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The Alliance of Rarities: Sturge-Weber, Dyke-Davidoff-Mason, and Developmental Venous Anomalies

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Abstract **Case Report**

We report an exceptional case of association between Sturge-Weber syndrome, Dyke-Davidoff-Mason syndrome, and developmental venous anomalies in a 24-year-old patient admitted to the emergency department for generalized epileptic seizures, associated with a left fronto-orbital port-wine stain, cognitive impairment, facial asymmetry, and ipsilateral glaucoma. Multimodal imaging (CT and MRI) revealed gyriform cortical calcifications, cerebral hemiatrophy with compensatory hyperostosis, as well as intraparenchymal venous anomalies consistent with impaired venous drainage. Management with antiepileptic therapy and ocular hypotensive agents resulted in satisfactory seizure control and improved quality of life. This rare association suggests shared pathophysiological mechanisms and highlights the major role of imaging in diagnosing these complex neurovascular interactions.

Keywords: Sturge-Weber Syndrome, Dyke-Davidoff-Mason Syndrome, Developmental Venous Anomalies, Cerebral Hemiatrophy, Port-wine Stain.

INTRODUCTION

Sturge-Weber syndrome is a rare congenital neurocutaneous disorder characterized by a facial capillary malformation associated with leptomeningeal vascular anomalies, often leading to cortical calcifications and a wide spectrum of neurological manifestations [1,2].

Dyke-Davidoff-Masson syndrome is defined by cerebral hemiatrophy associated with compensatory osseous changes, including calvarial thickening and hyperpneumatization of the frontal sinuses [3].

The coexistence of these two entities, particularly in association with developmental venous anomalies, is exceedingly rare [4–8]. We report the case of a 24-year-old woman in whom this association was revealed following generalized seizures.

CASE REPORT

A 24-year-old woman was admitted to the emergency department for generalized tonic-clonic seizures.

Initial clinical examination revealed a left fronto-orbital port-wine stain, significant cognitive and psychomotor delay, and facial asymmetry. No focal motor deficit was identified on neurological examination.

Ophthalmological evaluation showed preserved visual acuity with ipsilateral ocular hypertension measured at 28 mmHg, compared to 14 mmHg in the contralateral eye, along with visual field defects. Fundoscopic examination revealed optic disc cupping without evidence of choroidal hemangioma. These findings were consistent with glaucoma.

Non-contrast brain CT demonstrated left hemispheric cortical atrophy, predominantly in the parieto-occipital region, associated with extensive gyriform calcifications and ipsilateral calcified choroid plexus hypertrophy.

Following contrast administration, dilated left temporal subcortical veins were observed, draining into a prominent subependymal vein along the ipsilateral

temporal horn, which subsequently drained into a dilated right internal cerebral vein.

These vascular abnormalities, in association with cortical calcifications and cutaneous and ocular findings, supported the diagnosis of Sturge-Weber syndrome.

Additionally, serpiginous intraparenchymal venous structures were identified within the corpus callosum and the right lateral ventricular atrium, draining into the right internal cerebral vein and the vein of Galen, consistent with developmental venous anomalies.

Bone window analysis revealed left frontoparietal calvarial thickening and ipsilateral frontal sinus hyperpneumatization, consistent with Dyke-Davidoff-Masson syndrome and explaining the observed facial asymmetry.

Brain MRI confirmed these findings and additionally demonstrated bilateral frontal white matter hypointensities on T2- and FLAIR-weighted sequences, suggestive of chronic microvascular injury.

The patient was managed with antiepileptic drugs and topical prostaglandin analogs for glaucoma. No surgical intervention was indicated.

Clinical evolution was favorable, with complete seizure control and a reduction of intraocular pressure to 18 mmHg after four weeks of treatment. Overall clinical status remained stable, although neuropsychological follow-up was required.

Written informed consent was obtained from the patient for the publication of this case report and the accompanying clinical and radiological images.

