Posterior Reversible Encephalopathy Syndrome

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Outline

- Case
- Brief overview of PRES
- Typical Imaging features with anatomy review
- Imaging Differential Diagnosis
Case: Presentation

• 41 year old female presented to the ED with headache, blurred vision, and unclear left leg weakness

• Recent history of falls and vertigo

• Possible seizure episodes consisting of urinary incontinence and “foaming at the mouth” over the past 5 days
Case

- Physical exam: limited due to lack of patient effort and functional overlay.

- Pertinent positives: Subjective left leg weakness; Anion gap acidosis on routine labs

- Vital signs: T: 97 HR: 89-130 RR: 16 SaO2: 95% on room air.

- BP was 128/75 upon admission, but later spiked to 200/100 requiring transfer to the MICU.
Case: Differential

- Infectious—encephalitis/meningitis
- Ischemic/Infarct
- Trauma/hemorrhagic
- Cerebral vein thrombosis
- CNS vasculitis
- Demyelinating condition
- Malignancy
- …the list goes on…. 
Case: Imaging

• Step 1: rule out acute hemorrhage!
Imaging—Non-Contrast Head CT

- No evidence of acute bleed
- No evidence of evidence of fracture
- No midline shift or evidence of intracranial hypertension
- Multiple subcortical white matter hypoattenuating regions involving the occipital, parietal, and frontal lobes
Differential Diagnosis

- Trauma/hemorrhagic
- Ischemic/Infarct
- Infectious—encephalitis/meningitis
- Cerebral vein thrombosis
- CNS vasculitis
- Demyelination
- Malignancy
- ...
Imaging

• Step 1: rule out acute hemorrhage with non-contrast head CT
• Step 2: More refined information with MRI
FLAIR

Hyper-intensities in the bilateral occipital, bilateral posterior frontal, posterior right cingulate, and bilateral basal ganglia. Most likely diagnosis—PRES.

Images from PACS, BIDMC
PRES: Overview

- Posterior Reversible Encephalopathy Syndrome
- Does not have to be posterior; not always reversible
- Two theories as to the pathophysiology of PRES
  - Acute increase in blood pressure \( \rightarrow \) Hyper-perfusion
  - Endothelial dysfunction \( \rightarrow \) Hypo-perfusion \( \rightarrow \) reflex hypertension
- Believed that edema is concentrated in the posterior hemispheres due to decreased autoregulation of blood pressure and sympathetic tone in the posterior fossa

Petrovic et al. 2011
PRES: Aggravating factors

- Sudden increases in blood pressure
- Immunosuppressant drugs
- Renal Failure
- Certain Chemotherapies
- Autoimmune disorders (50% of people with PRES)

Rykken & McKinney 2014; Petrovic et al. 2011
Symptoms of PRES

- Headache
- Visual Disturbances
- Confusion
- Tonic-Clonic Seizures
- Focal Neurologic Deficits
Diagnostic Complications

- Clinical criteria is non-specific
- No gold standard with only retrospective information
- Diagnosis does change management
- This makes imaging extremely important!
Typical Imaging Features of PRES

• Bilateral vasogenic edema in the parietal-occipital regions

• Affects both the cortical and the sub-cortical regions

• Bright on T2 sequences, FLAIR is the most sensitive.

• Contrast enhancement is inconsistent

• Hallmark ➔ imaging changes are reversible

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Petrovic et al. 2011
Typical Imaging Features of PRES

T1: Hypointense
T2: Hyperintense
FLAIR: Hyperintense
CT: Hypoattenuating

All images from PACS, BIDMC
Follow-up imaging: our patient

Original CT: 12/2015
Follow up: 12/2016

Resolution of prior imaging findings!

