The Case for Conservative Management of Venous Angiomas

Douglas Kondziolka, Peter K. Dempsey and L. Dade Lunsford

ABSTRACT: Venous angiomas (developmental venous anomalies) are vascular malformations increasingly recognized in general neurosurgical or neurological practice. They are associated with intracranial hemorrhage, seizures, or progressive neurological deficits or found as incidental findings in patients who present with headaches or have neurological imaging studies for investigation of unrelated neurological disorders. Since venous angiomas drain normal cerebral tissue within a functionally normal arterial territory, resection can lead to venous infarction. This report studies 27 patients with venous angiomas, all of whom had conservative treatment. The venous angioma was considered to be responsible for the onset of neurological symptoms in 14 patients (7 with hemorrhage, 3 with hemorrhage and seizures, 2 with seizures, one with an extrapyramidal movement disorder, and one with motor deficit). Thirteen patients had incidental lesions (8 with headache, and 5 with unrelated neurological symptoms). Ten venous angiomas were in the posterior fossa; seven in the cerebellum. Location did not correlate with symptomatic presentation. No patient with hemorrhage required surgical evacuation of the hematoma. No patient died or had significant morbidity during the follow-up interval (mean of 3.7 years). Venous angiomas are low flow, low resistance vascular malformations, many of which are not associated with neurological sequelae. Our series supports the concept that surgical removal or radiosurgical obliteration should not be performed unless a patient has a second life threatening hemorrhage.

RÉSUMÉ: En faveur du traitement conservateur des angiomes veineux. Les angiomes veineux (anomalies du développement veineux) sont des malformations vasculaires de plus en plus souvent découvertes en pratique neurochirurgicale ou neurologique générale. Ils sont associés à l'hémorragie intracrânienne, aux convulsions ou à des déficits neurologiques progressifs ou sont une découverte fortuite chez des patients qui se présentent pour céphalée ou qui subissent des examens de neuro-imagerie lors d'investigations d'affections neurologiques sans lien avec cette malformation. Comme les angiomes veineux drainent du tissu cérébral normal dans un territoire artériel fonctionnant normalement, leur résection peut mener à l'infarctus veineux. Ce compte rendu étudie 27 patients avec angiome veineux qui ont tous été traités de façon conservatrice. L'angiome veineux a été considéré comme responsable du début des symptômes neurologiques chez 14 patients (7 patients avec hémorragie, 3 avec hémorragie et convulsions, 2 avec convulsions, un avec un désordre du mouvement de nature extra-pyramydale et un avec un déficit moteur). Treize patients avaient des lésions incidentes (8 avec céphalée et 5 avec des symptômes neurologiques non reliés). Dix angiomes veineux étaient situés dans la fosse cérébrale postérieure; sept dans le cervelet. La localisation n'était pas corrélée avec la symptomatologie qui avait amené le patient à consulter. Aucun patient qui a subi une hémorragie n'a requis une évacuation chirurgicale de l'hématome. Aucun patient n'est décédé ou n'a eu une morbidité significative pendant l'intervalle du suivi (moyenne de 3.7 ans). Les angiomes veineux sont des malformations qui ont un flux sanquin et une résistance vasculaire faibles, et plusieurs ne sont pas associés à des séquelles neurologiques. Notre série supporte le concept qu'une exérèse chirurgicale ou une obliteration radiochirurgicale ne devrait pas être effectuée à moins qu'un patient ne subisse une deuxième hémorragie menant sa vie en danger.

major post-operative morbidity has been reported but other studies indicate a high incidence of partial resection, venous infarction and mortality. The present report details the conservative management of 27 patients with venous angiomas. Venous angiomas are not symptomatically innocuous in all patients, and although not of high-flow, can be the cause of intracranial hemorrhage. We believe that symptoms alone should be treated, because of the risks associated with obliteration of the venous angioma itself. This report summarizes our experience in the evaluation and conservative management of patients with venous angiomas.

