Imaging of Cerebrovascular Disease in Sickle Cell Patients

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Our Patient

- 3 year old girl with Hg SS Sickle Cell Disease
- History of splenic sequestration crisis at 7 months, s/p splenectomy
- Multiple admissions for pain crises, infections
- No history of acute chest
- Developmentally normal, no hx of neurological issues
- Baseline HCT 20-25
The Problem: SCD and Cerebrovascular Disease

- 0.61-0.76% risk of CVA per year
- 300 times > age-matched population
- 46% of children with Hg SS Disease have brain injury visible on MRI
- 64% have vasculopathy visible on MRA
- Both large and small vessels are affected

Source: Uptodate online: “Cerebrovascular Disease in Sickle Cell Disease”
Menu of Tests to Image Cerebrovascular Disease

- Trans-cranial Doppler Ultrasound
- MRA
- Angiography
- MRI
- CT
Trans-Cranial Doppler

- Measures blood velocity in large intra-cranial vessels (MCA and distal internal carotid artery)
- Velocity inversely proportional to vessel diameter squared
- >170 cm/sec considered abnormal
- RR of stroke is 44 in patients with abnormal TCD
- TCD Screening is standard of care for all SC patients < 16

Source: http://www.hemodynamic.com/edu.html
TCD screening is standard of care!

- STOP Trial: 130 patients with TCD velocity >200 cm/sec randomized to standard care or transfusion
  - Relative risk reduction = 90%
  - Absolute risk reduction = 9%

Data from Adams, RJ, McKie, VC, Hsu, L et al, N Engl J Med 1998; 339:5
Our Patient’s Screening TCD

RT MCA velocity = 214 cm/sec

Courtesy of Dr. Ellis Neufeld and Dr. Karen Lee, Children’s Hospital, Boston
Our Patient’s Screening TCD

Left MCA velocity = 241 cm/sec

Courtesy of Dr. Ellis Neufeld and Dr. Karen Lee, Childrens’ Hospital, Boston
TCD Results

- Abnormal screening TCD bilaterally
- Patient receives transfusions to reduce risk of stroke, with goal of Hg SS concentration < 30% of total Hg.
- MRA and MRI ordered to look for further neurological disease
MRA Findings

“Significant drop-off of signal intensity in the proximal MCA and ACA segments with the right greater than left. There is decreased filling of the middle cerebral artery vessels on the right diffusely. The caliber of bilateral internal carotid arteries is diminished which is again more apparent on the right.”

Courtesy of Dr. Ellis Neufeld and Dr. Karen Lee, Childrens’ Hospital, Boston
MRI Results: T2 Weighted Images

Courtesy of Dr. Ellis Neufeld and Dr. Karen Lee, Childrens’ Hospital, Boston
MRI Results: T2 Weighted FLAIR Images

SE: 9
IM: 17
x: 31.06

IR
TR: 10002
ETL: 0
TE: 169.88
EC: 1
HFS
8HR BRAIN
SP: 6
THK: 5
FA: 90
NEX: 0.5
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MRI Results: DWI and ADC

Images

Bright on DWI

Dark on ADC

Courtesy of Dr. Ellis Neufeld and Dr. Karen Lee, Childrens’ Hospital, Boston
MRI Results

- Abnormal increased T2 signal intensity in right corona radiata and right frontal white matter
- Area of high signal intensity in right posterior parietal region
- Vascular changes consistent with Moyamoya disease: severe bilateral stenosis or occlusion of the arteries around the circle of Willis with prominent collateral circulation.
- No evidence of acute infarction

**Diagnosis: Multiple Silent Infarcts**
Spectrum of Cerebrovascular Disease in Sickle Cell

- Vasculopathy
- Moyamoya
- TIA
- Silent Infarct
- Ischemic Stroke
- Hemorrhagic Stroke
- Recurrent Stroke
Subtle Right Temporo-Parietal Changes in a 7 year old Girl

Right distal ICA, Proximal MCA and ACA Stenosis on Angiography

CT: L. Caudate Hematoma & Intraventricular Hemorrhage

Source: Uptodate Online, “Moyamoya disease.”
Extensive left MCA infarct in 13 year old boy with SCD
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