Table 1: Summary of previously reported cases of SWS associated with DDMS

Author (Year)	Patient Age/Sex	Side of Lesions	Clinical Presentation
Corey *et al.,* (2005) [5]	38, Female	Right	Intractable seizures, facial and arm nevus
Zamora *et al.,* (2015) [6]	13, Male	Left	Long-standing seizures, right hemiparesis, glaucoma
Bekci *et al.,* (2016) [7]	14, Male	Right	Seizures, left hemiparesis, mental retardation
Lin *et al.,* (2024) [8]	2, Female	Right	Seizures, left hemiparesis, DDMS secondary to SWS

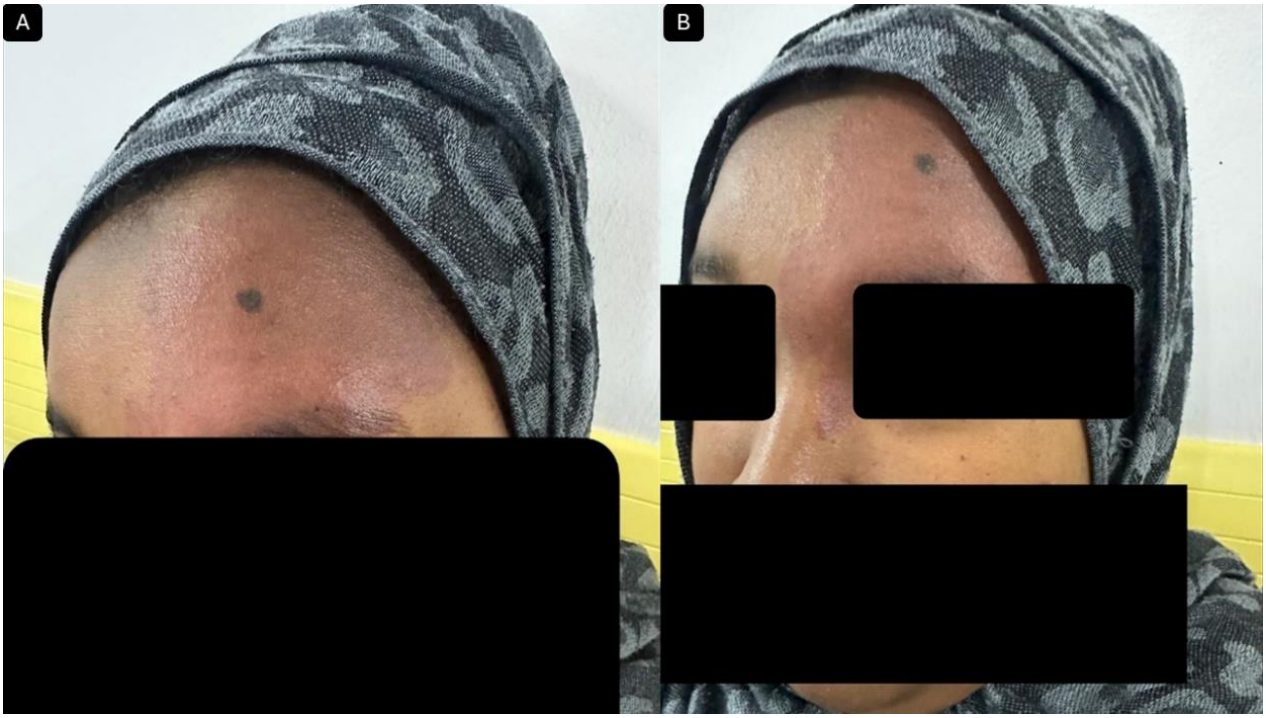

Figure 1: Clinical photograph showing a left fronto-orbital facial angioma, characteristic of Sturge-Weber syndrome

