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Spontaneous Obliteration of Pial Arteriovenous Malformations: A Review of 27 Cases

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BACKGROUND AND PURPOSE: **Brain arteriovenous malformations (AVMs) occur in approximately 0.14% of the population. The most common presentations are hemorrhage (50%) and seizures (25%). Although they are congenital abnormalities, their angioarchitecture may vary over time. A rare but well-recognized phenomenon of AVMs is that of spontaneous obliteration. It is not known what factors predispose to spontaneous obliteration. The purpose of our study was to determine whether spontaneous thrombosis of AVMs can be predicted by their angioarchitecture and whether there is any risk of recurrence once obliteration has occurred.**

METHODS: **We retrospectively reviewed the angiographic and cross-sectional imaging data amassed over an 18-year period, including follow-up imaging studies and mail surveys of referring and family physicians. A control group was obtained from contemporaneous AVMs of a similar size.**

RESULTS: **We identified 28 cases of spontaneous obliteration in a series of 2162 patients. The mean time between initial diagnostic angiography and angiographic obliteration was 10 months, during which time there was no intervention and no history of repeat hemorrhage; nor had hemorrhage recurred during the follow-up period (mean, 53 months). Most of the AVMs were deep (22/27) with only one draining vein (21/27) and few feeding arteries. In more than half the cases (15/27) drainage was exclusively into the superficial venous system.**

CONCLUSION: **Spontaneous obliteration is rare (1.3%). Common features include hemorrhagic presentation and few arterial feeding vessels. Although we found no instance of repeat hemorrhage during the follow-up period, AVMs can recanalize, and follow-up is therefore recommended.**

Spontaneous obliteration of pial arteriovenous malformations (AVMs) is a rare but well-recognized event. Pathogenic factors that predispose to spontaneous obliteration are not well recognized, in part because of the rare nature of these events. Consequently, the largest reported series to date includes only six cases (1). Furthermore, the long-term outcome for these patients is unknown and hence there is no consensus about the length and nature of follow-up. We examined the angiographic anatomy and presenting symptoms in a large series of patients with spontaneously obliterating AVMs to determine whether spontaneous obliteration can be predicted and, once it occurs, whether it is likely to be permanent.

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Methods

We reviewed the AVM database, which includes all patients with AVMs accepted for stereotactic radiosurgery (STRS) at our institution, for cases of spontaneous obliteration of AVMs. To be accepted for treatment at our institution, patients had to have had a cerebral angiogram showing a pial AVM. From these patients we selected those in whom the AVM was no longer visible on angiograms at the time of their arrival for treatment. None of the patients had had a further neurologic event between their referral angiogram and their arrival at our institution. The patients' case notes, angiograms, and cross-sectional imaging studies (if available) were reviewed. Standard biographical data were collected for each patient. Information about presentation, timing of angiography relative to ictus (most presented with hemorrhage), and timing of angiography showing obliteration was also recorded. The timing and nature of any surgical procedures (including endovascular treatments) were collected. Details of the AVM (size, location, number and source of feeding arteries, number and direction of draining veins) were obtained from the angiograms and cross-sectional studies. Owing to variation in the quality of the angiograms, it was not possible to reliably determine details of the angioarchitecture of the nidus (eg, the presence of in-

tranidal aneurysms) in all cases, and therefore this information was not collected. Follow-up data were obtained from the case notes at our institution and, in addition, the referring clinician (neurosurgeon or neurologist) and the patient's primary care physician were contacted directly and asked to provide details of subsequent neurologic events and follow-up imaging. In those patients who had undergone an intervention before referral there was always a postprocedure angiogram depicting the continued presence of the AVM. The control group consisted of the nearest (in time) patients both before and after the index patient with a STRS treatment volume of less than 2000 mm^3 (to reflect the size of the AVMs of our index cases). The index cases and the control group were compared using the χ^2-test.

Results

Demographic Features

The study population included all patients referred to our facility for STRS from its inception (September 1985) to the start of our investigation (December 1998). The frequency of spontaneous obliteration of a pial AVM was 1.3% (28/2162 patients). Full angiographic data were available in 27 of these 28 cases, and these patients were included in the study. This group included 13 females (age range, 10–58 years; mean age, 35 years) and 14 males (age range, 13–66 years; mean age, 35 years). Two of the patients were under the age of 18 at presentation. The control group included 22 females (age range, 9–64 years; mean age, 29 years) and 32 males (age range, 8–64 years; mean age, 39 years).

