AMSER Case of the Month December 2019

71 yo F presents with altered mental status, facial droop, extremity weakness, and left homonymous hemianopsia on a background of hypertension

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Patient Presentation

- HPI: 71 year old female presents with new onset altered mental status, left sided facial droop, and limb weakness.
  -Admitted one month prior for recurrent atrial fibrillation and underwent ablation.
  -Hospital course complicated by bacteremia secondary to esophageal perforation and pulmonary vein thrombosis.
- PMH: Hypertension, dyslipidemia, paroxysmal atrial fibrillation, aortic insufficiency
- PSH: Cardiac ablation for atrial fibrillation, stenting for esophageal perforation
- PE:
  - Right upper limb weakness, bilateral lower limb weakness
  - Left homonymous hemi-anopsia
- A non-contrast CT of the head on the day of presentation was negative, but neurological manifestations continued to Day 2 of admission.

What Imaging Should We Order?

What Imaging Should We Order, one day after the negative CT?

Variant 4: **New focal neurologic defect, fixed or worsening. Longer than 6 hours. Suspected stroke.**

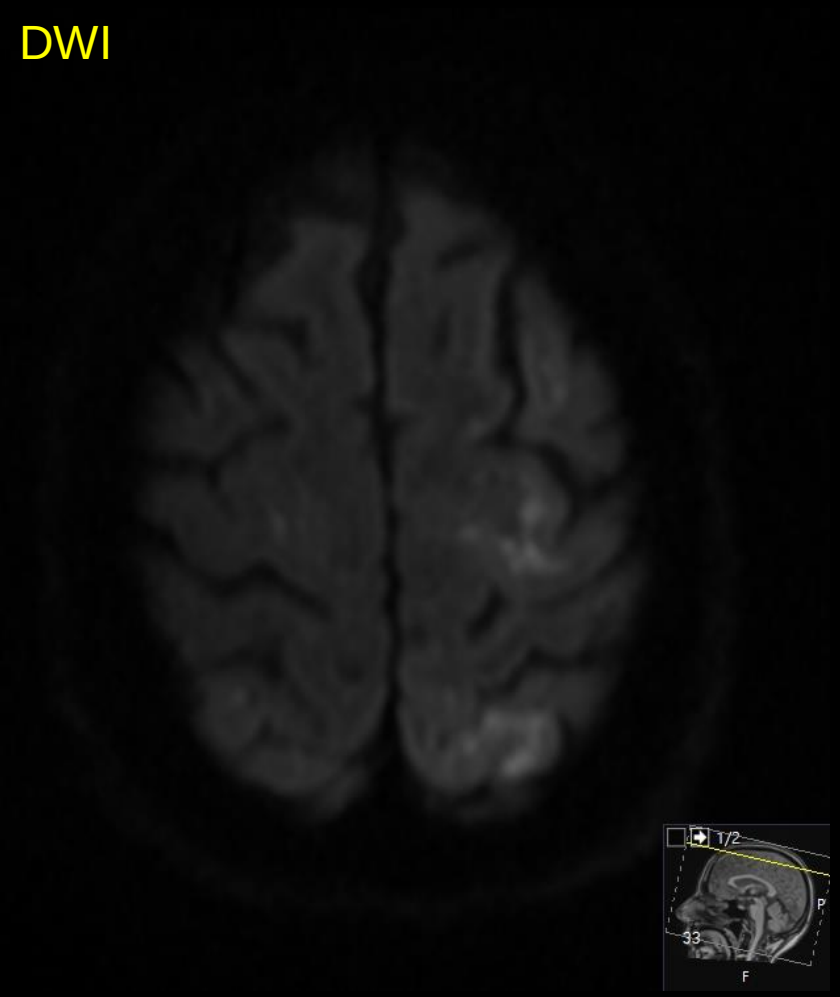
Radiologic Procedure	Rating	Comments	RRL*
MRI head without IV contrast	8	Parenchymal brain imaging and CT or MR vascular imaging of the head and neck should be considered. Noncontrast head CT is often obtained first to assess for hemorrhage or large infarct. Can be useful if there is a contraindication to contrast. MRI is more sensitive than CT for acute infarct.	O
MRI head without and with IV contrast	8	Parenchymal brain imaging and CT or MR vascular imaging of the head and neck should be considered. Noncontrast head CT is often obtained first to assess for hemorrhage or large infarct. MRI is more sensitive than CT for acute infarct.	O
MRA head and neck without IV contrast	8	Can be obtained in conjunction with MRI head. Preferred MR vascular imaging of the head and neck includes noncontrast head MRA and contrast-enhanced neck MRA. May be useful in patients with renal failure or contrast allergies.	O
MRA head and neck without and with IV contrast	8	Can be obtained in conjunction with MRI head. Preferred MR vascular imaging of the head and neck includes noncontrast head MRA and contrast-enhanced neck MRA.	O
		Noncontrast head CT is often obtained	

Obtained after no improvement in neurological manifestations.

