

## Vascular Malformations: Brain



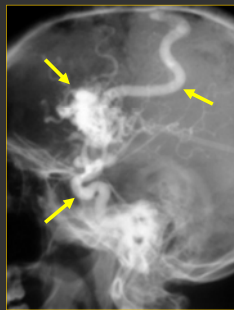
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## Vascular Malformations

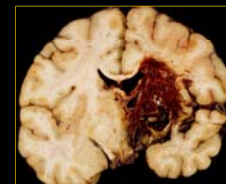
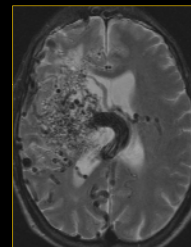
- Arteriovenous malformation (AVM)
- Dural arteriovenous fistula (dAVF)
- Vein of Galen Malformation (VOG)
- Cavernous angioma
- Developmental venous anomaly (DVA)
- Capillary telangiectasia
- Sinus pericranii

## AVM

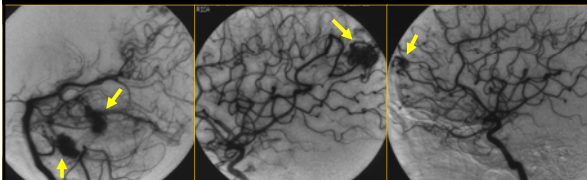
- Nonneoplastic vascular abnormalities
- Arteriovenous shunting & no intervening capillary bed
  - congenital
  - Usually neural tissue in between
- Occur anywhere in brain or spinal cord
- 98% solitary
  - Multiple AVMS usually syndromic:
    - Hereditary hemorrhagic telangiectasia (HHT)



## AVM



## HHT



## AVM

- Peak age: 20-40 year old
- Risk of hemorrhage: 2-4%/year
  - ~50% present with symptoms of hemorrhage
- Spontaneous obliteration rare: < 1%
- Dysregulated angiogenesis → continued vascular remodeling

### AVM Grading: Spetzler - Martin Scale

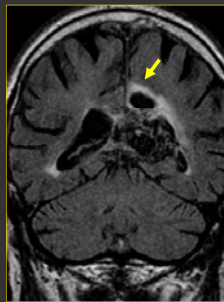
- Size
  - Small (<3cm) = 1
  - Medium (3-6 cm) = 2
  - Large (>6 cm) = 3
- Location
  - Noneloquent = 0
  - Eloquent = 1
- Venous drainage
  - Superficial = 0
  - Deep = 1

### AVM Imaging: CT

- Variable Hemorrhage
- Calcification: 25-30%
- Enhance post-contrast
- CTA: Enlarged arteries & draining veins

### AVM Imaging: MRI

- Flow Voids: “Bag of worms”
- Variable hemorrhage
  - “Blooming” on T2\* GRE
- T2: Increased signal → gliosis
- Contrast: Strong enhancement
- MRA/MRV



### AVM Imaging: Conventional Angiography

- Best method of imaging
- Must image ICA, ECA & vertebral circulations
  - 27-32% of AVMs have dual arterial supply
- Dural arterial supply via leptomeningeal anastomoses or transdural anastomoses
  - Transdural anastomoses affects treatment decisions

### AVM: Associated Abnormalities

- Flow-related aneurysm on feeding artery: 10-15%
- Intranidal “aneurysm”: >50%
- Vascular “steal”: Ischemia in adjacent brain



### Increased Risk of Hemorrhage

- Central venous drainage
- Intranidal aneurysm
  - Difficult to detect by MR
- Peri- or intraventricular location
- +/- obstruction of venous outflow

## AVM: Treatment

- Embolization
- Radiation: Stereotaxic radiosurgery
- Microvascular surgery

## dAVF

- Arteriovenous shunts within dura
- 10-15% of intracranial vascular malformations
- 2 types:
  - Adult: Tiny vessels in wall of thrombosed dural venous sinus → typically middle aged & older patients
    - Usually acquired - trauma
  - Infantile: Multiple high-flow AV-shunts involving several thrombosed dural sinuses

## dAVF Grading: Cognard Classification

- Type I: In sinus wall, normal antegrade venous drainage
- Type II: In main sinus
  - A: Reflux into sinus
  - B: Reflux into cortical veins: 10-20% hemorrhage
- Type III: Direct cortical drainage
  - 40% hemorrhage
- Type IV: Direct cortical drainage + venous ectasia
  - 2/3 hemorrhage
- Type V: Spinal perimedullary venous drainage
  - Progressive myelopathy

## dAVF

- Most common near skull base
  - Transverse sinus most common
- Hemorrhage incidence: 2-4% per year
- Spontaneous closure rare
  - Most are type I

## dAVF Imaging: CT

- NECT: May be normal
- CECT: May see tortuous dural feeders & enlarged dural sinus

