Bilateral Cavernous Hemangiomas in Sturge-Weber Syndrome

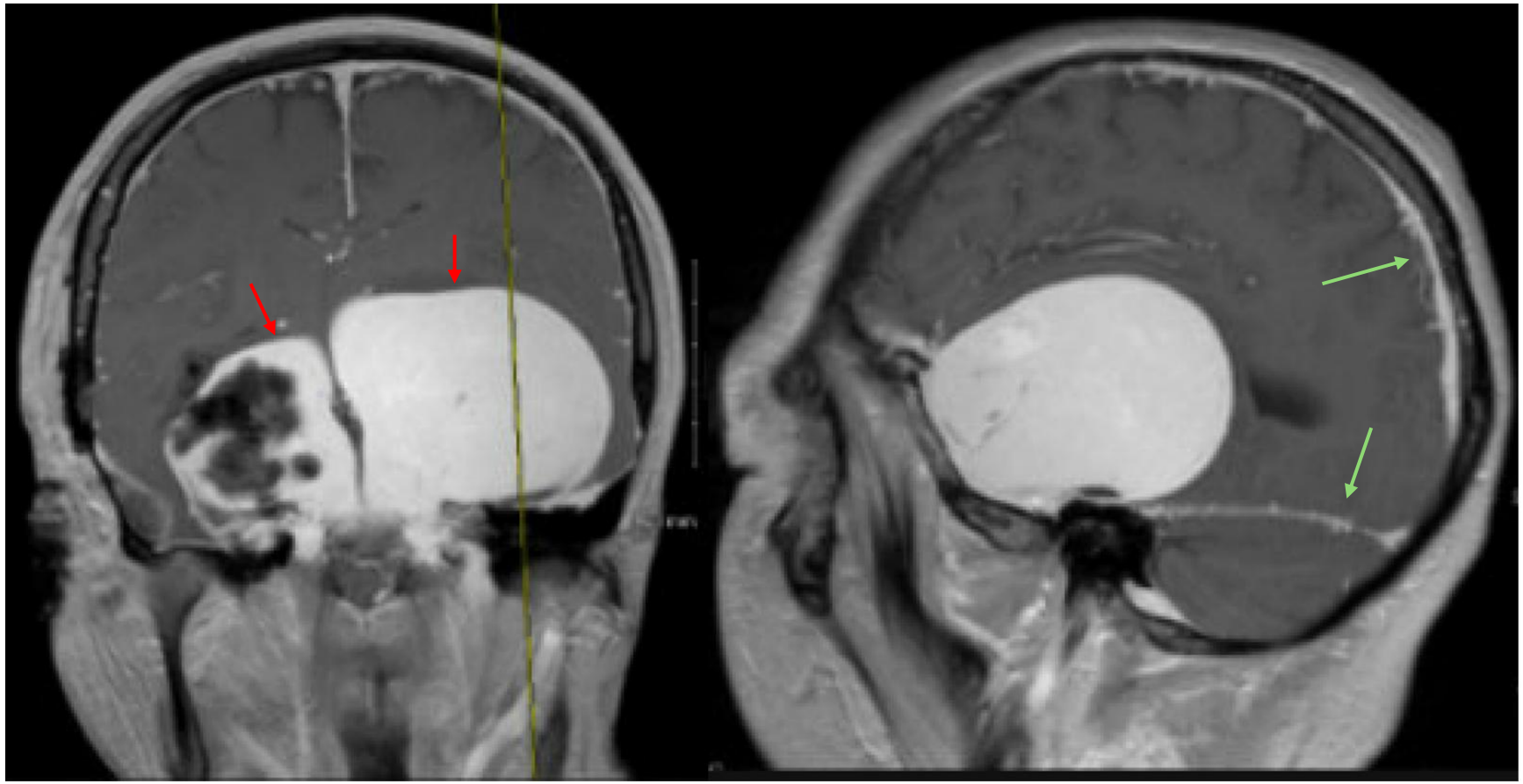
Anastasiya Boutko, Marlee Ghobrial , Megan Ghobrial, Ryan Murtagh.

