Moyamoya disease:
An unusual presentation of stroke

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Dr. Gillian Lieberman

Image from http://www.sensitivelight.com/smoke2/
1. **Index patient:** we will follow a patient with an unusual presentation of stroke.

2. Our patient’s story will be our basis to review several *broad topics in neuroradiology*, including:
   - Common imaging modalities used to evaluate intracranial hemorrhage
   - Interpretation of radiologic findings in hemorrhagic stroke
   - Differential diagnosis of intracranial hemorrhage based on radiologic appearance

3. After we have discussed those broad topics, we will **return to our patient** to narrow down the differential diagnosis based on her clinical history and learn how radiology is used in our patient’s particular diagnosis, treatment, and follow-up.
Our patient: clinical presentation

**CC:** Our patient is a 44 year-old Vietnamese woman who woke up at 9:00 am unable to move her left side.

**HPI:** She was in her usual state of health at 8:30 pm the night before. She went to sleep and woke up the next morning with left-sided paralysis. She and her family denied any recent trauma, drug use, or recent sexual activity.

**PMH:** None.

**Meds:** None. No oral contraceptives or over-the-counter medications. No drug allergies.

**SH:** No drugs, alcohol, smoking, recent sexual activity. Urine toxicology screen was negative.

**ROS:** No headache, loss of vision, shortness of breath, chest pain, recent illness.
Our patient: clinical presentation

**Vitals:** T 98, HR 60, BP 101/50, RR 15, O2 100%

**Pertinent features of her physical exam:**
- Decreased strength in left face, arm, and leg
- Decreased sensation of left face, arm, and leg
- Reflexes brisker L>R
- Babinski sign positive (upgoing toes) on left, negative (downgoing toes) on right
What next?

Based on our patient’s presentation - acute onset of left-sided weakness and sensory loss - we highly suspect a stroke in her right brain.
Neuroimaging guides clinical decisions

Step 1: Determine whether there is an acute hemorrhage with head CT

Suspect stroke?
- Make sure patient’s airway, blood pressure, circulation, and neurological status are not rapidly deteriorating

Hemorrhagic?
- Avoid tPA administration

Will it need to be evacuated?

What is the etiology of the bleed?

Is there an associated infarction?

Within the time window for thrombolysis with tPA?

Risks and benefits of medical or mechanical thrombolysis?

Other treatments – Anticoagulation? Monitoring?

Workup to determine treatment plan: etiology, comorbidities, extent of damage

Ischemic?

What portion is infarcted?
- What portion is at risk? Is it salvageable?

Workup for tumor, encephalitis, other stroke mimics

Stroke mimic?
Non-contrast head CT: a good initial study to detect acute intracranial hemorrhage

- Faster and more widely available than MRI

- 20% patients have contraindications to MRI (can’t hold still; have pacemakers, etc.) Singer OC, Sitzer M, duMensil de Rochemont R, Neumann-Haeffelin T. Practical limitations of acute stroke MRI due to patient-related problems. Neurology 2004; 62:1848-9.

- Appearance of hemorrhage depends only the degree that x-rays are attenuated by blood products

- Factors that influence the degree of x-ray attenuation include:
  - Hematocrit
  - Degree of clot retraction
  - Hemoglobin content

- Hemorrhages remain hyperdense for approximately 1 week, then fade
  - Notable exception: anemic patients with hemoglobin <8%; hemorrhages may be isodense
Our patient’s initial non-contrast head CT (C-CT)

<table>
<thead>
<tr>
<th>We can be less suspicious of:</th>
<th>Appearance on NCCT:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute/subacute infarction only</td>
<td>Isodense</td>
</tr>
<tr>
<td>Chronic infarction only</td>
<td>Hypodense</td>
</tr>
<tr>
<td>Tumor only</td>
<td>Hypodense</td>
</tr>
<tr>
<td>Infection only</td>
<td>CT: hyperdense on CT with contrast</td>
</tr>
</tbody>
</table>

Recent hemorrhage (hyperdense) with surrounding vasogenic edema (hypodense)

Axial C-Head CT
What caused our patient’s intracranial hemorrhage?

- On non-contrast head CT, we saw focal hyperdense lesion in the region of the right thalamus and internal capsule. It was surrounded by a hypodense rim. These findings were consistent with an acute hemorrhage (high x-ray attenuation) with surrounding vasogenic edema (low x-ray attenuation).