**MATERIALS AND METHODS**

From August 1987 to August 1990, 27 patients with venous angiomas were referred to the Specialized Neurosurgical Center at the University of Pittsburgh, usually for consideration of stereotactic radiosurgery. These 27 patients represented a subset of over 750 patients with cerebral vascular malformations reviewed at our multi-disciplinary arteriovenous malformation (AVM) conference during this three year interval. All patients were reviewed by specialists in neurosurgery, neuroradiology and radiation oncology. Clinical information and pertinent neuroimaging studies were required for review.

Fourteen patients were male (mean age = 37 years; range = 6-72), and 13 were female (mean age = 34 years, range = 15-56). The locations of venous angiomas are detailed in Table 1. Nine were cortical, one was subcortical, seven were diencephalic, and ten were located in the posterior fossa. Conservative management (ie. neither neurosurgery nor radiosurgery) was recommended in all patients. Follow-up clinical information was obtained by phone conversation with patients or their referring physicians. Clinical observation under the supervision of a neurologist or neurosurgeon ranged from 3 months to 26 years (average of 3.7 years), totalling 100 patient-years. These patients are being studied longitudinally to provide information on the long-term natural history of venous angiomas.

**RESULTS**

**Clinical Presentation**

The clinical presentations of all patients are detailed in Table 2. All patients presenting with chronic headaches only were considered to have incidental venous angiomas (n = 8). An additional five patients had incidental venous angiomas found in the context of an evaluation of various neurological symptoms: anxiety disorder and depression, syncope, hearing loss, or cerebral ischemia. Six patients had focal neurological deficits, of which four were related to prior intracranial hemorrhage. Two patients had a history of chronic headache prior to their presentation with hemorrhage. Ten patients (36%) sustained at least one intracranial hemorrhage. The hemorrhage could not be correlated with any clinical episode likely to increase intracranial venous pressure. Two patients were diagnosed during pregnancy, but neither had bled.

**Neuroimaging Evaluation**

Diagnosis was confirmed by cerebral angiography in 24 patients and by MRI in three patients. Cerebral angiograms carried out into the late venous phase showed evidence of a radiating collection of medullary veins deep within the brain parenchyma (Figure 1). Angiography was diagnostic of a venous angioma in all patients studied.

MRI was performed in 26 patients; a characteristic venous angioma was identified in 20. The appearance on MRI was usually that of a tubular hypointensity corresponding to the draining vein, radially surrounded by low signal intensity.

**Table 1: Location of Venous Angiomas (n=27)**

<table>
<thead>
<tr>
<th>Location</th>
<th>Number of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebellum</td>
<td>7 (25%)</td>
</tr>
<tr>
<td>Pons</td>
<td>2 (7.1%)</td>
</tr>
<tr>
<td>Quadrigeminal Plate</td>
<td>1 (3.6%)</td>
</tr>
<tr>
<td>Basal Ganglia</td>
<td>5 (17.9%)</td>
</tr>
<tr>
<td>Thalamus</td>
<td>2 (7.1%)</td>
</tr>
<tr>
<td>Frontal Lobe</td>
<td>6 (21.4%)</td>
</tr>
<tr>
<td>Medial Temporal Lobe</td>
<td>2 (7.1%)</td>
</tr>
<tr>
<td>Medial Parietal Lobe</td>
<td>1 (3.6%)</td>
</tr>
<tr>
<td>Centrum Semiovale</td>
<td>1 (3.6%)</td>
</tr>
</tbody>
</table>

**Table 2: Clinical Characteristics of Patients with Venous Angiomas (n=27)**

<table>
<thead>
<tr>
<th>Presentation</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemorrhage (single episode)</td>
<td>5</td>
</tr>
<tr>
<td>Hemorrhage (two episode)</td>
<td>2</td>
</tr>
<tr>
<td>Hemorrhage and Seizures</td>
<td>3</td>
</tr>
<tr>
<td>Seizures</td>
<td>2</td>
</tr>
<tr>
<td>Neurological deficit without hemorrhage</td>
<td>2</td>
</tr>
<tr>
<td>Incidental:</td>
<td></td>
</tr>
<tr>
<td>Headache</td>
<td>8</td>
</tr>
<tr>
<td>Stroke</td>
<td>2</td>
</tr>
<tr>
<td>Syncope</td>
<td>1</td>
</tr>
<tr>
<td>Hearing Loss</td>
<td>1</td>
</tr>
<tr>
<td>Anxiety/Depression</td>
<td>1</td>
</tr>
</tbody>
</table>