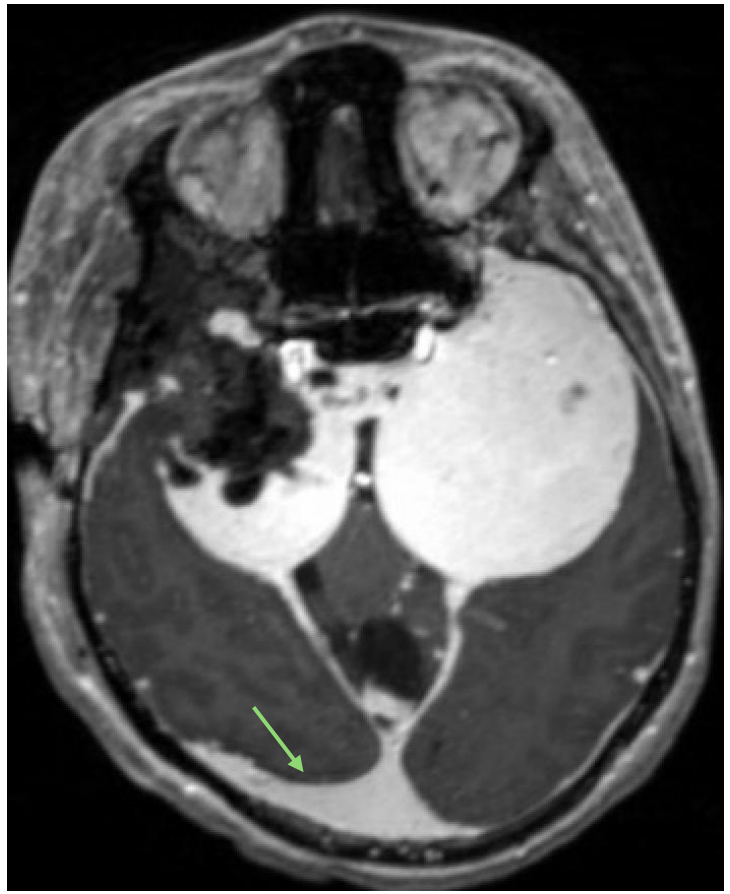
USF Health Morsani College of Medicine, Tampa, FL

Clinical Presentation

- 22-year-old male with known Sturge-Weber Syndrome (SWS), glaucoma, retinal detachment
- Symptoms: 2 weeks of worsening fatigue, headache, poor appetite → acute lethargy & nonverbal in last 1-2 days.
- Physical exam: bilateral facial port-wine stains
- Initial CT revealed bilateral masses prompting further evaluation

Initial MR Imaging





Pre-treatment.

1. Solid enhancing masses (→) from the cavernous sinus causing anterior temporal extra-axial mass effect and obstructive hydrocephalus.
2. Additional findings: Leptomeningeal angiomatosis and dural enhancement (→).

Post right partial resection.

Management and outcome

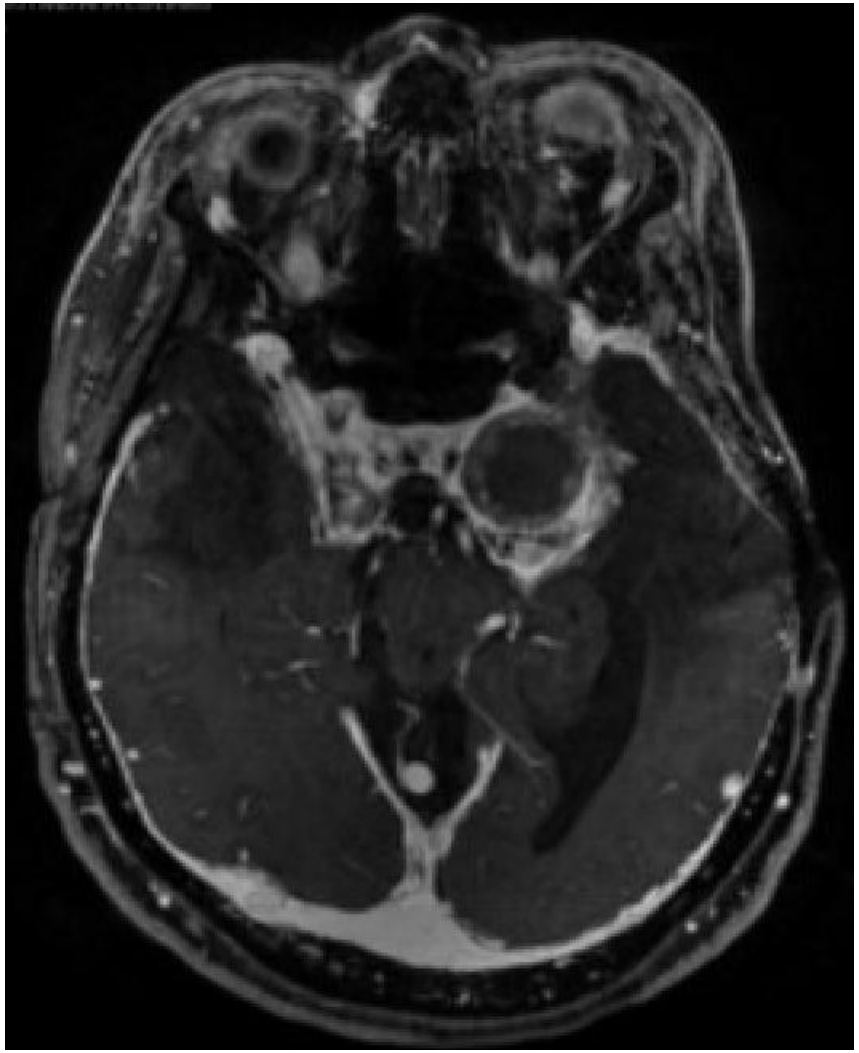
Typical SWS treatment:

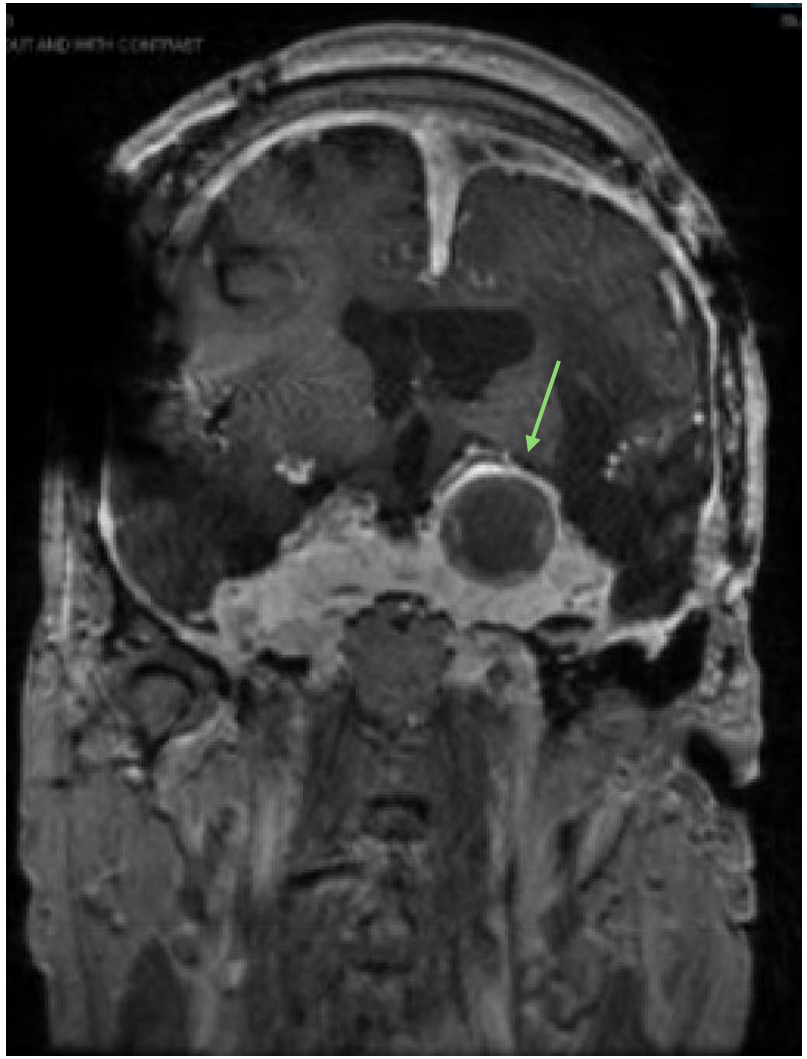
- Primarily seizure control
- Surgical resection only considered in refractory epilepsy.
- Ophthalmological exam for glaucoma/ocular involvement.

Our patient's course:

- Pathology confirmed cavernous hemangiomas
- VP shunts → symptom improvement
- Partial resections and radiation therapy over following years

Most recent follow-up MR





Stable residual cavernous hemangiomas post multiple treatments (several partial resections, shunt placements, and radiation therapy), with a retained cotton ball (→).

