Images in...

Dyke-Davidoff-Masson syndrome

Rajesh Verma, Ritesh Sahu

Department of Neurology, Chhatrapati Shahuji Maharaj Medical University, Lucknow, Uttar Pradesh, India

Correspondence to Professor Rajesh Verma, drrajeshverma32@yahoo.com

DESCRIPTION

An 18-month-old baby presented with decreased movements of right upper and lower limb since birth. The birth history revealed perinatal insult in the form of delayed cry that was followed by right-sided complex partial seizures. The patient exhibited delayed milestones. There is no history of cyanosis, jaundice or head injury in the neonatal period. At present, the patient is unable to walk without support.

On examination, the vitals were stable. General examination revealed comparative shortening of right upper and lower limbs. The cranial nerve examination showed normal findings. The tone on the right side was increased in the form of spasticity. She preferably uses her left hand for holding objects. The reflexes were brisk on the right side and plantar response was extensor on the right side. The painful stimulus provoked withdrawal response. Other system examination did not reveal any abnormality. MRI

of cranium, demonstrated left cerebral hemisphere atrophy with dilated ventricles on same side (figures 1 and 2). The clinical evaluation and neuroimaging findings suggested the diagnosis of Dyke-Davidoff-Masson syndrome (DDMS).

The characteristic features of DDMS are convulsions, contralateral hemiplegia, mental retardation and hemiatrophy of the brain.¹

In 1933, Dyke, Davidoff and Masson presented a series of nine patients who were characterised clinically by hemiparesis, facial-asymmetry, seizures and mental retardation where they described the plain skull radiographic and pneumatoencephalographic changes.² Shen *et al*³ described three patterns of MRI showing cerebral hemiatrophy: MRI pattern I corresponds to diffuse cortical and subcortical atrophy; pattern II corresponds to diffuse cortical atrophy coupled with porencephalic cysts and pattern III corresponds to previous infarction with gliosis in middle cerebral artery territory.

Figure 1 T2 FLAIR axial image showing left cerebral hemiatrophy with dilated left lateral ventricle.

Figure 2 T2-weighted axial image showing left cerebral hemiatrophy with significant dilated left lateral ventricle.

Learning points

- ▶ Dyke-Davidoff-Masson syndrome is a rare epilepsy syndrome with predominant manifestations of seizures, facial asymmetry, hemiatrophy, contralateral hemiparesis, skull vault thickening and mental retardation.
- ▶ The common causes of cerebral hemiatrophy are congenital anomalies, ischaemic cerebrovascular disease, perinatal hypoxia, intracranial haemorrhage, cerebral injury and infections.
- ▶ Perinatal hypoxia is quite common in developing nations, including India, due to inadequate obstetric care.

Competing interests None.

Patient consent Obtained.

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