Arteriovenous Malformations of the Posterior Fossa

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Checklist: Surgical Treatment of Posterior Fossa AVMs

Equipment needed	Procedural steps
Anesthesia • Arterial line • Mannitol • ICG if used • Blood transfusion if needed Radiology • C-Arm • Intraoperative angiography Neurosurgery • Microscope • Neuro-navigation setup • Microdissection tray • Irrigating bipolar cautery • Aneurysm clips, temporary and permanent	Preparation • Recognize angle of approach from preoperative MRI and DSA Localizing and positioning • Head position according to lesion location: prone if vermian or hemispheric location, park bench if approaching via cerebellopontine angle • Intraoperative navigation as needed AVM resection • Wide bone removal to foramen magnum to open cisterna magna and allow cerebellar relaxation • Begin with arachnoid dissection to identify any feeding artery and draining veins on the cortical surface • Circumferential dissection of AVM by separation of plane between AVM nidus and surrounding normal tissue • Cauterize feeding arteries while preserving major draining vein as last pedicle • Occlude draining vein after occlusion of all feeding arteries • Intraoperative angiography to confirm obliteration of AVM • Hemostasis

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Complication Avoidance Flowchart

Complication	Cause	Remedy	Avoidance
Post-operative hemorrhage	Residual AVM	Reoperation and/or salvage radiosurgery	Intraoperative DSA to confirm obliteration
	Inadequate Intraoperative Hemostasis	Consider reoperation	Meticulous Intraoperative Hemostasis
	Normal Perfusion Pressure Breakthrough	Consider reoperation	Assessment of hemodynamics before treatment Consider pre-operative staged embolization if high risk
Infarction	Intraoperative occlusion of en passage arteries	None	Ensure knowledge of angioarchitecture to identify non-feeding arteries

Introduction

Posterior fossa arteriovenous malformations (pAVMs) or infratentorial AVMs are a relatively rare subset comprising approximately 7–15% of all AVMs [1–3]. These AVMs draw particular attention as they are presumed to be more aggressive with respect to the natural history of hemorrhage and may sustain increased risk of treatment complications due to their proximity to critical functional structures such as the brainstem and cranial nerves [1].

The pAVMs consist mainly of cerebellar AVMs and brainstem AVMs, although there have been reports describing cerebellopontine angle (CPA) AVMs as another distinct location [4]. Despite being classified together in the posterior fossa location, the pretreatment assessment, treatment strategy, approach, postoperative care, and associated complications for AVMs vary between the cerebellar location and brainstem location and therefore need to be separately discussed. In general, AVMs in the superficial cerebellar location have a favorable risk profile and may be safely treated with surgical resection; conversely, deep cerebellar AVMs or brainstem AVMs have a significantly higher risk of postoperative complications and are preferred to be managed with radiosurgery or embolization [2].

Within the context of the two major locations (brainstem and cerebellum), pAVMs can be further categorized in detail based on the specific locations of the nidus. In a review article by Almeida et al., the authors classified pAVMs into eight distinct groups, which elucidate the anatomical relationships between each subgroup and affected cranial nerves. The classification scheme is demonstrated in Table 16.1 [2].

Table 16.1 Classification of posterior fossa arteriovenous malformations

Name	Location	Arterial supply	Cranial nerves
Midbrain	Midbrain	SCA	IV and V
Pontine	Pons	SCA/AICA/PICA	VI, VII, VIII
Medullary	Medulla	PICA	IX, X, XI, XII
Suboccipital	Suboccipital surface of the cerebellum	PICA	IX, X, XI, XII
Petrosal	Petrosal surface of the cerebellum	AICA	VII and VIII
Tentorial	Tentorial surface of the cerebellum	SCA	IV and V
Vermian	Vermis	SCA/PICA	–
Tonsillar	Cerebellar tonsils	PICA	–

Abbreviations: *SCA* superior cerebellar artery, *AICA* anterior inferior cerebellar artery, *PICA* posterior inferior cerebellar artery
Source: Almeida JP et al., Management of posterior fossa arteriovenous malformations, 2015, Surgical Neurology International

