Dyke Davidoff Masson: A Case Report

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Abstract

Dyke Davidoff-Masson syndrome is a rare neurological condition that results from brain injury during early childhood stages. The precise incidence of this condition is unknown, with a slight male predominance, and adult forms have been documented. Imaging findings reveal hemisphere atrophy along with ipsilateral compensatory skull changes and hyper-pneumatization of mastoid cells. The treatment approach involves anti-epileptic medications and hemispherectomy is reserved for cases with intractable seizures. This case report delineates the clinical manifestation and therapeutic approach employed in an 8-year-old male patient exhibiting pharmaco-resistant left hemi-body convulsive seizures. The magnetic resonance imaging (MRI) findings revealed right cerebral hemiatrophy, mesencephalon atrophy, ipsilateral calvarial hypertrophy and hyperpneumatization of mastoid cells. The objective of this study is to contribute to the existing literature by presenting this rare case report. We propose that in cases involving pediatric pharmaco-resistant epilepsy, it is essential to conduct further investigations to establish a comprehensive management strategy.

Keywords

Dyke Davidoff Masson, pediatric cerebral hemi-atrophy, refractory epilepsy

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Introduction

Dyke Davidoff Masson syndrome (DDMS) is a rare neurological disorder primarily observed in the pediatric population. It is characterized by hemispheric cerebral atrophy, accompanied by ipsilateral compensatory osseous changes, and represents a rare underlying cause of pharmaco-resistant epilepsy.[1] Clinically, patients may present with contralateral hemiparesis, seizures, and learning disabilities, imaging features include cerebral hemi-atrophy with ipsilateral osseous changes as well as hypoplasia of basal ganglia and mesencephalon can be present.[1]

Due to its infrequent occurrence, this disorder is often subject to misdiagnosis or underreporting. Given the absence of specific clinical symptoms associated with DDMS, the importance of imaging findings cannot be overstated.[2]

Notable differential diagnoses for DDMS include Sturge–Weber syndrome, Hemimegaloencephaly, Fishman syndrome, Silver–Russell syndrome, and Rasmussen encephalitis. The primary focus of treatment is symptom management.[2]

Case Report

We present a case involving an 8-year-old male patient with a history of anoxic birth who presented infantile spasms in the first year of his life. Electroencephalogram (EEG) findings revealed high-amplitude slow waves and irregular spikes, indicative of Hypsarrhythmia which was compatible with west syndrome. Initial treatment consisted of monotherapy with sodium valproate which proved ineffective as partial seizures persisted. In terms of laboratory assessments, the results were within the normal range and no imaging examinations were performed.

As the patient progressed in age, he exhibited pharmaco-resistant left hemi-body convulsive seizures along with developmental and intellectual impairment. Consequently, the parents sought further investigation

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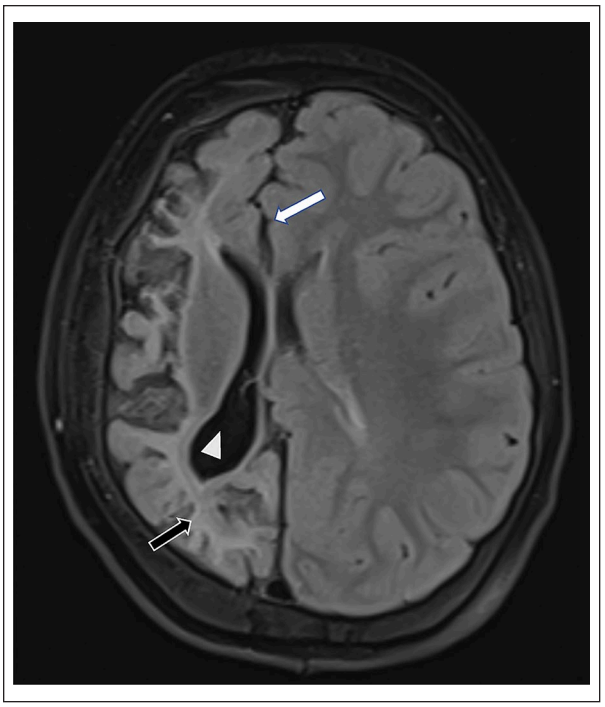

Figure 1. Axial FLAIR sequence showing a right hemi atrophy with enlargement of the right lateral ventricle (White arrow head) and a midline shift (White arrow), Hyperintensity of the subcortical white matter on the effected side (gliotic changes) (Black arrow).

