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RESEARCH ARTICLE

DYKE-DAVIDOFF-MASSON SYNDROME (DDMS)

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ABSTRACT

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare medical condition caused by brain damage that may occurring in utero, perinatally or in early infancy. The diagnosis is usually difficult and is based on patient's history , clinical presentation and correlation with CT findings. We present a case of a 25-year-old male admitted with recurrent seizures, right-sided facial deviation, speech difficulties, right-hand stiffness, and progressive walking difficulty since age 12.CT imaging showed left-sided cerebral hemiatrophy, ventriculomegaly, hyperpneumatization of the sinus, reduced cortical vein caliber, and right-sided skull thickening, all indicative of DDMS. The diagnosis was based on the patient's history, clinical presentation, and CT findings. Diagnosing DDMS is challenging due to limited awareness and varied symptoms. While CT is crucial for diagnosis, early signs may not be apparent. There is no standardized treatment; management is primarily symptomatic. Early diagnosis is vital for supporting mental and physical development through a multidisciplinary approach.

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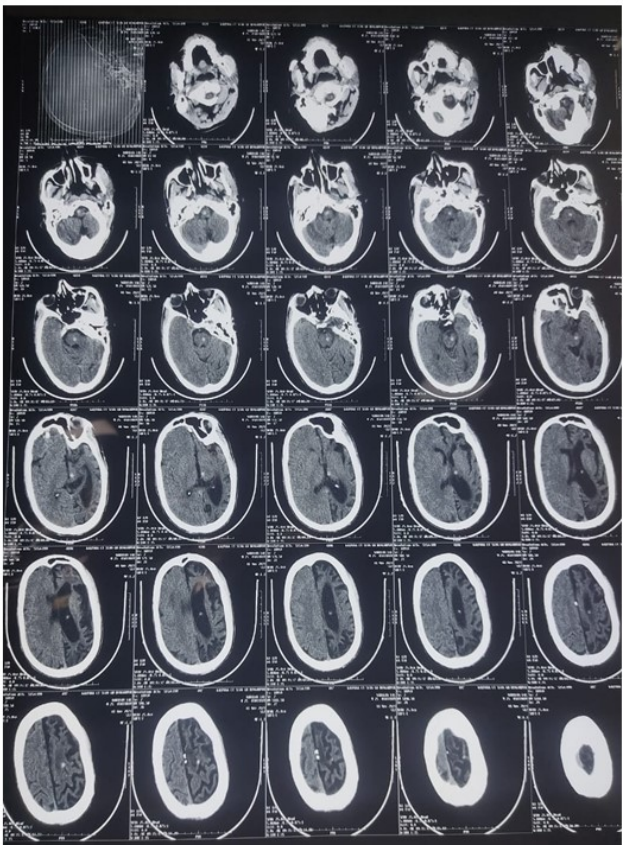
INTRODUCTION

Dyke-Davidoff-Masson Syndrome (DDMS) was first identified by Dyke, Davidoff, and Masson in 1933. It is a neurological condition marked by hemiparesis, facial asymmetry, seizures, learning disabilities, and intellectual impairment. The syndrome can be either congenital or acquired, typically emerging in childhood due to alterations in brain development during intrauterine life or early childhood. Radiographic indicators on CT often include cerebral hemiatrophy, ipsilateral ventriculomegaly, compensatory skull enlargement, and pronounced sulcal spaces. DDMS due to its rarity has the potential for misdiagnosis , given the limited documentation in medical literature. The syndrome can profoundly affect patient's devlopment and overall quality of life.

Case presentation: 25-year-old male resident of Dungarpur, Rajasthan ,presented with complaint of seizures, speech difficulty, facial deviation, and progressive right-sided hemiparesis presented to the medicine outpatient department . The patient was born at full term to non-consanguineous parents with no significant antenatal or perinatal complications. He achieved all developmental milestones and appeared normal till 12 years of age. Thereafter, he experienced a two day febrile episode followed by multiple focal seizures that later became generalized to tonic-clonic seizures, each lasting 2-3 minutes, characterized by stiffening of the right upper extremity and upward gaze deviation. He was admitted to a private hospital, treated, and started on antiepileptic medication. He has had multiple seizure episodes since, attributed to non-adherence to antiepileptic medication. Gradually, the patient developed a progressive right-sided weakness in both upper and lower extremities, facial deviation, and difficulties in walking and writing. . Despite these neurological impairments, his parents reported no hearing or visual problems. On physical examination the patient was alert, oriented, and responsive to commands.He had an intelligence quotient (IQ) of 55, indicating mild mental retardation. Vital signs were within normal range: Blood pressure 110/72 mmHg, Heart rate 82 beats per minute, Respiratory rate 20 per minute, Temperature 96°F, and Oxygen saturation 98%. His height was 180 cm and weight was 63 kg, and body mass index (BMI) of 19.4 kg/m². Vision and hearing assessments were normal. Cranial nerve and sensory examinations were unremarkable, and no neurocutaneous markers were observed.However, his gait was abnormal. Neurological examination demonstrated 3/5 muscle strength in the right upper and lower extremities, brisk reflexes, and a flexor plantar response, with 2/5 power at the right wrist joint. The left side upper and lower extremities scored 5/5. spasticity was present in the right upper and lower limbs. Sensation and proprioception were intact on both side. No signs of neck rigidity, cerebellar dysfunction, or bladder-bowel involvement were noted. The laboratory investigations, including Complete Blood Count and Comprehensive Metabolic panel (renal and liver function, CRP, and electrolytes), were within normal limits.

