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Case Presentation

A Case of Cerebellar Hemiatrophy and Discussion of Cerebral Hemiatrophy: Dyke-Davidoff-Masson Syndrome

Amy Avakian[1], Ijeoma Chijioke[1], Zachary I. Merhavy[1*], Garrett Barfoot[1], and David De Bruin[2]

[1]*Ross University School of Medicine, Barbados*

[2]*Mount Sinai Hospitals - Chicago, USA*

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[*]**Corresponding author:** Zachary I. Merhavy, Ross University School of Medicine, Bridgetown, Barbados

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Abstract

A 63-year-old female presents with a one-day history of abnormal twitching movements of the left upper extremity. Her past medical history of hypertension, type 2 diabetes mellitus, chronic systolic congestive heart failure, situs inversus with dextrocardia, seizures, gastroesophageal reflux disease, arthritis, and cervical degenerative disc disease presents with a one-day history of abnormal twitching movements of the left upper extremity. Physical examination was significant for left-sided hemiparesis and chronic mental impairment. Imaging was limited due to claustrophobia, but emergent CT showed no intracranial hemorrhage, mass effect, or midline shift. Multiecho multiplanar MRI of the brain performed under sedation before and after administration of gadolinium-based contrast showed long standing right cerebellar atrophy and decreased size of the right posterior fossa with associated calvarial thickening. A diagnosis of cerebellar hemiatrophy was established.

Keywords: Dyke-Davidoff-Masson Syndrome; Cerebellar Hemiatrophy; Cerebral Hemiatrophy; DDMS; Radiology; Radiologic Findings; MRI; CT Dyke-Davidoff-Masson Syndrome

Background

Dyke-Davidoff-Masson Syndrome (DDMS) typically presents with a combination of neurological deficits that can include hemiparesis, seizures, facial asymmetry, and intellectual disability. The syndrome is characterized by its association with cerebral hemiatrophy, which leads to the aforementioned clinical features [1]. Patients often present in childhood, although the exact age of onset can vary depending on the severity of brain insult and subsequent development of symptoms. Seizures are frequently the presenting symptom and can range from focal to generalized seizures [2]. Hemiparesis, or weakness on one side of the body, is another common presentation and may be noticed early in infancy or childhood as delayed milestones in motor development.

DDMS is a rare condition with no specific prevalence data due to its varied presentation and potential underreporting [3]. It affects both males and females equally and has been documented worldwide. The syndrome can occur as a consequence of various prenatal, perinatal, or early postnatal

insults, which can affect the developing brain leading to the characteristic hemiatrophy [4].

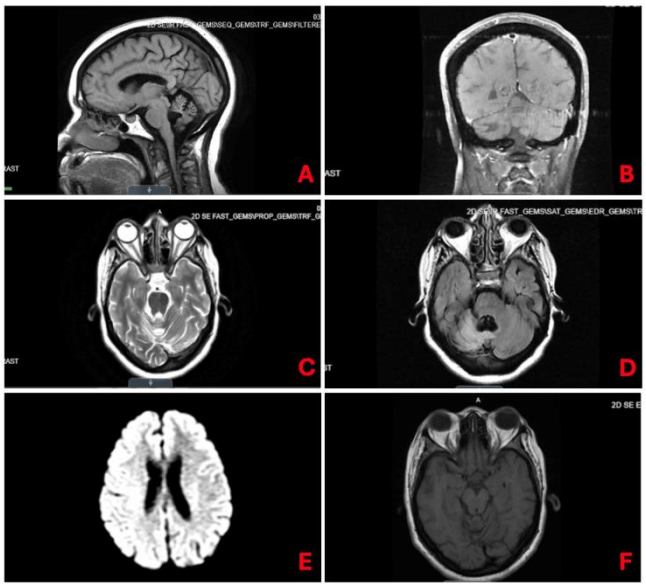
The etiology of DDMS is multifactorial, often linked to events that cause damage to the developing brain. These events can be classified as prenatal (intrauterine infections, congenital anomalies, and vascular insults), perinatal (birth trauma, hypoxic-ischemic encephalopathy), or postnatal (head trauma, infections such as meningitis or encephalitis) [1]. The underlying mechanism involves loss of brain parenchyma which leads to compensatory changes in the skull, such as hyper-pneumatization of the frontal sinuses and elevation of the petrous ridge and greater wing of the sphenoid bone [1].