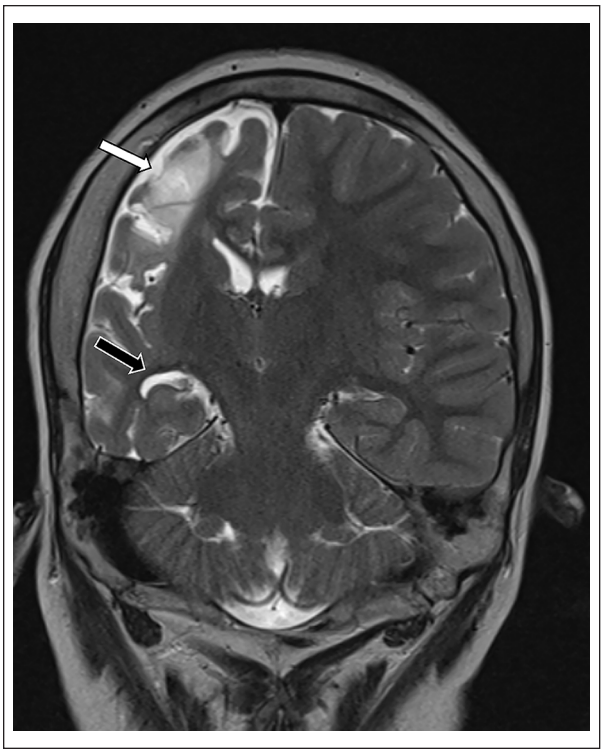

Figure 2. Coronal T2 of MRI sequence demonstrating an enlargement of the right sulcal spaces (White arrow) and ipsilateral choroidal fissure (Black arrow).

into his condition, leading to the performance of a magnetic resonance imaging (MRI) scan at our facility.

Magnetic Resonance Imaging (MRI) investigations revealed cerebral hemi-atrophy affecting the right cerebral hemisphere (Figure 1). This was characterized by the widening of cortical sulci, subarachnoid spaces, and the sylvian fissure (Figure 2). Furthermore, signal abnormalities in the subcortical white matter manifested as hyperintensities on T2 and FLAIR sequences, indicative of gliotic changes (Figure 1). Additionally, there was an ipsilateral enlargement of the lateral ventricle (LV) and a midline shift toward the atrophic side as well as a mesencephalon atrophy (Figure 3). The MRI findings also indicated compensatory calvarial hypertrophy (Figure 4) and hyper-pneumatization of the mastoid cells on the affected side (Figure 5). The coronal T2 sequence illustrated an elevation of the petrous ridge on the atrophic side (Figure 6).

The diagnosis of DDMS was established based on the clinical presentation and specific imaging features and patient is currently undergoing combination therapy, consisting of sodium valproate and carbamazepine. Close observation has revealed a significant decrease in both the frequency and duration of convulsive seizures.

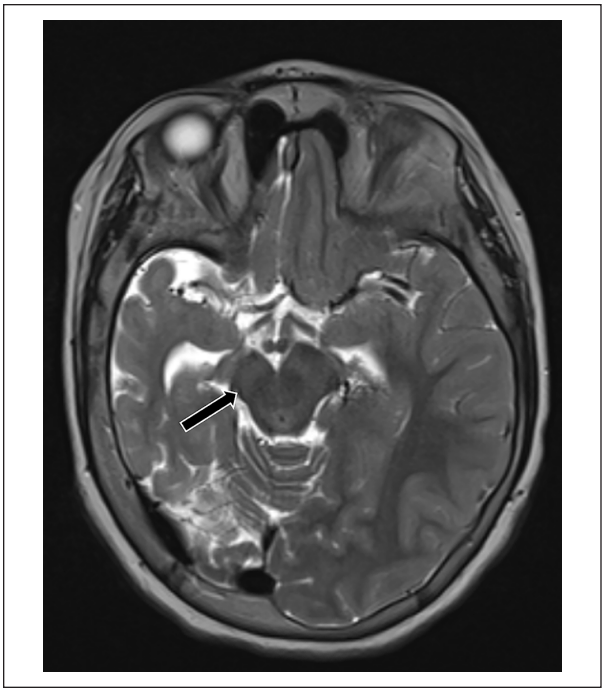

Figure 3. Axial T2 sequence of MRI showing a mesencephalon atrophy (Black arrow).

