Dyke Davidoff Masson Syndrome Presenting with Recurrent Focal Seizures: A Case Report

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Abstract:

Dyke Davidoff Masson Syndrome (DDMS) is a rare neurological disorder characterized by unilateral cerebral hemiatrophy, compensatory calvarial hypertrophy, and chronic neurological deficits including epilepsy, hemiparesis, and cognitive impairment. The syndrome may be congenital, secondary to intrauterine vascular insults, or acquired following perinatal or postnatal ischemic injury. We report a 17-year-old female presenting with recurrent focal motor seizures with impaired awareness, characterized by left-sided tonic stiffening lasting three minutes followed by transient postictal confusion. Seizures began at age 12, with a history of febrile seizures in infancy. Neurological examination revealed left-sided hemiparesis, hyperreflexia, and a positive Babinski sign. Brain MRI demonstrated right cerebral hemiatrophy with cortical-subcortical encephalomalacia in the temporo-parietal region, mesencephalic atrophy with Wallerian degeneration (Stage 4), right basal ganglia atrophy, frontal sinus enlargement, and compensatory skull vault thickening — all consistent with DDMS. Hippocampal atrophy and an anomalous AICA looping (Type 2, CHAVDA classification) were additionally identified. Electroencephalography revealed continuous slowing activity and asymmetric background rhythm attenuation in the right hemisphere, indicative of an underlying epileptogenic focus. The patient was treated with phenytoin and carbamazepine, achieving improved seizure control. This case underscores the importance of early recognition of characteristic neuroimaging findings, comprehensive neurophysiological assessment, and a multidisciplinary management approach — including optimized antiseizure therapy and neurorehabilitation — to enhance functional outcomes and quality of life in patients with DDMS.

Figure/Scheme: Brain MRI (TIFF/JPEG) showing right cerebral hemiatrophy with cortical-subcortical encephalomalacia, mesencephalic atrophy, and compensatory skull thickening consistent with DDMS.

Keywords:

Dyke-Davidoff-Masson Syndrome, Focal seizure, Cerebral hemiatrophy, Neuroimaging, Epilepsy.