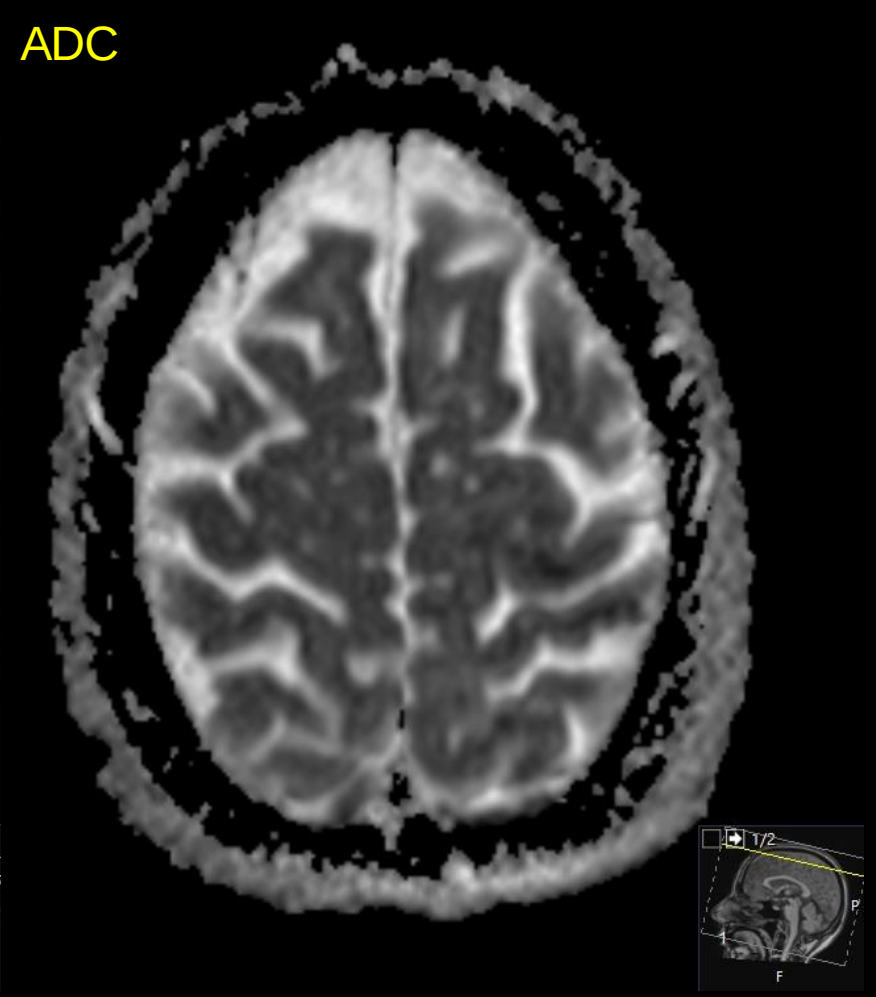
American College of Radiology.

MRI Findings (unlabeled)

