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CASE REPORTS

A RARE PRESENTATION OF DYKE-DAVIDOFF-MASSON SYNDROME

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ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological condition occurring due to an insult to the developing brain. We describe an infant with subtle hemiplegia and cerebral hemi-atrophy. With physiotherapy and occupational therapy, improvement in tone and power was noted. Early diagnosis and rehabilitation of DDMS improves prognosis.

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KEYWORDS

hemiplegia, cerebral hemi-atrophy, Dyke-Davidoff Masson

Case Report

An 8-month-old male infant, born of a non-consanguineous marriage, with normal perinatal history, presented with predominant usage of the left upper and lower limbs. There was no history of trauma or fever or seizures or cognitive decline. He was developmentally normal with normal vision and hearing.

On examination, he had right sided spastic hemiplegia, exaggerated deep tendon reflexes, fisting movements of the right hand. Power on the right side was 2/5 whereas that on the left was >3/5. There was no facial asymmetry, neurocutaneous markers, limb length or girth discrepancy or facial dysmorphism.

Baseline blood investigations were normal. MRI brain showed diffuse left sided cerebral hemi-atrophy, cystic encephalomalacic changes, ipsilateral wallerian degeneration, subtle atrophy of the midbrain, pons and medulla, dilated lateral ventricle, falcine displacement, left sided mild calvarial thickening and contralateral cerebellar hemisphere atrophy (Figure 1, 2). Left middle cerebral artery attenuation and hypoplasia of left intracranial internal carotid artery was also noted. Features were suggestive of Dyke-Davidoff-Masson syndrome.

Discussion

Dyke-Davidoff-Masson syndrome (DDMS), a rare neurological condition, first described in 1933 in 9 patients, using plain skull radiographs. The exact incidence for DDMS is unknown. It is characterized by cerebral hemi-atrophy, skull/ facial asymmetry, contralateral hemiplegia, seizures and developmental delay occurring with varying degrees of severity.[1] These occur due to brain injury during the intrauterine, neonatal or early childhood period.

The aetiology of Dyke-Davidoff Masson syndrome in this child could be either vascular insult during intrauterine life or acquired causes like trauma, infection, or intracranial haemorrhage in the perinatal period or shortly thereafter causing hemi cerebral atrophy. Unlike the other cases[2,3] reported, this child did not have associated seizures, facial asymmetry, or developmental delay. Other radiological findings reported in this syndrome include hyperpneumatization of the paranasal sinuses and mastoid cells[4,5], which was not seen in this child probably due to his age.

Facial asymmetry has also been described in literature. Male preponderance and left sided cerebral involvement has been described in other studies.[5,6] Other findings reported include café au lait spots, ocular lipodermoid, dental manifestations- delayed eruption of teeth, hypoplasia, and taurodontism.[7] Psychiatric symptoms have been described in older children and adults.[7,8] Treatment is conservative and includes physiotherapy, occupational and speech therapy, and treatment of seizures and other comorbidities, if associated. Hemispherectomy, as a treatment for

intractable disabling seizures, with good success rate has been reported.[9]

This child was advised physiotherapy and occupational therapy, which was taught to the care givers and was continued at home. The child is on regular follow up. At follow up, improvement was noted in the tone and power of the affected limbs. This child did not have associated seizures or cognitive decline on follow up. At follow up, the child was noted to be gaining milestones appropriately.

Therefore, knowledge of the presenting features and radiological findings of this syndrome help in early diagnosis and rehabilitation, thereby improving prognosis and quality of life.

Compliance with Ethical Standards
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Conflict of Interest None

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