Cerebellar venous angioma: "benign" entity?

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Cerebellar Venous Angioma: "Benign" Entity?

Four patients with bleeding venous angiomas in the cerebellar hemispheres are reported. All of the hemorrhages were subacute; three were recurrent. A literature review substantiated venous angioma as a recognized source of spontaneous cerebellar hemorrhage. Cerebellar venous angiomas may be more prone to bleed than venous angiomas in other locations. Their natural history seems to be similar to that of arteriovenous malformations. Though a minority bleed, the hemorrhagic event can be acute or subacute, recurrent, or catastrophic. Thus the clinical course is not always innocuous.

Venous angiomas are vascular malformations in which veins are the predominant vascular constituent. They were described as distinct clinical entities by Cushing and Bailey [1] in 1928 and since then have been reported sporadically. Because venous angiomas are infrequently symptomatic, their clinical significance remains unclear. When symptomatic, they are associated mainly with seizures or bleeding, although bleeding is considered to occur rarely [2]. During the past 2 years we have examined four patients with cerebellar venous angiomas who had intracerebellar bleeding. We describe the clinical and radiographic features of these cases, and review the pertinent literature to help clarify the natural history of these lesions.

Case Reports

Case 1

A 22-year-old woman had right-sided frontooccipital headaches that began abruptly and persisted for 4 days. When examined, she had mild papilledema; mild appendicular ataxia, especially of the right leg; dysdiadochokinesia of the right arm; and decreased tone in the right extremities. A computed tomographic (CT) scan without contrast enhancement revealed a moderate-sized right cerebellar hematoma. After contrast administration, a curvilinear enhancing structure was identified along the posterior border of the lesion (figs. 1A and 1B). A cerebral angiogram demonstrated right cerebellar mass effect in the arterial phase and a branching collection of enlarged and tortuous medullary veins in the venous phase (fig. 1C), consistent with a venous angioma. Eight days after the headaches began, she became confused and had a right hemiparesis, right seventh nerve paresis, and right gaze paresis. A repeat CT scan showed enlargement of the hematoma from new hemorrhage and hydrocephalus (fig. 1D). An operation was performed immediately. A large cerebellar hematoma was evacuated, the venous angioma was removed, and a ventriculostomy was inserted. Postoperatively, she had residual right-sided dysmetria and dysdiadochokinesia. A postoperative CT scan revealed no residual angioma. The pathology specimen was a vascular malformation composed predominantly of veins. The occurrence of previous and recent hemorrhage around and within the lesion was confirmed by the presence of hemosiderin and hematoidin.
Case 2

A 12-year-old boy was admitted after 2 weeks of progressive headaches, vertigo, ataxia, and vomiting. He also had visual blurring and right arm weakness. He was lethargic, with papilledema, right lateral-gaze nystagmus, right-sided dysmetria and dysdiadochokinesia, and a positive Romberg test. CT revealed an inhomogeneous, hyperdense right cerebellar hemispheric mass. The posterior aspect of the mass enhanced as a curvilinear, branching structure (fig. 2). Moderate hydrocephalus was present. A ventriculoperitoneal shunt was inserted and his ventricles diminished, but his clinical condition did not improve substantially. A craniectomy was performed, and a large hematoma and venous angioma were removed. Postoperatively, his recovery was uneventful and his only neurologic deficit was transient right arm dysmetria. Histologically, the specimen was a venous angioma with evidence of recent and previous hemorrhage. Postoperative CT showed no residual angioma.

Case 3

A nonhypertensive 57-year-old man was evaluated after 1 month of occipital headache, diplopia, and ataxia. On examination, he had right lateral-gaze nystagmus. A CT scan without contrast enhancement revealed an inhomogeneous posterior fossa mass. After contrast administration, fairly well defined regions of enhancement were seen along the posterior, medial, and anterior sides of the mass (figs. 3A and 3B). A vertebral angiogram demonstrated the typical circumferential pattern of enlarged veins of a venous angioma (fig. 3C). An operation was performed, confirming the presence of a venous angioma with an associated hematoma that consisted of new and old blood. Resection was incomplete but bleeding did not recur. The patient recovered without deficit.

