Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Dyke-Davidoff-Masson Syndrome in A Teenager: Radiological Clues to A Rare Neurological Entity

A. El Hadri[1]*, M. Darfaoui[2], Y. Bouktib[2], A. El Hajjami[2], B. Boutakioute[2], M. Ouali Idrissi[2], N. Cherif Idrissi Guennouni[2]

[1]Department of Radiology, Arrazi Hospital, Mohammed VI University Hospital Center of Marrakech
[2]Faculty of Medicine and Pharmacy, Marrakech Cadi Ayad University, Morocco

DOI: https://doi.org/10.36347/sjmcr.2025.v13i08.042 | **Received:** 17.06.2025 | **Accepted:** 20.08.2025 | **Published:** 29.08.2025

***Corresponding author:** A. El Hadri
Department of Radiology, Arrazi Hospital, Mohammed VI University Hospital Center of Marrakech

Abstract | **Case Report**

Dyke-Davidoff-Masson syndrome is a rare clinico-radiological entity, characterized clinically by seizure activity, facial asymmetry, and hemiparesis. The clinical presentation may also include motor, cognitive, and behavioral impairments. Neuroimaging, particularly computed tomography (CT) and magnetic resonance imaging (MRI) plays a fundamental role in the diagnosis and management of this condition. MRI enabled a straightforward diagnosis of the syndrome, revealing cerebral hemiatrophy, thickening of the cranial vault, and hyperpneumatization of the frontal sinus. We report a case of a teenager presenting with seizures highlighting the pivotal role of imaging in identifying this syndrome.

Keywords: Dyke-Davidoff-Masson, seizure, cerebral MRI, cerebral hemiatrophy, hyperpneumatization of the frontal sinus.

INTRODUCTION

Dyke-Davidoff-Masson syndrome is a rare neurological condition first described in 1933 by C. G. Dyke, L. M. Davidoff, and C. B. Masson. It results from cortical brain lesions and is most frequently reported in the pediatric population. Among the approximately 100 cases described in the literature, only 21 involved adults. [1] Historically, the syndrome has been associated with ischemic or hemorrhagic strokes occurring in early childhood, a period during which the brain is undergoing rapid development. [2]

The syndrome manifests with unilateral neurological signs, combining seizures, facial asymmetry, and hemiparesis, often accompanied by motor, cognitive, and behavioral issues. Neuroimaging, especially MRI, is essential for diagnosis, typically revealing cerebral hemiatrophy, cranial vault thickening, and frontal sinus hyperpneumatization and often accompanied by cerebral asymmetry, which is well documented through imaging studies. [3-4].

CASE REPORT

The patient is a 19-year-old male, born to second-degree consanguineous parents, with no significant past medical history. Symptom onset dates back to the age of 10, marked by the development of poorly managed focal epilepsy.

Clinical examination revealed progressively worsening spastic hemiparesis, accompanied by muscle wasting of the left hemibody and a dysmorphic syndrome, including asymmetry of the palpebral fissures, a short neck, and a scoliotic posture of the thoracolumbar spine (Figure 1). Neuropsychological assessment showed mild intellectual disability.

Brain MRI revealed right cerebral hemiatrophy, with widening of the cortical sulci and dilatation of the ipsilateral lateral ventricle, accompanied by midline shift, subcortical porencephalic cavities, and thickening of the right cranial vault with hyperpneumatization of the frontal sinus (Figure 2). The MR angiography sequence showed no abnormalities.

Demonstrating right cerebral hemiatrophy, as evidenced by widening of the cortical sulci and dilatation of the ipsilateral lateral ventricle with midline shift. Associated findings include subcortical porencephalic cysts, thickening of the right cranial vault, and hyperpneumatization of the frontal sinus.

The patient was initially started on phenobarbital, but continued to experience seizures. The

treatment was then adjusted with the addition of carbamazepine, leading to a notable clinical improvement.

