

## Association of Cerebral Arteriovenous Malformation and Cerebral Aneurysm. Diagnosis and Management

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### Summary

Between 1979 and 1989, 7 patients were admitted, with cerebral arteriovenous malformations (AVM) and associated aneurysms (7% of the AVM patients and 2% of the aneurysm patients admitted during the same period). 6 of these patients were admitted because of an intracranial haemorrhage (in 3 of them the AVM, angiographically occult, was discovered at surgery). The last patient was referred for seizures.

Preoperatively it was supposed that the haemorrhage was related to the aneurysm in 3 cases, and to the AVM in 3 cases. But surgery allowed one to correct this supposition. Haemorrhage was due to AVM rupture in all 6 cases, and no aneurysm had ruptured. Overall three situations were demonstrated in this series: aneurysm and occult AVM (3 cases); AVM and independent aneurysm in the same area (2 cases); large AVM and aneurysm on a feeding artery (2 cases). All 6 patients admitted for haemorrhage were operated upon, at one operation in 5 of them. Both the malformations were excluded in these six patients. For the patient admitted for seizure, intra-vascular embolization of the AVM was performed, the aneurysm was not treated. The pathogenesis of the association AVM-aneurysm is discussed. In the authors' opinion, haemodynamic relationship should be considered in two cases (large AVM and aneurysm on a feeding vessel). For the other 5 cases, both the AVM and the coexisting aneurysm may be the end-result of a common congenital vascular malformation syndrome.

**Keywords:** Cerebral aneurysm; cerebral arteriovenous malformation; occult vascular malformation; association of aneurysm with AVM.

### Introduction

The association of intracranial aneurysm and arteriovenous malformation (AVM) has been thoroughly reported in the literature. According to Yaşargil<sup>18</sup>, this association has been initially described by Lowes in 1925 and later further documented by Stewart and Ashby in 1930 and 1931. The incidence of coexisting aneurysm and AVM in patients harbouring an AVM, ranges from 2.7% to 16.7% considering the different

authors (Table 1). This incidence, however, may be underestimated if small size aneurysms, overlooked by angiographic investigations, are taken into account as pointed out by Yaşargil<sup>18</sup>. The frequency of this pathological condition in aneurysm patients, although less frequently addressed in previous reports, was estimated at only 0.1%<sup>10–15</sup>. In this paper, we report our experience in seven patients, and discuss the different therapeutic modalities by a review of the pertinent literature.

### Clinical Material

Between 1979 and 1989, 105 patients harbouring an AVM – according to the classification of Mc Cormick<sup>9</sup> – were evaluated and treated in our department. During the same period, 349 cases of intracranial aneurysm were managed by the authors. In 7 of these patients, a coexisting AVM and aneurysm was demonstrated so that the incidence of the association was 7% out of the AVM patients, and 2% out of the aneurysm patients.

Four of these patients were female and 3 were male. Patients ranged in age from 33 to 52 years. All but one patient were admitted because of an intracranial haemorrhage – intracerebral haematoma in 5 instances and intraventricular haemorrhage in one case. One patient was referred to our department because of seizures. Patients data are summarized in Tables 2 and 3.

Among the patients admitted for bleeding, three patients presented with a temporal intracerebral haematoma (patients 1, 2, and 3). In all three instances, cerebral angiography could only reveal an aneurysm of the middle cerebral artery bifurcation. The misleading diagnosis of ruptured aneurysm was, therefore, made so that these patients were scheduled for clipping of their aneurysm. Exposure of an unruptured aneurysm led to a thorough exploration of the cavity left by the clot previously removed: the angiographically occult associated AVM, lying in the haematoma wall, could be finally discovered and resected. In patients 4 and 5, who were admitted for intracerebellar haematoma and intracerebral haematoma with intraventricular haemorrhage respectively, diagnosis of coexisting AVM and aneurysm was achieved by means of cerebral angiography. Preoperatively the origin of the haemorrhage could only be defined

Table 1. *Incidence of Coexisting Aneurysm and AVM*

	N	Ratio AVM/aneurysm	Haemorrhage incidence	Mortality
Suzuki <i>et al.</i>	9	6%	78%	0%
Perret and Nishioka	37	8%	78%	41%
Yaşargil	10	3%	100%	13%
Batjer <i>et al.</i>	22	14%	41%	23%
Okamoto <i>et al.</i>	5	3%	80%	0%
Personal series	7	7%	86%	0%