- We saw a subtle midline shift, but both her symptoms and follow-up non-contrast head CTs remained stable over time, so mass effect and herniation were not an urgent concern.

- Now, the most important question to determine our treatment plan is: what caused our patient’s hemorrhage?

- Let’s move on to generate a differential diagnosis for the hemorrhage we saw on our patient’s head CT.
Differential diagnosis of hemorrhage on head CT: location matters  
(All images below are axial C-head CTs)

Intraparenchymal:
Pathology of small penetrating arteries and arterioles

Epidural:
Pathology of meningeal vasculature

Subdural:
Pathology of bridging veins

Subarachnoid:
Pathology of Circle of Willis

Our patient has an intraparenchymal hemorrhage. What is the differential diagnosis of intraparenchymal hemorrhage?
Differential diagnosis of intraparenchymal hemorrhage
(All images below are axial C- head CTs)

1. **Primary - spontaneous (80%)**:
   1. Most commonly associated with hypertension; often seen in deep gray matter
      Our patient has always had low blood pressure (90s/50s).
   2. Is also associated with cerebral amyloid angiopathy in the elderly; more often seen in lobar regions
      Our patient is young (44 years old).

2. **Secondary to other pathologies (20%)**:
   1. Hemorrhagic transformation of ischemic stroke
   2. Infection
      Our patient had no clinical signs of infection.
   3. Cerebral venous or sinus occlusion
   4. Trauma
      Our patient experienced no trauma prior to symptom onset.
   5. Drug use (i.e. sympathomimetics)
      Our patient had a negative urine toxicology screen.
   6. Tumor (primary or metastasis)
   7. Vascular malformations
   8. Rupture of aneurysms
   9. Coagulopathy
      Our patient had normal labs and no history of coagulopathy.

What imaging modality will allow us to differentiate between these diagnoses?

MRI: a great way to work up an atypical hemorrhage

- Non-invasive

- The appearance of hemorrhage depends upon two factors
  - Magnetic state of blood and breakdown products
  - MRI settings used to generate images (magnetic field strength, pulse sequence used)

- Manipulating the MRI settings can highlight acute hemorrhage (T2-weighted Gradient Echo (GRE) pulse sequence) or changes in the hemorrhage over time

- Manipulating MRI settings also allows us to selectively highlight different tissues to evaluate:
  - Structure of brain parenchyma (T1 images)
  - Edema and infarct (T2 FLAIR, Diffusion weighted images)
  - Vasculature (Axial 3D Time of Flight)
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# A comparison: the radiologic appearances of intracranial hemorrhage on CT and MRI

<table>
<thead>
<tr>
<th>Stage</th>
<th>Pathogenesis</th>
<th>CT</th>
<th>T1 MRI</th>
<th>T2 MRI</th>
<th>T2 Fluid-attenuated inversion recovery (FLAIR) MRI</th>
<th>T2 Gradient-weighted Echo (GRE) MRI</th>
</tr>
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<tbody>
<tr>
<td>Hyperacute (&lt;12 h)</td>
<td>Oxygenated intracellular hemoglobin. Development of clot Clot retracts; serum/reactive edema surrounds it</td>
<td>Hyperdense Density increases</td>
<td>Isointense or mildly hyperintense</td>
<td>Hyperintense Hyperintense</td>
<td>Hypointense Hyperintense Hypointense rim</td>
<td></td>
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<tr>
<td>Acute (12 hrs –2 days)</td>
<td>Deoxygenated intracellular hemoglobin</td>
<td>Hyperdense</td>
<td>Isointense or hypointense</td>
<td>Hypointense Hypointense Hypointense</td>
<td>Hypointense rim</td>
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<tr>
<td>Early subacute (2 –7 days)</td>
<td>Intracellular hemoglobin $\rightarrow$ Intracellular methemoglobin</td>
<td>Hyperdense</td>
<td>Hyperintense</td>
<td>Hypointense Hypointense Hypointense</td>
<td>Hypointense Hypointense</td>
<td></td>
</tr>
<tr>
<td>Late subacute (8 days –1 mo)</td>
<td>RBC lysis $\rightarrow$ Extracellular methemoglobin Intracellular methemoglobin $\rightarrow$ Extracellular methemoglobin Vasogenic edema slowly subsides</td>
<td>Decreasing density until isodense</td>
<td>Hyperintense Hyperintense</td>
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<td>Hypointense</td>
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<tr>
<td>Chronic (&gt;1 mo – years)</td>
<td>Macrophages, astrocytes surround and phagocytose hematoma. Extracellular methemoglobin stored in macrophages; converted into hemosiderin and ferritin. Hematoma resolve completely, leaves fluid-filled cyst or collapsed brain defect.</td>
<td>Hypodense hematoma Hypodense defect</td>
<td>Hypointense Hypointense Hypointense</td>
<td>Hypointense Hypointense Hypointense</td>
<td>Slit-like hyperintense/isointense core, surrounded by hypointense rim</td>
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T2 Gradient Echo (GRE) Susceptibility MRI can detect acute hemorrhage

