AMSER Rad Path Case of the Month:

29 year old male with black out spells

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AT THE FOREFRONT UChicago Medicine

AMSER

29-year-old male presents with “black-out spells.”

- Episodes began several months prior “out of the blue,” now occurring at least 3 times per week.
- Described as unresponsive “blank stare forward” associated with “crying, lip smacking, finger rolling, and tongue movements.”
- Endorses tongue biting and urinary incontinence associated with episodes observed to last 3-5 minutes, along with headaches afterwards.
- Denies aura and triggers; does not remember the episodes.

History

- Medical: HIV (diagnosed in 2021), remote TBI with a brick
- Surgical: None
- Meds: Bictegravir-emtricitabine-tenofovir-alafenamide
- Social: Active cigarette (3 pack-years) and marijuana use (6 blunts daily), social alcohol use
- Family: No family history of stroke or aneurysms
- Pertinent (+): Retro-orbital headaches
- Pertinent (–): Dizziness, tremors, syncope, facial asymmetry, speech difficulty, weakness, paresthesia
- Physical Exam: Unremarkable

Initial Labs, Workup, and Management

- CBC: **WBC 3.7 (↓), Hgb 13.2 (↓)**, PLT 273
- Coagulation Studies: **PTT 37.7 (↑)**, INR 1.0
- Lupus anticoagulant: **Positive**
- EEG: No epileptiform or lateralizing activity, no active seizures recorded
- Medical Therapy: Levetiracetam 500 mg BID provided inadequate control, subsequently added with Oxcarbazepine 300 mg BID
- Next Step: MRI with/without contrast

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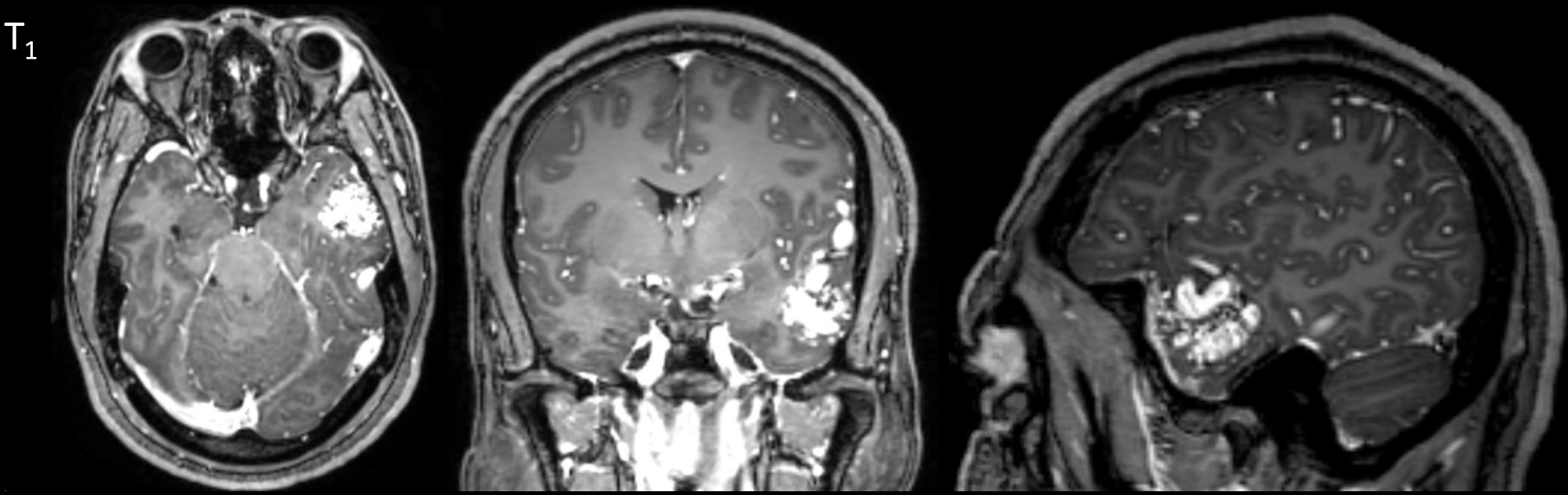
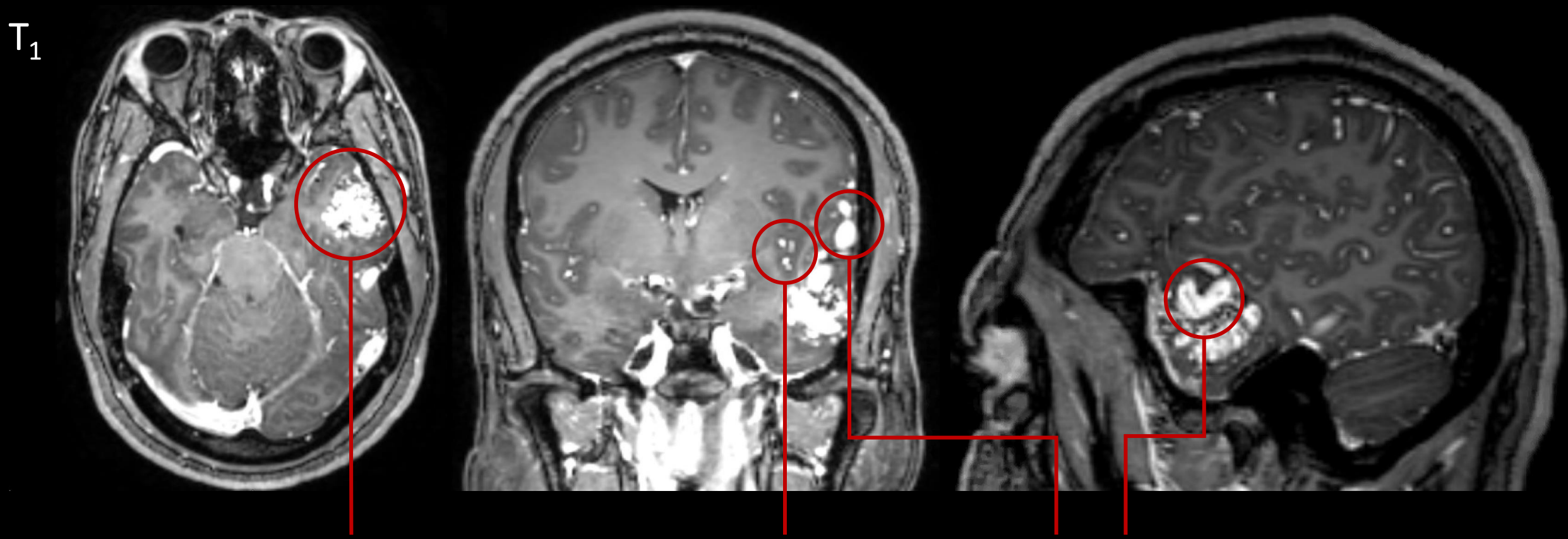


