Cerebral Arteriovenous Malformation (AVM)

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Abstract: An arteriovenous malformation (AVM) is another cause of brain hemorrhage and it comprises about 15% of intracerebral hematomas. It often occurs at a young age. Bleeding from an AVM most often occurs between the ages of 10 and 30 years. It often causes a cerebral hematoma in the frontal lobe, temporal lobe, occipital lobe or parietal lobe, and in some cases, even within the ventricle. Cerebellar AVMs are less frequent. Besides an ICH, another type of presentation of an AVM is seizure disorders, and it is commonly partial seizure. Investigations of a cerebral AVM include a CT scan, MRI of the brain, CTA, MRA and DSA, and sometimes tractography and fMRI are necessary for a diagnosis, as well as a complete and compact understanding of an AVM and the planning of management. The management options for a brain AVM are microsurgical excision, endovascular embolization, radiosurgery or any possible combination of the above three options. Microsurgery is the main option for curative treatment. In this chapter, the pathology, clinical presentation, investigation, grading of AVM, options for treatment, choosing option/s and a brief introduction of microsurgical excisions are mentioned.

Abbreviations

AOVM	angiographically occult vascular malformation	AV	Fistula–arteriovenous fistula
AVM	arteriovenous malformation	CCF	caroticocavernous fistula
CNS	central nervous system	CT	computed tomography
CT	angiogram-computed tomographic angiogram	DSA	digital subtraction angiogram
DVA	developmental venous anomaly	fMRI	functional magnetic resonance imaging
MR	angiogram–magnetic resonance angiogram	MR	scan–magnetic resonance scan
MR	tractography–magnetic resonance tractography		

1. Introduction

An AVM is a nonneoplastic vascular malformation of the CNS. An arteriovenous malformation is another cause of brain hemorrhage and it constitutes about 15% of intracerebral hematomas. It often affects the younger generation. Bleeding from an AVM most often occurs between the ages of 10 and 30 years, which is a little different from an SAH which is common in older or middle-age groups of people. It often causes a cerebral hematoma in the frontal lobe, temporal lobe, occipital lobe or parietal lobe, and in some cases, even within the ventricle. Cerebellar AVMs are less encountered now-a-days. Besides an ICH, another type of presentation of an AVM is seizure disorders, and it commonly partial seizure (Greenberg 2010).

2. Types Vascular Malformations

McCormick described vascular malformation in 1966 (McCormick 1966):

a. Arterio-venous malformation (AVM);
b. Cavernous malformation;
c. Developmental venous anomaly (DVA), formerly venous angioma;
d. Capillary telangiectasia.

Possible additional categories:

1. A direct fistula is also known as an arteriovenous fistula (AV fistula). One or numerous dilated arteriolar direct connections to a draining vein sans an intervening nidus is the pathology of an AV fistula. These have high pressure, as well as high flow with a low frequency of hemorrhage. They are generally amenable to endovascular techniques. Examples include the following:

   (i) Carotid–cavernous fistula (CCF);
   (ii) Vein of Galen malformation (aneurysm);
   (iii) Dural AVM.

2. For a mixed or unclassified angioma, 11% are angiographically occult vascular malformations (AOVMs) (July and Wahjoepramono 2019).

3. Components of an AVM

An AVM consists of the following (Figure 1):

- Feeding artery;
- Nidus;
- Draining vein/veins.

AVM-related arteries:

- Feeding artery—terminal artery of either the anterior, middle or posterior cerebral arteries.
- Transit artery (en passage)—branches to the AVM nidus but does not end into the nidus.

The choroidal feeding artery and perforating artery are examples of such a type.

- Bystander artery—normal artery that does not send supply to the AVM nidus but travels alongside or near to the nidus (Spetzler et al. 2015).

