Arteriovenous Malformation

Last Updated: October 1, 2018

Overview

Arteriovenous malformations (AVMs) are vascular lesions characterized by high flow pathologic shunting between the arterial and venous circulations with an intervening nidus of dysplastic vascular channels but no capillary bed. AVMs vary in degree of arterial supply, nidal size and location, flow-related aneurysms, and venous outflow. Lesions are typically within the subpial space supplied by the cerebral arteries, though AVMs often parasitize dural branches.

Changes in the adjacent or underlying parenchyma identified on MRI and have been hypothesized to result from vascular steal phenomenon and venous congestion. Clinical symptomatology is variable and generally related to intracranial hemorrhage. Occasionally, AVMs are discovered incidentally. Brain AVM, pial AVM, cerebral AVM, and non-Galenic CAVM are synonymous.

Imaging

- General features
  - Subpial space nidus, distinct from dural and subarachnoid AV shunts
  - Supplied predominantly by pial arteries, may
parasitize dural branches

- AVMs are solitary and sporadic lesions in the vast majority of cases (98%)
- Location: supratentorial (~ 85%), posterior fossa (~ 15%)
- Multiple AVMs are rare, usually syndromic
- Cerebral (Cerebrofacial) Arteriovenous Metameric Syndrome (CAMS)
  - Segmental craniofacial AVMs, often very complex lesions

- **CT**
  - **NECT**
    - Parenchymal hematoma and/or IVH (SAH only occasionally to rarely)
    - Isodense to hyperdense serpiginous structures representing the nidus
    - Portions may be calcified
    - Mass effect by AVM directly if very large, or hematoma if present
    - Small AVMs that have not hemorrhaged are likely to be over-looked
    - Status post embolization, liquid embolics appear very hyperdense
    - If there is evidence of rupture and hemorrhage, proceed to contrast-enhanced
study

- **CT Angiography**
  - MIP and 3D reformats useful in characterization
  - Depicts hypertrophied arteries and draining veins
  - Allows good approximation of distribution of arterial supply
  - Strongly enhancing nidus
  - High flow shunts result in early draining veins and contrast-opacified dural sinuses demonstrated on arterial phase

- **Limitations**
  - Beam hardening artifact
  - Bone (Skull base and posterior fossa)
  - Metallic artifact
  - High attenuation of endovascular coils and liquid embolics can obscure residual nidus
  - 3D reconstruction and surface shading software may produce artifacts
  - Limited or judicious use in patients with iodine-based allergy and/or renal dysfunction
  - May require contrast dose-adjustment considerations for subsequent catheter angiography
Figure 1: Typical NECT (top row left) appearance of an AVM with relatively ill-defined isodense to hyperdense vessels replacing brain parenchyma with evidence of small adjacent parenchymal hemorrhage in the right temporal lobe. CTA (top row right, bottom row) increases the conspicuity of the underlying AVM, revealing abnormal right temporal nidiform vessels.
• **MRI**
  - **Findings**
    - Excellent sensitivity and specificity for detection and relative staging of blood products
    - Tangle of serpiginous flow voids
    - Adjacent parenchyma likely to demonstrate gliosis and/or venous congestion
  - **Typical pulse sequences:**
    - **T1**
      - Signal varies with flow rate, direction, and stage of blood products from hemorrhages
      - Tangle of serpiginous black flow voids representing the nidus
    - **T2 and FLAIR**
      - Tangle of serpiginous black flow voids representing the nidus
      - Variable T2 hyperintensity and volume loss representing gliotic change in the underlying/adjacent parenchyma
      - Variable T2 hyperintensity and expansion representing venous congestion edema in the adjacent parenchyma, typically in the territory of draining veins and dural
sinuses

- T2* GRE and SWI
  - Susceptibility artifact (blooming artifact) representing hemorrhage if present

- T1WI C+
  - Enhancement of nidus
  - Rapid flow may demonstrate signal loss without enhancement (vascular flow voids) on traditional spin echo T1 postcontrast images
  - Can greatly increase the characterization of the enhancing vessels of an AVM when IRSPGR 3-D sequence is obtained

- MR Angiography:
  - Technique
    - 3D time-of-flight (TOF)
      - Spatial resolution of modern 3D TOF MRA on the order of 1 mm3
    - Axial source images
    - Maximum intensity projection (MIP) images derived from source data
  - Contrast-Enhanced MRA
    - Useful in surveillance, staged treatment follow-up
  - Metal suppression techniques can be employed
- MRA should generally be considered complementary to conventional MRI
- Best to include CE-MRA as well as GRE or SWI sequences
- Feeding arteries are often easy to identify (because of location and dilatation)
- Draining veins can be identified by their larger caliber relative to arteries and drainage into deep or cortical veins

- **Limitations**
  - Prone to multitude of artifacts that can be both helpful and difficult to interpret
  - Pulsation artifacts may be seen with AVMs
  - Pulsation artifacts become more pronounced on contrast-enhanced images
  - Generally less cost-effective, less availability
  - CT is more easily performed in uncooperative, combative patients