Our patient’s acute hemorrhage appears to have a hypointense rim on T2 GRE MRI. The dark rim will progress toward the center over time.

Serum and vasogenic edema appear bright on T2 weighted MRI images.

CSF appears bright on T2 weighted MRI images.
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T1 MRI produces good images for evaluating brain parenchyma (Images below are sagittal T1 weighted sagittal MRIs)

The edema surrounding our patient’s hemorrhage is visualized in her right thalamus.

CSF appears dark on T1 weighted MRI images.

Cut through left thalamus

Cut through right thalamus
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MR Angiogram
(Image below: axial 3D Time of Flight MRI)

There is increased vascularity around our patient’s hemorrhage (right thalamus), compared to her left thalamus.

Does the asymmetry in our patient’s vasculature concern you?

Let’s orient ourselves to cerebral vasculature and take a closer look at our patient’s vessels.
Normal MR Angiogram

Anterior cerebral artery (ACA)

Middle cerebral artery (MCA)

Internal carotid artery (ICA)

Vertebral artery

Posterior cerebral artery (PCA)

Basilar artery

Anterior circulation

Posterior circulation

Patient 6
PACS, BIDMC

Image from www.mdconsult.com
Normal MR Angiogram

Anterior cerebral artery (ACA)
Middle cerebral artery (MCA)
Internal carotid artery (ICA)
Posterior cerebral artery (PCA)
Basilar artery
Vertebral artery

Anterior circulation

Posterior circulation

Patient 6
PACS, BIDMC

Normal

Our patient

- Internal carotid artery (ICA)
- Middle cerebral artery (MCA)
- Anterior cerebral artery (ACA)
- Basilar artery
- Posterior cerebral artery (PCA)
- Vertebral artery

Stenosis of the right ICA

ACA not visualized?

Abnormally robust, tortuous posterior cerebral arteries

MCA not visualized?
MR angiogram: sagittal view of the right internal carotid artery

Normal

Our patient

Anterior cerebral artery (ACA)
Middle cerebral artery (MCA)
Internal carotid artery (ICA)
Point where ICA enters cranium over the petrous bone

Abnormal MCA and ACA
Artifact: portion of the basilar artery was not subtracted from this image
ICA stenosis

Patient 6
PACS, BIDMC

Index patient
PACS, BIDMC
We need a better look at the vessels.

- Increased vascularity seen near hemorrhage?
- Narrowing of MCA/ACA lumens?
- Abnormally robust posterior circulation?
- What’s going on?
Menu of tests to further evaluate vasculature

- **CT Angiogram**
  - Less invasive
  - CTA > MRA in detailed vessel imaging of Circle of Willis and tributaries

- **Conventional angiogram**
  - Indicated in stroke patients without clear underlying etiology or patients with hemorrhages in unusual locations (like our patient CN)
  - Angiogram > CTA – visualization distal vasculature and subtle findings in the deep gray matter or brainstem.
Menu of tests to further evaluate vasculature

- Conventional angiogram
  - Indicated in stroke patients without clear underlying etiology or patients with hemorrhages in unusual locations (like our patient CN)
  - Angiogram > CTA – visualization distal vasculature and subtle findings in the deep gray matter or brainstem.
Conventional angiogram

Contrast is injected into the artery of interest. Radiographs are taken sequentially to visualize the vessel of interest and its branches as the contrast flows through them.