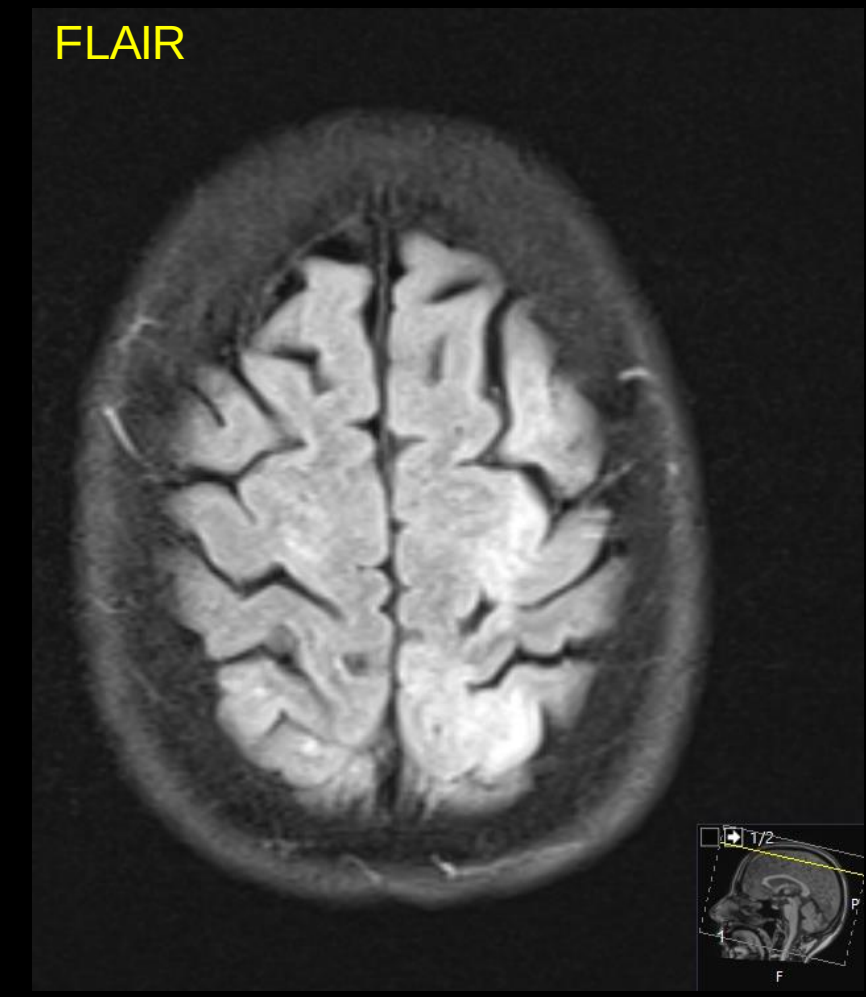
DWI



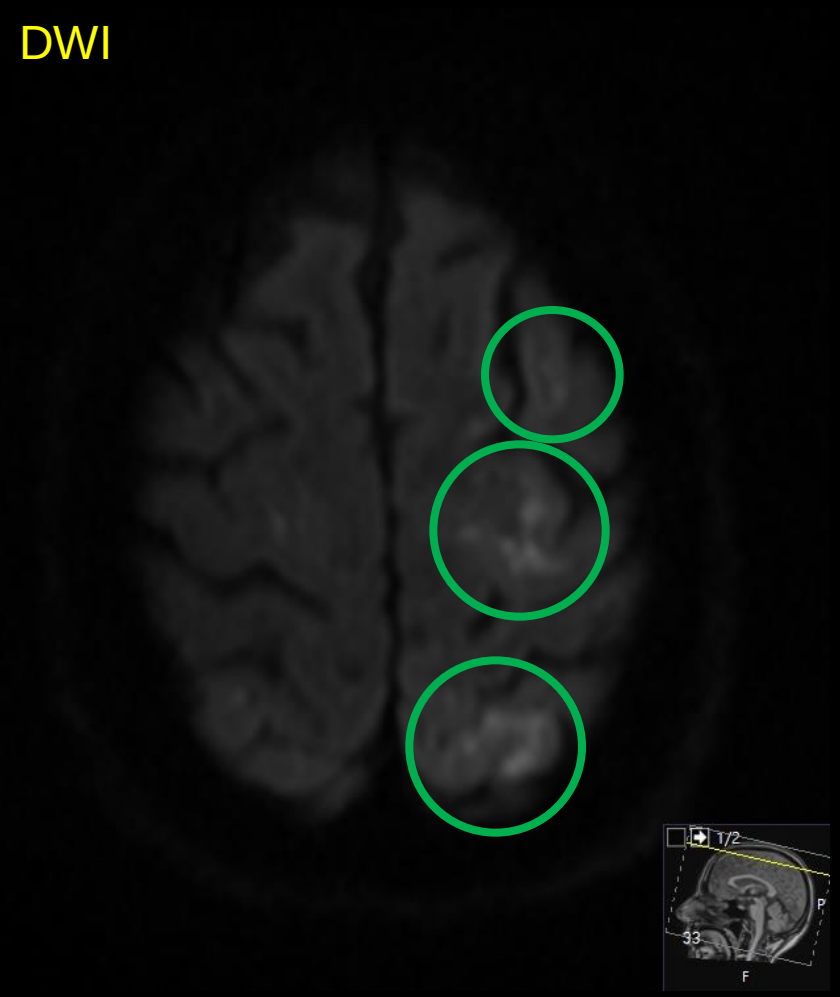
ADC

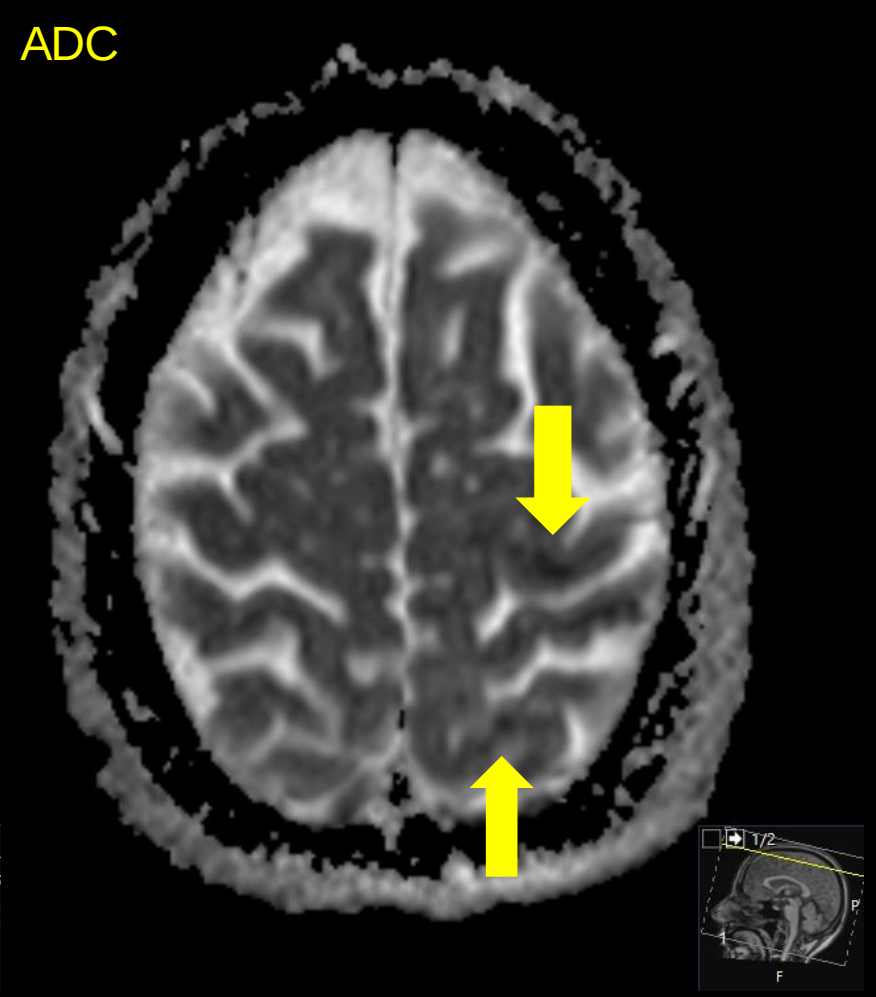


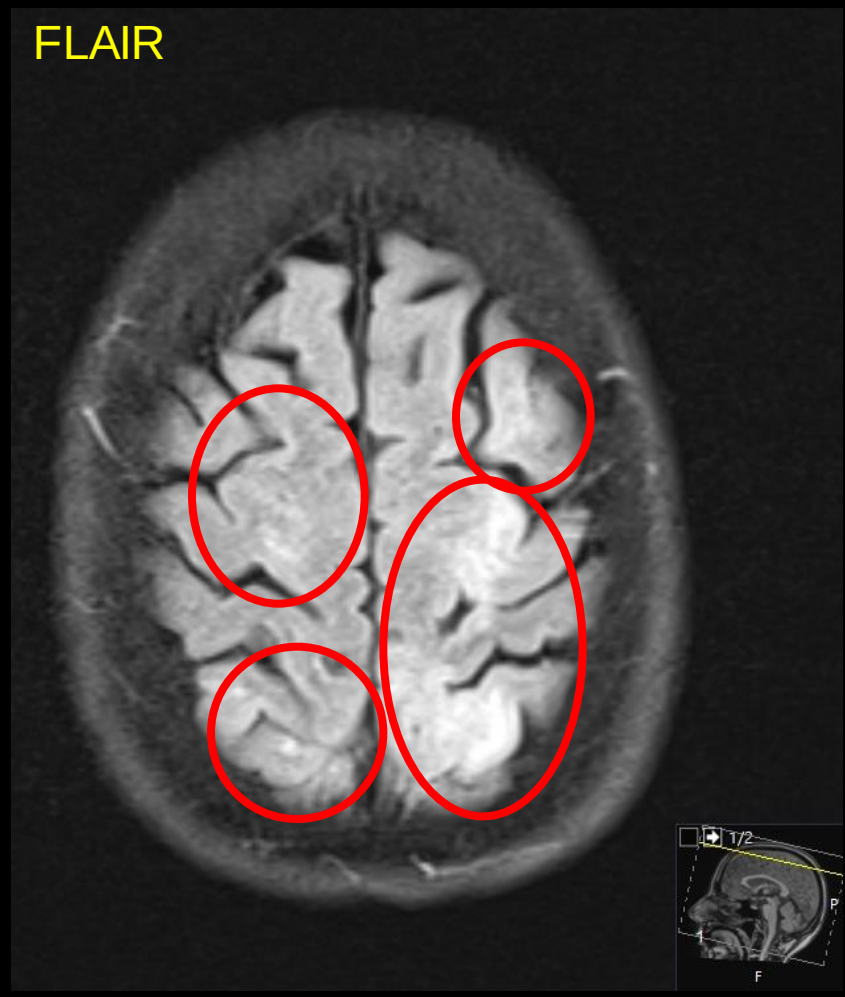
FLAIR



MRI Findings (labeled)







Diffuse areas of edema (hyperintensity on FLAIR images) with scattered areas of diffusion restriction [hyperintensity on DWI and hypointensity on ADC (apparent diffusion coefficient) images].

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Differential Diagnosis for Diffusion Restriction

- Ischemia
  - Infarction: arterial or venous, thrombotic microangiopathies (DIC, HUS, TTP)
  - Reversible: Posterior reversible encephalopathic syndrome (PRES), status epilepticus
- Infectious: Creutzfeldt-Jakob Disease, encephalitis, abscess (fungal, pyogenic, parasitic), toxoplasmosis
- Metabolic: hypoglycemia, Wernicke's encephalopathy
- Demyelinating: multiple sclerosis, osmotic myelinolysis
- Poisoning: carbon monoxide
- Neoplasm: metastasis, primary tumor, lymphoma

Finelli, 2012.

Final Dx:

Acute hypertensive encephalopathy
popularly known as
Posterior Reversible Encephalopathy Syndrome
(PRES)

PRES

- All age groups susceptible, females > males
- Pathogenesis
  - Primary hypothesis: failure of appropriate arteriolar autoregulation with hypertension resulting in cerebral vasogenic edema
  - Recently suggested hypothesis: since occurs even in normotensive persons, may be more due to immune system activation resulting in endothelial damage and vascular instability, explaining several systemic risk factors
- Risk Factors
  - Hypertension, pre-eclampsia, sepsis, autoimmune conditions (eg. lupus, scleroderma, granulomatosis with polyangiitis), vasculitis, immunosuppressive therapy, and renal disease

