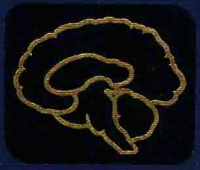
CURRENT NEUROSURGICAL PRACTICE



Intracranial Arteriovenous Malformations

Edited by

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CURRENT NEUROSURGICAL PRACTICE

Intracranial Arteriovenous Malformations

Foreword

Long overdue is a comprehensive volume with a distinguished authorship to review the nature and treatment of intracranial arteriovenous malformations (AVM's). Although cerebral AVM's are at times considered to have a cloak of benignity, they are dangerous lesions, as we are reminded in these chapters. Their bleed and rebleed rates approach those of healed or intact aneurysms, and they differ from aneurysms only in regard to their somewhat reduced arterial pressure, a virtual absence of ischemic calamities associated with vasospasm, and consequently lower mortality rates. In contrast to most aneurysms, the risk of operative morbidity in resecting large AVM's from eloquent cortical regions, or even smaller ones from the basal ganglia, is much higher than is the overall potential for morbidity in the lesion's natural course. What this volume addresses, in part, is this prognostic dilemma and the options open to the neurosurgeon in confronting it. Microsurgical techniques have expanded considerably the potential for reasonably safe surgical resection of larger AVM's and AVM's in critical cerebral regions. Still, there is no consensus about whether surgical intervention is indicated for a patient with a precariously situated AVM whose only symptom is headache or a convulsive disorder. Alternative approaches by embolization and high energy radiation therapy, used either alone or as adjuvants, are evolving also—although currently their long-term benefit is not assured unless the AVM has been obliterated. Omitting only a discussion of the frustrating inadequacy of the instrumentation currently available for hemostasis, the results and complications of proton therapy, and variant techniques for embolization, this authoritative volume represents the current state of the art in the treatment of AVM's.

CHARLES G. DRAKE, M.D.

Preface

To inaugurate this new series on *Current Neurosurgical Practice*, I chose intracranial arteriovenous vascular malformations (AVM's) as the subject of the first volume. Setting aside my own interests, I compiled a list of neurosurgical conditions in which current practice differs significantly from practice in the past with respect to knowledge about the particular condition, therapeutic advances, and operative techniques. For a number of my chosen topics, recently published monographs precluded the usefulness of a redundant publication. From the shortened list, I selected vascular malformations as the area in which a volume on the current status of diagnosis and therapy would be most timely.

With the precision afforded by computerized tomography (CT), increasing numbers of vascular malformations are being identified. Malformations suspected on the basis of CT scans are being confirmed by angiography; and arteriovenous, racemose, and venous malformations that cannot be identified angiographically can be detected by CT scanning. Surgical and anesthetic techniques have broadened the indications for operations on these anomalies, and they are performed now with a lower risk of morbidity or mortality. The pathophysiology of disturbed blood flow in "normal" brain surrounding AVM's has been clarified from the standpoints of neurologic manifestations and therapeutic implications. Finally, endovascular techniques have emerged as a major therapeutic advance, providing definitive treatment in some instances, and reducing the blood flow as a preoperative step in others.

With a raison d'être, I approached Dr. Bennett Stein, whose enthusiasm for the subject equaled my own. We trust that our readers will agree with our decisions regarding the manner of presentation and the authors invited to contribute to this volume.

Acknowledgments

The editors acknowledge with deep gratitude the yeoman's task of editing performed by Susan Eastwood-Berry. Her charge has been particularly demanding because of the many authors and their variety of writing styles. Respecting their depth of personal experience and applying her talent, skill, and her tenacity to detail, she has blended these authors' contributions into a comprehensive and easily flowing text on the subject of vascular malformations of the brain.

J. Lawrence Pool, whose vast experience in the treatment of AVM's at a time when surgical skill and his ability to communicate this skill to his pupils were the only weapons against these devastating lesions, has been a prevailing influence on the editors in the compilation of this book. Many of his precepts in the surgical treatment of AVM's formed the foundation of surgical techniques described herein. Without his stimulus in the treatment of these lesions and his thrust into microsurgery at its infancy, we would not be where we are today.