Morphologic Characteristics

In the study group, the nidus was less than 2 cm in size in 93% (25/27) of the cases and less than 1 cm in size in 70% (19/27) of the cases. The AVMs were supratentorial in location in 78% (21/27) of the study population and in 83% (45/54) of the control group. Five (19%) of the 27 AVMs in the study group were superficially located (defined as gyral or less than 1 cm from the brain surface); 44% (12/27) had a single feeding artery (vs 44% [24/54] in the control group), 48% (13/27) had two feeding arteries (26% [14/54] in the control group), and one had four feeding arteries (in one case the number of feeding arteries could not be determined from the angiograms supplied). In the control group, 30% (16/54) had three or more feeding arteries.

Among the study population, 78% (21/27) had a single draining vein, 11% (3/27) had two draining veins, and 3% (1/27) had more than three draining veins. Corresponding numbers in the control group were 85% (46/54), 13% (7/54), and 2% (1/54), respectively. In the study group, drainage was exclusively into the deep venous system in 30% (8/27) and exclusively into the superficial venous system in 56% (15/27); corresponding figures for the control group were 24% (13/54) and 76% (41/54), respectively. In one case drainage was into both systems, and in three cases drainage was indeterminate (Tables 1–3,2,3 and Figs 1 and 2).

TABLE 1: Location and number of vessels associated with spontaneously obliterated AVMs

Case No.	Location	No. of Feeding Arteries	No. of Draining Veins
1	Genu corpus callosum	1	1
2	Superior vermis	1	2
3	Posterior pericallosal	1	1
4	L occipital lobe	2	2
5	L foramen of Monroe	2	1
6	L midbrain	2	1
7	R occipital lobe	1	1
8	L pulvinar (thalamus)	2	1
9	L frontal lobe	1	1
10	L internal capsule	1	Varix, vein not seen
11	R parietal lobe	1	1
12	L occipital lobe	1	1
13	Cingulate gyrus	2	1
14	R occipital lobe	1	>3
15	L central white matter	4	1
16	L perisylvian	2	1
17	L thalamus/midbrain	2	1
18	L medial temporal lobe	2	1
19	R frontal lobe	2	2
20	L cerebellum	2	1
21	R perisylvian	1	1
22	Vermis	1	Varix, vein not seen
23	R frontal lobe	2	1
24	Suprasellar	Indeterminate	1
25	R cerebellum	2	1
26	R frontal lobe	1	1
27	R thalamus	2	1

Presentation

Among the study population, 89% (24/27) presented with a hemorrhagic event (66% [36/54] in the control group), one had epilepsy, and two patients were asymptomatic. Of those with a hemorrhagic presentation, 20 had had a single event, two had had two events, one had had three events, and one had had four events. Seventy-four percent (20/27) of the patients had had no surgical or endovascular procedure with administration of glue before referral for STRS (including one asymptomatic patient and one who presented with epilepsy). In those patients who presented with hemorrhage, the mean time between hemorrhage and angiographic demonstration of an AVM was 7 days (median, 4 days), excluding the three very delayed cases in which the intervals were 3 months, 2 years, and 5 years, respectively. Of the seven patients who had had intervention before the present referral, two had had evacuation of a posterior fossa hematoma on the day of presentation, two had had attempted surgical removal of the AVM, and three had had partial embolization. In the control group, 13% (7/54) had had a prior neurosurgical procedure and 5% (3/54) had had embolization. Three patients had angiographic demonstration of an AVM at least 1

TABLE 2: Presentation, morphologic characteristics, and follow-up data for patients with spontaneously obliterated AVMs