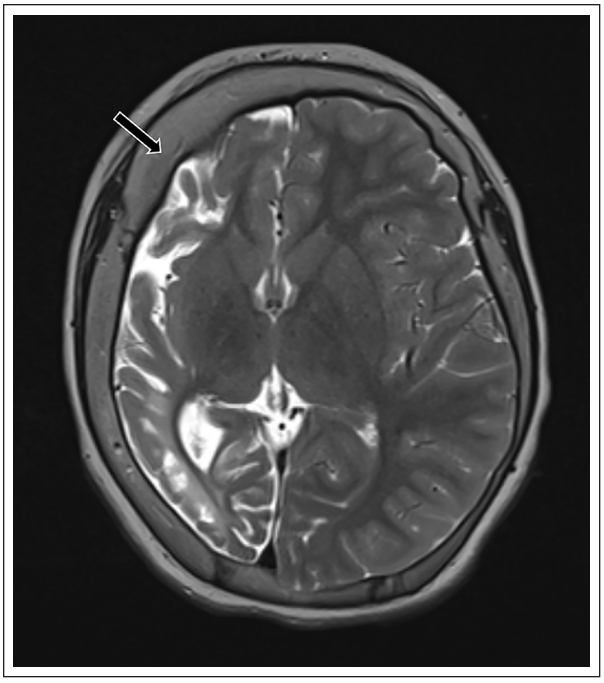

Figure 4. Axial T2 of MRI sequence demonstrating a calvarial thickening interesting the affected side (Black arrow).

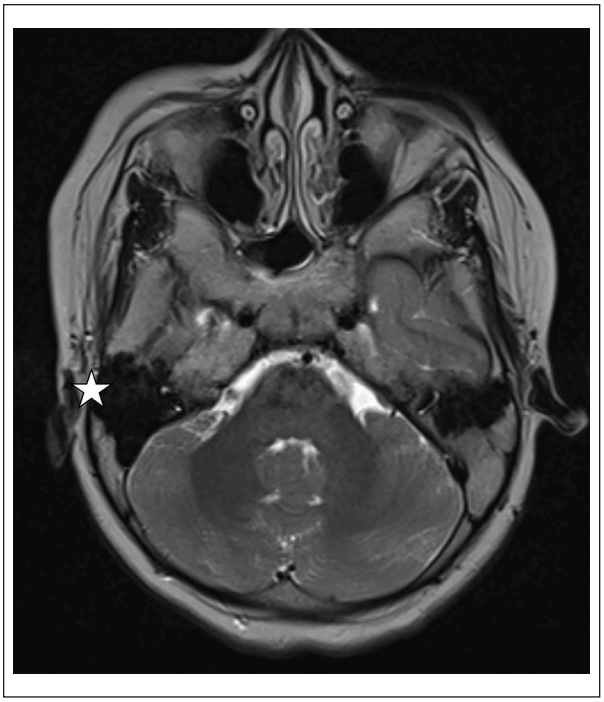

Figure 5. Axial T2 sequence of MRI showing hyper-pneumatization of the right mastoid cells (White star).

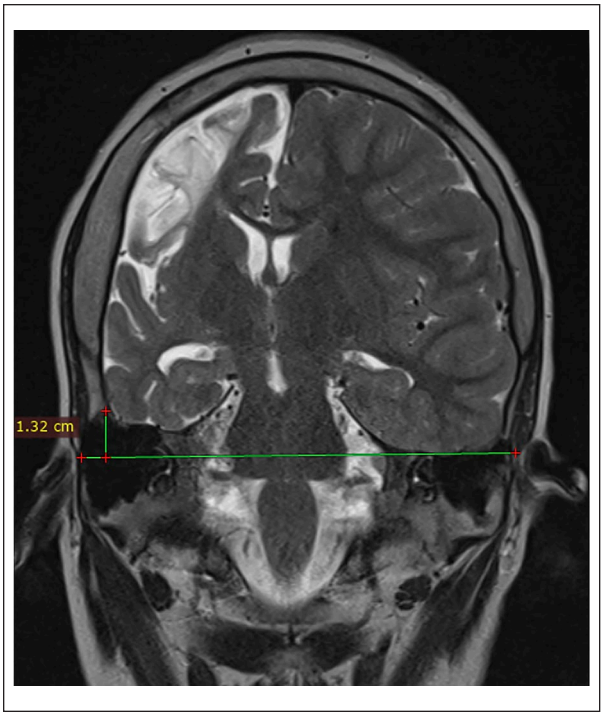

Figure 6. Coronal T2 sequence of MRI showing an elevation of the right petrous ridge (Affected side).