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CHAPTER ONE

Neurological Manifestations and Factors Related to Therapeutic Decisions

Jay P. Mohr, M.D.

In view of the referral patterns for patients with an arteriovenous malformation (AVM), it is not surprising that the larger series are reported mainly from surgical clinics, and that these are the source of most of the data available regarding AVM's. Because of the low incidence of these lesions, most physicians and surgeons rarely encounter them. As a result, the published experience with AVM's is concentrated in reports from remarkably few clinics. The largest single series is the 545 cases reported from the Cooperative Study (47), an effort involving many centers; and among series of AVM's published over a period of almost half a century, hardly more than a dozen comprise more than 100 cases (10, 14, 19, 31, 35, 36, 43, 44, 47, 48, 50, 56, 61, 63). The time required to accumulate most of these major series has been a period of decades. As referring physicians have become more aware that definitive therapy for AVM's is possible, the data base is changing.

INCIDENCE AND PREVALENCE

In the Cooperative Study (47), symptomatic AVM's were found in 545 of 6368 cases, which yields an incidence of 8.6% of subarachnoid hemorrhage (SAH). By extrapolation, as SAH accounts for roughly 10% of strokes, approximately 1% of all strokes are associated with AVM's. These figures for the *incidence* of AVM's were reflected in our experience in the Harvard Cooperative Stroke Registry project (34). In a prospective study recently completed in South Alabama in an eligible population of 100,000 studied over a period of 3 yr, we encountered nine AVM's among 494 cases of stroke from all causes, yielding an incidence of 1.8%. Data on the *prevalence* of AVM's are more difficult to obtain but are equally important, especially in efforts to assess the risk for stroke in asymptomatic cases. For example, a high ratio of asymptomatic to symptomatic AVM's might encourage less aggressive management for the asymptomatic cases, which are being detected more frequently as computerized tomography (CT) scanning becomes generally available.

The clinical awareness of AVM's has increased over the past few decades. Early studies suggested a very low prevalence of AVM's, 0.8% in the autopsy studies by Courville in 1945 (6). The prevalence reported by Jellinger (22) in 1972 confirmed the figure of 0.8%, whereas that reported in 1978 by Sarwar and McCormick (51) was 4.05%. Their study (51) revealed 165 vascular malformations of all types among 4069 consecutive autopsies. Although their data were based on autopsy findings rather than findings in a clinical population, they represent a careful effort to document the prevalence of vascular malformations, whether symptomatic or not. Only 24 of the 165 malformations were arteriovenous (AVM's) in type, which represents 0.59% of the total cases; and the largest group, 105 cases, were venous malformations. The majority of the AVM's had produced symptoms; 19 had caused hemorrhage, which was massive in 16 cases.

Age of Onset

As referring physicians become more aware that AVM's can be detected by radiographic methods and effectively treated with surgery, AVM's are being diagnosed in greater numbers of older patients. Consequently, the presumed age of onset of

AVM's has shifted upward (Table 1.1); and although most hemorrhages from AVM's occur in the younger age group, this lesion is no longer considered a disorder mainly involving the young.

Prevalence by Sex

A predominance of AVM's among males has been noted in virtually all series (Table 1.2). This apparent predilection of the lesion is not easily explained by patterns of referral, and probably reflects a true characteristic of AVM's.

Family History

As AVM's are presumably congenital, it might be expected that there would be many cases of a family history of AVM—but such cases appear to be rare (Table 1.3). Only seven such families, involving 15 people in all, were reported as of 1981. The mode of inheritance has not been explained. Despite the general predisposition of males for AVM's, members of both sexes are represented equally in the sparse data on family history.

CLINICAL FEATURES

AVM's are well known to produce the triad of hemorrhage, seizures, and recurrent headaches. Each of these features is thought to have a character distinctive enough to suggest a diagnosis of AVM on the strength of a single symptom. However, the basis for this notion is not documented.