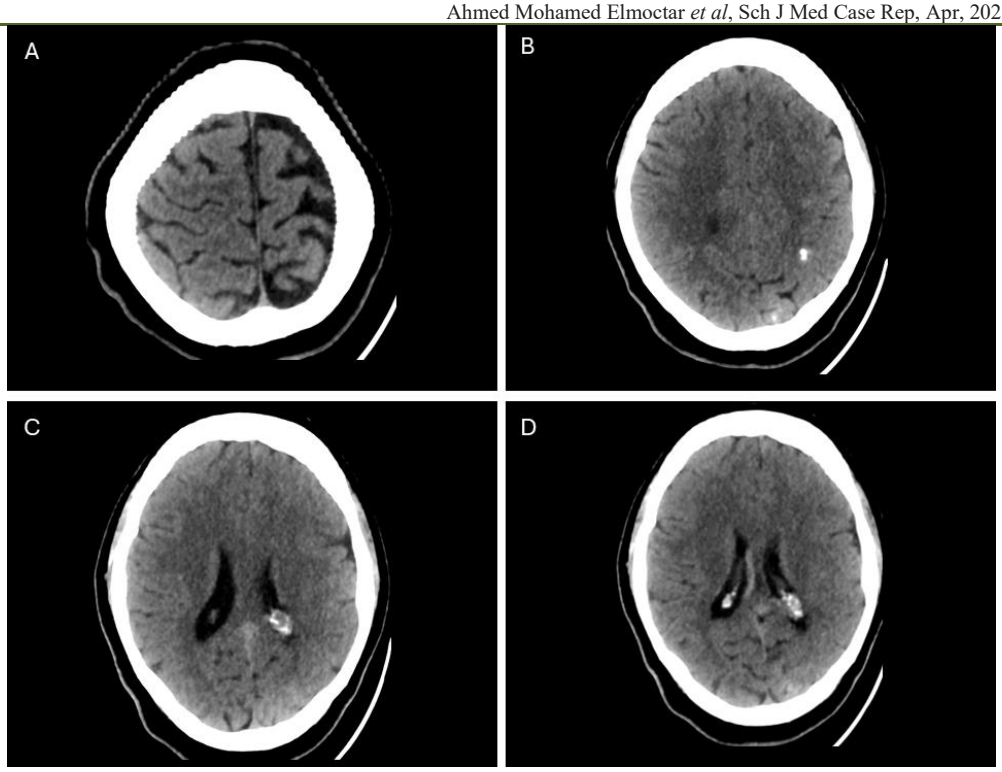

Figure 2: Non-contrast brain CT images.
(a) Axial image showing marked left hemispheric cortical atrophy, predominantly involving the parieto-occipital region
(b) Axial image demonstrating extensive gyriform cortical calcifications in the same territory
(c, d) Axial images illustrating hypertrophy and calcifications of the left choroid plexus

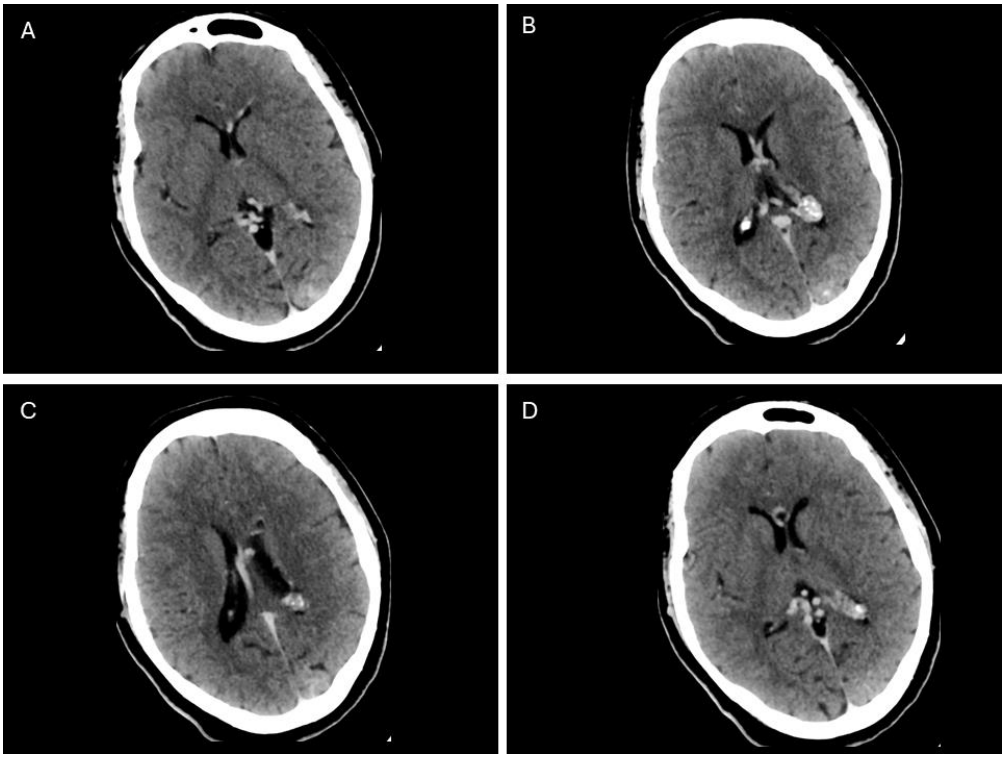

Figure 3: Post-contrast brain CT images
(a) Axial image showing ectatic left temporal subcortical veins
(b) Coronal reformation demonstrating drainage into a prominent subependymal vein ascending along the temporal horn
(c) Coronal image showing subsequent drainage into an enlarged right internal cerebral vein
(d) Axial image revealing additional serpiginous venous structures at the corpus callosum and the right atrium of the lateral ventricle