Case 4

A nonhypertensive 57-year-old woman was admitted for the second time in 18 months. Both admissions were to evaluate occipital headache, tinnitus, vertigo, gait ataxia, and incoordination lasting several days. Her first examination disclosed rotatory nystagmus, lateral-gaze nystagmus, left intention tremor, and a positive Romberg sign. A CT scan during the first admission showed a moderate-sized cerebellar hematoma, with branchiak enhancement at its posterior aspect (figs. 4A and 4B). An angiogram demonstrated a medusoid conglomeration of dilated medullary veins draining into a large transcerebellar vein, indicative of a venous angioma. No operation was performed. A CT scan during the second admission revealed a similarly enhancing structure at the periphery of a resolving hematoma (figs. 4C and 4D). The angiogram was unchanged (figs. 4E and 4F). Again no operation was performed and her symptoms improved. She was still stable after 11 months with ataxia that varied in severity.
Fig. 3.—Case 3. A, Noncontrast CT scan. Ringlike area of lucency (arrowheads) around resolving left cerebellar hematoma. B, Postcontrast scan. Several well-defined areas of enhancement around periphery of hematoma. C, Venous phase, after unremarkable arterial phase. Typical pattern of venous angioma. Enlarged medullary veins (small arrows) drain into large transcerebellar vein (large arrow), which ultimately drains into petrosal vein.

Fig. 4.—Case 4. A, Resolving hematoma in left cerebellar hemisphere extends to cerebellopontine angle (arrows). B, Posterior part of hematoma enhances in branching pattern (arrow). C, 18 months later. Another resolving hematoma is in exactly same location as previous one. D, Enhanced scan. Prominent branching vascular structures posterior to hematoma (arrow). Frontal (E) and lateral (F) venous phase vertebral angiograms demonstrate venous angioma. Posterosuperior aspect of angioma correlates with branching structures seen on B and D.
Discussion

Venous angiomas were once considered uncommon lesions. Until recently there were few autopsy reports of these angiomas and even fewer surgical reports [1, 3, 6]. However, large autopsy series suggest that venous angiomas are relatively common incidental findings [7, 8]. In fact, McCormick and Sarwar [9, 10] have shown them to be the most common type of angioma found at autopsy, occurring in about 3% of cases. Despite this relatively frequent occurrence, the number of reported cases of symptomatic venous angiomas is low. Most seem to be clinically silent. When symptomatic they most commonly cause seizures, and are said to bleed rarely [2].

Venous angiomas have been diagnosed more frequently since the advent of magnification angiography and CT. The angiographic hallmarks of venous angiomas are enlarged medullary veins. These veins usually converge to form a large central channel that courses transcerebrally or transcerebellar to end in a cortical vein, dural sinus, or less commonly into the deep venous system [11–18]. The arterial phase is usually completely normal, but occasionally a poorly circumscribed blush, early draining vein, or slightly enlarged arterial branch can be seen [15, 17, 18]. Of interest, a higher frequency of bleeding has been reported for angiomas with typical angiographic features (i.e., no early appearing vessels) as opposed to those with early appearing vessels [14]. The angiograms of our three cases were typical, with normal arterial phases and a caput medusa of veins merging into large central veins, which drain superficially into the petrosal vein.

CT complements angiography in the detection of venous angiomas. Several recent reports describe the appearance of venous angiomas on CT scans [13, 14, 19–26]. Enhancement is the key feature in these reports and takes the form of a distinct round or linear density. There is no mass effect, unless hemorrhage has occurred. The confluence of linear structures (veins) can be seen occasionally, and the large transparenchymal draining vein(s) may be followed on serial scan slices [20, 26]. The CT characteristics alone are not specific enough to diagnose venous angiograms, and angiography is usually required to distinguish them from arteriovenous malformations (AVMs) or small neoplasms.

CT was undeniably helpful in the evaluation of our patients. It accurately identified the location and character of the hemispheric hematomas and identified hematomas as the cause of a worsening clinical condition. Enhanced scans were most informative. In all the cases there was well defined enhancement that was distinct from the hematoma. Although this was a nonspecific finding that could have been mimicked by an AVM or tumor of the cerebellum [27, 28], findings were more specific in two cases. Cases 2 and 4 showed branching enhancing densities that were highly suggestive of vascular structures. In case 4, a large vascular structure could be identified coursing toward the petrosal sinus (not shown), suggesting a transparenchymal draining vein.

Spontaneous cerebellar hemorrhages are usually hypertensive in origin. Angiomatosmal malformations are the second most frequent cause, with aneurysms, blood dyscrasias, and infection being responsible for a minority of cerebellar hemorrhages [23, 29–37]. Hypertensive hemorrhages occur most often in the region of the dentate nucleus of one hemisphere, whereas hemorrhages from other causes occur anywhere in the hemisphere or vermis [30, 33, 34]. Angiomatous hemorrhages of any etiology (arteriovenous, venous, cavernous, etc.) are usually located deep in the cerebellar hemisphere, but may extend superficially to the cortex [33]. Hypertensive hemorrhages occur in older age groups, while angiomatous malformations are responsible for most bleeds in younger age groups [38].

Any type of angioma can bleed. In the cerebellum, the angioma that bleeds most often is the AVM. Venous angiomas, however, are second most common [8]. The actual frequency of hemorrhage in cerebellar venous angiomas is uncertain. McCormick et al. [8], in their large autopsy series of posterior fossa angiomas, found evidence of hemorrhage in three (15%) of 20 cases of cerebellar venous angioma. Considering the overall frequency of this type of angioma, hemorrhage may not be as rare as was once believed.

