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ORIGINAL ARTICLE

The "focus on aneurysm" principle: Classification and surgical principles of management of concurrent arterial aneurysm with arteriovenous malformation causing intracranial hemorrhage

Vikas Jha, Sanjay Behari, Awadhesh K. Jaiswal, Kamlesh Singh Bhaisora, Yogesh P. Shende, Rajendra V. Phadke[1]

Departments of Neurosurgery and [1]Radiology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

ABSTRACT

Context: Concurrent arterial aneurysms (AAs) occurring in 2.7-16.7% patients harboring an arteriovenous malformation (AVM) aggravate the risk of intracranial hemorrhage.

Aim: We evaluate the variations of aneurysms simultaneously coexisting with AVMs. A classification-based management strategy and an abbreviated nomenclature that describes their radiological features is also proposed.

Setting: Tertiary care academic institute.

Statistics: Test of significance applied to determine the factors causing rebleeding in the groups of patients with concurrent AVM and aneurysm and those with only AVMs.

Subjects and Methods: Sixteen patients (5 with subarachnoid hemorrhage and 11 with intracerebral/intraventricular hemorrhage; 10 with low flow [LF] and 6 with high flow [HF] AVMs) underwent radiological assessment of Spetzler Martin (SM) grading and flow status of AA + AVM. Their modified Rankin's score (mRS) at admission was compared with their follow-up (F/U) score.

Results: Pre-operative mRS was 0 in 5, 2 in 6, 3 in 1, 4 in 3 and 5 in 1; and, SM grade I in 5, II in 3, III in 3, IV in 4 and V in 1 patients, respectively. AA associated AVMs were classified as: (I) Flow-related proximal (n = 2); (II) flow-related distal (n = 3); (III) intranidal (n = 5); (IV) extra-intranidal (n = 2); (V) remote major ipsilateral (n = 1); (VI) remote major contralateral (n = 1); (VII) deep perforator related (n = 1); (VIII) superficial (n = 1); and (IX) distal (n = 0). Their treatment strategy included: Flow related AA, SM I-III LF AVM: aneurysm clipping with AVM excision; nidal-extranidal AA, SM I-III LF AVM: Excision or embolization of both AA + AVM; nidal-extranidal and perforator-related AA, SM IV-V HF AVM: Only endovascular embolization or radiosurgery. Surgical decision-making for remote AA took into account their ipsilateral/contralateral filling status and vessel dominance; and, for AA associated with SM III HF AVM, it varied in each patient based on diffuseness of AVM nidus, flow across arteriovenous fistula and eloquence of cortex. Follow up (F/U) (23.29 months; range: 1.5-69 months) mRS scores were 0 in 12, 2 in 2, 3 in 1 and 6 in 1 patients, respectively.

Conclusions: Patients with intracranial AVMs should be screened for concurrent AAs. Further grading, management protocols and prognostication should particularly "focus on the aneurysm."

Key words: Aneurysm, arteriovenous malformation, classification, intracerebral hematoma, subarachnoid hemorrhage, surgery

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Address for correspondence:
Dr. Sanjay Behari, Department of Neurosurgery,
Sanjay Gandhi Postgraduate Institute of Medical Sciences,
Lucknow - 226 014, Uttar Pradesh, India.
E-mail: sbehari27@yahoo.com

Introduction

Hemorrhage is a serious complication in approximately 50% of patients having an intracranial arteriovenous malformation (AVM).[1,2] Concurrent arterial aneurysms (AAs), occurring in nearly 2.7-16.7% patients harboring an AVM, significantly increase the risk of hemorrhage.[3] In this situation, a non-invasive diagnostic evaluation utilizing either a computed tomography (CT) or magnetic resonance (MR) angiography often fails to pinpoint the precise source of the bleeding. No uniform classification system exists that qualifies the features of the aneurysm (such as size, direction of its neck or fundus, the proximity of the two entities and the dominance of parent vessel) and correlates it with the characteristics of the parent malformation. The association of aneurysms with AVMs also significantly influences treatment decisions and management protocols.

This study attempts to evaluate the variations of AAs coexisting with AVMs in patients who presented with either subarachnoid or intracerebral hemorrhage. It also attempts to formulate a treatment strategy for concurrent aneurysms with AVMs that incorporates both their individual management considerations as well as their relationship to one another and with the parent vessels.

Subjects and Methods

Inclusion criteria

Sixteen patients presenting with subarachnoid hemorrhage (SAH), intracerebral hematoma or both and harboring a concurrent intracranial AA with AVM who underwent either surgery or an interventional procedure between January 2006 and December 2011 were included in this study. Their clinical presentation and timing from the onset of ictus/first symptom were noted. The angioarchitecture of the aneurysm as well as the AVMs and their supporting arteries and the draining veins were assessed utilizing CT/MR angiogram as well as digital subtraction angiogram (DSA).

Classification of aneurysms

The aneurysms were classified into 9 categories depending upon their location relative to the coexisting AVM. This was based upon a modification of the classifications suggested by Redecop *et al*. and Cunha e Sa *et al*. [Table 1].[4,5] *Flow related proximal aneurysms* were located on the proximal arteries that were specifically supplying the AVM and included the supraclinoid internal carotid artery (ICA) and the circle of Willis, the middle cerebral artery (MCA) until and including the primary bifurcation, the A1 segment of anterior cerebral artery (ACA) and the anterior communicating artery (A Comm A), or the vertebrobasilar trunk [Figures 1 and 2]. *Flow related distal aneurysms* were located on the arteries supplying the AVM and beyond the location of these major vessels but with a significant distance from the actual nidus of the AVM. *Intranidal aneurysms* were located within the AVM nidus and could be distinguished from arterial ectasias and venous aneurysms by their early filling during the arterial phase of the angiogram [Figure 3]. *Intra- and extra nidal aneurysms* included the simultaneous existence of both intranidal aneurysm and aneurysm in close proximity to the nidus of the AVMs [Figure 4]. The latter arose from arterial branches that formed one of the peripheral vessels of the conglomeration of the AVM nidus and could be easily distinguished from it on early arterial phase of angiography. In contrast to the flow related distal aneurysms that have the possibility of being at a considerable distance from the AVM, the intra and extranidal aneurysms were situated in and around the main nidus, respectively and were always surgically accessible along with the AVM nidus. *Remote major ipsilateral aneurysms* were situated on a major artery of circle of Willis unrelated to the AVM but were getting filled on ICA/vertebral injection ipsilateral to the side of AVM [Figures 5 and 6]. *Remote major contralateral aneurysms* were also situated on a major artery of the circle of Willis unrelated to the AVM but were getting filled on ICA/vertebral injection contralateral to the

Table 1: Classification of arterial aneurysms associated with AVMs

Arterial aneurysms associated with AVMs	
A.	Arterial aneurysms associated with AVMs
I:	Flow related proximal: Proximally situated on the major artery of circle of Willis that supplied the AVM
II:	Flow related distal: Situated on distal branches of circle of Willis that supplied the AVM
III:	Intranidal: Situated within nidus of the AVM filling early during angiography
IV:	Both extra and intranidal: Two or more aneurysms, one intranidal and the other (s) situated on arterial branches that formed one of the peripheral vessels of the conglomeration of the AVM nidus and is located in close relation to the AVM
V:	Remote major ipsilateral: Situated on a major artery of circle of Willis unrelated to AVM but ipsilateral to the side of AVM
VI:	Remote major contralateral: Situated on a major artery of circle of Willis unrelated to AVM but contralateral to side of AVM
VII:	Deep perforator related: Situated on deep perforator vessels proximal and ipsilateral to side of AVM
VIII:	Superficial: Situated on superficial cortical artery supplying the AVM
IX:	Distal: Situated on the major feeding artery distal to the location of the AVM
B.	Spetzler Martin grade of AVM I, II, III, IV, V
C.	AVM flow I: High flow II: Low flow
D.	Hemorrhage (designated by*) I: None II: From aneurysm III: From AVM IV: Not distinguishable from either aneurysm or AVM V: From remote source

Spetzler and Martin, 1986.[6] AVM – Arteriovenous malformation

side from where the vessels that were supplying the AVM were filling [Figure 7]. *Deep perforator related aneurysms* were situated on deep perforator vessels proximal and ipsilateral to the side of AVM [Figure 8]. *Superficial aneurysms* were aneurysms arising from one of the abnormal unnamed arterial branches supplying the AVM and traversing superficially on the cortical surface. Finally, *distal aneurysms* were situated on the feeding artery beyond the location of the AVM.