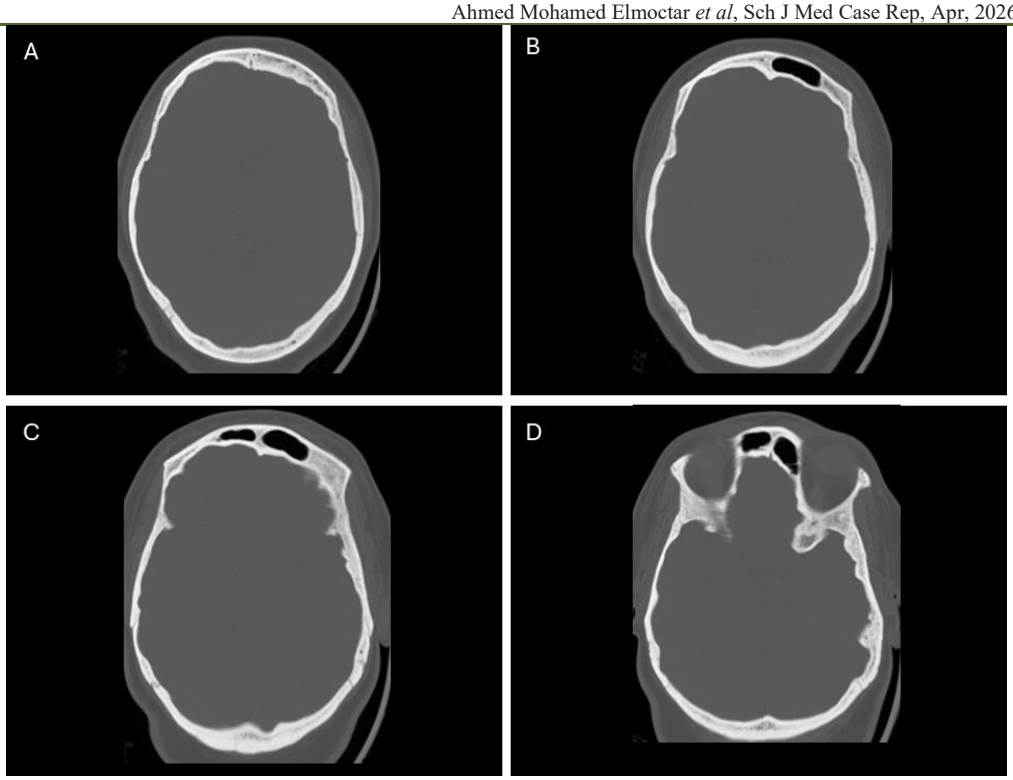

Figure 4: Bone window brain CT images
(a, b) Axial images demonstrating compensatory left frontoparietal calvarial thickening
(c, d) Axial images showing hyperpneumatization of the left frontal sinus

