Journal of Neonatal Surgery
ISSN(Online): 2226-0439
Vol. 14, Issue 32s (2025)
https://www.jneonatalsurg.com

OPEN ACCESS

Dyke-Davidoff-Masson Syndrome Without Motor Deficits: A Case Report Highlighting Diagnostic Challenges

Dr. Joel Devasia[1], Dr. Esha Nobbay[2*], Dr. Karthik J[3], Dr. Madhusudan J[4]

[1]Junior Resident, Department of Psychiatry, Adichunchanagiri Institute of Medical Sciences

[2*]Junior Resident, Department of Psychiatry, Adichunchanagiri Institute of Medical Sciences

[3]Senior Resident, Department of Psychiatry, Adichunchanagiri Institute of Medical Sciences

[4]Assistant Professor, Department of General Medicine, Adichunchanagiri Institute of Medical Sciences

***Corresponding Author:**

Dr. Esha Nobbay,

Junior Resident, Department of Psychiatry, Adichunchanagiri Institute of Medical Sciences

Cite this paper as: Dr. Joel Devasia, Dr. Esha Nobbay, Dr. Karthik J, Dr. Madhusudan J, (2025) Dyke-Davidoff-Masson Syndrome Without Motor Deficits: A Case Report Highlighting Diagnostic Challenges. *Journal of Neonatal Surgery*, 14 (32s), 330-334.

ABSTRACT

Background: Dyke-Davidoff-Masson syndrome (DDMS) is a rare condition characterized by cerebral hemiatrophy, calvarial thickening, and neurological deficits, typically secondary to early cerebral injury. Classical presentations include seizures, hemiparesis, and cognitive impairment, though atypical cases without motor deficits have been reported.

Case Report: A 19-year-old male presents with a long history of recurrent generalized seizures and cognitive impairment. Neurological examination revealed no focal motor deficits or hemiparesis. Brain imaging demonstrated marked left cerebral hemiatrophy, ipsilateral ventricular dilatation, prominent sulci, asymmetric calvarial thickening, and hyperpneumatization of the right frontal sinus, confirming DDMS despite the absence of motor weakness.

Discussion: This case highlights an atypical DDMS presentation without hemiparesis, expanding the phenotypic spectrum of the syndrome. The absence of motor deficits may be attributed to selective non-motor cortical involvement or compensatory neuroplasticity during early brain development. The imaging findings of left cerebral atrophy, ventricular enlargement, and ipsilateral osseous changes are pathognomonic for DDMS and likely reflect chronic hemispheric volume loss due to perinatal or childhood vascular insult. Notably, the seizure semiology and cognitive impairment align with prior reports, reinforcing the role of functional reorganization in masking motor deficits. This case also underscores the diagnostic challenge in patients without classic hemiparesis, necessitating a high index of suspicion when neuroimaging reveals unilateral cerebral atrophy.

Conclusion: This atypical presentation of Dyke-Davidoff-Masson syndrome without hemiparesis expands the recognized phenotype. Persistent refractory epilepsy and cognitive deterioration should prompt high-resolution neuroimaging for definitive diagnosis, individualized antiseizure-psychotropic regimens, and early multidisciplinary neurorehabilitation. Mitigating pharmacological interactions, socioeconomic constraints, and caregiver burden through cost-effective, integrated strategies may significantly enhance long-term functional outcomes.

Key Words: *Dyke-Davidoff-Masson syndrome, cerebral hemiatrophy, refractory epilepsy, calvarial thickening*

1. INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) was first described in 1933 by Dyke, Davidoff, and Masson through their radiographic studies of seven patients exhibiting characteristic skull changes and cerebral hemiatrophy. (1) This rare neurological condition represents a distinct clinicoradiological entity resulting from insults to the developing brain, typically occurring before two years of age. (2) The syndrome has since been recognized as an important cause of refractory epilepsy and neurological disability in pediatric and young adult populations. The pathogenesis of DDMS involves a complex interplay between early cerebral injury and subsequent compensatory changes. The primary insult, which may include perinatal hypoxia-ischemia, intrauterine infections, trauma, or cerebrovascular accidents, leads to unilateral cerebral

hypoplasia or atrophy. (3) This triggers several secondary adaptations: the calvaria thickens ipsilaterally due to decreased intracranial pressure, the frontal sinus hyperpneumatizes as a compensatory phenomenon, and the petrous ridge elevates. These changes reflect the brain's remarkable plasticity during early development and its capacity for morphological adaptation to injury.