Table 2. *Patient Datas – Clinical Presentation and Preoperative Diagnosis*

Presentation	Preoperative diagnosis
1. V. S. 52 years intracerebral haematoma right temporal	aneurysm of right MCA
2. L. D. 33 years intracerebral haematoma left temporal	aneurysm of left MCA
3. C. G. 49 years intracerebral haematoma right temporal	aneurysm of right MCA
4. G. A. 39 years haematoma of left cerebellar hemisphere	aneurysm of left IAA AVM fed by left AICA, PICA, and SCA
5. D. A. 52 years intracerebral haematoma left frontal and IVH	aneurysm of left MCA AVM fed by left ACA
6. B. E. 39 years intraventricular and subarachnoid haemorrhage	aneurysm of basilar artery AVM fed by left PCA and MCA
7. R. B. 43 years seizures	aneurysm of basilar artery AVM fed by left MCA and PCA

ACA = anterior cerebral artery, AICA = antero-inferior cerebellar artery, IAA = internal auditory artery, MCA = middle cerebral artery, PCA = posterior cerebral artery, PICA = postero-inferior cerebellar artery, SCA = superior cerebellar artery, IVH = intra-ventricular haemorrhage.

Table 3. *Patient Datas – Management and Outcome*

Treatment	Outcome and follow-up
1. V. S. 52 years 1. clipping of unruptured aneurysm and resection of occult AVM	good 34 months
2. L. D. 33 years 1. clipping of unruptured aneurysm and resection of occult AVM	good 3 months
3. C. G. 49 years 1. clipping of unruptured aneurysm and resection of occult AVM	good 39 months
4. G. A. 39 years 1. multiple embolizations of AVM 2. clipping of aneurysm and AVM resection	good 34 months
5. D. A. 52 years 1. clipping of unruptured aneurysm and resection of AVM	fair 63 months
6. B. E. 39 years 1. clipping of unruptured aneurysm 2. embolization of AVM 3. resection of AVM	poor 36 months
7. R. B. 43 years 1. partial embolization of AVM	good 6 months

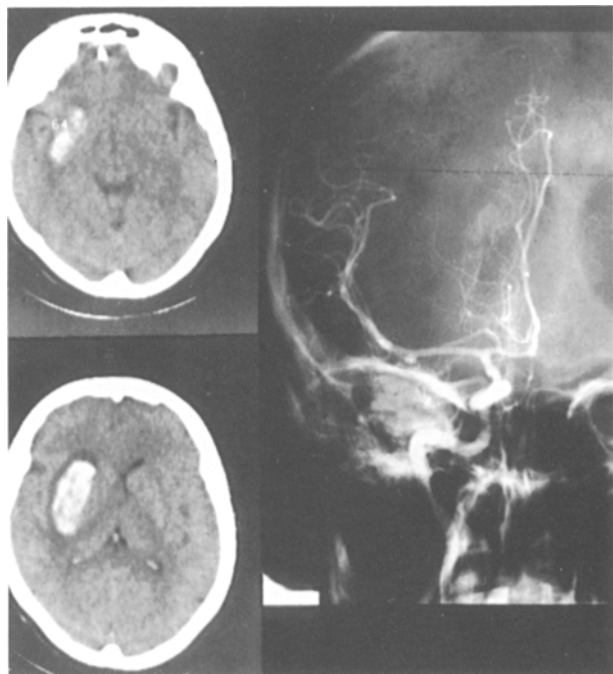


Fig. 1. Patient number 1. Upper and lower left = intracerebral haematoma of the right temporal lobe (CT scan without contrast). Right = right MCA aneurysm, directed inferiorly and anteriorly

clearly in patient 5, in whom a frontal intracerebral haematoma obviously originated from the AVM fed by the anterior cerebral artery. In patient 4, however, the associated lesions were so closely located that such a distinction was not possible. According to their relative vicinity, both the lesions were approached and treated in a one-stage procedure. In these two patients, the AVM appeared at surgery as the cause of the haemorrhage. Patient 6 was admitted because of intraventricular haemorrhage. Cerebral angiography disclosed a huge AVM fed by the middle and posterior cerebral arteries,

associated with an aneurysm of the basilar bifurcation. Clipping of the aneurysm was first carried out. Treatment of the AVM was secondarily addressed. Because of the large size and the deep localization of the lesion multiple embolizations of the AVM were performed, allowing later surgical resection of the malformation.