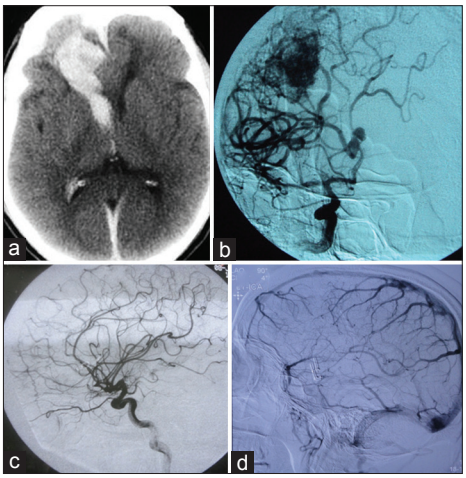

Figure 1: Patient 1 - (a) Axial images of plain CT scan showing hemorrhage in the gyrus rectus, basifrontal region and frontal horn of lateral ventricle, (b) A right internal carotid artery (ICA) digital subtraction angiogram (DSA) showing the posterior frontal arteriovenous malformation (AVM) (Spetzler Martin grade 3; size = 2, venous drainage = 1, eloquence = 0) supplied by anterior and middle cerebral artery with a low flow through the nidus. The aneurysm had ruptured causing the intracerebral hematoma, (c) Follow-up arterial, (d) Venous phase of right ICA DSA lateral view showing the clipped aneurysm with excised AVM

Classification of AVMs

The AVMs were assigned Spetzler-Martin (SM) grade (by evaluating their size, number of draining veins and eloquence of the part of the brain they were situated in).[6] The classification of the AVMs (based upon the degree of arteriovenous shunt present) into a high flow (HF) and a low flow (LF) state was determined based upon the length of time required for venous filling to occur after arterial filling according to the criteria suggested by Brown *et al*.[7] Thus, HF AVMs had an almost simultaneous filling of their arteriovenous compartment; in the LF AVMs however, there was a considerable delay (more than 2 s) in the time to venous filling after the arterial filling was complete.

An abbreviated nomenclature for concurrent aneurysms with AVMs presenting with intracranial hemorrhage

The CT/MR imaging was studied to determine whether the aneurysm or the AVM had undergone hemorrhage. Following a detailed assessment of their radiological imaging and the four parameters [anatomical relation of aneurysm with AVM,

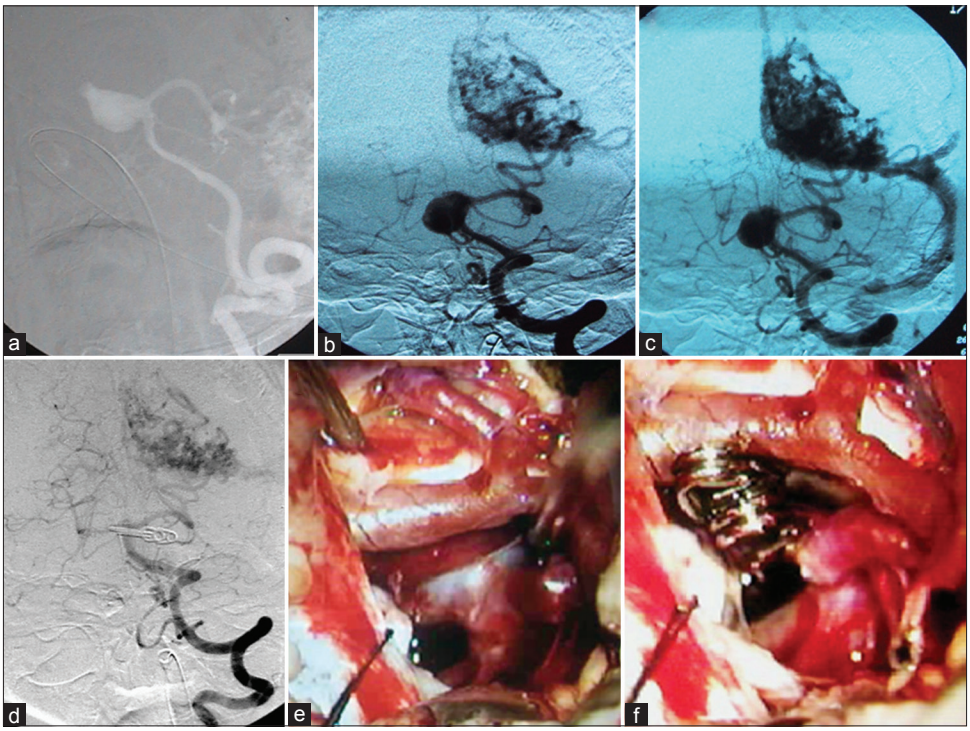

Figure 2: Patient 2 - (a) Lateral, (b and c) Oblique views of vertebral digital subtraction angiogram (DSA) showing a giant basilar artery aneurysm high above dorsum sellae. The occipital high flow arteriovenous malformation (Spetzler Martin grade IV; size = 2, venous drainage 1; eloquence = 1) is supplied by left posterior cerebral artery and superior cerebellar artery and drains by multiple feeders into the superior sagittal sinus and left transverse sinus, (d) Postoperative right vertebral DSA showing adequate clipping of basilar apex aneurysm, (e) Operative image showing the aneurysm and the pituitary stalk draped around it, (f) Successful clipping of the aneurysm

SM grading, high/low flow status of the AVM and the origin of subarachnoid/intracerebral hemorrhage being from either aneurysm or AVM; Table 1], these patients were assigned an abbreviated nomenclature that helped to determine their therapeutic intervention. The aneurysm (an) associated with the AVM was assigned a numerical value based upon its category given in Table 1. The AVM was also assigned

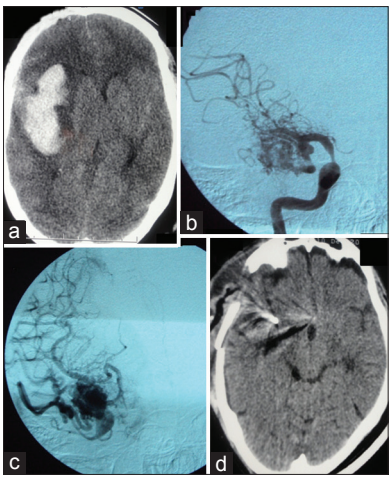

Figure 3: Patient 4 - (a) Plain computed tomography (CT) axial images showing frontotemporal perisylvian bleeding with midline shift, (b) AP view of right internal carotid artery digital subtraction angiogram showing the right temporal and perisylvian high flow arteriovenous malformation (AVM) (Spetzler Martin grade 3; size 2, venous drainage = 0, eloquence = 1) supplied by M2 and M3 branches of right middle cerebral artery with an intranidal AA, (c) The venous drainage into superficial cortical vein, (d) Postoperative contrast axial CT scan showing the aneurysmal clip artefact and total AVM excision

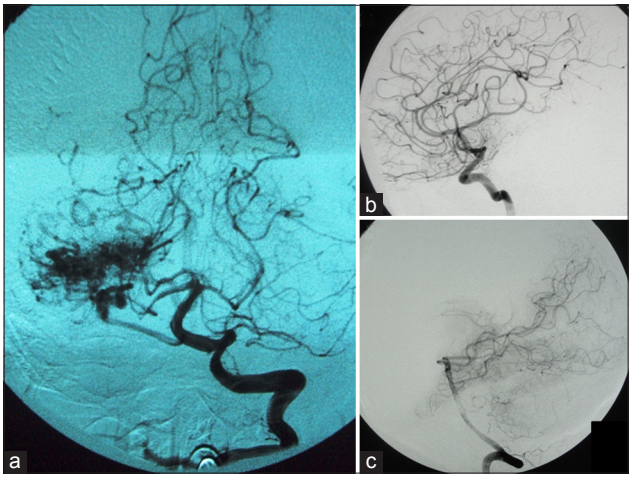

Figure 4: Patient 5 - (a) Right vertebral digital subtraction angiogram (DSA) AP view showing intra and extranidal aneurysms arising from arteriovenous malformation (AVM) supplied by right anterior inferior cerebellar artery, (b) Postoperative lateral internal carotid artery, and (c) Vertebral DSA showing total excision of AVM and aneurysm excision

