Venous Angiomas with Arteriovenous Shunts: Report of Three Cases and Review of the Literature

Masaki Komiyama, M.D., Kazuhiro Yamanaka, M.D., Yoshiyasu Iwai, M.D., Toshihiro Yasui, M.D.

Department of Neurosurgery, Osaka City General Hospital, Osaka, Japan

OBJECTIVE AND IMPORTANCE: In spite of recent recognition of the benign nature of venous angioma (VA), only limited information is available on the clinical features of VA with arteriovenous shunt (AVS). The purpose of this study was to elucidate the clinical profile of VA with AVS.

CLINICAL PRESENTATION AND INTERVENTION: We describe three patients having a VA with AVS and review the clinical features of 31 patients reported in the literature, including our three patients. The patients included 12 women and 19 men, ranging in age from 18 to 54 years. Seven patients (22.6%) presented with intracranial hemorrhage, and none of 16 patients developed a new or recurrent hemorrhage (mean follow-up period, 11 months). Treatment was conservative in 14 patients, lobectomy or partial resection of the VA in 6, removal of hematoma in 4, operation only for coexisting aneurysm or arteriovenous malformation in 4, and not known in 3. The outcome was reported as good recovery in 19 patients, persistent neurological deficits in 2, death or deterioration not related to the VA in 3, and not known in 7.

CONCLUSION: Although there remains some uncertainty as to the clinical features of VA with AVS, its prognosis seems to be essentially as benign as that of VA without AVS. Thus, conservative treatment is recommend except for patients with a large hematoma or with a coexisting arteriovenous malformation or a symptomatic, accessible cavernous angioma, which may be treated by surgical intervention. Further collection of data is required to establish definite treatment guidelines.

(Neurosurgery 44:1328–1335, 1999)

Key words: Angiography, Arteriovenous shunts, Cavernous angioma, Early venous filling, Venous angioma

Vascular malformations in the central nervous system are classified into five groups on the basis of their microscopic features: telangiectasia, varix, cavernous angioma, arteriovenous malformation (AVM), and venous angioma (VA) (18). VA has the following characteristic angiographical features: absence of feeding arteries, demonstration of dilated medullary veins and collecting dilated central veins (either transcortical or subependymal veins) only in the venous phase, and aplastic or hypoplastic venous system in the neighborhood of the angioma (4, 11, 14, 20, 28, 41, 42).

There is a type of VA that has an arteriovenous shunt (AVS) and/or capillary blush in the late arterial phase on angiogram. This entity has been given a variety of designations, such as VA with AVS, VA with early filling vessels (21, 35), medullary venous malformation with an arterial component (11), high-flow medullary venous malformation (26), arterialized venous malformation (2), AVM with venous predominance (41, 42), transitional form between a VA and an AVM (22), or a mixed vascular malformation of a VA and an AVM (2, 8).

Twenty-eight such cases have been reported in the literature. There is no consensus on this entity as to whether it differs from a typical VA without AVS. We report three additional cases having a VA with AVS and review the pertinent literature to clarify the clinical features of VA with AVS.

CASE PRESENTATION

Patient 1

A 26-year-old man underwent magnetic resonance imaging (MRI) to evaluate for numbness in both hands for several years. The patient was neurologically normal, and his history was not contributory. The MRI examination revealed a signal-void lesion in the right basal ganglia suggestive of a vascular lesion. He underwent cerebral angiography. A right carotid angiogram showed dilated lenticulostriate and anterior thalamoperforating arteries, capillary blush in the basal ganglia and in the right anterior temporal lobe, dilated medullary veins, early venous filling of the right markedly dilated inferoventricular vein and basal vein and the right normal-sized internal cerebral vein (deep drainage) (*Fig. 1*). A left vertebral angiogram showed dilated posterior thalamoperforating arteries, capillary blush in the right thalamus, and early venous filling of the dilated medullary veins in the thalamus draining to the right superior thalamic vein (deep drainage). Instead of the right hypoplastic superior striate veins, the right inferior striate veins and temporal pole vein were well developed. These angiographic findings were consistent with a VA with AVS. The patient was treated conservatively. There was no cerebrovascular event during a follow-up period of 6 years.

