Pediatric Headaches: When should we image?

A Case of a 15 year old boy with sudden onset of headache

Erin West, Harvard Medical School Year III
Gillian Lieberman, MD
BIDMC Radiology Core Clerkship
February 2011
Outline

– Pediatric Headaches: Classification and Causes
– Indications for Imaging
– Menu of Tests
– Case Presentation/Brief Neuroanatomy Review
– Differential Diagnosis
– Brief Discussion of Patient’s Diagnosis
Classification of Pediatric Headaches

- **Acute**
  - Localized
    - Associated with URI (sinusitis, Otitis media) or viral infection (influenza)
    - Post-traumatic
    - Related to oral cavity (dental abscess, TMJ dysfunction)
    - First migraine
  - Generalized
    - Systemic infection (influenza, meningitis)
    - Intracranial hemorrhage
    - Exertional
    - First migraine
- **Acute and Recurrent**
  - Migraine headache
  - Cluster headache
- **Chronic and Non-Progressive**
  - Tension type headache
  - Psychiatric (depression, school phobia)
- **Chronic and Progressive**
  - Idiopathic intracranial hypertension
  - Space occupying lesion (tumor, abscess, hemorrhage, hydrpcephalus)

**Primary Causes**
- Infection/Inflammatory
- Neoplasm
- Vascular
- Trauma
- Psychosocial

**Secondary**—everything else

**Headache Pain Mechanism:**
- Traction on meningeal pain fibers
- Increased intracranial pressure (ICP)
- Blood in the cerebrospinal fluid

Pediatric Headaches: Goal of Initial Care

- Most causes of pediatric headache are benign
- Goal of initial care is to identify cases that require urgent diagnostic evaluation and treatment. These include:
  - Subarachnoid hemorrhage (SAH) from ruptured aneurysm
  - Intracranial bleeding from an arteriovenous malformation
  - Neoplastic lesions
  - Encephalitis
  - Meningitis
  - Intracranial venous sinus thrombosis
  - Vasculitis
  - Metabolic disorders

Field et al.
Overview of Pediatric Headaches

- **Most common cause according to setting of presentation**
  - In the emergency room: viral infection, upper respiratory infection, minor head trauma\(^1\)
  - In the primary care setting: psychosocial (family or school problems) or infectious \(^2\)
- **Neuroimaging for pediatric headaches is controversial**
  - Majority are benign and majority of imaging studies in patients with headache are normal\(^3\)
  - Very high prevalence of headaches but low prevalence of brain tumors (annual incidence is 3 per 100,000 children)\(^4\), or other space-occupying lesions

\(^{1}\text{Kan et al.}\)
\(^{2}\text{Van der Wouden}\)
\(^{3}\text{Schwedt et al.}\)
\(^{4}\text{The Childhood Brain Tumor Consortium}\)
Important Historical Data to Gather in a Child with Headache

- Age at onset
- Presence or absence of aura and prodrome
- Frequency, severity, and duration of pain
- Time and mode of onset
- Quality, site, and radiation of pain
- Associated symptoms and signs
- Family history of headache or other neurologic disorder
- Precipitating and relieving factors
- Effect of activity on pain

- Relationship with food/alcohol
- Response to any previous treatment
- Recent change in weight or vision
- History of recent trauma
- Recent changes in sleep, exercise, or diet
- State of general health
- Change in school or home environment
- Association with environmental factors
When should we image a child with headache?

