Vascular Disease and Intracranial Hemorrhage: Case Based Review

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Disclosures

• Nothing to disclose.
All the following are likely etiologies for SAH except:

- Trauma
- Arteriovenous malformation
- Aneurysm
- Vasculitis
- Dissection
- Stroke

Case #1

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Case #1
Intracranial Aneurysms

Axial 3D TOF MRA: Left supraclinoid ICA aneurysm and basilar artery aneurysm.

Case #1

Which of the following is true?

- Posterior communicating artery origin aneurysms can result in subdural hemorrhage
- Aneurysms rarely grow
- There is a higher rate of recurrence with clipped aneurysms as compared to coiled aneurysms
- Calcified aneurysms do not pose any additional treatment risk
Which Aneurysm Ruptured?

Cerebral aneurysm

- Three types of cerebral aneurysms
  - Saccular
  - Fusiform
  - Dissecting
Saccular (Berry) Aneurysm

- “True” aneurysms: frequently arise at intracranial branch points, rounded
- 1-5% incidence on angiography
- Multiple aneurysms in 15-20%
- Age at presentation: 40-60 years
- Most common presentation: SAH
- Typically Asx until rupture
  - May cause cranial neuropathies, sz, headache, infarct
- Risk of rupture: 1-2% per year

Case #1

Saccular Aneurysm

- Presentations: SAH
- Location: At bifurcations
  - 30-35% ACOM
  - 30-35% ICA/PCOM
  - 20% MCA bifurcation
  - 5% basilar
- Etiology
  - Once thought to be congenital
  - More likely hemodynamically induced degenerative vascular injury
  - Other causes: trauma, infection, drug abuse, high flow states (AVM/AVF)
  - Associated with connective tissue d/o, FMD and PCKD
Most common cause of death/morbidity in aneurysm rupture?

- Complications of vasospasm
- Cerebral edema
- Brain herniation
- Hydrocephalus
- Hypotension
- Infection

Etiologies for stroke include all except?

- Plaque rupture/artery embolus
- Cardiac embolus
- Vasculitis
- Hypoperfusion
- Venous thrombosis
- Reversible Cerebral Vasoconstriction Syndrome (RCVS)
- Arachnoid Cyst
Differential Includes the following Except?

- Moyamoya Disease
- Atherosclerosis
- Vasculitis
- RCVS
- Vasospasm

Most Likely Diagnosis?

- Moyamoya Disease
- Atherosclerosis
- Vasculitis
- Reversible Cerebral Vasoconstriction Syndrome (RCVS)
- Vasospasm
Most Likely Diagnosis?

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What is the correct treatment answer?

- Symptomatic Moyamoya disease should be first treated medically
- Reversible cerebral vasoconstriction syndrome is surgically managed
- Infectious vasculopathy should be treated with a trial of steroids for diagnosis
- First line management for stenotic atherosclerosis is surgical bypass
- Primary CNS vasculitis is treated with immunomodulators
### Vasculitis

- The Calabrese criteria for PACNS diagnosis:
  - an acquired or otherwise unexplained neurological deficit
  - angiographic or histopathologic features of angiitis within the CNS
  - absence of another systemic disorder to explain these features

- Secondary: Any systemic vasculitis can affect the CNS

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### Diagnosis

- Systemic vasculitis with cerebral manifestations
  - diagnosis depends on serum tests and body tissue biopsies.

- Diagnosis of an isolated CNS manifestation can be more challenging.
  - CSF studies
  - Neuroimaging
  - Definitive diagnosis may require brain biopsy
Imaging Findings

– Luminal Imaging (CTA, MRA, DSA):
  • Multi-focal arterial stenoses with beaded appearance
    – Nonspecific
  • Luminal imaging may be normal
    – DSA sensitivity between 50-90% for CNS vasculitis

– Non-contrast CT findings
  – SAH and IPH
  – Signs of acute and chronic ischemia

MRI Findings

• Conventional MRI most sensitive imaging exam for vasculitis.
  – Negative MRI can essentially rule out vasculitis
  – MRI abnormalities in 90–100% of vasculitis
  – Findings of ischemia:
    • DWI, T2 FLAIR, enhancement findings of different ages
  – IPH and SAH: FLAIR and SWI
• MRI non-specific for the diagnosis of vasculitis
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Cerebral Vasculitis

Hemorrhage
  vessel wall inflammation
  arterial rupture

Infarct
  vessel wall inflammation
  high-grade stenosis
  intraluminal thrombus

Case #2

T1 POST
T2 FLAIR
Vessel Wall Imaging Findings

Circumferential, intense enhancement of the vessel wall.