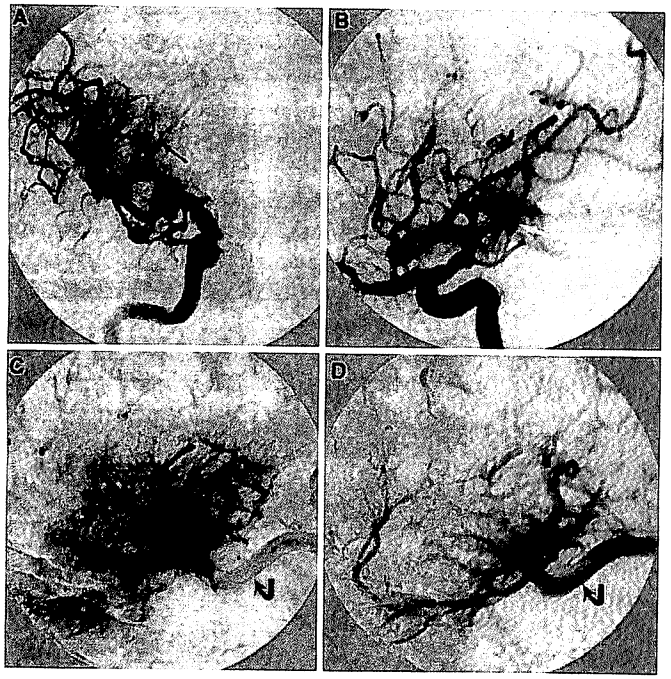

FIGURE 1. Patient 1, a 26-year-old man. Right internal carotid injection. *A*, anteroposterior view; *B–D*, lateral views. *A* and *B* are in the midarterial phase, *C* is in the late arterial phase, and *D* is in the venous phase. Dilated lenticulostriate arteries (*black arrow*) and anterior thalamoperforating arteries (*white arrow*) are observed. Capillary blush in the basal ganglia and anterior temporal lobe is also seen. Dilated medullary veins and early venous filling of the inferoventricular vein and the basal vein (*curved arrows*) are demonstrated.

Patient 2

A 24-year-old woman suddenly developed vertigo, nausea, and vomiting. These symptoms improved within 2 days. When she came to our hospital about 1 month later, she was neurologically normal. Her history was unremarkable. An MRI examination revealed a recent infarction in the bilateral cerebellar hemispheres and a linear signal void in the left medioparietal lobe suggestive of a vascular lesion. A cardiac examination and laboratory data were normal. A left vertebral angiogram showed the aortic origin of the left vertebral artery and its caliber change at the C1 level, suggesting arterial dissection. A right vertebral angiogram showed retrograde filling of the left posteroinferior cerebellar artery and a normal basilar artery. The angiogram also showed slightly dilated left posterior pericallosal and left parieto-occipital arteries, which produced capillary blush in the medioparietal lobe and early venous filling of the dilated medullary veins, which drained to the left medioparietal vein (superficial drainage) and left medioatrial vein (deep drainage). A left carotid angiogram demonstrated dilated left inferior internal parietal and pericallosal arteries, which fed the above-mentioned vascular lesion (*Fig. 2*). The diagnosis was left vertebral artery dissection causing cerebellar infarction and an incidentally discovered VA with AVS. This patient was conservatively treated. There was no recurrence or new cerebrovascular event during a follow-up period of 1½ years.

Patient 3

A 22-year-old man experienced sudden dizziness and mild headache. This patient was admitted to a local hospital. A computed tomographic scan revealed intraventricular hemorrhage. Cerebral angiograms were interpreted as an AVM. Two weeks later, the patient was transferred to us for further examination and possible gamma knife surgery. At admission, this patient was neurologically normal without a remarkable history. Repeated cerebral angiograms demonstrated dilated left inferior internal parietal and left parieto-occipital arteries, capillary blush, and early venous filling of the medioparietal vein (superficial drainage). The collecting medioparietal vein showed varicose change before the entrance to the superior sagittal sinus (*Fig. 3*). The angiograms were interpreted as revealing a VA with AVS in the left parietal lobe. This patient was conservatively treated, and a repeated cerebral hemorrhage has not occurred during a follow-up period of 3 years.

Summary of 31 reported cases of VA with AVS

In the literature, there have been 31 patients, including our three patients, who were diagnosed as having a VA with AVS (*Table 1*) (2, 3, 8, 11, 12, 20, 21, 26–28, 31–33, 35, 36, 38–40, 42–44). There were 12 women and 19 men, ranging in age from 18 to 54 years (mean, 36 years). Locations of the lesions were frontal in 15 patients, temporal in 2, parietal in 6, occipital in 1, basal in 3, cerebellum in 1, and in the larger region in 3. VA with AVS was located in the both supra- and infratentorial locations in one patient. Intracerebral or intraventricular hemorrhage occurred in 7 (22.6%) of the 31 patients, although a coexisting cavernous angioma could not be completely ruled out. None of the 16 patients whose clinical data were available developed new or recurrent hemorrhage in follow-up periods of 1 month to 6 years (mean, 11 months). No treatment was performed in 14 patients, whereas lobectomy or partial resection of the VA was performed in 6 patients, hematoma removal in 4, and operation only for AVM or aneurysm in 4. No data on treatment were available for three patients. The outcomes included good recovery in 19 patients, persistent neurological deficits in 2, death not related to the VA in 2, deterioration not related to the VA in 1, and not known in 7.

