Cerebellar Venous Angioma Associated with Angiographically Occult Brain Stem Vascular Malformation. Report of Two Cases

Masamitsu Abe, M.D., Wilson T. Asfora, M.D., Antonio A.F. DeSalles, M.D., and Raymond N. Kjellberg, M.D.

Department of Neurosurgery, Massachusetts General Hospital, Harvard Medical School, Boston, Massachusetts

Abe M, Asfora WT, DeSalles AAF, Kjellberg RN. Cerebellar venous angioma associated with angiographically occult brain stem vascular malformation. Report of two cases. Surg Neurol 1990;33: 400–3.

We studied two patients with angiographically documented cerebellar venous angioma (malformation) and angiographically occult vascular malformation of the brain stem. One patient had recurrent hemorrhage in the pontine tegmentum. The second patient had recurrent hemorrhage in the midbrain. None had hemorrhage originating from the abnormal cerebellar venous channels. The more benign nature of angiographic venous malformations is supported by the cases we are presenting, as well as from a review of the literature, which includes an autopsy study of similar cases. The management of multiple vascular brain lesions is contingent on the verification of symptomatic pathological blood vessels. It is emphasized that angiographically occult vascular malformation could possibly exist in the vicinity of angiographic venous malformation when the patient with intracerebral hemorrhage, especially in the posterior fossa, was diagnosed as having venous malformation.

KEY WORDS: Cerebral angiography; Vascular malformation; Venous angioma; Brain stem; Cerebellum

Venous angioma (malformation) is often diagnosed on the sole basis of angiographic features. The clinical significance of angiographic venous malformation is still controversial, especially if it is located in the posterior fossa [2,10,13,15,16,18]. We are reporting two cases of angiographically documented cerebellar venous malformation. Recurrent hemorrhages occurred in the brain stem and not in the cerebellum, where a prominent venous abnormality was located. The significance of this is discussed.

Address reprint requests to: Masamitsu Abe, M.D., Department of Neurosurgery, Saga Medical School, Saga, 840-01, Japan.

Received December 12, 1989; accepted January 30, 1990.

Case Reports

Case 1

A 42-year-old man experienced the acute onset of blurred vision and weakness of the left limbs at age 14. Similar symptoms recurred at the age of 15 and 17. Subsequently, he did well until 22 years later when he noted right facial weakness and blurred vision that progressed over a few weeks.

Examination. His neurologic deficits on admission were slurred speech, bilateral horizontal gaze paresis, incomplete right peripheral facial palsy, hypesthesia on the left (sparing face), incoordination of the right limbs, and ataxic gait, all suggestive of a lesion in the pontine tegmentum and right cerebellar peduncle.

Investigations. The right vertebral angiogram revealed multiple spokelike venous structures in the right cerebellar hemisphere that drained into a dilated petrosal vein (Figure 1 *A* and *B*). A computed tomography (CT) scan with contrast demonstrated an enhancing, irregularly defined abnormality in the pons and right middle cerebellar peduncle (Figure 1 *C* and *D*). Magnetic resonance imaging (MRI) demonstrated an area of signal abnormality with little mass-effect in the pontine tegmentum and right brachium pontis. The signal abnormality consisted of an area of markedly decreased signal intensity felt to represent hemosiderin deposits surrounding punctate foci of high signal intensity, representing products of old hemorrhage (Figure 1 *E*).

The image abnormalities on angiogram, CT scan, and MRI were transposed to a lateral skull film and compared to one another [5]. The location of the abnormalities on CT scan and MRI related closely; however, they were different from that of the venous change on the angiogram.

Case 2

A 15-year-old boy, at the age of 7 years, developed weakness of all limbs. A right oculomotor nerve palsy

