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?

Obtained after no improvement in neurological manifestations.
MRI Findings (unlabeled)
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].
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

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 (cystotoxic edema).
  • May resemble other pathology since watershed zones are involved.

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:

• Neill TA. UpToDate: Reversible posterior leukoencephalopathy syndrome. 2018.