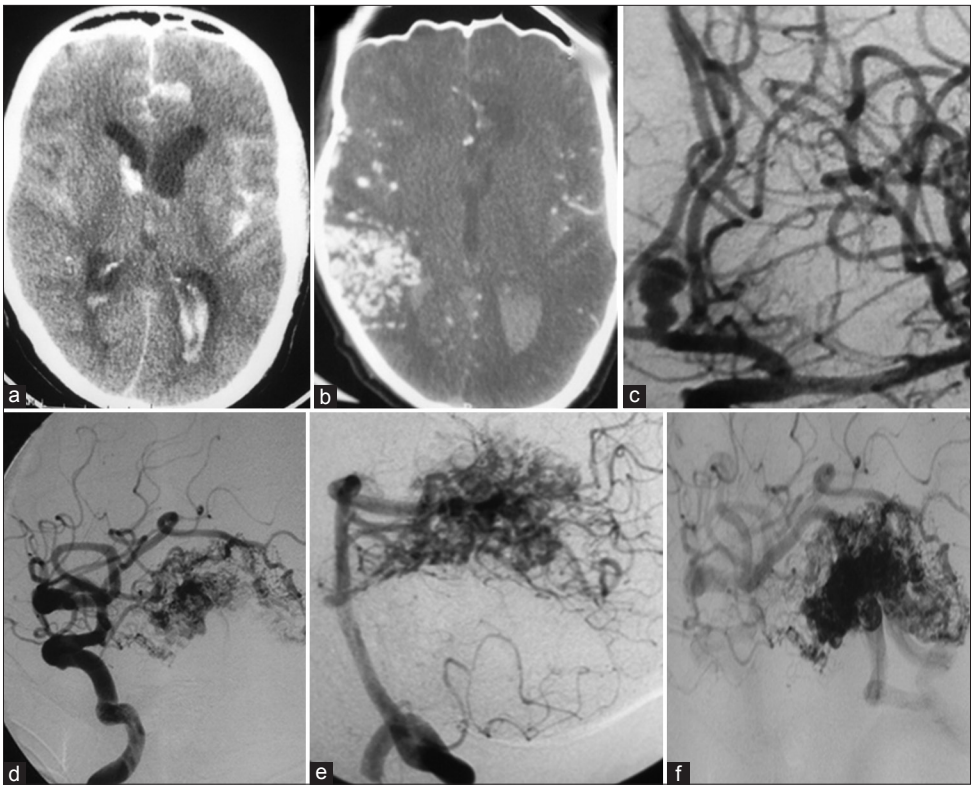

Figure 5: Patient 6 - (a) Axial contrast enhanced computed tomography scan showing blood in the interhemispheric fissure, bilateral sylvian cisterns and lateral ventricles, (b) Serpigenous contrast enhancing hyperdensity indicative of arteriovenous malformation (AVM) in the right posterior temporal region, (c) AP view of internal carotid artery (ICA) digital subtraction angiogram (DSA) showing anterior communicating artery aneurysm, (d) Lateral view of ICA DSA showing the posterior temporal AVM supplied by right middle cerebral artery, (e) Lateral view of right vertebral DSA showing AVM supply also from right posterior cerebral artery, (f) Venous drainage into the right transverse sinus

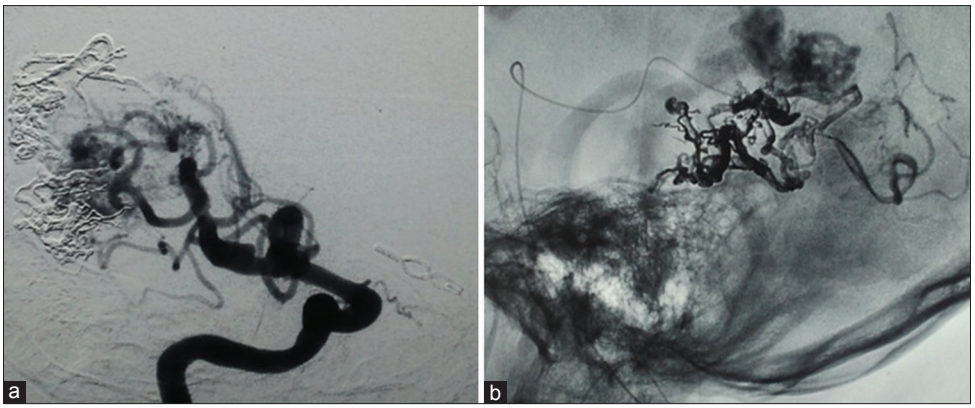

Figure 6: Patient 6 - (a) Right internal carotid artery digital subtraction angiogram (DSA) AP view showing partial embolization of arteriovenous malformation (AVM). The clip applied on anterior communicating artery aneurysm is also seen, (b) Right vertebral DSA lateral view showing partial embolization of AVM being filled from the posterior cerebral artery

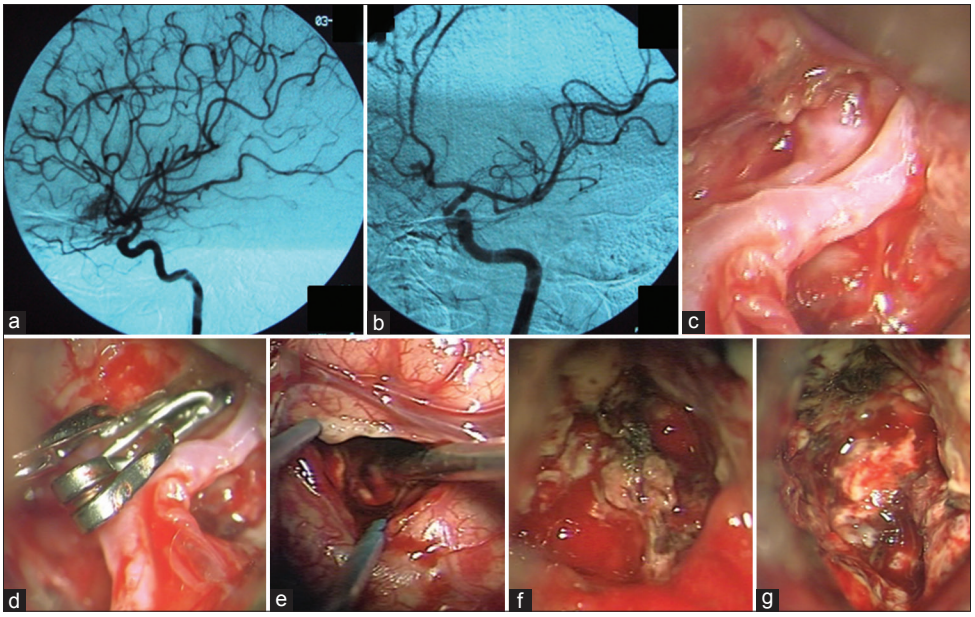

Figure 7: Patient 7 - (a) Right internal carotid artery (ICA) digital subtraction angiogram (DSA) lateral view showing the small arteriovenous malformation (AVM) in the right frontal region, (b) Left ICA DSA AP view showing the filling of the anterior communicating artery (A Comm A) aneurysm, (c) operative image showing the A comm A aneurysm after removal of small part of gyrus rectus, (d) Clip applied to neck of the aneurysm, (e) Evacuation of hematoma surrounding the AVM from left frontal cortex, (f) Coagulation of the AVM, (g) Its excision

a numerical value based upon the SM grade and, the "HF" or "LF" status based upon the prevalent high or low flow arteriovenous shunting, respectively, in them. The asterix designated whether the aneurysm or the AVM had been responsible for the intracranial hemorrhage. If the hemorrhage was not distinguishable as being from either the aneurysm or the AVM (especially in cases of nidal aneurysms), the asterix was assigned to both the aneurysm and the AVM category. If the hemorrhage was remote from the primary site of both the aneurysm or the AVM, the asterix was omitted. However, none of our patients had bleeding away from the site of the AVM and the aneurysm.