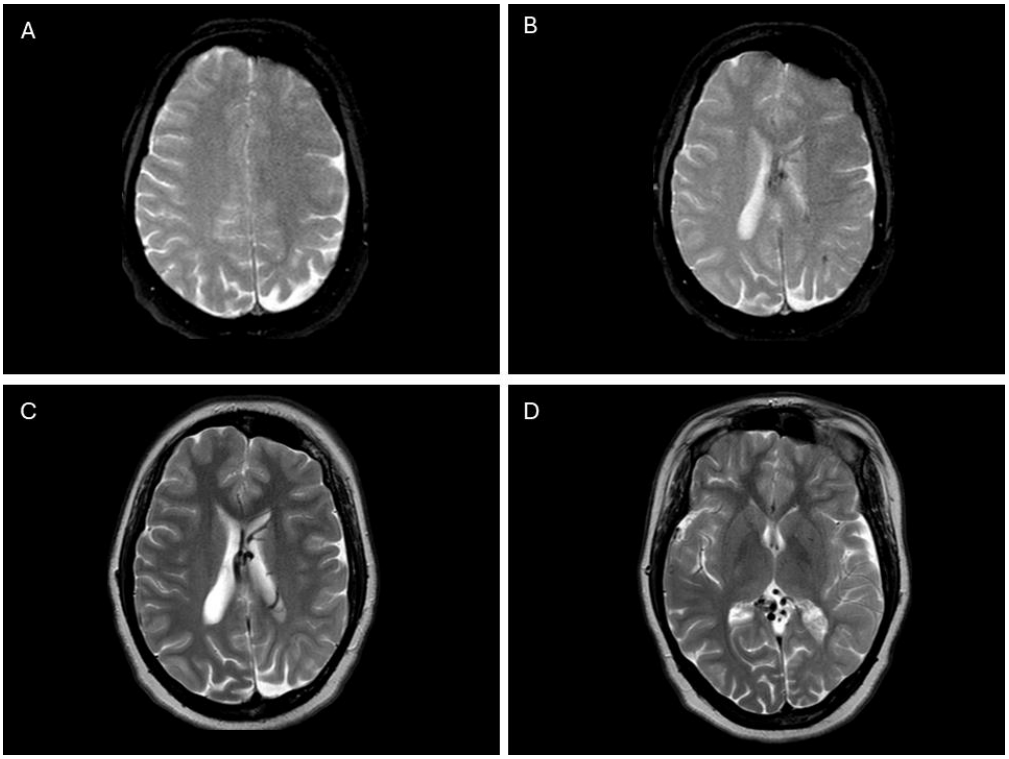

Figure 5: Brain MRI findings
(a) Axial T1-weighted image confirming left frontoparietal cortico-subcortical atrophy
(b) Axial post-contrast T1-weighted image showing abnormal pial enhancement corresponding to vascular malformations
(c) Axial SWI (or GRE) sequence highlighting extensive cortical calcifications
(d) Axial FLAIR sequence demonstrating bilateral frontal white matter hypointensities suggestive of chronic microvascular injury

DISCUSSION

Sturge-Weber syndrome is a rare neurocutaneous disorder with an estimated incidence of 1 in 20,000 to 1 in 50,000 live births [1]. It results from a somatic mutation in the GNAQ gene, leading to intracranial vascular malformations and cortical calcifications. Clinically, it is characterized by a facial capillary malformation, neurological manifestations, and ocular involvement, including glaucoma or choroidal hemangioma [2].

In our case, the presence of facial and cerebral angiomatosis associated with glaucoma is consistent with type I SWS. The observed gyriform calcifications reflect abnormal angiogenesis and impaired cortical venous drainage, which are hallmark features of the disease [4].

Dyke-Davidoff-Masson syndrome is characterized by a triad of cerebral hemiatrophy, calvarial thickening, and frontal sinus hyperpneumatization [3]. The absence of contralateral hemiparesis in our patient highlights the clinical variability of this condition, which depends on the timing and severity of the initial cerebral insult. Chronic cerebral hypoperfusion is considered the main underlying mechanism.

The association between SWS and DDMS is extremely rare but may be explained by shared neurovascular mechanisms. SWS is primarily a disorder of venous dysplasia occurring during early embryogenesis. Failure of regression of the primitive venous system leads to venous stasis, chronic hypoxia, and secondary cortical atrophy. This early unilateral hypoperfusion likely constitutes the pathophysiological basis for DDMS.

The developmental venous anomalies observed in this case play a compensatory role by establishing alternative deep venous drainage pathways. However, these abnormal venous channels may paradoxically contribute to persistent hypoxia and exacerbate neurological dysfunction [4,9].

Only a limited number of cases describing the coexistence of SWS and DDMS have been reported in the literature [5–8]. These observations support the hypothesis that early and severe hemispheric ischemia in SWS leads to the compensatory osseous change's characteristic of DDMS. Our case is particularly notable for the multimodal imaging demonstration of multiple deep DVAs compensating for impaired superficial venous drainage.

Management is primarily symptomatic, focusing on seizure control, treatment of glaucoma, reduction of facial angioma using pulsed dye laser therapy, and provision of rehabilitative and psychological support to optimize patient outcomes [1,2].

CONCLUSION

This case highlights an exceptional association between Sturge-Weber syndrome and Dyke-Davidoff-Masson syndrome, supporting a shared pathophysiological mechanism dominated by early impairment of cerebral venous drainage.

It emphasizes the central role of chronic hypoperfusion in the development of parenchymal and osseous abnormalities, as well as the contribution of developmental venous anomalies as compensatory mechanisms.

These findings underscore the major value of multimodal imaging in the characterization of these rare entities and in guiding diagnostic and prognostic assessment.

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