Clinical Presentation

Hemorrhage is the most common presentation in patients with pAVMs. Compared to an overall hemorrhage risk of 20–50% in all AVMs [5], numerous reports have documented an increased risk in pAVMs, with 63.9–92% of all pAVMs patients presenting with hemorrhage [1, 6, 7]. Some studies attributed the higher risk of hemorrhagic presentation to sampling bias, as pAVM patients generally present with hemorrhage because fewer other acute symptoms such as seizures occur [2, 6, 7]. Regardless of etiology, the high proportion of hemorrhagic presentation in pAVM is particularly concerning as the limited capacity for hematoma expansion due to close proximity to critical anatomical structures and associated obstructive hydrocephalus may result in rapid neurological compromise that requires immediate neurosurgical interventions. Herniation and brainstem compression due to mass effect from acute posterior fossa ICH are more frequent than supratentorial ICH and can be catastrophic (Fig. 16.1). Additionally, obstructive hydrocephalus frequently occurs in conjunction with infratentorial hematoma as a result of direct compression on the fourth ventricle or casting within the aqueduct or fourth ventricle due to concurrent intraventricular hemorrhage (IVH). Infarction may also develop from compression of posterior circulation vessels due to significantly increased intracranial pressure in the infratentorial space.

In contrast to supratentorial AVMs where seizure is generally the most frequent manifestation followed by hemorrhage, patients with pAVMs rarely present with seizure; instead, approximately 28% of all unruptured pAVMs patients present with progressive neurological deficits [7]. Cranial nerve palsies can be present due to mass effect of the nidus or from ischemic attacks due to blood steal phenomenon of high-flow AVMs. The affected cranial nerve is closely related to the location of the AVM, as shown in Table 16.1. Other non-specific symptoms such as headache, imbalance, or weakness may be present, but these are relatively rare compared to the aforementioned clinical presentations.

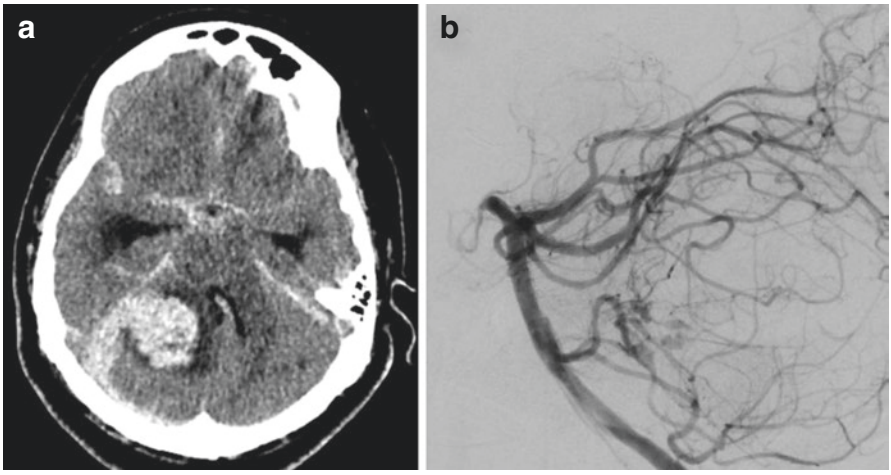

Fig. 16.1 (**a**) Axial CT of a 56-year-old woman presenting with large hemispheric ICH combined with IVH and SAH and surrounding edema causing leftward displacement of fourth ventricle and supratentorial hydrocephalus with dilated temporal horns. (**b**) Subsequent DSA demonstrated a small (1.5 cm) AVM nidus fed by AICA

Natural History of Hemorrhage and Risk Factors

Several studies examining the natural history of hemorrhage have revealed an annual hemorrhage rate of 4.7–11.6% [2, 6–8], which is significantly higher than the reported risk of hemorrhage of AVMs in the general population (2–4% per year), suggesting that pAVMs are indeed more prone to rupture than their supratentorial counterparts. Although the mechanism is unclear, some have proposed that increased frequency of deep venous drainage and venous outflow tortuosity in posterior fossa AVMs may contribute to their increased risk of rupture. Similar to AVMs in other locations, the risk of hemorrhage may be significantly altered by clinical and angioarchitectural characteristics. These factors include previous history of rupture, presence of intranidal/prenidal aneurysms, venous stenosis or ectasia, and deep location [6]. Understanding the impact of these factors is critical in the assessment of overall hemorrhagic risk of pAVMs and may play a critical role in the decision-making process for treatment selection.