- In a 4 year retrospective study by Medina et al, 7 independent predictors of surgical lesions in patients with headache were identified. Positive correlation was found between the number of predictors and risk of space-occupying lesion (neoplasms, hemorrhagic vascular malformations, arachnoid cyst)
  - 4% of patients had surgical space-occupying lesions
  - **Independent multivariate predictors of a surgical lesion**
    - Strongest predictors:
      - Sleep-related headache (awaken from sleep, occur on awakening)
      - Absent family history of migraine
    - Vomiting
    - Absence of visual symptoms
    - Headache of less than 6 months duration unresponsive to medical treatment
    - Confusion or disorientation
    - Abnormal neurologic examination findings (papilledema, nystagmus, gait or motor abnormalities)

Medina et al.
Useful predictors of positive imaging evaluation in a child with headache

• Positive Neurologic Signs or Symptoms
• Supporting Patient Historical Data
  – Diplopia
  – Morning vomiting
  – Headaches that awaken from sleep
• Sudden severe headache “thunderclap” headache
  – Associated with subarachnoid hemorrhage and intracranial hemorrhage that may occur with aneurysms or AVMs
• Trauma
# American College of Radiology (ACR) Appropriateness Criteria

## Child with Headache

### Variant 2. Headaches with positive neurologic signs or symptoms

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Appropriateness Rating</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT, head</td>
<td>8</td>
<td>CT or MRI should be performed in every patient</td>
</tr>
<tr>
<td>MRI, head</td>
<td>8</td>
<td>CT or MRI should be performed in every patient</td>
</tr>
<tr>
<td>Catheter angiography</td>
<td>2</td>
<td>Unless indicated by other studies</td>
</tr>
<tr>
<td>SPECT, head</td>
<td>2</td>
<td>Only helpful when further evaluating the lesion</td>
</tr>
</tbody>
</table>

Note: Appropriateness Criteria® scale: 1 = least appropriate; 9 = most appropriate. CT = computed tomography; MRI = magnetic resonance imaging; SPECT = single photon-emission computed tomography.

### Variant 3. Headaches with supporting historical data (diplopia, morning vomiting, headaches that awaken patient from sleep)

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Appropriateness Rating</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>MRI, head</td>
<td>9</td>
<td>MRI is recommended, but CT is acceptable alternative</td>
</tr>
<tr>
<td>CT, head</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>MRA, head</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Catheter angiography</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>SPECT, head</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

Note: Appropriateness Criteria® scale: 1 = least appropriate; 9 = most appropriate. CT = computed tomography; MRA = magnetic resonance angiography; MRI = magnetic resonance imaging; SPECT = single photon-emission computed tomography.

ACR Appropriateness Criteria
Child with Headache

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Appropriateness Rating</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT, head</td>
<td>9</td>
<td>Noncontrast preferred</td>
</tr>
<tr>
<td>MRI, head</td>
<td>8</td>
<td>Either CT or MRI, not both</td>
</tr>
<tr>
<td>CTA, head</td>
<td>7</td>
<td>Indicated if subarachnoid or parenchymal blood is identified on CT, MRI, or LP; either CTA or MRA, not both</td>
</tr>
<tr>
<td>MRA, head</td>
<td>7</td>
<td>Indicated if subarachnoid or parenchymal blood is identified on CT, MRI, or LP; either CTA or MRA, not both</td>
</tr>
<tr>
<td>Catheter angiography</td>
<td>6</td>
<td>If MRA or CTA not available or if intervention is considered</td>
</tr>
<tr>
<td>SPECT, head</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

Note: Appropriateness Criteria® scale: 1 = least appropriate; 9 = most appropriate. CT = computed tomography; CTA = computed tomographic angiography; MRA = magnetic resonance angiography; MRI = magnetic resonance imaging; SPECT = single photon-emission computed tomography.

http://www.acr.org/secondarymainmenucategories/quality_safety/app_criteria.aspx
Menu of Tests for Pediatric Headache

- **CT Head without contrast**
  - Primary modality in acute or emergent cases
    - Fast, cost-effective, and readily available
  - Benefits: Screens for acute or subacute hemorrhage (without contrast), edema, herniation, fractures, hypoxic-ischemic injury, focal infarction, hydrocephalus, tumor mass, or abnormal collection (eg, pneumocephalus, abscess, empyema)
  - Disadvantages: Not very sensitive for ischemic changes prior to 24 hours, not sensitive for gray-white matter differentiation, exposure to ionizing radiation