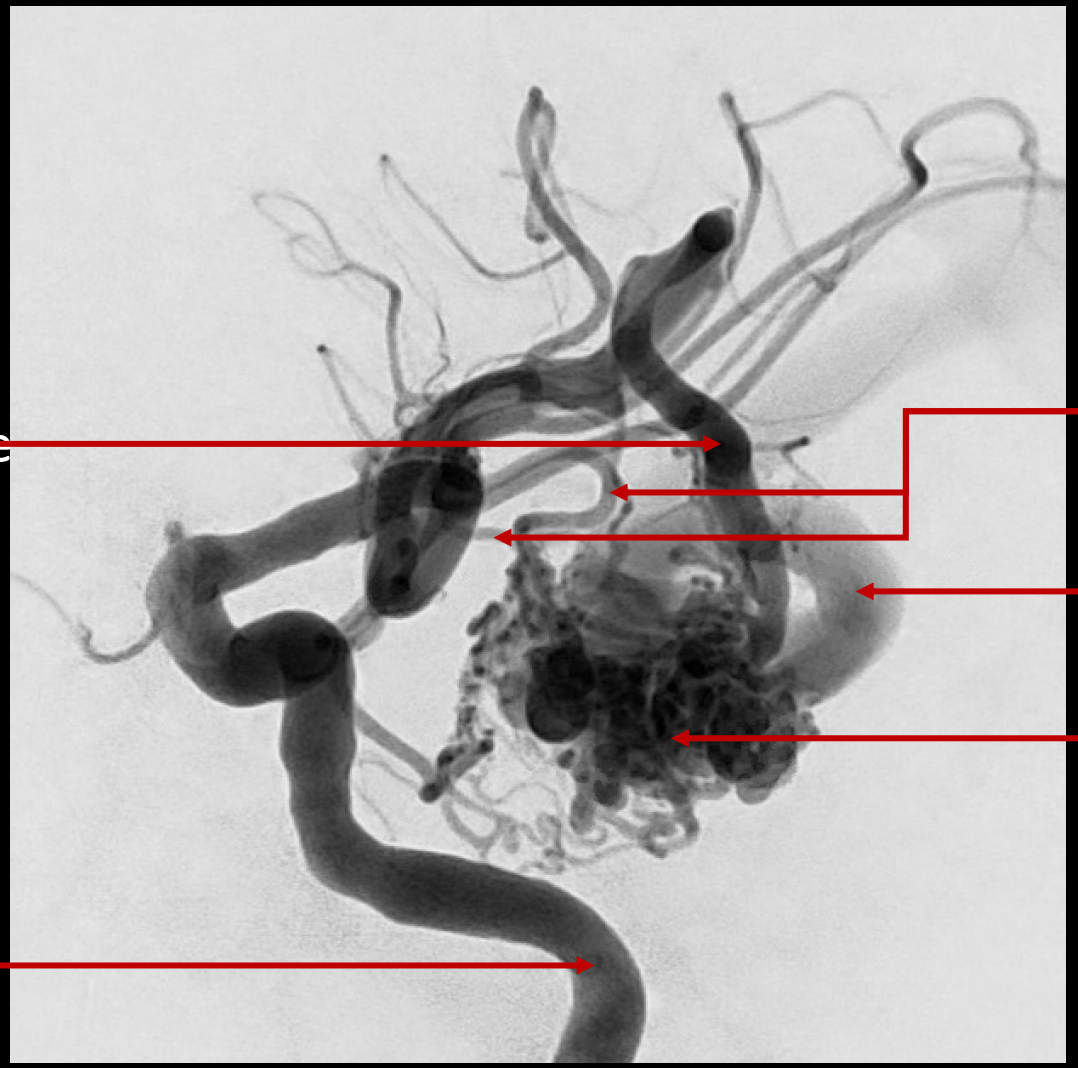
MRI wwo revealed possible left temporal arteriovenous malformation (AVM)



