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CASE REPORT

# A Rare Case in a Pediatric Patient: Dyke-Davidoff-Masson Syndrome

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**Abstract**

Dyke-Davidoff-Masson syndrome (DDMS) is a clinical condition characterized by epilepsy, hemiplegia or hemiparesis, mental retardation, facial asymmetry, psychiatric disorders, sensorineural hearing loss, cerebral atrophy on neuroimaging, excessive enlargement and air increase in the paranasal sinuses, and unilateral skull thickening. In this paper, a 16-year-old male patient who was followed up with diagnoses of epilepsy, intellectual disability, and left hemiparesis, and who was diagnosed with DDMS upon the detection of right cerebral atrophy, thickening in the right hemiclavarial bone structures, and increased aeration of the frontal sinuses on neuroimaging, is presented. DDMS, a rare case in the literature, is emphasized for consideration in the differential diagnosis of cerebral hemiatrophy.

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## Introduction

Dyke-Davidoff-Masson syndrome (DDMS) was first described by Dyke, Davidoff, and Masson in 1933 and is a clinical condition characterized by epilepsy, hemiplegia or hemiparesis, mental retardation, facial asymmetry, psychiatric disorders, sensorineural hearing loss, cerebral atrophy on neuroimaging, excessive enlargement and air increase in the paranasal sinuses, and unilateral skull thickening.[1-2] In this paper, a 16-year-old male patient diagnosed with DDMS, which is rare in the literature, is presented.

## Case

A 16-year-old male patient, followed up for epilepsy, left hemiparesis, and intellectual disability, presented to our clinic with increased irritability and restlessness in recent times. His medical history revealed that he was born at term, did not receive incubator care, and had normal developmental milestones until one year of age. At one year old, he experienced generalized tonic-clonic seizures, for which levetiracetam treatment was initiated. Subsequently, he exhibited developmental delays, spoke his first meaningful word and started walking at 2.5 years of age. As his seizures continued during that period, sodium valproate was added to his treatment. He received special education due to intellectual disability. There was no history of consanguinity between his parents, and no history of epilepsy or other neurological diseases in his family.

On neurological examination, he was conscious, had limited cooperation, aggressive behaviors and stereotypies, facial asymmetry, and left hemiparesis. His cognitive development was observed to be delayed compared to his peers.

Electroencephalography showed focal epileptic abnormalities originating from the right frontocentrotemporal region. Cranial magnetic resonance imaging (MRI) revealed encephalomalacic changes in the right occipital, temporal, and frontal regions, dilatation in the occipital horn of the right lateral ventricle secondary to volume loss, thickening in the right hemiclavarial bone structures, and increased aeration of the frontal sinuses (Figure-1a, b).

Based on the findings of left hemiparesis, facial asymmetry, intellectual disability, epilepsy, and right cerebral hemiatrophy on MRI, the patient was diagnosed with DDMS.