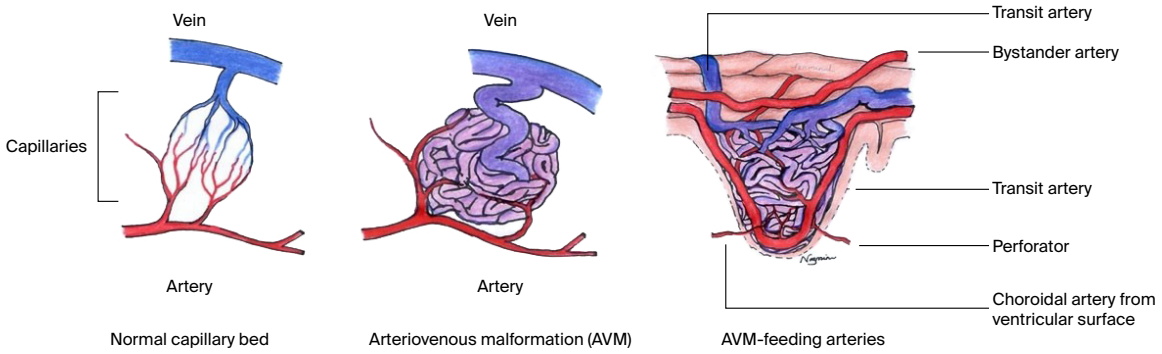

Figure 1. Schematic drawing of AVM. Source: Figure by authors.

4. Classification of AVMs

1. Pure dural AVM.
2. Parenchymal AVMs (discussed below) subclassified as:

   (i) Pial;
   (ii) Subcortical;
   (iii) Paraventricular;
   (iv) Combined.

3. Mixed parenchymal and dural (rare) (Spetzler et al. 2015).

Compact AVMs: The nidus of an AVM may be twisted tightly or loosely, according to preference. A compact AVM (Figure 2) has defined boundaries, no intermingled brain parenchyma, and is well apart from the surrounding brain, all of which help in defining a parenchymal dissection. Compact AVMs are simple to understand and follow.

Diffuse AVMs: With ambiguous boundaries, intermingled brain and poor distinctiveness, a diffuse AVM is twisted loosely, as if torn apart or unraveled, complicating parenchymal dissection. The neurosurgeon must determine the plane of demarcation between the AVM as well as the brain while dealing with diffuse AVMs. A diffuse AVM's border may be an uncontrolled periphery which has to be surrounded either broadly at the cost of the adjacent cerebral parenchyma, or narrowly enough to cross more of this vascular fringe. The interaction of eloquence as well as hemostasis pulls in and pushes back the circum-dissection, presenting a problem to the vascular neurosurgeon in determining the proper dissection distance. This decision may invite the dissection too near to the nidus, causing hemorrhage or leaving an aberrant piece of the nidus behind. Conversely, this decision may push the dissection much far away from the nidus, culminating in a full and simpler AVM excision, although more cerebral parenchyma removal than required (Spetzler et al. 2015).

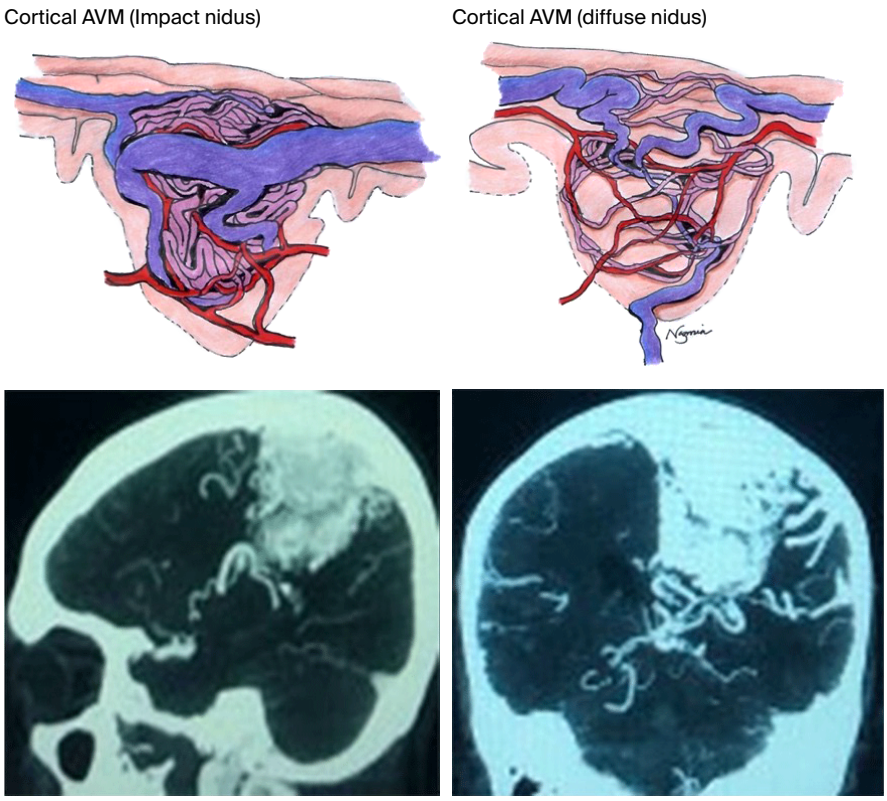

Figure 2. Image showing the compactness of an AVM. Source: Figure by authors.