Differential Diagnoses

- AVM
- Cerebral cavernous malformation (CCM)
- Capillary telangiectasias
- Dural AV fistula
- Pial AV fistula

Diagnostic cerebral angiogram confirmed left temporal AVM



Dominant arterial feeding branch from inferior division of left middle cerebral artery (L MCA)

Multiple arterial feeding branches from L MCA

Cortical venous drainage

Nidus with maximum diameter 3.5 cm

Left internal carotid artery

Assessment and Plan

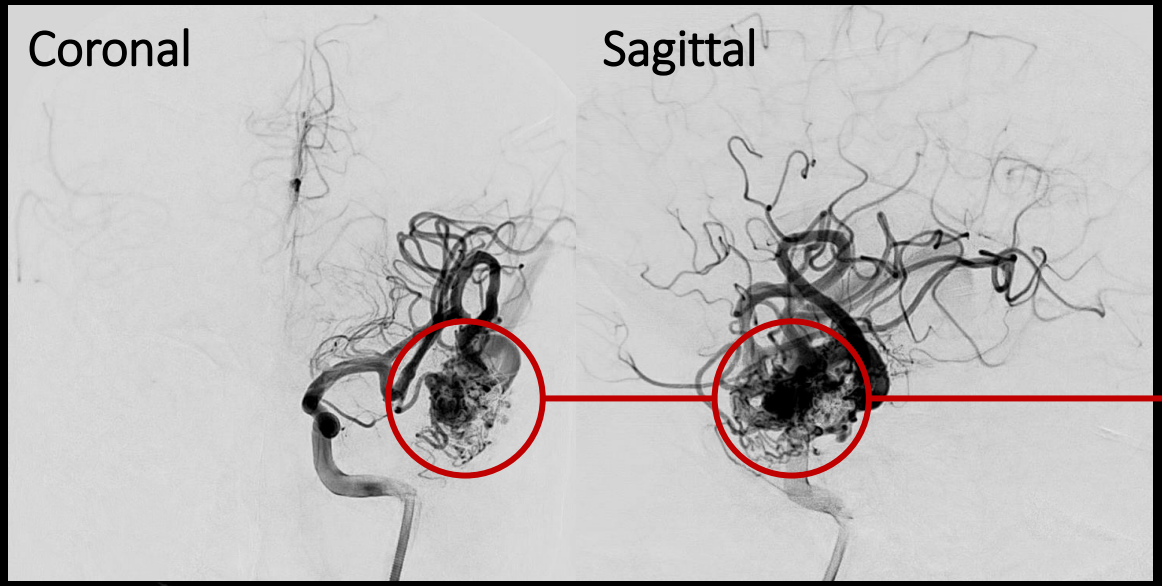
- 29-year-old male with focal impaired awareness epileptic seizures in setting of left temporal Spetzler-Martin Grade III AVM
    - Medium size (3.5cm) → 2 points
    - Adjacent to eloquent region (Broca's area) → 1 point
    - Superficial cortical venous drainage → 0 points

- Planned Intervention:
    - Two-stage embolization
    - Neurosurgical resection

- Resection favored over stereotactic radiosurgery due to nidus size and risk of radiation complications exacerbating poorly-controlled seizures