Figure

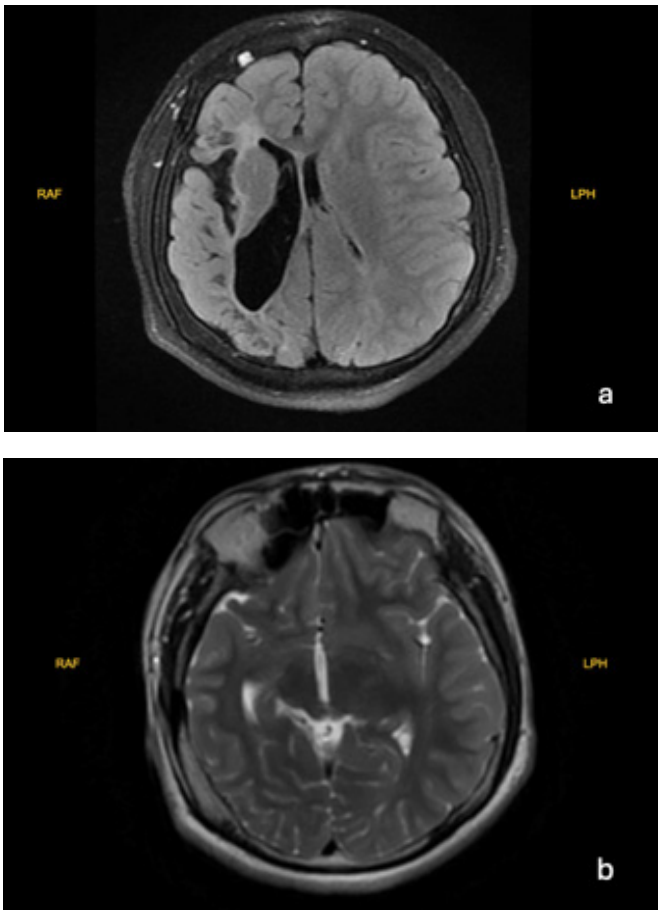


Figure-1 Cranial magnetic resonance imaging shows encephalomalacic changes in the right occipital, temporal, and frontal regions accompanied by thickening in the right hemiclavarial bone structures in a; increased aeration of the frontal sinuses is observed in b.

## Discussion

The characteristic symptoms of DDMS were first described by Dyke, Davidoff, and Masson in 1933 in a series of nine cases.[3] According to the literature, it is a rare neurological disorder diagnosed more frequently in pediatric patients. It is characterized by epileptic seizures, contralateral hemiparesis or hemiplegia, intellectual disability, learning problems, facial asymmetry, language and speech disorders, contralateral choreic movements, sensory disturbances, and unsteady gait. In our case, most of the symptoms of DDMS, such as epileptic seizures controlled with antiepileptic treatment, hemiparesis, intellectual disability, and facial asymmetry, were present.

In a literature review by Rondo et al., 70% of cases had contralateral hemiparesis or hemiplegia, 46% had intellectual disability, and 31% had facial asymmetry. In the study, there was a male predomi-

nance of 55.3%, similar to our case.[4]

DDMS has two etiologies: congenital and acquired. The congenital type is generally caused by brain damage occurring in the intrauterine or neonatal period due to vascular causes where maturation is incomplete. In these cases, symptoms may appear perinatally or in infancy. A compatible genetic model has not been identified for the DDMS clinic.[5]
Acquired DDMS causes include infection, intracerebral hemorrhage or ischemia, trauma, neoplasms, and immunological disorders. The onset of the clinical picture in acquired DDMS may extend to late childhood or adolescence, depending on the timing and nature of the etiological factors.[6] In our case, the patient's clinical findings became evident in early childhood.

In neuroimaging of DDMS, unilateral cerebral volume loss, calvarial thickening, hyperpneumatization of the paranasal sinuses and mastoid cells, enlarged sulci, ipsilateral enlargement of the ventricles, and encephalomalacia have been demonstrated. The encephalomalacic changes and increased calvarial thickness detected in our case's imaging support the role of congenital causes in the etiology of the case.[4-7]

The irritability, anger outbursts, and increased psychomotor activity, which were the reasons for our patient's admission to the outpatient clinic, were found to be similar to the literature.[8]

DDMS treatment should be multidisciplinary; seizures should be controlled with antiepileptic drugs, and motor and cognitive rehabilitation support should be provided. In addition, hemispherectomy can be considered as a treatment method for refractory seizures and hemiplegia. In our case, seizures were controlled with a single antiepileptic drug. Special education and physical therapy exercises were given.[9-10]

Differential diagnoses of DDMS include Sturge-Weber syndrome, hemimegalencephaly, linear sebaceous nevus syndrome, Rasmussen encephalitis, and Silver-Russell syndrome, which also show cerebral hemiatrophy.[11]

Neuroimaging should be performed in cases with complaints such as mental retardation, hemiplegia or hemiparesis, and facial asymmetry accompanied by epilepsy, and the detection of findings such as cerebral hemiatrophy and skull thickening in imaging should suggest DDMS. Given that the diagnosis of DDMS is established through radiological imaging in addition to clinical findings, patients with cerebral hemiatrophy should be more carefully evaluated for the syndrome. In patients where DDMS is suspected, the initiation of early motor and cognitive rehabilitation support is crucial for improving quality of life through early diagnosis and rehabilitation interventions.

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