In the remaining patient – patient 7 – seizures were the presenting symptom. Cerebral angiography disclosed a huge temporal AVM fed by Sylvian vessels, associated with a basilar artery aneurysm. A first attempt of embolization of the malformation resulted in minor and completely reversible ischaemic complications, postponing further treatment.

#### *Illustrative Cases*

*Patient 1:* This 52-year-old female presented with sudden onset of headache, vomiting and hemiplegia on the left side. CT scan demonstrated a huge intracerebral haematoma of the right temporal lobe (Fig. 1). Cerebral angiography disclosed an aneurysm of the right MCA directed inferiorly and anteriorly. Surgery denied the previous diagnosis of ruptured aneurysm and revealed a minute AVM which lay on the wall of the haematoma and was extirpated. Final outcome was good and postoperative angiography satisfactory.

*Patient 4:* This 39-year-old male was referred for an aneurysm of the left internal auditory artery associated with an AVM of the left cerebellar hemisphere responsible for a cerebellar haematoma (Fig. 2). Cerebral angiography demonstrated the feeders of the AVM, mainly the left AICA, SCA, and PICA. After selective catheterization of the AICA and preoperative embolization of the AVM, clipping of the aneurysm and resection of the AVM, were carried out during the same procedure. The AVM was found responsible for the haemorrhage. The patient had a good recovery and postoperative angiography confirmed the obliteration of both the malformations.

*Patient 6:* Intraventricular haemorrhage, as demonstrated by the CT scan, revealed the malformative association in this 39-year-old female. Cerebral angiography depicted a huge AVM fed by the MCA and the PCA as well as an aneurysm located on the basilar bifurcation (Fig. 3 A). At the first step, attempt at balloonization of the aneurysm was made, with no success and clipping of the aneurysm was carried out. Five months later, 3 embolizations of the AVM were performed (Fig. 3 B) authorizing final and complete extirpation of the AVM. The patient however, remained severely disabled.

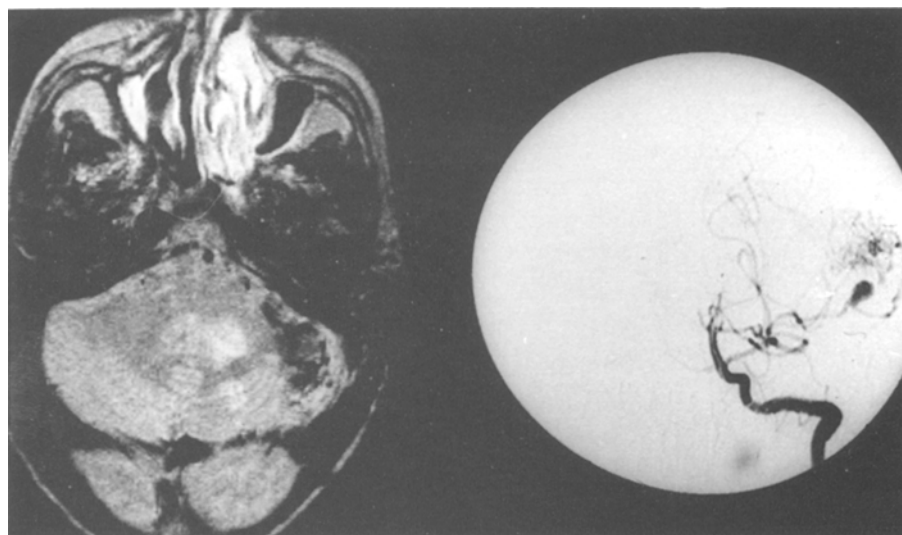


Fig. 2. Patient number 4. On the left, AVM of the left cerebellar hemisphere (MRI). On the right, aneurysm of the left internal auditory artery

