

**DYKE DAVIDOFF MASSON SYNDROME IN A FUNCTIONALLY INDEPENDENT ADULT
WITH CHILDHOOD ONSET HEMIPARESIS: A CASE REPORT****Dr. Hruthik Kumar T.^{1*}, Dr. Alekya Katti², Dr. Sai Kaushik Rathod³**¹Post Graduate, MD Internal Medicine, CMRIMS, Kandlakoya, Hyderabad, Telangana, India.²Associate Professor, Consultant, Department of Internal Medicine, CMRIMS, Kandlakoya, Hyderabad, Telangana, India.³Post Graduate, MD Internal Medicine, CMRIMS, Kandlakoya, Hyderabad, Telangana, India.***Corresponding Author: Dr. Hruthik Kumar T.**

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ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological disorder characterized by cerebral hemiatrophy, contralateral hemiparesis, facial asymmetry, and recurrent seizures. It typically presents during childhood, while diagnosis in adulthood is uncommon, particularly in individuals who remain functionally independent. We report the case of a 37-year-old male with childhood-onset hemiparesis who remained ambulatory and independent in activities of daily living. The diagnosis of DDMS was established incidentally on magnetic resonance imaging (MRI) during evaluation for an unrelated illness. This case highlights the variable clinical spectrum of DDMS and emphasizes that mild cases may remain undiagnosed until adulthood.

INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) was first described by Dyke, Davidoff, and Masson in 1933.^[1] It is an uncommon neurological disorder, predominantly affecting the pediatric population, although its exact prevalence remains unknown. DDMS is characterized by cerebral hemiatrophy or hypoplasia, contralateral hemiparesis, facial asymmetry, and seizures, resulting from an insult to the developing brain during fetal life or early childhood.^[2]

Adult presentation is unusual and has been only rarely reported in the literature.^[3] We report the case of a 37-year-old male in whom DDMS was diagnosed incidentally. As functionally independent adults with previously undiagnosed DDMS are rarely described, this case adds to the limited literature and broadens the recognized clinical spectrum of this uncommon condition.

CASE PRESENTATION

A 37-year-old male presented to the Department of General Medicine with complaints of fever. During neurological examination, mild right-sided hemiparesis

was noted, which had been present since early childhood. Despite the weakness, he remained ambulatory, performed all activities of daily living independently, worked as a daily-wage laborer, and was married with two children.

There was no history of seizures, cognitive impairment, progressive neurological deterioration, or worsening of weakness, unlike the typical clinical presentation of this syndrome.^[4]

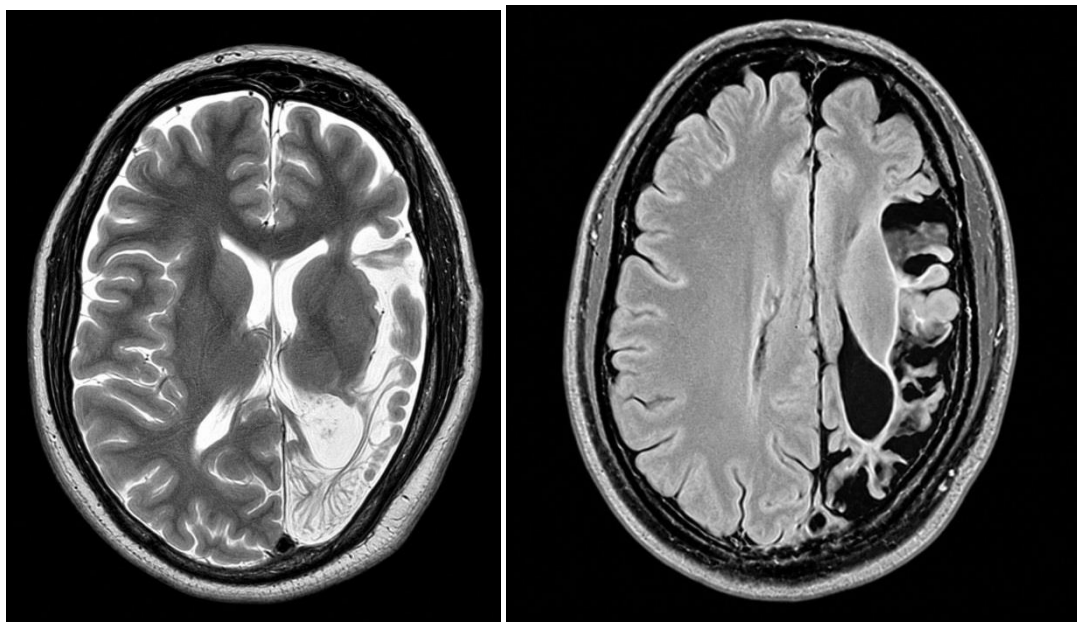
Magnetic resonance imaging (MRI) of the brain demonstrated marked unilateral cerebral hemiatrophy with imaging features consistent with Dyke-Davidoff-Masson syndrome. The diagnosis had never been established previously because of his relatively preserved functional status and the absence of significant neurological disability.

Only a few cases have been reported in which DDMS remained undiagnosed until adulthood because of mild or absent clinical manifestations.^[5] Adult presentations of DDMS without significant neurological disability are particularly rare.

DISCUSSION

Dyke-Davidoff-Masson syndrome is a rare neurological disorder characterized by cerebral hemiatrophy or hypoplasia involving one cerebral hemisphere, associated with ipsilateral ventricular dilatation,

prominence of the cortical sulci and cisternal spaces, reduced size of the ipsilateral cranial fossa, and varying degrees of midline shift. Clinically, it commonly presents with contralateral hemiparesis, seizures, facial asymmetry, and developmental delay.^[7]



The syndrome is reported to occur slightly more frequently in males, with left cerebral hemisphere involvement being more common.^[8]

DDMS results from cerebral insult during fetal development or early childhood and is broadly classified into congenital and acquired forms. In the congenital type, cerebral vascular anomalies leading to fetal cerebral injury are considered the primary cause.^[9] The acquired type results from central nervous system injury occurring during the perinatal period or secondary to conditions such as trauma, tumors, infections, ischemia, hemorrhage, or prolonged febrile seizures.^[10]

Clinical manifestations include varying degrees of facial asymmetry, contralateral hemiparesis, seizures, impaired speech, and intellectual disability.^[11] Neuropsychiatric manifestations have also been reported in some patients.^[12]

Management of DDMS is primarily symptomatic and depends on the patient's clinical presentation. Antiepileptic medications are indicated for seizure control, while physiotherapy, occupational therapy, and speech therapy play important roles in improving functional outcomes.

Although asymptomatic or minimally symptomatic adults with DDMS have been described, only a limited number of such cases have been reported in the literature.^[13] Our patient remained functionally independent despite childhood-onset hemiparesis and had never experienced seizures or progressive

neurological deficits. This case underscores the broad clinical spectrum of DDMS and highlights the importance of considering this diagnosis in adults with longstanding non-progressive hemiparesis, even in the absence of seizures or significant disability.

CONCLUSION

Dyke-Davidoff-Masson syndrome is a rare neurological disorder that is typically diagnosed during childhood, while functionally independent adult cases remain exceptionally uncommon. This case highlights that DDMS can remain undiagnosed until adulthood in individuals with minimal neurological deficits and no history of seizures. Recognition of such atypical presentations broadens the clinical spectrum of this rare condition and emphasizes the importance of considering DDMS in adults with longstanding, non-progressive hemiparesis. Further reporting of similar cases will improve understanding of its varied clinical presentation.

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