Overview of Management Strategy and Outcomes

The major goal of treatment is to eliminate the risk of hemorrhage while preserving the functional status of the patient. Provided with the heterogeneity of AVMs, selection of treatment strategy should involve multidisciplinary teams, and an individualized approach is encouraged for each patient. For patients with ruptured presentation or those who are deemed to have high risk of hemorrhage, definitive treatment is

warranted. Such treatment may include a multimodality approach with single or combined therapy of surgery or radiosurgery with or without endovascular embolization. Unfavorable treatment outcomes are associated with AVMs involving critical brain structures, high Spetzler-Martin grades, or poor functional status at presentation. In these patients, if hemorrhagic risk is presumed to be low, conservative management may be considered in order to avoid harm to the patient. On the contrary, for high risk patients, if the treatment benefit outweighs the risk, definitive treatment with radiosurgery as the preferred modality may be warranted. Several studies have noted satisfactory treatment outcomes in pAVM patients with good functional outcomes at presentation, as well as in elderly patients with favorable surgical risk profile [1, 9]. It should be emphasized that in patients without absolute contraindications, the threshold to initiate treatment should be reasonably lowered in pAVMs since the consequence of hemorrhage is more likely to be catastrophic in these patients.

The presence of prenidal aneurysms complicates the treatment course, and the relative rupture risk of the aneurysm weighed against the AVM should be cautiously evaluated to determine the sequence of staged treatment. It is evident in the literature that prenidal aneurysms both portend a risk of hemorrhage on their own and increase the risk of AVM hemorrhage. Therefore, for patients presenting with subarachnoid hemorrhage (SAH) with angiographic evidence of both AVM and prenidal aneurysm, bleeding from the aneurysm should be suspected; additionally, even with isolated intracerebral hemorrhage (ICH) presentation, hemorrhage from the prenidal aneurysm should be considered before concluding that the primary source of hemorrhage is the AVM.

Accurate identification of hemorrhage source is important for optimal treatment selection. If the AVM has a higher risk of hemorrhage compared to the prenidal aneurysm, an AVM-first strategy should be adopted, and flow-related aneurysms have been frequently reported to occlude spontaneously after AVM obliteration. On the contrary, if the aneurysm heralds a higher risk, it is more likely to be treated first, and an endovascular approach is frequently utilized as it provides preoperative embolization and aneurysm obliteration in a single treatment session. Notably, some literature specifically noted a higher incidence of aneurysmal hemorrhage in posterior inferior cerebellar artery (PICA) supplied AVMs [10]. In these cases, it is necessary to secure the aneurysm before considering treatment of the AVM.

Treatment outcome is affected by a variety of factors, such as the treatment modality, location of the pAVM, and Spetzler-Martin grade of the lesion. For cerebellar AVMs, surgical treatment may achieve a 92–100% obliteration rate with 75–80% favorable outcomes. For brainstem AVMs, surgical series report a 70–80% obliteration rate with 22–25% morbidity, while radiosurgery series achieve 3-year obliteration rate of 43.8–73% with 73.3–95% of patients with improved or unchanged clinical status [11, 12]. For Spetzler-Ponce Class B or C (Spetzler-Martin grade 3–5) pAVMs, the obliteration rate is as low as 52%. Overall reported posttreatment mortality is approximately 7.7%, whereas morbidity is 16.3% [7, 13].

Procedural Overview

Microsurgical Resection for pAVMs

Preoperative Evaluation

Microsurgical resection is the most commonly adopted treatment modality in definitive treatment of pAVMs. Preoperative evaluation includes magnetic resonance imaging (MRI) and digital subtraction angiography (DSA). Among these, DSA is considered the gold standard for diagnosis and assessment of the angioarchitecture of the AVM. Superselective DSA is usually performed during evaluation to provide arterial view of the nidus, locate and characterize feeding arteries and draining veins, and identify intranidal aneurysms. This information is essential for developing a strategic operative approach to the AVM and to determine whether preoperative embolization is warranted. External carotid injection should also be considered as on rare occasions the pAVMs may be concomitantly supplied by external carotid vessels. Both computed tomography angiography (CTA) and MRA are regarded as secondary options in addition to the DSA study and are likely to have been obtained prior to the DSA. Three-dimensional cone-beam CT (3D-CBCT) provides enhanced-contrasted thin-slice images with simultaneous imaging of both arterial and venous phases, which renders it an excellent adjunct to DSA and is helpful for radiosurgery planning. Brain MRI is essential for localization, defining nidus relationships to ventricles and pial surfaces, and is highly sensitive and specific for detection of concomitant silent brain hemorrhage, which factors into decision-making regarding risk of treatment. The advantage of supplementing DSA with MRI is the capability to assess anatomical relationships between the nidus and critical functional structures.

Intraoperative Adjuncts

Adjuncts include intraoperative monitoring of brainstem somatosensory-evoked potentials (SSEP) and brainstem auditory-evoked potentials (BAEP). Electromyography (EMG) and direct stimulation may be used to determine safe territories of intraoperative manipulation. The use of neuronavigation may be useful during the early phases of the approach, such as for the craniotomy to determine venous sinus location, and is less helpful in pAVMs given sufficient anatomical landmarks and brain shift from CSF drainage, but may be used to determine the borders of deep niduses in the cerebellum. Intraoperative or postoperative DSA is strongly recommended to confirm obliteration of the AVM after microsurgical resection.