5. Clinical Presentation

- Hemorrhage (50%):
  - Intracerebral (82%);
  - SAH;
  - Intraventricular;
  - Subdural.
- Seizure.
- Progressive neurological deficit (cerebral steal syndrome).
- Mass effect.
- Headaches.
- Hydrocephalus.

Of AVMs, 90% are supratentorial. The annual hemorrhage risk is 2–4%. If present with intracranial hemorrhage, then the annual hemorrhagic risk is 50% and epileptic seizure risk is 10–30% (Greenberg 2010; Nader et al. 2014; Knopman and Stieg 2014).

6. Natural History of Diseases

AVMs are associated with a 2.4% risk of bleeding per annum. The risk was the greatest in the 1st five years following diagnosis of an AVM and gradually reduced after that. Young age, past rupture, deep and infratentorial placements, and predominantly deep venous drainage are all possible causes for a recurrent AVM hemorrhage. Previous rupture, a big AVM, and infratentorial and deep placements are all risk factors on their own (Spetzler et al. 2015).

7. Radiological Assessment and Classification

- CT scan (Figure 3);
- CT angiogram;
- MR scan;
- Cerebral DSA (Figure 4);
- MR angiogram (Figure 5);
- MR tractography;

- Functional MRI (fMRI) (Greenberg 2010).

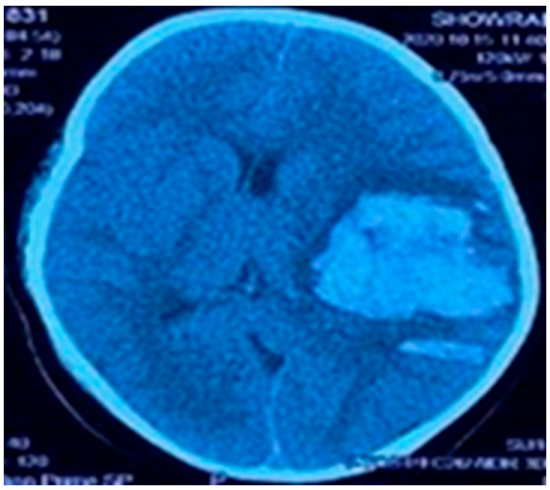

Figure 3. CT scan showing a lobar hematoma from a ruptured AVM. Multimodality imaging is needed in a cerebral AVM for the diagnosis, flow, feeders, drainage, location, size and compactness. Source: Figure by authors.

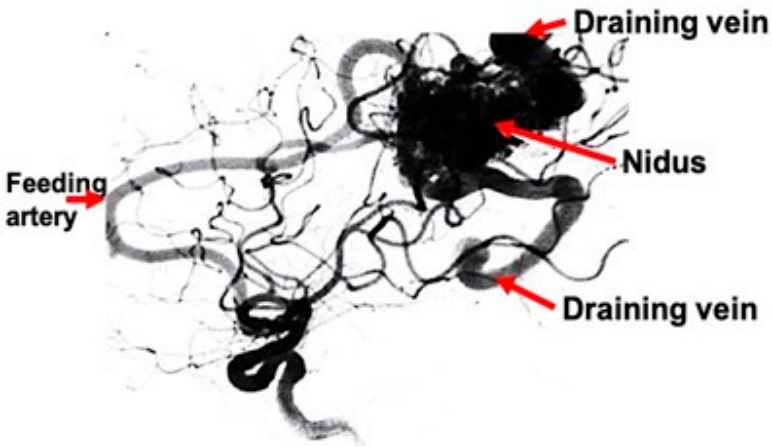

Figure 4. Angiography showing a compact nidus feed by the pericallosal artery and draining toward the deep venous system. Source: Figure by authors.

