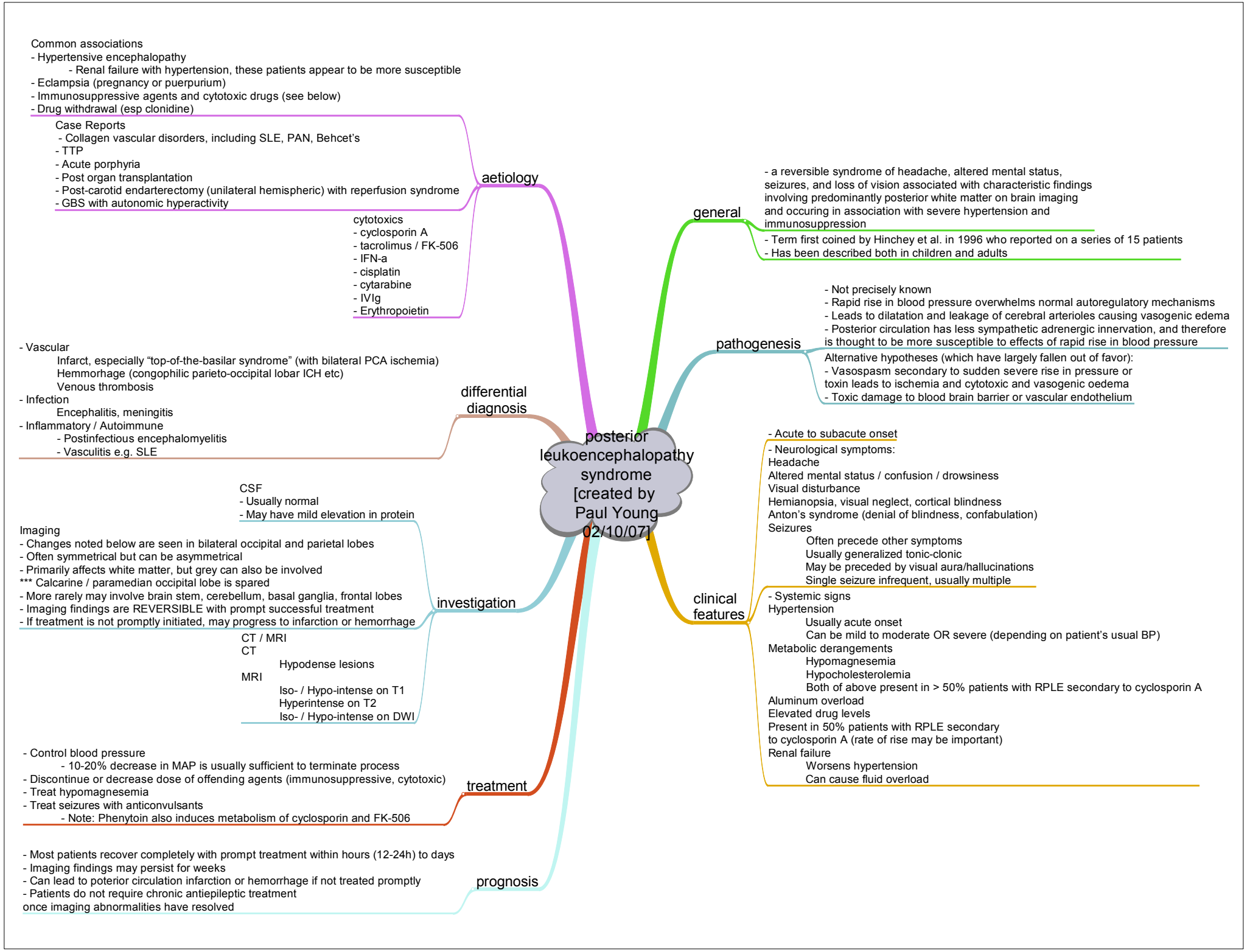
posterior leukoencephalopathy syndrome [created by Paul Young 02/10/07]
aetiology
Common associations
- Hypertensive encephalopathy
- Renal failure with hypertension, these patients appear to be more susceptible
- Eclampsia (pregnancy or puerpurium)
- Immunosuppressive agents and cytotoxic drugs (see below)
- Drug withdrawal (esp clonidine)
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
cytotoxics
- cyclosporin A
- tacrolimus / FK-506
- IFN-a
- cisplatin
- cytarabine
- IVIg
- Erythropoietin
general
- 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 occuring in association with severe hypertension and immunosuppression
- Term first coined by Hinchey 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
Alternative hypotheses (which have largely fallen out of favor):
- Vasospasm secondary to sudden severe rise in pressure or toxin leads to ischemia and cytotoxic and vasogenic oedema
- Toxic damage to blood brain barrier or vascular endothelium
clinical features
- Acute to subacute onset
- Neurological symptoms:
Headache
Altered mental status / confusion / drowsiness
Visual disturbance
Hemianopsia, visual neglect, cortical blindness
Anton's syndrome (denial of blindness, confabulation)
Seizures
Often precede other symptoms
Usually generalized tonic-clonic
May be preceded by visual aura/hallucinations
Single seizure infrequent, usually multiple
- Systemic signs
Hypertension
Usually acute onset
Can be mild to moderate OR severe (depending on patient's usual BP)
Metabolic derangements
Hypomagnesemia
Hypocholesterolemia
Both of above present in > 50% patients with RPLE secondary to cyclosporin A
Aluminum overload
Elevated drug levels
Present in 50% patients with RPLE secondary to cyclosporin A (rate of rise may be important)
Renal failure
Worsens hypertension
Can cause fluid overload
investigation
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
*** Calcarine / paramedian occipital lobe is spared
- 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
CT / MRI
CT
Hypodense lesions
MRI
Iso- / Hypo-intense on T1
Hyperintense on T2
Iso- / Hypo-intense on DWI
differential diagnosis
- Vascular
Infarct, especially "top-of-the-basilar syndrome" (with bilateral PCA ischemia)
Hemmorhage (congophilic parieto-occipital lobar ICH etc)
Venous thrombosis
- Infection
Encephalitis, meningitis
- Inflammatory / Autoimmune
- Postinfectious encephalomyelitis
- Vasculitis e.g. SLE
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 poterior circulation infarction or hemorrhage if not treated promptly
- Patients do not require chronic antiepileptic treatment
once imaging abnormalities have resolved