- **CT Angiogram:**
  - Recommended if CT without contrast shows subarachnoid hemorrhage or intraparenchymal bleeds
  - Recommended for the evaluation of suspected or known vascular malformation, infarction, neoplasm, abscess, or empyema, and in patients with head and neck masses.
Menu of Tests continued

- MRI
  - Preferred modality for detecting cranial pathology in children in non-emergent cases
    - Demonstrates sellar lesions, craniocervical junction lesions, posterior fossa lesions, white matter abnormalities, and congenital anomalies more accurately than does CT
  - Benefits: no ionizing radiation, finer parenchymal detail than CT, better white-gray matter differentiation, more sensitive for detection of ischemic changes (especially diffusion-weighted MRI) and vascular abnormalities
  - Disadvantages: costly, time-consuming, may require sedation ➔ not ideal in emergent cases or for children

- Conventional angiography
  - Cerebral angiogram is used to further delineate vascular malformations and for treatment planning
  - Invasive and requires sedation
Now that we know the etiologies of headaches in the pediatric population, the clinical predictors of a positive neuroimaging study, and the menu of tests available, we are ready to meet our patient!
Our Patient: Clinical Presentation

- **Clinical Presentation**
  - **History of Present Illness**
    - 15 year old previously healthy boy
    - Sudden onset of severe headache 10/10 while in shower the night prior to presentation to ED
    - No alleviation with aspirin
    - Associated with emesis X 2
    - No sleep due to pain
    - Continued, steady → worsening pain until morning of presentation
  - **Past Medical History**
    - History of mild headaches
  - **Pertinent ROS**
    - Denied photophobia, visual changes, weakness or numbness
    - Denied fever, recent illness, neck pain or stiffness
    - Denied recent head trauma
  - **Physical Exam**
    - Vital signs normal, no signs of increased intracranial pressure
    - Neuro exam: alert but sleepy, no focal neurologic deficits, CN2-12 grossly intact, sensory and motor function intact, no ataxia, reflexes 2+ throughout.
Which imaging study should we order given our patient’s clinical presentation?

Let’s look what the ACR appropriateness criteria suggest...
ACR Appropriateness Criteria
Child with Headache

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Appropriateness Rating</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT, head</td>
<td>9</td>
<td>Noncontrast preferred</td>
</tr>
<tr>
<td>MRI, head</td>
<td>8</td>
<td>Either CT or MRI, not both</td>
</tr>
<tr>
<td>CTA, head</td>
<td>7</td>
<td>Indicated if subarachnoid or parenchymal blood is identified on CT, MRI, or LP; either CTA or MRA, not both</td>
</tr>
<tr>
<td>MRA, head</td>
<td>7</td>
<td>Indicated if subarachnoid or parenchymal blood is identified on CT, MRI, or LP; either CTA or MRA, not both</td>
</tr>
<tr>
<td>Catheter angiography</td>
<td>6</td>
<td>If MRA or CTA not available or if intervention is considered</td>
</tr>
<tr>
<td>SPECT, head</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

Note: Appropriateness Criteria® scale: 1 = least appropriate; 9 = most appropriate. CT = computed tomography; CTA = computed tomographic angiography; MRA = magnetic resonance angiography; MRI = magnetic resonance imaging; SPECT = single photon-emission computed tomography.

http://www.acr.org/secondarymainmenucategories/quality_safety/app_criteria.aspx
Before we review our patient’s non-contrast CT head, the imaging study of choice to rule out intracranial bleeds, let’s briefly review the appearance of hemorrhage on CT and MRI.
Appearance of Hemorrhage in Brain on CT and MRI

- **CT**
  - Acute hemorrhage (<3 days)
    - Hyperdense (80-100 HU) relative to brain (40-50 HU)
    - High density caused by protein-hemoglobin combination
    - Not hyperdense if hematocrit is low
  - Subacute hemorrhage
    - Hyper, iso, or hypodense relative to brain
    - Degradation of hemoglobin complex evolves from margins to inside
  - Chronic hemorrhage (> 2 weeks, depending on size)
    - Hypodense relative to brain