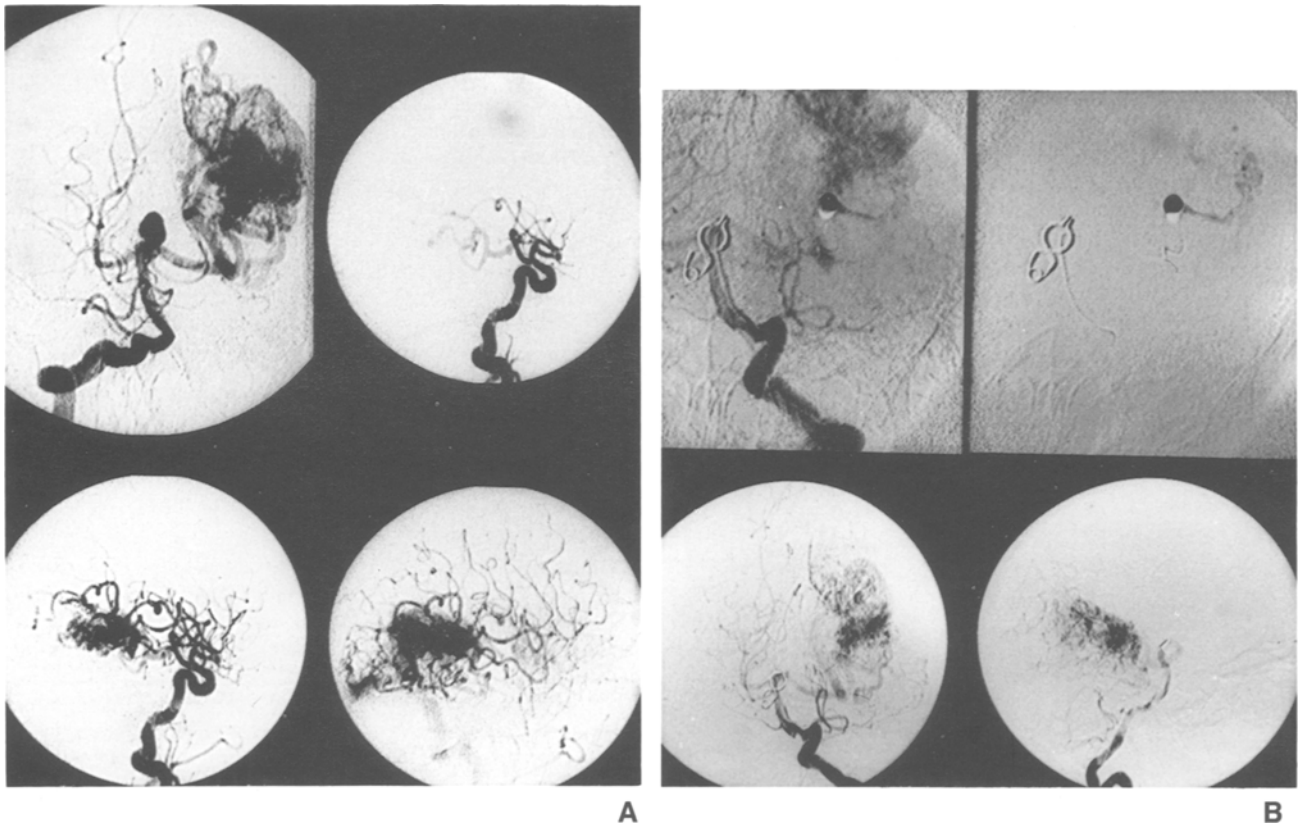


Fig. 3. Patient number 6. A) Left temporo occipital AVM and aneurysm of the basilar bifurcation. B) After clipping of the aneurysm, the AVM is partially embolized through femoral catheterization. See the catheter going through the fenestrated clip into the left posterior cerebral artery

## Results

All the patients admitted for bleeding were operated on and radical cure of both the malformations could be achieved in each case. In all the patients but one (patient 6), this could be performed in a one-stage procedure. Patients were available for follow-up from 4 to 63 months. There was no fatality in this series. All the patients improved after treatment except patient 6 who was severely disabled by tremendous bleeding from the AVM during surgery, despite the preoperative embolization. The follow-up of patient 7, at 6 months is still uneventful.

## Discussion

The pathogenesis of the association AVM-aneurysm has drawn increasing attention throughout the literature. To date, three different theories have been proposed to explain the coexistence of intracranial aneurysms with AVM<sup>10</sup>: increased blood flow induced by the AVM, casual occurrence of aneurysms in AVM patients and common congenital malformation syndrome.