DISCUSSION

Pathogenesis of VA with and without AVS

Lasjaunias et al. (14) designated a VA as a "developmental venous anomaly," because this lesion represents an extreme but normal anatomic variant of venous drainage without a venous occlusive condition in early fetal life. The same concept of the VA representing a compensatory drainage system was presented by others (32, 34). Goulao et al. (7), Lasjaunias and Berenstein (13), and

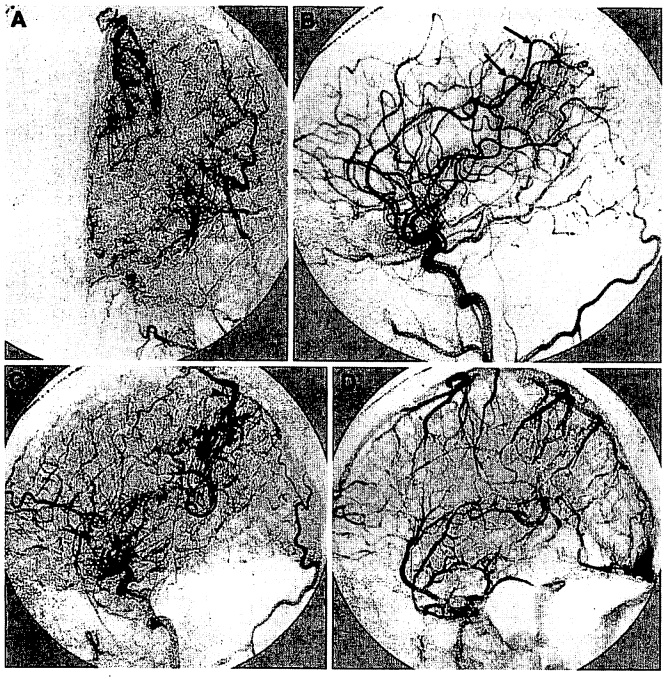

FIGURE 2. Patient 2, a 24-year-old woman. Left common carotid injection. *A*, anteroposterior view; *B–D*, lateral views. *A* is in the late arterial phase, *B* is in the midarterial phase, *C* is in the capillary phase, and *D* is in the venous phase. The left internal parietal arteries (*arrows*) are slightly dilated. Dilated medullary veins and capillary blush are observed. Arteriovenous shunts to the collecting veins of left medioparietal vein (*arrowhead*) and medioatrial vein (*open arrow*) are also observed.

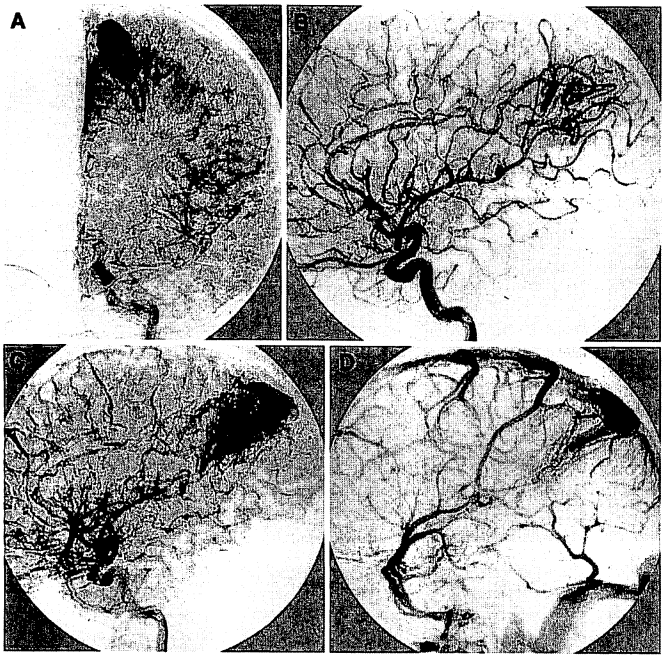

FIGURE 3. Patient 3, a 22-year-old man. Left internal carotid injection. *A*, anteroposterior view; *B–D*, lateral views. *A* and *C* are in the midarterial phase, *B* is in the late arterial phase, and *D* is in the venous phase. Dilated medullary veins and early venous filling are observed. Varicose change (*arrow*) is noted before the entrance to the superior sagittal sinus.

Lasjaunias et al. (14) assert that the associated ectatic veins in some cases of VA are related to additional, acquired dural or venodural anomalies and reflect a hemodynamic obstacle to drainage and that the hemorrhage in association with VA is caused by the associated cavernous angioma or reduced flexibility of the venous system to venous hemodynamic changes. They think that a significant venous disorder (such as thrombosis) at an early stage of development must be associated with some neural tissue abnormality. Lasjaunias proposes that the difference between VAs with and without AVS is simply a "transit time" within the VAs (P Lasjaunias, personal communication, 1998). Thus, VA with AVS and VA without AVS are the same clinical entity with the same clinical picture.

Huang et al. (10) postulated that when there is a partial, incomplete, repetitive collapse and reopening of pial, medullary, or subependymal vein(s) or dural sinus(es), occurring during early fetal life in the uterus, any increase in venous pressure of the involved veins may extend in retrograde fashion to the neighboring venules, capillaries, arterioles, and arteries. This mechanism may lead to the formation of a VA with involvement of medullary veins only, VA with involvement of capillovenous components, VA with medullary arterial components, VA with arterial components, or frank malformation of localized or even diffuse form, depending upon the degree and extent of the retrograde involvement. Huang (9) explained that dead cells derived from any injury can be removed by cell motility in the fetus, and thus the lesion is left as an "anomaly" owing to the lack of tissue abnormality.