Therapeutic interventions

In this series, only patients associated with the simultaneous presence of AVM and aneurysm who presented with SAH/intracerebral hematoma were included. Evacuation of the clinically significant hematoma to relieve raised intracranial pressure, surgical clipping of the aneurysm (or occasionally, excision of the intra-nidal or extra-intra nidal aneurysm with the AVM), excision of the AVM, direct or staged embolization of the AVM and stereotactic radiosurgery were the therapeutic options available. The patients were assigned a modified Rankin's score (mRS) [Table 2][8,9] and a Fisher's score (based upon the CT/MR image in the patients presenting with SAH) at admission and compared with the score at the last follow-up.

Results

Demographic profile

The sixteen patients having the simultaneous presence of an intracranial aneurysm and AVM represented 2.56% of patients

who had undergone surgical clipping for isolated intracranial aneurysms (n = 623); or 17.5% (n = 91) of patients who had undergone either surgical excision (n = 72) or therapeutic embolization (n = 29) for their isolated AVMs during the same time frame. Out of the later, 75 patients with an isolated AVMs had undergone an intracranial (subarachnoid or lobar) hemorrhage.

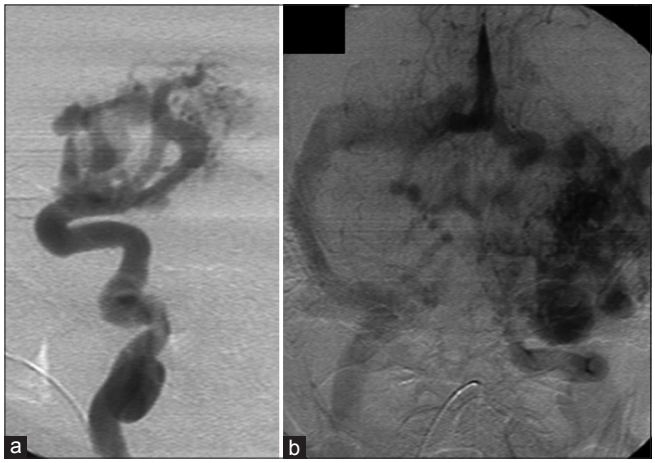

Figure 8: Patient 8 - (a) Right internal carotid artery (ICA) digital subtraction angiogram showing multiple aneurysms within the deep-seated arteriovenous malformation nidus supplied by perforators of ICA and middle cerebral artery, (b) Venous phase showing multiple draining veins into transverse and sigmoid sinus and, internal cerebral vein and straight sinus

Clinical spectrum

The mean age of presentation in the series was 38.31 years (age range 12-60 years). The male:female ratio was 10:6. Their

Table 2: Modified Rankin's score

Score	
0:	No symptoms at all
1:	No significant disability despite symptoms; able to carry out all usual duties and activities
2:	Slight disability; unable to carry out all previous activities, but able to look after own affairs without assistance
3:	Moderate disability; requiring some help, but able to walk without assistance
4:	Moderately severe disability; unable to walk without assistance and unable to attend to own bodily needs without assistance
5:	Severe disability; bedridden, incontinent and requiring constant nursing care and attention
6:	Dead

Rankin, 1957[9]

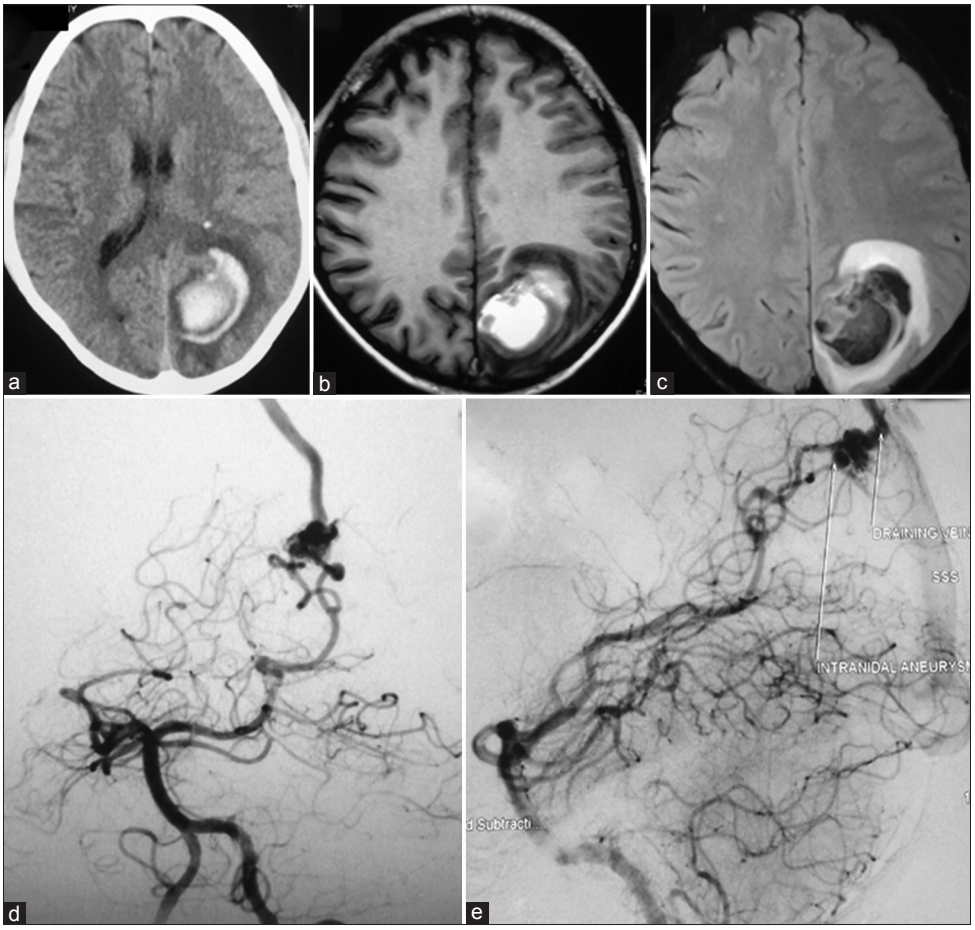

Figure 9: Patient 11 - (a) Axial computed tomography scan, (b and c) Axial magnetic resonance scan showing the left posterior parietal intracerebral hematoma, (d) AP view of left vertebral angiogram showing the arteriovenous malformation with intranidal aneurysm in the left parietal region, (e) Draining into superior sagittal sinus

clinical presentation is summarized in Table 3. All of them presented with a sudden onset of symptoms (sudden headache with recurrent vomiting in 15 occasionally with neck stiffness or grand mal seizures and , sudden onset hemianopia in 1 patient) heralding the emergent development of either an intracerebral/intraventricular hematoma ($n = 10$) or SAH ($n = 5$). Transient loss of consciousness, alteration of sensorium, hemiparesis, ataxia and hemianopia were the associated clinical manifestations. Their mRS at admission was 0 in 5, 2 in 6, 3 in 1, 4 in 3 and 5 in 1 patients, respectively.[8,9] The mean duration of presentation was 199.43 days but the range was extremely variable (2-190 days). Noteworthy was the fact that 9 out of the 16 patients presented for DSA and definitive treatment for their underlying aneurysm and AVM after a delay of 2 weeks or more following their initial ictus. One of them (patient 10) presented nearly 6 years after the first symptom appeared.

Radiological features

Five patients presented with SAH. The location of the bleed included interhemispheric fissure with gyrus rectus ($n = 2$), sylvian fissure, cistern magna or interpeduncular and suprasellar cistern ($n = 1$ each, respectively). Three of these 5 patients having SAH were in Fisher's grade IV with intraventricular extension of the hemorrhage and one each were in grade II and III, respectively. Eleven other patients presented with intracerebral hematoma that usually included surfacing hematomas in the frontal, temporo-occipital or posterior parietal regions and the infratentorial supracerebellar surface. Two of these patients, however, presented with deep seated hematomas in the basal ganglionic region and the caudate nucleus-lateral ventricular location; and, two others with a primary intraventricular hematoma [Table 4]. In 5 patients, aneurysmal rupture and in 2 patients, rupture of the AVM, caused the subarachnoid/intracerebral hemorrhage. In 9 patients, the concurrent AVM and aneurysm were in close proximity and so it was not possible to distinguish the etiology of hemorrhage. Six of these patients had an intranidal aneurysm and one each respectively, had an intra- and extranidal aneurysm, a deep seated perforator related aneurysm with AVM and an aneurysm on a superficial cortical branch in close proximity to the AVM.

