posterior leukoencephalopathy syndrome (created by Paul Young 02/10/07)

**Clinical Features**
- A reversible syndrome of headache, altered mental status, seizures, and loss of vision associated with characteristic findings involving predominantly posterior white matter on brain imaging and occurring in association with severe hypertension and immunosuppression

**General**
- Term first coined by Hinchee et al. in 1996 who reported on a series of 15 patients
- Has been described both in children and adults

**Pathogenesis**
- Not precisely known
- Rapid rise in blood pressure overwhelms normal autoregulatory mechanisms
- Leads to dilatation and leakage of cerebral arterioles causing vasogenic edema
- Posterior circulation has less sympathetic adrenergic innervation, and therefore is thought to be more susceptible to effects of rapid rise in blood pressure

**Differential Diagnosis**
- Vascular
  - Infarct, especially "top-of-the-basilar syndrome" (with bilateral PCA ischemia)
  - Hemorrhage (congiopphilic parieto-occipital lobar ICH etc)
- Venous thrombosis
- Encephalitis, meningitis
- Inflammatory / Autoimmune
  - Postinfectious encephalomyelitis
  - Vasculitis e.g. SLE

**Investigation**
- Imaging
  - Changes noted below are seen in bilateral occipital and parietal lobes
  - Often symmetrical but can be asymmetrical
  - Primarily affects white matter, but grey can also be involved
  - More rarely may involve brain stem, cerebellum, basal ganglia, frontal lobes
  - Imaging findings are REVERSIBLE with prompt successful treatment
  - If treatment is not promptly initiated, may progress to infarction or hemorrhage

**Treatment**
- Control blood pressure
  - 10-20% decrease in MAP is usually sufficient to terminate process
- Discontinue or decrease dose of offending agents (immunosuppressive, cytotoxic)
- Treat hypomagnesemia
- Treat seizures with anticonvulsants
  - Note: Phenytoin also induces metabolism of cyclosporin and FK-506

**Prognosis**
- Most patients recover completely with prompt treatment within hours (12-24h) to days
- Imaging findings may persist for weeks
- Can lead to posterior circulation infarction or hemorrhage if not treated promptly
- Patients do not require chronic antiepileptic treatment once imaging abnormalities have resolved

**Aetiology**
- A reversible syndrome of headache, altered mental status, seizures, and loss of vision associated with characteristic findings involving predominantly posterior white matter on brain imaging and occurring in association with severe hypertension and immunosuppression

**Cytoxics**
- Cyclosporin A
- Tacrolimus / FK-506
- IFN-α
- Cisplatin
- Cytarabine
- IVIG
- Erythropoietin

**Immunosuppressive agents and cytotoxic drugs (see below)**
- Drug withdrawal (esp clonidine)
- Hypertensive encephalopathy
- Renal failure with hypertension, these patients appear to be more susceptible
- Eclampsia (pregnancy or puerperium)
- Collagen vascular disorders, including SLE, PAN, Behcet's
- TTP
- Acute porphyria
- Post organ transplantation
- Post-carotid endarterectomy (unilateral hemispheric) with reperfusion syndrome
- GBS with autonomic hyperactivity

**Systemic Signs**
- Hypertension
  - Usually acute onset
  - Can be mild to moderate OR severe (depending on patient's usual BP)

**Metabolic derangements**
- Hypomagnesemia
- Hyperchloremia
- Hypocholesterolemia
- Both of above present in > 50% patients with RPLE secondary to cyclosporin A

**Case Reports**
- Collagen vascular disorders, including SLE, PAN, Behcet's
- TTP
- Acute porphyria
- Post organ transplantation
- Post-carotid endarterectomy (unilateral hemispheric) with reperfusion syndrome
- GBS with autonomic hyperactivity

**Pathogenesis**
- Not precisely known
- Rapid rise in blood pressure overwhelms normal autoregulatory mechanisms
- Leads to dilatation and leakage of cerebral arterioles causing vasogenic edema
- Posterior circulation has less sympathetic adrenergic innervation, and therefore is thought to be more susceptible to effects of rapid rise in blood pressure

**CSF**
- Usually normal
- May have mild elevation in protein

**Imaging**
- Changes noted below are seen in bilateral occipital and parietal lobes
- Often symmetrical but can be asymmetrical
- Primarily affects white matter, but grey can also be involved
- More rarely may involve brain stem, cerebellum, basal ganglia, frontal lobes
- Imaging findings are REVERSIBLE with prompt successful treatment
- If treatment is not promptly initiated, may progress to infarction or hemorrhage

**Imaging Findings**
- Hypodense lesions on CT / MRI
- Iso- / Hypo-intense on T1
- Hyperintense on T2
- Iso- / Hypo-intense on DWI