The treatment of associated intracranial aneurysms and arteriovenous malformations

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Cerebral arterial aneurysm associated with arteriovenous malformation (AVM) has been described with a variable incidence, averaging 10% of AVM cases. The present series includes 39 patients with this association, derived from a total of 400 patients with AVM's evaluated and treated since 1970. The aneurysms are classified into four major groups, each carrying particular therapeutic implications. Optimum treatment of these lesions is based in part on a knowledge of the hemodynamic alterations associated with the AVM's. In most of these cases, the symptomatic lesion was treated first; occasionally, when feasible, both lesions were treated during the same operation. All patients had some form of treatment, either surgical or endovascular, directed to at least one of the two types of lesions. All symptomatic lesions were treated and all ruptured aneurysms were obliterated. There were no deaths in this series.

KEY WORDS: aneurysm · arteriovenous malformation · subarachnoid hemorrhage · embolization · craniotomy · cerebral blood flow

The association of intracranial arterial aneurysm and arteriovenous malformation (AVM) has been reported with an incidence of approximately 10% of AVM cases.1-4,7-9,11,13-15,21-25,28,29,34-40,45,51,53 Most authors have analyzed this coexistence from the aspect of pathophysiology and epidemiology. Less emphasis has been given to the therapy of these combined lesions.4,7,9,18-21,25,26,34-40,51,53 The consensus of opinion now favors early clipping of symptomatic intracranial aneurysms. With an increasing understanding of the natural history and hemodynamic changes associated with AVM's, a tendency is developing to treat these lesions whether they are symptomatic or not.5,6,18,30,32,33,35-50 When feasible, as in most of our cases, it has been our policy to recommend surgical resection of an AVM.49,50

Clinical Material and Methods

Incidence and Classification of Lesions
Our experience is based on 39 patients with both intracranial arterial aneurysms and AVM's, representing 9.75% of 400 patients with cerebral AVM's treated between 1970 and 1990. There were 13 (33%) males and 26 (67%) females. The patients' age at the time of presentation ranged between 12 and 69 years (average 38.4 years). Excluding infundibular dilatations (< 3 mm), 84 aneurysms were identified by angiography. Twelve patients (30.8%) harbored multiple aneurysms, ranging from two to five. The fact that there was a high incidence of posterior circulation aneurysms (25 cases (39%)) as well as distal and "bizarre" (see below) aneurysms (18 cases (28%)) indicated that the location of these aneurysms was different from those not associated with AVM's.26 Furthermore, the number of aneurysms arising from the anterior cerebral arteries (nine cases (14%)) was lower than expected.

We have expanded the traditional scheme of classifying aneurysms associated with AVM's to five types, including one subcategory (Fig. 1, Table 1).19,23,35 Among the group of aneurysms arising from arteries related to the AVM, four categories were distinguished: Type I aneurysms were seen in 39 cases (61%) and occurred proximally on major ipsilateral arteries contributing to the AVM (Fig. 2). The six cases (9.4%) of aneurysms occurring contralateral to the malformation but bearing an important indirect relationship via the circle of Willis were included in Type IA. There were 10 cases (15.6%) of Type II aneurysms, occurring distally on superficial feeders to the AVM. Type III aneurysms (eight cases (12.5%)) occurred proximally or distally involving deep arteries feeding the AVM (Fig.
3). This latter group included aneurysms that we have designated as “atypical” or “bizarre” because they involved deep arteries such as the posterior or anterior choroidal, lenticulostriate, and thalamoperforating arteries (unusual locations for aneurysms). One Type IV aneurysm (1.6%) occurred on an artery anatomically and hemodynamically unrelated to the AVM.

There was excellent correlation between the location of the AVM and that of the aneurysms, as can be concluded by the almost exclusive distribution of cases in Types I, IA, II, and III. Giant aneurysms, rarely described in association with AVM’s, were found in three of our cases, two of Type I and one of Type IV. The AVM’s were separated into three categories according to size: large (> 5 cm, 20 cases), medium (2.5 to 5 cm, 11 cases), and small (< 2.5 cm, eight cases). No significant correlation was found between the size of the AVM’s and the type, number, or size of the aneurysms with which they were associated.