Hemorrhage

Approximately 50% of AVM's present clinically as an intracranial hemorrhage (35, 36, 43, 44, 47, 48, 56, 58, 63). Most frequently, the hemorrhage is primarily parenchymatous. This occurs in approximately 63% of cases. SAH occurs in 32% of cases, and ventricular hemorrhage is the least frequent, occurring in 6% of cases (47, 48). At surgery, about 30% of AVM's show evidence of prior hemorrhage (33); most frequently these are smaller AVM's (35, 43, 48, 59). Such a high incidence of prior bleeding clearly indicates that many bleeding events escape clinical detection.

Most authors (14, 19, 24, 35, 43, 47, 64) concur that the smaller AVM's appear to be

Table 1.1.
Age at Onset of AVM Symptoms

Year	Authors	Decade in Life >10	>20	>30	>40	>50	>60	>70
1980	Parkinson & Bachers (43)	4	10	7	15	10	4	2
1979	Pertuiset et al. (48)	10	27	43	44	21	15	2
1979	Nornes et al. (39)	4	18	10	9	11	6	5
1970	Moody & Poppen (35)	12	15	27	21	16	12	2
1966	Perret & Nishioka (47)	15	56	66	70	48	39	10
1965	Svien & McRae (59)	13	22	26	19	11	4	
1958	Dimsdale (9)	5	8	11	12	8	5	1
1956	Paterson & McKissock (44)	11	38	26	23	7	4	
1953	Mackenzie (33)	5	24	6	9	5	1	
1948	Olivecrona & Riives (41)	4	14	13	6	6		

Table 1.2.
Predilection of AVM's by Sex

Year	Authors	Male < Female		Ratio
1980	Guidetti & Delitala (19)	89	56	1.59
1979	Pertuiset et al. (48)	102	60	1.70
1979	Nornes et al. (39)	40	23	1.74
1973	Morello & Broghi (36)	88	66	1.33
1972	Forster et al. (14)	99	51	1.94
1970	Moody & Poppen (35)	65	40	1.63
1966	Perret & Nishioka (47)	236	217	1.09
1956	Paterson & McKissock (44)	63	47	1.34

Table 1.3.
Family Histories of Patients with AVM's[a]

Year	Authors	Age/sex	Relationship	AVM site
1947	Kidd & Cumings (23)	22 M	Cousins	Right temporal
		? F		Left parietal
1951	Griepentrog (18)	? M	Brothers	Left parietal
		? M		Left temporal
1974	Laing & Smith (28)	16 F	Sisters	Right frontal
		29 F		Right temporal
1977	Stoll & Wolfram (58)	66 M	Fathers	Left temporal
		37 F	Daughters	Left parietal
1978	Barre et al. (4)	30 M	Brothers	Left basal ganglia
		39 F	Sisters	Right frontotemporal
1979	Snead et al. (53)	17 F	Half-sisters	Left parietal
		11 F	Sisters	Lateral ventricular floor
		14 M	Brothers	Deep thalamic
1981	Aberfield & Keshav (1)	23 F	Sisters	Left frontal
		32 M	Brothers	Left parietal

[a] Data from Aberfield DC, and Keshav RR: Familial arteriovenous malformation of the brain. *Neurology* 31:184, 1981.

more dangerous than the giants. In Morello and Broghi's series (36), rupture occurred in 86% of small AVM's and in 75% of medium-size AVM's, but in only 46% of giant AVM's. These findings, which are consistent with those in other series, suggest that the larger the lesion, the longer it has been present and the less likely it is to rupture. Another factor that has been associated with the rupture of AVM's is physical exertion. However, some authors have found no correlation between such activity and AVM rupture (45) and argue against advising asymptomatic patients to live a restrictive and sedentary life.