0090-3019/90/$3.50

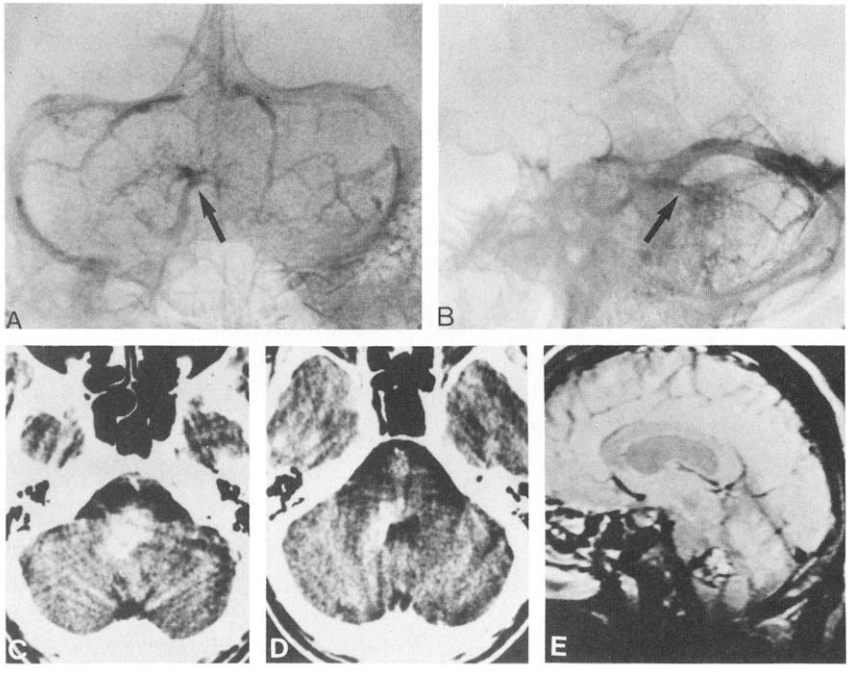

Figure 1. *Case 1.* **(A)** *Anteroposterior and* **(B)** *lateral views of the right vertebral angiograms in the venous phase demonstrate multiple spokelike venous structures (arrows) in the right cerebellar hemisphere that drain into a dilated petrosal vein.* **(C)** *A CT scan with contrast shows an enhancing, irregularly defined abnormality in the pons and right middle cerebellar peduncle.* **(D)** *There is prominence of vein posterior and superior to the lesion.* **(E)** *An MRI demonstrates an area of signal abnormality with little mass-effect in the pontine tegmentum and right brachium pontis.*

was also noted. At the age of 14 years he had an intense headache followed by worsening of his left hemiparesis and diplopia.

Examination. His neurologic deficits were a right partial oculomotor nerve palsy, double hemiparesis (left weaker than right), rotatory nystagmus on horizontal gaze, upbeat nystagmus on upgaze, and downbeat nystagmus on downgaze. These findings suggested a predominantly right midbrain lesion.

Investigations. Late venous films of the left vertebral angiogram demonstrated enlarged veins with numerous dilated medullary veins adjacent to the fourth ventricle (Figure 2 *A* and *B*). A CT scan demonstrated an area of high attenuation with microcalcifications in the right

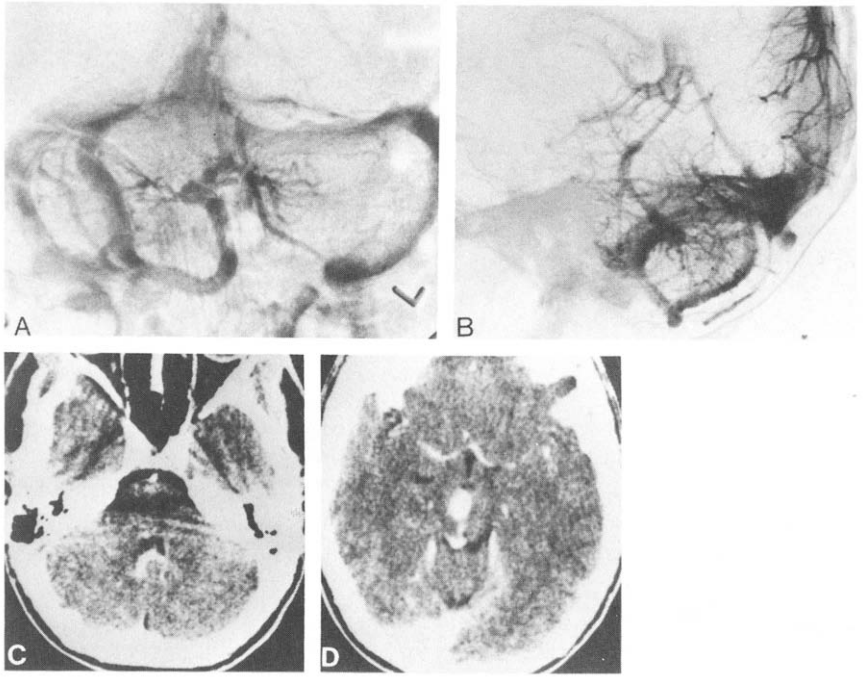

Figure 2. *Case 2.* **(A)** *Anteroposterior and* **(B)** *lateral views of the left vertebral angiograms in the venous phase demonstrate enlarged veins with numerous dilated medullary veins adjacent to the fourth ventricle.* **(C)** *A CT scan with contrast shows prominence of vein of lateral recess of fourth ventricle and* **(D)** *an area of high attenuation with microcalcifications in the right midbrain.*

midbrain. The vein of lateral recess of fourth ventricle, the precentral vein, and the posterior mesencephalic vein were prominent on the enhanced scan (Figure 2 *C* and *D*). An MRI demonstrated an area of heterogeneous signal intensity in the midbrain.