- **MRI**
  - Acute hemorrhage
    - T1: isointense
    - T2: hyperintense (only in hyperacute (<12 hours) bleeds—as soon as hemoglobin becomes deoxygenated, the signal is hypointense)
  - Subacute hematoma. ICH appears as high signal intensity on T1 due to the presence of methemoglobin, particularly in the periphery. The appearance on T2 is initially dark, then later becomes bright as the red blood cells lyse and methemoglobin becomes extracellular.
  - Chronic hematoma. Hemosiderin is produced by phagocytes ingesting methemoglobin; this appears as a low signal on T2 and T1, enhanced by susceptibility weighted images such as GRE sequences

Reference: Weissleder and Wittenberg
CT/MR appearance of Hemorrhage in Brain

- **CT scan**: hyperdense hemorrhage with surrounding area of hypodense edema
- **T1**: central portion of the hematoma is isointense to brain parenchyma
- **T2**: central portion is hyperintense to brain parenchyma
- **T2-weighted and T2* gradient echo images**: periphery of the hemorrhage is hypointense, consistent with deoxygenation that occurs more rapidly at the borders.
- **T2 weighted image**: hyperintense surrounding tissue, consistent with vasogenic edema
- **Note**: All images are of patients with hemorrhage age <2 hours or a hyperacute bleed.
Our Patient: Initial Non-contrast CT Head—Axial View

- Sylvian fissure with CSF (low attenuation)
- Intraparenchymal hemorrhage centered in the right frontal lobe
- Supracellar cistern with low attenuation CSF (normal)
- Blood in 4th ventricle
Our Patient: Non-contrast Head CT—Axial View

- Right frontal lobe hemorrhage
- Blood in 3rd Ventricle
- Quadrigeminal cistern with normal low attenuation CSF
Our Patient: Initial Non-contrast CT Head—Axial View

Right Frontal Lobe Hemorrhage

Bilateral Lateral Ventricles with blood

Small white arrows refer to the rim of hypodensity, representative of vasogenic edema
Our Patient Non-contrast CT Head: Axial View

Subfalcine herniation with leftward midline shift

Bilateral Lateral Ventricles with blood

Mechanism of Injury from Intraparenchymal Hemorrhage
1. Primary direct, mechanical injury from expanding clot
2. Increased intracranial pressure
3. Mass effect leading to herniation
Neuroanatomy Review: The Ventricular System

CSF FLOW

Lateral ventricles (Choroid plexus) → 3rd Ventricle → Aqueduct of Sylvius → 4th Ventricle → Magendie and Lushka → Subarachnoid space
Cisterns are CSF collections surrounding the brain. 4 key cisterns must be examined for blood, asymmetry, and effacement (occurs with increased ICP).

- **Circummesencephalic** - ring around the midbrain
- **Suprasellar** - (Star-shaped) Location of the Circle of Willis
- **Quadrigeminal** - W-shaped at top of midbrain
- **Sylvian** - Between temporal and frontal lobes
Anatomic Basis of Herniation Syndromes

(1) Cingulate gyrus herniation under the falx
(2) Downward transtentorial (central) herniation
(3) Uncal herniation over the edge of the tentorium
(4) Cerebellar tonsillar herniation into the foramen magnum.

Coma and death can result with 2,3, or 4.
Childhood Stroke

Childhood Stroke
2-3/100,000*

Hemorrhagic Stroke
1.1/100,000

Ischemic Stroke
1.2/100,000

Intraparenchymal Hemorrhage (IPH)
0.8/100,000

Subarachnoid Hemorrhage (SAH)
0.3/100,000

Our Patient

*Per 100,000 children per year

Adapted from Jordan et al. Incidence numbers from Fullerton et al.
Differential Diagnosis for Spontaneous Intracranial Hemorrhage

