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Case Report Status Epilepticus in a 45-Year-Old Male with Dyke-Davidoff-Masson Syndrome

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Abstract: A 45-year-old male presented with status epilepticus and was subsequently diagnosed with Dyke-Davidoff-Masson Syndrome (DDMS), a rare neurological disorder characterized by cerebral hemiatrophy, seizures, hemiplegia, and cranial asymmetry. This case highlights the clinical features, imaging findings, and management of DDMS in adulthood, contributing to the limited literature on late presentations of this syndrome. The patient's acute condition was managed with antiepileptic therapy, and follow-up strategies were established to address chronic neurological deficits. This report underscores the need for heightened awareness of DDMS in adult seizure presentations and provides insights into its long-term management.

Keywords*: *Dyke-Davidoff-Masson Syndrome, Status Epilepticus, Cerebral Hemi atrophy, Seizures, Hemiplegia.

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I. INTRODUCTION

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare condition first described in 1933, characterized by cerebral hemi atrophy, seizures hemiparesis, and intellectual disability. While predominantly diagnosed in childhood or adolescence, adult presentations, particularly with status epilepticus, are uncommon. The syndrome may result from prenatal, perinatal, or early childhood insults to the brain, leading to compensatory skull and sinus changes visible on imaging.

This case report discusses the presentation of a 45-year-old male with DDMS who developed status epilepticus. We aim to provide a detailed clinical account and discuss management challenges in adult cases.

II. CASE PRESENTATION

A 45-year-old male presented to the emergency department with generalized tonic-clonic status epilepticus lasting over 30 minutes, not responding to benzodiazepines, levetiracetam, and phenytoin. The patient was intubated, placed on ventilator support, and started on midazolam infusion, following which seizures were terminated. The patient was extubated on day 5, and there were no further episodes of seizures.

The patient had a history of recurrent focal seizures since childhood, which were poorly controlled due to non-compliance with antiepileptic medication. He also had poor scholastic performance with intellectual disability since childhood.

He had known left-sided hemiplegia from early adulthood but had no documented diagnosis of DDMS. He had been receiving treatment from a local practitioner and was managed as a case of stroke with antiplatelet. There was no significant family history of epilepsy or neurological disorders.

On clinical examination, there was hemiplegia on the left side with muscle wasting and hyperreflexia. Hemi facial asymmetry was more pronounced on the left, with spasticity in the left upper and lower limbs.

III. INVESTIGATIONS

- *Brain MRI*

Findings consistent with left-sided cerebral hemi atrophy, ventricular enlargement (ipsilateral ventriculomegaly), hyper pneumatization of the left frontal sinus, and compensatory skull changes.

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➢ *Electroencephalogram (EEG)*

- Include findings, if available.
- Laboratory Workup Serum electrolytes, blood glucose, and renal function tests were within normal limits.

IV. DISCUSSION

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological condition characterised by a triad of clinical features: hemiplegia or hemiparesis, seizures, and intellectual disability. Radiological findings include cerebral hemi atrophy, ipsilateral compensatory skull thickening, and hyperpneumatization of the frontal sinuses.

➢ *Pathophysiology*

DDMS results from significant brain injury occurring during prenatal, perinatal, or early childhood periods. Congenital cases are attributed to intrauterine events such as vascular occlusion, hypoxia, or infection, while acquired cases result from trauma, infections like meningitis or encephalitis, cerebrovascular accidents, or prolonged seizures.

➢ *Clinical Spectrum*

DDMS typically presents during childhood or adolescence with seizures, hemiplegia, and cognitive impairment. However, adult-onset cases, particularly presenting as status epilepticus, are rare and require a high index of suspicion.

➢ *Management*

Management focuses on symptom control and improving the quality of life.

- Acute management**:** Benzodiazepines and antiepileptic drugs.
- Long-term seizure control**:** Maintenance of antiepileptic drugs; surgical options (e.g., hemispherectomy) for refractory cases.
- Rehabilitation: Physical therapy for motor deficits, speech therapy for cognitive

➢ *Prognosis*

Prognosis varies, with early-onset cases associated with worse outcomes. Late-onset cases, as in this patient, may achieve a reasonable quality of life with appropriate management.

➢ *Acknowledgements*

None

➢ *Declarations*

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- *Conflict of Interest***:** None declared
- *Ethical Approval***:** Not required

