The natural history of intracranial venous angiomas

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Cerebral venous angiomas are congenital anomalies of the intracranial venous drainage. Many believe that they are associated with a high risk of hemorrhage and neurological dysfunction, but newer neurodiagnostic imaging techniques are showing not only that they are more common than previously known but also that many have no associated symptoms. In this retrospective study, the natural history of venous angiomas was examined in 100 patients (48 males and 52 females) with radiographically identifiable lesions treated over a 14-year period. Information on the natural history of the lesion was obtained from clinical records and follow-up data. Imaging studies included angiography, computerized tomography, and magnetic resonance imaging. Angioma locations were classified as frontal (42 cases), parietal (24 cases), occipital (4 cases), temporal (2 cases), basal or ventricular (11 cases), cerebellar (14 cases), or brain stem (3 cases); 47 lesions were on the left side.

Headache as a presenting symptom was common (36 patients) and often led to other radiographic studies, but this appeared to be related to the vascular lesion in only four patients. Other possibly related complications were hemorrhage in one patient, seizures in five, and transient focal deficits in eight. Fifteen patients had no neurological signs or symptoms. The mean patient age at last contact was 45.3 years (range 3 to 94 years). All patients have been managed without surgery. It is concluded that significant complications secondary to venous angiomas are infrequent and that surgical resection of these lesions and of surrounding brain is rarely indicated.

Key Words • angioma • hematoma • vascular malformation • pregnancy • diagnostic imaging

Cerebral venous angiomas are congenital anomalies of intracranial venous drainage. Although autopsy series have demonstrated venous angiomas to be among the most common of the intracranial vascular malformations22 (WF McCormick, personal communication, 1987) and a frequent incidental autopsy finding, many surgical reports suggest that these lesions are associated with a significant risk of hemorrhage, seizures, or focal neurological deficit. This suggestion has resulted in much discussion and confusion regarding the appropriate management of these vascular lesions, particularly those that are found incidentally on neurodiagnostic studies. Lack of documented information regarding the true natural history of venous angiomas has further hampered a rational assessment of treatment options.

The purpose of this study was to identify and review a series of patients with radiographically identifiable venous angiomas in an attempt to gain information on the natural history of this particular vascular lesion.
a 0.5-tesla MR imaging system;† after that date, MR images were obtained at 1.5 tesla. All MR imaging performed after July 6, 1988, was carried out both before and after the intravenous administration of gadolinium-diethylenetriaminepenta-acetic acid at a dose of 0.1 mmol/kg. Standard angiographic, CT, or MR imaging criteria were used to diagnose a venous angioma.

Following radiographic identification of a venous angioma, hospital charts and any additional radiographic or pathological studies from these patients were reviewed, and telephone and office interviews with the patients were made to obtain additional clinical information. Special note was made of a history of headache, seizures, intracranial hemorrhage, or other focal neurological deficits. The quality of the symptoms or signs was determined, including their occurrence or variation during pregnancy. Assessment of the relationship of the symptom or sign to the venous angioma was based on the location of the lesion and the results of diagnostic or pathological studies.

Descriptive statistics (means, standard errors of the means, and frequencies) of patient characteristics were calculated. Incidence rates of hemorrhage were estimated utilizing the total person-years of follow-up review for this series of patients.

Results

A total of 4340 cerebral angiograms, 25,600 CT scans, and 8200 MR images performed during the 14-year study period were reviewed, and 100 patients (48 males and 52 females) were identified whose studies met the stringent radiographic criteria for the diagnosis of venous angioma. The mean patient age at the time of initial diagnosis (± standard error of the mean) was 42.9 ± 1.8 years (range 1 to 92 years). The locations of the angiomas are shown in Table 1; 50 of the hemispheric lesions were on the right, 47 on the left, and three in the brain stem.

† Magnetic resonance imaging systems manufactured by Picker International, Highland Heights, Ohio.

The majority of patients had undergone multiple diagnostic studies: CT in 80 patients, MR imaging in 58, arteriography in 30, and electroencephalography (EEG) in 43. Lesions in the first 50 patients in this consecutive series were diagnosed primarily by CT and arteriography; lesions in the second 50 primarily by high-field MR imaging. Computerized tomography was the only diagnostic test performed in 21 patients, MR imaging the only diagnostic test in 34 patients, and arteriography the only diagnostic test in eight patients.