Angiographic and 3D reconstruction images demonstrated stage 1 embolization

Cerebral angiogram

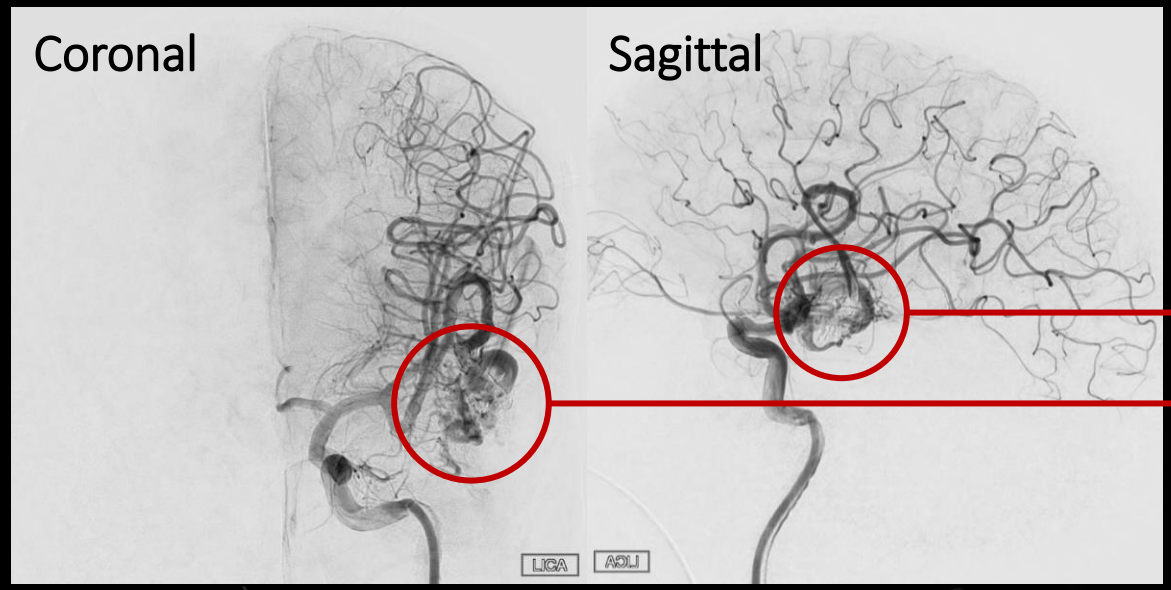


Partial embolization
(50% of nidus)

3D reconstruction

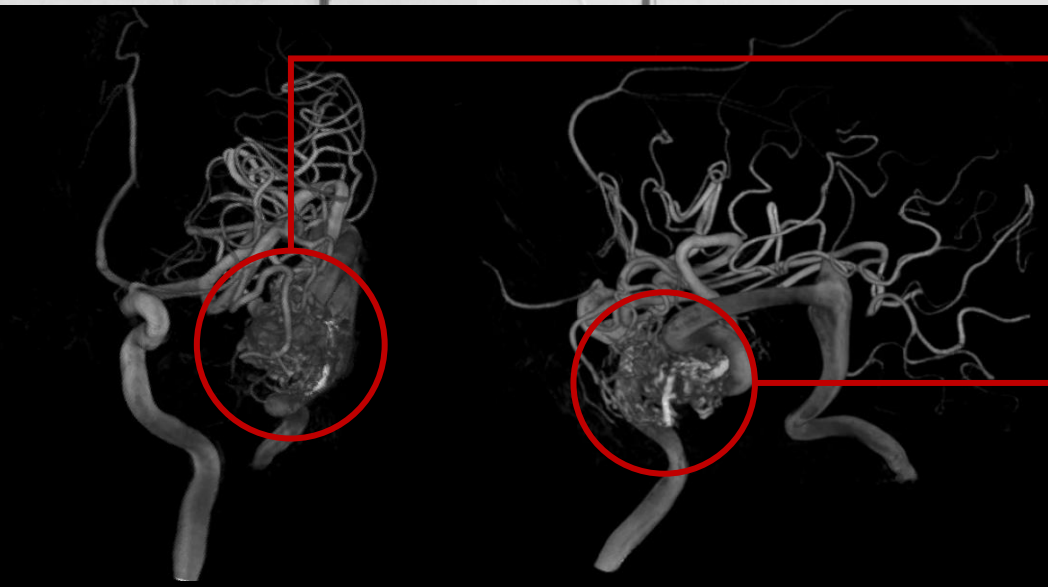
Angiographic and 3D reconstruction images demonstrated stage 2 embolization