**Clinical Features**

Regarding initial presentation, hemorrhage occurred in 62% of cases, seizures in 21%, headaches in 5%, and mass effect in 8%. When identified, the hemorrhage was due to the aneurysm in 46% of cases and to the AVM in 33%. Eleven patients presented with aneurysmal rupture into the subarachnoid space. Seven of these patients had Type I and four had Type II aneurysms. The single Type IV aneurysm had not bled. In 21% of patients, imaging data and surgical observation did not pinpoint the cause of the hemorrhage. These were patients with parenchymal hemorrhage who harbored

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**TABLE 1**

<table>
<thead>
<tr>
<th>Type</th>
<th>Location of Aneurysm</th>
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<tbody>
<tr>
<td>I</td>
<td>proximal on ipsilateral major artery feeding the AVM</td>
</tr>
<tr>
<td>IA</td>
<td>proximal on major artery related but contralateral to the AVM</td>
</tr>
<tr>
<td>II</td>
<td>distal on superficial artery feeding the AVM</td>
</tr>
<tr>
<td>III</td>
<td>proximal or distal on deep artery feeding the AVM (“bizarre”)</td>
</tr>
<tr>
<td>IV</td>
<td>on artery unrelated to the AVM</td>
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*AVM = arteriovenous malformation.

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FIG. 1. Artist’s drawing of Type I to III aneurysms associated with arteriovenous malformations. For a description of the categories see Table 1.

FIG. 2. Left: Left carotid angiogram, lateral view, in a 60-year-old woman with intraparenchymal ventricular hemorrhage related to an arteriovenous malformation (AVM). A posterior temporal AVM (arrow-heads) and a proximal Type I large posterior communicating artery aneurysm (arrow) are evident. Right: Intraoperative photograph of the second operation following previous complete removal of the AVM showing partial thrombosis of the aneurysm sac with flow visible only through a small portion of the neck (arrow).
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TABLE 2
Outcome in the 39 treated patients in this series

<table>
<thead>
<tr>
<th>Outcome</th>
<th>No. of Cases</th>
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<tbody>
<tr>
<td>normal</td>
<td>24</td>
</tr>
<tr>
<td>mild neurological deficit, independent</td>
<td>12</td>
</tr>
<tr>
<td>moderate to marked neurological deficit, dependent on others</td>
<td>3</td>
</tr>
<tr>
<td>death</td>
<td>0</td>
</tr>
</tbody>
</table>

Type II or III aneurysms, which arose in the vicinity of, adjacent to, or even within the nidus of the AVM, making the etiology of the hemorrhage difficult to determine. The eight patients with Type III aneurysms had deep AVM's: four thalamic, one cingulate, one temporal, one sylvian, and one fronto-parietal. No other correlation between the location of the AVM's and the number or type of the aneurysms was established. No difference in age was found between groups with aneurysmal rupture as opposed to those with AVM-related hemorrhage.

Results

Our policy, when feasible, is to treat the symptomatic lesion first. All symptomatic lesions were treated. In 13 patients (33%), both lesions were treated at the same time. These included Type II aneurysms in five cases and Type III in seven cases (Fig. 4). All patients underwent surgery or endovascular procedures for at least one of the two types of lesions. Twenty-seven patients (69%) had both lesions totally eradicated by surgery, and all ruptured aneurysms were obliterated. Eleven patients (28%) underwent embolization of their malformation, either as the only form of treatment (four cases) or as an adjuvant for surgery (seven cases). Six patients had aneurysm obliteration as the result of embolization of their AVM's (Fig. 5). Fourteen patients (36%) had both types of lesions treated in staged operations, one operation for resection of the AVM and the other for clipping of the aneurysm.

Nine patients had no treatment for their AVM's. These decisions were related to our criteria for operability of AVM's and in most cases were based on the fact that the AVM was too large and strategically located to be resected safely. There were nine unruptured aneurysms that were not obliterated. Six of these aneurysms regressed with treatment of the AVM and three remained unchanged. Unfortunately, the expectation that hemodynamically related aneurysms will regress following removal of the associated AVM was not always realized; however, none of the residual aneurysms increased in size or ruptured during the follow-up periods after removal of the AVM.

