brain tissue in a nonhemorrhagic case and in partially gliotic brain parenchyma in hemorrhagic cases. We recommend designating these lesions “venous-predominant parenchymal AVMs,” because they have more prominent draining veins than arterial feeding vessels, and these are intermingled with parenchyma without typical nidus formation. Three histologically con-
firmed cases can be used to argue in favor of considering the vascular malformations in our series to be AVMs.

Clinical manifestations appear to be associated with hemodynamic factors characterized by flow rates and the size and number of arteriovenous shunts.

In the present study, clinically silent lesions had low flow rates and slow circulation on angiography. In patients with lesions that had prominent arterial components and very early venous drainage, the malformations tended to follow an aggressive clinical course, and the 3 cases in which patients experienced rebleeding episodes strongly support this suggestion.

Based on our observations, these AVMs presumably follow a clinical course typical of AVMs. Symptoms, when they occur, were usually hemorrhage or epilepsy. Radical treatment should be recommended in patients who have suffered a massive or recurrent hemorrhage, as supported by our experiences of 2 cases of postoperative hemorrhage. Treatment of these AVMs may be challenging because neither arterial feeding vessels nor a localized nidus is clearly depicted during the operation, and incomplete resection may lead to a catastrophic hemorrhage. Postoperative catastrophic hemorrhagic infarctions in our 2 cases are believed to reflect an abruptly interrupted venous flow in draining veins without complete obliteration of feeding vessels.
Moreover, enlarged draining veins may be critical in enabling the venous drainage of surrounding structures. Gamma Knife radiosurgery for this type of vascular malformation has been considered as a therapeutic option, and has been proven to be efficient by some authors.\(^2,13,16\)

This type of AVM, with its angiographically ill-defined arterial components but with relatively predominant venous channels, may be easily overlooked and may be incorrectly diagnosed as a DVA. In such cases, symptoms such as hemorrhage or focal neurological deficits can be erroneously attributed to DVA. We stress the importance of detailed angiographic evaluation in patients with a symptomatic DVA to rule out the presence of this type of vascular malformation.

**Conclusions**

In our view, the vascular malformations described in this study should be viewed as a distinct clinicopathological entity of the AVM subtype. Correct recognition of this subtype of AVM is critical for proper management, and radical treatment should be recommended for symptomatic patients, especially those who have experienced a massive or recurrent hemorrhage.

**References**


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