- **MRA Pitfalls**
  - Intrinsic T1 shortening, such as in subacute hemorrhage, may simulate vascular flow on TOF MRA
  - Subtle early draining veins may be the only finding in otherwise occult, largely thrombosed AVMs
Figure 2: T1 (top row left), FLAIR (top row right), and SWI (bottom row) axial MR images illustrate mixed stage blood products of subacute parenchymal hematoma within the superior right cerebellar hemisphere with associated subarachnoid hemorrhage within the adjacent cerebellar sulci and crossing midline. Nidiform flow voids are present at the medial aspect of the hematoma on the FLAIR image consistent with underlying cerebellar AVM.
Figure 3: Top row: Axial T2 (top row left) and axial T1 (top row middle) reveal large dilated serpiginous and clustered nidiform flow voids centered on the central sulcus at the right frontoparietal junction. Axial T1 C+ (top row right) demonstrates robust postcontrast enhancement in a pattern consistent with high flow AVM. Bottom row: Sagittal T2 (bottom row left) and Coronal T1 C+ (bottom row right) again demonstrate the right frontoparietal AVM centered on the central sulcus involving eloquent cortex (Spetzler-Martin grade 4).

- Digital Subtraction Angiography
  - Findings
- Best characterization and delineation of AVM angioarchitecture

- **Location/Depth**
  - Hemispheric, lobar, basal ganglia, thalamus, callosal, intraventricular, cerebellar hemispheric, cerebellar vermis, brain stem, perimesencephalic

- **Nidus size and compactness**

- **Flow physiology**
  - Predominant nidal flow or
  - Fistula (shunt) predominant or
  - Mixed

- **Arterial supply**
  - Organized by major feeding territory
  - 25-33% may have pial and dural arterial supply

- **Associated aneurysms**
  - Number and location
  - Nidal (>50% of cases)
  - Flow-related (of the feeding arteries, 10-15% of cases)
  - Proximal aneurysms at the level of the circle of Willis branch points
  - Venous

- **Degree of pial collaterals and Moyamoya changes**
Venous characteristics

- Early draining veins ± venous stenoses due to high-flow venopathy (may increase intracranial hemorrhage (ICH) risk)
- Number of draining veins
- Superficial versus deep drainage
- Presence of venous angiopathy such as hypertrophy, outflow stenosis

Eloquence

- Several regions of the brain defined by Spetzler and Martin that, when damaged, result in more debilitating deficits

- Staging, Grading, & Classification
  - Spetzler-Martin scale, derived by DSA findings
    - Global assessment and estimated surgical risk, from 1-5

- Size
  - Small (< 3 cm) = 1
  - Medium (3-6 cm) = 2
  - Large (> 6 cm) = 3

- Location
  - Noneloquent area = 0
  - Eloquent brain = 1
- Eloquent areas: sensorimotor cortex, visual cortex, hypothalamus, thalamus, internal capsule, brainstem, cerebellar peduncles, deep nuclei

- **Venous drainage**
  - Superficial only = 0
  - Deep = 1

Figure 4: 16-year-old male with cerebellar AVM. Top row: Axial T2-weighted image (top row left) and TOF MRA images (top row middle and right) reveal an AVM with parenchymal nidus within the right superior cerebellum and large varix within the quadrigeminal plate cistern. Bottom row: DSA images obtained from arterial to venous phase at high frame
(left to right) demonstrate the angioarchitecture of the superior cerebellar AVM with direct supply by the bilateral PCA and SCA as well as en passant supply by the right AICA. There is early opacification of the internal cerebral veins, vein of Galen, and straight sinus.

Figure 5: 6-year-old female with seizure. Top row – Axial NECT (left image) demonstrates an ill-defined hyperdensity at the left mid-frontal convexity. Postcontrast CTA axial (middle images) and sagittal MIP (right image) reveal an abnormal nidiform cluster of vessels and two dilated varices in close proximity. Bottom row – Precontrast coronal T2 MRI (left image) and postcontrast T1 MRI (center image) place the lesion in the left pre-rolandic region; there is characteristic
flow-related signal loss within the larger varix on T2 as well as numerous dilated flow voids surrounding the nidus. Dynamic CE-MRA image demonstrates the spatial relationships of the left pre-rolandic AVM and varices; enlarged cortical veins are identified emerging superiorly toward the superior sagittal sinus and inferolaterally toward the superficial sylvian territory and vein of Labbé.
Figure 6: Same 6-year-old female with seizure. Left internal carotid arteriography, left anterior oblique stereoscopic images (stereo image pair obtained at slightly different projection angles, viewed by crossing eyes) and lateral projection image (bottom left) illustrates the complexity of the left pre-rolandic AVM with direct supply from the prefrontal division of the left middle cerebral artery and en passant supply from the precentral division and venous
drainage via two main conduits, each associated with dilated venous varix.

Left internal carotid arteriography lateral projection (bottom right image) status post endovascular embolization (Onyx) and surgical resection demonstrates no residual arteriovenous shunting or nidiform vessels.

For more information, please see the corresponding chapter in Radiopaedia, and the Arteriovenous Malformation chapter within the Brain Tumor Msimics sub-volume within the Neurosurgical Atlas.

Contributor: Daniel Murph, MD

DOI: https://doi.org/10.18791/nsatlas.v2.03.03.04.02

References


Kim H, Pourmohamad T, Westbroek EM, McCulloch CE,


Submit Your Complex Case to be reviewed by the Atlas team.
Dr. Aaron Cohen-Gadol is donating part of royalties from the COHEN™ Bipolar Forceps to the Neurosurgery Research & Education Foundation (NREF).