Significant intracranial lesions other than the venous angioma were identified in 18 patients: cavernous angioma in two, arteriovenous malformation in one, arachnoid cyst in two, Chiari malformation in two, infarct in four, primary brain tumor in six, and a persistent trigeminal artery in one.

Thirty-six patients had suffered significant headaches and in four of those a causal relationship with the venous angioma could not be excluded. A seizure disorder was recorded for 23 patients; on the basis of the focality of seizures, the EEG localization, and the lack of other lesions, a possible relationship of the seizures to the venous angioma was believed to exist in five of them. Focal neurological deficits, usually transient and ranging from dizziness to hemiparesis, were recorded for 41 patients; a possible causal relationship with the venous angioma was suspected in eight of these. Intracranial hemorrhage had occurred in six patients; however, the hemorrhage had apparently been due to the venous angioma in only one of them. Fifteen patients exhibited no signs or symptoms that could be attributable to their venous angiomas. A total of 68 uncomplicated pregnancies and deliveries had occurred in 39 patients without apparent change in reported cerebral signs or symptoms.

No patient in this series underwent resection of the venous angioma. Neurological sequelae have been managed nonoperatively in all but one patient. The one

### TABLE 1

<table>
<thead>
<tr>
<th>Location of Lesion</th>
<th>No. of Lesions</th>
<th>Total</th>
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</thead>
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<tr>
<td></td>
<td>Left Side</td>
<td>Right Side</td>
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<tr>
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<tr>
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</tr>
<tr>
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<td>2</td>
<td>0</td>
</tr>
<tr>
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<td>7</td>
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<tr>
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<td>5</td>
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<td>0</td>
</tr>
<tr>
<td>total lesions</td>
<td>47</td>
<td>50</td>
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### TABLE 2

<table>
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<th>Location &amp; Type</th>
<th>No. of Angiomas</th>
<th>No. Ruptured</th>
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<tr>
<td></td>
<td>location of lesion</td>
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</tr>
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<td>11</td>
</tr>
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<td>4</td>
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<tr>
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<td>5</td>
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<td>1</td>
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<td>1</td>
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<td></td>
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<td></td>
</tr>
<tr>
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<td></td>
</tr>
<tr>
<td>cavernous</td>
<td>21</td>
<td></td>
</tr>
</tbody>
</table>

* Data derived from a consecutive autopsy series of 7007 patients (WF McCormick, personal communication, 1987). Of the 309 patients with angiomas, 16 had multiple angiomas, resulting in 327 lesions.
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exception had suffered a hemorrhage, and a ventriculo-peritoneal shunt had been placed.

The mean period from diagnosis to last contact was 2.46 years; the mean patient age at last contact or death was 45.3 ± 1.86 years (range 3 to 94 years). Eleven patients had died at the time of last contact: two from central nervous system (CNS) tumors, six from tumors unrelated to the CNS, and three from miscellaneous causes. Six patients were disabled due to problems unrelated to the venous angioma; all others were doing well at last contact.

Discussion

The ideal management of patients with intracranial venous angiomas has been confused by the wealth of conflicting information regarding the incidence of complications associated with this vascular malformation. Before improved neurodiagnostic imaging techniques were available, venous malformations were frequently recognized only at autopsy or following the development of a significant complication, the latter occurrence leading to the belief that complications secondary to venous angiomas were frequent. However, the advent of CT and MR imaging has facilitated the pre-mortem diagnosis of these lesions, allowing the acquisition of information regarding their true natural history. Utilizing data from this series as well as from other reported series, we will discuss five specific areas: the incidence, pathology, diagnosis, complications, and management of venous angiomas.

Incidence of Venous Angiomas

Venous angiomas appear to be among the most common intracranial vascular malformations. In early autopsy series, such as that published in 1968 by McCormick, et al.,13 venous angiomas accounted for 15% of vascular malformations. With the increasing recognition of this lesion, Sarwar and McCormick12 concluded in 1978 that venous angiomas were the most common intracranial vascular malformation, accounting for 63% of the 165 malformations found in 4069 consecutive autopsies. A more recent autopsy review by WF McCormick (personal communication, 1987) showed that venous angiomas represented 205 of the 327 vascular lesions identified (Table 2). Venous angiomas appear to be the most common vascular malformation identified on radiographic studies as well. A recent review of over 8200 craniospinal MR images performed at our institution yielded identification of 50 venous angiomas, 33 cavernous angiomas, and 17 arteriovenous malformations. Indeed, the incidental finding of a venous angioma on high-field MR studies appears to be quite high. It is likely that the incidence of these perceived lesions will continue to increase concomitantly with the increased use of CT and MR imaging.