Cerebral angiogram



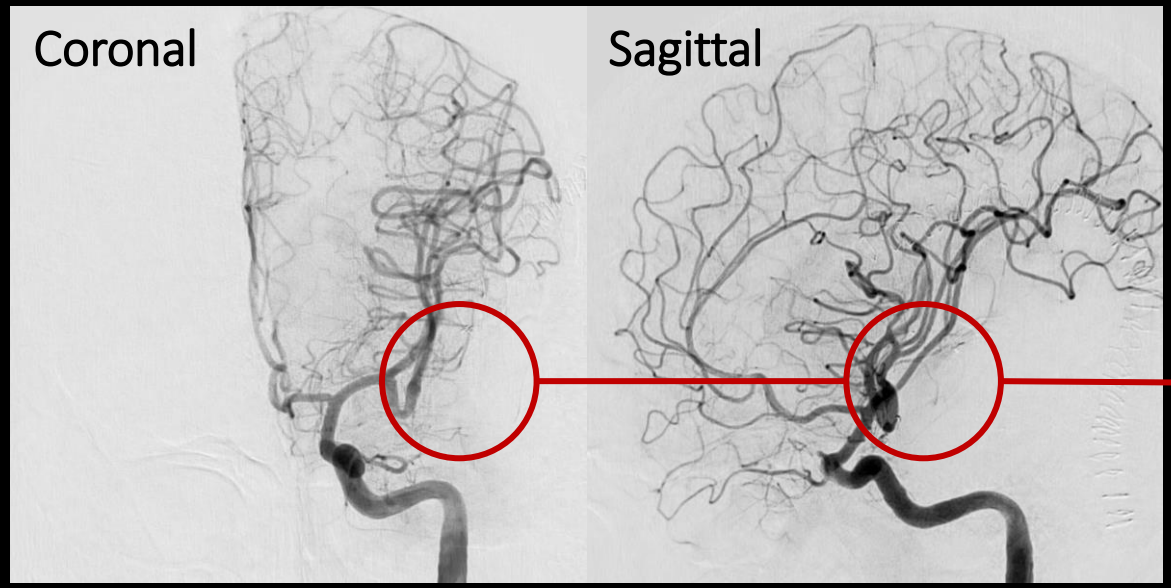
Complete embolization

3D reconstruction



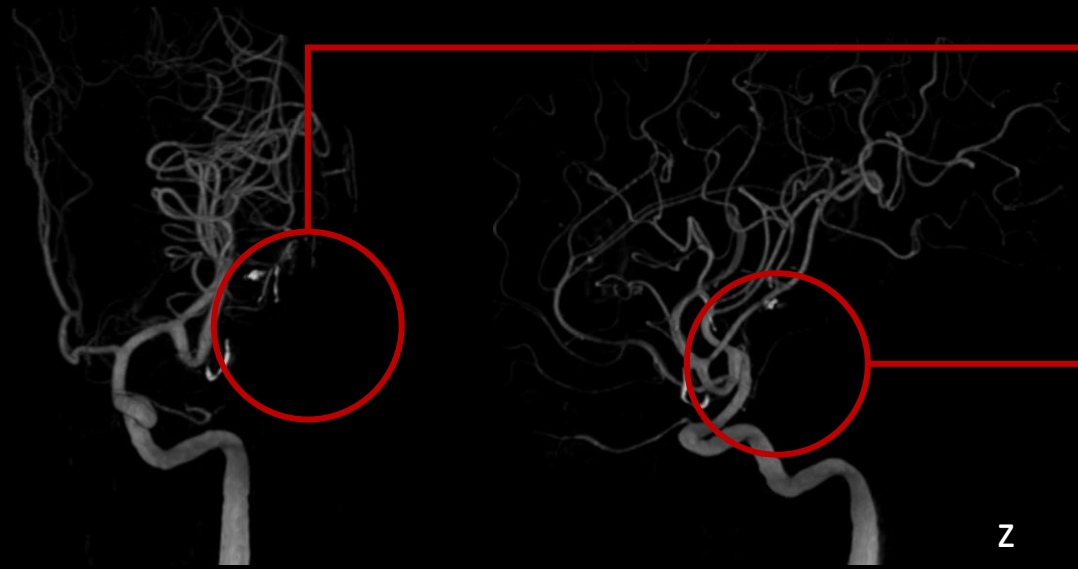
Angiographic and 3D reconstruction images demonstrated complete resection of AVM