Mullan et al. (23) postulated that VA is the result of developmental failure of the venous system in its fourth (last) developmental stage and proposed that an AVM is a fistulized VA. Mullan et al. (22) and Hirata et al. (8) postulated further that a VA with AVS is a transitional form between a VA and an AVM. Mullan et al. (22) emphasized the angiographic features of the "arteriovenous fistula" and "venous pattern" in a VA with AVS and speculated that the venoocclusive process plays some role in the pathogenesis of the transitional form between the VA and the AVM. Awad et al. (2) theorized that a VA with AVS is a mixed vascular malformation of a VA and an AVM, and they also termed it an arterialized VA.

There are not sufficient direct data to support each of the above-mentioned ideas on the pathogenesis of VAs without or with AVS, and it can therefore be said that, at present, there is no consensus on their pathogenesis.

Clinical features of VA

Although VA was believed to be a rare lesion, owing to the paucity of clinical reports in the 1960s and 1970s, it is

now recognized as a not uncommon incidental lesion at autopsy, and its incidence is reported to be 2.6% (105 of 4069 patients) (33). The nonpathognomonic nature of the VA is emphasized, on the one hand (2, 6, 13, 14, 29, 32), although the propensity toward bleeding from a VA also has been emphasized (17, 30). Huang et al. (11) reported that 24% of 62 patients with a VA manifested intracranial hemorrhage. Rothfus et al. (30) reported that a VA in the posterior fossa is more likely to bleed, in comparison to those in the supratentorial location. Rigamonti et al. (29) reported that the VA is essentially a silent lesion and is frequently associated with other, more symptomatic conditions, including cavernous angioma. Garner et al. (6) reported that bleeding among their 100 patients with VA occurred in one patient (1%) and that seizures and focal neurological deficits were unusual associated findings. McLaughlin et al. (19) reported that among their 80 patients with a VA, the retrospective bleeding rate was 0.61% per year and the prospective bleeding rate was 0.68% per year (symptomatic rate, 0.34% per year). Although subsequent bleeding of a VA may occur (5, 17, 19, 30), the rate of further episodes of bleeding has not been established. Judging from the data of recent studies (6, 19), the prognosis of VA is relatively benign, with a significantly low bleeding rate.

Clinical features of VA with AVS

Because venous filling in the frontoparietal region is normally earlier than that in the rest of the brain, earlier venous filling and/or capillary blush in a VA located in the frontoparietal region can be regarded as normal venous filling (13, 32, 41). Confusion as to interpretation of early venous filling in the frontoparietal region leads to confusion in the diagnosis of VA with or without AVS. We have included in this report all of the well-documented cases of VA, in which the authors observed "AVS" or "capillary blush" in the late arterial phase.

Computed tomographic and MRI features of VA with AVS are essentially the same as those of VA without AVS. Thus, it is impossible to differentiate VAs with and without AVS solely by computed tomography or MRI (2). If the prognosis and treatment strategy of VA with AVS were significantly different from those of VA without AVS, cerebral angiography would be required. Awad et al. (2) proposed performance of angiography for symptomatic VA because they believe that the prognosis of VA with AVS is different from that of VA without AVS.

Differential diagnosis of a VA with AVS from an AVM with medullary components is important. AVMs with medullary components usually exhibit a wedge shape and a streaming pattern of medullary arteries and veins, and they have grossly enlarged feeding arteries with more rapid drainage through multiple venous channels, whereas VA with AVS frequently exhibits transcerebral drainage and absence of deep or superficial connecting venous segments (11, 42).

It is not clear whether the clinical features of VA with AVS are different from those of VA without AVS. Moritake et al. (21) reported that VA with AVS has no predilection for intracerebral hemorrhage. Hirata et al. (8) thought, however, that the incidence of bleeding is higher in VA with AVS than without AVS because of the similarities of the characteristics of an AVM and a VA with AVS. Four patients with an AVM that drained into a coexisting VA have been reported (11, 15, 25). Tomura et al. (40) reported the case of a VA with AVS coexisting with an AVM. These cases suggest common pathogenesis or causation-evolution among VA without AVS, VA with AVS, and AVM. It is important to differentiate VA with AVS from the coexistence of VA and an AVM because the therapeutic strategies may be different. Four patients had coexisting cerebral aneurysms (26, 32, 36, 39). Patient 2 in our series presented with vertebral artery dissection (dissecting aneurysm) and an incidental VA with AVS.

The clinical data on 31 patients having VA with AVS, including 3 of our patients, indicated that 7 (22.6%) of 31 patients presented with hemorrhage, none of 16 patients whose clinical data were available experienced subsequent bleeding, and the outcome was generally good. These data cannot simply be compared with the recent reports of benign prognosis and low bleeding rate of the VA without AVS, because of the difference in the population studied (6, 19). Although the prevalence and clinical profile of VA with AVS can be revealed only by prospective study, the prevalence and clinical profile seem to be similar to those of VA without AVS.

