International Journal of Contemporary Pediatrics
Rajan AR et al. Int J Contemp Pediatr. 2025 Nov;12(11):1886-1888
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20253498

Case Report

Dyke Davidoff Masson syndrome: rare cause of hemiparesis

A. R. Rajan[1]*, Prisha Chordia[1], A. A. Gautam[2], Priyanka Bansode[1]

[1]Department of Pediatrics, Symbiosis Medical College for Women and Symbiosis University Hospital and Research Center, Symbiosis International (Deemed University), Pune, Maharashtra, India
[2]Department of Radiodiagnosis, Symbiosis Medical College for Women and Symbiosis University Hospital and Research Center, Symbiosis International (Deemed University), Pune, Maharashtra, India

Received: 14 September 2025
Accepted: 14 October 2025

***Correspondence:**
Dr. A. R. Rajan,
E-mail: aramanrajan@gmail.com

ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare clinical entity caused by a neurological disorder that results from cerebral injury in -utero or in early infancy resulting in unilateral brain atrophy. The resulting clinical presentation is with hemiplegia, seizures, developmental delay and behavioural disorders. Our case is a 12-year-old boy who presented with hemiparesis, poor scholastic performance and seizures. The MRI brain revealed a decreased volume of the right cerebral hemisphere with gliotic change in the frontal region. The right cerebral peduncle appeared small in size with cystic encephalomalacia and thinning of the corpus callosum. MRI findings were suggestive of DDM syndrome. The electroencephalogram was abnormal. An early recognition will help reduce disability with physiotherapy and anticonvulsant therapy.

Keywords: Hemiparesis, Dyke Davidoff Masson syndrome, Vascular insult, Gliosis, Cystic encephalomalacia

INTRODUCTION

DDMS, is a rare cause of hemiparesis in children. Also known as cerebral hemiatrophy, it was initially documented by Dyke, Davidoff, and Masson in 1933. The syndrome develops following brain injury and is characterized by underdevelopment (hypoplasia) of one cerebral hemisphere.[1] Dyke-Davidoff-Masson syndrome manifests in two distinct forms: congenital and acquired. The congenital (infantile) variant emerges early in infancy, typically resulting from prenatal brain injuries such as vascular occlusion or middle cerebral artery anomalies. The acquired form develops later during childhood and stems from various conditions that compromise cerebral blood flow, including infections, prolonged febrile seizures, traumatic brain injury, haemorrhage, or ischemic events.[2] The clinical presentation varies according to the severity and extent of brain damage, encompassing a range of neurological manifestations including motor deficits (hemiparesis or hemiplegia), epileptic seizures, cognitive impairment or developmental delays, speech and language disturbances, and craniofacial asymmetry.[3]

CASE REPORT

Our patient was a 12-year-old male child born 2nd in order to non-consanguineous parents at term, by caesarean section after an uneventful antenatal period. The birth weight was 2900 grams. There were no intrapartum complications. The neonatal period was also unremarkable and the baby was discharged with breast feeding and immunization advice. By the age of one year the parents noted that as compared with his peers he had a delay in achieving motor milestones like rolling, sitting, standing and walking. The mother also noted that the boy preferentially used his right hand from infancy onwards. She however did not seek any medical advice for the above during his infancy. There was no delay in social or language domains of development. At the age of

8 years, he came to medical attention due to poor school performance and frequent falls while walking and running. He was diagnosed as a case of left sided hemiparesis but was not evaluated due to financial constraints. He reported to our hospital at the age of 12 years with a single episode of generalized tonic-clonic seizure. General examination findings were unremarkable and neurocutaneous markers were absent. Vital parameters were normal. He was normotensive. Craniofacial asymmetry was observed with smaller left hemi cranium and flattened left temporal region. The neurological examination revealed reduced bulk on the left side in both upper and lower limbs (difference 3cm lower limb and 2 cm upper limb) (Figure 1) with reduced power (grade 4/5) with increased tone (spasticity) and exaggerated reflexes on the left side).