Cerebral angiogram



Absence of flow through nidus shows successful resection

3D reconstruction

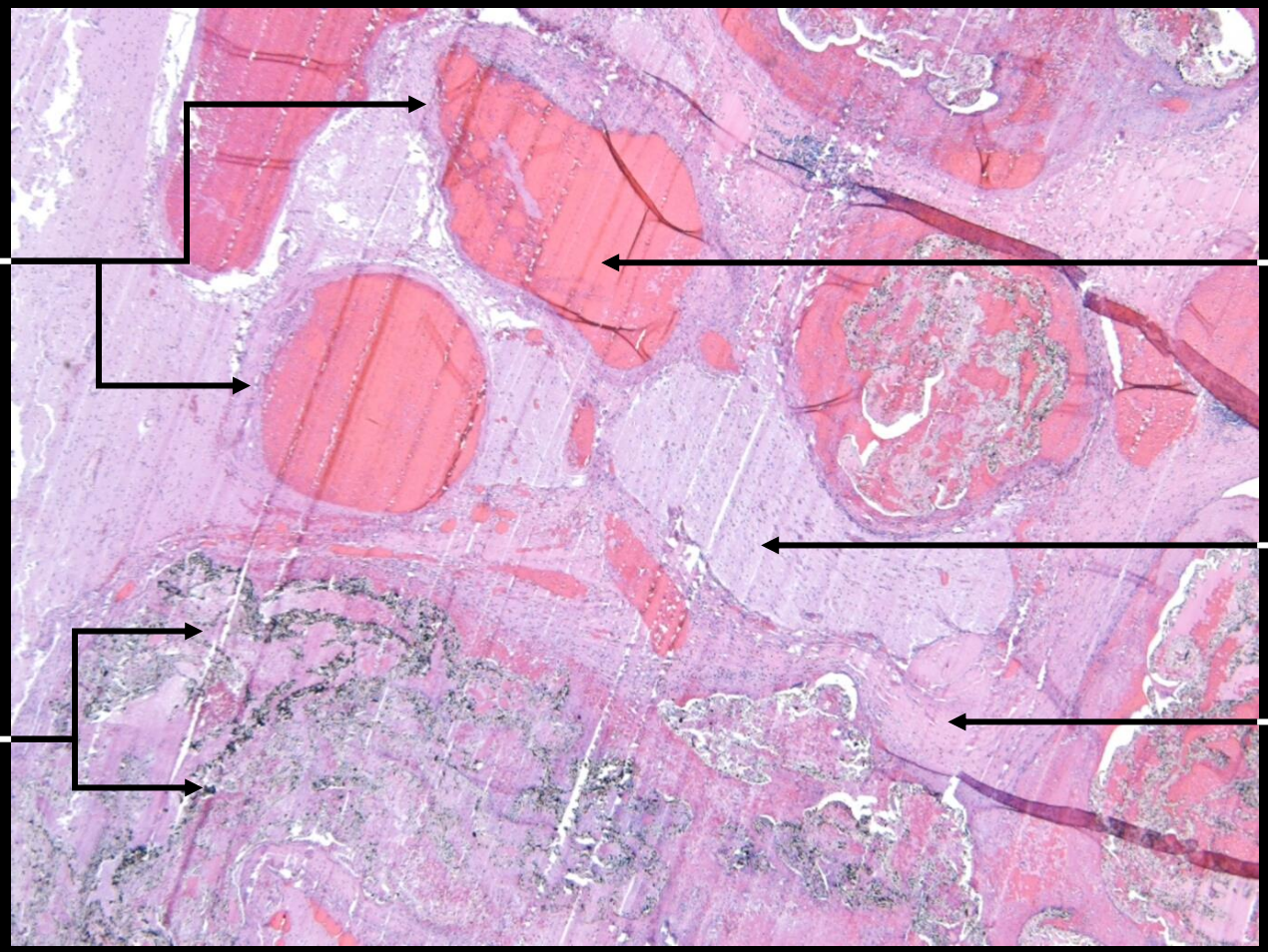


2-week follow-up visit and CT demonstrated seizure-free recovery without residual lesion

Hyperdensities consistent with prior embolization

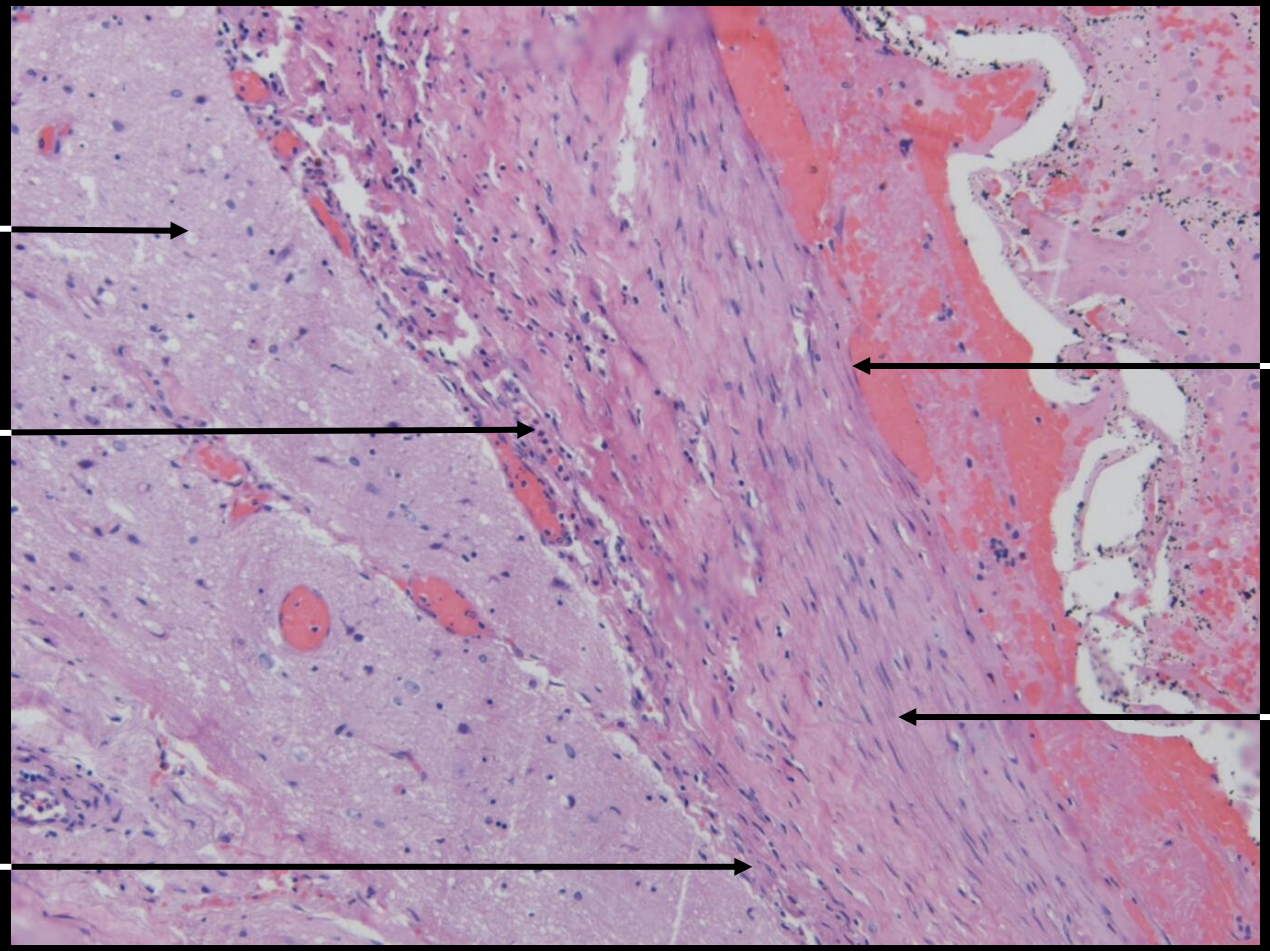
Slightly hypoattenuated areas consistent with resolving postoperative gliosis and/or vasogenic edema