Pathological Findings

In 1887, Pfannenstiel17 first described an intracranial vascular malformation composed entirely of veins. In 1963, Courville5 delineated the morphological detail of 22 small venous angiomas found at autopsy. Using a silver stain, he confirmed their anomalous venous character and described the "venous shunt," which subsequently has been recognized as the draining vein of the lesion.5 McCormick12 described the classic anatomical features of venous angiomas: a composition entirely of veins with interspersed neural parenchyma and commonly thickened and hyalinized veins, having minimal smooth muscle and elastic tissue. These features distinguish venous angiomas from the other commonly recognized vascular malformations: arteriovenous malformations, cavernous angiomas, and capillary telangiectasias.

Venous angiomas appear to be congenital malformations or anomalies of normal venous drainage. This is supported by the finding of venous angiomas in infants and children (in this series, venous angiomas were diagnosed in eight patients before the age of 16 years, the youngest of whom was 18 months old). This assumption regarding the congenital nature of venous angiomas is also supported by the complete absence of normal draining veins in the region of the malformation as seen at autopsy, in surgery, and on angiography. Further evidence supporting the role of venous drainage in these malformations is provided by the important observation that venous infarction of the adjacent brain occurs following removal of the venous angioma.24 Indeed, some authors have suggested that the name "venous angioma" be changed to "developmental venous anomaly" to reflect this characteristic more precisely.10 Others prefer to use the term "venous malformation," again making an attempt to define the anomalous, rather than the neoplastic, character of the lesion.

The derivation or etiology of venous angiomas remains unclear. Saito and Kobayashi11 have suggested that an intrauterine accident occurs during the formation of the medullary veins or their tributaries, either by thrombosis or by some other unknown mechanism, and a collateral pathway is then formed to drain the area of the brain so affected. It is clear from Padget's work16 on the development of the circulatory system that abnormal development at the appropriate gestational stage could result in an anomalous draining vein supplementing the usual drainage. It appears then that venous angiomas are venous anomalies that are present at birth and that remain largely static throughout life. Additional long-term monitoring with CT or MR imaging should help to confirm this assumption.

Diagnostic Neuroradiological Studies

Angiographic criteria used to diagnose a venous angioma were similar to those previously described.3,6,29 In the late venous phase, dilated medullary veins are seen converging on a large central vein, which drains either centripetally to the deep venous system or cen-
and a hypo- or hyperintense signal is seen on T2-weighted sequences.5,9,18,26 (Figs. 3 and 4).

**Complications**

Vascular malformations of the brain are known to be associated with, and occasionally responsible for, headaches, seizures, hemorrhages, and focal neurological deficits. However, the relative frequency of those complications appears to vary with the pathological type of vascular malformation. Attempts to ascertain the risks associated with a particular malformation (the natural history of the lesion) have been hampered by the inability to diagnose and follow the lesion before the development of significant complications. The issue has been further confused by the frequent misnaming or misdiagnosing of malformations. This is demonstrated by the finding that many of the "venous angiomas" identified in the earlier literature were subsequently proved to be arteriovenous malformations.5 This situation has resulted in the misconception that venous angiomas are associated with a particularly high incidence of neurological complications.

**Hemorrhage.** Hemorrhage has been reported as a relatively frequent complication of venous angiomas. Although hemorrhage from arteriovenous malformations is distinctly more common, McCormick et al.13 found in a large autopsy series that three (8%) of 39 venous angiomas in the posterior fossa and two (17%) of 12 venous angiomas above the tentorium had hemorrhaged. Sarwar and McCormick22 reported three cases of subarachnoid hemorrhage and one of intracerebral hemorrhage (29%) among 14 patients with angiographically proven venous angiomas. Reviewing the literature in 1985, Handa and Moritake6 found a 17% incidence of intracerebral hematoma. Malik, et al.,11 reported an even higher incidence of hemorrhage, with nine (43%) of 21 venous angiomas having bled. Other reports have

