(International Print/Online Journal)

SJIF IMPACT FACTOR: 6.829
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 9, Issue 2 , Page No: 301-304
March-April 2026

IJMSCR

A Shrinking Hemisphere, A Growing Challenge: Dyke-Davidoff-Masson Syndrome

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Type of Publication: Original Research Paper
Conflicts of Interest: Nil

Abstract

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological condition characterized by cerebral hemiatrophy, contralateral hemiplegia, facial asymmetry, and seizures, compensatory hypertrophy of the skull and sinuses. It results from an insult to the developing brain in the fetal or early childhood period

We report the case of a 7-year-old male child, born of a non-consanguineous marriage with a normal perinatal history. The child was asymptomatic until 1.5 years of age, when he was presented with refractory seizures followed by left-sided hemiparesis and right sided facial asymmetry. Even on multiple antiepileptics due to non compliance he presented again at 7 years of age with recurrence of focal seizures, left upper limb weakness. Magnetic Resonance Imaging (MRI) of the brain revealed prominence of cortical sulci and thinning of gyri in the right cerebral hemisphere, ipsilateral ventricular dilatation, and mild right-sided calvarial thickening.

The findings were consistent with a diagnosis of Dyke-Davidoff-Masson Syndrome. This case highlights the importance of neuroimaging in children presenting with hemiparesis and seizures and emphasizes the critical role of medication compliance and physiotherapy in managing this static but debilitating condition.Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological disorder marked by cerebral hemiatrophy, paralysis on the opposite side of the body (contralateral hemiplegia), facial asymmetry, and seizures. It is also characterized by the skull and sinuses showing compensatory overgrowth (hypertrophy). This condition arises from damage to the developing brain, either in utero or during early childhood.

We present the case of a 7-year-old boy from a non-consanguineous marriage with an unremarkable history around birth. He was symptom-free until 1.5 years old, when he started having refractory seizures, which were followed by left-sided weakness (hemiparesis) and right-sided facial asymmetry. Despite being prescribed multiple antiepileptic medications, his non-compliance led to a recurrence of focal seizures and weakness in his left upper limb at the age of 7. Magnetic Resonance Imaging (MRI) of the brain revealed enlarged cortical sulci and thinning of the gyri in the right cerebral hemisphere, along with ipsilateral ventricular dilation and minor thickening of the right calvaria (skull bone). These findings confirmed the diagnosis of Dyke-Davidoff-Masson Syndrome. This case emphasizes the critical necessity of neuroimaging for children presenting with a combination of hemiparesis and seizures, and it underlines the vital importance of adhering to medication regimens and physical therapy in managing this static yet debilitating disorder.

Keywords: Dyke-Davidoff-Masson Syndrome, Cerebral Hemiatrophy, Hemiparesis, Refractory Seizures, Calvarial Thickening

Introduction

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare clinical entity first described by Dyke, Davidoff, and Masson in 1933. They observed specific skull radiographic changes in a series of nine patients presenting with hemiparesis and cranial asymmetry. The syndrome is defined by a triad of clinical features: seizures, facial asymmetry, and contralateral hemiplegia or hemiparesis, often accompanied by mental retardation or learning disabilities.

The etiology of DDMS is broadly classified into two categories: congenital and acquired. The congenital type results from vascular occlusion or maldevelopment during intrauterine life. The acquired type, as seen in our case, occurs due to brain insults such as trauma, infection, intracranial hemorrhage, or ischemia during the perinatal period or early childhood typically before the age of two or three years.

The pathogenesis involves the atrophy or hypoplasia of one cerebral hemisphere. Because this volume loss occurs during a period of skeletal growth, the cranial vault undergoes compensatory changes to fill the resulting vacuum. Characteristic radiological features include cerebral hemiatrophy, dilatation of the ipsilateral lateral ventricle, and compensatory hypertrophy of the homolateral skull (calvarial thickening) and paranasal sinuses.

While the clinical presentation can vary, seizures are a predominant feature and can be refractory to treatment. Mental retardation is not universally present and correlates with the extent of brain injury. Diagnosis is confirmed via Computed Tomography (CT) or Magnetic Resonance Imaging (MRI), which visualize the parenchymal loss and compensatory skeletal changes.

This case report discusses a 7-year-old male with the acquired form of DDMS, highlighting the clinical course, characteristic imaging findings, and the importance of long-term management strategies involving antiepileptic therapy and rehabilitation.Dyke-Davidoff-Masson Syndrome (DDMS), a rare clinical entity, was first documented in 1933 by Dyke, Davidoff, and Masson. Their initial observations focused on distinct skull radiographic changes in nine patients who presented with hemiparesis and cranial asymmetry.

The syndrome is classically defined by a clinical triad: contralateral hemiplegia or hemiparesis, seizures, and facial asymmetry. Intellectual disability or learning difficulties often accompany these core features.

Etiology and Pathogenesis

DDMS is categorized as either congenital or acquired. The congenital form stems from vascular occlusion or developmental issues that occur prenatally. In contrast, the acquired form, as exemplified by the case discussed here, results from brain injuries such as trauma, ischemia, intracranial hemorrhage, or infection sustained during the perinatal period or early childhood, typically before the age of two or three years.

The underlying pathogenesis involves atrophy or hypoplasia of one cerebral hemisphere. Because this loss of brain volume occurs while the skeleton is still growing, the cranial vault compensates to fill the vacant space. This leads to characteristic radiological findings, including cerebral hemiatrophy, dilation of the ipsilateral lateral ventricle, and compensatory hypertrophy (thickening) of the homolateral skull (calvaria) and paranasal sinuses.