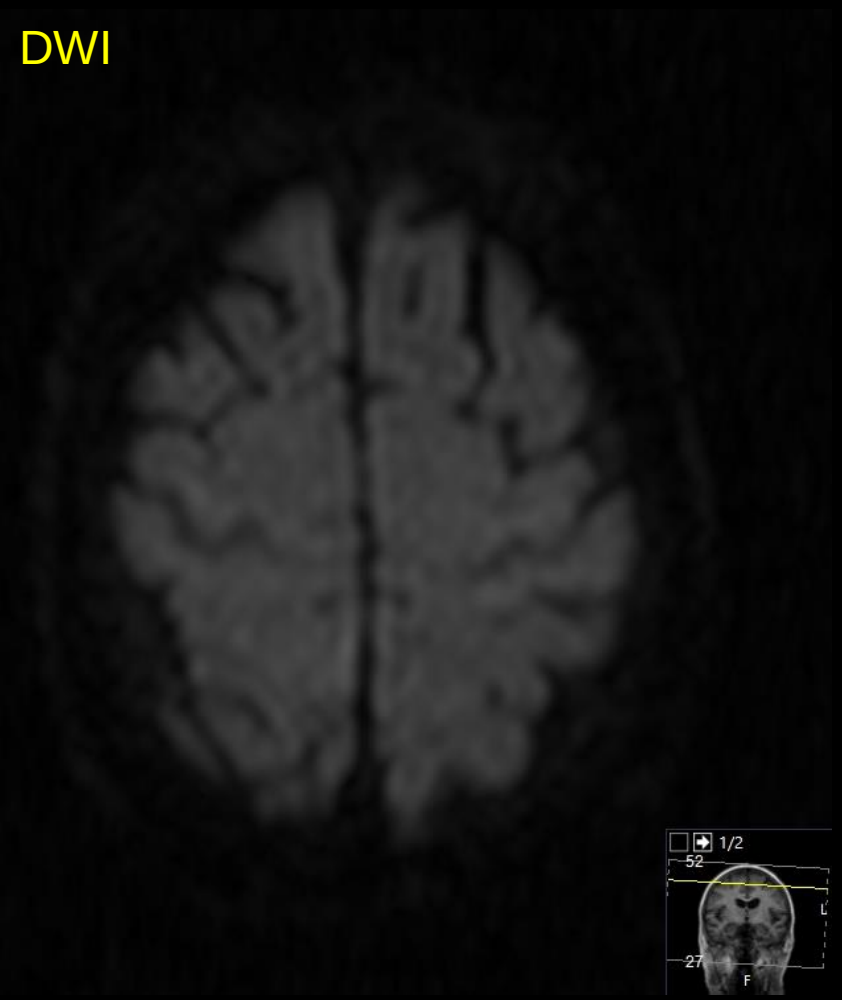
Neill, 2018; Bartynski, 2008; Hugonnet, 2013.

PRES

- Clinical Manifestations
  - Headaches, altered level of consciousness, visual disturbances, seizures, nausea, coma
  - Limb weakness is uncommon
- Radiologic Diagnosis
  - Neuroradiographic findings, while not specific, may strongly support a diagnosis given an appropriate clinical setting
  - On CT, may show diffuse hypodense areas in affected regions which correspond to:
  - On MR, areas of low signal intensity on T1 and high signal intensity on T2 in affected regions.
  - MRI (specifically, T2 weighted imaging) is most appropriate – shows evidence of widespread vasogenic edema; typically in parietal and occipital lobes
  - Diffusion hyperintensity on DWI images, though uncommon, may signal irreversible infarction and is associated with poorer outcomes . If decreased, ADC values tend to be not as low as with infarction (cytotoxic edema).
  - May resemble other pathology since watershed zones are involved.

Neill, 2018; Bartynski, 2008; Hugonnet, 2013.

Patient Follow-Up



- Continued follow-up by internal medicine and neurology with continued management of bacteremia and targeted management of hypertension.
- Neurological manifestations, including limb weakness and visual symptoms, continued for 1.5 months.
- On day 47 after presentation, after neurological manifestations resolved, a follow-up MRI was obtained, in which no pathology was identified, further supporting the diagnosis of PRES.

References:

- ACR Appropriateness Criteria for cerebrovascular disease. American College of Radiology.
- Finelli PF. Diagnostic approach to restricted-diffusion patterns on MRI imaging. Neurol Clin Pract 2012 Dec;2(4):287-29.
- Hugonnet E, Da Ines D, Boby H et al. Posterior reversible encephalopathy syndrome (PRES): Features on CT and MR imaging. Diagn Interv Imaging 2013 Jan;94(1):45-52.
- Neill TA. UpToDate: Reversible posterior leukoencephalopathy syndrome. 2018.
- Bartynski WS. Posterior Reversible Encephalopathy Syndrome, Part 1: Fundemental Imaging and Clinical Features. Am J Neuroradiol 2008;29(6):1036-42.
- Bartynski WS. Posterior Reversible Encephalopathy Syndrome, Part 2: Controversies Surrounding Pathophysiology of Vasogenic Edema. Am J Neuroradiol 2008;29(6):1043-49.