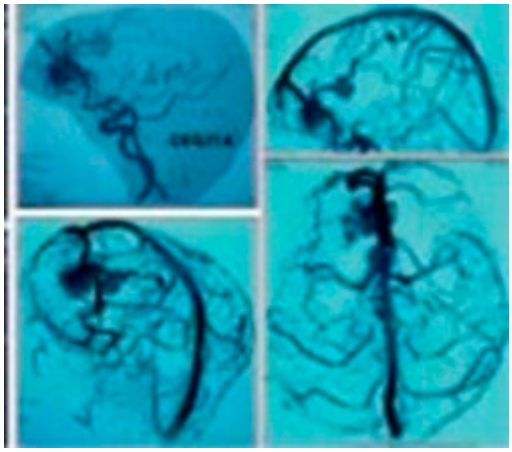

Figure 5. MRA showing a right frontal AVM feed by anterior circulation arteries and multiple drainages toward the superior sagittal sinus. Source: Figure by authors.

Yasargil classification of AVMs:

- Convexity (pallial);
- Central AVMs.

Limbic AVMs:

- Amygdalohippocampal;
- Parasplenial;
- Cingular;
- Callosal.

Medio-basal temporal AVMs:

- Amygdala;
- Anterior;
- Middle;
- Posterior;

Surgical exposure of different AVMs are shown in Table 1. Free surface of AVM is important for surgical resection (Table 2).

Table 1. Summary of the exposures to AVMs.

Type	Subtype	Craniotomy
Frontal AVM	Medial frontal	Bifrontal
	Lateral frontal	Frontal
	Paramedian frontal	Bifrontal
	Basal frontal	Orbital pterional
	Sylvian frontal	Pterional
Temporal AVM	Basal temporal	Temporal
	Lateral Temporal	Temporal
	Medial temporal	Orbitozygomatic
	Sylvian temporal	Pterional
Pareto-occipital AVM	Lateral parieto-occipital	Parieto-occipital
	Medial parieto-occipital	Torcular
	Paramedian parieto-occipital	Biparieto-occipital
	Basal occipital	Torcular
Ventricular/periventricular AVM	Callosal	Bifrontal
	Atrial	Parietal
	Ventricular body	Bifrontal
	Temporal horn	Temporal
Deep AVM	Anterior Midbrain	Orbito-zygomatic
	Posterior midbrain	Torcular
	Lateral pontine	Retrosigmoid
	Anterior Pontine	Retrosigmoid
	Anterior medullary	Suboccipital
	Lateral medullary	Far lateral
Cerebellar AVM	Suboccipital cerebellar	Suboccipital
	Vermian cerebellar	Torcular
	Tentorial cerebellar	Torcular
	Tonsillar cerebellar	Suboccipital
	Petrosal cerebellar	Retrosigmoid

Source: Authors' compilation based on data from Spetzler et al. (2015).

Table 2. Free surfaces of an AVM.

Free Surface on Convexity	AVM Subtype
Supratentorial	
Frontal convexity	Lateral frontal AVM, paramedian frontal AVM
Temporal convexity	Lateral temporal AVM
Parieto-occipital convexity	Lateral parieto-occipital AVM, paramedian parieto-occipital AVM
Infratentorial	
Cerebellar convexity	Suboccipital cerebellar AVM
Free Fissure surface	**AVM subtype**
Supratentorial	
Subfrontal plane	Basal frontal AVM
Subtemporal plane	Basal temporal AVM
	Medial temporal AVM (posterior)
Sylvian fissure	Frontal Sylvian AVM, temporal sylvian AVM, pure Sylvian AVM, insular AVM
Interhemispheric fissure	Medial frontal AVM, paramedian frontal AVM Medial parieto-occipital AVM
	Paramedian parieto-occipital AVM, callosal AVM
	Ventricular body AVM, basal ganglion AVM
Choroidal fissure	Ventricular body AVM Thalamic AVM
Supratentorial–infraoccipital	Basal occipital AVM
Infratentorial	
Sylvian fissure	Anterior midbrain AVM
Supracerebellar–infratentorial fissure	Tentorial cerebellar AVM, vermian cerebellar AVM, posterior midbrain AVM
Cerebello-mesencephalic fissure	Tentorial cerebellar AVM
	Posterior midbrain AVM
Cerebellopontine fissure	Petrosal cerebellar AVM, anterior pontine AVM, lateral pontine AVM
Cerebello-medullary fissure	Tonsillar cerebellar AVM, lateral medullary AVM

Source: Authors' compilation based on data from Spetzler et al. (2015).