The clinical presentation of DDMS typically includes a triad of symptoms: seizure disorders (present in 95% of cases), contralateral hemiparesis (reported in 85-90% of patients), and varying degrees of cognitive impairment. (4) Seizures often begin in childhood and are frequently refractory to multiple antiepileptic medications. The motor deficits range from mild weakness to complete hemiplegia, often accompanied by spasticity and hyperreflexia. Cognitive manifestations may include intellectual disability, learning difficulties, and in some cases, psychiatric comorbidities such as behavioral disorders or mood disturbances. Diagnostic confirmation of DDMS relies heavily on neuroimaging findings. Computed tomography (CT) and magnetic resonance imaging (MRI) typically reveal: unilateral cerebral hemiatrophy, ipsilateral ventricular enlargement (ex vacuo dilatation), thickening of the cranial vault, and elevation of the petrous bone MRI provides additional valuable information, often demonstrating gliosis, porencephalic cysts, or evidence of prior ischemic injury in the affected hemisphere. Advanced imaging techniques such as diffusion tensor imaging have begun to reveal more subtle white matter changes in recent studies.

While the classic triad of seizures, hemiparesis, and cognitive impairment characterizes most DDMS cases, approximately 5-10% of reported cases present without motor deficits, creating diagnostic dilemmas. (5) These atypical presentations may be attributed to several factors: the timing of the initial insult (with very early injuries potentially allowing greater functional reorganization), the specific regions of cortex affected (sparing motor pathways), or individual differences in neuroplasticity. The absence of hemiparesis often delays diagnosis, particularly in resource-limited settings where neuroimaging may not be readily available. (6) The present case of a 19-year-old male with long-standing refractory epilepsy and cognitive impairment but no motor deficits contribute valuable insights to the DDMS literature. This presentation challenges the conventional diagnostic criteria that emphasize hemiparesis as an essential feature. Furthermore, the case highlights the importance of considering DDMS in the differential diagnosis of young patients with unexplained epilepsy and cognitive decline, even in the absence of motor symptoms. Detailed neuroimaging evaluation remains crucial for accurate diagnosis and appropriate management of such atypical cases

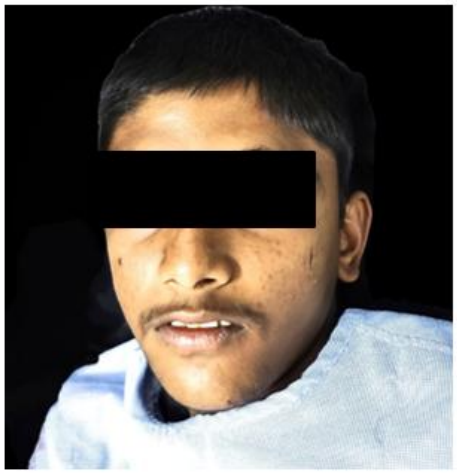
2. CASE REPORT

A 19-year-old male presented to our emergency department following a witnessed generalized seizure characterized by unprovoked clonic movements of the left extremities progressing to bilateral tonic-clonic activity with upward eye deviation. The 8-10 minute episode was followed by postictal lethargy persisting for one hour. This represented one of several breakthrough seizures occurring over six months despite antiepileptic therapy, without identifiable metabolic or infectious triggers.

The patient was born at 39 weeks via vaginal delivery but suffered severe birth asphyxia requiring 28 days of NICU care. Born to consanguineous parents (first cousins), he achieved normal developmental milestones until age 4 when he experienced significant regression following a febrile illness, coinciding with new-onset generalized epilepsy.His epilepsy course featured 2-3 monthly generalized tonic-clonic seizures with partial response to valproate monotherapy. Breakthrough seizures consistently correlated with medication non-adherence. Cognitive assessment revealed severe global developmental delay with current functioning at a 5-6 year level. Behavioral comorbidities included disruptive mood dysregulation disorder diagnosed at age 12, managed with risperidone/olanzapine combination therapy showing partial efficacy against aggression and emotional lability.During evaluation, the patient appeared generally well-nourished but exhibited persistent postictal lethargy limiting full cooperation. Neurological assessment revealed:

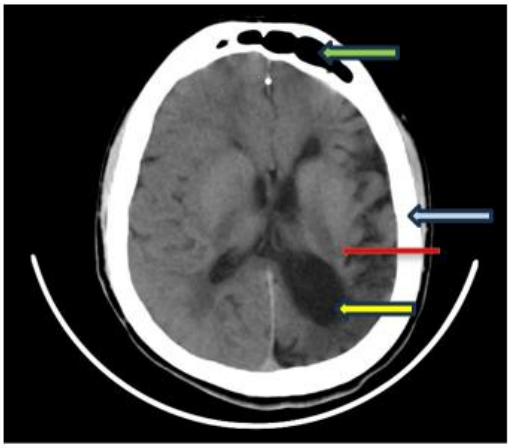
- Significantly impaired higher mental functions (memory, orientation, and judgment)
- Markedly restricted speech output
- Intact cranial nerves without gross abnormalities
- Preserved motor strength and tone bilaterally
- Symmetrical deep tendon reflexes with bilateral flexor plantar responses
- Responsiveness to tactile stimuli without focal deficits
- Formal coordination testing and comprehensive sensory examination proved impossible due to reduced cooperation. Routine laboratory studies, including glucose and electrolytes, were within normal limits.

Figure 1.



Absence of facial asymmetry despite underlying hemiatrophy

Figure 2.

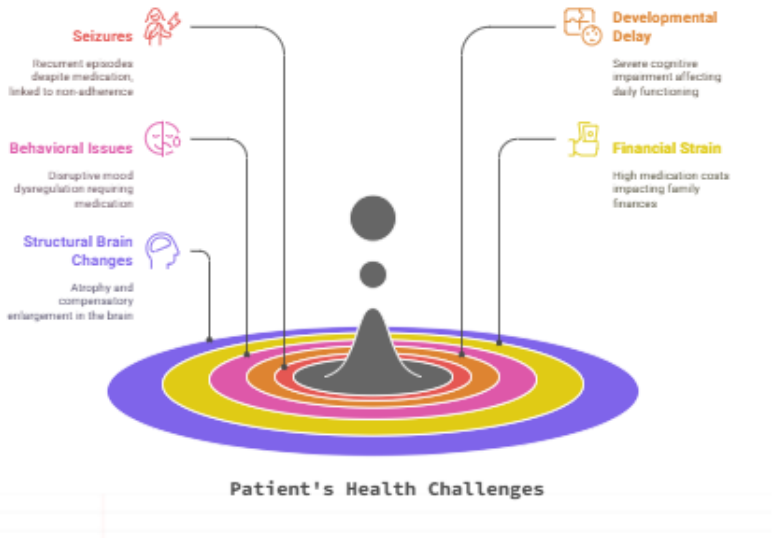


Non-contrast CT brain showing left cerebral hemiatrophy (red arrow) with compensatory:
(a) dilatation of left lateral ventricle (yellow arrow);
(b) mild calvarial thickening (blue arrow);
(c) hyperpneumatization of left frontal sinus (green arrow).

Non-contrast computed tomography of the brain revealed significant atrophy of the left cerebral hemisphere, with the most pronounced volume loss affecting the frontoparietal cortical regions. Associated compensatory ex vacuo enlargement of the ipsilateral lateral ventricle was noted. The characteristic osseous changes linked to DDMS were clearly evident, including thickening of the left cranial vault, marked hyperpneumatization of the left frontal sinus, and increased prominence of the diploic spaces (figure 1.). Additional structural changes included atrophic changes affecting the right cerebral peduncle and posterior corpus callosum. Chronic inflammatory changes were noted within the left mastoid air cells, consistent with the overall pattern of unilateral involvement. The constellation of findings provided definitive radiological confirmation of DDMS, supporting the clinical impression despite the notable absence of contralateral motor weakness and facial asymmetry (figure 1.) in this patient.

Residing in a rural area with limited specialist access, the family spent 20% of their income on medications, a burden exacerbated by the father's chronic illness and subsequent death. The patient required complete assistance for activities of daily living and communicated solely through basic gestures.

Figure 3.