The first theory, proposed by Patterson and Mc Kissock<sup>12</sup>, suggests that aneurysms may develop from malformative vessels, as a consequence of an increased blood flow within the feeding vessels. The localization of some aneurysms, located on a feeding vessel of a huge AVM, may support this theory, more especially as aneurysms associated with an AVM often arise from arteries usually spared by aneurysmal processes—*i.e.* lenticulo-striate arteries, terminal branches of pericallosal artery, anterior choroidal artery and cerebellar arteries<sup>12</sup>. A haemodynamic relationship between the increased blood flow and the development of aneurysms is further supported by Shenkin<sup>14</sup> reporting 3 cases of carotid aneurysm formation, following carotid ligation on the opposite side. In these patients, it may be assumed that the development of an aneurysm on a previously normal carotid vessel is likely as a consequence of an abrupt increase in the blood flow. The haemodynamic changes in the AVM vessels may induce a weakening of the arterial wall and therefore reveal some asymptomatic arterial defect, resulting in the development of an aneurysm.

Though a haemodynamic factor should be considered in the pathogenesis of aneurysms arising from AVM feeding vessels, aneurysms developed on arteries haemodynamically independent from the AVM, can not be so easily explained by the former theory. In the co-operative study, as reported by Nishioka and Perret<sup>13</sup>, only 37% of the aneurysms associated with an AVM were located distally on a feeding vessel of the AVM, 21% proximally on a feeding artery while 42% were found to be unrelated to the AVM. In our series, in two patients only (patient 6 and 7), a haemodynamic correlation could be demonstrated between the AVM and the artery supporting the aneurysm. In our opinion, these observations draw further interest on alternative aetiological factors.

In 1959, Boyd-Wilson<sup>3</sup> suggested that aneurysms may occur in a random fashion in the whole population as well as AVM patients, and therefore that their association could be fortuitous. However, the incidence of aneurysms in post-mortem studies is significantly lower than that reported in AVM patients (Table 1), so that this theory appears rather unlikely<sup>18</sup>.

The third hypothesis, developed by Arieti and Gray<sup>1</sup>, supports that both the AVM and the co-existing aneurysm may be the end-result of a common congenital vascular malformation syndrome, occurring during the angiogenesis. This theory is more likely to be able to explain the association of two malformations haemodynamically independent. In 3 of our patients, an aneurysm of the bifurcation of the MCA was associated with an occult deep Sylvian AVM. Since occult AVM's are not usually considered as high-flow malformations, a common congenital abnormality is the most likely mechanism of formation of such a pathological condition. To the best of our knowledge, this association – *i.e.* occult AVM and aneurysm – has never been reported before. Our experience with these 3 cases, however, proves that such an entity does exist and should be looked for. In each instance, the haemorrhage was initially related to aneurysmal bleeding, which was denied by the operative finding of an unruptured aneurysm. Exploration of the walls of the cavity, left by evacuation of the haematoma, confirmed the diagnosis by discovering the AVM. Diagnosis, therefore, often appears as a challenge, since the AVM may be overlooked by radiological studies. However, critical review of the radiological data may draw attention on some anatomical discrepancies between the haemorrhage and the so-called “ruptured” aneurysm. In particular, on the CT scan the absence of blood in the Sylvian cistern as well as the deep localization of

the haematoma, remote from the aneurysm fundus, may suggest an alternative cause for the haemorrhage. Furthermore, the direction of the fundus of the aneurysm pointing anteriorly and inferiorly as depicted in patient 1, should reinforce the likelihood of an “occult malformation”. The diagnostic value of the MRI, though well reported for the evaluation of larger AVM's, is still to be defined in this particular instance.

Clinically, the association of aneurysm and AVM is characterized by the high incidence of haemorrhage (Table 1). In 6 of our patients (86%) as well as in all the patients reported by Yaşargil<sup>18</sup>, an intracranial bleeding was the presentation. This frequency is significantly higher than that reported by the Cooperative Study<sup>13</sup> – *i.e.* 68% – for isolated AVM's. More difficult, however, is to determine the respective responsibility of the associated lesions as the origin of the bleeding. In this series, the AVM was the cause of the haemorrhage in all these instances. This clinical prominence of the AVM was also reported by Suzuki<sup>15</sup> for posterior fossa lesions. Conversely, in the 22 patients reported by Batjer<sup>2</sup>, the haemorrhage could be related to the AVM in only 22%. According to Wilkins<sup>16</sup>, AVM's appeared as the cause of the bleeding in a third of the patients, while simultaneous rupture of both the associated lesions could be demonstrated in another third. Similar observations were made by Okamoto<sup>11</sup>.

