### **Case Report**

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20205528

# Dyke Davidoff Masson syndrome: a rare cause of cerebral hemiatrophy in 11 months old

Amit Vatkar\*, Ipsita Vashishtha, Vinaykumar P. Hedaginal, B. Revanth Sai Madhav

Department of Paediatrics, DY Patil University, School of medicine, Navi Mumbai, Maharashtra, India

**Received:** 21 October 2020 **Accepted:** 05 December 2020

## \*Correspondence: Dr. Amit Vatkar,

E-mail: vatkaramit@yahoo.com

**Copyright:** © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

#### **ABSTRACT**

Dyke-Davidoff-Masson syndrome (DDMS) is an uncommon condition, characterized radiologically by cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses, of unknown frequency resulting from brain injury due to large no of causes; especially in early life. Mostly presents early in life with seizures, learning difficulty, contralateral hemiparesis and facial symmetry. Here we present a case of 11 months old female child with developmental delay, visual abnormality, microcephaly and spastic hemiplegia. CT-brain done which was suggestive of infantile type of cerebral hemiatrophy or DDMS.

**Keywords:** Dyke-Davidoff-Masson syndrome, Hemiatrophy, Spastic hemiplegia

#### INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) is an uncommon disease with unknown frequency with available literature mostly from case reports/series. It refers to atrophy/hypoplasia of one cerebral hemisphere, due to an insult to the developing brain in fetal or early childhood period.<sup>1</sup>

Hageman et al proposed the terms cerebral hemi hypoplasia or unilateral cerebral hypoplasia for primary (congenital) cerebral atrophy owing to the fact that there is a lack of cerebral development rather than atrophy.<sup>2</sup>

DDMS is occasionally seen in clinical practice. It has been reported that DDMS is caused by cerebral insult that may occur in utero when the maturation of calvarium has not been completed, or during early life due to brain damage (usually traumatic). The clinical findings may be of variable degree according to the extent of brain injury.<sup>3</sup>

More commonly they present with recurrent seizures, facial asymmetry, contralateral hemiplegia, mental

retardation or learning disability, and speech and language disorders.

It was initially described as changes in the skull seen on skull X-ray in patients with cerebral hemiatrophy but is now applied more broadly to cross-sectional imaging. It was initially described by C. G. Dyke, L. M. Davidoff and C. B. Masson in 1933.<sup>4</sup>

Radiographic features may include spectrum of findings: atrophy in basal ganglia and/or brain stem, calvarial thickening on affected side, Wallerian degeneration of the mesencephalon and middle fossa hypoplasia, capillary malformations (in some situations), hyper pneumatisation of mastoid cells on affected side.

Some authors divide the condition into two types mainly dependent on clinical presentation age.<sup>5</sup>

Congenital or infantile: Patient becomes symptomatic in the perinatal period or infancy. Variable causes such as neonatal or gestational vascular occlusion involving the middle cerebral artery, infection, coarctation of the mid aortic arch and unilateral cerebral arterial circulation anomalies.

Acquired: Tumor, infection, trauma, ischemia, prolonged febrile seizures and haemorrhage.

#### **CASE REPORT**

A 11 months old female child, born full term, to nonconsanguineous parents presented with developmental delay mostly in the motor and language domain child is not able to stand or walk with support and not able to speak a single word, only cooing present. He had no history significant antenatal history suggestive of bilateral lateral ventricular dilatation for which parents were advised abortion by some doctor but chose to continue with the pregnancy, no perinatal complications. He had a microcephaly with head circumference of 40 cm (<3<sup>rd</sup> percentile) without any neurocutaneous marker or facial asymmetry. Child cannot sit but has attained neck holding partial, recognizes mother and father, speechcooing. These are delayed development as per age. vision-bilateral nystagmus, squint. The bilateral carotid pulsations were normal with no bruit. Neurological examination bilateral upper limb spastic hemiparesis with brisk tendon reflexes and extensor planter response, other cranial nerves and systemic examinations being normal.

On fundus examination bilateral mild disc pallor present with subtle pigmentary changes.

Based on antenatal diagnosis, USG Brain done showing dilated 3<sup>rd</sup> and both lateral ventricles.

A plain CT brain done shown parenchyma of the left cerebral hemisphere is mildly atrophic with asymmetrical enlargement of the left lateral ventricle. The atrium of right lateral ventricle is mildly prominent with dilatation of the 3<sup>rd</sup> ventricle. The aqueduct of Sylvius and the 4<sup>th</sup> ventricle are normal. Non-enhanced CT brain findings consistent with unilateral cerebral atrophy with resultant dilatation of the left lateral ventricle (Figure 1).



Figure 1: Unilateral cerebral Atrophy with dialation of left lateral ventricle.