The CT scan of the brain revealed atrophy of the left cerebral hemisphere, ipsilateral ventricular dilation, and calvarial thickening of the left hemicranium. There was also sulcal widening and gyral atrophy in the left cerebral hemisphere. These findings suggested a final diagnosis of Dyke-Davidoff-Masson Syndrome (DDMS),that was corroborated by the clinical presentation. The diagnosis of DDMS was made after excluding other neurological conditions presenting with similar symptoms adolscent population. Patient was started on Tab Phenytoin 200mg BD, Tab Baclophen 5mg TDS. His parents were educated about DDMs and its potential progression. The patient was advised to undergo 6 monthly neurological follow-ups to monitor symptomatic changes and to consider interventions such as physiotherapy as the disease progresses



DISCUSSION

Dyke-Davidoff-Masson Syndrome (DDMS) was first described in 1933 by Dyke, Davidoff, and Masson. This rare neurological disorder is characterized by cerebral hemiatrophy or hypoplasia. Its etiology can be either congenital or acquired, often resulting from prenatal or early childhood brain injuries, typically traumatic in nature. Clinical presentations vary depending on the extent of brain injury and commonly include facial asymmetry, recurrent seizures, contralateral hemiplegia, learning disabilities, intellectual disability, and delayed developmental milestones. In some cases, psychiatric disorders such as schizophrenia and schizoaffective disorder have also been reported. Typical radiographic findings include cerebral hemiatrophy with ipsilateral ventriculomegaly, compensatory skull enlargement, and significant sulcal spaces. Other radiologic observations may include hyperpneumatization of the frontal sinuses, ethmoid air cells, mastoid air cells, and air cells of the petrous temporal bone on the affected side. These are thought to be compensatory changes due to congenital or acquired brain injuries that decrease cerebral cortical volume.
The congenital form is attributed to in-utero cerebral insults from vascular malformations, cerebral infarction, mid-aortic arch coarctation, gestational vascular occlusion, and infections. The acquired form is linked to perinatal or postnatal cerebral insults such as hypoxia, birth trauma, tumors, infections, prolonged febrile seizures, and intracranial hemorrhage. DDMS can affect either hemisphere in both sexes, but it is more commonly seen in males and the left hemisphere. Diagnosing DDMS can be challenging due to low awareness and varied clinical presentations. For instance, the patient initially presented with seizures but later developed facial deviation, left-sided hemiparesis, speech difficulty, and mental deterioration. In such cases, a clinician might only treat the seizures without identifying the underlying condition, especially in underprivileged areas where advanced imaging like MRI is less accessible. Although CT and MRI are the gold standards for diagnosing DDMS, early disease manifestations are better appreciated on MRI due to its detailed cross-sectional imaging.

A comprehensive history, extensive neurological and cognitive assessment, along with characteristic radiologic findings, are essential for an accurate diagnosis of DDMS. Differential diagnoses include Rasmussen encephalitis, Sturge-Weber syndrome (SWS), Fishman syndrome, basal ganglia germinoma, linear nevus syndrome, and Silver-Russell syndrome, as they share similar imaging findings and clinical presentations with DDMS. Rasmussen encephalitis is an immune-mediated brain disorder in children, characterized by cognitive defects and seizures, with hemispheric atrophy but no calvarial changes. SWS is marked by a triad of glaucoma, port-wine nevus, and leptomeningeal angiomas, leading to stroke-like symptoms, seizures, hemiparesis, intellectual disability, and developmental delays. Fishman syndrome is a rare congenital neurocutaneous disorder featuring intellectual disability, cerebral calcification, seizures, ipsilateral cerebral malformation, unilateral temporofrontal lipomatosis, and leptomeningeal lipomatosis. Basal ganglia germinoma may present with cerebral hemiatrophy and progressive hemiparesis. Linear nevus syndrome features facial nevus, cyclic refractory seizures, growth restriction, intellectual disability, and unilateral ventriculomegaly.

Silver-Russell syndrome is an imprinting gene disorder characterized by severe intrauterine and postnatal growth restriction, distinct facial features, fifth finger clinodactyly, and hemihypertrophy without affecting mental capacity. There is no standard protocol for managing DDMS; treatment is primarily symptomatic, including anticonvulsants for seizures. In cases of intractable disabling seizures, hemispherectomy is an available neurosurgical option with an 85% success rate. Long-term management involves physiotherapy for hemiparesis, occupational therapy, speech therapy for speech defects, psychiatric counseling, and medications as needed.

CONCLUSION

DDMS is a rare syndrome with limited case reports in medical literature, making it prone to misdiagnosis by less experienced clinicians if not thoroughly investigated. Early detection and diagnosis are crucial for supporting the child's mental and physical development through a multidisciplinary approach. Imaging studies are essential for identifying such conditions, and while MRI is more costly, it should be considered alongside CT scans to ensure accurate diagnosis and avoid missed cases. Increased awareness of DDMS is necessary so that it is included in differential diagnoses and not overlooked. The absence of a standardized management protocol for DDMS and social stigma highlights the need for further research and a deeper understanding of the syndrome.

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