

Dyke-Davidoff-Masson Syndrome: A Case Report

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Abstract

Background : Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological disorder that results from brain injury in intrauterine or early years of life. This condition is characterised by hemicerebral atrophy/hypoplasia secondary to brain insult and is accompanied by ipsilateral compensatory osseous hypertrophy and contralateral hemiparesis.

Case Presentation : Here we describe about a 17 months old female child who presented with unable to stand or walk without support till date and left sided weakness since birth. Neurological examination revealed left-sided spastic hemiparesis with brisk tendon reflexes and equivocal planter response. MRI of brain revealed mild focal parenchymal volume loss in left superior parietal lobule with evidence of gliotic changes in the adjacent cerebral parenchyma. Left cranial fossa appeared to be smaller in size with slightly thickened skull bones and left lateral ventricular dilatation. Body and splenium of the corpus callosum was grossly hypoplastic. Focal parenchymal volume loss bordered by T2-flair hyperintensity was noted at right parieto-temporo-occipital region along with exvacuo dilatation of adjacent part of right lateral ventricle, suggesting encephalomalacic changes.

Conclusion : As our patient presented with hemiparesis and developmental delay, her parents were counselled to take her to child development center for developmental therapy with regular follow up.

Key words: Contralateral hemiparesis; Dyke-Davidoff-Masson Syndrome (DDMS); Hemicerebral atrophy.

INTRODUCTION

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological condition that was first described by Dyke, Davidoff, and Masson in a series of nine patients with hemiplegia and plain skull X-ray changes.^{1,2} The syndrome is characterized by facial asymmetry, seizures, hemiplegia or contralateral hemiparesis, and mental retardation.^{3,4,5} These clinical features can present with diverse combinations and severity. The etiology can be congenital or acquired as a result of an alteration in the cerebral perfusion during prenatal, perinatal or early childhood periods.^{3,5,6,7,8} Specific imaging findings include unilateral brain volume loss, ventriculomegaly and compensatory bone hypertrophy resulting in cerebral hemiatrophy. In addition, calvarial thickening and hyperpneumatization of frontal sinuses may occur.^{5,9}

CASE PRESENTATION

A 17 months old, female child, 1st issue of non-consanguineous parents, presented on 10th December 2021 at Chattogram Maa Shishu -O-General Hospital with the complaints of unable to stand or walk without support till date and slight less

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movement of left lower limb since birth. She had no history of convulsion. She was delivered at term via vaginal delivery in a Upazilla Health complex and had history of hospitalization after birth due to Birth Asphyxia (Hypoxic Ischemic Encephalopathy Stage-I) with Early Onset Neonatal Sepsis. She had delayed milestones of development in the form of not able to stand or walk without support till date. She had microcephaly (< 3rd centile) without any neurocutaneous marker or facial asymmetry. Her vision, hearing were normal and could say 8-10 words.

Neurological examination revealed left-sided spastic hemiparesis with brisk tendon reflexes and equivocal planter response. Cranial nerves were intact and other systemic examinations were normal.

MRI of brain revealed mild focal parenchymal volume loss in left superior parietal lobule with evidence of gliotic changes in the adjacent cerebral parenchyma. Left cranial fossa appeared to be smaller in size with slightly thickened skull bones and left lateral ventricular dilatation. Body and splenium of the corpus callosum was grossly hypoplastic. Focal parenchymal volume loss bordered by T2-flair hyperintensity was noted at right parieto-temporo-occipital region along with exvacuo dilatation of adjacent part of right lateral ventricle, suggesting encephalomalacic changes.

All these features were suggestive of Dyke-davidoff Masson Syndrome. After diagnosis was made, parents were counselled accordingly and developmental therapy was advised for the child with regular follow up.

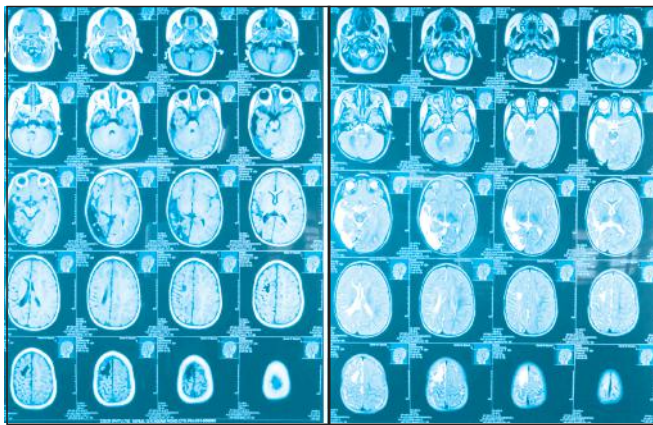


Figure 1 MRI of brain

DISCUSSION

Dyke-davidoff-masson syndrome is a condition characterised by hemispheric atrophy/hypoplasia secondary to brain insult usually in fetal or early childhood period and is accompanied by ipsilateral compensatory osseous hypertrophy and contralateral hemiparesis.

It is characterised by:

- Thickening of the skull vault (Compensatory)
- Enlargement of the frontal sinus (Also ethmoidal and mastoid air-cells)
- Elevation of the petrous ridge
- Ipsilateral falcine displacement
- Capillary malformations.

Clinically, patients may have seizures, mental retardation, contralateral hemiparesis, and facial asymmetry. Our patient had delayed milestones of development, contralateral hemiparesis and microcephaly. Birth history revealed hypoxic-ischemic injury. But there was no history to suggest intra-uterine/perinatal infection. MRI of brain was suggestive of Dyke-davidoff Masson Syndrome. Though our patient had no history of seizure but seizures do not always appear during early childhood and often begin months or years after the hemiparesis onset.^{4,5}

Sometimes patients display refractory epilepsy, and the treatment must focus on seizure control with anticonvulsant medication being either mono or polytherapy.⁶ Children with refractory epilepsy and hemiplegia are potential candidates for hemispherectomy, with a success rate of 85%. There is evidence that 30%-50% of patients with partial epilepsy may experience a decrease in convulsions in over 50% of the cases.⁴ The prognosis is better if the hemiparesis appears after the first 2 years of age and in absence of recurrent and prolonged epilepsy.

Even though an established protocol for management is lacking, there is the indication for therapy including anticonvulsants and surgery in specific cases. In addition, physical therapy, occupational therapy and language therapy play significant roles in the long term management of the patients.⁶

CONCLUSION

Due to its rarity, Dyke-Davidoff Mass on Syndrome may easily be missed by the majority of treating clinicians. Knowledge of its features on imaging enables timely and accurate diagnosis allowing appropriate management.

DISCLOSURE

All the authors declared no competing interest.

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