Figure 1 — Lateral subtraction angiogram (mid-venous phase) showing a venous angioma (narrow arrows). The radially arranged venous channels (caput medusa) drain into a single trans-hemispheric vein (wide arrow).
(Figure 2). With gadolinium-DTPA enhancement, a starburst pattern of deep white matter veins was seen. In six patients, MRI was consistent only with the diagnosis of a vascular malformation without further classification. This usually showed a heterogeneous lesion of mixed signal intensity on short and long TR sequences, without delineation of specific venous channels. CT scan was performed in 16 patients; a diagnosis of venous angioma (radial arrangement of venous channels in the deep white matter after administration of contrast) was suggested in eight (Figure 3). Contrast-enhanced CT scan in the eight remaining patients showed only a circumscribed area of increased density. MRI tended to have greater specificity than CT in confirming the diagnosis of venous angioma.

Conservative Management

During the follow-up interval of 100 patient-years, no patient died or developed new neurological morbidity. Headache did not change in those patients presenting with this symptom; no patient developed the new onset of headache during follow-up. Seizures were controlled with anti-convulsant medication; no patient developed intractable epilepsy. Two patients sustained a second hemorrhage during follow-up. One patient with re-hemorrhage from a venous angioma of the pons had temporary worsening of facial paresthesiae. A second patient with a caudate nucleus malformation suffered a small new hemorrhage identified on a follow-up imaging study; this patient remained asymptomatic.

DISCUSSION

Developmental Anomalies of Venous Drainage

The developmental absence of normal venous drainage for a region of normal brain tissue can lead to the embryological formation of a venous angioma in that region.\(^5,16,18,22\) These anomalous vascular channels may be structurally weaker than normal cerebral veins. The normal venous outflow of the brain represents a balance between the superficial and deep venous drainage systems in both the supratentorial and infratentorial compartments. Anastomotic channels exist between the deep and superficial systems.\(^10,16\) Venous angiomas are located at the junction between the superficial and deep venous systems, and perhaps represent transhemispheric anastomotic pathways. Noran noted that venous angiomas have a pyramidal shape with the apex toward a ventricle.\(^23\) Venous angiomas often represent the dominant venous collection system of a specific brain.
region, without the presence of significant anastomoses in the brain outside the lesion. Their drainage is usually into a normal venous sinus or bridging vein.16

Although venous angiomas of the posterior fossa have been considered uncommon,15 a predisposition for location in the cerebellum has been noted by many authors.5,9,12,15,17 In the posterior fossa, these lesions are commonly located at the junction between the superficial and deep venous systems of the cerebellum. Rarely, venous angiomas are present only in the deep system. Goulao et al described five routes of drainage for posterior fossa venous angiomas: the deep system by a trans-pontine vein, the vein of the lateral recess, the pre-central vein, the superficial system by the vermian vein, and the hemispheric cerebellar veins.5 Matsushima et al studied the microsurgical anatomy of the posterior fossa venous system and described three groups of drainage: the Galenic system, the petrosal system, and the torcular system via the tentorial sinuses.10 Neurological sequelae after surgical venous occlusion were believed to be infrequent, due to the presence of diffuse anastomotic channels in this region. However, many such channels are subarachnoid in location and not parenchymal.

Imaging Characteristics of Venous Angiomas

The characteristic angiographic appearance of venous angiomas is a radiating collection of medullary veins deep within the brain parenchyma.19 These veins drain centrally into one or more larger veins that terminate in a superficial cortical vein or deep sinus.24 They are best visualized during the early to mid-venous and occasionally late venous phases with normal circulation time.2

The characteristic appearance of the caput medusa can be visualized on CT and MRI scans as well as on angiograms. The usual appearance on CT scan is that of a round distinct area of contrast enhancement.17,25 Agnoli and Hildebrandt reported a series of 15 patients with venous angiomas and found that 6 had a normal CT scan.26 The MRI appearance of an “angiographically occult vascular malformation” can include patent or thrombosed arteriovenous malformations, cavernous angiomas and venous angiomas.1 As a result, for lesions being considered for surgical resection or radiosurgery, angiography is mandatory in order to exclude the specific diagnosis of a venous angioma.