Discussion

The clinical manifestations of these two cases suggested brain stem vascular lesions that were then verified by CT scan and MRI. The angiogram showed adjacent cerebellar venous malformations but no brain stem vascular anomaly. It was concluded that these patients had symptomatic angiographically occult vascular malformations (AOVM) of the brain stem and unruptured venous malformations of the cerebellum.

Vascular malformations of the brain stem are often angiographically occult. In a series of 927 patients with vascular malformations of the brain, we observed 63 cases to be in the brain stem, of which 30 cases were AOVM [1]. Parenchymal hemorrhage is the main mechanism for clinical manifestations of AOVM. In most cases, however, the onset of symptoms is not apoplectic. Magnetic resonance imaging aids in determining the nature of the lesions, distinguishing them from multiple sclerosis or tumors. Pathological studies of these lesions have shown, predominantly, cavernous angiomas. Small arteriovenous malformations with thrombosis of some vessels, capillary telangiectases, and venous malformations have also been encountered [3,4,17].

Reports on the nature of angiographic cerebellar venous malformations are controversial. Patients with cerebellar hemisphere venous malformations that had bled were reported by Rothfus et al [13] and Malik et al [10]. They thought the natural history of these lesions was similar to that of arteriovenous malformations. Lasjaunias et al [8], however, proposed that venous angiomas should be renamed developmental venous anomalies. They drain normal cerebral tissue within a functionally normal arterial territory, they are associated with absence of a venous pathway that would normally drain the territory, and they, therefore, illustrate well-known transhemispheric anastomotic pathways that have developed in response to hemodynamic need. Senegor et al [16] reported a patient with a cerebellar venous malformation who suffered massive hemorrhagic venous infarction following surgical resection. Martin et al [11] and Biller et al [2] also reported a similar experience. They suggested that the potential adverse effects of iatrogenic occlusion of the venous drainage of a normal territory should be carefully considered.

Hemorrhage in our patients was caused by AOVM in the brain stem, and whether these AOVM are related to the cerebellar venous malformations remains uncertain. In view of their proximity, conceivably, they share some veins and are embryonically related. We believe that this association is more than a fortuitous one. Table 1 shows similar cases reported [6,7,9,12]. In all, recurrent hemorrhage occurred in the brain stem, and cerebral angiogram revealed a radially oriented group of veins in the cerebellar hemisphere.

In the cases submitted to pathological analysis [6,12], brain stem cavernous angioma associated with hemorrhage was encountered. Thin-walled vessels intermixed with normal brain parenchyma without evidence of hemorrhage were identified in the cerebellar white matter with extension into the pontine tegmentum. These were classified as telangiectases by pathologists. The histological distinction between venous malformation and

Table 1. *Brain Stem Vascular Malformation Associated with Angiographic Cerebellar Venous Malformation*

Case no.	Authors	Age/sex	Mode of presentation	Location of symptomatic lesion	Pathology
1	Roberson et al [12]	35/F	Hemorrhage	Pons and medulla	Cavernous angioma with hematoma in pons and medulla; telangiectases in cerebellum and pontine tegmentum
2	Diamond et al [6]	48/M	Progressive neurologic deficit	Pontine tegmentum	Cavernous angioma with hematoma and thrombosis in pons; telangiectases in pons (no section of cerebellum taken)
3	Diamond et al [6]	38/F	Hemorrhage	Pontine tegmentum	Large hemorrhage with tissue destruction in pons; telangiectases in cerebellum and pons
4	Gomori et al [7]	18/M	Hemorrhage	Pontine tegmentum	(−)
5	Lasjaunias et al [9]		Hemorrhage	Pons	( )
6	Present case 1	42/M	Hemorrhage	Pontine tegmentum	(−)
7	Present case 2	15/M	Hemorrhage	Midbrain	(−)

(−), pathological exam not carried out.

telangiectasis is somewhat arbitrary [6,14]. The angiographic appearance of the patients discussed herein are typical of venous malformation. Both lesions rarely give rise to hemorrhage while cavernous angiomas have a strong tendency to bleed and are generally angiographically occult.