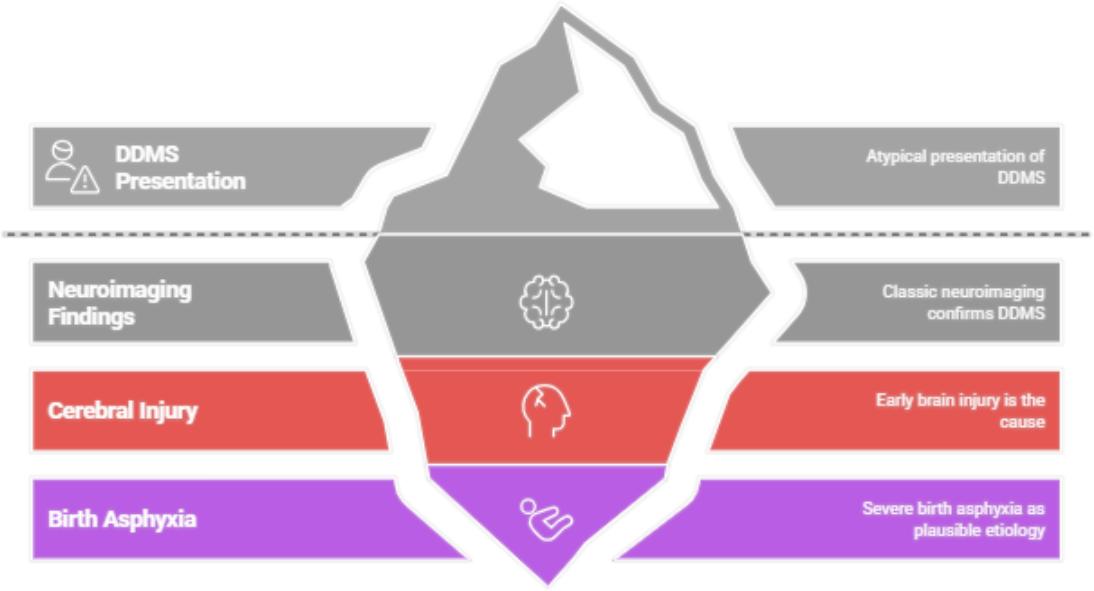
The evaluation was complicated by several factors:

- Limited cooperation restricting comprehensive neurological examination
- Delayed advanced neuroimaging due to financial constraints
- Absence of prior genetic workup despite consanguinity
- Narrow therapeutic window for antiepileptics with history of toxicity
- Significant drug interactions between antiepileptics and psychotropics

3. DISCUSSION

This case presents an atypical presentation of Dyke-Davidoff-Masson syndrome (DDMS) characterized by classic neuroimaging findings but absence of the hallmark motor deficits, contributing to the evolving understanding of this rare condition. The patient's clinical presentation aligns with several key features described in the literature while challenging conventional diagnostic expectations. The unilateral cerebral hemiatrophy with ipsilateral calvarial thickening and frontal sinus hyperpneumatization observed in our patient (figure 2.) are pathognomonic radiographic features of DDMS, as originally described by Dyke, Davidoff and Masson in 1933. (7) These findings typically result from antenatal, perinatal or early postnatal cerebral injury, with our patient's history of severe birth asphyxia providing a plausible etiology. The timing of injury correlates with the developmental window of vulnerability described by Shetty et al. (2017), where insults before age 2 years are most likely to produce the characteristic osseous changes. (8)

Figure 4.



Unveiling the atypical presentation of Dyke-Davidoff-Masson syndrome.

Notably, our case lacked contralateral hemiparesis, which is reported in 85-90% of DDMS cases. (4) This absence may be explained by several mechanisms proposed in recent literature. Chunchu et al. (2020) suggest that very early insults (before 28 weeks gestation) may allow greater interhemispheric reorganization of motor pathways. (9) Alternatively, selective sparing of the primary motor cortex with predominant injury to association areas could account for preserved motor function, as demonstrated in diffusion tensor imaging studies. (10) The patient's refractory epilepsy and cognitive impairment are consistent with prior DDMS reports. Thakkar et al. found that 95% of DDMS patients develop epilepsy, with 60% showing poor response to first-line antiepileptic. (11) The cognitive profile in our case - with severe impairment but preserved basic motor functions - mirrors the "cognitive-predominant" variant recently described by Dakka et al in their series of atypical DDMS cases. (12)

The behavioral comorbidities observed in this patient extend the known phenotypic spectrum of DDMS. While psychiatric

manifestations have been previously reported, the combination of aggressive behaviors and mood dysregulation in our case suggests possible involvement of frontolimbic circuits, consistent with the left frontotemporal atrophy seen on imaging. This correlation between neuroanatomical changes and behavioral phenotype has been increasingly recognized in neurodevelopmental disorders. (13) The management challenges in this case highlight important clinical considerations. The narrow therapeutic window for antiepileptics and significant drug interactions observed in our patient have been similarly reported by Mallik et al (2014) in their review of DDMS complications. (14) Furthermore, the socioeconomic barriers to care experienced by this patient reflect the global health disparities in managing chronic neurological conditions, particularly in resource-limited settings.