Case No.	Age/Sex	Presenting Event	Nidus Size (cm)	Mode of Follow-up	Follow-up Duration (mo)
1	42/F	SAH, ICH, IVH	<1	C	67
2	37/F	ICH	1–2	C	3
3	55/F	SAH × 3	1–2		0
4	27/F	ICH	1	C, X, A	70
5	18/F	IVH	<1	C, A	104
6	44/M	ICH	<1	C, X	62
7	41/M	ICH	<1	C	4
8	30/F	IVH	<1	C, A	48
9	30/F	ICH	<1	C	10
10	43/M	IVH	<1	C	5
11	42/M	EP	2–3	C	31
12	58/F	ICH	1–2	C	32
13	52/M	SAH	<1	C	3
14	46/M	ICH	2–3	C	68
15	52/F	ICH, IVH	1–2	C	26
16	33/F	SAH	2–3	C, A	50
17	13/M	ICH, IVH	<1	C	56
18	51/F	IVH	1–2	C, X, A	64
19	61/F	Asymptomatic	<1	C, A	39
20	52/F	ICH	<1	C	39
21	54/M	Asymptomatic	<1	C	70
22	66/M	SAH	<1	C, X	112
23	19/F	ICH	<1	C, X	102
24	48/M	ICH	1–2	C	123
25	26/M	ICH	<1	C	149
26	10/F	SAH	<1	C	81
27	42/F	IVH	<1	C	1

Note.—SAH indicates subarachnoid hemorrhage; ICH, intracerebral hemorrhage; IVH, intraventricular hemorrhage; C, clinical; X, cross-sectional imaging; A, angiography.

month after the ictus or surgical intervention (at 3 months, 2 years, and 5 years, respectively). Two patients were asymptomatic at presentation. The first (a 61-year-old woman) presented with a left intracerebral hematoma and angiographic demonstration of separate, bilateral AVMs. The left-sided AVM was no longer present after hematoma removal and the right-sided (asymptomatic AVM) was no longer present at the time of STRS. The second asymptomatic patient (a 54-year-old man) had had STRS for a posterior fossa AVM and at follow-up angiography (3 years later) was noted to have a new right perisylvian AVM. At the time of his admission for STRS, the perisylvian AVM was found to be obliterated.

Follow-up Studies

Clinical follow-up was obtained in 96% (26/27) of cases (one patient could not be traced). Six patients had follow-up angiography and five patients had follow-up cross-sectional imaging. Only one patient had evidence of recurrence of an AVM either on imaging studies or at clinical examination throughout the maximum follow-up period of 149 months (range, 0–149 months; mean, 53 months). This patient (case 6) originally presented with midbrain hemorrhage and was found to have a midbrain AVM that drained into the vein of Galen. The lesion was found to be obliterated 4 months later, at the time of STRS. Five years later, the patient reported light-headedness and palpitations, but there was no clinical evidence of hemorrhage. Angiography showed a small (1-cm) AVM in the same location. The bulk of the venous drainage was now into the superior vermian vein with some drainage into the vein of Galen. At this writing, the patient is awaiting STRS (Fig 3).

Our study uncovered two adult patients (ages 30 and 54 years, respectively) in whom new AVMs developed during the period of surveillance. One of these patients had had a previous posterior fossa AVM treated with STRS while the second patient, with a left thalamic AVM that had spontaneously obliterated, was found to have a new cerebellar AVM at check-up angiography 4 years after her previous angiogram. At this writing, she is asymptomatic from this AVM and is awaiting STRS.

We found statistically significant differences between the control and study groups with regard to the frequency of hemorrhagic presentation ($P < .05$) and between the number of cases with two or fewer feeding arteries and those with three or more feeding arteries ($P < .01$). No other differences between the control and study groups were statistically significant.

Discussion

We prefer the term spontaneous obliteration to spontaneous thrombosis (as is used frequently in the literature), because the mechanism involved is usually not known and the latter term implies causality. Spontaneous obliteration of cerebral pial AVMs is a rare event that is said to occur in 0.8% to 20% of cases (1, 2). The incidence in our series, 1.3%, lies at the lower end of the reported range and is in keeping with the incidence reported in the larger of the previously reported series. We cannot claim that this is a true incidence, as our population was highly selected, in that it consisted of patient who were referred (introducing a referral bias) and suitable for STRS (introducing a size and location bias). Although spontaneous obliteration does occur with larger AVMs (3) it is more common in small ones and therefore the overall true incidence for AVMs of all sizes is probably lower than that which we report. This would tie in with our own clinical observations from non-STRS AVMs, for which we believe that the incidence of spontaneous obliteration is less than 1.3%. Therefore, this phenomenon cannot be relied upon in management of pial AVMs, in which the annual incidence of hemorrhage lies in the range of 2% to 4% (4, 5). This is particularly true in as much as no authors (including ourselves) have been able to identify reliable anatomic or angiographic predictors for spontaneous obliteration.