In his review of 50 AVM patients, Wilkins<sup>16</sup> found a bleeding incidence of 18% during the first year decreasing to 3% after 5 years and then to 2% after 10 years responsible for a mortality as high as 14% for the first haemorrhage and 21% for rebleeding. Since the risk of bleeding is even higher in AVM's associated with aneurysm, as noted above, surgical treatment should be carried out. This assumption is supported by the poor results yielded by conservative management as demonstrated by Nishioka and Perret<sup>13</sup> and later, by Gamache<sup>5</sup> in their series. To date, however, there is no clear definition of what should be the surgical attitude<sup>10</sup>. In most of the instances, surgery is directed towards the malformation responsible for the haemorrhage. Extirpation of the AVM only or clipping of the aneurysm only may thereafter lead to an increased risk of bleeding from the remaining lesion. As observed by Batjer<sup>2</sup> in one of his patients, the resection of the AVM alone may lead to a sudden increase in vascular resistance and therefore produce a distention of the aneurysm fundus. Similar experiences were reported by Drake [personal communication, may 1984] and Luesenhop<sup>8</sup>.

Conversely, Shenkin<sup>14</sup> and later Yaşargil<sup>18</sup> and Hi-

gashi<sup>6</sup> have noted cases of aneurysmal occlusion following extirpation of the associated AVM. These authors suggested that the decrease in blood flow may lead to thrombosis of the aneurysm. However, the high risk of rupture and bleeding from an aneurysm precludes as a rule conservative management of this type of lesion, even if asymptomatic.

Whenever the aneurysm appears as the cause of the haemorrhage, clipping represents undoubtedly the first surgical step. Conversely, the treatment of the remaining AVM is often delayed or even neglected. However, Fults and Kelly<sup>4</sup> and more recently Wilkins<sup>16</sup>, reviewing cases of AVM, found that the risk of rebleeding from an AVM was higher than previously reported by the Cooperative Study. Our study further demonstrates an increased tendency to bleed whenever the AVM is associated with an aneurysm, so that, in our opinion, surgery should be directed towards both lesions, provided that such a procedure is technically feasible.

Practically and according to the size of the lesions and their respective participation in the clinical manifestations, three distinct situations may be considered. The first situation is that of a large AVM associated with an aneurysm located on a feeding vessel of the malformation. This condition is a difficult one. Ideally, radical cure of both the malformations should be attempted. However, the hazards of radical treatment of large or huge AVM's are well known. Clipping (or ballooning) of the aneurysm, as proposed by Batjer<sup>2</sup>, should initiate the procedure, whether because of previous rupture of the aneurysm or in order to prevent abrupt changes in the tension applied over the aneurysmal wall. Besides, whenever embolization of the AVM is considered, initial clipping of the aneurysm – or its endovascular occlusion – seems mandatory for the same reasons.

The second situation is represented by an aneurysm and an AVM located in territories haemodynamically independent, but not remote one from the other. Surgical treatment, in this instance, should focus on the lesion responsible for the haemorrhage, but the management of the remaining lesion should be performed during the same procedure, when feasible.

The third situation is that of an occult AVM associated with an aneurysm. The main issue is a thorough and careful exploration of the haematoma after clipping of the aneurysm recognized as unruptured, in order to disclose the responsible lesion. This emphasizes the importance of correct, although difficult, preoperative diagnosis, affording appropriate planning of the surgical approach. Considering the high risk of bleed-

ing associated with this type of AVM, as demonstrated by Lobato<sup>7</sup>, disregarding the AVM, exposes the patient to deleterious complications.

Independently from clinical or morphological specificities, the association of an AVM-with an intracranial aneurysm should be regarded as a pathological entity characterized by a severe morbidity. Radical cure of both the malformations by means of surgery represents the safest warrant of the outcome, otherwise often compromised. Moreover, the results obtained by Suzuki<sup>15</sup> as well as those presented in this study, suggest that a one-stage procedure makes for an optimal neurological outcome. Finally and despite its relative rarity, the association of an occult AVM and an aneurysm justifies in our opinion a critical and careful reading of radiological studies whenever even discrete discrepancies are noted between the bleeding and the so-called ruptured aneurysm. Since such a review failed to completely clear the doubt, the surgical approach should include the discovery of the responsible underlying lesion.

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