Therapeutic consideration of VA with and without AVS

Because the natural history of VA without AVS is still not fully understood, the treatment strategy is difficult to formulate. Recent knowledge of the benign nature of the VA without AVS encourages a more conservative approach than was previously taken (1, 6, 19, 37). Classically, surgical resection of the VA is contraindicated because resection of the large collecting venous system, which drains normal venous return, may cause catastrophic venous infarction (5, 6, 19, 24, 29, 34). Only removal of the hematoma is indicated, when necessary, or the VA in the "silent" region or epileptogenic region may be removed in selected patients (2, 4, 35, 42). However, some groups, who believe that the VA has a significant potential to bleed and/or rebleed, advocate surgical extirpation of the lesion when the VA is symptomatic (17, 21, 30). Since the VA is frequently associated with a cavernous angioma, it is now recommended to resect an associated, accessible cavernous angioma with preservation of the VA in case of hemorrhage (2, 19, 29).

Although Hirata et al. (8) emphasized the need for surgical removal of the VA with AVS, it is not clear that treatment strategies similar to those for the typical VA can be applied to the VA with AVS. Because the prognosis of VA with AVS seems to be essentially as benign as that of VA without AVS, we believe that conservative treatment is recommended, except for cases with a large hematoma or with a coexisting AVM or a symptomatic, accessible cavernous angioma, which may be treated by surgical intervention.

In the literature, two patients having VAs with AVS were reported to be treated by stereotactic radiosurgery (2,

TABLE 1. Characteristics of 31 Reported Cases of Venous Angioma with Arteriovenous Shunt[a]

No.	Series (Ref. No.)	Age (yr)/Sex	Location 1	Location 2	Symptoms	Bleed from Angioma	Operation for Venous Angioma	Subsequent Bleeding	Outcome	Follow-up Period	Remarks
1	Wolf et al., 1967 (43)	52/M	Supra-	L-frontal, temporal L-parietal, R-frontal	ICH	No	No	—	Dead	—	Multiple venous angiomas ICH not related to angioma
2	Wendling et al., 1976 (42)	28/M	Supra-	R-frontal	Seizure	No	Lobectomy	—	—	—	
3	Wendling et al., 1976 (42)	49/M	Supra-	L-frontal	Seizure	No	No	No	Good recovery	3 yr	
4	Preissig et al., 1976 (28)	23/F	Supra-	R-frontal	Headache	No	Block resection	No	Good recovery	2 yr	
5	Michels et al., 1977 (20)	54/F	Supra-	L-frontal	Epilepsy	No	No	No	Good recovery	—	
6	Sarwar and McCormick, 1978 (33)	38/M	Supra-	L-basal	Hepatic coma	No	No	—	Dead	—	Death due to hepatic coma
7	Suganuma et al., 1978 (38)	28/M	Supra-	L-frontal	Seizure	No	Partial removal	—	Good recovery	—	
8	Cabanes et al., 1979 (3)	58/F	Supra-	L-frontal	Headache	No	Lobectomy	No	Good recovery	3 yr	
9	Cabanes et al., 1979 (3)	20/F	Infra-	R-, L-cerebellum	V, VI, VII nerve palsy	No	No	No	No change	2 yr	
10	Moritake et al., 1980 (21)	39/M	Supra-	R-frontal	Incidental	No	No	No	Deterioration	2 yr	Quadriparesis due to nonrelated infarction
11	Pardatscher et al., 1980 (27)	46/M	Supra-	L-occipital	IVH	No	No	—	Good recovery	—	
12	Saito and Kobayashi, 1981 (32)	27/F	Supra-	L-basal	Headache	No	No	—	—	—	
13	Saito and Kobayashi, 1981 (32)	25/M	Supra-	L-frontal	Incidental	No	No	—	—	—	
14	Saito and Kobayashi, 1981 (32)	41/F	Supra-	R-frontal	SAH	No	No	—	—	—	Operation for MCA aneurysm
15	Huang et al., 1984 (11)	39/M	Supra-	R-temporal	Tremor	No	—	—	—	—	
16	Huang et al., 1984 (11)	52/M	Supra-	L-parietal	Incidental	No	—	—	—	—	
17	Hirata et al., 1986 (8)	18/M	Supra-	R-parietal	ICH	Yes	Operation for ICH	No	Mild hemiparesis	2 yr	
18	Shiroyama et al., 1986 (35)	34/F	Supra-	L-frontal	ICH	Yes	Operation for ICH	—	Good recovery	—	
19	Yasargil et al., 1988 (44)	27/M	Supra-	R-frontal	ICH	Yes	—	—	—	—	
20	Sonoda et al., 1988 (36)	24/F	Supra-	R-parietal	IVH	No	No	No	Good recovery	1 mo	Bleed from choroidal aneurysm
21	Kurimoto et al., 1989 (12)	26/M	Supra-	R-frontal, parietal	ICH	Yes	Operation for ICH	No	Good recovery	3 yr	
22	Tashiro et al., 1989 (39)	49/M	Supra-	R-frontal	SAH	No	No	—	Good recovery	—	Operation for ruptured ICPC aneurysm

