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A rare case of spastic hemiplegic Cerebral Palsy: Dyke Davidoff Masson Syndrome

[1]Dr. Ipsita Vashishtha, Senior Resident, Department of Paediatrics, D.Y.Patil University and School of Medicine, Nerul, Navi Mumbai, Maharashtra, India

[2]Dr. Srikanth Velupula, Junior Resident, Department of Paediatrics, D.Y.Patil University and School Of Medicine, Nerul, Navi Mumbai, Maharashtra, India

[3]Dr. Neelu Elon, Assistant Professor, Department of Paediatrics, D.Y.Patil University and School Of Medicine, Nerul, Navi Mumbai, Maharashtra, India

[4]Dr. Prithi Inamdar, Associate Professor, Department of Paediatrics, D.Y.Patil University and School of Medicine Nerul, Navi Mumbai, Maharashtra, India

[5]Dr. Rajesh Rai

Corresponding Author: Dr. Srikanth Velupula, Junior Resident, Department of Paediatrics, D.Y.Patil University and School Of Medicine, Nerul, Navi Mumbai, Maharashtra, India

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Abstract

Dyke Davidoff Masson Syndrome (DDMS) is a rare clinical entity characterised by clinical features of hemiparesis, seizures, facial asymmetry and mental retardation with contralateral cerebral hemiatrophy and homolateral hypertrophy of skull and sinuses. Hereby, reporting a case of DDMS in a 9 yrs old male child who presented with spastic hemiparesis of right upper and lower limbs, seizures, global developmental delay with microcephaly , facial asymmetry and MRI brain suggestive of characteristic diagnostic features of DDMS.

Keywords: Spastic Hemiparesis, Cerebral Hemiatrophy, Microcephaly

Introduction

Dyke-Davidoff-Masson syndrome is a rare neurological condition, refers to atrophy or hypoplasia of one cerebral hemisphere called hemiatrophy due to an insult to a developing brain either in the fetal or early childhood period.[1] DDMS could be either congenital or acquired, congenital forms presents either in neonatal or early infancy period secondary to vascular insult in the antenatal or neonatal period and acquired form is result of the trauma, hemorrhage or ischemic injury to the brain.[2] Clinical features may vary depending upon the extent of brain parenchyma involved, most commonly presenting with recurrent seizures, contralateral spastic hemiparesis, facial asymmetry and developmental delay involving motor and speech domain.

Radiologically, varying degrees of cerebral atrophy of the affected hemisphere along with ipsilateral dilatation of the lateral ventricle and sulcal prominence accompanied by homolateral hypertrophy of the skull and sinuses with elevation of the petrous ridge have been noted.[3]

Other imaging findings may include such as[3] :

- Wallerian degeneration of the mesencephalon and middle fossa hypoplasia
- Atrophy in basal ganglia
- Atrophy in the brain stem
- Capillary malformations: may be detected in some situations
- Calvarial thickening (affected side)
- Hyperpneumatization of mastoid cells (affected side)

Case Report

Here we report a case of an 8 years old boy who was brought with a complaint of right sided hemiparesis with recurrent generalised tonic clonic seizures since 4 yrs age. There was a history of head trauma leading to intracranial hemorrhage(details not known by the mother) at 5 months of age followed by 1 episode of generalised tonic clonic convulsion post which the child had loss of consciousness which lasted for almost 15-20 mins. Child was admitted for 7 days and was started on an antiepileptic drug, (details not known).The Child was on medication till 2 years of age after which he abruptly stopped the medicines.

The child remained convulsion free for another 2 years before he started convulsing again at 4 years of age. The patient was started on sodium valproate.

However, continued to have seizures on and off. His mother revealed a history of developmental delay in all 4 domains (gross motor, fine motor, language and social) post trauma before which child had achieved age appropriate milestones. Patient had a history of right sided weakness of both upper and lower limb, noticed by the mother at 3 yrs of age, which was non progressive in nature. There was no significant antenatal history or similar illness in any other sibling or family member.

On examination, the patient was conscious with an intelligence quotient of 60 indicating mild intellectual disability, with head circumference 46 cms <3rd centile for his age and sex. Neurological examination revealed spastic hemiperesis with 4/5 power in the right upper and lower limb with brisk reflexes. Plantar reflex was extensor (babinski sign positive). There were no signs of meningial irritation such as neck rigidity, any sensory deficit, cranial nerve or bowel bladder involvement.

All routine hematological, biochemical and metabolic workup was within normal range.

Magnetic Resonance Imaging (MRI) Brain done showed multiple large gliotic areas involving the entire left cerebral hemisphere with sparing of the basal ganglia and thalamus with prominence of the adjacent cortical sulci and ex vacuo dilatation of the adjacent left lateral ventricle.

Significant resultant volume loss of the left cerebral hemisphere with shift of midline towards left.