Microsurgical Techniques

A wide craniotomy in treatment of pAVMs facilitates sufficient exposure of the nidus and increases working distances for navigating through surgical corridors. The choice of approach is dependent on the location of the nidus. For lesions involving midline and paramedian structures including vermian or tonsillar cerebellum, a posterior midline approach from the occiput to C1 is commonly adopted. For posterolateral lesions such as cerebellar hemispheric or lateral pontine AVMs, access

can be achieved through an extended retrosigmoid approach. Retrosigmoid craniectomy with or without far-lateral extension can be used for CPA AVMs or medullary AVMs, and a supracerebellar approach can be used for superficial cerebellar AVMs abutting the tentorium and for posterior midbrain AVMs. All posterior approaches should be particularly aware of the location of transverse and sigmoid sinuses to avoid incidental injury. In all cases, opening of the foramen magnum to allow easy access to opening the arachnoid of the cisterna magna for cerebellar relaxation is recommended.

The resection of the nidus follows general principles for AVM resection in other locations. Adequate exposure, meticulous microdissection, rigorous hemostasis, circumferential approach, and protection of major venous drainage before nidus extirpation are critical for complete obliteration while minimizing intraoperative complications. Fixed retraction should be avoided if possible to prevent extensive stretching of cranial nerves. Preemptive packing of the large subdural spaces with Gelfoam sponges to contain the dissemination of blood into the posterior fossa and cervical subdural spaces as the resection proceeds decreases the risk of subsequent hydrocephalus. Intraoperative identification of feeding arteries and draining veins through careful dissection is the first step and key to avoiding premature interruption of venous drainage. Superficial, arterialized, dilated veins overlying the plane of approach may be commonly seen and should be cautiously dissociated from the arachnoid and gently mobilized away from the focus of resection. Small, temporary clips may be applied on suspicious arteries that are potentially feeding the nidus, which are useful for recognizing and preventing injury to *en passage* vessels. Once identified, feeding arteries should be skeletonized with sharp dissection and occluded definitively with coagulation. Of note, the response to coagulation on perforators arising from perforating feeding arteries may be suboptimal given the lack of smooth muscle cell layer, and application of permanent aneurysm clip is warranted for definitive occlusion; nevertheless, hemostasis during pAVM resection should be compulsively pursued given limited tolerance for hematoma extension in the infratentorial space. A combination of bipolar coagulation, pressure, and aneurysm clips may be used to achieve satisfactory hemostasis. Separation of the nidus from the parenchyma should follow a circumferential pattern with spiral progression, creating perinidal corridors to allow mobilization of the nidus while occluding the feeders. The margin of nidus can be established through distinction between normal parenchyma and gliotic tissues or hemosiderin stains from previous hemorrhages. The nidus can be safely removed after confirmation of occlusion of all feeding arteries followed by the last step of disconnecting all major draining veins.

Radiosurgery for pAVM

Radiosurgery is the preferred definitive treatment for nonsurgical pAVMs. Overall obliteration for pAVMs in radiosurgery series reaches 75–80% at the 5-year mark. However, given the persistent risk of hemorrhage of 1–3.6% during the latency interval between treatment and obliteration [8, 14], it is generally considered as the

secondary option for patients with acceptable surgical risks. Before radiation delivery, high-resolution axial MRI images of the AVM and Dyna-CT are first registered to the treatment planning system to allow contouring of the target. Treatment margins should be cautiously determined and conformal for avoidance of irradiating critical structures. The development of an optimal treatment plan involves multidisciplinary efforts from both neurosurgery and radiation oncology. Treatment of pAVMs is usually achieved by stereotactic radiation using Gamma Knife or CyberKnife in a single session with a median dose at approximately 18–21 Gy. The utility for pre-radiosurgery embolization for AVMs is controversial since multiple studies have suggested decreased obliteration rate after radiosurgery for embolized patients.