![Fig. 1. Cerebral arteriograms demonstrating the typical appearance of a venous angioma in the right frontal region (A and B) and a more subtle venous angioma in the left posterior frontal region (C and D). A: Arteriogram, lateral projection in the late venous phase, showing a large dilated medullary vein (arrow) draining anteriorly toward the anterior sagittal sinus. B: Arteriogram, anteroposterior projection, demonstrating the "caput medusae" of the lesion (arrow); the confluence of multiple dilated medullary veins converging on the draining vein. C: Arteriogram, lateral projection in the late venous phase, showing several "curved" veins draining into the sagittal sinus superiorly, typical of cortical surface veins. One large vein (arrow) appears to penetrate straight down toward the ventricle, consistent with a venous angioma. D: Arteriogram, anteroposterior projection, showing the venous angioma draining the subependymal region of the left lateral ventricle (arrow), coursing superiorly toward the sagittal sinus. Trifugally to cortical veins or dural sinuses; the arterial phase and circulation times are usually normal (Fig. 1). Computerized tomographic criteria for the diagnosis of venous angioma are as follows. On the contrast-enhanced CT scan, the lesion appears as a linear, transcerebral, enhancing density entering either the deep venous system, a major dural sinus, or a cortical vein. The unenhanced scan usually appears normal, although it may occasionally demonstrate a rounded density in the white matter. There should be no associated mass or edema and no other major vessels in the region of the malformation (Fig. 2). The "caput medusae" of the lesion (the confluence of the multiple dilated medullary veins converging on the draining vein) occasionally may appear as a rounded density adjacent to the draining vein.6,14,27 Magnetic resonance imaging criteria for the diagnosis of venous angioma are similar to those for CT. A tubular area of decreased signal intensity appears in the white matter of the brain on T2-weighted MR images, and a hypo- or hyperintense signal is seen on T2-weighted sequences.5,9,18,26 (Figs. 3 and 4).
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Fig. 3. Magnetic resonance images demonstrating the typical appearance of a venous angioma in the right cerebellum. A: Sagittal T1-weighted image without contrast showing a subtle hypointense linear signal (arrow) in the cerebellum. B: Sagittal T1-weighted image following administration of intravenous contrast medium (gadolinium-diethylenetriaminepenta-acetic acid) revealing an enhancing linear structure (arrow) coursing toward the surface, apparently formed by the confluence of several smaller enhancing structures centrally. C: Axial T1-weighted image with contrast medium demonstrating the confluence of smaller vessels (the "caput medusae") and the larger draining vein centrifugally (arrow). D: Axial T1-weighted image showing a mixed hypointense-hyperintense signal (arrow) corresponding to the venous angioma.

suggested that the risk of hemorrhage is even greater during pregnancy\textsuperscript{11} or with lesions in the posterior fossa;\textsuperscript{11,19,20,28} neither of these suggestions is supported by the results of our study.

In our series of 100 patients with venous angiomas, hemorrhage secondary to the lesion was distinctly uncommon, occurring in only one patient. This patient was a 15-year-old girl who developed a generalized headache while singing in a choir and quickly became comatose. The initial CT scan showed a left parietal intracerebral hemorrhage with associated intraventricular hemorrhage (Fig. 5); subsequent scans showed resolution of the hemorrhage but development of hydrocephalus. Cerebral angiography performed 4 weeks after the hemorrhage demonstrated a left parietal venous angioma. The patient was treated with a ventriculoperitoneal shunt and has continued to do well without evidence of further hemorrhage during a 5-year follow-up period. Recent CT scans and MR images show no change in the appearance of the angioma (Fig. 6).

Assuming that venous angiomas are congenital lesions and that the risk of hemorrhage is equal throughout the life of the individual, the risk of hemorrhage for our patient population can be estimated to be 0.22%/year (one hemorrhage per 4498 person-years of follow-up period). The great discrepancy in incidence and risk of hemorrhage between our patient population and those of previously reported series is presumably explained by the inclusion in our series of all patients with identifiable venous angiomas, and not just those presenting after the development of significant complications that led to diagnosis. As our population represents a selected group (those patients undergoing neurodiagnostic studies at a university medical center), extrapolating these figures to the general population should yield an even lower risk estimate, allowing for the incorporation of greater numbers of asymptomatic individuals.