Figure 6 are showing frontal convexity AVM.

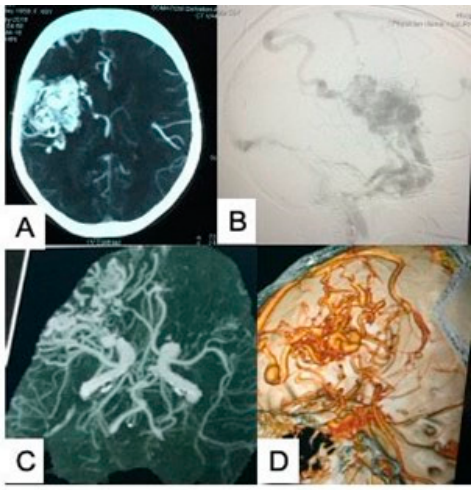

Figure 6. (**A**,**B**) CT angiogram showing a compact AVM of the lateral frontoparietal region fed by an MCA and venous drainage toward the superior sagittal sinus. (**C**,**D**) CT angiogram showing the right frontal diffuse AVM fed by branches from the middle cerebral artery and venous drainage toward the superior sagittal sinus. Source: Figure by authors.

The mesial parietal AVM is depicted in Figure 7.

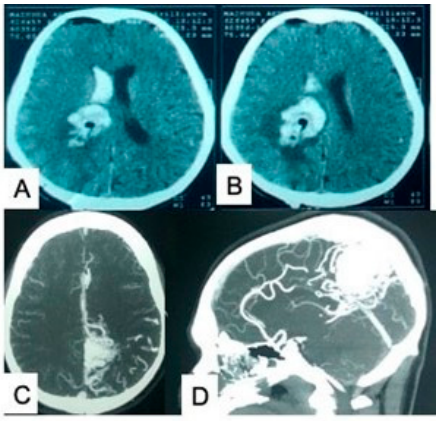

Figure 7. (**A**,**B**) CT scan of the brain showing an intraventricular along with a medial parietal bleed. (**C**,**D**) CT angiogram revealed a compact medial parietal (paracentral lobule) AVM fed by branches from the pericallosal artery and venous draining toward the superior sagittal sinus. Source: Figure by authors.

The posterior medial temporal AVM and Sylvian fissure AVM are demonstrated in Figures 8 and 9, respectively.

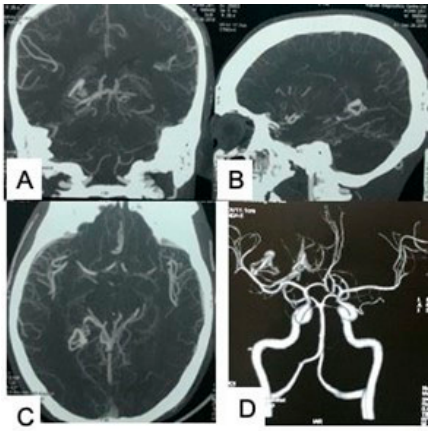

Figure 8. (**A**–**D**) CT angiogram showing a posterior medial temporal AVM fed by branches from the posterior cerebral artery and venous drainage toward the deep venous system. Source: Figure by authors.

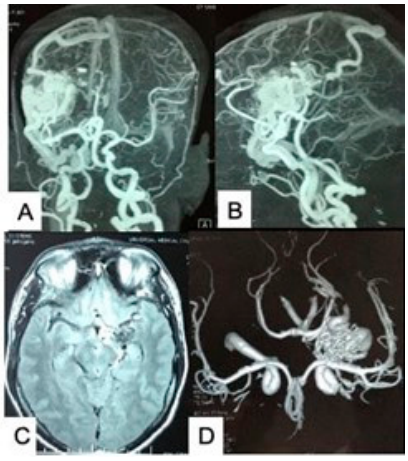

Figure 9. (**A**,**B**) CT angiogram showing a compact Sylvian fissure AVM fed by branches from the MCA and venous drainage toward the superior sagittal sinus. (**C**,**D**) MRA revealing a mesial temporal AVM fed by branches from the PCOM and anterior choroidal artery. Source: Figure by authors.