Figure 1: Muscle atrophy of the left hemibody (A, B), associated with mild scoliotic posture (C)

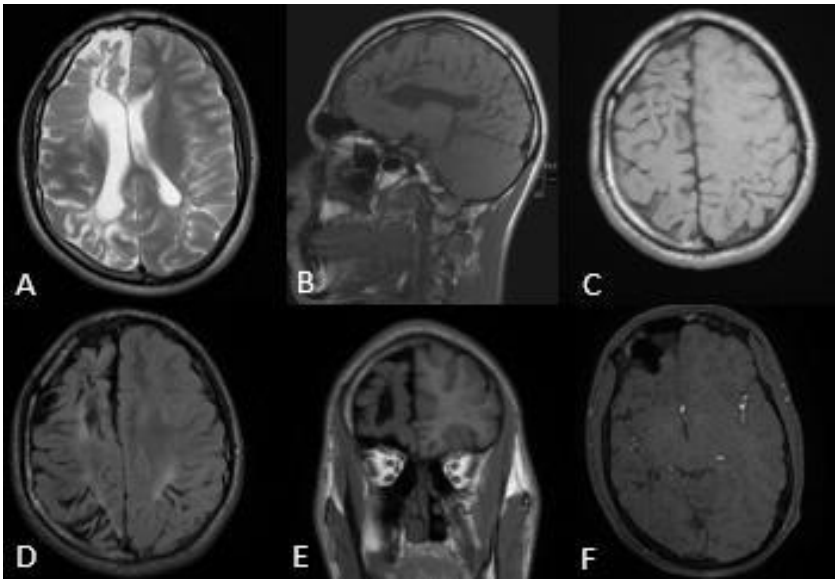

Figure 2: Brain MRI – Axial T2 sequences (A), sagittal T1 (B), axial T1 (C), axial T2 FLAIR (D), coronal T1 FLAIR (E), and TOF FS (F)

DISCUSSION

Dyke-Davidoff-Masson syndrome is a rare clinico-radiological entity first described in 1933 by C. G. Dyke, L. M. Davidoff, and C. B. Masson through nine cases that included hemiparesis, facial asymmetry, critical manifestations such as epilepsy, and radiological findings of cerebral hemiatrophy, cranial thickening, and hyperpneumatization of the frontal sinuses. [5] Approximately one hundred cases have been published in the literature to date, the majority of which are pediatric.

The etiopathogenesis remains unclear, but the condition can have congenital or acquired causes. In congenital forms, clinical manifestations are either neonatal or discovered early and result from occlusion of the middle cerebral artery, unilateral cerebral vascular anomalies, aortic coarctation, or infectious origins. Acquired forms are primarily due to neonatal distress, prolonged febrile seizures, trauma, infections, and cerebrovascular accidents (both hemorrhagic and ischemic) [5, 4].

The severity of clinical manifestations depends on the extent of cerebral involvement. These manifestations include critical impairments (partial or generalized seizures), facial asymmetry, contralateral motor deficits, and intellectual disability [3]. School difficulties and cognitive impairments, particularly in visuospatial functions, have also been reported [3]. A polymorphic malformation syndrome (including pes cavus, cleft palate, hemitruncal atrophy, or limb abnormalities) has been described in the literature [6].

In our patient, focal epilepsy was associated with left-sided spastic hemiparesis and facial asymmetry,

while neuropsychological assessment revealed mild intellectual disability.

Radiologically, computed tomography (CT) and brain MRI remain the primary imaging modalities used in diagnosing Dyke-Davidoff-Masson syndrome. Radiological signs vary from one patient to another, depending on the severity of cerebral involvement, including cerebral hemiatrophy, dilatation of the ipsilateral lateral ventricle, cranial bone thickening, and hyperpneumatization of the sinuses (particularly the frontal sinus) [3]. These radiological manifestations become more pronounced with age [7]. Some studies have suggested compensatory thickening of the calvarium due to intrauterine or early postnatal cerebral injury, especially within the first three years of life. Brain MRI in our patient showed right cerebral hemiatrophy, widening of the cortical sulci, dilatation of the ipsilateral lateral ventricle with midline shift, subcortical porencephalic cavities, and thickening of the right cranial vault with hyperpneumatization of the frontal sinus, which was consistent with the findings in the literature. The MR angiography sequence showed no abnormalities [7-8].

