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Teaching NeuroImages: Dyke-Davidoff-Masson in Sturge-Weber syndrome

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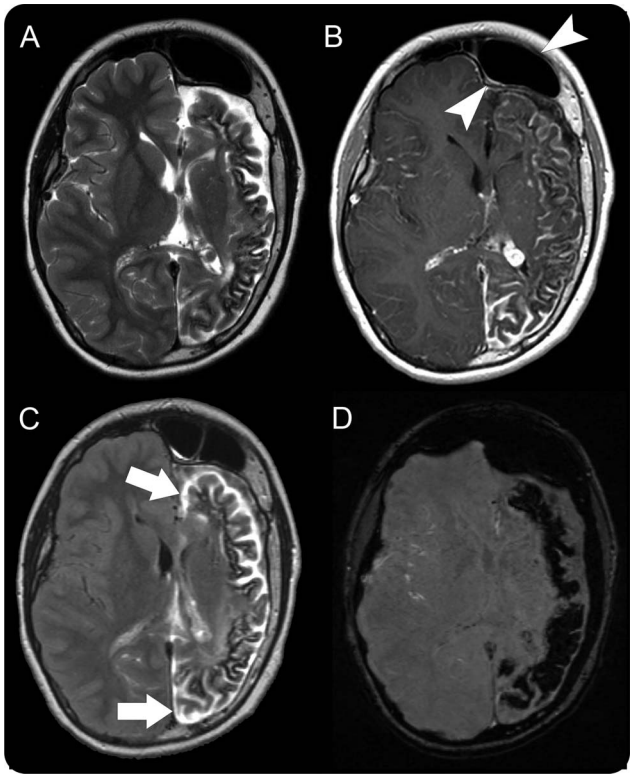
Figure MRI findings in Dyke-Davidoff-Masson syndrome and Sturge-Weber syndrome



(A) T2 and (B) contrast-enhanced T1 and (C) fluid-attenuated inversion recovery images show marked left hemispheric atrophy with engorgement of ipsilateral choroid plexus and extensive leptomeningeal enhancement (arrows, C). Note calvarial thickening with marked expansion of the left frontal sinus (arrowheads, B) and extensive susceptibility due to cortical calcification on susceptibility-weighted imaging (D).

A 13-year-old boy with long-standing seizures presented with a port wine stain involving the left V1 trigeminal distribution, right hemiparesis, and left-sided glaucoma. MRI showed typical manifestations of Sturge-Weber syndrome (SWS) with cerebral atrophy and extensive pial angiomatosis[1] (figure). Images also demonstrated findings of Dyke-Davidoff-Masson syndrome (DDMS) with compensatory calvarial expansion as a consequence of long-standing cerebral hemiatrophy.[2] DDMS usually results from early insults to the developing brain. Symptoms reflect the underlying injury and include seizures, mental retardation, hemiparesis, and facial asymmetry. Seizure management in SWS is challenging and may include medical therapy or surgery in refractory cases.

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AUTHOR CONTRIBUTIONS

Carlos Zamora: study concept, analysis of MRI data, revising the manuscript, and final approval. Marinos Kontzialis: analysis of MRI data, drafting and revising the manuscript, and final approval.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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