TABLE 3: Intervention and timing of obliteration associated with spontaneously obliterated AVMs

Case No.	Interval from Ictus to First Angiogram	Interval Between Angiograms (mo)	Other Intervention
1	2 d	13	· · ·
2	28 d	3	Attempted surgical removal (no AVM identified)
3	14 d	23	· · ·
4	1 d	5	Evacuation of hematoma
5	2 d	10	· · ·
6	3 d	4	· · ·
7	22 d	4	· · ·
8	3 d	7	· · ·
9	2 d	7	· · ·
10	1 d	4	· · ·
11	· · ·	28	· · ·
12	15 d	11	Attempted embolization (no glue administered)
13	24 m	4	Embolization (partial) × 2
14	3 m	3	Attempted embolization (no glue administered)
15	1 d	8	· · ·
16	5 d	49	Embolization (partial) × 3
17	6 d	4	· · ·
18	5 d	3	· · ·
19	· · ·	4	Embolization (partial)
20	10 d	7	· · ·
21	· · ·	9	· · ·
22	11 d	16	Attempted surgical removal
23	3 d	6	· · ·
24	60 m	12	· · ·
25	4 d	5	Evacuation of hematoma
26	10 d	18	· · ·
27	2 d	6	· · ·

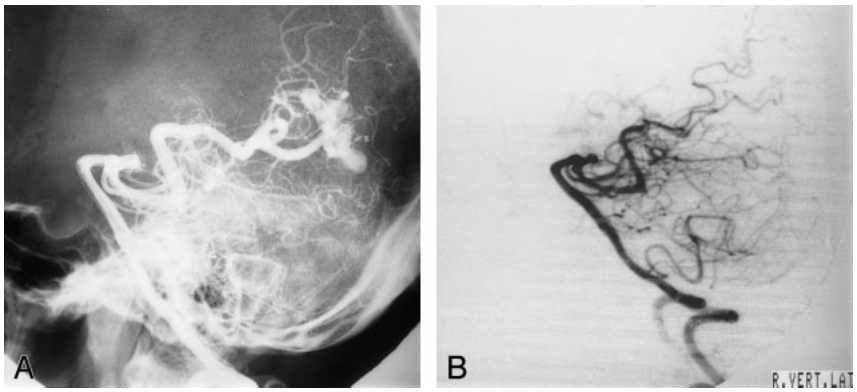

FIG 1. Case 14.
A and *B,* Vertebral angiograms (lateral projections). The first angiogram (*A*), 3 months after an intracerebral hemorrhage, shows a 2- to 3-cm right occipital AVM. The second angiogram (*B*), after a further 3 months, at the time of admission for STRS, shows absence of the previously visible AVM.

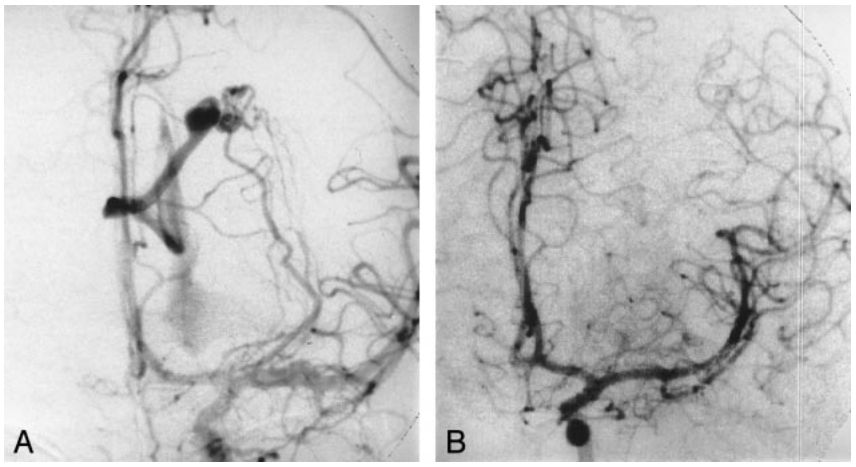

FIG 2. Case 15.
A and *B,* Left internal carotid angiograms (frontal projections) show absence of the AVM after an interval of 8 months. The patient presented with hemiplegia (Glasgow Coma Scale score = 4) associated with an intracerebral hematoma.