• Most common etiologies of spontaneous (non-traumatic) ICH
  – Adults: hypertensive vasculopathy
  – Elderly: cerebral amyloid angiopathy
  – **Children: vascular malformations**
  – Additional causes of nontraumatic ICH include:
    • Hemorrhagic infarction (including venous sinus thrombosis)
    • Septic embolism, mycotic aneurysm
    • Brain tumor
    • Bleeding disorders, anticoagulants, thrombolytic therapy
    • Central nervous system (CNS) infection (eg, herpes simplex encephalitis)
    • Moyamoya
    • Vasculitis
    • Drugs (cocaine, amphetamines)
Differential Diagnosis of Spontaneous Intracranial Hemorrhage (SICH) in Children

- In a retrospective analysis of 50 consecutive pediatric patients with SICH, the most common causes of SICH were:
  - Arteriovenous malformation (AVM) rupture (most common)
  - Aneurysm
  - Moyamoya
  - Tumor
  - Hematologic disorder
  - Cavernoma
  - Unknown (possibly vasculitis)

Kumar et al.
Given that parenchymal and intraventricular blood was seen on the non-contrast CT, which test is appropriate?

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Appropriateness Rating</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT, head</td>
<td>9</td>
<td>Noncontrast preferred</td>
</tr>
<tr>
<td>MRI, head</td>
<td>8</td>
<td>Either CT or MRI, not both</td>
</tr>
<tr>
<td>CTA, head</td>
<td>7</td>
<td>Indicated if subarachnoid or parenchymal blood is identified on CT, MRI, or LP; either CTA or MRA, not both</td>
</tr>
<tr>
<td>MRA, head</td>
<td>7</td>
<td>Indicated if subarachnoid or parenchymal blood is identified on CT, MRI, or LP; either CTA or MRA, not both</td>
</tr>
<tr>
<td>Catheter angiography</td>
<td>6</td>
<td>If MRA or CTA not available or if intervention is considered</td>
</tr>
<tr>
<td>SPECT, head</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

Note: Appropriateness Criteria® scale: 1 = least appropriate; 9 = most appropriate. CT = computed tomography; CTA = computed tomographic angiography; MRA = magnetic resonance angiography; MRI = magnetic resonance imaging; SPECT = single photon-emission computed tomography.
Our Patient
Contrast-Enhanced CT Head

Subfalcine Herniation

Contrast enhances cerebral vasculature and further evaluates enhancement pattern of hematoma

Blood extending to lateral Ventricles

Area of hyperdensity within intraparenchymal hemorrhage, possibly representing the nidus of AVM

Images from Children’s Hospital Boston, PACS
Review
Anatomy of
Major Cerebral Arteries

http://www.accessmedicine.com
Copyright © The McGraw-Hill Companies, Inc. All rights reserved.
Our Patient
CT Head Angiogram—Maximum Intensity Projections (MIPS)

Major Cerebral Vessels (arrows)
- Posterior cerebral arteries
- Middle Cerebral Arteries
- Anterior cerebral arteries

Large branching vessel from distal right ACA draping over the anterior frontal lobe with a blush of contrast emerging from it at the anterior aspect of the intraparenchymal hemorrhage (consistent with AVM?)

Images from Children’s Hospital Boston, PACS
Given that there is question of an AVM on CTA and intervention is considered, which imaging study should we order?

http://www.acr.org/secondarymainmenucategories/quality_safety/app_criteria.aspx
Cerebral Angiography: Gold Standard for AVM diagnosis

- **Interventional radiology procedure**
  - Under general anesthesia, selective catheterization of artery of interest allows direct injection of contrast
  - Radiographic images (CT or MR) are then obtained in sequence during arterial, capillary and venous phases

- **Use in Evaluation of Brain AVMs**
  - Gold Standard for Diagnosis
  - Planning for Treatment/Surgical Intervention
  - Follow-up Evaluation of AVM
  - Evaluates:
    - Nidus configuration
    - Relationship to surrounding vessels
    - Localization of draining or efferent portion of the brain AVM
    - Contrast transit time allows evaluation of the flow state of lesion
Our Patient: Cerebral angiogram

