

Dyke-Davidoff-Masson Syndrome-A Rare Entity. A Case Report of 11 Year Old Male Child

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ABSTRACT

A 11 year old male child presented with recurrent generalized seizures, spastic hemiplegia, microcephaly and had developmental delay in motor and speech domains. CT of the brain revealed characteristic features diagnostic of infantile type of cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome.

Keywords: Dyke-Davidoff-Masson syndrome, hemiatrophy, hemiparesis.

INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) results from hypoplasia of one cerebral hemisphere (hemiatrophy) because of insult in congenital, neonatal or early infantile period to the developing brain. It is characterized clinically by variable degrees of facial asymmetry, contralateral hemiplegia or hemiparesis, mental retardation, recurrent seizures and speech and language disorders. The typical radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses. The syndrome had been documented in mainly adolescents and adults along with children also.^[1,2]

CASE REPORT

A 11-year-old boy was referred for management of recurrent seizures and right hemiparesis. He was born full-term, after a normal delivery in the hospital. At 22 day of life, he developed high grade following which he had seizure activity leading to paralysis of right upper and lower limbs. He subsequently had delayed sitting, standing, and speech milestones and was poor in scholastic performance. He always had 2-3 episode of generalised tonic clonic seizure per year for which no treatment was taken. On examination, he had

microcephaly, relative atrophy of the right half of the body, high arched palate, right-sided torticollis and thoracic scoliosis with convexity toward left [Figure 1]. Neurological examination revealed spastic weakness of right upper and lower limbs with a spastic deformity of the right upper limb and a spastic right hemiplegic gait with brisk tendon reflexes and extensor plantar response, other systemic examination within normal limits. His computerised tomography (CT) showed diffuse atrophy of the left cerebral hemisphere and left half of the cerebral peduncle, pons and medulla with thickening of the overlying calvarium and hyperaeration of the frontal sinuses [Figure 2]. There was also a large ill-defined area showing encephalomalacic changes involving the left temporo-parietal region with loss of adjacent white matter and dilatation of the ipsilateral lateral ventricle with a midline shift of 8mm toward the left side.



Figure 1: Right hemiplegia, spastic posturing of right upper limb, right sided torticollis (a) and scoliosis with convexity toward the left (b)

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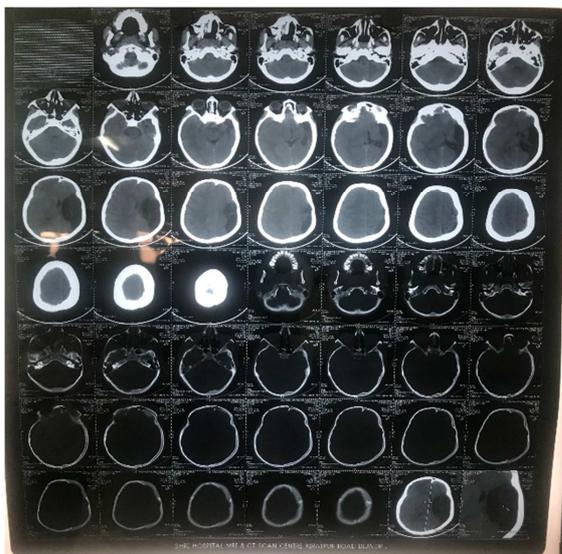


Figure 2: CT showed diffuse atrophy of the left cerebral hemisphere and left half of the cerebral peduncle, pons and medulla with thickening of the overlying calvarium and hyper-aeration of the frontal sinuses. There was also a large ill-defined area showing encephalomalacic changes involving the left temporo-parietal region with loss of adjacent white matter and dilatation of the ipsilateral lateral ventricle with a midline shift of 8mm toward the left side.

Our patient, therefore, had non-progressive infantile hemiplegia, mental retardation and early-onset generalised seizures and CT findings characteristic of the rare syndrome known as DDMS with additional musculoskeletal changes and was started on Sodium Phenytoin.

DISCUSSION

Dyke-Davidoff-Masson syndrome is a syndrome characterized by cerebral hemiatrophy and a contralateral hemiplegia, hemiatrophy, epilepsy and mental retardation with characteristic skull changes.^[3] After the first report of the syndrome given by Dyke, Davidoff and Masson in 1933, Alpers and Dear defined two types of cerebral hemiatrophy in 1939, according to the probable etiology and the time of the insult. In the primary or congenital type, an entire cerebral hemisphere is atrophic or hypoplastic while in the secondary or acquired type there is a localized volume loss of the brain parenchyma.^[1,4] In the primary type, a potential insult occurs in an early pregnancy, causing a development of an entire brain hemisphere to slow down. There is no brain volume loss, but the brain never develops to the normal size, therefore this condition is known as hemihypoplasia or unilateral cerebral hypoplasia.^[5,6] The vast majority of the primary type is of an unknown origin (idiopathic) and the cerebrovascular origin in the early pregnancy is thought to be the most probable cause of the disorder. In the secondary type, cerebral insults occur in the perinatal period or later and a possible

cause could be either cerebrovascular insult, both ischemic or hemorrhagic, inflammatory, infectious process or head trauma. Any of these factors can cause a delay in the development of the brain,^[6] most probably due to reduced perfusion and hypoxic-ischemic injury.

The differential diagnosis for the triad of hemiplegia, hemiatrophy and epilepsy include Rasmussen encephalitis, Sturge-Weber syndrome, hemimegalencephaly, Linear nevus syndrome, Fishman syndrome, Silver-Russell syndrome and unilateral complete occlusion of the middle cerebral artery (MCA) could mimic DDMS on imaging.

CONCLUSION

For DDMS cases presenting in early childhood, refractory seizures remain the usual concern. Accordingly, hemispherectomy is the treatment of choice with a success rate of 85% in selected cases.^[7] Further longitudinal studies are required to ascertain the natural course of this syndrome which would help in planning strategies regarding the time and nature of interventions and management accordingly.

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