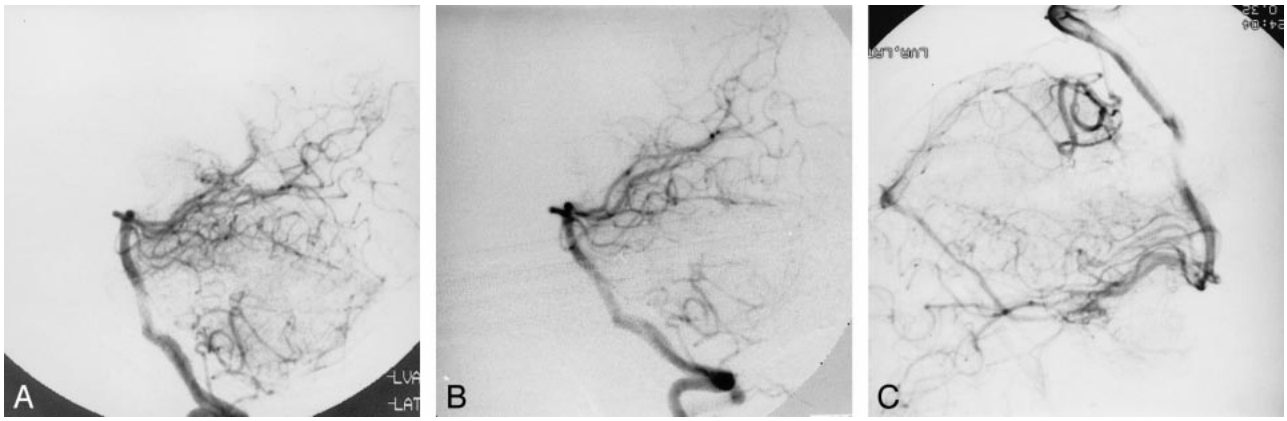

FIG 3. Case 6.

A–C, Vertebral angiograms (lateral projections) at presentation (*A*), at presentation for STRS 4 months later (*B*), and at time of recurrence 6 years later (*C*). A barely visible AVM is shown with early shunting of contrast medium into the straight sinus (*A*). No AVM is seen 4 months later (*B*). Six years later, the angiogram shows an AVM in the same location but with predominate early filling of the superior vermian vein (*C*).

Postulated causes of spontaneous obliteration include premature atherosclerosis (6), embolus (7), turbulence in feeding vessels (8), elevated estrogens (9), hemodynamic changes associated with surgery (10), or kinking of feeding vessels as a result of gliosis (11). Since we did not find a preponderance of females in our cohort we find it difficult to support the theory that estrogens play a role in spontaneous obliteration. Similarly, while we cannot disprove the notion that hemodynamic changes after surgery may lead to obliteration, the majority of our patients had no surgical intervention and there was no statistical difference in the number of spontaneous obliterations between the control group and the index cases. It is certainly possible that surgery either for hematoma evacuation or for intended resection may remove enough nidus for an alteration in flow pattern to take place and lead to obliteration; however, unless patients undergo angiography immediately before and after surgery, and at later intervals, this cannot be proved. Furthermore, the fact that of the 2162 cases treated with STRS at our institution 24% had had previous surgery and that in the spontaneously obliterating subgroup only 15% (4/27) had had surgery would support the view that surgery does not contribute a late protective effect. We do not think that our data shed further light on any of the other proposed theories.