The outcome following obliteration or resection of the cerebellar venous malformation may be catastrophic [2,11,16]. The cerebellar venous malformation in our cases presented were treated conservatively once it was established that these were not the site of hemorrhage. The symptomatic vascular lesions were treated at our institution by stereotactic Bragg peak proton beam radiotherapy using subnecrotic doses. The effectiveness of this treatment modality is not discussed here because of relatively short follow-up period.

In summary, we have described the association of cerebellar venous angioma and angiographically occult brain stem vascular malformation, the latter being the source of hemorrhage. Angiographic venous malformations appear to be a more benign entity and on some occasions may support the presence of AOVM in the proximity to them. We emphasize the need for accurate localization of symptomatic pathological blood vessels through the combination of CT, MRI, and angiogram, especially if treatment is planned, whether it is by surgical resection, stereotactic radiation, or particle beam therapy.

We thank R.M. Moffet and C. Ideguchi for assistance in the preparation of this manuscript.

References

1. Abe M, Kjellberg RN, Adams RD. Clinical presentations of vascular malformations of the brainsrem: comparison of angiographically positive and negative types. J Neurol Neurosurg Psychiatry 1989;52:167–75.
2. Biller J, Toffol GJ, Shea JF, Fine M, Azar-Kia B. Cerebellar venous angiomas. Arch Neurol 1985;42:367–70.
3. Britt RH, Connor WS, Enzmann DR. Occult arteriovenous malformation of the brainstem simulating multiple sclerosis. Neurology 1981;31:901–3.
4. Cohen HCM, Tucker WS, Humphreys RP, Perrin RJ. Angiographically cryptic histologically verified cerebrovascular malformations. Neurosurgery 1982;10:704–14.
5. DeSalles AAF, Asfora WT, Abe M, Kjellberg RN. Transposition of target information from magnetic resonance and computed tomographic scan images to the conventional x-ray stereotactic space. Appl Neurophysiol 1987;50:23–32.
6. Diamond C, Torvik A, Amundsen P. Angiographic diagnosis of telangiectases with cavernous angioma of the posterior fossa. Acta Radiol Diag 1976;17:281–8.
7. Gomori JM, Grossman RI, Goldberg HI, Hackney DB, Zimmerman RA, Bilaniuk LT. Occult cerebral vascular malformations: High-field MR imaging. Radiology 1986;158:707–13.
8. Lasjaunias P, Burrows P, Planet C. Developmental venous anomalies (DVA): so-called venous angioma. Neurosurg Rev 1986;9: 233–44.
9. Lasjaunias P, Terbrugge K, Choi IS. Trans-mesencephalic arteries and veins: angiographic aspects in tectal vascular lesions. Acta Neurochir 1988;92:138–43.
10. Malik GM, Morgan JK, Boulos RS, Ausman JI. Venous angiomas: an underestimated cause of intracranial hemorrhage. Surg Neurol 1988;30:350–8.
11. Martin NA, Wilson CB, Stein BM. Venous and cavernous malformations. In: Wilson CB, Stein BM, eds. Intracranial arteriovenous malformations. Baltimore: Williams & Wilkins, 1984: 234–45.
12. Roberson GH, Kase CS, Wolpow ER. Telangiectases and cavernous angiomas of the brainstem: "cryptic" vascular malformations. Report of a case. Neuroradiology 1974;8:83–9.
13. Rothfus WE, Albright AL, Casey KF, Latchaw RE, Roppolo HMN. Cerebellar venous angioma: "benign" entity? AJNR 1984;5:61–6.
14. Russel DS, Rubinstein LJ. Tumors and hamartomas of the blood vessels. In: Pathology of tumors of the nervous system. 4 ed. London: Edward Arnold, 1977:116–45.
15. Saito Y, Kobayashi N. Cerebral venous angiomas. Radiology 1981;139:87–94.
16. Senegor M, Dohrmann GJ, Wollmann RL. Venous angiomas of the posterior fossa should be considered as anomalous venous drainage. Surg Neurol 1983;19:26–32.
17. Stahl SM, Johnson KP, Malamud N. The clinical and pathological spectrum of brain-stem vascular malformations. Long-term course stimulates multiple sclerosis. Arch Neurol 1980;37:25–9.
18. Wendling LR, Moore JS, Kieffer SA, Goldberg HI, Latchaw RE. Intracerebral venous angioma. Radiology 1976;119:141–7.