Sturge-Weber Syndrome

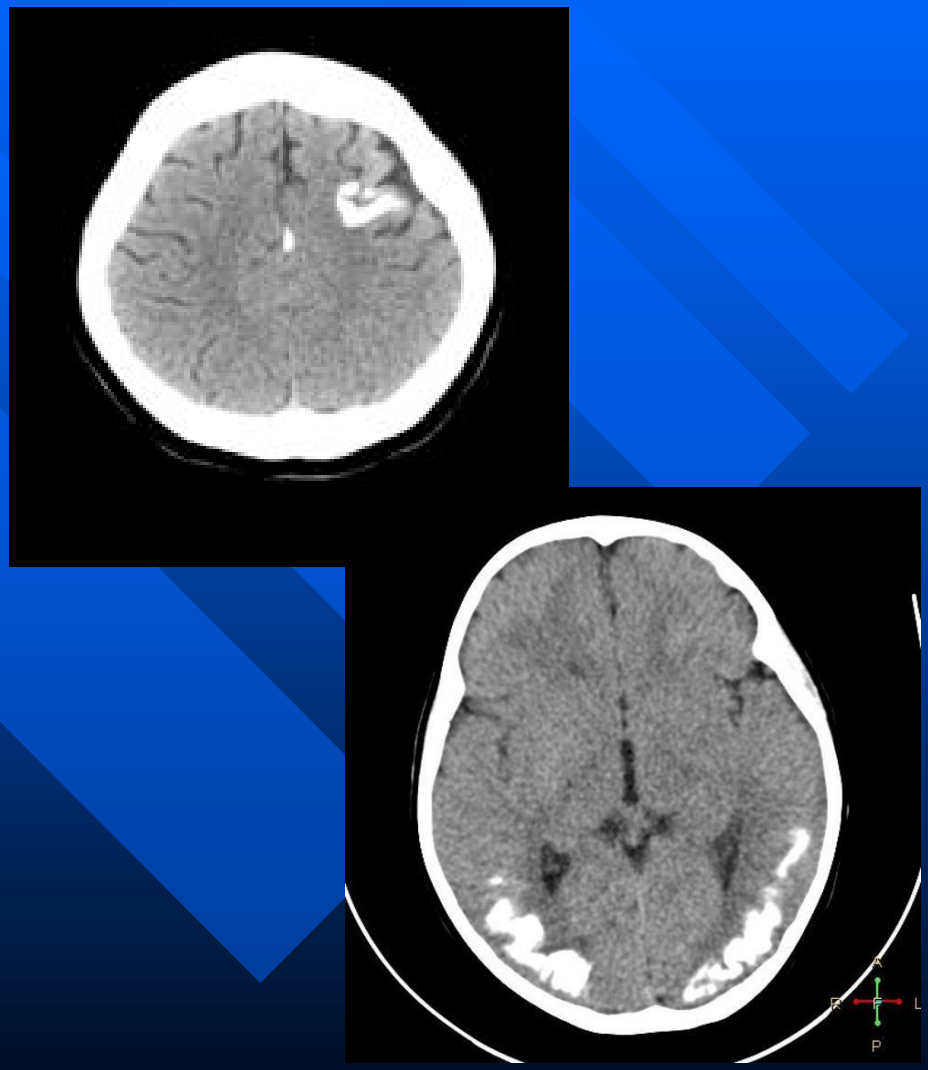
- Encephalotrigeminal angiomatosis
  - is a phakomatosis characterised by facial port wine stains and pial angiomas.
- Part of a wide spectrum of possible phenotypes included in the craniofacial arteriovenous metameric syndrome (CAMS).

Sturge-Weber Syndrome

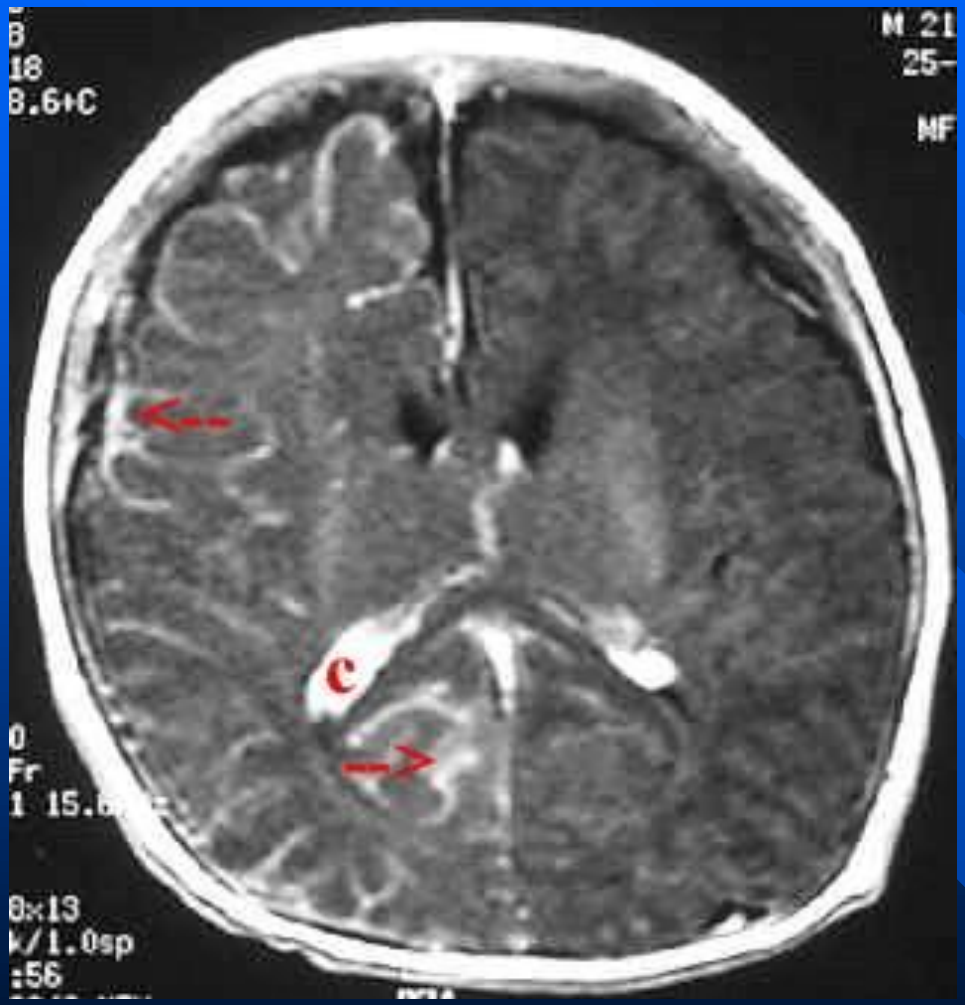
- Usually obvious on account of a congenital facial cutaneous haemangioma (also known as port wine stain or facial naevus flammeus).
- Feature is almost always present and usually involves the ophthalmic division (V1) of the trigeminal nerve; if this territory is not involved, SWS is unlikely
- Most common clinical manifestation
  - Childhood seizures, present in 71-89% of cases 2, that are often refractory to medical therapy

CT Findings

- Subcortical calcification at an earlier age than plain film and can also demonstrate associated parenchymal volume loss
- 'tram-track' subcortical calcification
- Calvarial and regional sinus enlargement may be evident
- Ipsilateral choroid plexus may be enlarged



Sturge-Weber Syndrome



Associations:

- **Coarctation of aorta**
- **Paragangliomas**