In this image, contrast was injected into the internal carotid artery (ICA) and a radiograph was taken while contrast was still in the arteries (arterial phase). We can see its branches, the anterior and middle cerebral arteries, perfusing their respective brain territories.
Our patient’s **right common carotid artery** angiogram – sagittal view

**Normal**

- Internal carotid artery (ICA)
- Middle cerebral artery (MCA)
- Anterior cerebral artery (ACA)

**Our patient**

- Branches of the middle meningeal artery – supplied by the external carotid artery
Our patient’s **right common carotid artery angiogram** – coronal view

**Our patient**

- Abnormally robust middle meningeal artery circulation (branch of external carotid)
- Abnormal middle cerebral artery (MCA) circulation
- Anterior cerebral artery (ACA) not visualized

**Normal**

- Normal MCA circulation
- ACA visualized
Our patient’s angiogram findings give us her diagnosis.

Abnormally robust middle meningeal artery circulation (branch of external carotid)

Abnormal middle cerebral artery (MCA) circulation

Anterior cerebral artery (ACA) not visualized

Normal MCA circulation

ACA visualized
Diagnosis: **Moyamoya disease**

(Official name: “Spontaneous occlusion of the circle of Willis” Not nearly as cool, therefore, less used.)

*Moyamoya*: the Japanese word for “something hazy, just like a puff of cigarette smoke drifting in the air”
Diagnosis: **Moyamoya disease**

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*Moyamoya*: the Japanese word for “something hazy, just like a puff of cigarette smoke drifting in the air”
Moyamoya disease: definition

- **Moyamoya disease:**
  An idiopathic cerebrovascular condition that predisposes affected patients to stroke, defined by two characteristics:
  1. Progressive stenosis of the intracranial internal carotid arteries

- **Moyamoya syndrome:**
  Stenosis of intracranial internal carotid arteries and development of collateral vasculature secondary to other conditions. These conditions include:
  - Radiotherapy to the head or neck
  - Down’s syndrome
  - Neurofibromatosis type 1
  - Sickle cell disease
Moyamoya disease becomes more severe over time. Categorization of the severity of moyamoya depends upon the degree to which the intracranial ICA has become stenosed. The greater the occlusion of the ICA, the higher the grade.

A. Normal lateral-projection ICA angiogram

B. Suzuki Grade I-II - Narrowing of internal carotid artery

C. Suzuki grade III-IV – Significant narrowing of ICA and small, capillary “puff of smoke” collaterals

D. Suzuki grade V-VI – Obliteration of ICA flow. Cortical perfusion is entirely supplied by posterior circulation (basilar artery) and collateral vessels of the external carotid artery.

For diagnosis and grading of moyamoya, conventional cerebral angiogram is the gold standard.
Our patient has stage IV-V moyamoya disease: severe stenosis of her right ICA with collateral meningeal circulation.
Our patient developed **collateral circulation via her middle meningeal artery** (branch of external carotid artery).
Our patient also developed robust collateral vasculature via her posterior circulation.
Moyamoya disease: symptoms

1. **Progressive stenosis of intracranial internal carotid arteries**

   - **Decreased flow to brain → symptoms of ischemia**
     - Ischemic stroke
     - Transient ischemic attacks
     - Seizures

2. **Collateral vasculature develops**

   - **Increased flow rate through collateral vasculature → symptoms of abnormal flow**
     - Aneurysm development
       - Turbulent flow through vessels
     - Hemorrhage
       - High flow through small fragile collateral vessels
     - Headache
       - Stretching of pain-sensitive meningeal vasculature
Moyamoya disease: epidemiology

• More common in southeast Asia than elsewhere
  □ Most common pediatric cerebrovascular disease in Japan (3/100,000)
  □ European incidence approx 1/10 that of Japan

• Female:Male ratio nearly 2:1

• Bimodal peaks in incidence
  □ Children approx 5 years old
  □ Adults in mid-40s

• Adults are more likely to present with hemorrhage (42%) than children (2.8%)
Moyamoya disease: \textit{treatment}

Goal of treatment: prevent future ischemic strokes.

\textbf{Revascularization surgery}: make collateral circulation more robust to prevent future ischemic events; i.e., increase the responsibilities of the external carotid artery

1. \textbf{Direct}: Anastamose a branch of the internal carotid artery with a branch of the external carotid artery to bypass ICA stenosis
Moyamoya disease: treatment

Internal carotid artery – superficial temporal artery (STA) anastamosis

In this drawing, the superficial temporal artery (STA), which is a branch of the external carotid artery, is anastomosed to the cortical conducting branches of the internal carotid artery.