What is the most likely diagnosis in this 20 year old female?

- Moyamoya disease
- Reversible Cerebral Vasocostriction Syndrome
- Atherosclerosis
- Dissection
- Vasospasm
What is the most common pediatric/adult presentation of Moyamoya?

- Hemorrhage/Ischemia
- Dementia/Ischemia
- Hemorrhage/Dementia
- Ischemia/Hemorrhage
- PRES/Dementia
- Infection/PRES

Moyamoya Vasculopathy

- Progressive steno-occlusive vascular disease affecting the carotid termini with development of extensive basal collaterals
- Moyamoya disease: Idiopathic disease, bimodal distribution (<10 yo and 40’s), most commonly in Japanese
- Moyamoya syndrome: Secondary process- Atherosclerosis, vasculitis, radiation, genetic disorders (sickle cell, Downs, NF-1, etc.)
Moyamoya Presentation

- MMS more frequently seen in US populations
- Pediatric
  - Ischemia as collaterals are not well developed
- Adults
  - Hemorrhage relating to rupture of fragile collaterals
- Treatment
  - Peds: Indirect ECA to MCA bypass
  - Adults: Heterogeneous, but for symptomatic-direct ECA to MCA bypass

Imaging Findings

- MRI/CT
  - Ischemia and intracranial hemorrhage
  - Chronic ischemic changes/atrophy
  - Loss of flow voids
- Luminal Imaging
  - Occlusion of the carotid termini with extensive collaterals
Imaging Findings

- MRI/CT
  - Ischemia and intracranial hemorrhage
  - Chronic ischemic changes/atrophy
  - Loss of flow voids
- Luminal Imaging
  - Occlusion of the carotid termini with extensive collaterals

Case #3

All the following disease/treatment possibilities are correct except:

- Vasculitis-Immunosuppressive
- RCVS- Calcium channel blockers
- Moyamoya Disease-Aspirin
- Atherosclerosis-Statins/Anti-platelet

Case #4
35 year old female presented with thunderclap headache, diagnosis?

- Vasculitis
- RCVS
- Moyamoya disease
- Aneurysm with rupture
- Atherosclerosis
- Dissection

Case #4

RCVS

- Presentation:
  - Thunderclap headache that is intermittent
  - Young and middle aged females
  - Inciting etiology: SSRI, sympathomimetic agents, coitus, cold shower, eclampsia, post-partum, activity, alcohol, etc.

- Intracranial vasospasm that tends to resolve after 1-3 months
RCVS- Imaging Findings

• MRI/CT
  – Multifocal infarcts
  – Subarachnoid and IP hemorrhage
  – PRES-like appearance

• Luminal Imaging
  – Beaded arterial appearance
    • Alternating segments of narrowing and dilatation
    • Indistinguishable from vasculitis
  – Findings will progressively improve on CTA/MRA/DSA
30 year old with parenchymal hemorrhage, what is the most likely diagnosis?

- Cerebral amyloid
- Ischemic infarct
- Anterior communicating aneurysm rupture
- Ruptured developmental venous anomaly
- Arteriovenous malformation

Which of the following syndromes is associated with AVM?