The aneurysms associated with AVMs in the present series belonged to the following categories: (I) Flow related proximal [$n = 2$, Figures 1 and 2]; (II) flow related distal ($n = 3$); (III) intranidal [$n = 5$, Figures 3, 9 and 10]; (IV) both extra- and intranidal [$n = 2$, Figure 4]; (V) remote major ipsilateral [$n = 1$, Figures 5 and 6]; (VI) remote major contralateral [$n = 1$, Figure 7]; (VII) deep perforator related [$n = 1$, Figure 8]; (VIII) superficial ($n = 1$); and (IX) distal ($n = 0$) [Table 1]. Apart from a single giant basilar top aneurysm, the rest were small to medium sized aneurysms.

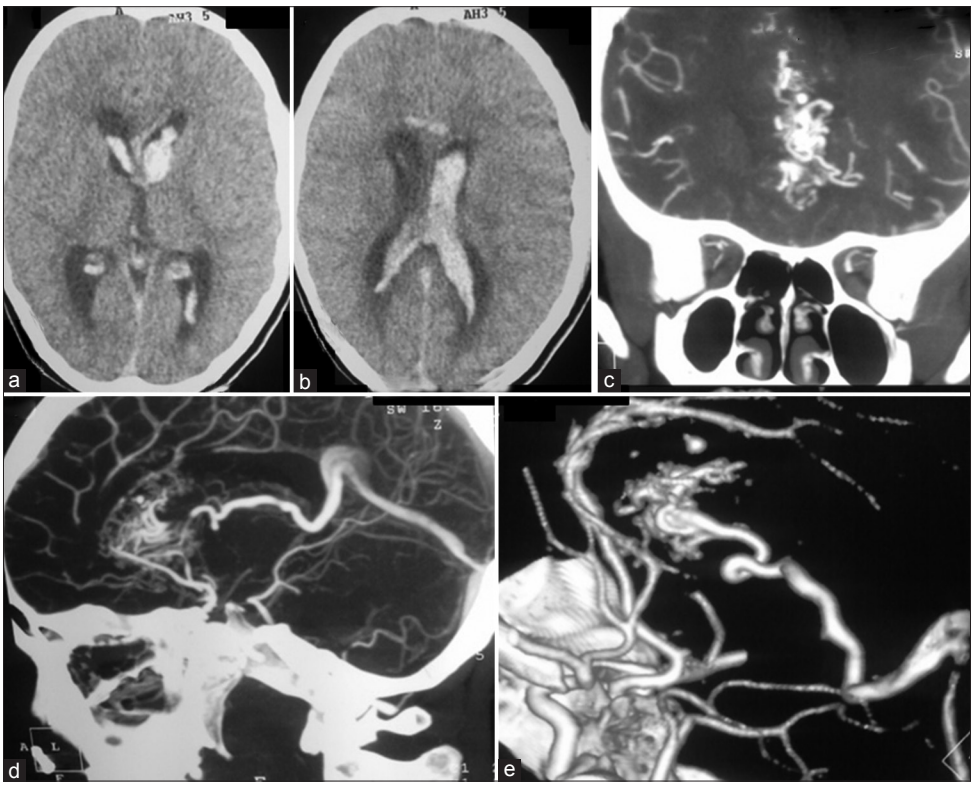

Figure 10: (Patient 12): (a and b) Axial CT scan showing intraventricular and corpus callosal bleed; (c) AP view; and, (d and e) lateral view of CT angiogram showing an intranidal aneurysm and deep seated caudate nucleus and intraventricular AVM supplied by ACA and its perforators and draining into the superior sagittal sinus and via the internal cerebral vein into the straight sinus

Table 3: Clinical spectrum of patients with simultaneous presence of aneurysm and AVM

Patient no.	Age (in years)/ sex	Clinical presentation	Duration of presentation (in days)
1 [Figure 1]	45Y/M	Sudden severe headache, vomiting, seizures	45
2 [Figure 2]	47Y/M	Sudden severe headache, vomiting, neck stiffness	14
3	40Y/M	Sudden severe headache, vomiting, altered sensorium, follows simple command, neck stiffness	19
4 [Figure 3]	45Y/F	Sudden severe headache, vomiting	14
5 [Figure 4]	45Y/F	Sudden severe headache, vomiting, right cerebellar ataxia	2
6 [Figures 5 and 6]	60Y/M	Sudden severe headache, vomiting, hemiparesis	7
7 [Figure 7]	33Y/M	Sudden onset headache, vomiting, altered sensorium	7
8 [Figure 8]	40Y/M	Sudden severe headache, recurrent vomiting	8
9	45Y/F	Sudden severe headache, vomiting, altered sensorium	2
10	45Y/F	Sudden severe headache, vomiting, transient unconsciousness	2190 (6 years ago)
11 [Figure 9]	29Y/M	Sudden onset right homonymous hemianopia	90
12 [Figure 10]	12Y/F	Sudden severe headache, recurrent vomiting, transient unconsciousness	14
13	17/F	Sudden severe headache	9
14	35/M	Grand mal seizure 2 years, sudden onset headache and transient unconsciousness 1 month ago	730 (2 years)
15	30/M	Headache, sudden unconsciousness 3 h	10
16	45/M	Headache, vomiting	30

AVM – Arteriovenous malformation; No – Number

The aneurysms had a fairly widespread location on blood vessels of the circle of Willis or their branches, namely the A comm A ($n = 3$), MCA ($n = 5$), posterior cerebral artery ($n = 4$), distal ACA ($n = 1$), basilar bifurcation ($n = 1$), anterior inferior cerebellar artery ($n = 1$) and posterior inferior cerebellar artery ($n = 1$).

The AVMs were located in the frontal ($n = 2$), temporal/perisylvian ($n = 5$), parietal ($n = 2$), occipital ($n = 2$), basal ganglionic/lateral ventricular ($n = 3$); or cerebellar ($n = 2$) locations [Table 4]. Their SM grade was I in 5, II in 3, III in 3, IV in 4 and V in 1. Ten patients had a LF and six, a HF AVM [Table 1]. In 7 of these patients, the AVM was located in the eloquent cortex.

The procedures performed are summarized in Table 5. Clipping of the aneurysm and excision of the AVM was performed in 4 patients (Patient 1, 3, 4, and 7) and excision of both AVM and aneurysm in 2 patients (Patient 5 and 11). The procedure was staged and two separate craniotomies made in a patient who had ACA dominance and A Comm A aneurysm filling from the side contralateral to that harboring the frontal AVM [Patient 7, Table 5, and Figure 7]. Two patients with an intranidal HF AVM in deep seated basal ganglionic/intraventricular location and in SM grade 4 and 5 respectively [patients 8 and 12, Figures 8 and 10] were only administered stereotacic radiosurgery. Five patients (4 with intranidal aneurysms [patients 9, 13, 14, 15] and one with aneurysm on superficial cortical branch supplying the AVM [patient 16]) underwent direct embolization of the AVM as well as the aneurysm. One patient each respectively, underwent clipping of the aneurysm and stereotactic radiosurgery of the AVM (Patient 2), and clipping of aneurysm with staged embolization of AVM (patient 6) [Table 5].

One patient who underwent excision of the AVM and clipping of the A Comm A aneurysm died due to development of post-operative extradural hematoma and septicemia. One patient had improved significantly in neurological status at discharge (modified Rankin's grade 2) and at first follow-up with resolution of the ganglionic hematoma and had been referred for stereotactic radiosurgery but was later on lost to long-term follow up. At follow-up ranging from 1.5 to 69 months (mean follow-up: 23.29 months), 12 patients had normalization of neurological disability (modified Rankin's grade 0), 2 were in grade 2 and 1 in grade 3, respectively [Table 5].[7-9] One other patient, following successful clipping of the A comm A aneurysm, is undergoing a staged embolization of the AVM with onyx glue [patient 6, Figures 5 and 6]. The patients were referred to another center for radiosurgery where the following protocol for radiosurgery was utilized. The radiation used for SM grade 4 and 5 was 17-20 Gy with follow-up every 6 months for the 1st year and then every year thereafter. A residual AVM usually warranted a repeat radiosurgery and/or microsurgery depending upon its accessibility and grade. A pre-radiosurgery embolization was also a part of the armamentarium for the multimodality strategy for these AVMs. The rationale for decision making due to the simultaneous presence of aneurysm with AVM is summarized in Table 6.