This allows blood from the external carotid artery to flow past an intracranial ICA stenosis and enter cerebral branches of the ICA that are not receiving blood.
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1. Direct: Anastamose a branch of the internal carotid artery with a branch of the external carotid artery to bypass ICA stenosis

2. Indirect: Vascularized tissue supplied by external carotid artery is placed contact with the brain → new ingrowth of blood vessels
   1. Pial synangiosis (STA-pia)
   2. Encephaloduroarteriosynangiosis (STA-dura)
   3. Encephalomyoarteriosynangiosis (Temporal muscle-dura)
Moyamoya disease: treatment

Pial synangiosis

In pial synangiosis, the superficial temporal artery (STA) is placed underneath the arachnoid mater, directly upon the pia. The goal of this treatment is for vessel growth into the brain so that the external carotid artery can supply brain parenchyma via the STA. It is commonly used in children whose vessels are too small for direct ICA-STA anastamosis.
Moyamoya disease: treatment

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Outcome of revascularization surgery: Areas of brain underperfused by MCA and ACA (branches of ICA) will be supplied by collateral vessels that develop from branches of the external carotid artery (ECA).
The posterior branch of the superior temporal artery ( Older ) was placed upon the pia during pial synangiosis.

One year after surgery, we see a threefold dilatation of vessels supplied by external carotid artery.

We also see that cerebral blood flow to entire hemisphere is supplied by external carotid artery (instead of the stenosed internal carotid artery) via the craniotomy site.
Moyamoya disease: treatment

Revascularization surgery can help patients with moyamoya develop collateral vasculature when their internal carotid arteries are stenosed. This reduces the risk of symptoms from ischemic stroke.
Moyamoya disease: treatment

Benefits:

Reduction of ischemic symptoms
A meta-analysis of 1156 moyamoya stroke patients who underwent revascularization surgery showed that 87% of patients with moyamoya had symptomatic benefit from surgical revascularization. Indirect, direct, and combined techniques were equally effective.

Reduction of symptomatic progression:
Moyamoya disease progresses in severity in most patients. Up to 67% of patients with moyamoya have symptomatic progression over a 5-year period, and the outcome is poor (severe stroke) without treatment. In contrast, an estimated 2.6% of patients have symptomatic progression after surgery.


Risks:

The risk of stroke is highest during the first 30 days after surgery (4%/hemisphere).

Risk decreases considerably after the first month. After the first month, patients have a 96% probability of remaining stroke free over the next 5-years.

What happened to our patient?

- Our patient is being followed by neurology. She is regaining strength and sensation, but still has some weakness on her left side.

- She will consult with neurosurgery to weigh the risks and benefits of having revascularization surgery when she already has collateral vasculature.
Summary

- Radiology is essential for the diagnosis, treatment, and follow-up of stroke
- In this presentation, we focused on the imaging of intracranial hemorrhage:
  - Discussed common imaging modalities used to diagnose and work-up intracranial hemorrhage and saw examples of hemorrhage on CT and MRI (T1, Gradient Echo (GRE) susceptibility, and 3D Time of Flight)
  - Learned that the appearance of hemorrhage on CT is dependent upon the density of the blood/clot, and the appearance on MRI is dependent upon both (1) the paramagnetic properties of the blood and its breakdown products and (2) the MRI settings
  - Learned that the differential diagnosis for intracranial hemorrhage is different based on location of hemorrhage, and saw examples of each:
    - Intraparenchymal
    - Subarachnoid
    - Subdural
    - Epidural
  - Discussed the differential for intraparenchymal hemorrhage, which is divided into primary (spontaneous) and secondary (to other pathologies). The differential can be narrowed based on the hemorrhage’s location within the parenchyma and patient’s clinical history
- We followed the course of a patient with moyamoya disease who presented with intraparenchymal hemorrhage
  - Learned that moyamoya disease is a rare idiopathic cerebrovascular disease that predisposes patients to ischemic and hemorrhagic stroke; moyamoya syndrome is secondary to other causes
  - Learned that the gold standard for diagnosis of moyamoya is conventional angiogram
  - Learned the defining features, epidemiology, treatment, and follow up of moyamoya
  - Saw examples of MR angiograms and conventional angiograms of patients with normal vessels and patients with moyamoya disease prior to and after revascularization surgery
Many thanks for your wonderful teaching, suggestions, and help.

- Dr. Aarti Sekhar
- Dr. Gul Moonis
- Dr. R. Michael Scott
- Dr. D. Eric Searls
- Maria Levantakis
- Dr. Gillian Lieberman
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