MRI brain (plain and contrast) done suggestive of mildmoderate dilatation of bilateral lateral ventricle (left>right) and the 3<sup>rd</sup> ventricle with paucity of bilateral periventricular white matter and mild thinning of corpus callosum (Figure 2).



Figure 2: Paucity of bilateral peri-ventricular white matter and mild thinning of corpus callosum.

From the above CT and MRI findings, diagnosis of DDMS is made.

Patient was started on tab baclofen (10 mg) ½ tab BD followed by sessions of physiotherapy. To which child seems to be improving as spasticity is decreasing and child is now able to sit without support.

#### DISCUSSION

Brain reaches half of its adult human size during the first year of life and the surface of the hemisphere remains smooth and uninterrupted until early in the fourth month of gestation. By the end of the eighth month, all the important sulci can be recognized.6 In 1933, Dyke, Davidoff and Masson first described the syndrome in plain radiographic and pneumoencephalographic changes in a series of nine patients.<sup>4</sup> It is characterized by asymmetry of cerebral hemispheric growth with atrophy or hypoplasia of one side and midline shift, ipsilateral osseous hypertrophy with hyper pneumatisation of sinuses mainly frontal and mastoid air cells with contralateral paresis.<sup>7</sup> The developing brain presses outward on the bony skull table resulting in gradual increase in head size and shape. When the brain fails to grow properly, the other structures grow inward resulting in increased width of diploic spaces, enlarged sinuses, and elevated orbital roof.8

DDMS is caused by an insult to growing brain either cerebrum in utero when maturation of calvarium has not been completed, or during early life due to damage to brain. It is characterized by asymmetry of cerebral hemispheric growth with hypoplasia or atrophy of one side. Other features are dilatation of ipsilateral ventricle,

cisternal space, decrease in size of ipsilateral cranial fossa, enlargement of ipsilateral sulci and unilateral thickening of skull

Clinical features and age of presentation depends on the time of insult and characteristic changes, child may present with any of the following clinical features seizures (generalized or focal), mental retardation, contralateral hemiparesis, learning disabilities, improved speech, variable degree of facial asymmetry etc. Left side involvement with male gender is more common.

A clinical history and radiological findings (CT or MRI) are needed for right diagnosis.

Differential diagnosis-Sturge-Weber's syndrome, basal cell germinoma, Fishman syndrome, Silver-Russell syndrome, linear nevus syndrome, and Rasmussen encephalitis. 9,10

Here in our case, the findings of left cerebral hemiatrophy with enlarged cortical sulci, microcephaly, and presentation at the age of 11 months reflect an onset of brain insult after the completion of sulci formation, probably of vascular origin involving left middle cerebral artery.

Treatment is symptomatic and should target treating seizures, hemiparesis, hemiplegia, learning difficulties, Speech therapy, visual stimulation effects for visual defects, etc.

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

#### **REFERENCES**

- Sharma S, Goyal I, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff-Masson Syndrome. Ind J Radiol Imag. 2006;16:165-6.
- 2. Hageman G, Gooskens RH, Willemse J. A cerebral cause of arthrogryposis: Unilateral cerebral hypoplasia. Clin Neurol Neurosurg. 1985;87:119-22.
- Goyal J, Shah V, Rao S, Jindal N. Dyke–Davidoff– Masson syndrome in children. Internet J Pediatr Neonatol. 2009;10:101-7.
- 4. Dyke CG, Davidoff LM, Masson CB. Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. Surg Gynecol Obstet. 1933;57:588-600.
- 5. Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. J of neurosciences in rural practice. 2012;3(3):411-3.
- 6. Graham A, Molnar Z. Development of the Nervous system. In: Standring S, editor. Gray's Anatomy. 40th ed. London, Churchill Livingstone: Elsevier. 2008;385.
- 7. Pendse NA, Bapna P, Menghani V, Diwan A. Dyke-Davidoff-Masson Syndrome. Indian J Pediatr. 2004;71:943.
- 8. Parker CE, Harris N, Mavalwala J. Dyke-Davidoff-Masson Syndrome: Five case studies and deductions from dermatoglyphics. Clin Pediatr. 1972;11:28.
- 9. Rao KC. Degenerative diseases and hydrocephalus. In: Lee SH, Rao KC, Zimmerman RA, editors. Cranial MRI and CT. New York: McGraw-Hill. 1999:212–4.
- 10. Zilkha A. CT of Cerebral hemiatrophy. Am J Roentgenol. 1980;135:259-62.

Cite this article as: Vatkar A, Vashishtha I, Hedaginal VP, Madhav BRS. Dyke Davidoff Masson syndrome: a rare cause of cerebral hemiatrophy in 11 months old. Int J Contemp Pediatr 2021;8:187-9.