[a] **Supra-, supratentorial; infra-, infratentorial; L, left; R, right; ICH, intracerebral hemorrhage; IVH, intraventricular hemorrhage; SAH, subarachnoid hemorrhage; TIA, transient ischemic attack; MCA, middle cerebral artery; ICPC, internal carotid and posterior communicating arteries; Acom, anterior communicating artery; AVM, arteriovenous malformation; —, not reported.**

TABLE 1. *Continued*

No.	Series (Ref. No.)	Age (yr)/Sex	Location 1	Location 2	Symptoms	Bleed from Angioma	Operation for Venous Angioma	Subsequent Bleeding	Outcome	Follow-up Period	Remarks
23	Ochi et al., 1990 (26)	48/F	Supra-	L-frontal	Headache	No	No	No	Good recovery	6 mo	Operation for Acom aneurysm
24	Tomura et al., 1990 (40)	47/M	Supra-	L-frontal	ICH	No	No	—	Good recovery	—	Operation for bleeding AVM
25	Awad et al., 1993 (2)	39/F	Supra-	L-frontal	Seizure or TIA	No	Gyrectomy	No	Good recovery	2 yr	
26	Awad et al., 1993 (2)	36/F	Supra-	parietal	ICH	Yes	Operation for ICH + radiosurgery	No	Good recovery	1 yr	
27	Awad et al., 1993 (2)	54/M	Supra-	L-temporal	ICH	Yes	Gyrectomy	No	Good recovery	8 mo	
28	Sagoh et al., 1996 (31)	24/M	Supra-, infra-	R-cerebrum, cerebellum	Hemiparesis (TIA)	No	No	—	Good recovery	—	
29	Current report, Patient 1	26/M	Supra-	R-basal	Incidental	No	No	No	Good recovery	6 yr	
30	Current report, Patient 2	24/F	Supra-	L-parietal	Infarction	No	No	No	Good recovery	1 yr	Infarction due to vertebral dissection
31	Current report, Patient 3	22/M	Supra-	L-parietal	IVH	Yes	No	No	Good recovery	3 yr	

22). Lindquist et al. (15) reported that radiosurgery for the VA without AVS resulted in total obliteration in 11% (one of nine patients) and partial obliteration of the VA in 33% (three of nine patients), with radiation-related complications in 30% of patients overall. Judging from the disappointing results of radiosurgery for VA and the inherent benign nature of VA, radiosurgery may have no role in the treatment of VA without AVS at present (16), and possibly even for treatment of VA with AVS. We believe that long-term follow-up and collection of data on the natural history of this entity are necessary to formulate definite treatment guidelines for VA with AVS.

Received, October 30, 1998.
Accepted, February 5, 1999.
Reprint requests: Masaki Komiyama, M.D., Department of Neurosurgery, Osaka City General Hospital, 2-13-22, Miyakojima-Hondouri, Miyakojima, Osaka 534-0021, Japan.

REFERENCES

1. Awad IA: Radiosurgery and venous malformations. **J Neurosurg** 80:171–173, 1994 (letter).
2. Awad IA, Robinson JR, Mohanty S, Estes ML: Mixed vascular malformations of the brain: Clinical and pathogenetic considerations. **Neurosurgery** 33:179–188, 1993.
3. Cabanes J, Blasco R, García M, Tamarit L: Cerebral venous angiomas. **Surg Neurol** 11:385–389, 1979.
4. Dias PS, Forster DMC, Bergvall U: Cerebral medullary venous malformations: Report of four cases and review of the literature. **Br J Neurosurg** 2:7–21, 1988.
5. Fujii K, Matsushima T, Inumura T, Fukui M: Natural history and choice of treatment in forty patients with medullary venous malformation (MVM). **Neurosurg Rev** 15:13–20, 1992.
6. Garner TB, Del Curling O Jr, Kelly DL Jr, Laster DW: The natural history of intracranial venous angiomas. **J Neurosurg** 75:715–722, 1991.
7. Goulao A, Alvarez H, Garcia Monaco R, Pruvost P, Lasjaunias P: Venous anomalies and abnormalities of the posterior fossa. **Neuroradiology** 31: 476–482, 1990.
8. Hirata Y, Matsukado Y, Nagashiro S, Kuratsu J: Intracerebral venous angioma with arterial blood supply: A mixed angioma. **Surg Neurol** 25:227–232, 1986.
9. Huang YP: Large diffuse arteriovenous malformations affecting the right cerebral and cerebellar hemispheres: Case report. **Surg Neurol** 46: 127–128, 1996 (commentary).
10. Huang YP, Okudera T, Fukusumi A, Maehara F, Stollman AL, Mosesson R, Lidov M, Mitty HA: Venous architecture of cerebral hemisphere white matter and comments on pathogenesis of medullary venous and other cerebral vascular malformations. **Mt Sinai J Med** 64:197–206, 1997.
11. Huang YP, Robbins A, Patel SC, Chaudhary M: Cerebral venous malformations and a new classification of cerebral vascular malformations, in Kapp JP, Schmidek HH (eds): *The Cerebral Venous System and Its Disorders.* Orlando, Grune & Stratton, 1984, pp 373–474.
12. Kurimoto M, Oka N, Otsuji T, Endo S, Takaku A: Medullary venous malformation with marked early venous filling: A case report [in Japanese]. **Rinsho Hoshasen** 34:735–738, 1989.
13. Lasjaunias P, Berenstein A: Intracranial venous system, in *Surgical Neuroangiography: Functional Vascular Anatomy of Brain, Spinal Cord and Spine.* Berlin, Springer-Verlag, 1990, vol 3, pp 223–296.
14. Lasjaunias P, Burrows P, Planet C: Developmental venous anomalies (DVA): The so-called venous angioma. **Neurosurg Rev** 9:233–244, 1986.
15. Lindquist C, Guo W-Y, Karlsson B, Steiner L: Radiosurgery for venous angiomas. **J Neurosurg** 78:531–536, 1993.
16. Lindquist C, Karlsson B, Guo W-Y, Steiner L: Radiosurgery and venous malformations. **J Neurosurg** 80:174–175, 1994 (letter).
17. Malik GM, Morgan JK, Boulos RS, Ausman JI: Venous angiomas: An underestimated cause of intracranial hemorrhage. **Surg Neurol** 30:350–358, 1988.
18. McCormick WF: The pathology of vascular ("arteriovenous") malformations. **J Neurosurg** 24:807–816, 1966.
19. McLaughlin MR, Kondziolka D, Flickinger JC, Lunsford S, Lunsford LD: The prospective natural history of cerebral venous malformations. **Neurosurgery** 43:195–201, 1998.