Right Internal Carotid Artery Injection, Arterial Phase, Sagittal view

Go to next slide for coned in view of abnormality

Abnormality

Anterior cerebral artery

Middle cerebral artery

Posterior cerebral artery

Right internal carotid artery

Basilar artery

Children’s Hospital Boston, PACS
Our Patient: Cerebral Angiogram
AVM close-up

Single, superficial draining vein with early contrast enhancement (no intervening capillary network between feeding artery and draining vein)

Feeding artery off of anterior cerebral artery

☆ Small AVM nidus
Pattern Analysis of Cerebral Vessels on Angiography: Differential Diagnoses

- **Differential Diagnosis for Hypervascularity**
  - Arteriovenous malformation, vein of Galen aneurysm
  - Collateral circulation
  - Congenital variant
  - Neoplasm

- **Differential for Decreased Transit Time and Early Venous Filling**
  - Arteriovenous malformation
  - Increased pCO₂
  - Infarction
  - Neoplasm

- **Differential Diagnosis for Intracranial Arteriovenous Shunting and Early Venous Filling on Cerebral Angiography**
  - **Common**
    - AV malformation, congenital or acquired (carotid-cavernous fistula, vein of Galen “aneurysm”)
    - Infarction of brain
    - Occlusive vascular disease
    - Malignant neoplasm of brain, primary or metastatic
  - **Uncommon**
    - Cerebral arteritis
    - Contusion of brain
    - Epilepsy, focal idiopathic

Reeder and Felson’s Gamuts in Radiology
Brain Arteriovenous Malformations
Radiographic Features

• Radiographic features
  – Serpiginous high and low signal (depending on flow rates) within feeding and draining vessels best seen by MRI/MRA
  – AVMs replace but do not displace brain tissue (mass effect is uncommon)
  – No edema unless there is recent hemorrhage or venous thrombosis with infarction

• Complications
  – Parenchymal hemorrhage
  – Subarachnoid hemorrhage
  – Intraventricular hemorrhage

Weissleder et al.
The findings on cerebral angiogram confirmed the presence of a right frontal AVM and our patient was then brought to the OR for surgical resection. The following slides show a CT with and without contrast as well as a repeat cerebral angiogram to confirm resolution of our patient’s vascular lesion.
Operative Course of Our Patient: Surgical Resection of AVM

- **Procedures:**
  - Right frontal craniotomy for resection of arteriovenous malformation
  - Evacuation of intraparenchymal clot
  - Placement of external ventricular drain with duraplasty, microdissection, intraoperative fluorescein angiography, intraoperative ultrasonography, frameless stereotaxy.

- During procedure, duplex ultrasonography was used to guide procedure
  - prior to entering dura to identify AVM vessels
  - Identification of ventricular system entry for drain placement

- Catheter placed into the frontal horn near foramen of Monro in anticipation of potential hydrocephalus postoperatively
Our patient
Post – Surgical CTs

Post-Operative Changes:
- craniotomy (only seen on bone windows)
- mild extra-axial blood (shown with small yellow stars)
- pneumocephalus
- evidence of right frontal surgical track (not shown on these slices)
- right frontal external ventricular drainage catheter (tip is in region of foramen of Monro)
- persistent intraventricular blood in lateral ventricle
- mild decrease in mass effect and midline shift compared to prior CT
- persistent mild subfalcine herniation

Images from Children’s Hospital Boston, PACS
Our Patient

Post-Procedure Cerebral Angiogram

No evidence of right frontal AVM (previously fed by ACA)

No evidence of early venous enhancement of draining vein

Right Internal Carotid Artery Injection, Arterial Phase, Sagittal view

Images from Children’s Hospital Boston, PACS
We have now used a combination of non-contrast CT, CTA, and cerebral angiography to diagnosis and to guide the surgical treatment of our patient.