Case Presentation

A 63-year-old female presented with a one-day history of abnormal twitching movements of the left upper extremity. Her past medical history of hypertension, type 2 diabetes mellitus, chronic systolic congestive heart failure, situs inversus with dextrocardia, seizures, gastroesophageal reflux disease, arthritis, and cervical degenerative disc disease presents with a one-day history of abnormal twitching movements of the left upper extremity. Physical examination was significant for left-sided hemiparesis and chronic mental impairment. Imaging was limited due to claustrophobia, but emergent CT showed no intracranial hemorrhage, mass effect, or midline shift. Multiecho multiplanar MRI of the brain performed under sedation before and after administration of gadolinium-based contrast showed long standing right cerebellar atrophy and decreased size of the right posterior fossa with associated calvarial thickening. A diagnosis of cerebellar hemiatrophy was established.

Imaging Features

Imaging, particularly MRI and CT scans, play a crucial role in the diagnosis of DDMS. Characteristic imaging features include unilateral cerebral atrophy, ipsilateral compensatory osseous hypertrophy, and enlargement of the frontal sinuses [2]. MRI is particularly useful for assessing the extent of cerebral parenchymal loss, gliosis, and changes in the ventricular system. Typical findings include a shrunken and atrophic hemisphere, dilated lateral ventricle on the affected side, and compensatory hypertrophy of the contralateral hemisphere [4]. CT scans can reveal additional bony changes such as calvarial thickening and sinus expansion [5].



MRI Images courtesy of the Department of Radiology, Mount Sinai Hospital, Chicago

A. Sagittal plane - Right cerebellar hemiatrophy; Right posterior fossa with associated calvarial thickening
B. Coronal plane - Right cerebellar hemiatrophy
C. Axial plane - Right cerebellar hemiatrophy; Right posterior fossa with associated calvarial thickening
D. Axial plane - Right cerebellar hemiatrophy; Right posterior fossa with associated calvarial thickening
E. Right cerebellar hemiatrophy
F. Axial plane - Right cerebellar hemiatrophy; Right posterior fossa with associated calvarial thickening

Discussion

The etiology of DDMS varies but is usually a combination of cerebral insults in infancy or childhood. In this patient, it is unclear given the lack of documented brain insults typically associated with the syndrome. However, the chronic nature of this patient's atrophy suggests a longstanding process, potentially linked to undiagnosed or undocumented early life events.

The prognosis for patients with DDMS varies widely depending on the severity of brain atrophy and the timing and effectiveness of intervention [5]. Early diagnosis and comprehensive management can significantly improve the

quality of life and functional outcomes. Children with milder forms of the disease who receive appropriate medical and therapeutic interventions can achieve relatively good control of seizures and motor function [4]. However, severe cases associated with extensive brain damage may result in persistent neurological deficits and a greater degree of disability [5].

Management of DDMS is symptomatic and supportive, focusing on controlling seizures, managing hemiparesis, and addressing developmental delays [3]. Antiepileptic drugs are commonly used to manage seizures, with the choice of drug tailored to the type and severity of seizures [3]. Physical and occupational therapy are essential for improving motor function and aiding in the development of skills necessary for daily activities [4]. In severe cases of refractory epilepsy, surgical interventions such as hemispherectomy or corpus callosotomy may be considered [5]. Early intervention and a multidisciplinary approach are crucial for optimizing patient outcomes [3].The chronic mental impairment noted in the patient prompted a multidisciplinary approach to optimize her quality of life.

Conclusion

DDMS is an uncommon condition that typically presents in childhood with a spectrum of neurological manifestations including hemiparesis, seizures, and intellectual disability. In this report, the atypical initial presentation in a patient with a complex medical history prompted further investigation into her symptoms and revealed this diagnosis. Despite the patient's extensive medical history, which may have contributed to or exacerbated her neurological symptoms, the diagnosis of cerebellar atrophy was established through imaging studies. These imaging findings, in conjunction with the patient’s clinical presentation, led to the definitive diagnosis. This case illustrates the variability in clinical presentation, emphasizing the need for heightened awareness among clinicians of this condition even in older adults. It also highlights the role of comprehensive imaging studies in diagnosing DDMS, particularly when initial imaging is inconclusive. Early recognition and a thorough approach are essential for managing the complex needs of patients with DDMS and mitigating the impact of this condition. Future research should focus on improving diagnostic techniques and therapeutic approaches to better support patients with DDMS and similar neurodevelopment disorders.

Disclosures & Consents

The authors declare no conflicts of interest or funding to disclose. This case was prepared with all necessary informed consent from the patient for educational purposes.

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