All images from PACS, BIDMC
Alternative Imaging Features

• Main distribution patterns
  – 1. Parietal-occipital bilaterally
  – 2. Watershed distribution over the full hemisphere
  – 3. Superior frontal sulcus
Alternative Imaging Features

Parietal Orbital Pattern

Image obtained by Petrovic et al. 2011
Alternative Imaging Features

Watershed Distribution

Image obtained by Petrovic et al. 2011
Alternative Imaging Features

Superior Frontal Sulcus Pattern

Image obtained by Petrovic et al. 2011
Best Imaging Modalities for PRES

• No strict criteria due to limited research
  – MRI FLAIR is considered the most sensitive
  – Many patients receive a CT because of acute onset headache
  – Additional imaging (such as CTV or MRV) may be necessary depending on the specific clinical presentation

Petrovic et al. 2011
Differential Diagnosis of Imaging

- Trauma/hemorrhagic  ➔ Rule out with CT
- Ischemic/Infarct
- Cerebral vein thrombosis
- CNS vasculitis
- Progressive Multifocal Leukencephalopathy
- Autoimmune encephalitis: Acute disseminating encephalomyelitis
- Malignancy: Gliomatosis cerebri

Fugate & Rabinstein, 2015; Hugonnet et al. 2013
Differential Diagnosis of Imaging

Infarct

• Ways to differentiate
  – Vascular territories
  – DWI restriction

Hugonnet et al. 2013
Differential Diagnosis of Imaging

Infarct

Our Patient
PACS, BIDMC

Vascular Territories
 Obtained from www.radiopedia.com

PRES is less likely to fall in a strict vascular distribution
Differential Diagnosis of Imaging

Infarct

PRES on DWI
Obtained from Covarrubias, Luetmer, and Campeau 2002

Infarct on DWI
Dr. Gagandeep Choudhary
https://radiopaedia.org/cases/left-middle-cerebral-artery-territory-infarct

PRES is not diffusion restricted on DWI
Differential Diagnosis of Imaging

- Trauma/hemorrhagic → Rule out with CT
- Ischemic/Infarct → Vascular distribution and DWI
- Cerebral venous thrombosis
- CNS vasculitis
- Progressive Multifocal Leukencephalopathy
- Autoimmune encephalitis: Acute disseminating encephalomyelitis
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Fugate & Rabinstein, 2015; Hugonnet et al. 2013
Differential Diagnosis of Imaging
Cerebral Venous Thrombosis

• Symptoms: vague and similar to PRES
  – Headache
  – Nausea
  – Seizures

• MRI Imaging
  – Can develop edema and show a similar imaging pattern to PRES
  – Should not be bilateral, may show hemorrhage
  – Magnetic resonance venography or CTV can confirm diagnosis if unclear

Petrovic et al. 2011
Differential Diagnosis of Imaging
Cerebral Venous Thrombosis

PRES on T2 MRI
Our Patient
PACS, BIDMC

PRES on FLAIR MRI
Obtained from Stevens and Haran, 2012

CVT on T2 MRI
Obtained from Saposnik et al. 2011
Differential Diagnosis of Imaging

- Trauma/hemorrhagic → Rule out with CT
- Ischemic/Infarct → Vascular distribution and DWI
- Cerebral venous thrombosis → MRV or CTV
- CNS vasculitis
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Fugate & Rabinstein, 2015; Hugonnet et al. 2013
Differential Diagnosis of Imaging

- **CNS vasculitis**
  - Infarcts in different vascular territories
  - Will not be reversible
  - MRV/CTV to confirm

- **Progressive Multifocal Leukencephalopathy**
  - Spares the cortex
  - Would not be reversible

- **Acute Disseminating Encephalomyelitis (ADEM)**
  - Also has bilateral T2 and FLAIR hyperintense lesions
  - Follows clinical signs of infection

- **Gliomatosis Cerebri**
  - Tends to be more asymmetric
  - Would not be reversible

Hugonnet et al. 2013
Follow-up

• Remove aggravating factors
  – Control blood pressure
  – Take off potentially causal drugs in the acute setting
  – Treat associated conditions (i.e. renal disease)
  – Treat seizures with anti-epileptics
Prognosis

- Generally a good prognosis
- Complications include
  - Intracranial and subarachnoid hemorrhage (10-25%)
  - Brainstem compression
  - Increased intracranial pressure
- Mortality rates are variable due to the unclear diagnostic criteria for PRES
Summary

• PRES is an uncommon, but important clinical entity

• Diagnostic presentation is inconsistent and vague, giving imaging a greater role in diagnosis

• If PRES is treated it can be entirely reversed, however delayed treatment can lead to permanent neurologic injury and death
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Imaged obtained from http://amaranthinesplendour.blogspot.com/2013/02/analyzing-brain-ct.html
References