- Kearns-Sayre Syndrome
- Hereditary Hemorrhagic Telangiectasia
- Down’s Syndrome
- Morquio Syndrome
- Kartagener Syndrome
Cerebral AVM

- Congenital lesions
  - Feeding artery(ies)
  - Intervening nidus
  - Draining vein(s)

Case #5

- Presentation
  - Hemorrhage most common (42-72%)
  - Seizures (25%)
  - Headaches (15%)
  - Focal neurologic deficit
    - Steal phenomenon
    - Pulsatile tinnitus
  - Thought as congential lesions

- Associations with initial hemorrhage
  - Deep brain location
  - Deep venous drainage
  - Associated aneurysms
  - Posterior fossa

- Risk for repeat hemorrhage
  - Prior hemorrhage
Imaging Findings

- **CTA/MRA**
  - Tangle of vessels with large feeding arteries and draining veins

- **Catheter Angiography**
  - Tangle of vessels/nidus
  - Feeding arteries
  - Early appearance of draining veins
Imaging Findings

- **NC CT**
  - Curvilinear or speckled calcifications
  - Hyperattenuating serpentine vascular structures
  - Hemorrhage
  - Surrounding hypoattenuation

- **MR**
  - Arterial, venous and nidal flow voids
  - Perinidal T2 hyperintensity
  - Enhancement on post-contrast MRI
  - AVM may be compressed by hematoma
    - May not be seen on any imaging modality early
    - Repeat imaging once the hematoma has reduced in mass effect may be necessary
Case #6

What is the most likely diagnosis?

- Fibromuscular dysplasia
- Dissection
- Arteriovenous fistula
- Atherosclerosis
- Thromboembolism
Arterial Dissection

- **Subintimal**
  - Intimal tear: Collection of blood in subintimal space
  - Complications:
    - Hypoperfusion from luminal narrowing
    - Embolic events

- **Subadventitial**
  - Blood collects in subadventitial space
  - Secondary to vasa vasorum rupture or extension of subintimal dissection

Luminal Imaging Findings

- **DSA/CTA**
  - Very good at depicting subintimal dissections
    - Double lumen appearance
    - Stenosis of true lumen
  - Not as good with subadventitial dissections
    - Especially if do not result in luminal stenosis
MRI Findings

- Sensitive for detection of thrombus within subintimal or subadventitial dissections
- T1/PD fat sat
  - Crescentic hyperintensity adjacent to lumen

What is abnormal?

- Left cerebellum
- Suprasellar cistern
- Interpeduncular cistern
- Frontal lobe white matter
- Right MCA
What is the diagnosis?

- Subarachnoid hemorrhage
- Subdural hemorrhage
- Hyperacute ischemic infarct
- Meningitis w/ vasculitis
- Moyamoya Disease

What is the diagnosis?

- Subarachnoid hemorrhage
- Subdural hemorrhage
  - Hyperacute ischemic infarct
- Meningitis w/ vasculitis
- Moyamoya Disease
Case #7

DSA before and after thrombectomy

Case #7

Thrombus in Stent Retrieval Device
Hyperacute Infarct

- CT
  - Hyperdense MCA
  - Loss of gray-white distinction
  - Loss of basal ganglia
- MRI
  - DWI abnormality with matching ADC abnormality
  - Decreased CBV/Prolonged transit time on MRP
  - No T2 FLAIR parenchymal signal abn
  - Hyperintense arteries (slow or no flow)

Subacute Infarct

- CT
  - Increased mass effect/herniation
  - Edema
  - Sulcal effacement
  - May see hemorrhagic conversion
- MRI
  - Infarct enhancement
  - Pseudonormalization of ADC
  - T2/FLAIR signal abn
  - T2 fogging effect
Subacute Infarct

- **CT**
  - Increased mass effect/herniation
  - Edema
  - Sulcal effacement
  - May see hemorrhagic conversion

- **MRI**
  - Infarct enhancement
  - Pseudonormalization of ADC
  - T2/FLAIR signal abn
  - T2 fogging effect
Chronic Infarct

- Volume loss
  - Shrinkage of parenchyma and ventricular enlargement
- Loss of overlying cortex
- CSF attenuation/signal encephalomalacia with surrounding gliosis
- SWI/GRE: Hemosiderin deposition due to hemorrhage

Thank you!

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