4. CONCLUSION

Dyke-Davidoff-Masson syndrome (DDMS) represents a rare but clinically significant cause of refractory epilepsy and neurodevelopmental impairment. (7) This case illustrates several important considerations for clinicians. Dyke–Davidoff–Masson syndrome can present without the full triad of seizures, hemiparesis, and cognitive impairment; our motor-sparing case broadens its phenotypic spectrum. Neuroimaging is therefore pivotal, and clinicians should suspect DDMS in children with refractory seizures, cognitive delay, unilateral cerebral atrophy, or craniofacial/osseous asymmetry. Early recognition allows optimized antiepileptic therapy, neurorehabilitation, and coordinated multidisciplinary care. Continued study of such atypical presentations will clarify pathophysiologic variability and refine management strategies

REFERENCES

[1] Verhandlungen der deutschen Gesellschaft für Kreislaufforschung VIII Tagung. Arch Intern Med. 1936 Jul 1;58(1):186.

[2] Pollak N, Soni RS. Endoscopic excision of a tympanic paraganglioma: Training the next generation of ear surgeons. World J Otorhinolaryngol - Head Neck Surg. 2017 Sep 22;3(3):160–2.

[3] Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. J Neurosci Rural Pract. --;3(3):411–3.

[4] Urs GM, Doddabele HR. Dyke-Davidoff-Masson Syndrome: A Case Report. Int J Med Stud. 2022 Apr 13;10(1):82–5.

[5] Pradeep U, Chiwhane A, Acharya S, Kumar P, Kasat P. Dyke-Davidoff-Masson syndrome: A rare case of hemiatrophy with chronic seizure disorder. Radiol Case Rep. 2025 Mar 1;20(3):1470–4.

[6] Hamid M, Cherradi S, Satte A, Bourazza A. Dyke-Davidoff-Masson syndrome: A case report with a literature review. Radiol Case Rep. 2022 Aug;17(8):2616–8.

[7] Dyke, C.G., Davidoff, L.M. and Masson, C.B. (1933) Cerebral Hemiatrophy with Homolateral Hypertrophy of the Skull and Sinuses. Surgery, Gynecology & Obstetrics, 57, 588-600. - References - Scientific Research Publishing [Internet]. [cited 2025 May 30]. Available from: https://www.scirp.org/reference/referencespapers?referenceid=1488123

[8] Shetty DS, Lakhkar BN, John JR. Dyke-Davidoff-Masson syndrome. Neurol India. 2003 Mar;51(1):136.

[9] Dyke-Davidoff-Masson Syndrome: A Case Report | Cureus [Internet]. [cited 2025 May 30]. Available from: https://www.cureus.com/articles/134274-dyke-davidoff-masson-syndrome-a-case-report#!/

[10] Kessi EMCJ, Amrani SE, Dghoughi B, Touarsa F, Fikri M, Jiddane M, et al. Dyke–Davidoff–Masson syndrome: A typical case of late diagnosis. Int J Case Rep Images IJCRI. 2024 Jun 22;15(1):103–6.

[11] Thakkar PA, Dave RH. Dyke-Davidoff-Masson syndrome: A rare cause of cerebral hemiatrophy in children. J Pediatr Neurosci. 2016;11(3):252–4.

[12] Dakka A, Vyas A, Laurent BP. A Rare Cause of Seizures and Hemiparesis - Dyke Davidoff Masson Syndrome: A Case Report. J Clin Trials. 6(5):1–3.

[13] Rwililiza E M, C. M, F S, A S, R.D H., S K, et al. Familial occurrence of Dyke-Davidoff-Masson syndrome in two African siblings with unexplained parotid enlargement. Int J Case Rep Images. 2016;7(6):354.

[14] Malik P, Garg R, Gulia A kumar D, Kario J. Dyke-Davidoff-Masson Syndrome- a rare cause of refractory epilepsy. Iran J Psychiatry. 2014 Mar;9(1):42–4