CLINICAL SYNDROMES OF HEMORRHAGE

Because the pathologic features of AVM's differ markedly from those of aneurysms and hypertensive hematomas, one might imagine that a clinical differentiation between these lesions should pose no problem. Because many AVM's involve central white matter and subcortical areas of the cerebrum, but usually extend also to the ventricle or cerebral surface, they may produce parenchymatous, subarachnoid, or ventricular hemorrhage, or a combination of these. Because bleeding may originate on the venous side of the arteriovenous shunt, as a rule the effect of the hemorrhage is less violent than is that from an aneurysm; and unlike aneurysmal hemorrhage, which characteristically occurs within seconds, often it evolves over a protracted period of time. Because the hemorrhage arises within the bulk of the AVM, it has a less disruptive impact on cerebral function than does hypertensive hemorrhage. Vasospasm, often a complication of ruptured aneurysms, occurs infrequently with AVM's because the SAH from an AVM is located away from the base of the brain and is associated with a smaller volume of blood entering the subarachnoid cisterns.

Despite all these differences, few cases reported in the literature are described well enough to enable one to draw a characteristic clinical picture of AVM rupture. Even the few descriptive clinical reports that are available make the syndromes of hemorrhage seem decidedly pedestrian. In the most detailed account, which appeared in 1953, Mackenzie (33) states simply that, "in most cases there has been nothing remarkable about the history, the incident being simply one of sudden onset of severe headache, accompanied by neck stiffness, vomiting and perhaps pyrexia."

Reports of deep hematomas from AVM's in the basal ganglia (7, 16, 48, 66) describe the same smooth onset, hemiparesis, sensory disturbance, ocular motility disorders, and language and mental defects that are encountered in cases of hypertensive hemorrhage (20). Three such cases in the series of

Wilson et al. (67) were diagnosed only after surgical exploration, a finding that lends support to Sarwar and McCormick's (51) contention that cryptic AVM's are more prevalent than has been supposed. There are data to indicate that AVM's may cause as few as 10% of parenchymatous hematomas (29), but the frequency reported in other series is as high as 35% to 44% (25), which should warn against complacency in attempting to make an etiological diagnosis on clinical grounds alone.

PROGNOSIS AFTER HEMORRHAGE

It appears that a good prognosis can be expected in cases of AVM hematomas. In Pia's series (49), among 16 patients with hematomas ranging from 100 to 250 ml, only 1 died and 14 regained full capacity. Four patients had slight to moderate hemiparesis, and two were asymptomatic. In Pia's experience, ". . . particularly impressive was the reversibility or marked improvement of severe deficits even in cases with large hematomas . . . extensive lesions of the basal ganglia and thalamus seemed to be tolerated quite well and were connected with relatively little or no neurologic deficits. . . ."

A few patients have had remarkable functional improvement, despite hemorrhage and extensive surgery. Garrido and Stein (16) described a 29-yr-old man with an extensive lenticulostriate AVM that presented as a hematoma causing left hemiparesis, hemianopsia, and poor memory. After removal of this deep-seated lesion, the patient eventually improved enough to return to work. Schlachter et al. (52) reported a 27-yr-old man with initial flaccid hemiplegia resulting from hemorrhage from a parietal AVM. Postoperatively, he was able to ". . . regain most of his lost function." The outlook may not be so encouraging for all patients who have had hemorrhage, however, Pertuiset et al. (48) found that 19 of their patients with aphasia caused by hematoma made no postoperative improvement; as best can be inferred from this text, the aphasias were of the Broca or Wernicke types.

The preceding discussion suggests that the outlook for patients with AVM's may be better than that for patients with parenchymal hemorrhage from other causes. It may be incorrect, however, to consider that hematomas from AVM's are unique in this respect, because a favorable outcome can follow deep hypertensive hematomas if their size is small (20). As yet, no study has compared the outlook for hematomas of the same size from AVM's and other causes. Should AVM's have a better prognosis for recovery of function, it may be appropriate to deal more aggressively with such hematomas than is routinely the case in many institutions.

