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Case Report

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DYKE DAVIDOFF MASSON SYNDROME

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ABSTRACT

Dyke-Davidoff- Masson syndrome (DDMS) is a rare congenital malformation that was first described by Dyke, Davidoff, and Masson in 1933. It is characterised by hemiparesis or hemiplegia, seizures, mental retardation, facial asymmetry. Radiological features of this syndrome includes hemiatrophy of cerebral hemisphere, ipsilateral compensatory hypertrophy of skull and sinuses. Aetiology of cerebral hemiatrophy can be divided into congenital or primary and acquired. Congenital causes can be idiopathic or intrauterine cerebral vascular injury. Acquired causes are birth trauma, perinatal intracranial haemorrhage, Rasmussen encephalitis, infection, Sturge Weber syndrome. DDMS, thus can be congenital or acquired and it is commonly due to vascular insult involving middle cerebral artery. Treatment of DDMS is symptomatic targeting convulsion, hemiparesis, learning difficulties, etc. Here we report a 29 year old male born via full-term uncomplicated normal vaginal delivery to a second degree consanguineously married parents presented to the emergency department with recurrent generalized tonic clonic seizures since 8 months of age.

KEYWORDS: Dyke-Davidoff- Masson syndrome, cerebral hemiatrophy, Rasmussen encephalitis, Sturge Weber syndrome

INTRODUCTION

Dyke-Davidoff- Masson syndrome (DDMS) is a rare congenital malformation that was first described by Dyke, Davidoff, and Masson in 1933. It is characterised by hemiparesis or hemiplegia, seizures, mental retardation, facial asymmetry. Radiological features of this syndrome includes hemiatrophy of cerebral hemisphere, ipsilateral compensatory hypertrophy of skull and sinuses.

CASE REPORT

A 29 year old male born via full-term uncomplicated normal vaginal delivery to a second degree consanguineously married parents presented to the emergency department with recurrent generalized tonic clonic seizures since 8 months of age. He had global developmental delay and developed left sided hemiparesis past 20 years. Patient was on oral phenobarbitone, phenytoin and carbamazepine. There was no history of fever, headache, vomiting, trauma in head and any significant abnormal birth.

There was facial asymmetry with lesser development of left side of face. The muscle bulk of left side of the body was less compared with the right and the limb circumferences of left upper and lower limbs were 3 cm less than the right. All the deep tendon jerks of left side were exaggerated with extensor plantar response. Neuropsychiatric evaluation revealed that the patient had mental retardation. His electrolytes, haemogram, renal and liver functions were normal. Screening for infectious agents were negative.

CT Brain showed subtle atrophic changes involving frontal lobe, high parietal lobe on right side and contralateral cerebellum- probably crossed cerebellar diaschisis/atrophy (figure 1 and figure 2). Hyperpneumatisation of frontal and sphenoid sinuses and subtle calvarial thickening on the right side. Patient was started on oral Levetiracetam and was discharged.

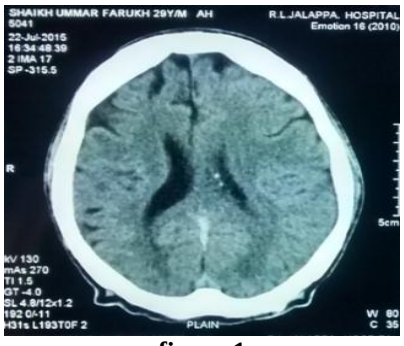

figure 1

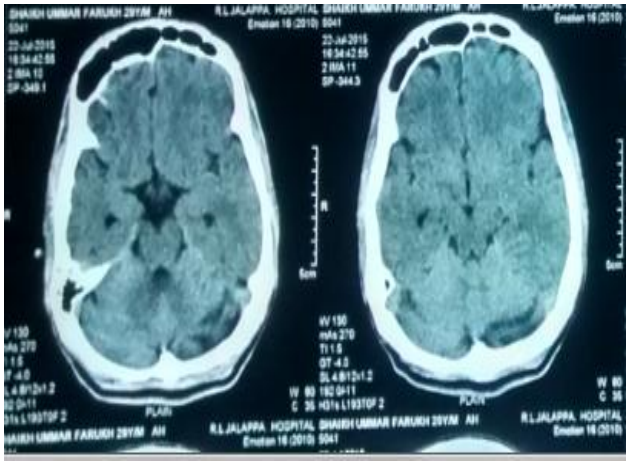

figure 2

DISCUSSION

CT scan or MRI of brain is gold standard investigation for the diagnosis of this syndrome. There is hemiatrophy of brain with compensatory hypertrophy of opposite hemisphere, resulting in midline shift of brain to the affected side. Other features are unilateral thickening of skull, dilatation of ventricle and cisternal space, enlargement of ipsilateral sulci, dilatation of ipsilateral frontal and ethmoid sinuses. Aetiology of cerebral hemiatrophy can be divided into congenital or primary and acquired. Congenital causes can be idiopathic or intrauterine cerebral vascular injury.

Acquired causes are birth trauma, perinatal intracranial haemorrhage, Rasmussen encephalitis, infection, Sturge Weber syndrome. DDMS, thus can be congenital or acquired and it is commonly due to vascular insult involving middle cerebral artery.

Treatment of DDMS is symptomatic targeting convulsion, hemiparesis, learning difficulties, etc. Patients with intractable disability and refractory seizures are candidates for hemispherectomy with 85% success rate.

CONCLUSION

DDMS can be diagnosed by the assessment of patient by complete clinical history and examination along with radiological features on CT/MRI. In patients of recurrent seizures, CT/MRI should be considered as early as possible.

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