Discussion

Combined culpability of concurrent AVM and aneurysm

The simultaneous presence of an aneurysm with an AVM introduces several additional management dilemmas apart from those related to their individual natural histories. Firstly, aneurysms and AVMs are both prone to bleed; the presence of concurrent aneurysms either within the nidus, close to it or even remotely situated, increases the risk of hemorrhage associated with AVMs several folds.[2,10,11] An escalation in annual risk of 7% for intracranial hemorrhage in patients harboring unruptured AVMs with a concurrent AA compared with a 1.7% annual risk for patients with an AVM but without an aneurysm has been reported.[7] From the time of its diagnosis, a 9.8% hemorrhage rate of an AA is significantly higher than the 2-4% annual risk of hemorrhage

Table 4: Radiological characteristics of aneurysm and AVM and grading according to present classification

Patient number, site and size of aneurysm	CT/MR	Fisher grade	AVM location	Spetzler Martin grade of AVM	Arterial supply to AVM	Venous drainage of AVM	AVM flow	Aneurysm/ AVM/both ruptured*	Grading according to present classification
Pt. 1: Medium sized anterior communicating A aneurysm with right A1 dominance	Blood in gyrus rectus, anterior interhemispheric fissure, orbito-frontal cortex	IV	Right posterior frontal	2 (size: 2; venous drainage: 0; eloquence: 0)	Anterior communicating artery and MCA	Single vein into superior sagittal sinus	Low	Aneurysm*	An1*, AVM 2, LF
Pt. 2: Giant basilar apex aneurysm high above dorsum sellae pointing anteriorly	Blood in inter-peduncular cistern, supra-sellar cistern	II	Left occipital	4 (size: 2; venous drainage 1; eloquence: 1)	PCA and SCA	Multiple veins into superior sagittal sinus and transverse sinus	High	Aneurysm*	An1*, AVM 4, HF
Pt. 3: Two aneurysms on tonsillo-medullary segment of left PICA present proximal to AVM	Cistern magna, IVth and lateral ventricular bleed with hydrocephalus	IV	Left CP angle cistern and left cerebellar hemisphere	1 (size: 1; venous drainage: 0; eloquence: 0)	PICA	Transverse sinus	Low	Aneurysm*	An2*, AVM1, LF
Pt. 4: Intranidal aneurysm arising from right MCA	Right perisylvian fronto-parietal cortical bleed	III	Right temporal and perisylvian AVM	3 (size: 2; venous drainage: 0; eloquence: 1)	M2 and M3 segments of right MCA	Cortical vein into vein of Labbe	Low	Not distinguishable from aneurysm or AVM	An3*, AVM3*, LF
Pt. 5: Intra and extranidal aneurysms arising from AVM supplied by right AICA	Infratentorial superior cerebellar surface large hematoma	No SAH	AVM on right superior cerebellar surface	2 (size: 2; venous drainage: 0 eloquence: 0)	Right AICA	Cerebellar cortical vein	Low	Not distinguishable from aneurysm or AVM	An4*, AVM2*, LF
Pt. 6: Anterior communicating artery aneurysm situated remotely from AVM (aneurysm being filled by the ipsilateral ICA injection supplying both aneurysm and AVM)	Blood in interhemispheric fissure, septum pellucidum and lateral ventricles	IV	AVM in right posterior temporal region	4 (size: 3; venous drainage: 1; eloquence: 0)	Right MCA and PCA	Transverse sinus and superior sagittal sinus	High	Aneurysm*	An5*, AVM4, HF
Pt. 7: Anterior communicating artery aneurysm situated remotely from AVM (aneurysm being filled by the left ICA contralateral to the side supplying the right frontal AVM)	Right frontal hematoma	No SAH	AVM in right frontal region	1 (size: 1; venous drainage: 0; eloquence: 0)	Right frontopolar branch of ACA	Ophthalmic vein	Low	AVM*	An6, AVM1*, LF
Pt. 8: Multiple aneurysms associated with deep seated AVM supplied by perforators of right MCA and ICA	Right basal ganglionic hematoma	No SAH	AVM in right basal ganglionic region	5 (size: 3; venous drainage: 1; eloquence: 1)	Right M1 segment of MCA and ICA perforators	Multiple draining veins into transverse and sigmoid sinus, internal cerebral vein and straight sinus	High	Not distinguishable from aneurysm or AVM	An7*, AVM5*, HF
Pt. 9: Multiple intranidal aneurysms supplied by M2 branches of right MCA	Right temporal hematoma	No SAH	AVM in right temporal lobe	3 (size: 2; venous drainage: 1; eloquence: 0)	M2 branches of right MCA	Superior sagittal sinus, superficial and deep middle cerebral veins	High	Not distinguishable from aneurysm or AVM	An3*, AVM3*, HF
Pt. 10: Aneurysm on P4 segment of right distal PCA	Right medial temporo-occipital hematoma	No SAH	AVM in right medial temporo-parietal region	1 (size: 1; venous drainage: 0; eloquence: 0)	P4 segment of right distal PCA	Cortical vein on medial temporal region draining into straight sinus	Low	Aneurysm*	An2*, AVM1, LF

Contd...

Table 4: Contd...

Site and size of aneurysm	CT/MR	Fisher grade	AVM location	Spetzler Martin grade of AVM	Arterial supply to AVM	Venous drainage of AVM	AVM flow	Aneurysm/ AVM/both ruptured*	Grading according to present classification
Pt. 11: Intranidal aneurysm within AVM in posterior parietal interhemispheric region supplied by distal left PCA artery	Left posterior parietal hematoma	No SAH	AVM in left posterior parietal region	1 (size: 1; venous drainage: 0; eloquence: 0)	P4-5 segment of distal PCA	Cortical vein to superior sagittal sinus	Low	Not distinguishable from aneurysm or AVM	An3*, AVM1*, LF
Pt. 12: Intranidal aneurysm in deep seated caudate nucleus and intra-lateral ventricular AVM supplied by ACA	Intraventricular hematoma	No SAH	Caudate nucleus and intraventricular lateral ventricular AVM	4 (size: 2; venous drainage: 1; eloquence: 1)	Distal ACA	Vein to superior sagittal sinus and internal cerebral vein to straight sinus	High	Not distinguishable from aneurysm or AVM	An3*, AVM4*, HF
Pt. 13: Two intranidal aneurysms in deep seated peritrigonal AVM supplied by Rt MCA	Intraventricular hematoma	No SAH	Rt peritrigonal deep seated AVM	4 (size: 2; venous: 1; eloquence: 1)	M3 branches of Rt MCA	Vein to superior sagittal sinus	Low	Not distinguishable from aneurysm or AVM	An3*, AVM4*, LF
Pt. 14: Medium sized nidal-extranidal aneurysms on P3 segment of Lt PCA	Parietal hematoma	No SAH	Left parietal AVM	1 (size: 1; venous drainage: 0; eloquence: 0)	Lt posterior choroidal artery and P3 segment of left PCA	Vein to superior sagittal sinus	Low	AVM	An4, AVM1*, LF
Pt. 15: Lt MCA medium sized aneurysm with AVM	Lt frontotemporal parenchymal hematoma	No SAH	Lt temporal and perisylvian AVM	2 (size: 1; venous drainage: 0; eloquence: 1)	M3 segment of Lt MCA	Vein to transverse-sigmoid junction	Low	Not distinguishable from aneurysm or AVM	An 2*, AVM2*, LF
Pt. 16: Rt occipital aneurysm situated on superficial cortical branch of PCA with AVM supplied by PCA	Occipital hematoma	No SAH	Rt occipital AVM	3 (size: 2; venous drainage: 0; eloquence: 1)	Rt PCA	Vein to superior sagittal sinus	High	Not distinguishable from aneurysm or AVM	An8*, AVM3*, HF