RECURRENT RUPTURE

Once hemorrhage has occurred, the risk of rebleeding is greater, but the extent and the timing of the later hemorrhages cannot be predicted. In 81 cases of recurrent hemorrhage reported in the Cooperative Study (47), 13 were the third, and 4 were the fourth, hemorrhage. In another study, Krayenbühl and Yasargil (25), found that of 53 recurrent hemorrhages, 12 were at least the second recurrent hemorrhage. It is generally acknowledged that approximately 30% of operated AVM's show evidence of prior hemorrhage when the surgical specimen is inspected (35, 48, 56).

Vasospasm

The rarity of vasospasm in association with AVM's—whether the vasospasm is symptomatic or noted on angiography—has been a source of special interest, but to date, it has not been fully explained (3, 17, 43, 49, 60). In contrast to SAH from intracranial aneurysms, SAH from AVM's ordinarily does not result in a massive accumulation of blood in the basal cisterns that encases the larger cerebral arteries. It may be this difference that accounts for the high incidence of spasm following aneurysm rupture and the rarity of spasm after rupture of an AVM.

Seizures

Seizures may alert the physician to the AVM before it ruptures. As a presenting feature of AVM's, their incidence varies from 28% (47) to 67% (49) (Table 1.4). The frequency of seizures correlates so poorly with the location of the AVM that, at present, no specific relationships have been defined. Similarly, reports vary too greatly to allow definition of relationships between the

Table 1.4.
Seizures as the First Sign of the AVM in Percent of Cases

Year	Authors	No. cases	Total	Alone	+ Hemorrhage	Generalized	Focal
1981	Stein[a]	121	43.8	36.3	7.4		
1980	Parkinson & Bachers (43)	100	67.0				
1979	Pertuiset et al. (48)	162	37.6	25.3	12.3	41.0	59.0
1973	Morello & Broghi (36)	154	35.0	20.1	14.9		
1970	Moody & Poppen (35)	105	50.5	40.0	10.5	55.0	45.0
1967	Tönnis (61)	215	48.3				
1966	Perret & Nishioka (47)	406	28.0				
1956	Paterson & McKissock (44)	110	46.4				

[a] Bennett M. Stein (personal communication, 1981).

size of the lesion (35), the ease with which seizures can be controlled with medication (54), and the probability of hemorrhage.

CLINICAL FEATURES OF SEIZURES

The types of seizures occurring with AVM's are not described in most reported series. Among those in which they are described, focal spells constitute from 45% (35) to 59% (46) of seizures. In the Cooperative Study (47), of 102 cases involving seizures, 45% were focal, 42% were generalized, 8% were psychomotor, and 7% were of an unspecified type.

Olivecrona and Riives (41) recorded more variation in the type and frequency of seizures from AVM's than in those from cryptogenic or traumatic epilepsy. Mackenzie (33) made three pertinent observations: (*a*) in all but one of 16 cases, the attacks displayed focal features at some time or other; (*b*) the periodicity of focal seizures varied widely, ranging up to 20 yr; (*c*) when the initial seizure was generalized, there was no period of remission longer than 3 yr. Ozer et al. (42), however, were unable to distinguish between the focal seizures from AVM and those of other etiology.

Troost and Newton (62) described focal seizures associated with occipital AVM's in 5 of 26 patients and reviewed the related literature on these lesions. The aurae experienced by patients vary, some reporting "sudden dimming of everything in the right side of vision," "swirling spots of brightly colored lights," "dimming of vision," and "red spots," and others seeing as if through "frosted glass." In several cases, patients subsequently had a generalized seizure. Unfortunately, each of these symptoms may occur with occipital seizures of any cause, and none seems to be unique to occipital AVM's.

RELATION OF SEIZURE TO HEMORRHAGE

The incidence of seizures alone as compared to that of seizures occurring in association with hemorrhage varies among the few series in which this relationship has been documented: 36.3 to 7.4% in one series (56); in others 25.3 to 12.3% (46), 20.1 to 14.9% (36), and 40.1 to 10.5% (35). The association of seizures and hemorrhage is high in cases of AVM's involving the surface of the brain, especially for centroparietal lesions (35, 48, 65), but is unusual for deep AVM's (47). Hemorrhage occurred within 1 yr in only 15% of 90 cases of seizure reported in the Cooperative Study. Whether the character of the seizure differs when associated with hematoma has not been established.