Spetzler–Martin Grading of AVM is shown in Table 3.

Table 3. Grading of AVMs.

Spetzler–Martin Grading	Points	Supplementary Grading
Size		Age, years
<3 cm	1	Less than 20
3–6 cm	2	20–40
>6 cm	3	More than 40
Venous drainage		**Hemorrhage**
Superficial	0	Yes
Deep	1	No
Eloquent brain		**Compactness**
Non-eloquent	0	Yes
Eloquent	1	No
Total Grade	5	

Note: Sensorimotor, language, visual cortex, hypothalamus, thalamus, internal capsule, brain stem, cerebellar nuclei, cerebellar peduncles or regions directly adjacent to these structures are eloquent brain. Source: Authors' compilation based on data from Spetzler et al. (2015) and Spetzler and Martin (1986).

A three-tier classification of brain AVMs was proposed, along with a management paradigm (Table 4).

Table 4. Three-tier classification of cerebral AVMs with management options.

Class	Spetzler-Martin Grade	Management
A	I and II	Resection
B	III	Multimodality treatment
C	IV and V	No treatment

Source: Authors' compilation based on data from Spetzler and Ponce (2011).

8. Treatment

There are various ways of treatment for AVMs depending on the Spetzler–Martin grading (Greenberg 2010; Spetzler et al. 2015; Nader et al. 2014; Knopman and Stieg 2014; Spetzler and Martin 1986; Feghali and Huang 2020; Flemming and Lanzino 2017; Pezeshkpour et al. 2020; van Beijnum et al. 2011; Ding et al. 2013).

The treatment options for AVMs are as follows:

- Observation;
- Embolization;
- Radio surgery;
- Microneurosurgery.

8.1. Microneurosurgical Treatment

8.1.1. The Fundamental Principles of AVM Surgery

1. Find, coagulate and divide arterial feeders;
2. Dissect the nidus of the AVM circumferentially;
3. Divide the main drainage vein or veins.

It is critical to keep the primary venous drainage open until the very end to avoid the AVM expanding and causing spontaneous bleeding.

The aim is to excise the AVM without damaging the normal parenchyma and its blood supply (Nader et al. 2014).

8.1.2. General Technique

Large craniotomy:

- To inspect the cortical vascular anatomy;
- To inspect the gyral and sulcal anatomy;
- To compare the location of feeder arteries, as well as draining vein/s with an angiogram.

Craniotomy with a margin of a few cm around the AVM.

- Be aware of trans-osseous feeders.

Just enough dural opening.
Exercise extreme caution when reflecting the dura;

- Inspect and coagulate dural feeders in large and giant AVMs;
- Careful reflection over large draining veins.

Identify as many feeders as possible on the surface;
Begin dissection to open the arachnoid adjacent to arteries and veins;
Open sulci to find feeders and relax the brain;
Follow feeders to the AVM nidus through the sulcus where they usually hide before reaching the AVM.
Differentiate feeding arteries from draining veins (veins that are larger and more delicate, and have thinner walls).
Systematically open every sulcus around the AVM to identify smaller feeders (Nader et al. 2014).

8.1.3. Surgical Outcomes

The surgical outcomes depend on the following:

- Approach with multimodal options;
- Intraoperative imaging technologies with guidance;
- Appropriate microsurgical planning;
- Rigorous training as well as expertise in microsurgery;
- Comprehensive micro-anatomical knowledge.

The majority of the scientific literature reports positive outcomes with microsurgical treatment of temporal AVMs (mortality 0–5%, morbidity 5–25%).

Complete obliteration should be 100%.

Regarding micro-neurosurgery, we need to carry out a wide craniotomy to expose the AVM generously. A frontal craniotomy, parietal craniotomy, temporal craniotomy or occipital craniotomy commonly is required according to the location of the AVM.

The initial aim is to find the feeding artery with preservation of the draining vein. We need to perform a pial incision (Figure 10) by insulin syringe or a 3 cc hypodermic syringe.