Clinical Presentation and Natural History

The natural history of venous angiomas is not known. No estimated risks for hemorrhage have been made, and large clinical series are uncommon. Saito reviewed the literature and found only 24 angiographically diagnosed venous angiomas from 1968 to 1981.22 Sarwar and McCormick reported 105 venous angiomas in 4,069 autopsies (2.6%).27 Lobato et al reported 241 angiographically occult vascular malformations; 26 (9.9%) were venous angiomas.1 Although, many reports have stressed the incidental and innocuous nature of venous angiomas, especially in regard to causation of headache, several reported cases confirm the risk of intracranial hemorrhage.

The etiology of seizures in relation to venous angiomas is controversial. Although focal areas of microhemorrhage surrounding the lesion could be epileptogenic, such a theory is not well substantiated. To date no evidence supports the concept that ischemic phenomena result from untreated venous angiomas. Saito reported a comparison study between the electroencephalographic localization of seizures and venous angioma location; in eight out of eleven patients, the locations did not match.22

In the present series, two patients presented with neurological syndromes in the absence of hemorrhage. One had an extrapyramidal movement disorder associated with a venous angioma in the left caudate nucleus, and one had a contralateral hemiparesis in association with a lesion in the posterior internal capsule. The mechanism of production of neurological symptoms in these patients remains elusive, since repeat imaging studies failed to discover any evidence of hemorrhage or mass effect.

In this series, chronic headache was considered an incidental symptom in all patients. The location or hemodynamics of the malformation would be unlikely to cause headache. In no patient did angiography suggest a high-flow lesion, and in no patient did the venous angioma abut on sensory-innervated structures.

The Role of Surgical Resection

Surgical resection has been advocated for the treatment of surgically accessible venous angiomas, usually after intracerebral hemorrhage.1,7,9,11-13,28 Malik et al resected ten venous angiomas in a series of 21 patients. Nine had prior hemorrhage.9 They were able to achieve complete resection in 3 patients, and concluded that surgical extirpation should be considered in all patients with cerebral venous angiomas associated hemorrhage. Although they felt that an asymptomatic lesion should be treated conservatively, they suggested that surgery should be considered in women of child-bearing age.
In contrast to reports of surgical resection with minimal morbidity, Biller et al reported surgical removal of a cerebellar venous angioma in a patient who died from venous infarction of the brainstem and cerebellum. Senegol et al described a patient who died four days after surgical resection of a venous angioma of the cerebellum and fourth ventricle. Sadah et al detailed the case of a patient who died four months after resection of a venous angioma of the medulla; this patient underwent surgery in poor neurological condition after multiple hemorrhages. Cabanes et al reported the successful removal of a frontal lobe venous angioma via a frontal lobectomy. This may have been successful due to the removal of a large region of normal brain tissue that represented the venous drainage region of the malformation.

In the present series, no patient was found to have the association of an angiographically occult vascular malformation (AOVM) adjacent to a venous angioma. The developmental relationship between these two lesions is not understood. If the AOVM is found to be the cause of symptomatic hemorrhage, we would consider treatment of this malformation only. The adjacent venous angioma should be left intact.

The majority of patients in the present series were referred for consideration of stereotactic radiosurgery. Agnoli and Hildebrandt suggested that radiation might be useful in the treatment of venous angiomas; no evidence substantiates this hypothesis. Although we have treated patients with angiographically occult vascular malformations who sustained two or more hemorrhages, in all patients the diagnosis of venous angioma was excluded by high-resolution angiography. In the absence of normal venous anastomotic channels, we believe that delayed radiosurgical occlusion of venous angiomas likely entails the same risk of venous infarction and neurological deficit as microsurgical removal.

Conservative non-microsurgical, non-radiosurgical management should remain the present management strategy for venous angiomas. Venous angiomas represent developmental anomalies of venous drainage for normally functioning brain tissue. Surgical resection should be considered only in patients who sustain recurrent symptomatic hemorrhage in association with progressive neurological deterioration.

REFERENCES