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MRI Revelations in Dyke-Davidoff-Masson Syndrome: Understanding Hemiatrophy through Imaging

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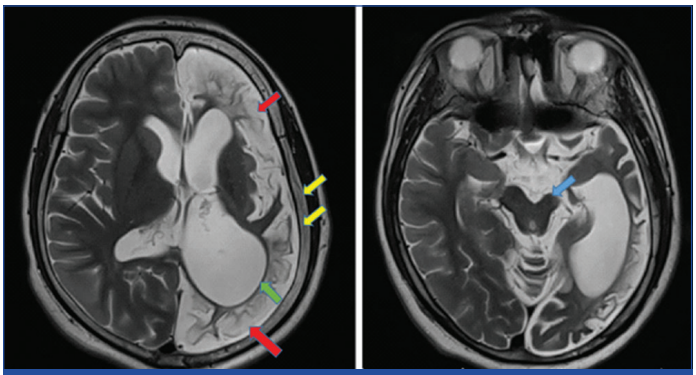
A 22-year-old male presented with involuntary limb movements for two days, characterised by pronation-supination of the upper limbs. The patient had a history of perinatal hypoxic-ischaemic insult, as reported by his family. He had epilepsy for the past eight years and was on regular carbamazepine therapy, previously at a maintenance dose of 20 mg/kg/day, divided into two doses. Despite medication, he continued to experience breakthrough seizures approximately every 2-3 months.

The developmental history was significant for delayed motor and language milestones. The patient also had academic difficulties during schooling, suggestive of mild cognitive impairment. The mother reported microcephaly and subtle facial asymmetry since early childhood. None of the other siblings had similar facial features or neurological symptoms. There was no history of head trauma, Central Nervous System (CNS) infection, or previous neurosurgery.

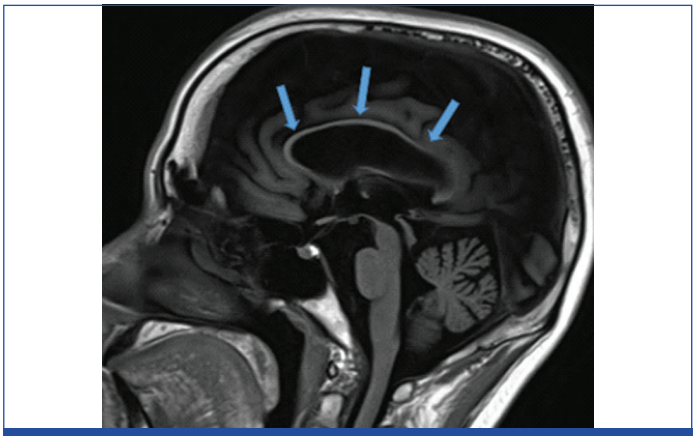
On neurological examination, right-sided hemiparesis was noted, more prominent in the upper limb, with spastic tone. Motor strength was graded 3/5 in the right upper limb and 4/5 in the right lower limb. Cranial nerves were intact, pupils were equal and reactive and auditory and visual functions were age-appropriate.

An earlier Computed Tomography (CT) scan demonstrated left cerebral hemiatrophy with calvarial thickening. A subsequent Magnetic Resonance Imaging (MRI) scan, performed three weeks after symptom onset, confirmed atrophy of the left cerebral hemisphere, with ipsilateral ventricular dilation, widened sulci and an enlarged Sylvian fissure. A leftward midline shift and compensatory calvarial hypertrophy (3 mm thicker than the contralateral side) were also noted. Quantitative MRI revealed over a 40% reduction in cortical thickness of the left hemisphere [Table/Fig-1-3].

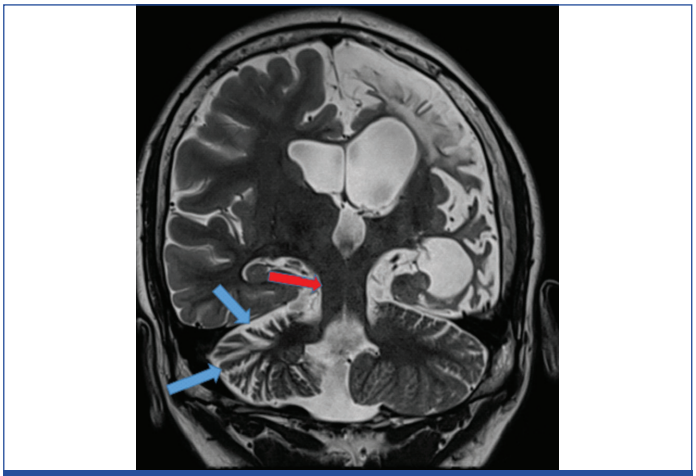
Based on the clinical and imaging findings, a diagnosis of Dyke-Davidoff-Masson Syndrome (DDMS) was established. The patient's carbamazepine was adjusted to 15 mg/kg/day, with weekly titration to 30 mg/kg/day, resulting in a favourable seizure response. He also began physiotherapy and occupational therapy and has remained seizure-free for six months on follow-up.



[Table/Fig-1]: T2WI axial slices of the brain showing diffused left-sided hemiatrophy with encephalomalacia and gliotic changes seen with abnormal prominence of sub-arachnoid spaces (red arrows), Severe left ex vacuo-ventriculomegaly was also noted (green arrow) with mild sub-falcine shift of midline structures towards the left and left-sided calvarial hypertrophy were seen (yellow arrows). Hemiatrophy of the left cerebral peduncle of midbrain was noted (blue arrows).



[Table/Fig-2]: T1WI sagittal slice of the brain showing thinned out corpus callosum (blue arrows).



[Table/Fig-3]: T2WI coronal slice of the brain showing right cerebellar hemisphere (blue arrows) and brainstem (red arrow) also showing atrophic - crossed cerebellar diaschisis.

The DDMS is characterised by cerebral hemiatrophy accompanied by compensatory cranial changes. First described in 1933 [1], DDMS presents with seizures, facial asymmetry, contralateral hemiparesis and variable cognitive deficits [2,3].

The condition is categorised into congenital and acquired forms. The congenital variant arises from in utero vascular insults, while the acquired form results from early childhood injuries, including perinatal hypoxic-ischaemic encephalopathy, infections, or trauma [3,4]. In the present case, the probable etiology is acquired DDMS secondary to perinatal hypoxic-ischaemic injury, as suggested by the history and the neuroimaging features that became more apparent in adolescence.

The MRI plays a central role not only in diagnosing DDMS but also in distinguishing congenital from acquired forms. Imaging signs include ipsilateral calvarial thickening, frontal sinus hyperpneumatisation and petrous ridge elevation, which typically evolve as the patient ages. Intellectual impairment, as seen in this patient, is also a commonly reported feature [4-6].

Although most patients have refractory epilepsy, some may respond well to optimised anticonvulsant therapy. In drug-resistant cases with disabling hemiparesis, hemispherectomy may be curative in up to 85% of cases [5,7].

The DDMS remains under-recognised, particularly in atypical age groups such as adolescents and young adults, where earlier signs may have been subtle or misattributed. The present case reinforces the importance of recognising characteristic MRI findings—not only for diagnosis but also for prognostic evaluation and treatment planning. MRI is instrumental in differentiating congenital from acquired DDMS, guiding therapeutic decisions such as medication adjustments or surgical referrals. Early identification enables tailored interventions, including seizure control, neurorehabilitation and cognitive support, thereby improving long-term outcomes in these patients.

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