Fig. 4. Magnetic resonance (MR) images demonstrating a venous angioma in the left posterior frontal region. These MR images correspond to the arteriograms seen in Fig. 1C and D. Left: Axial T1-weighted image, with contrast enhancement, showing only a small round hyperintense signal (arrow) in the white matter. This represents the "end on" appearance of the draining vein of the venous angioma. Right: The venous angioma (arrow) is better seen in this MR image in the coronal plane.

Seizures. The role of venous angiomas in the initiation of seizures is more difficult to ascertain and understand. Of the 23 patients presenting with seizures in this series, there were five in whom a possible relationship with the malformation (or surrounding parenchyma) was suspected. This presumed relationship was based primarily on the localization of the EEG findings or of the clinical features of the seizure to the region of the venous angioma and on the absence of other lesions in that region. Surgical resection of the lesion was not performed in any of these patients, and all have been managed successfully with antiepileptic medications.

Focal Deficits. Focal neurological deficits were also common in this population, being identified in 41
patients either at presentation or at follow-up examination. As with seizures, it is difficult to ascertain the relationship of the venous angioma to the neurological deficit; however, there were eight patients in this series in whom a potentially causal relationship was initially suspected. No patients were debilitated by their focal deficits, and most no longer reported their deficits at the time of last contact.

**Headaches:** Headaches were also reported frequently in this population undergoing diagnostic cranial MR imaging. Although the relationship is difficult to assess, venous angiomas were implicated as a possible cause of the headache in only four of the 36 patients.

**Management of Venous Angiomas**

Primarily on the basis of the previously perceived high incidence of hemorrhagic complications associated with venous angiomas, resection of all surgically accessible venous angiomas has been recommended by some. We are opposed to that approach because of the relatively benign natural history of the lesion and our belief that venous angiomas represent anatomically disordered but physiologically essential venous drainage of the surrounding brain. This concept is well demonstrated by the patient reported by Senegor, et al., who underwent uneventful resection of a posterior fossa venous angioma, but died 4 days postoperatively; au-

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**FIG. 5.** Computerized tomography (CT) scans of the one patient in the current series who presented with intracerebral hemorrhage secondary to a venous angioma. **Upper:** Axial CT scans, without contrast enhancement, revealing a large hematoma in the left parieto-occipital area, with extension into the ventricular system and resultant hydrocephalus. **Center:** Axial CT scans, following administration of intravenous contrast medium, demonstrating a linear enhancing structure coursing from the region of the hematoma toward the surface, consistent with the appearance of a venous angioma. **Lower:** Follow-up axial CT scans obtained approximately 4 weeks later revealing encephalomalacia in the region of the previous hemorrhage, persistent hydrocephalus, and the venous angioma. Arteriography confirmed the presence of a venous angioma.
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topsy revealed a venous infarction of the brain stem and cerebellum.

Clearly, resection of uncomplicated or asymptomatic venous angiomas is not indicated, although the rare occurrence of hemorrhage may necessitate the removal of a hematoma to relieve intracranial hypertension. In such instances, the decision of whether to disrupt or resect the angioma itself is more of a problem. Although insufficient information exists as to the risk of rebleeding with venous angiomas, that risk seems to be quite low. Thus, the most prudent approach appears to be to leave the angioma intact, unless the location is such that resection of the lesion and surrounding brain will not result in unacceptable neurological deficits. This approach has been advocated by others as well.\textsuperscript{2,10,23,25}

Reporting a series of 11 patients with venous angiomas, Numaguchi, \textit{et al.},\textsuperscript{13} included two with posterior fossa hemorrhage; all patients were managed without resection of the lesion, the result being neurological improvement in six patients and no change in five. As previously stated, our series suggests that the risk of hemorrhage during pregnancy is not significant and does not warrant the surgical resection of venous angioma.

Conclusions

Venous angiomas are frequent serendipitous findings on neurodiagnostic studies. We present a series of 100 patients with radiologically identifiable venous angiomas and review their clinical course. Hemorrhage secondary to the lesion occurred in only one patient (1%); seizures and focal neurological deficits were unusual associated findings. This study supports the concept that the natural history of venous angiomas is relatively benign, and therefore surgical resection of these venous anomalies is rarely indicated.

References

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Manuscript received August 14, 1990. Accepted in final form April 29, 1991.
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