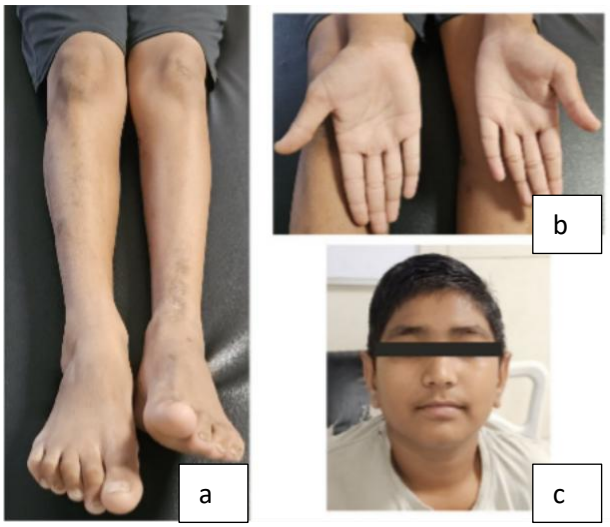

Figure 1 (a-c): Reduced bulk on left side of face, lower and upper extremity with limb length discrepancy.

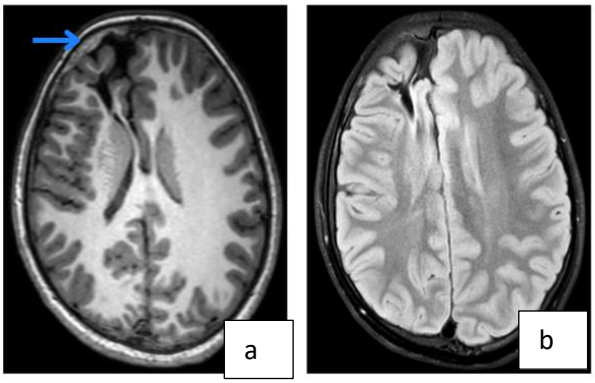

Figure 2 (a and b): MRI brain showing encephalomalacia and asymmetry in size of cerebral hemispheres.

Babinski's sign was positive on the left. There was no cranial nerve involvement and cerebellar signs were absent. The haematological tests were normal with normal liver and renal functions. The coagulation profile was normal. Magnetic resonance imaging (MRI) brain revealed partial gliosis, volume loss with cystic encephalomalacia of the right cerebral hemisphere. There was gliotic change in the right frontal region and thinning of the corpus callosum. The right cerebral peduncle appeared small in size. The right frontal sinus was enlarged and there was right sided calvarial thickening in the frontal bone with hyper pneumatization. An electroencephalogram done was abnormal suggesting seizure semiology.

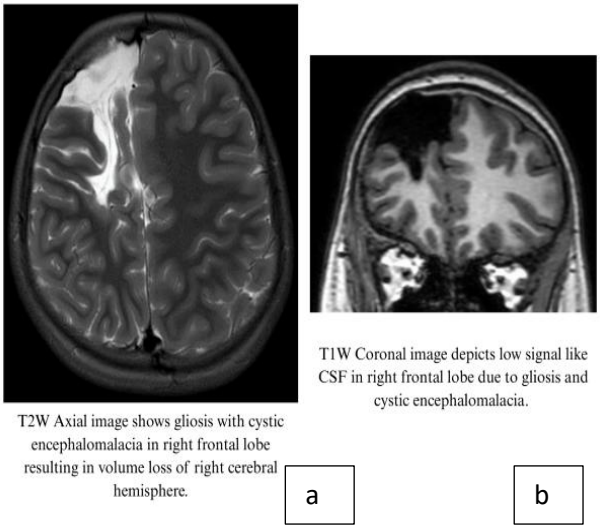

Figure 3 (a and b): MRI showing cystic encephalomalacia and volume loss on right cerebral hemisphere.

In view of the clinical evidence of left sided hemiparesis along with typical MRI findings a diagnosis of Dyke-Davidoff-Masson Syndrome was made. The child was started on anti-epileptic medication (Sodium Valproate) and physiotherapy. Parents were counselled regarding the outcome of the evaluation and likely prognosis as well as the requirement of long term anti-convulsant therapy and follow up. This case highlights the critical role of clinical awareness of DDMS as well as understanding the neuroimaging features to establish the diagnosis of this rare entity. Timely diagnosis can help plan optimal treatment stratergies.in these cases to limit disabilities.