We made an extensive literature search for cases of bleeding cerebellar venous angiomas in order to compare them with our four cases. In all, 20 cases were found that had been documented by histology (autopsy or surgical), by angiography (typical radiologic features discussed above), or both [6–8, 14, 18, 20, 21, 23, 28, 33, 39–41]. These 20 represent a large proportion (i.e., 36%) of the total number of cerebellar venous angiomas described in the literature [2, 11, 18, 22, 23, 25, 26, 42, 43]. In addition, these 20 cases constitute almost half of the total number of reported hemorrhagic venous angiomas, both infral- and supratentorial [1, 3, 5, 11, 13, 16–18, 23, 25, 26, 44–46] (in contrast to a 3:1 predominance of cerebral venous angiomas to cerebellous venous angiomas [7, 8]). These facts imply a possible propensity of cerebellar venous angiomas to bleed and contrast with previous reports that these lesions have a benign clinical course [2, 12, 17, 42].

The 20 patients were 7–65 years old, however, most were in the younger age groups. The most common location of both angioma and hemorrhage was the cerebellum hemisphere. Symptoms sometimes occurred with catastrophic severity, sometimes slowly worsened over several days, and sometimes occurred as multiple episodes over months or years. The sporadic symptoms reported in several cases suggest that some cerebellar venous angiomas bleed subacutely and intermittently. Pathologic proof of subacute bleeding is provided in at least one of the published cases [18]. It is important to note that operative removal of the hematoma was successful in all three of the surgical cases [21, 28, 40].

Our four cases accent several points about hemorrhagic cerebellar venous angiomas. Venous angioma, although uncommon, must be considered in the differential diagnosis of spontaneous cerebellar hemorrhage in any age group. Only two out of the four patients were young. The other two were symptomatic in the sixth decade of life, an age group in which hypertensive rather than angioma would normally be considered responsible for spontaneous cerebellar hemorrhage. The initial symptoms in all of our cases included a headache, usually occipitofrontal, with associated cerebellar and brain-
stem signs. Most reported cases have had similar signs and symptoms. All of our cases involved the cerebellar hemisphere, usually near the cerebellolopontine angle. Most reported cases were also in the cerebellar hemisphere.

All of our patients had a subacute clinical picture. Two patients (cases 1 and 2) presented with fairly rapidly deteriorating courses. A third (case 3) had a slowly deteriorating course, and the fourth had symptomatic episodes spaced 18 months apart, but without clinical deterioration. All of these patients had subacute bleeds documented by CT scans (resolving hematoma) or pathology specimens (hemosiderin). Acute rebleeding was a significant feature of cases 1, 2, and 4 and was also confirmed by CT or pathology. Rebleeding occurred several days to 18 months postictus. The findings in our cases and those in the literature suggest a natural history much like that of AVMs. That is, some cerebellar venous angiomas (eight [15%] of 55 reported cases) bleed extensively at first, causing rapid, catastrophic clinical deterioration. Others (11 [20%] of 55 reported cases) bleed less profusely and remain static for a few days or years, after which they may rebleed. Apparently, most angiomas do not bleed at all.

The relation of size of venous angioma to bleeding is unclear. All of our patients had moderately large angiomas. Although several authors noted a positive correlation between the size of vascular malformations of any type and frequency of hemorrhage [2, 8], small, occult malformations are known to bleed [33, 39].

Given the possible propensity for cerebellar venous angiomas to bleed and the chance that rebleeding will occur, operative intervention becomes an important consideration. It is, as yet, impossible to predict what course an asymptomatic cerebellar venous angioma will take. Size and location may be indicators of possible future hemorrhage, but larger series are necessary to define prognostic factors. Once an angioma has bled, rebleeding may occur at any time. The extent and rate of resultant clinical decline is thus unpredictable. Indeed, a rebleed may be fatal, as several cases in the literature indicate [6, 14, 18, 40]. It would seem then that expeditious removal of a recognized hemorrhagic cerebellar hemisphere venous angioma may be indicated.

Surgery has been successful in cases of spontaneous (hypertensive and angiomatous) cerebellar hematoma [29, 31, 36, 38, 47, 48]. Both acute and chronic hematomas have been treated by suboccipital craniectomy and evacuation. Most cases with favorable outcome have been those with a good preoperative level of consciousness and those with slowly evolving clinical signs. Because venous angiomas can present with subacute bleeding, their clinical course seems to fit with those cases that predictably would do well with operation. The favorable outcome in our three operated cases substantiates the effectiveness of surgical management of subacutely bleeding cerebellar hemisphere venous angioma, a subgroup of spontaneous cerebellar hemorrhage that may, in fact, be cured by resection.

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