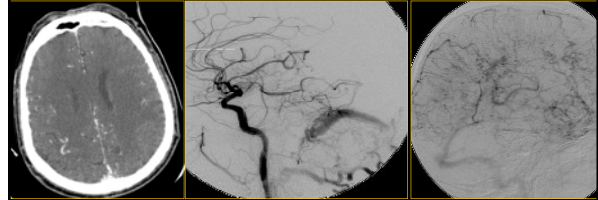
## dAVF Imaging: MRI

- Flow voids around dural venous sinus
- Thrombosed sinus
- Dilated cortical veins without parenchymal nidus
- T2: Focal hyperintensity in adjacent brain → retrograde leptomeningeal venous drainage/venous perfusion abnormalities
- MRA: May be negative
- MRV: Occluded sinus, collateral flow

## dAVF: Conventional Angiography

- Multiple arterial feeders
  - Dural/transosseous branches from ECA: most common
  - Tentorial/dural branches from ICA or VA
- Involved dural sinus frequently thrombosed
- Flow reversal in dural sinus/cortical veins
  - progressive symptoms, risk of hemorrhage
- Tortuous engorged pial veins
  - "pseudophlebitic pattern"

## dAVF



## Carotid Cavernous Fistula (CCF)

- dAVF second most common site
- Abnormal communication between carotid artery & cavernous sinus
  - Enlarges cavernous sinus
  - Usually see enlarged superior ophthalmic vein
    - CCF may be contralateral to dilated SOV
- Classified by arterial supply & venous drainage (Barrow):
  - A: Direct ICA-cavernous sinus high-flow shunt
  - B: Dural ICA branches-cavernous shunt
  - C: Dural ECA-cavernous shunt
  - D: ECA/ICA dural branches shunt to cavernous sinus

## CCF: Imaging

- **CT:**
  - Marked dilation & enhancement of cavernous sinus
  - May see prominent SOV
- **MRI:**
  - Abnormal flow voids in cavernous sinus
  - Enlargement of cavernous sinus



Non-Contrast

## CCF



## dAVF: Treatment

- Endovascular
- Surgical resection
- Stereotaxic radiosurgery
- Observation: Type 1

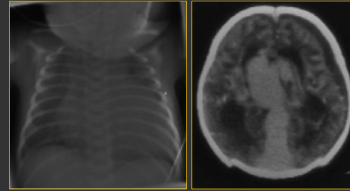
## Vein of Galen Malformation (VOG)

- Arteriovenous fistula involving aneurysmal dilatation of median prosencephalic vein
- Neonatal > infant presentation
  - Rare adult presentation
- Classification:
  - Choroidal: Multiple feeders from pericallosal, choroidal, & thalamoperforating arteries
  - Mural: Few feeders from collicular or posterior choroidal arteries



## VOG

- Newborns: Most common extracardiac cause of high-output congestive heart failure
- < 1% of cerebral vascular malformations

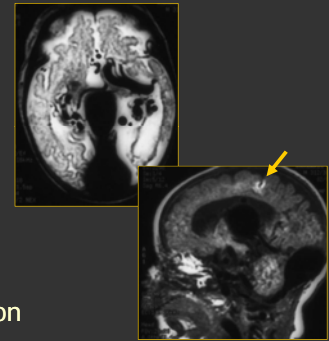


## VOG: CT Findings

- Venous pouch
- May have hydrocephalus
- Atrophy
- Parenchymal atrophy
- Intraventricular hemorrhage: Rare
- Post contrast: Avid enhancement of feeding arteries and vein

## VOG: MR Imaging

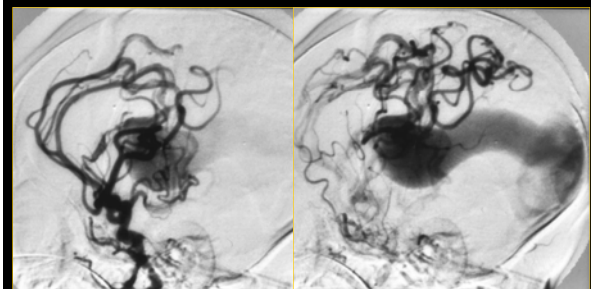
- Flow voids
- T1 hyperintensity
  - In pouch → thrombus
  - In brain → ischemia, calcification
- DWI: Restricted diffusion if acute ischemia, infarction



## VOG: Angiography

- Choroidal or mural
- Dural venous sinus anomalies
  - Falcine sinus in 50%
  - +/- absence or stenosis of other sinuses

## Choroidal



### VOG: Treatment

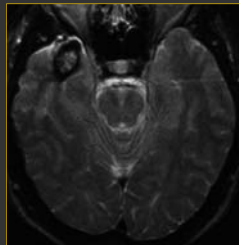
- Choroidal
  - Medical therapy for congestive heart failure until 5 or 6 mo
  - 5-6 mo: Transcatheter embolization
    - Arterial more effective than venous
- Mural
  - Transcatheter embolization performed later