Clinical Features and Diagnosis

While the clinical picture is variable, seizures are a primary and often treatment-resistant feature. Mental retardation is not a universal finding; its presence correlates with the extent of the brain injury.

Diagnosis is confirmed using Computed Tomography (CT) or Magnetic Resonance Imaging (MRI), which clearly show the parenchymal volume loss and the associated compensatory skeletal changes.

Case Report

This report details the case of a 7-year-old male with the acquired form of DDMS. It aims to highlight the clinical course, characteristic imaging findings, and the critical need for long-term management strategies, which encompass antiepileptic therapy and rehabilitation.

Case Report

A 7-year-old male child presented with complaints of recurrent focal seizures involving the left upper limb and a decrease in power on the left side. The child was born at full term to non-consanguineous parents with an uneventful antenatal and perinatal history. He was developmentally normal and asymptomatic until 1.5 years of age. When he was presented with refractory

Volume 9, Issue 2; March-April 2026; Page No 301-304
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seizures lasting for 7 days, followed by hemiparesis involving upper limb more than lower limb with facial asymmetry, characterized by deviation of the mouth to the left side and loss of the left nasolabial fold.

He was initiated on antiepileptic medication and remained seizure-free for approximately 5 years. symptoms recurred at 7 years of age due to non compliance. The parents reported focal seizures involving the left upper limb accompanied by a staring look. These episodes occurred 4-5 times per day, lasted 1-2 minutes, and were self-aborting. The child was developmentally normal for his age.On general examination, the child appeared malnourished with Anthropometric measurements (Height-111cm, weight-15kg) revealed significant atrophy of the left upper limb, with a mid-arm circumference of 15 cm compared to 16 cm on the right side, head circumference of 49cm. No neurocutaneous markers. Gait was normal. BCG scar was present

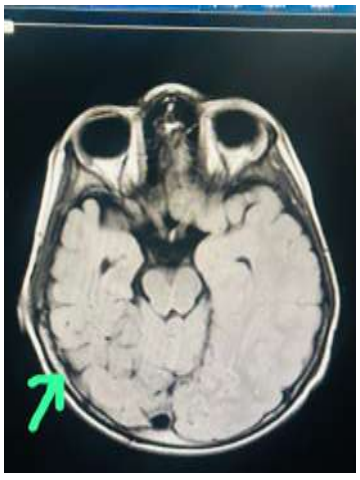
Neurological Examination revealed upper motor neuron right-sided facial nerve involvement, evidenced by deviation of the angle of the mouth to the left. Other cranial nerves were intact. Fundus examination was normal. Decreased power (4/5) was noted in the left upper limb, with proximal weakness being more prominent. Power in the other limbs was normal. Deep tendon reflexes were brisk in the left upper limb (specifically the biceps jerk). Reflexes in the other limbs were normal. Sensory System Intact. Cardiovascular, respiratory, and abdominal examinations were within normal limits. With all the above findings we came to differential diagnosis of structural brain anamoly and vascular events affecting right side of brain

Neuroimaging Magnetic Resonance Imaging (MRI) of the brain (Plain + Contrast) was performed. The scan revealed:

1. Prominence of the cortical sulci in the right cerebral hemisphere.
2. Thinning of the gyri, predominantly in the right high parietal region.
3. Asymmetrical prominence (dilatation) of the right lateral ventricle.
4. Mild thickening of the calvarium on the right side.

These findings—cerebral hemiatrophy with ipsilateral ventricular dilatation and compensatory calvarial thickening—are consistent with a diagnosis of right-sided Dyke-Davidoff-Masson Syndrome.

Mri Brain Showing Calvarial Thickening With Cerebral Hemiatrophy



Discussion

Although this condition is considered rare in general literature,our institute has reported 3 cases,suggesting that it is not uncommon in our Patient population

In a case series by Chen and colleagues involving seven patients,hemiparesis, cognitive impairment, seizures, and intellectual disability were the most common clinical features..Our patient showed a similar pattern, presenting with early-onset focal seizures and weakness on the opposite side of the body. MRI of the brain revealed classical features of DDMS, including unilateral cerebral atrophy, enlargement of the ipsilateral ventricle, and calvarial thickening. Typical radiological findings were present in all cases, in case series by chen and colleagues

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emphasizing the importance of neuroimaging in confirming the diagnosis.

In a case series from central India where mental retardation and facial asymmetry were commonly reported, our case exhibited preserved cognitive and speech development, which may reflect a later or less extensive brain insult

Refractory epilepsy is emphasized in some reports (e.g., cases requiring hemispherectomy reported on Griessenauer CJ at all), our patient achieved seizure control with antiepileptic therapy, underscoring the seizure severity and response to antiepileptics in DDMS

According to dr vijaybaburao Sonawane et.al mental retardation was present, but seizures developed at the age of 2 years. In our patient seizures presented at 1.5 year of age and there is no mental retardation

According to Dr. Sowjan et al. 9-year-old child with DDMS who had developmental delay and speech impairment, which contrasts with the preserved neurodevelopment seen in our patient

Conclusion

Dyke-Davidoff-Masson Syndrome should be considered in the differential diagnosis of any child presenting with hemiparesis and seizures. While the condition is static, early diagnosis via neuroimaging is crucial for prognostication. This case emphasizes that with strict adherence to antiepileptic medication and consistent physiotherapy, patients can achieve a good quality of life and functional independence.

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