There were no deaths in this series. Follow-up study lasted from 1 to 192 months (mean 55 months). Twelve patients had mild deficits; of these patients, seven displayed partial visual field cuts, two mild dysphasia, one transient Type III nerve palsy, one slight hemiparesis, and one mild memory problem and unilateral blindness. One patient had a major stroke related to angiography prior to the institution of treatment and has expressive aphasia and right hemiparesis, not worsened by surgery. One patient made a slow partial recovery from an acute postoperative hypotremic state causing prolonged coma from brain swelling; she is now ambulatory and able to care for herself but with marked cognitive deficits. One patient developed a hemorrhage in the AVM bed in the immediate postoperative period; as a result, she was in prolonged coma and now has seizures and a left arm paresis but is otherwise normal. Outcome is summarized in Table 2.

Fig. 3. Vertebral angiogram, anteroposterior view, in a 17-year-old girl with intraparenchymal ventricular hemorrhage and coma. A Type III "bizarre" aneurysm (arrow) is seen arising from a posterior choroidal branch feeding an arteriovenous malformation (AVM, arrowheads). The AVM and aneurysm were successfully resected during the same operation.

Fig. 4. Right carotid angiogram, lateral view, in a 21-year-old woman with left hemiparesis. An arteriovenous malformation (AVM, small arrowheads) is seen, with a Type III "bizarre" aneurysm (large arrowhead) arising from one of the thalamoperforators feeding the AVM. The AVM and associated aneurysm were successfully resected during the same operation.
Illustrative Case

The following case illustrates the complex problems presented by tandem lesions and the various treatment options that are available.

This 20-year-old man suffered a deep periventricular hemorrhage. Angiography showed a large dominant-hemisphere AVM (inoperable) and a small deep Type III aneurysm on a lenticulostriate artery adjacent to the apex of the AVM. Because of an intervening septicemia, we were unable to institute therapy. Follow-up angiography showed that the aneurysm (presumed to be the site of the hemorrhage) had markedly enlarged. Treatment was immediately undertaken in the form of embolization with occlusion of the artery containing the aneurysm. The AVM was subsequently reduced but not obliterated by staged embolization (Fig. 6). The follow-up period of 8 years was without incident, and the patient's neurological status is normal.

Discussion

We consider the sequencing of treatment for patients presenting with an aneurysm and an AVM to be of paramount importance. Two major considerations are the hemodynamic issues and the identification and priority given to treatment of the symptomatic lesion.

Hemodynamic Considerations

It has been hypothesized that aneurysms arise from a congenital defect of the artery wall.\(^1\) This theory is still accepted as valid for some aneurysms.\(^8,55\) Similarly, AVM's are thought to be congenital. Boyd-Wilson\(^8\) regarded the association as coincidental, except when aneurysms occurred on direct feeders to the AVM (hemodynamic stress might be a contributing factor). Paterson and McKissock\(^26\) linked the occurrence of aneurysms to increased blood flow in nutrient arteries to the AVM. This theory has gained extensive support from more recent series.\(^27,13,14,15,19,23,36\) Besides contributing to the development of aneurysms by flow-related factors, the presence of the AVM also causes enlargement of the feeding arteries and probable pathological changes in the arterial wall, which may represent important anatomical factors.\(^27,23,25,50\) The assumption is that the etiology for this association is multifactorial, involving one or more of the mechanisms discussed above.\(^5,34,38,53\)

The reported incidence of aneurysms in association with AVM's has varied significantly, from 2.7% to 17.8%.\(^7,11,19,23,24,40,49,51,55\) This discrepancy may be attributed to a sampling error in relation to the small size of most series and the different criteria used for distinguishing aneurysm from infundibular dilatation.\(^29,36\) Our series has an incidence approximating 10% of AVM cases, which is higher than that of aneurysms found at autopsy in an unslected patient population.\(^10,20\) All but one of the aneurysms have been located on major arterial trunks supplying the AVM. Perret and Nishioka\(^26\) described 34 patients harboring this association, of whom 57% had aneurysms that were hemodynamically related to the AVM. Okamoto, et al.\(^36\) studied the difference in the distribution of aneurysms alone or with AVM's. They showed that the presence of the AVM altered the normal distribution of aneurysms, a higher number occurring on direct feeders to the malformation. Nonetheless, for aneu-
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Fig. 6. Left carotid artery angiograms, anteroposterior view, in a 20-year-old man presenting with coma, intracerebral hemorrhage, and subsequent sepsis. A and B: Serial angiograms obtained on different days prior to the institution of treatment showing marked enlargement of a Type III “bizarre” aneurysm (arrowhead) on a lenticulostriate branch feeding a large parietal arteriovenous malformation. C: Postembolization angio-
gram demonstrating proximal obliteration of the lenticulostriate artery (arrow) and obliteration of the aneurysm.