**Summary of Image Findings**
The non-contrast CT demonstrated the presence of the intraparenchymal hemorrhage while the CTA allowed direct visualization of the abnormal vessels. The cerebral angiogram then further delineating the cerebral vasculature and guided treatment decisions.

Let’s learn more about brain arteriovenous malformations!
Brain Arteriovenous Malformations (AVMs)

- **General Facts**
  - Sporadic congenital (but can be acquired) developmental vascular lesions
  - Brain AVM is the most common cause of spontaneous intraparenchymal hemorrhage in children, excluding hemorrhages of prematurity and early infancy
  - Consist of direct arterial to venous connection without intervening capillary network
  - High flow predisposes to aneurysm and arterialization of venous limb
  - Higher prevalence associated with hereditary hemorrhagic telangiectasia (HHT; Osler-Weber-Rendu syndrome)

- **Epidemiology**
  - Occur in about 0.1% of population
  - Supratentorial lesions 90%
  - 10% posterior fossa lesions
  - Annual hemorrhage rates are between 3-5% and after initial bleed, rates increase to 8%

- **Clinical Presentation**
  - Usually between ages 10-40 with intracranial hemorrhage, seizure, headache, and focal neurologic deficit.
  - *Hemorrhage is most common presentation, especially in children*

Singer et al
Brain AVMs: Diagnosis and Treatment

• **Diagnosis**
  – CT with contrast can demonstrate:
    • Flow voids in and around the region of the nidus
    • Intraparenchymal hemorrhage without significant surrounding edema
    • Hematoma can compress nidus and preclude CT diagnosis
    • *CT-angiography can improve sensitivity in acute setting*
  – MRI
    • Very sensitive for delineating location of brain AVM nidus and associated draining vein, and for demonstrating remote bleeding related to the lesion
    • Dark flow voids are seen on T1 and T2 weighted images
    • Valuable for following patients post-treatment
  – Angiography
    • Gold standard for diagnosis and treatment

• **Treatment options:**
  – Microsurgical resection
    • Gold standard of accessible pediatric AVMs, especially in cases that present with hemorrhage
  – Newer modalities better for deeper-seated lesions that are unresectable with microsurgical techniques alone
    • Embolization
    • Stereotactic radiosurgery
  – Multimodal approach is often used
  – Must consider patient’s age, lesion location and size, and prior history of intracranial hemorrhage when determining treatment

Niazi et al.  
Singer et al.  45
Learning Points

• Pediatric headaches
  – Headaches are extremely common in the pediatric population, and are often due to benign causes and usually do not require imaging.
  – Specific clinical predictors are useful to identify those patients that would benefit from neuroimaging

• Imaging Modalities for evaluation of Spontaneous Intracranial Hemorrhage
  – CT without contrast is the test of choice in emergent cases to identify acute cranial bleeds.
  – CT-angiogram is indicated if subarachnoid hemorrhage or parenchymal blood is identified on CT, MRI or LP.
  – MRI with and without contrast is very sensitive and specific for delineating vascular and hemorrhagic disorders, but is time-consuming and costly. Therefore, it is not often used in the acute setting when intracranial bleed is suspected.

• The most common causes of spontaneous intracranial bleeds in children include:
  – AVMs, aneurysm, moyamoya, tumor, hematologic disorder, and cavernoma

• Brain Arteriovenous Malformations
  – AVMs are rare congenital vascular malformations that most often present with intraparenchymal hemorrhage, especially in children.
  – Cerebral angiography is the gold standard of diagnosis for brain AVMs and can guide treatment and is useful for follow-up.
  – Treatment options for AVM include surgical resection, radiosurgery and endovascular embolization.
References

• Bonthius DJ, Lee AG. Approach to child with headache. UpToDate Article.
References continued


Acknowledgements

Behroze Vaccha, MD for her generous offering of time and assistance with image acquisition and film review.

Gillian Lieberman, MD for leading our core radiology clerkship and her outstanding teaching.

Emily Hanson for all her assistance and organization for the radiology core clerkship.

My wonderful classmates for making this rotation so much fun!