When Febrile Seizures Are Not Benign: An Unusual Cause of Dyke–Davidoff–Masson Syndrome

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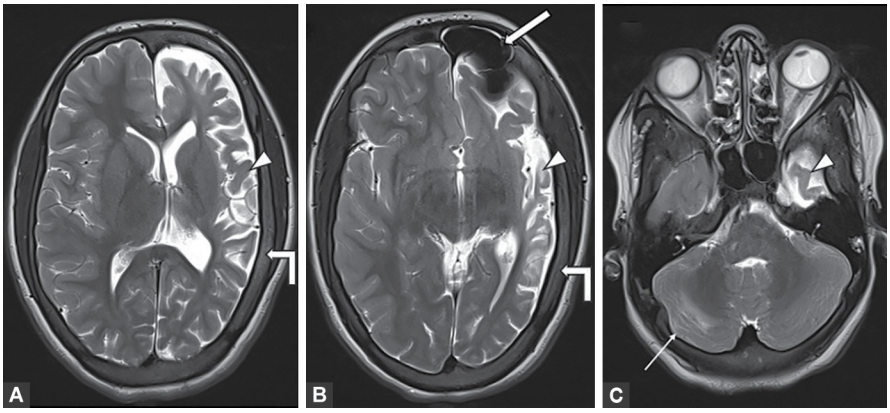
Received: 27 December 2025; *Accepted:* 05 March 2026

Journal of The Association of Physicians of India (2026): 10.59556/japi.74.1588

A 14-year-old girl presented with a history of recurrent seizures over the past year, varying in type—generalized tonic-clonic and right-sided focal seizures involving both upper and lower limbs. Birth and family history were unremarkable, and early developmental milestones were appropriate. However, her academic performance declined over the past 2 years, eventually leading her to drop out of school. At 7 months of age, she had an upper respiratory tract infection followed by febrile status epilepticus lasting 20 minutes and requiring 5 days of hospitalization. Neuroimaging, cerebrospinal fluid analysis, and blood cultures were normal. There were no focal neurological deficits at discharge. Antiepileptic medications were discontinued after two seizure-free years. Current examination showed mild right-sided spasticity with increased deep tendon reflexes and an extensor plantar response. Magnetic resonance imaging (MRI) revealed left frontal sinus hypertrophy, left calvarial thickening, left cerebral atrophy, and right-sided crossed cerebellar atrophy, suggestive of Dyke–Davidoff–Masson syndrome (DDMS) (Fig. 1). Electroencephalography (EEG) revealed intermittent sharp waves and background slowing over the left frontotemporal region. She was started on antiepileptic therapy, physiotherapy, and occupational and speech therapy.

Dyke–Davidoff–Masson syndrome is a rare neurological disorder characterized by cerebral hemiatrophy, hemiparesis, developmental delay, and seizures.[1] It can be congenital, often from intrauterine vascular insults, or acquired, secondary to perinatal trauma, infection, vascular events, or hemorrhage.[1] In the absence of antenatal, perinatal, or early developmental risk factors, the prolonged febrile seizure at 7 months represents the most likely neurological insult. Its temporal association with the characteristic neuroimaging findings supports a diagnosis of DDMS secondary to febrile status epilepticus. Febrile seizures are typically benign and self-limiting, with most children achieving complete neurological recovery. The association between febrile seizures and DDMS is rare and sparsely reported in the literature.[2] The proposed pathophysiological mechanism linking DDMS and febrile status epilepticus involves prolonged hypoxic-ischemic injury leading to reduced levels of brain-derived neurotrophic factors, impaired neuronal survival, and progressive cerebral atrophy.[3] Crossed cerebellar atrophy is an uncommon feature in DDMS, typically observed in patients with longstanding, severe unilateral cerebral damage.[4] It results from disruption of the corticopontocerebellar pathways connecting the affected cerebral hemisphere and the contralateral cerebellum.[4] The presence of crossed cerebellar atrophy in our case suggests a chronic and progressive process rather than an acute insult alone. Our case is notable for several atypical features, including delayed clinical presentation in adolescence, crossed cerebellar atrophy, and an association with febrile status epilepticus. DDMS should be differentiated from conditions such as Sturge–Weber syndrome, basal ganglia germinoma, Silver–Russell syndrome, Fishman syndrome, linear nevus sebaceous syndrome, hemiconvulsion-hemiplegia-epilepsy (HHE) syndrome, and Rasmussen encephalitis, based on clinical features and characteristic neuroimaging findings.[1] Prognosis is better if onset is after 2 years of age and seizures are well controlled. Hemispherectomy may be considered in selected patients with intractable epilepsy and hemiplegia. Long-term management requires a multidisciplinary approach, including antiepileptic therapy, physiotherapy, occupational therapy, speech therapy, and educational support.[1–3] Through this case, we highlight a rare presentation of DDMS following febrile status epilepticus and emphasize the need for long-term neurological follow-up.



Figs 1A to C: Axial T2-weighted MRI images: (A) Left cerebral atrophy (arrowhead) with ipsilateral calvarial thickening (bent arrow); (B) Left frontal sinus enlargement (thick arrow); (C) Contralateral cerebellar atrophy (thin arrow)

Conflict of Interest

None.

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How to cite this article: Ahammed J, Zaki SA, Reddy KK. When Febrile Seizures Are Not Benign: An Unusual Cause of Dyke–Davidoff–Masson Syndrome. J Assoc Physicians India 2026;74(7):88–89.

Financial Disclosure

None.

Authors' Contribution

Concept and design of study or acquisition of data or analysis and interpretation of data: SAZ, JA, KKR.

Drafting the article or revising it critically for important intellectual content: SAZ, JA, KKR.

Final approval of the version to be published: All authors approved the version of the manuscript to be published.

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