Aneurysms arising in vessels proximal to the AVM or at several branching points away from the AVM, the incidence and distribution of aneurysms was similar to that of aneurysms alone. Our findings were similar, with the exception of a higher incidence of posterior circulation and Type III (“bizarre”) aneurysms.

Unpredictably, some aneurysms that are hemody-namically related to an AVM may decrease in size or disappear following obliteration of the AVM;16,25,33,45 conversely, the growth of aneurysms when both lesions are left untreated has been documented,1,4,23,52 as well as the case of aneurysmal rupture following resection of the AVM, raising the issue of perfusion overload factors.7 In our series, an incidence of multiple aneurysms higher than in the normal population and a concentration of Type I, II, and III (AVM flow-related) aneurysms support the role of the hemodynamic factors in their development. On the other hand, the fact that the Type III aneurysms were found in younger age groups, along with their unusual location (beyond the bifurcations of parent arteries), might indicate in some cases that congenital factors were involved.2,3,13,38,43,55

Our Type IV aneurysm was thought to be coincidental. There is ample information from intraoperative, post-
operative, and postembolization observations to suggest that an increase in intraluminal pressure,2,16,33,34,35 as well as dilatation of the feeding arteries,12,16,17,31,41,42,50,57 occurs following obliteration of the AVM. It is theoretically possible that these changes, which are abrupt but slow to normalize, could have an adverse effect on unruptured aneurysms proximal to the AVM, even causing aneurysmal rupture following the AVM resection or reduction by embolization. In our experience, this has not occurred and is of theoretical interest only.

After the elimination of the malformation, flow velocity is reduced and redirected to the high-resistance capillary bed, temporarily raising intraluminal arterial pressure which returns to normal as autoregulation reappears.30,32,33,54,55 This phenomenon has been offered as an explanation for the gradual reduction in size of some aneurysms following AVM removal.43 In some cases, it may be important in altering the natural history of these aneurysms, thereby justifying exclusion of the malformation alone when the aneurysm is asymptomatic. A more radical option is the obliteration of both lesions during the same operation.42,39,54 This strategy has been pursued in those cases of aneurysms located adjacent to the AVM. In our series, these aneurysms were all Type II or III. In a few of these cases, the embolic obliteration of both lesions can also be achieved.

Treatment Strategy

Although our policy has been to treat the sympto-
matic lesion first, an exception in our series was a patient presenting with an intracerebral hemorrhage from an AVM whose associated aneurysms were clipped first. This was an early case, and clipping was thought necessary to permit safe embolization of the AVM for which the catheters had to be passed through the lumen of the arteries harboring the aneurysms. Based on our experience since then, caution is unjusti-
fied and we have used embolization techniques via the arteries that contain untreated aneurysms without incident. Also to be considered is the morbidity and mortality from bleeding associated with aneurysmal rupture, which is significantly higher than that for an AVM. Furthermore, the risk of repeated hemorrhage from an AVM is significantly lower than that for aneurysms. This has been used as the strongest argument in recommending treatment of the aneurysm first, regardless of the circumstances. In our series, we found these considerations pertinent only when the cause of the hemorrhage is definite.

In identifying the cause of intracranial hemorrhage, computerized tomography scans are most helpful. Intraparenchymal clots are generally due to AVM's, whereas subarachnoid hemorrhage is more consistently associated with aneurysmal rupture. The five patients in our series who had hemorrhage of uncertain origin all had Type II (one case) or Type III (four cases) aneurysms. By virtue of the proximity of these aneurysms to the nidus of the malformation, which itself was deep, the hemorrhage resulting from the rupture of either of these lesions would be parenchymal. The close anatomical relationship to the AVM in these cases enabled both lesions to be treated simultaneously.

Conclusions

A five-tiered scheme of classification for the aneurysms was developed to consider hemodynamic relationships between the AVM's and the aneurysms and the implications for treatment. The criteria used for the treatment of the malformations in this group of patients were identical to those we apply to AVM's in general. These criteria prescribe that the symptomatic lesion, when determined, is treated first. Whenever it was possible to safely exclude both the aneurysm and the malformation during the same operation without unreasonably increasing the risks of the procedure, this option was undertaken.

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