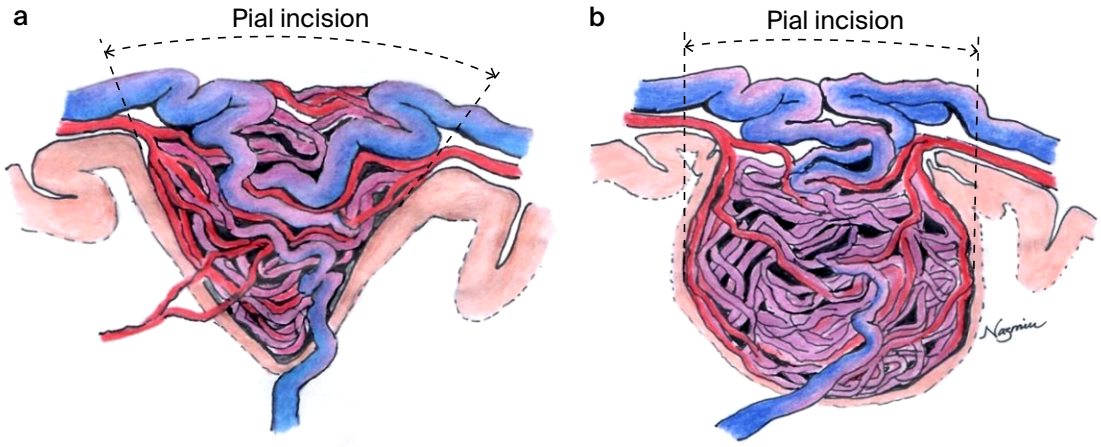

Figure 10. Pial incisions should optimize parallel exposure of AVM sides, (**a**) with tight incisions around conical AVMs and (**b**) wide incisions around spherical AVMs that optimize the visualization of deeper planes by resecting some of the overlying cortex (shaded areas). Source: Figure by authors.

Application of AVM clips or cautery of the feeding artery for proper hemostasis (Nader et al. 2014).

Circumferential dissection of the AVM nidus performed with nonstick irrigating or nonirrigating bipolar cautey. Usually, the nidus is conical in shape and the cone is directed toward the ventricle. The vessels become thinner and more fragile, so it is very difficult to carry out cautery and control bleeding.

Silver clips or AVM clips are useful in such a situation (Figures 11 and 12).

When using bipolar cautery, shrinkage of the feeding vessels is caused. Failure of proper hemostasis will cause hematoma formation as brain swelling. Thus, whenever there is brain swelling, we must check the cleavage between

the brain parenchyma and AVM nidus by removal of the cottonoid. At the end, the draining vein needs to coagulate and cauterization is performed for the resection of the AVM nidus (Nader et al. 2014). Proper hemostasis must be carried out before closure of the dura by raising the blood pressure plus/minus Valsalva mechanism.

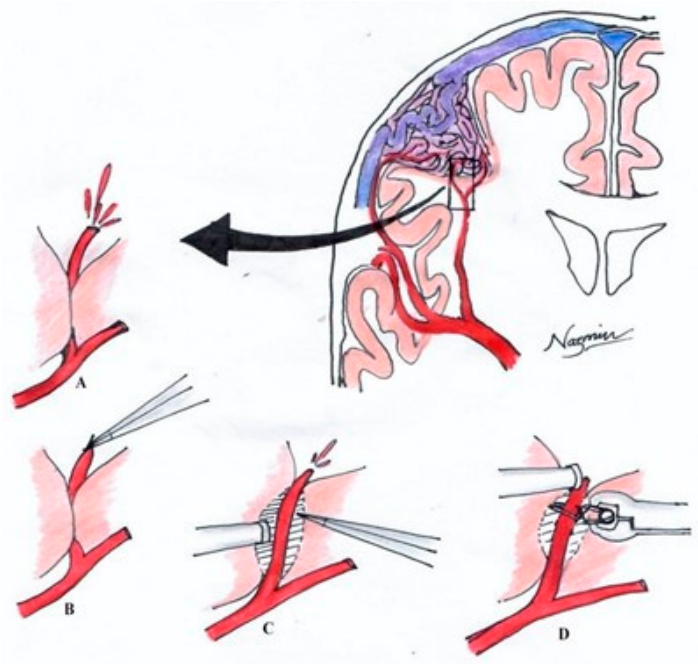

Figure 11. Controlling a deep perforating artery: (**A**) drying the area on a thin cottonoid; (**B**) attempting to coagulate it with bipolar cautery; (**C**) proximal dissection into white matter to free a segment of the artery proximal to the bleeding point, sometimes dissecting it into the sucker; and (**D**) applying the microclip. Source: Figure by authors.