DISCUSSION

DDMS as a cause of hemiplegia was initially described by Dyke, Davidoff and Masson in 1933 when on skull radiographic findings nine patients with facial asymmetry, hemiparesis and seizures.[1] Our case also presented with seizures and left hemiparesis along with wasting and facial asymmetry. In early onset DDMS internal carotid and middle cerebral artery abnormalities are associated findings. Though our case was early in onset there may be other causes like infection, vascular malformations (carotid artery stenosis and atherosclerosis), trauma, and intracranial bleeding that

can cause cerebral atrophy later in the perinatal period.[4] In utero vascular compromise can result in cerebral hemisphere hypoplasia. The consequence of vascular insults results in reduced neurologically derived neurotrophic factors which in turn cause atrophy of the cerebral hemisphere.[3] While an early insult results in absence of sulci and gyri subsequent acquired insults can result in encephalomalacia and large sulci in the atrophic cortex.[5] The recurrent seizures are attributed to neuronal irritability as a sequela of cortical gliosis and encephalomalacia. The delayed motor milestones and intellectual impairment is essentially due to vascular compromise during neurological development thus causing unilateral cortical damage with resulting in motor disabilities.[6]

Imaging hallmarks in DDSM include atrophic cortex on the affected side with hyper pneumatization of the paranasal sinuses and osseous hypertrophy. All these radiological features will become more obvious with increasing age.[7] The common differential diagnosis of DDMS includes Silver Russel syndrome, Rasmussen encephalitis, Germinoma of basal ganglia, Fishman syndrome and Sturge –Weber syndrome. Rasmussen encephalitis is of post viral etiology without any calvarial alterations. Sturge-Weber syndrome presents with similar neurological findings however it has characteristic leptomeningeal angiomatosis and port-wine nevus on face and tram track appearance on MRI. In Silver -Russel triangular face, prominent forehead, micrognathia, clinodactyly, feeding difficulties and growth retardation are usually present.[8] Thus, findings on neuroimaging are essential for establishing diagnosis and ruling out these conditions. Cerebral hypoplasia or hemiatrophy with hyper pneumatized sinuses along with osseous hypertrophy as a compensatory fallout are common findings on neuroimaging. With increasing age, the radiological findings become more obvious.

The treatment options are targeted at seizure control with antiepileptic drugs and motor disability limitation by regular exercise and physiotherapeutic interventions. Only if seizures are refractory surgical options like hemispherectomy can be considered. An early and definitive diagnosis is essential for disability limitation.[3]

CONCLUSION

Dyke Davidoff Masson Syndrome is a rare clinical entity that must be considered in early and late onset hemiparesis in children. Our case had hemiparesis, seizures and cerebral hemiatrophy on MRI confirming the diagnosis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Rondão MBA, Hsu BRRHS, Centeno RS, de Aguiar PHP. Dyke-Davidoff-Masson Syndrome: Main clinical and radiological findings- systematic literature review. Seizure. 2023;110:58-68.
2. Al Hatmi A, Almashaikhi T, Al Ajmi E. Imaging Features of Dyke-Davidoff-Masson Syndrome. Sultan Qaboos Univ Med J. 2023;23(1):122-4.
3. Arora R, Rani JY. Dyke-Davidoff-Masson syndrome: imaging features with illustration of two cases. Quant Imaging Med Surg. 2015;5(3):469-71.
4. Lim S, Westover C, Ratan RB, Guiahi M. Routine abortion training matters for obstetrics and gynecology residents. Open J Obstet Gynecol 2013;03(10):727–31.
5. Shrestha B. Acquired cerebral hemiatrophy: Dyke-Davidoff-Masson syndrome—a case report. Turk Neurosurg. 2011;23(1):117–21.
6. Holmes GL. Effect of seizures on the developing brain and cognition. Semin Pediatr Neurol. 2016;23:120–6.
7. Abdul Rashid AM, Md Noh MSF. Dyke-Davidoff-Masson syndrome: a case report. BMC Neurol. 2018;18(1):76.
8. Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. J Neurosci Rural Pract. 2012;3(3):411–3.

Cite this article as: Rajan AR, Chordia P, Gautam AA, Bansode P. Dyke Davidoff Masson syndrome: rare cause of hemiparesis. Int J Contemp Pediatr 2025;12:1886-8.