

## Clinical Image

## Imaging Features of Bilateral Involvement in Sturge-Weber Syndrome

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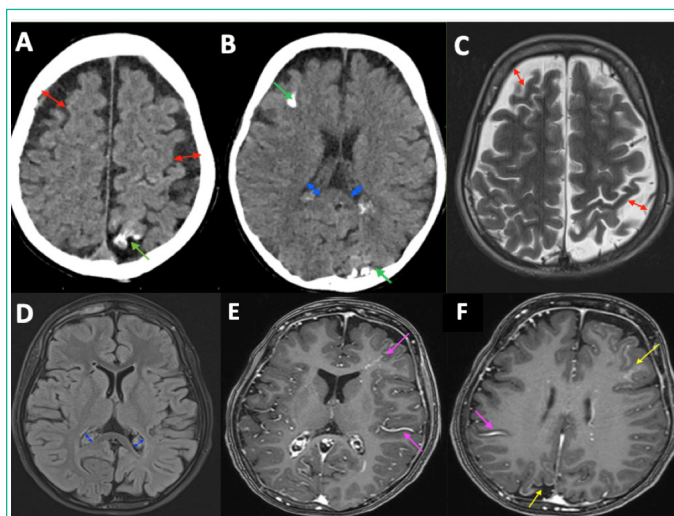
Sturge-Weber syndrome is a neurocutaneous phacomatosis due to an embryonic malformation of the foetal vascular system. It is characterised by two types of malformations: a congenital facial wine stain and a leptomeningeal capillary-venous angioma, associated with ocular manifestation [1-3] Generally located homolaterally in the classic form, although bilateral involvement is possible but remains rare [4].

The diagnosis of SWS relies massively on neuroimaging, specifically Magnetic Resonance Imaging (MRI), which is critical for identification abnormalities that precede neuro-ocular complications.

The diagnosis is usually obvious because of the cutaneous angioma, involving the ophthalmic branch (V1) of the trigeminal nerve. the main neurological manifestations are dominated by epileptic seizures, affecting between 75% and 90% of patients. Around half of patients also present with a motor deficit. associated with convulsions, headaches or migraines. Although psychiatric disorders and mental retardation have been reported, they remain rare [1,2]. Radiological findings can be seen on both CT scan and brain MRI, but the MRI is better than the CT scan, at detecting early signs, sometimes even before clinical symptoms appear [5,6] including:

- o Focal or hemispheric cerebral atrophy
- o Cortical or subcortical calcifications
- o Hypertrophy and calcification of the choroid plexus
- o Choroid plexus angioma
- o Leptomeningeal enhancement due to congested internal veins

Dilatation of trans-parenchymal veins



**Figure 1:** CT and MRI images in a 10-year-old patient with bilateral sturge-weber syndrome.

Non injected CT sections (A, B) show bilateral frontoparietal atrophy (red arrows), right frontal and left parietal calcifications (green arrows), and choroid hypertrophy (blue arrows)

Axial T2 sequence (C) shows bilateral frontoparietal atrophy.

Axial flair sequence (D) shows choroid plexus hypertrophy. Axial T1 sequence after gadolinium injection (E, F) with leptomeningeal contrast (yellow arrows) and bilateral dilatation of some intraparenchymal veins.

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