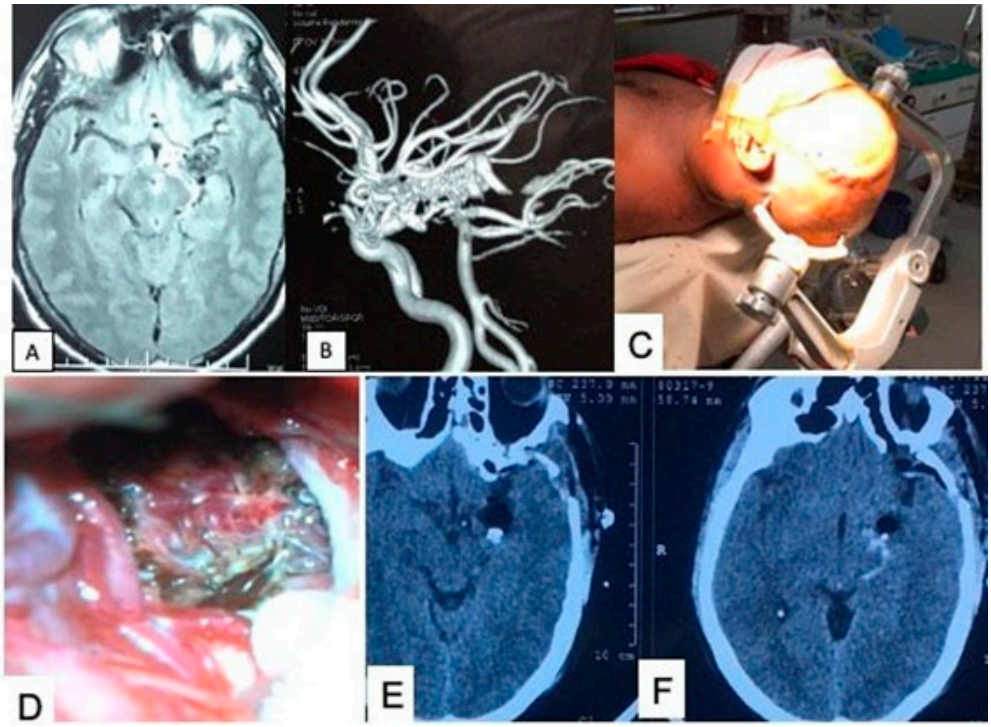

Figure 12. (**A**,**B**) MRI and MRA, respectively, showing a left mesial temporal AVM. (**C**) Patient was in a supine position, with the head end raised and tilting toward the contralateral side. (**D**) Preoperative picture showing an AVM in the mesial temporal area following the trans-Sylvian approach. (**E**,**F**) Postoperative picture showing a resection AVM from the mesial temporal area with a small amount hemorrhage in the temporal lobe. Source: Figure by authors.

8.1.4. Complications

Bleeding during surgery and brain swelling are the most common complications of an AVM resection.

An intraventricular bleed and subsequent hydrocephalus are other complications.

Seizure and impaired consciousness are not uncommon compilations.

Postoperative meningitis and late post-op hydrocephalus have to be kept in mind for some patients.

Hemiparesis or hemiplegia is another known complication of an AVM resection.

Intra-operative:

- Too wide margin of the resection;
- Parenchymal hemorrhage;
- Unrecognized intraventricular hemorrhage;
- Early occlusion of venous drainage;
- Occlusion of normal venous drainage;
- Retraction damage;
- Parenchymal damage from deep bleeding.

Postoperative:

- Hemorrhage from a residual AVM;
- Perfusion breakthrough;
- Seizures;
- Retrograde venous thrombosis;
- Retrograde arterial thrombosis;
- Vasospasm;
- Shocked brain (Nader et al. 2014).

9. Conclusions

Grade V and some grade IV AVMs should generally be treated conservatively.

Preoperative embolization should be used only to reduce the risk of the overall treatment.

Preoperative embolization should be guided by surgical considerations.

Indication for curative, palliative or pre-radiosurgery embolization is very limited.

Radiosurgery has a major role in AVM treatment and small AVMs with an unacceptable surgical risk (Spetzler et al. 2015).

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