However, the management of this syndrome remains symptomatic, focused on anticonvulsant therapy to control critical manifestations, as well as motor and speech rehabilitation [1]. In some cases involving hemiplegia with pharmacoresistant epilepsy, hemispherectomy may be indicated, with an 85% success rate [8]. Patients who experience prolonged and recurrent critical manifestations or those with established hemiparesis before the age of 2 years tend to have a poor prognosis [9].

Conclusion

Dyke-Davidoff-Masson syndrome is a rare and often underdiagnosed neurological condition, associated with clinical manifestations of varying severity, including seizures, hemiparesis, and intellectual impairment. Radiological findings, particularly CT and brain MRI, have facilitated an easy diagnosis of the syndrome, revealing cerebral hemiatrophy, cranial bone thickening, and hyperpneumatization of the sinuses. Management is primarily symptomatic, and surgery is rarely indicated.

Early identification allows for more appropriate management, including rehabilitation strategies and functional support.

References

1. Al-Smair A, Abdel Hafez S, Saadeh A, Al-Ali A. An Adult with Dyke-Davidoff-Masson Syndrome: A Case Report. Cureus. 2022 Mar 19;14(3):e23315. doi: 10.7759/cureus.23315. PMID: 35464553; PMCID: PMC9015058.
2. Dyke, C.G., Davidoff, L.M., Masson, C.M. (1933). Cerebral hemiatrophy associated with hemiplegia and mental defect: A report of four cases. *Archives of Neurology and Psychiatry*.
3. El Bahri-Ben Mrad, F., Mrabet, H., Ben Sghaier, R., Mrabet, A. (2005). Dyke-DavidoffMasson syndrome: A report of two cases. *J Neuroradiol*.Jan;32(1):50-3. French. doi: 10.1016/s0150-9861(05)83022-6. PMID: 15798614
4. M. A. Duncan*, S. Vázquez-Flores, E. B. Chávez-Lluévanos, A. C. Cantú-Salinas, L. de León-Flores, H. J. Villarreal-Velázquez Dyke-Davidoff-Masson syndrome: A case study. Medicina Universitaria 2014;16(63):71-73
5. Roy, U., Panwar, A., Mukherjee, A., & Biswas, D. (2016). Adult Presentation of DykeDavidoff-Masson Syndrome: A Case Report. Case Reports in Neurology, 8(1), 20–26. doi:10.1159/000443521
6. Rondão MBA, Hsu BRRHS, Centeno RS, de Aguiar PHP. Dyke-Davidoff-Masson Syndrome: Main clinical and radiological findings- systematic literature review. Seizure. 2023 Aug; 110:58-68. doi: 10.1016/j.seizure.2023.04.020. Epub 2023 Jun 3. PMID: 37327751.
7. Abdul Rashid AM, Md Noh MSF. Dyke-Davidoff-Masson syndrome: a case report. BMC Neurol. 2018 May 29;18(1):76. doi: 10.1186/s12883-018-1079-3. PMID: 29843624; PMCID: PMC5972440.
8. Arora R, Rani JY. Dyke-Davidoff-Masson syndrome: imaging features with illustration of two cases. Quant Imaging Med Surg. 2015 Jun;5(3):469-71. doi: 10.3978/j.issn.2223-4292.2014.11.17. PMID: 26029650; PMCID: PMC4426122
9. Al-Attas AA, Alwazna EO. Presentation of Dyke-Davidoff-Masson Syndrome in adult male. Neurosciences (Riyadh). 2023 Apr;28(2):143-147. doi:10.17712/nsj.2023.2.20220084. PMID: 37045464; PMCID: PMC10155480.