Endovascular Embolization for pAVM

Endovascular embolization is conventionally regarded as a non-definitive adjunct therapy to surgery and radiosurgery. It is particularly useful in obliterating high-flow or deep-seated parts of the AVM to avoid devastating intraoperative hemorrhage from these compartments. Additionally, for AVMs with associated prenidal aneurysms, both aneurysm and AVM can be managed in a single embolization session before surgical management. Embolization agent includes *N*-butyl cyanoacrylate (NBCA) and ethylene-vinyl alcohol copolymer (Onyx). Under rare circumstances, coils may be used to slow intra-arterial flow. Curative embolization of pAVMs remains controversial to date due to low obliteration rate and high risk of recurrent hemorrhage and AVM recanalization. Therefore, application of embolization should be concordant with primary treatment goals of preventing future hemorrhage and preserving functional status, so that aggressive pursuit of eliminating all pedicles in patients with favorable surgical or radiosurgical risk is currently not recommended.

Complication Avoidance

Hemorrhage is the most feared postoperative complication. Progressive hemorrhage within the infratentorial space quickly exceeds the capacity for hematoma expansion and may result in poor clinical course from rapid brainstem compression and catastrophic herniation. Hydrocephalus may also occur in conjunction with IVH as a consequence of obstructive intraventricular casting at the fourth ventricle or aqueduct. In surgical patients, the source of hemorrhage is most likely from residual lesion but can also originate from inadequate intraoperative hemostasis or bleeding from “normal perfusion pressure breakthrough” (NPPB) after large AVM resection. Prevention of postoperative hemorrhage therefore requires addressing all potential bleeding mechanisms. Residual AVM may result from incomplete resection of the AVM or simply from intraoperative judgment that part of the AVM cannot be safely resected. In both cases, thorough preoperative DSA evaluation of the AVM

angioarchitecture and proximity to surrounding critical structures prepares the operator and in turn significantly lowers the likelihood of incomplete resection. Intraoperative DSA provides a mechanism for confirming complete resection and remains the optimal modality for intraoperative detection of residual AVMs. However, if intraoperative angiography is not possible due to patient position, an immediate postoperative angiogram remains essential, with return to the OR for resection of residual if feasible. For hemorrhage of other causes, rigorous hemostasis should be pursued, and staged resection or presurgical embolization should be considered for large AVMs to prevent breakthrough bleeding. A brief post-resection moderate hypertensive challenge may be considered to test hemostasis. Strict blood pressure control is recommended postoperatively to prevent bleeding from the resection bed.

Postoperative ischemic events may also occur from intraoperative injury of *en passage* arteries; it is therefore critical to identify these arteries intraoperatively and avoid incidental occlusion. Prevention of vasospasm-induced ischemia can be achieved through aggressive medical intervention under vasospasm protocol in ruptured AVM patients. Prolonged fixed retraction should be avoided to lower the chances of cranial nerve injury.

Complication Management

Postoperative hemorrhage causing neurological deterioration or at risk for doing so requires rapid hematoma evacuation, and decompressive suboccipital craniectomy may also be warranted. If the source of bleeding is suspected as residual AVM, revised resection can be attempted during hematoma evacuation. Cautious scrutiny of the resection bed for bleeding source and aggressive hemostasis are crucial for avoidance of recurrent bleeding. If combined with IVH and suspected concurrent hydrocephalus, the threshold for insertion of an intraventricular catheter should be low. For residual AVM that is determined to be high risk for further resection, postsurgical radiosurgery should be considered for complete obliteration.

Conclusion

Posterior fossa AVMs represent a small cohort of the AVM population and are challenging to manage due to their proximity to critical brain structures, deep locations, and potentially aggressive clinical courses. Additionally, due to limited space to accommodate hemorrhage in the infratentorial compartment, hemorrhagic complications for pAVMs are generally less well tolerated compared to their supratentorial counterparts. Successful management of pAVMs therefore requires appreciation of the natural history of the disease, thorough evaluation of the angioarchitecture of the pAVM and relationship to critical posterior fossa anatomical structures, and optimal balancing of the risks and benefits associated with each treatment modality. A multidisciplinary team including neurosurgeons, neuroendovascular surgeons, and radiation oncologists should participate in

shared treatment decision-making. Longitudinal patient management may require physiatrists and neurologists. Surgical resection is considered the optimal treatment modality for patients with acceptable surgical risk profile, and postoperative hemorrhage may be avoided with meticulous surgical techniques, compulsive hemostasis, and intraoperative DSA for confirmation of AVM obliteration. Presurgical embolization is useful in selected cases for reducing the number of arterial feeding vessels and lesion size to optimize surgical outcome. Stereotactic radiosurgery may also achieve satisfactory occlusion rates in patients deemed to be non-operative. Finally, a subset of pAVM patients may ultimately fare better without intervention. The potential complications associated with each possible treatment should be anticipated before initiation of the management plan so that every effort is made to avoid such complications.

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