Mass effect due to parenchymal hemorrhage has also been suggested as a contributory factor (12), possibly as a consequence of stretching and narrowing of the feeding arteries, occlusion of the draining veins, or obliteration of the nidus. At our institution, 80% of AVMs present with hemorrhage, and in our series, 89% of the index cases but only 66% of the control subjects presented with hemorrhage. The difference in the rate of hemorrhagic presentation between the index cases and the control group is statistically significant, suggesting that there may be a link between hemorrhage and spontaneous obliteration. Quite what that link is is difficult to identify. Mass effect would seem to be the most plausible pathogenesis, but while 89% (24/27) of our cases presented with hemorrhage, only 58% (14/24) of those patients had parenchymal hematoma, the rest having exclusively subarachnoid or intraventricular hemorrhage, which would seem unlikely to have caused mass effect. Furthermore 21% (3/14) of those patients had angiographic demonstration of their AVM more than 28 days after the ictus, by which time any mass effect should have been diminishing and we would have expected obliteration, if it were due to mass effect, to have occurred.

Other authors have observed that spontaneous obliteration is more likely if the AVM is small, superficial, fed by a single artery, or drained by a single vein (2). In our series, all the AVMs were small (because of the nature of our referrals) and there was a preponderance of deep AVMs, namely 81% (22/27), but again this might be explained on the basis of selection bias (superficial AVMs are more likely to be treated by endovascular or surgical approaches). In 78% (21/27) of our cases there was only a single draining vein, and while 81% (22/27) were deep in location, 56% drained exclusively superficially. A solitary draining vein, which had a tendency to be long, was the most frequent anatomic finding in our patients, but this was also the case in the control group. This finding does not support previous suggestions that restriction of venous outflow on its own causes spontaneous obliteration, but a more complex relationship may exist, as it is also known that outflow restriction is a risk factor for hemorrhage from an AVM (13) and we have shown that hemorrhage may be linked to spontaneous obliteration. There was, however, a difference between the control and study groups with regard to the number of feeding arteries; although how hemorrhage (but not mass effect) and the presence of fewer feeding arteries (but no difference in the number of draining veins) are related is difficult to explain. We suspect that no in-

dividual factor will be found to account for spontaneous obliteration, but perhaps progressive venous restriction leading to venous occlusion in the presence of a protective factor may offer an explanation. Such protective factors may include, for example, a paucity of feeding vessels, or long feeding vessels with a large pressure drop along their length, or spasm of feeding vessels in the presence of hemorrhage. All these factors may prevent an increase in intranidal pressure during venous occlusion, thereby limiting the size of hemorrhage from the nidus. Large variations in feeding artery pressure have already been reported, with smaller AVMs having higher feeding artery pressure than larger ones and those that present with hemorrhage having higher feeding artery pressure than those with nonhemorrhagic presentations (14).

Follow-up studies of patients with AVMs that have been treated by surgery suggest that there is a risk of AVM recurrence, especially in children (15, 16). In our series, only one patient had a proved recurrence of an AVM during the maximum follow-up period of 149 months. We found just two reports of recurrence of an AVM after spontaneous obliteration (1, 12), and in both these cases there was at least some dural arterial supply to the nidus. We excluded cases of spontaneously obliterating dural arteriovenous fistulas (AVFs) (defined as having a dural supply) from our series (we found two cases in our database) in an effort to keep our results pure, although we accept that cortical AVMs may have a dural supply and that dural AVFs may recruit pial vessels. A criticism of our study would include the lack of angiographic follow-up in all patients, and, indeed, our results may be interpreted to show that the recurrence rate is 15% (one of the six cases with follow-up angiography had a recurrence, but that patient was symptomatic, which prompted angiography). We believe that the study length was such that if there was a tendency for rehemorrhage to occur we might reasonably have expected patients to have presented clinically.

Interestingly, two of our adult patients (ages 30 and 54 years, respectively) developed new AVMs (neither of which caused symptoms) during the follow-up period. Our database cannot be searched for such occurrences, but none of us can recall any other such events. In one of these patients, the new AVM obliterated spontaneously. These cases only serve to increase confusion as to the cause of pial AVMs, which are thought to be congenital lesions that are capable of growth during childhood and subject to remodeling thereafter. Our data do not shed further light on this debate, but merely add to the information available.

Conclusion

Spontaneous obliteration of pial AVMs is rare, with a prevalence of less than 1.3%. It is more likely to occur in small AVMs that present with hemorrhage and have few arterial feeders. In our series, no case of recurrent hemorrhage was seen, despite a maximum follow-up period of 149 months. However, recanalization may occur, and continued vigilance is recommended.

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