Thinning of the left cerebral peduncle suggestive of Wallerian degeneration.(fig1,2,3)

Twenty-one channel electroencephalography recordings under sedation done suggestive of right sided epilepsy. Based on characteristic radiological findings, the patient was diagnosed as a case of Dyke–Davidoff–Masson syndrome (DDMS) and was treated with Valproic acid and Levetiracetam. For rigidity, Tab baclofen along with sessions of physiotherapy.

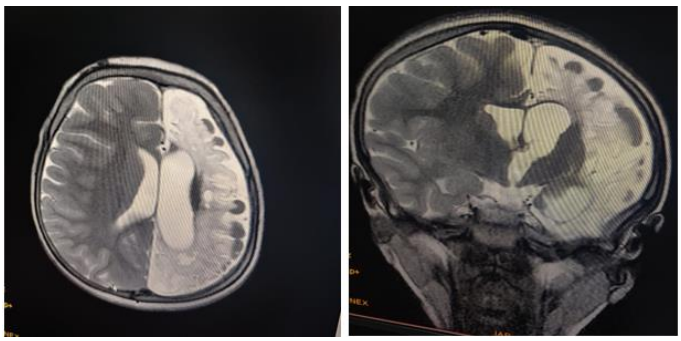

Fig 1,2-Multiple large gliotic areas involving the entire left cerebral hemisphere with sparing of the basal ganglia and thalamus with prominence of the adjacent cortical sulci and ex vacuo dilatation of the adjacent left lateral ventricle. Significant resultant volume loss of the left cerebral hemisphere with shift of midline towards left.

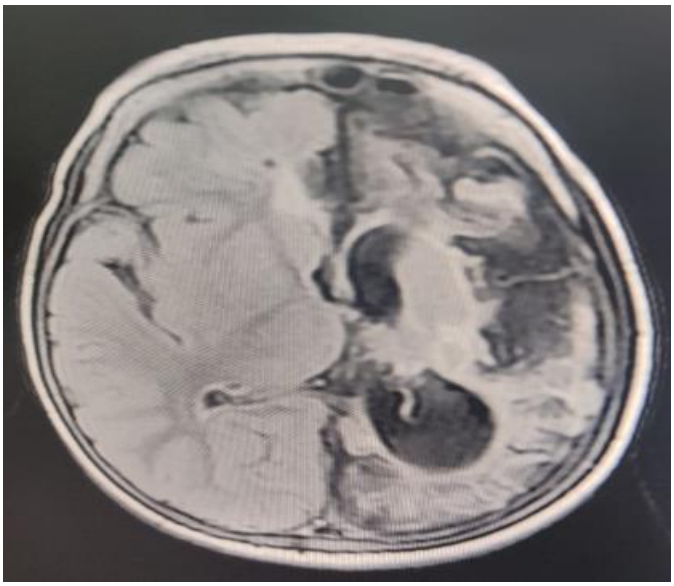

Fig 3: Thinning of the left cerebral peduncle suggestive of Wallerian degeneration

Discussion

In 1933, Dyke, Davidoff and Masson first described the syndrome in plain radiographic and pneumoencephalography changes in a series of nine patients [4]. 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.[5] Affected population is usually pediatrics but cases have been found in adult group also although rare[6].Congenital malformations, vascular malformations ,infarctions, infections and vascular occlusions are perinatal causes whereas traumatic birth history, infections, intracranial hemorrhages, tumors, hypoxia are both peri and postnatal causes.

In our case, the cause is most likely a postnatal trauma leading to an intracranial hemorrhage.

Clinically, patients may have seizures, contralateral hemiparesis, facial asymmetry and mental retardation. RI and CT scan are the imaging modalities of choice for diagnosis. RI can clearly differentiate between congenital and acquired types of DDMS. Midline shift of the structures towards the side of the disease is typically seen in congenital form. This helps to differentiate the cerebral atrophy occurring in early life. The atrophied hemisphere will show prominent sulcal spaces in acquired form which occurs after the end of sulcation, as in our case [7].Ipsilateral hypertrophy of calvaria, sinus enlargement, elevations of greater wing of sphenoid, petrous ridge, ipsilateral falcine displacement are common findings in those cases which develops early in life (first 2 years of life) which are the compensatory mechanisms for the hypoplastic cerebral hemisphere [8].Differential diagnosis of DDMS are Sturge weber syndrome, Fishmen syndrome, Rasmussen Encephalitis, Hemimegalencephaly. Rasmussen encephalitis doesn't show any calvarial changes whereas Sturge Weber syndrome is characterized by typical pattern of cortical calcifications and enhancing pial angiomas [9].

Only symptomatic treatment is available for seizures, paresis, hemiplegia and associated disorders. Better prognosis is seen with cases where hemiparesis occurs after 2 years of age and without repetitive seizures.

Hemispherectomy is advised for children with intractable disabling and hemiplegia with a success rate of 85% [10]

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