20. Michels LG, Bentson JR, Winter J: Computed tomography of cerebral venous angiomas. **J Comput Assist Tomogr** 1:149–154, 1977.
21. Moritake K, Handa H, Mori K, Ishikawa M, Morimoto M, Takebe Y: Venous angiomas of the brain. **Surg Neurol** 14:95–105, 1980.
22. Mullan S, Mojtahedi S, Johnson DL, Macdonald RL: Cerebral venous malformation-arteriovenous malformation transition forms. **J Neurosurg** 85:9–13, 1996.
23. Mullan S, Mojtahedi S, Johnson DL, Macdonald RL: Embryological basis of some aspects of cerebral vascular fistulas and malformations. **J Neurosurg** 85:1–8, 1996.
24. Nagata K, Kubo T, Fukushima T: Four cases of cerebral venous angioma: With particular reference to the operative indication and the importance of CT diagnosis [in Japanese]. **No Shinkei Geka** 11:1201–1208, 1983.
25. Nussbaum ES, Heros RC, Madison MT, Awasthi D, Truwit CL: The pathogenesis of arteriovenous malformations: Insights provided by a case of multiple arteriovenous malformations developing in relation to a developmental venous anomaly. **Neurosurgery** 43:347–352, 1998.
26. Ochi S, Hirano A, Kanwo K, Hashi K: High flow medullary venous malformation with aneurysm on the proximal artery: A case report [in Japanese]. **No To Shinkei** 42:361–366, 1990.
27. Pardatscher K, Fiore DL, Galligioni F, Iraci G: Diagnosis of cerebral venous angioma by rapidly enhanced CT scan. **Surg Neurol** 14:111–113, 1980.
28. Preissig RS, Preissig SH, Goree JA: Angiographic demonstration of a cerebral venous angioma: Case report. **J Neurosurg** 44:628–631, 1976.
29. Rigamonti D, Spetzler RF, Medina M, Rigamonti K, Geckle DS, Pappas C: Cerebral venous malformations. **J Neurosurg** 73:560–564, 1990.
30. Rothfus WE, Albright AL, Casey KF, Latchaw RE, Roppolo HMN: Cerebellar venous angioma: "Benign" entity? **AJNR Am J Neuroradiol** 5:61–66, 1984.
31. Sagoh M, Kodaki K, Ichikizaki K, Izumi C, Satoh T, Shiga H: Large diffuse arteriovenous malformation affecting the right cerebral and cerebellar hemispheres: Case report. **Surg Neurol** 46:122–127, 1996.
32. Saito Y, Kobayashi N: Cerebral venous angiomas: Clinical evaluation and possible etiology. **Radiology** 139:87–94, 1981.
33. Sarwar M, McCormick WF: Intracerebral venous angioma: Case report and review. **Arch Neurol** 35:323–325, 1978.
34. Senegor M, Dohrmann GJ, Wollmann RL: Venous angiomas of the posterior fossa should be considered as anomalous venous drainage. **Surg Neurol** 19:26–32, 1983.
35. Shiroyama Y, Kurokawa Y, Ueda H, Katayama S, Mitani T: Operated case of cerebral venous angioma with early venous filling on cerebral angiography: Case report [in Japanese]. **No Shinkei Geka** 14:1355–1360, 1986.
36. Sonoda H, Yoshida A, Ohtsuka T, Takaki S, Matsukado Y: Venous angioma with an arterial component and an associated aneurysm: Case report [in Japanese]. **Neurol Med Chir (Tokyo)** 28:838–842, 1988.
37. Spetzler RF, Hamilton MG: Radiosurgery and venous malformations. **J Neurosurg** 80:173–174, 1994 (letter).
38. Suganuma Y, Oie K, Tanigawa K, Matsushima Y, Inaba Y: A case of cerebral venous angioma [in Japanese]. **No Shinkei Geka** 6:77–83, 1978.
39. Tashiro Y, Takeno Y, Mizoguchi T, Maehara F: Medullary venous malformation with an arterial component associated with a ruptured aneurysm: Case report. **Neurol Med Chir (Tokyo)** 29:857–860, 1989.
40. Tomura N, Inugami A, Higano S, Uemura K, Hadeishi H, Yasui N: A case of medullary venous malformations with an arterial component coexisting with arteriovenous malformations. **Surg Neurol** 33:37–42, 1990.
41. Valavanis A, Wellauer J, Yasargil MG: The radiological diagnosis of cerebral venous angioma: Cerebral angiography and computed tomography. **Neuroradiology** 24:193–199, 1983.
42. Wendling LR, Moore JS, Kieffer SA, Goldberg HI, Latchaw RE: Intracerebral venous angioma. **Radiology** 119:141–147, 1976.
43. Wolf PA, Rosman NP, New PFJ: Multiple small cryptic venous angiomas of the brain mimicking cerebral metastases. **Neurology** 17:491–501, 1967.
44. Yasargil MG: Venous, cavernous and occult angiomas, in *Microneurosurgery: In 4 Volumes—AVM of the Brain, Clinical Considerations, General and Special Operative Techniques, Surgical Results, Nonoperated Cases, Cavernous and Venous Angiomas, Neuroanesthesia.* New York, Thieme Stratton, 1988, vol 3b, pp 405–438.