MCA – middle cerebral artery; AVM – arteriovenous malformation; LF – low flow; PCA – posterior inferior cerebellar artery; SCA – superior cerebellar artery; HF – high flow; PICA – posterior inferior cerebellar artery; CT – computed tomography; MR – magnetic resonance; CP – cerebellopontine; AICA – anterior inferior cerebellar artery; SAH – subarachnoid hemorrhage; ICA – internal carotid artery

associated with AVM alone.[5] Histology has also confirmed that these AAs are thin walled and devoid of elastic fibers or a thick muscular element. They are exposed to the same arterial pressures as arterial components of the AVM and are therefore prone to bleed.[12] Secondly, determining the etiology of the hemorrhage often becomes difficult.[5,13] In 9 of our patients with an intranidal, intra- and extranidal, deep perforator related or superficial aneurysms, it was not possible to distinguish whether the aneurysm or the AVM had bled. Thirdly, aneurysms and AVMs may occur on remote vessels and may represent a coincidental phenomenon or may represent an underlying vascular anomaly, hemodynamic alteration, vasoactive stimuli or the action of growth factors, leading to vascular remodeling.[3,7,14-16] The latter are conditions that may need to be identified and suitably modified to prevent recurrence. Fourthly, the rapidity of flow through the AVM and its SM grade would determine its resectibility. Institution of endovascular embolization or stereotactic radiosurgery as a primary or adjunctive therapy may often be necessary in higher grade AVMs with rapid fistulous connections.[12,16] This would fail to concurrently take care of the associated small but potentially risky aneurysms without an adequate neck that are often associated with AVMs. Fifthly, appropriate management of the aneurysm would often depend upon its distance from the coexisting AVM and its anatomical characteristics. Finally, it is not clear if the two entities need to be managed simultaneously or separately and which of conditions should be addressed first? Initial elimination of the AVM may initiate hemodynamic alterations that may lead to regression of the associated aneurysm;[3,17] conversely, AVM excision may increase the blood flow in feeding arteries and increase the risk of aneurysm rupture.[1]

The present classification

The traditional classification of concurrent AAs associated with AVMs proposing the categories of flow related aneurysms, nidal aneurysms and remote aneurysms has withstood the test of time and also has therapeutic and prognostic implications.[4,5,10,12,16,18] While retaining the basic framework of the original classification, in this study, a more comprehensive sub-categorization has been offered that perhaps will cover all clinical variations of AAs coexisting with AVMs. It has been recognized that any management protocol for concurrent aneurysms with AVMs does not depend solely on the type

Table 5: Intervention performed in patients with AVM and aneurysm and their outcome at follow-up

Patient number	Endovascular procedure			Surgery	Approach	Status of AVM	Stereotactic radiosurgery for AVM	Modified Rankin's score		Follow-up (in months)	Complications
	For aneurysm	For AVM	For both					At admission	At follow-up		
Pt. 1	-	-	-	Clipping of aneurysm and excision of AVM	Right pterional with right posterior frontal parasagittal approach	Total excision	-	2	0	42	-
Pt. 2	-	-	-	Clipping of aneurysm	Left fronto-temporal orbito-zygomatic osteotomy and subtemoral-infratemporal approach	-	Yes	3	0	38	-
Pt. 3	-	-	Micro-catheter-ization and embolization attempt failed due to vessel tortuosity	Clipping of aneurysms and excision of AVM	Midline suboccipital craniectomy	Total excision	-	4	3	6	Left cerebellar signs, IX-Xth nerve paresis. PICA not filling on follow-up DSA
Pt. 4	-	-	-	Clipping of aneurysm and excision of AVM	Right frontotemporo-zygomatic craniotomy and transsylvian approach	Total excision	-	2	0	45	-
Pt. 5	-	-	-	Excision of aneurysm and AVM	Rt temporal craniotomy, subtemoral transtentorial excision of AVM/ aneurysm	Total excision	-	2	0	69	-
Pt. 6	-	AVM partially embolized with onyx	-	Clipping of aneurysm and staged partial embolization of AVM	Left pterional craniotomy and trans-sylvian clipping of A Com A aneurysm (due to left ICA dominance)	Partial embolization	Yes	4	2	4	-
Pt. 7	-	-	-	Clipping of aneurysm and excision of AVM	Left pterional craniotomy and trans-sylvian clipping of A com A aneurysm (due to left ICA dominance) and right frontal craniotomy and excision of AVM	Total excision	-	4	6	-	Transtentorial herniation due to postoperative EDH; septicemia. Died
Pt. 8	-	-	-	-	-	No surgery	Yes	5	2	Lost to follow-up	-
Pt. 9	-	AVM embolized with onyx	-	Complete embolization of AVM	-	No surgery	-	2	0	29	-
Pt. 10	-	-	-	Clipping of aneurysm and excision of AVM	Right temporal craniotomy and subtemporal approach for clipping of aneurysm and excision of AVM	Total excision	-	2	0	24	-

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Table 5: Contd...

Patient number	Endovascular procedure			Surgery	Approach	Status of AVM	Stereotactic radiosurgery for AVM	Modified Rankin's score		Follow-up (in months)	Complications
	For aneurysm	For AVM	For both					At admission	At follow-up		
Pt. 11	-	-	-	Clipping of aneurysm and excision of AVM	Left parietal parasagittal craniotomy and excision of aneurysm and AVM	Total excision	-	0	0	1.5	-
Pt. 12	-	-	-	-	-	-	Yes	0	0	1.5	-
Pt. 13	-	AVM embolized with onyx		Complete embolization of AVM and aneurysm	-	No surgery	-	0	0	12	-
Pt. 14	-	AVM embolized with onyx; Aneurysm coiled		Complete embolization of AVM and aneurysm	-	No surgery	No	2	0	12	Rt hemiparesis, homonymous hemianopia and sensory loss on 2nd post-procedure day. All improved by 5th postoperative day. Asymptomatic at 1 year F/U
Pt. 15	-	AVM embolized with onyx		Complete embolization of AVM and aneurysm	-	No surgery	-	0	0	6	-
Pt. 16	-	AVM embolized with onyx. Aneurysm obliterated with AVM		Complete embolization of AVM and aneurysm	-	No surgery	-	0	0	36	-

MCA – middle cerebral artery; AVM – arteriovenous malformation; PICA – posterior inferior cerebellar artery; DSA – digital subtraction angiogram; ICA – internal carotid artery; EDH – extradural hematoma

Table 6: Rationale for decision making in the present series

Procedure performed	Patient number	Factors determining the procedure undertaken
Clipping of aneurysm and excision of AVM using same craniotomy	1	Proximal flow related aneurysm and AVM (SMG 2, LF) accessible by same craniotomy
	3	Distal flow related aneurysm and AVM (SMG 1, LF) accessible by same craniotomy
	4	Intranidal aneurysm and AVM (SMG 3, LF) accessible by same craniotomy
	10	Distal flow related aneurysm and AVM (SMG1, LF) accessible by same craniotomy
	11	Intranidal aneurysm and AVM (SMG 1, LF) accessible by same craniotomy)
Clipping of aneurysm and excision of AVM using staged craniotomy at 2 different sites	7	Remote major contralateral aneurysm and AVM (SMG 1, LF) surgically resectable. Contralateral dominance of ACA and direction of fundus of aneurysm directed approach to aneurysm and AVM from opposite sides
Excision of aneurysm and AVM	5	Intra/extranidal aneurysm and AVM (SMG2, LF) in same low flow nidus in non-eloquent location
Clipping of aneurysm with embolization of AVM	6	Remote ipsilateral aneurysm with AVM (SM_4, HF) with both not accessible by same approach
Clipping of aneurysm and SR for AVM	2	Giant basilar aneurysm causing brain stem compression was proximal flow related. AVM (SMG_4, HF) was not accessible by same approach
Only embolization	9, 13, 15	Intranidal/extranidal aneurysm with diffuse/deep seated AVM
	16	Aneurysm on superficial cortical branch of feeding vessel in close vicinity to aneurysm
	14	Both AVM and aneurysm accessible by endovascular route
Only SR	8	Deep seated perforator related aneurysm with basal ganglionic AVM (SM_5, HF) in eloquent cortex
	12	Deep seated basal ganglionic intranidal aneurysm with AVM (SMG_4, HF) in eloquent cortex

AVM – arteriovenous malformation; SMG – Spetzler Martin Grade; ACA – anterior cerebral artery; HF – high flow; SM – Spetzler-Martin; SR – stereotactic radiosurgery

of aneurysm present but also on whether the aneurysm or the AVM has bled[5] and the SM grade along with the flow status of the AVM.[6] A useful abbreviated nomenclature that succinctly summarizes the varying clinical situations in which intracranial AAs and AVMs coexist has also been included.

