Dyke-Davidoff-Masson syndrome: unusual cause of hemiplegic cerebral palsy

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DESCRIPTION

A 2-year-old-girl presented with recurrent episodes of right focal seizures. She had a history of prolonged febrile status epilepticus at 15 months, and subsequently had two more right focal motor seizures at 18 months of age. She was noted to have a paucity of movements of the right half of the body subsequent to the second seizure. She was born to non-consanguineous parents and had an uneventful antenatal and perinatal period. Her development was age appropriate. Her family history was unremarkable. On examination, she had normal anthropometric parameters, right upper motor type of facial palsy and right hemiparesis. She could speak two-word sentences and could walk with support. There were no asymmetric findings on physical examination. A clinical possibility of hemiconvulsion-hemiplegia-epilepsy (HHE) syndrome or Rasmussen encephalitis was considered.

An interictal electroencephalogram revealed frequent spikes and slow waves originating from the left frontotemporal region. A neuroimaging showed left hemispheric atrophy with thickening of the ipsilateral calvarium (figure 1). A diagnosis of Dyke-Davidoff-Masson syndrome (DDMS) was made, and the child continued on antiepileptic drugs. Physiotherapy and occupation rehabilitation was initiated.

DDMS was first described in 1933 by Dyke and colleagues. Classically, it presents with hemiparesis, focal or generalised epilepsy, facial asymmetry, varying degree of mental retardation, behavioural abnormalities, hemiatrophy and sensory disturbances. The two forms of the disease are congenital and acquired.[1] Congenital form presents in the neonatal period or infancy and considered secondary to vascular insult in the antenatal or neonatal period. The acquired form is consequent to haemorrhage, trauma and ischaemic injury to the brain.[1] The age of symptom onset depends on the timing of cerebral insult and the extent of brain parenchymal involvement.[2] The typical neuroimaging shows hemiatrophy, thickened ipsilateral calvarium, dilatation of ventricles and hyperpneumatisation of mastoid or frontal sinuses. Hypoplastic middle cranial fossa; thalamic and cerebellar peduncles atrophy; widening of the Sylvian fissure; cortical sulci and hypoplasia of lentiform nucleus are additional features.[3] The index child had the congenital form of DDMS, characterised by hemiatrophy of entire cerebral hemisphere and focal epilepsy. The congenital form of DDMS with focal status epilepticus should be differentiated from HHE syndrome and Rasmussen encephalitis, supported by lack of typical MRI features, the presence of thickened calvaria and hyperpneumatised sinuses and relative non-progressive focal deficits.

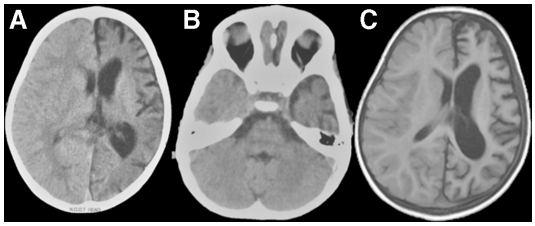

Figure 1 Neuroimaging of the child with Dyke-Davidoff-Masson syndrome. Axial sections of CT (A, B) and MRI (C) of the brain showing atrophy of left hemisphere with prominent sulci, atrophy of left caudate and ipsilateral dilatation of lateral ventricles. Note the thickening of ipsilateral calvarium (A–C) and hyperpneumatised left mastoid sinus (B).

Learning points

- Dyke-Davidoff-Masson syndrome is an unusual cause of hemiplegic cerebral palsy.
- Unilateral cerebral atrophy, thickened ipsilateral calvarium, dilatation of ventricles and hyperpneumatisation of mastoid or frontal sinuses are typical findings in neuroimaging.
- Congenital form is considered secondary to vascular insult in the antenatal or neonatal period.

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REFERENCES

1 Karuppiah S, Rodgman C, Lombard J. Dyke-Davidoff-Masson syndrome in postcerebral malaria. *J Child Neurol* 2009;24:487–90.
2 Zawar I, Khan A, Sultan T, *et al*. An unusual cause of status epilepticus. *Neurosciences* 2015;20:385–7.
3 Gökçe E, Beyhan M, Sade R. Radiological imaging findings of Dyke-Davidoff-Masson syndrome. *Acta Neurol Belg* 2017;117:885–93.

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