Low-power pathology of AVM demonstrates tangle of lesional arteries and arterialized veins

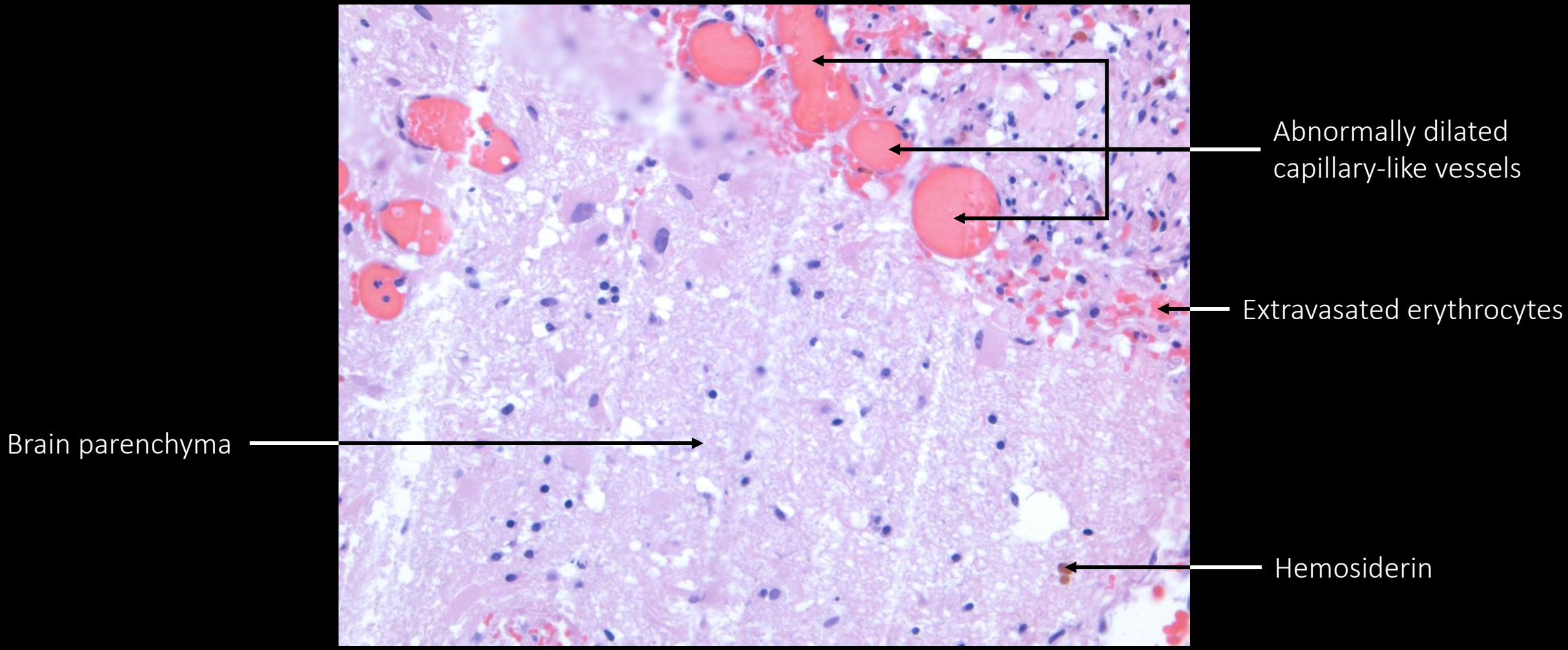


Striated cut pattern due to microtome blade resistance against embolic solution

Medium-power pathology of AVM demonstrates abnormal layers within arterialized vein



High-power pathology of AVM demonstrates abnormally dilated capillary-like vessels