The rationale for our classification was based on the differences between groups with regard to the approach and feasibility of surgical obliteration of the aneurysms and AVMs. The flow related proximal aneurysms existed on major arteries of the circle of Willis while the flow related distal aneurysms were based on distal branches of circle of Willis that were supplying the AVM but at a distance from the nidus.[4,5,10,18] Nidal aneurysms were situated within the AVM nidus while the extranidal aneurysms were situated on one of the peripheral nidal vessels with the aneurysm in close proximity but clearly distinguishable from the nidus of the AVM.[5] This differentiation would be necessary in a clinical scenario since it would often be possible to excise the intranidal and intra- and extranidal aneurysms along with the AVMs. Flow related aneurysms (whether proximal or distal), on the other hand, would usually require an additional focus on clipping/embolization of the aneurysm often utilizing a staged and/or separate approach for addressing the AVMs. Deep perforator related aneurysms may be associated with HF AVMs in eloquent regions of the brain and may not be suitable for surgical resection. A distal aneurysm uniquely occurs distal to the AVM on the feeding vessel while the superficial aneurysm occurs on one of the unnamed superficial cortical branches that has undergone a pathological dilatation and alteration of course to supply the AVM.[4]

Surgical planning for clipping of the aneurysm and excision of the AVM would also often be determined by the angiographic dominance of the circulation and the direction of fundus of the aneurysm. Thus, remote aneurysms could be divided into two categories: One where the aneurysm was being filled by the ipsilateral circulation harboring the AVM; and other, where the aneurysm was getting filled by the contralateral dominant circulation (so that the surgical approach would entail separate, staged craniotomies on either side to obliterate the two entities). This was well exemplified in our two patients with A Comm A aneurysms located on vessels remote from the feeding artery of the AVM (patients 6 and 7). In one of them, the circulation ipsilateral to, while in the other, the circulation contralateral to the side of the AVM was the dominant one and filling the aneurysm. The surgical approach, therefore, had to be tailored accordingly. It is also evident from this series that simultaneous decision making for the obliteration of AVMs would necessarily entail evaluating their flow status and SM grade. Thus, surgical excision in a LF SM grade I and II AVM would be preferable to endovascular obliteration; however, therapeutic embolization or stereotactic radiosurgery would be the more suitable option in SM grade IV and V, HF AVMs.

Emerging recommendations

Despite variations in management of individual patients in our series, some salient points emerged. Firstly, in SM grade I, II and III LF AVMs associated with flow related proximal or distal aneurysms that are surgically accessible through the same route as the coexisting AVM, surgical excision of the AVM with clipping of the aneurysm may be the most preferable option. If the AVMs and the aneurysms are not surgically accessible by the same route, a staged surgery using different corridors, endovascular embolization or surgery for the aneurysm and stereotactic radiosurgery for the AVM may be utilized. If the LF, grade I to III AVMs are associated with an intranidal or intra-extranidal aneurysm, then direct simultaneous excision of both the aneurysm and AVM may be performed. Decision making regarding flow related, intra- and intra-extranidal aneurysms associated with SM grade III, HF AVMs would, however, be variable and be based upon the eloquence of the cortex, their relative surgical accessibility, the diffuseness of the AVM and the rapidity of the arteriovenous fistula. Perhaps, therapeutic embolization of both; or, transarterial embolization of the aneurysm and steretotactic radiosurgery of the AVM would be the preferable option.

Surgical approach for remote ipsilateral or contralateral aneurysms associated with AVMs would need additional focus on the dominance of feeding vessels and the direction of fundus.

Finally, intranidal or intra-extranidal and deep perforator related aneurysms with a HF AVM in SM grade IV or V would not be considered for surgery and would solely require either therapeutic embolization or stereotactic radiosurgery or just observation.

Emphasis on surgical excision

In the present series, the role of surgery in facilitating immediate and definitive control of concurrent aneurysms with AVMS is emphasized.[4] However, wherever indicated, endovascular techniques or stereotactic radiosurgery were actively sought for their primary or adjunctive therapeutic potential.[13] Endovascular obliteration may induce thrombosis of aneurysms proximal to the AVM nidus as was observed in one of our patients (patient 9). It permits simultaneous access to flow related and remote aneurysms in conjunction with the AVM. Even staged partial embolization may help in eradicating the AVM nidus to prevent breakthrough bleeding.[12,18-20] It may also help to overcome the steal phenomenon by improving the regional cerebral blood flow around the AVM.[12,16,21,22]

We persisted with surgery in some of the cases due to several reasons. The AVMs associated with AAs were often having a LF; and, the aneurysm may be within or in close proximity to the AVM nidus. Tortuous arteries and multiple nidi of the AVM as well as small and multiple aneurysms within or around the AVM may often preclude its complete obliteration

utilizing the endovascular route. HF arteriovenous fistulae may increase the risk of distal embolism of the embolic material with its potential to obstruct end arteries of the brain. Both staged therapeutic embolization and primary radiosurgery are relatively expensive and require a long latency period to effect thrombosis of the AVM during which there is a continued risk of hemorrhage from the unobliterated nidus. Larger AVMs are less likely to completely thrombose with stereotacic radiosurgery also.[12]

The "focus on the aneurysm" principle

The present study clearly demonstrates that in contrast to the population that harbors an AVM alone, AVM with AA is much more common in older patients and is rare in children. In support of this finding, Lasjuanias *et al*. in their study observed that only 8% of patients less than 25 years presented with AAs, while 37% of patients between 25 and 50 years had an aneurysm associated with an AVM.[2,10] The presence of a concurrent aneurysm with the AVM, therefore, indicates an alteration in both the population characteristics as well as in the etiopathogenesis of the intracranial hemorrhage.

The simultaneous presence of an aneurysm with AVM often leads to a significant modification in the surgical planning. Aneurysms within or close to the AVM nidus may be too small to be clipped or coiled and may also not be obliterated utilizing endovascular technique or stereotacic radiosurgery. Aneurysms situated at a distance from the AVM, or even remotely (including those present contralaterally), may require additional/staged surgical or endovascular approaches.

The presence of AVM causing hemorrhage is not treated with the same degree of urgency as accorded to cases with aneurysmal SAH. This was aptly exemplified in our study where 9 out of the 16 patients harboring both an aneurysm and AVM were referred for DSA or MR/CT angiography and definitive treatment after a delay of 2 weeks or more following their initial ictus. This was despite the fact that the initial CT scan clearly pointed toward the presence of an AVM either due to the appearance of characteristic serpiginous vessel hyperdensities or due to the pattern of the lobar bleed. The emergent situation arising due to the simultaneous presence of an aneurysm and AVM with additional risk of hemorrhage is almost never considered under these circumstances and routine protocol is instituted for investigation and management of the suspected AVM. This study highlights that In the patients in whom an AVM is suspected as the primary cause of intracranial hemorrhage, special efforts must be undertaken to identify concurrent aneurysms. Further grading, institution of management protocols and prognostication should particularly "focus on the aneurysms."

Limitations of the study

The personal evaluating outcome were not blinded to the treatment administered and were participating in the care of the patients. While we were fortunate to include patients representing most of the categories of aneurysms with AVMs in the series, one of the groups (group IX: distal) went unrepresented. The patients included in this series were those who were admitted for surgical or endovascular obliteration of the aneurysm with AVM due to the presence of intracranial hemorrhage. Therefore, all of them underwent an interventional or surgical procedure and none of them were followed without treatment.

Outcome assessment in a larger number of patients would have added further credence to the validity of our classification in predicting recommendations for management of patients having concurrent aneurysms with AVMs. Finally, the protocols of management were not always consistent and varied between individual patients. A prospective assessment of the proposed grading with rigid treatment protocols would perhaps sanctify standard of care in these patients.

Conclusions

While encountering AVMs with intracerebral hemorrhage, special efforts must be undertaken to identify concurrent aneurysms. A comprehensive subcategorization of concurrent aneurysms as well as an abbreviated nomenclature has been offered that perhaps will cover all clinical variations of AAs coexisting with AVMs. A management protocol for concurrent aneurysms with AVMs has been proposed that depends on the type of aneurysm present, on whether the aneurysm or the AVM has bled and, the SM grade and the flow status of the AVM.

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