COMMENTS

The authors report a series of three patients having venous angiomas with the unique characteristic of arteriovenous shunting. In the first two patients presented, the venous angioma was found incidentally and was eventually followed up with contrast angiography, which confirmed the arteriovenous shunting. The third patient presented with sudden dizziness and mild headache and was found to have an intraventricular hemorrhage in addition to the venous angioma with arteriovenous shunting. The patients were all treated conservatively and had no subsequent cerebrovascular events during a period of 6 years, 1½ years, and 3 years, respectively. The authors then present a summary of 31 reported cases of venous angiomas with associated arteriovenous shunting, as well as extensive discussion of the pathogenesis of venous angioma with and without arteriovenous shunting, the clinical features of both, and a discussion of the therapeutic considerations of treating venous angiomas with and without arteriovenous shunting.

This is an excellent review of the literature. The authors present current and pertinent references. The images presented are clear and appropriate.

Although these patients have not had subsequent cerebrovascular events, this may not be representative of the natural history of these lesions if left untreated. Because they have clinical features similar to those of arteriovenous malformations, one would agree with Awad et al. (1) that these lesions should have a hemorrhagic incidence similar to that of arteriovenous malformations. Long-term follow-up and collection of data on the natural history of these lesions is necessary for us to further define other therapeutic options.

Donald W. Larsen
Interventional Neuroradiologist
Steven L. Giannotta
Los Angeles, California

---

1. Awad IA, Robinson JR, Mohanty S, Estes ML: Mixed vascular malformations of the brain: Clinical and pathogenetic considerations. **Neurosurgery** 33:179–188, 1993.

The authors provide an interesting report of three cases with a literature review regarding this unusual vascular anomaly. They present three distinct cases of patients who were found to have small-vessel arteriovenous shunting in the presence of a venous angioma. The authors then present a very lucent discussion of the various theories of pathogenesis of venous angiomas with and without the presence of arteriovenous shunting and base treatment recommendations on their review of the literature. The authors make the excellent point that when a venous angioma is associated with an intracerebral or intraventricular hemorrhage, suspicion of a coexisting cavernous malformation must be high. Cavernous malformations were probably significantly underdiagnosed before the introduction of magnetic resonance imaging. This is perhaps the singular flaw in the literature analysis, which mixes older and newer case reports encompassing both the pre- and post-magnetic resonance imaging eras.

The rate of subsequent bleeding quoted by the authors is significantly low as to affirm that these are indeed

relatively benign lesions. We have seen several lesions such as this which were initially misdiagnosed as either moya-moya disease, when they involved the proximal perforating vessels, or an arteriovenous malformation, when they were found distal on the arterial circulation. We agree that this entity is most likely a developmental hybridization of an arteriovenous malformation and a venous angioma. There is little to suggest that surgical removal is indicated unless necessitated by removal of a coexisting hematoma. There is also no evidence to suggest that radiosurgery has any role in the treatment of this entity. The authors bring to our attention an unusual vascular anomaly that can easily be misdiagnosed and, as best can be determined, has a relatively benign natural history.

Thomas A. Kopitnik
Duke S. Samson
Dallas, Texas