Sturge-Weber syndrome: MRI findings
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Patient: 21 years, female

Clinical History:

The patient presented with a history of medically controlled partial seizures. There was a dark port wine stain on the left upper eyelid.

Imaging Findings:

The patient presented with a history of medically controlled partial seizures, having suffered from seizures since the age of 2. She had been treated conservatively with anti-epileptic agents. There was a dark port wine stain on the patient's left upper eyelid; the rest of the face looked normal. Left hemianopia was established after ophthalmological examination. Mentally she was normal and physical examination did not reveal any neurological deficit.

MR imaging of the brain was performed. Axial T1W (TR/TE:420/20) and T2W (TR/TE: 2800/110) images revealed atrophy of the left temporal lobe and thickened cortex with few sulci. T1W axial and coronal images after the administration of paramagnetic agent showed superficial meningeal enhancement as well as crescentic enhancement of the posterior ocular wall. A prominent ipsilateral choroid plexus was also depicted. Calvarial thickening with enlarged diploic space was demonstrated in the cranial vault.

Discussion:

Neurocutaneous syndromes are a diverse group of disorders that involve both skin and nervous systems. Sturge-Weber syndrome (SWS), one of the neurocutaneous syndromes, is also known as encephalotrigeminal angiomatosis or meningofacial angiomasitosis. Intracranial angiomatosis is confined to the pia mater. It is typically located in the occipital lobe. Principal features include a port wine stain capillary vascular malformation of the face and a leptomeningeal vascular anomaly that results in ischaemia and consequent atrophy and calcification of the underlying cortex. SWS is almost always sporadic in occurrence. The defect most likely occurs at gestational weeks 4 to 8, when a primordial vascular mesenchyme lies adjacent to the neural tube and ectoderm that will overlie the head and face. Abnormal development of the blood vessels at this time can affect both the skin, the brain and the leptomeninges.

Up to 90% of patient with SWS have generalised or partial seizures. The patients are usually neurologically intact at birth but develop seizures within the first year of life. Developmental delay is common. Hemi-atrophy of the brain
results in hemiparesis in about 30% of cases. The vascular malformation of SWS is associated with impaired venous drainage and progressive ischaemia of the underlying brain. The cortex gradually loses cells and calcifies. Continued seizures may also contribute to metabolic damage. The occipitoparietal region is most commonly involved.

Skull radiographs show a tram-track appearance resulting from calcifications of gyri. The calcifications of the cortex can be identified early in life by CT, but rarely at birth. CT can show some white matter calcification as well. Regional atrophy in SWS is well demonstrated by MRI. White matter below the damaged cortex may show high signal intensity on T2W images as a result of ischaemia and gliosis. Gadolinium-enhanced MRI is highly sensitive to meningeal enhancement, which is a characteristic feature of SWS and is believed to represent leakage of contrast medium through the anomaloidal pial vessels that characterise the disease. Leptomeningeal enhancement need not be present in SWS, and the absence of this characteristic finding does not preclude the diagnosis. Areas of thickened cortex with few sulci, presumed to represent migration abnormalities are also well visualised with MRI.

Other features include persistence of prominent deep medullary and subependymal veins and enlarged deep venous structures. Absence of cortical veins with centripetal venous drainage into enlarged medullary veins or anomalous deep veins in Sturge-Weber syndrome has been described in the angiographic, computed tomographic, and MR literature. Several articles have described enlargement and calcification of the choroid plexus occurring on the same side as the hemisphere affected by Sturge-Weber syndrome. Enlargement of the choroid plexus is a recognised feature of Sturge-Weber syndrome and has been attributed to angiomatosis. Enlargement of the diploic space is another imaging feature indicating a prominent extraaxial involvement.

Abnormalities of the ipsilateral eye may occur in Sturge-Weber syndrome. This disease is a cause of buphthalmos, but glaucoma is the most common clinical finding. Retinal and choroidal detachments have been reported. Ocular haemangiomas involving the choroid are estimated to occur in approximately one-third of cases and these may be seen at fundoscopy. Ocular enhancement represents choroidal haemangiomas, which are known to occur in Sturge-Weber syndrome. MR examination of the choroidal haemangiomas shows thickening of the posterior wall of the globe on unenhanced T1-weighted images and abnormal signal on proton density–weighted images. After injection of contrast material, crescentic enhancement is noted, thickest posteriorly, extending to the anterior portion of the globe.

Despite less sensitivity for calcifications than CT, MR imaging of the brain is the most important imaging method in the examination of patients with Sturge-Weber syndrome, and this examination should include thorough investigation of the eyes.

Differential Diagnosis List: Sturge-Weber syndrome

Final Diagnosis: Sturge-Weber syndrome

References:


and intracranial imaging findings.
Figure 1

Description: Axial PD-weighted (TR/TE:2800/40) image showing left temporal cortical atrophy with thickened convolutions and few sulci in the anterior temporal lobe (long arrows). Note the high signal in the posterior ocular wall (short arrows). Origin:
**Description:** Axial post-contrast T1W (TR/TE:420/20) image showing leptomeningeal enhancement (arrows). Enhancement of the enlarged diploic space of the left sphenoid lesser wing is also depicted (arrowheads). **Origin:**
**Description:** Axial post-contrast T1W image at a higher level shows better strong enhancement of the posterior ocular wall (arrows). Note also the enhanced lesser sphenoid wing (arrowhead). **Origin:**
Description: Coronal T1W (TR/TE:360/20) image shows the leptomeningeal enhancement (small arrows), prominent left choroid plexus (thick arrow) and thickened left calvarium compared to the right (white arrow). Origin: