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**Case Report**

# Dyke-Davidoff-Masson Syndrome: A Case Report and Review of Literature

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**Abstract:** Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological disorder caused by brain injury in intrauterine or early years of life. Characteristic findings are prominent cortical sulci, dilated lateral ventricles, cerebral hemiatrophy, hyperpneumatization of the frontal sinus and compensatory hypertrophy of the skull. Hereby describing a male patient who presented with generalized tonic-clonic seizure and left-sided body weakness with neuroimaging findings of cerebral hemiatrophy, right lateral ventricle dilatation, hyperpneumatization of right frontal sinus, and asymmetric calvarial thickening. The above mentioned imaging features enables timely and accurate diagnosis, allowing appropriate management.

**Key words:** DDMS, cerebral atrophy, seizures, hemiplegia.



## INTRODUCTION

Cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome (DDMS) was described by Dyke, Davidoff, and Masson on nine patients having hemiplegia with plain skull X- ray changes [1, 2]. It is characterized by cerebral hypoplasia/hemiatrophy, seizures, facial asymmetry, and contralateral hemiplegia [1-3]. These clinical features may present with any combinations and severity. Imaging studies are helpful to make a diagnosis with its clinical features correlation. Certain imaging findings specifically like unilateral brain volume loss, ventriculomegaly, and compensatory bone hypertrophy results in cerebral hemiatrophy. Additionally hyperpneumatization of frontal sinuses and calvarial thickening may occur [3, 4]. As it is a rare disorder, it can be misdiagnosed and mismanaged.

## CASE REPORT

A case of a 21-years-old boy presented in medical emergency with complaints of generalized tonic-clonic seizures and right hemiparesis. The hemiparesis started at the age of twenty years and was non-progressive. He also had some behavioral changes like disturbed sleep, irrelevant talks, and irritability which started at the age of fourteen years, associated with frequent episodes of mild headache. No other family member or sibling had history of similar presentations. On general physical examination, there was emaciation with decreased cognitive functioning; 13/30 on the Mini-Mental State Examination. On neurological examination, the power was 3/5 on left side of the body along with decreased sensations, hypertonia, and brisk reflexes. Mild left-sided facial angle deviation was also observed. The rest of the systemic examination was unremarkable. Laboratory baseline investigations and a thorough workup for young stroke along with an autoimmune profile was done for the patient where in all laboratory investigations were within normal range. NCCT (non contrast computed tomography) scan of brain revealed atrophy of the right cerebral hemisphere, dilatation of the ipsilateral right lateral ventricle and prominence of extra-axial cerebrospinal fluid (CSF) spaces. It also showed low attenuation areas in the right frontal and right parietal lobes indicating possible subacute infarct in the right frontal and acute infarct in the right parietal region.

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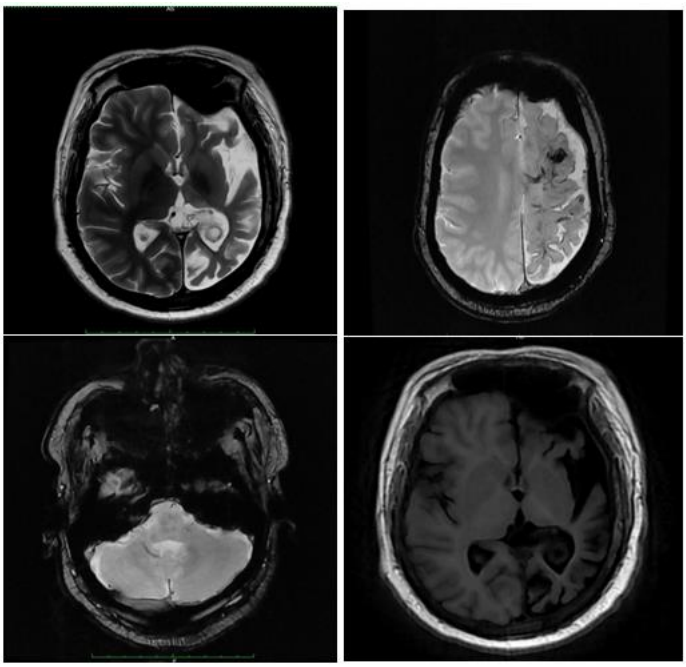

## DISCUSSION

This rare syndrome was first described by Dyke, Davidoff, and Masson back in 1933. They observed that few patients presented with specific group of symptoms including hemiparesis, facial asymmetry, seizures, and mental retardation. On skull radiography same patients had classical unilateral cerebral atrophy [5]. There is no established sex predilection or a specific hemisphere involvement but affecting the left side and in male gender are more common in the literature [6].

The clinical presentation of DDMS are seizures, contralateral hemiparesis of upper motor neuron type disease, facial asymmetry, and cognitive disabilities. Depending upon its etiology DDMS can be classified into two forms. The congenital subtype, which is symptomatic in infancy, and its pathogenesis include fetal vascular occlusion. The other is the acquired subtype, which presents in childhood. Its etiological factors include perinatal hypoxia, intracranial hemorrhage, infections, cranial trauma, and cerebrovascular lesion [7]. The possible mechanism of cerebral atrophy and the related progressive neuro deficit is hypothesized to be due to several ischemic episodes resulting from these factors, which reduce the production of brain-derived neurotrophic factors, and in turn leads to cerebral atrophy [8].

CT and MRI are the two gold standard imaging modalities that prove to be very significant in the diagnosis of DDMS. These two imaging modalities provide very detailed cross-sectional images. The typical imaging features for DDMS include prominent cortical sulci, dilated lateral ventricles, cerebral hemiatrophy, hyperpneumatization of the frontal sinus, and compensatory hypertrophy of the skull. These imaging findings become more obvious as the patient ages [9]. When the cerebral damage occurs during the intrauterine period or before the age of 3, compensatory calvarial involvement can be seen [10, 11]. In a patient with cerebral hemiatrophy, the differential diagnosis includes Rasmussen encephalitis, Fisherman syndrome, Sturge- Weber syndrome, basal ganglia dysgerminoma, and Silver-Russell syndrome. Detailed history and examination along with laboratory and imaging workup are required to differentiate these diseases. For the management of seizures, mono, or poly anticonvulsant medication is given. Children with refractory epilepsy and hemiplegia are potential candidates for hemispherectomy, with a success rate of 85%. Vagal stimulation is another alternative. Despite lacking any specific treatment algorithm, therapy with antiepileptics and surgery is indicated in specific cases. Long-term supportive management includes physical, language, and occupational therapy [4].

## CONCLUSION

DDMS is a rare neurological disorder leading to intractable seizures along a spectrum of disabilities. As it is a rare syndrome, it may be easily misdiagnosed by the inexperienced eye. Detailed history and examination are needed with imaging modalities to diagnose it. Physicians should be aware of signs and symptoms, risk factors, and diagnostic features of DDMS so that the patients could be managed properly.

**Compliance with ethical standards**

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**Ethical Approval**

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

**Informed Consent:** Informed consent was obtained from individual participant included in the study.

**Authors' contributions**

1. DR. SHREE VARSHINI.T (SV)
2. DR. ANBUMANI (AM)
3. DR. ARAVIND (AD)
4. DR. PRABHAKARAN.M (PM)

Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work –
Drafting the work or revising it critically for important intellectual content
Final approval of the version to be published -
Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved –

Dear Editor,
I, Dr. Shree varshini T, corresponding author submit this manuscript titled "A case report of Dyke-Davidoff-Masson Syndrome" to your esteemed journal for publication under QJM: EAS Journal of Radiology and Imaging Technology. Conflicts of interest and sources of financial support – Nil

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## REFERENCES

1. Paudel, K., & Venugopal, A. (2013). Dyke-Davidoff-Masson syndrome. *Journal of Nepal Medical Association*, *52*(189), 272-274.
2. Erdem, A., Vedat, A. C. I. K., Leventoğlu, A., Sarilar, C., & Cansu, A. (2009). Effect of Vagal Nerve Stimulation in Dyke–Davidoff–Masson Syndrome with Refractory Generalized Seizures–Case Report. *Turkish neurosurgery*, *19*(2), 197-199.
3. Aguiar, P. H., CHING, W. L., LEITÃO, H., Issa, F., Lepski, G., Figueiredo, E. G., ... & MARINO JR, R. A. U. L. (1998). MR and CT imaging in the Dyke-Davidoff-Masson syndrome: report of three cases and contribution to pathogenesis and differential diagnosis. *Arquivos de neuro-psiquiatria*, *56*, 803-807.
4. Behera, M. R., Patnaik, S., & Mohanty, A. K. (2012). Dyke-Davidoff-Masson syndrome. *Journal of Neurosciences in Rural Practice*, *3*(03), 411-413.
5. Dyke, C. G. (1933). Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. *Surg Gyn Obstet*, *57*, 588-600.
6. Ünal, Ö., Tombul, T., Çırak, B., Anlar, Ö., İncesu, L., & Kayan, M. (2004). Left hemisphere and male sex dominance of cerebral hemiatrophy (Dyke–Davidoff–Masson Syndrome). *Clinical imaging*, *28*(3), 163-165.
7. Pendse, N. A., Bapna, P., Menghani, V., & Diwan, A. (2004). Dyke-Davidoff-Masson syndrome (DDMS). *Indian journal of pediatrics*, *71*(10), 943-943.
8. Ono, K., Komai, K., & Ikeda, T. (2003). Dyke–Davidoff–Masson syndrome manifested by seizure in late childhood: a case report. *Journal of clinical neuroscience*, *10*(3), 367-371.
9. Rashid, A. M. A., & Noh, M. S. F. M. (2018). Dyke-Davidoff-Masson syndrome: a case report. *BMC neurology*, *18*(1), 1-3.
10. Arora, R., & Rani, J. Y. (2015). Dyke-Davidoff-Masson syndrome: imaging features with illustration of two cases. *Quantitative Imaging in Medicine and Surgery*, *5*(3), 469-471.
11. Atalar, M. H., Icagasioglu, D., & Tas, F. (2007). Cerebral hemiatrophy (Dyke–Davidoff–Masson syndrome) in childhood: clinicoradiological analysis of 19 cases. *Pediatrics International*, *49*(1), 70-75.

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