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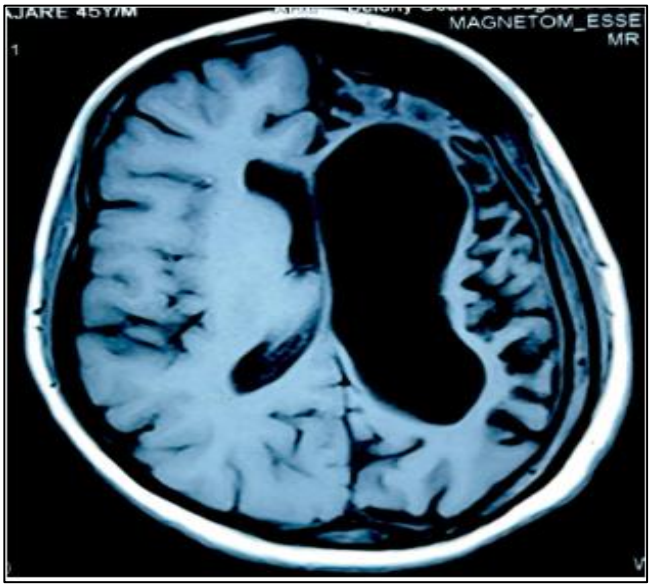
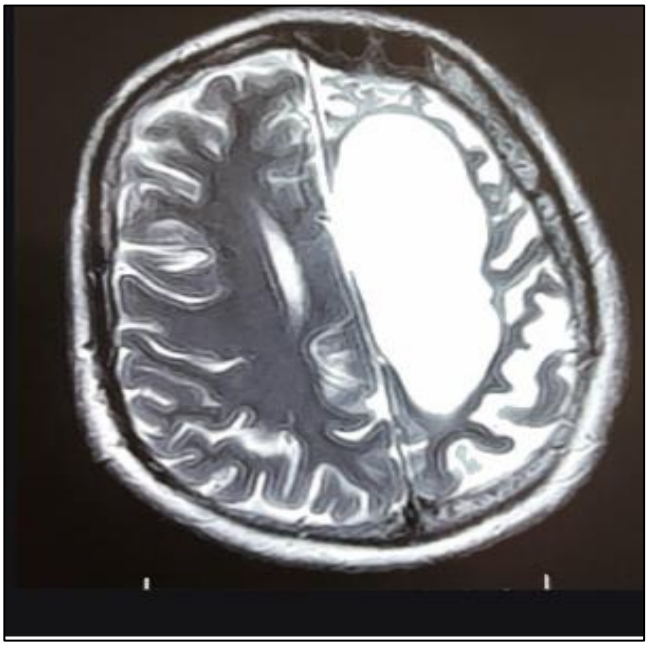
➢ *Figures and Legends*





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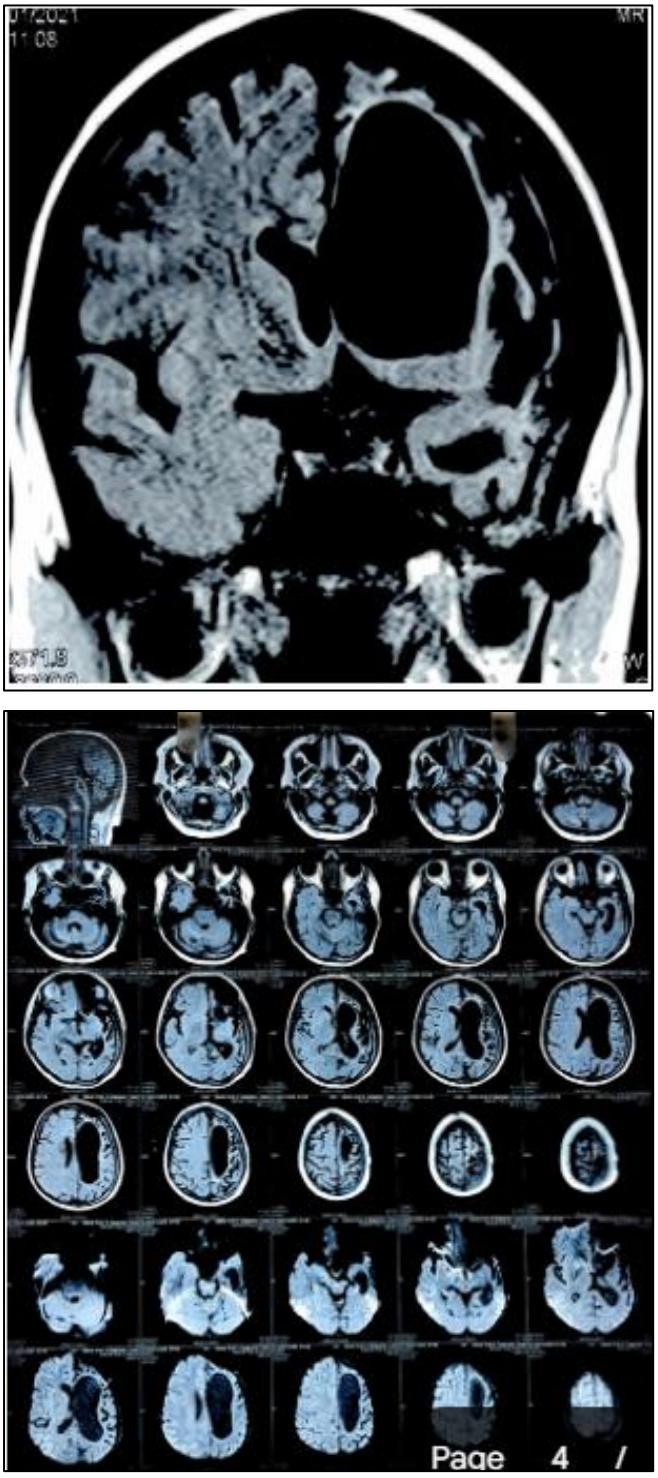

Fig 1 MRI images Showing Cerebral Hemi atrophy, Ventricular Enlargement (Ipsilateral Ventriculomegaly), Hyperpneumatization of the left Frontal Sinus, and Compensatory Skull Changes.