Discussion

Dyke Davidoff-Masson Syndrome (DDMS) is a rare neurological disorder classified among the spectrum of uncommon pathological conditions. It was first documented by Dyke Davidoff and Masson in 1933. This condition is characterized by hemi-atrophy of one cerebral hemisphere, which results from brain injury occurring either during fetal development or in early childhood.[3] DDMS can be categorized into 2 primary subtypes:

-Infantile DDMS, primarily linked to factors such as infection, vascular malformations, gestational vascular occlusion affecting the middle cerebral vascular region, or diminished blood flow through the carotid artery due to a coarctation of the aorta.[4]

-Acquired DDMS, typically stemming from factors like trauma, infection, neonatal hypoxic-ischemic brain injury, hemorrhage, or prolonged febrile seizures.[4]

In cases where cerebral injury occurs before the age of 3, the cranial skull structure undergoes inward growth, leading to the expansion of diploic spaces, paranasal

sinuses, and the elevation of the petrous ridge and orbital roof. These cranial changes serve as significant diagnostic indicators for this condition.[5]

The clinical manifestations of DDMS are nonspecific, with patients often presenting a range of symptoms, including contralateral hemiparesis, seizures, facial asymmetry, and intellectual disability. The severity of these symptoms varies depending on the extent of the brain injury, whether it occurred during the fetal phase or in early childhood. Some patients may also experience sensory issues and psychiatric disorders alongside the classical features.[5]

Imaging findings, using techniques such as CT and MRI, reveal either focal or diffuse hemi-atrophy, leading to volume loss of the ipsilateral cerebral parenchyma. This atrophy is frequently accompanied by compensatory calvarial thickening, especially in the diploic spaces and inner skull table. Moreover, there is ipsilateral widening of sulcal spaces and sylvian fissures, as well as lateral ventricle enlargement, resulting in midline shifting toward the atrophic cerebral hemisphere. Additionally, hypoplasia of various brain structures, such as the thalamus, lentiform nucleus, caudate nucleus, and mesencephalon, can be observed. Hyperpneumatization of frontal, ethmoid, and sphenoid sinuses and mastoid cells, along with elevation of the greater wing of the sphenoid and petrous ridge, may be present.[6] An important functional disconnection, known as crossed cerebral diaschisis, may also be identified, indicating a disconnection between the cerebellar hemisphere and cerebral cortex.[7]

Some studies suggest that unilateral cerebral arterial occlusion is the primary factor leading to DDMS. Time-of-Flight MR angiography can be performed to visualize occluded arteries and assess the extent of occlusion. It's worth noting that this sequence was not conducted in our case.[8]

Differential diagnoses for DDMS include Rasmussen encephalitis, which is a chronic inflammatory brain disease with no calvarial changes, and Sturge-Weber syndrome, a phakomatosis featuring facial port-wine stains and enhancing pial angiomas with a tram-track sign of cortical and subcortical calcifications. Other potential differential diagnoses include Silver-Russell syndrome, Fishman syndrome, and Hemimegaloencephaly.[4,8]

The treatment approach for DDMS primarily focuses on addressing the symptoms, including convulsions, hemiplegia, hemiparesis, and learning disabilities, through a comprehensive regimen of physiotherapy, occupational therapy, and speech therapy. A more favorable prognosis is associated with the onset of hemiparesis after the age of 2 years and the absence of prolonged or recurrent seizures. Hemispherectomy is the preferred therapeutic option for patients experiencing hemiplegia and intractable disabling seizures.[8]

Conclusion

Dyke-Davidoff-Masson Syndrome represents a rare cause of refractory epilepsy. This case report depicts the evolution of an 8-year-old child with pharmaco-resistant left hemi-body convulsive seizures with intellectual disability. Imaging has a definitive role in excluding alternative diagnoses, predicting the prognosis and adapting the treatment management.

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Author Contributions

Dr Boumeriem Khaoula: Contributed to conception or design, contributed to acquisition, analysis, and interpretation, drafted the manuscript, and agrees to be accountable for all aspects of work ensuring integrity and accuracy.
Dr Bourekba Iliass: Contributed to conception or design, contributed to acquisition, analysis, and interpretation, drafted the manuscript, critically revised the manuscript and agrees to be accountable for all aspects of work ensuring integrity and accuracy.
Pr Allali Nazik: Critically revised the manuscript, gave final approval and agrees to be accountable for all aspects of work ensuring integrity and accuracy.
Pr Chat Latifa: Authorized the author to use the case, critically revised the manuscript, gave final approval and agrees to be accountable for all aspects of work ensuring integrity and accuracy.
Pr El Haddad Siham: Critically revised the manuscript, gave final approval and agrees to be accountable for all aspects of work ensuring integrity and accuracy.

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Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

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