SWS Overview

- Rare, sporadic neurocutaneous syndrome caused by somatic GNAQ mutation.
- Characterized by abnormal vascular development in the face, eyes, and brain.
- Classic triad:
  - **Facial port-wine stains** (usually in trigeminal distribution)
  - **Leptomeningeal angiomas** (most often parieto-occipital, associated with seizures and progressive cortical damage)
  - **Ocular abnormalities** (glaucoma, choroidal hemangiomas, risk of retinal detachment)

Classic SWS Imaging [1,2]

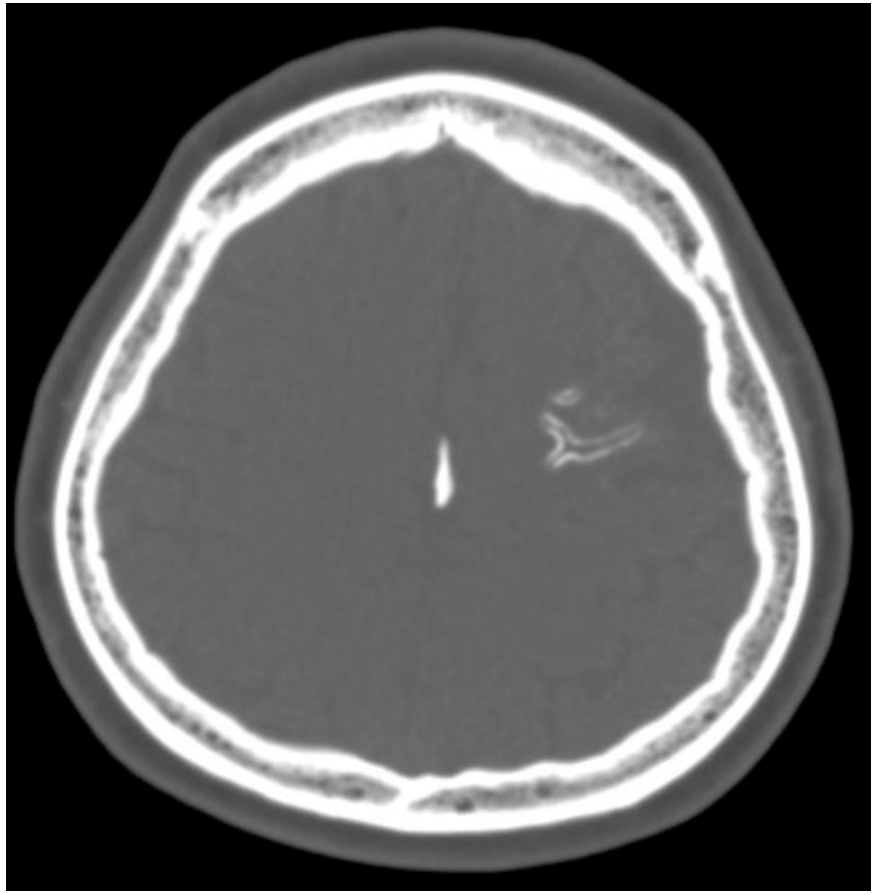
CT:

- **Tram-track gyriform calcifications**: linear/cortical calcifications, usually parieto-occipital
- **Cortical atrophy**: often ipsilateral to leptomeningeal angioma
- **Choroid plexus** enlargement and calcifications

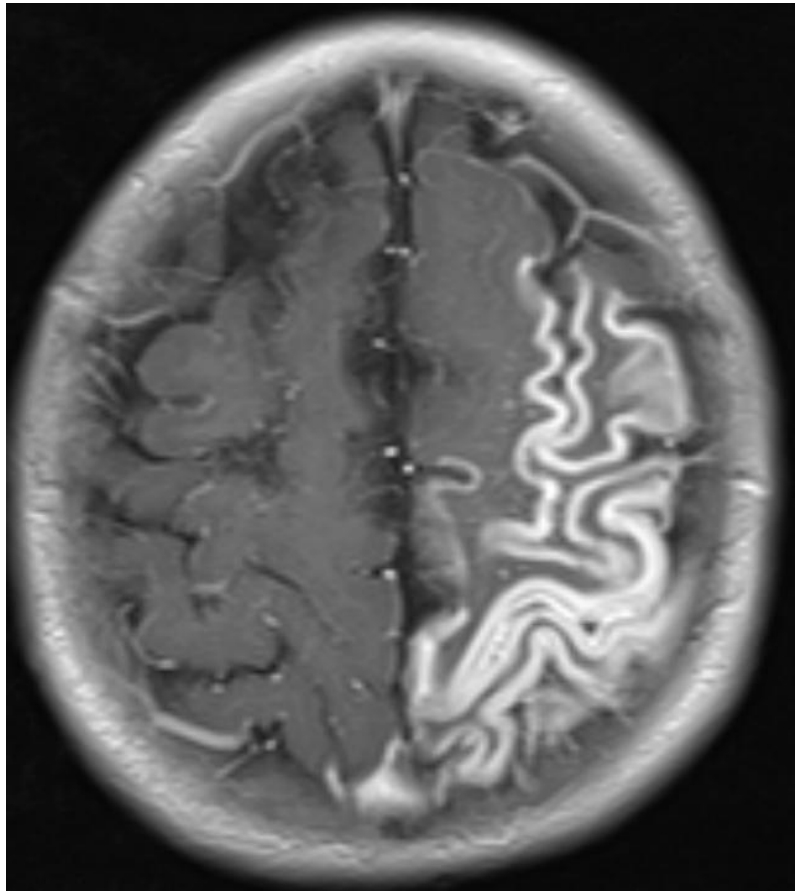
MRI:

- **Leptomeningeal angiomas**: pial enhancement after gadolinium, often unilateral and parieto-occipital
- **Venous anomalies**: enlarged deep medullary veins, transmedullary collateral pathways
- **Cortical/subcortical atrophy** with T2/FLAIR hyperintensity in underlying white matter
- **Choroid plexus angiomatosis**: thickened, enhancing

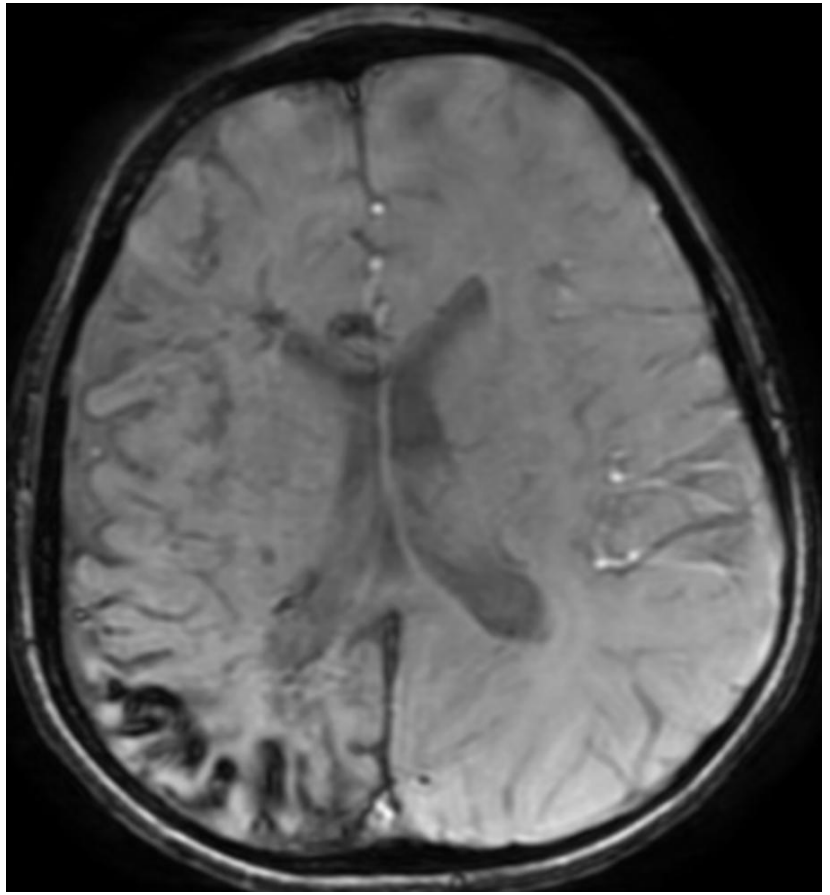
Classic SWS Imaging [3-5]



Tram-track gyriform calcifications



Leptomeningeal enhancement



Hemi-atrophy and tram-track calcifications

Take Home Points

- **Sturge-Weber syndrome**: characterized by leptomeningeal angiomas, cortical calcifications, and ocular involvement.
- **Neuroimaging hallmarks**: tram-track calcifications, cortical atrophy, leptomeningeal enhancement, and choroid plexus enlargement.
- **This case is unusual**: bilateral cavernous hemangiomas arising from the cavernous sinus — not a typical manifestation of SWS.
- **Clinical relevance**: In SWS patients with new acute neurologic decline, consider atypical vascular lesions in addition to seizures or leptomeningeal progression.

Thank you!

References:

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