Headache

The claim that recurrent unilateral headache and migraine are warnings of AVM or aneurysm has been a topic of interest. How notions such as this gain recognition is not easy to determine, but this particular one does not appear to be well documented (2, 5, 8, 9, 11, 21, 29, 40). Because of the difficulty in quantitating such a relationship,

no useful data on headaches was generated from the Cooperative Study.

UNILATERAL HEADACHE

The association of ipsilateral headache with AVM's may derive from Northfield's report (40) in 1940 that headache ". . . may affect only one side of the head, usually the side on which the angioma is situated," but there is scant data to support this claim. There were 11 patients with headache in Lees' series (29) of 70 with AVM's, but only 3 in whom it was consistently on the same side; in none were there alternating headaches typical of migraine. Mackenzie (33) described 12 cases in which the headache was persistently unilateral, but other focal neurologic signs were present, as well. In the later literature, no cases have been described in enough detail to pursue the point, although Parkinson and Bachers (43) recently reported finding no evidence, in their experience, that the incidence of headache has a specific relation to location of the AVM. The question seems to have been tabled; nonetheless, whether there is a correlation of AVM's with unilateral headache remains an unsettled issue.

MIGRAINE AND AVM HEADACHES

The early literature contained reports of migraine headaches accompanying occipital lobe AVM's; in some cases, the headache disappeared postoperatively (2, 5, 8, 9, 11, 21, 29). Although Troost and Newton (62) claimed that headaches caused by AVM's do not produce an aura of angular, scintillating figures, there can be no doubt that the symptoms in many of the early cases were migrainous. Enoksson and Bynke (11) described a patient who reported ". . . luminous crosses replaced by curved flashing lights with convexity upwards." Dimsdale's (9) patient described "flickering lights." Similarly, Lees' (29) patients described "flashing lights," and "flashes and black spots." Others simply described the headaches as being migrainous or atypical. As these are mostly reports of individual cases, it may be that the relation of migraine and AVM is only coincidental.

Paterson and McKissock's series (44) of AVM's occurring with headache was large enough to support the argument that the incidence of migraine in series of AVM's may simply reflect the incidence in the normal population. Common migraine has been estimated to occur in 10% of the population and classic migraine in 2%; these approximate the figures for patients with AVM's. Authors more recently have not been able to show any correlation between migrainous headache and AVM's (29, 42).

The most detailed description suggesting that AVM's cause an atypical form of migraine is found, once again, in Mackenzie's work: "With one exception it has been a symptom which appeared early. . . . Whereas in ordinary migraine the attacks follow a regular pattern in which the aura, lasting usually from a few minutes to 20 min or ½ hr, is then replaced by headache, in these cases the aura may be more prolonged. It may persist even after the headache has developed and sometimes the manifestations which one would normally regard as an aura may not develop until after the onset of headache" (33).

Cerebrovascular Steal Syndromes

Although hemorrhage, seizures, and headache in relation to AVM's are incompletely understood, these symptoms have at least been subjected to considerable study. By comparison, clinical information about steal syndromes is still in the anecdotal stage. It is presumed that the blood shunting through the fistula results in relative underperfusion of the adjacent brain, leading to focal or generalized symptoms. That shunting occurs has been amply documented by techniques and observations described in other chapters of this book and elsewhere (12, 56, 57). Yet there are remarkably few reports that describe the clinical deficits resulting from cerebrovascular steal. As early as 1949, Norlen (37) described a 19-yr-old male who had presented in 1946 with left-sided, spastic hemiparesis following hemorrhages in 1942 and 1945. Within 2 months after the AVM was excised, there was ". . . pronounced improvement in the paresis and the patient was in good condition," and he returned to full working capacity by 1948. Similarly, in Paterson and McKissock's series (44) of 110 patients with AVM's, there were eight whose progressive hemiparesis was ". . . gradual in onset, unaccompanied by features suggestive