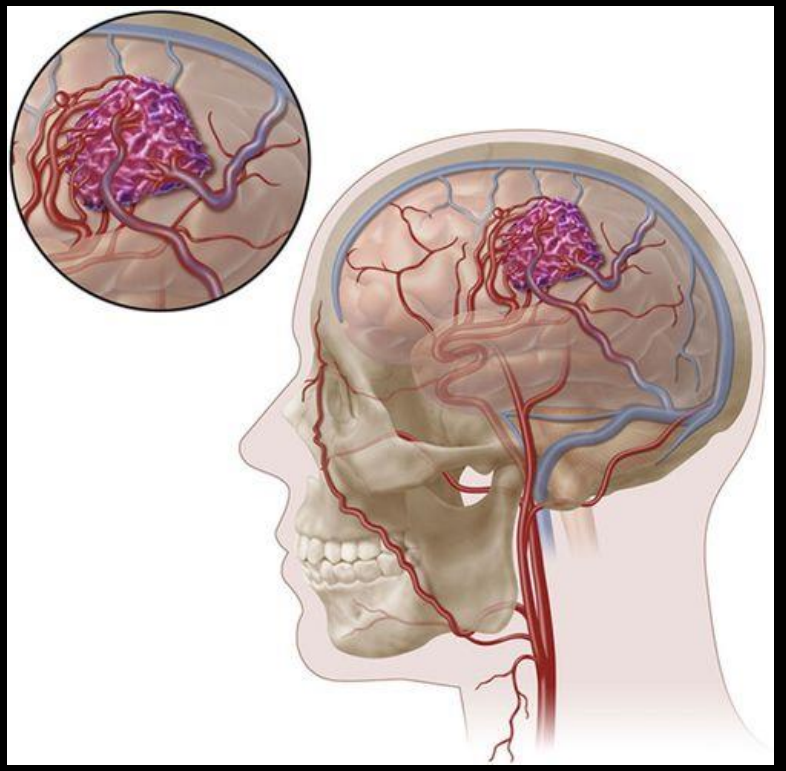


Final Diagnosis:

Left Temporal AVM

Brain AVMs are complex tangles of abnormal arteries directly connected to veins without true capillaries

- Epidemiology of brain AVMs:
  - Prevalence: 10-18 cases per 100,000 people
  - Incidence: 1 case per 100,000 person-years
- High-flow states within abnormal vessels predispose brain AVMs to rupture & hemorrhage:
  - Risk of hemorrhage: 2-4% per year
  - Overall mortality: 0.7-2.9% per year
- Leading cause of intracranial hemorrhage in children & young adults
- 38-68% of AVM patients present with first episode of hemorrhage



Rutledge *et al*., *Transl Stroke Res* (2014);
Abecassis et al., *Neurosurg Focus* (2014);
Artist's rendition of AVM from Derdeyn et al., *Stroke* (2017)

Clinical presentation of brain AVMs vary from incidental finding to devastating intracranial hemorrhage

- Presentations of brain AVMs include:
    - Intracranial hemorrhage (50% of cases)
    - Seizures (30%)
    - Headache (5-14%)
    - Focal neurologic deficits
    - Incidental discovery

- Typically, sporadic etiology

- Most common genetic etiology is hereditary hemorrhagic telangiectasia (HHT)
    - Autosomal dominant
    - Presentation includes recurrent epistaxis, mucocutaneous telangiectasias, incidentally discovered AVM(s)
    - Discovery of multiple AVMs is highly predictive of HHT

Abecassis et al., *Neurosurg Focus* (2014);
Krings *et al*., *AJNR Am J Neuroradiol* (2015)

Spetzler-Martin Grading Scale assesses morbidity and mortality risk of surgery in brain AVMs

- Grade = sum of points from each feature
  - Grade I-II: low-grade, low morbidity/mortality risk
  - Grade III: intermediate-grade, heterogenous risk based on features
  - Grade IV-V: high-grade, unacceptably high morbidity/mortality risk
  - Grade "VI": inoperable, almost inevitable total disability or death

- Supplementary grading system proposed but requires external validation

Graded Feature of AVM	Points
Size of Nidus	
Small (< 3 cm)	1
Medium (3 - 6 cm)	2
Large (> 6 cm)	3
Eloquence of Adjacent Brain	
Non-eloquent	0
Eloquent	1
Venous Drainage	
Superficial	0
Deep	1

Supplementary Grading	Points
Age	
< 20 years	1
20 - 40 years	2
>40 years	3
Rupture status	
Ruptured	0
Unruptured	1
Diffuse	
No	0
Yes	1
Perforating arterial supply	
No	0
Yes	1

Spetzler & Martin, *J Neurosurg* (1986);
Lawton *et al.*, *Neurosurgery* (2010);
Kim *et al.*, *Neurosurgery* (2015)

Treatment goal of AVM is to reduce lifetime risk of AVM-associated hemorrhage

- Management options:
  - Microsurgical resection → high success rate for high-risk and/or ruptured AVMs
  - Stereotactic radiosurgery → most successful for small AVMs
  - Endovascular embolization → typically used as adjunct prior to resection
  - Conservative management → lower 33-month risk of stroke/death in unruptured AVMs
    - Based on ARUBA – currently only randomized controlled trial on management of unruptured AVMs
    - ARUBA remains controversial
- Pharmaceutical therapy not yet available
- Intervention of AVMs does not necessarily reduce subsequent seizure/epilepsy risk

Solomon & Connolly Jr., *N Engl J Med* (2017);
Derdeyn et al., *Stroke* (2017);
Mohr et al., *Lancet* (2014)

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