### Cavernous Malformation

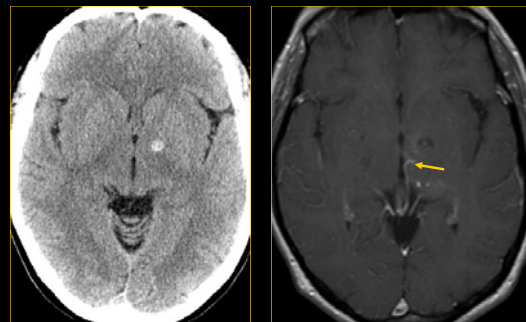
- AKA: Angioma, cavernoma, cavernous hemangioma
- Variable size intercapillary vascular spaces, sinusoids, & larger cavernous spaces
  - No intervening brain
  - "Blood sponge"
- 2 types:
  - Inherited: Multiple & bilateral
  - Sporadic

### Cavernous Malformation: Imaging

- Little or no mass effect
  - Unless complicated by hemorrhage
- May have internal areas of thrombosis or hemorrhage
  - Peripheral hemosiderin causes T2 shortening resulting in a black "halo" around the lesion

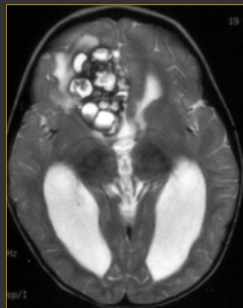


### Cavernous Malformation

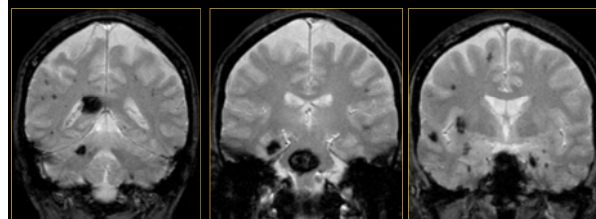


### Cavernous Malformation

- Risk of hemorrhage 0.25-0.7%/year
  - More common in posterior fossa lesions
  - In patients with prior hemorrhage annual rate of rehemorrhage → 4.5%
- Treatment:
  - Observation: Asymptomatic or inaccessible lesions
  - Surgical excision
  - Radiosurgery: Progressively symptomatic but surgically inaccessible



### Cavernous Malformation



MPGR

## Developmental Venous Anomaly (DVA)

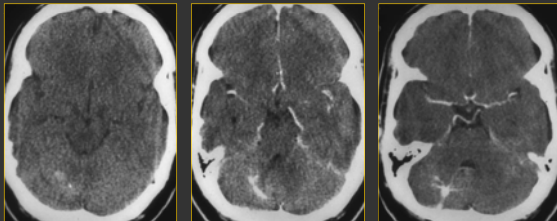
- May represent anatomic variant
  - Seen in up to 3% of autopsies
- Enlarged medullary veins
- Drain into dural sinus or deep ependymal vein
- Usually solitary
- “Medusa head” or “palm tree”

## Developmental Venous Anomaly

- Isolated or associated with cavernous angioma
- Hemorrhage unusual

## DVA Imaging: CT

- Calcification & ischemia may occur in the region drained → most likely due to chronic venous obstructive disease
  - Rare



## DVA

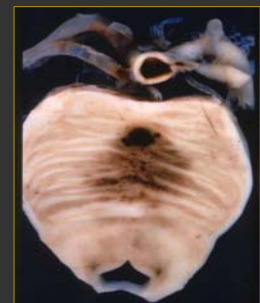


## DVA: Treatment

- NONE!
  - Removal may cause venous infarction

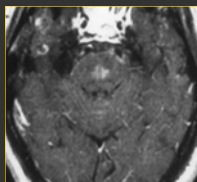
## Capillary Telangiectasia

- Dilated capillaries interspersed within normal brain
- Usually small, asymptomatic incidental findings
  - Rare reports of hemorrhage exist
- Most located in brainstem → Pons



## Capillary Telangiectasia

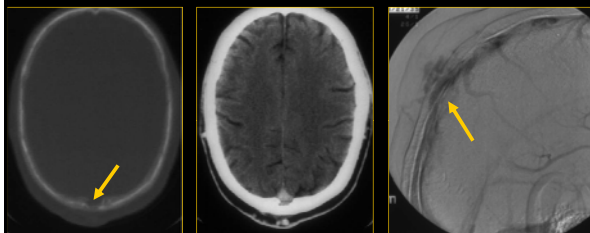
- T2: Increased signal
- T2\*: Low signal
- Ill defined enhancement after contrast administration
- Occult on angiography
- Treatment: None



## Sinus Pericranii

- Communication between extracranial venous system & dural venous sinus
- Rare
- May be congenital or acquired

## Sinus Pericranii



- CT: Single/multiple bone defects
- Vascular enhancement
- Conventional angiogram: Seen during venous phase

## Sinus Pericranii

- Spontaneous regression rare
- Risk of hemorrhage
- Treatment
  - Surgery
  - Endovascular

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