Dyke-Davidoff-Masson Syndrome- A Case Report

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Abstract :

Dyke-Davidoff-Masson syndrome was first described in 1933 in series of 9 patients characterized by hemiparesis, seizures, facial asymmetry and mental retardation. It is referred to atrophy or hypoplasia of one cerebral hemisphere which is secondary to brain insult in fetal or early childhood period. Clinically these patients present with seizures, mental retardation, contralateral hemiparesis and facial asymmetry. The treatment is symptomatic and should target convulsions, hemiplegia, hemiparesis and learning difficulties. In case of intractable hemiplegia, hemispherectomy can be considered. Our case a 11 year female presented with complaints of recurrent generalized tonic clonic seizures since 3 years of age, mental retardation and weakness of right side of body. On examination, the child was having mental retardation and hence higher functions could not be assessed. On central nervous system examinations, there was hemiparesis on the right side. MRI brain showed left frontal, temporal, parietal and occipital lobe atrophy with mild prominence of temporal horn of left lateral ventricle and prominence of CSF spaces in vicinity. There was also hyperpneumatisation of left frontal sinus.

Keywords: Dyke Davidoff Masson Syndrome, DDMS

Introduction:

Dyke- Davidoff-Masson syndrome (DDMS) is referred to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy) which is secondary to brain insult in fetal or early childhood period.[(1)] Hemiatrophy is not frequently encountered in Paediatric practice. Clinically these patients present with seizures, mental retardation, contralateral hemiparesis and facial asymmetry. MRI brain shows left frontal, temporal, occipital and parietal lobe atrophy with hyperpneumatised left frontal sinuses.[(2)]

Case Report:

A 11 year female presented with complaints of recurrent abnormal movements of limbs since 3 years of age. She also presented with mental retardation and weakness of right side of body since 3 years of age. The convulsions were generalized, involving all four limbs, associated with up-rolling of eyes and frothing from mouth. There is also history of passing urine and stool during the episode. These lasted for 2-3 min after which the patient remained unconscious for half an hour. There is history of recurrent episodes of such convulsions. She also gives history of weakness of the right half of the body which was progressive in nature. His mother revealed history of developmental delay including gross motor, fine motor, language, social and personal delay. There was no history of similar illness in any other sibling or family member. There is no significant birth history. No history of trauma.

On examination, the patient was conscious with an intelligence quotient of 64 indicating mild mental retardation. On central nervous system examinations, there was hemiparesis on the right side. Neurological examination revealed 4/5 power in the right upper and lower limb with brisk reflexes. Right planter was extensor. There was no any sensory deficit, cranial nerve, or bowel bladder involvement. All routine biochemical investigations including renal and liver function were within normal limits. Random blood sugar, serum electrolytes levels were within normal range. Routine blood investigations were normal. CSF was normal. MRI brain showed left frontal, temporal, parietal and occipital lobe atrophy with mild prominence of temporal horn of left lateral ventricle and prominence of CSF spaces in vicinity. **(Fig 1)** There was also hyperpneumatisation of left frontal sinus.

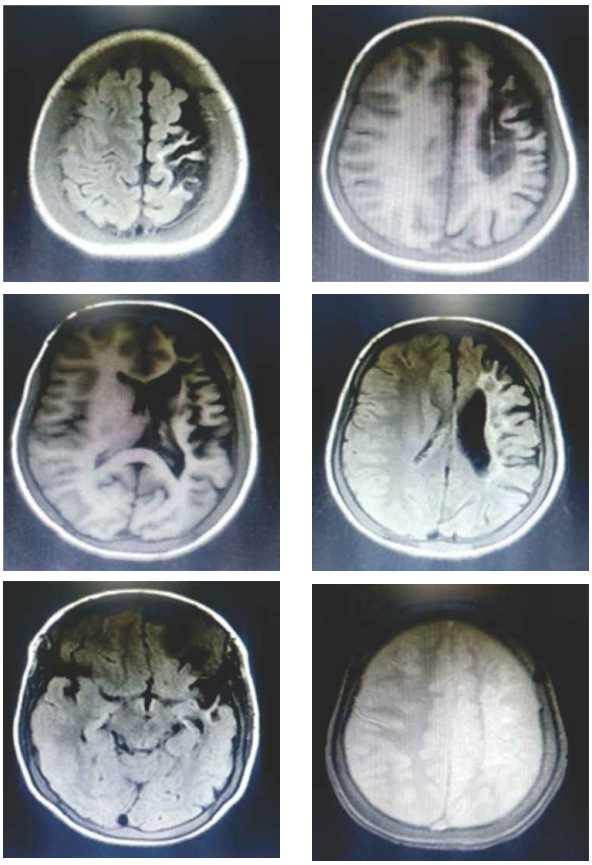

Fig 1 : Magnetic Resonance imaging scan revealing ipsilateral displacement of left falx cerebri, cerebral atrophy and elevation of petrous ridge with hyperpneumatization of left frontal sinus

Discussion:

Dyke-Davidoff-Masson syndrome(DDMS) was first described in 1933 in series of 9 patients characterized by hemiparesis, seizures, facial asymmetry and mental retardation.[3] DDMS is characterized clinically by variable degrees of facial asymmetry, recurrent seizures, contralateral hemiparesis, mental retardation, speech, and language disorders along with various learning disabilities. Seizures can be generalized or focal, however, in this case seizures were generalized. Behavioral problems have also been reported by previous researches as we found disturbed sleep, irritability, anger outburst, increased psychomotor activity, suspiciousness, and irrelevant talks in this case.[4] Causes for DDMS include congenital abnormalities, cerebral infarction, vascular malformations and infections. Perinatal causes include birth asphyxia, hypoxia and intracranial hemorrhage. Cerebral hemiatrophy can develop secondary to cerebral trauma, tumors, infections and prolong febrile seizures after birth.[4] Hageman et al. proposed the terms cerebral hemi-hypoplasia or unilateral cerebral hypoplasia for the congenital cerebral atrophy because there is a lack of cerebral development rather than atrophy.[5] When the insult occurs in-utero, it could be due to gestational vascular occlusion, primarily involving the middle cerebral vascular (MCA) territory. The treatment is symptomatic and should target convulsions, hemiplegia, hemiparesis and learning difficulties. Prognosis is better if the hemiparesis happens after 2 years of age and in absence of recurrent seizures. In case of intractable hemiplegia, hemispherectomy can be considered.[6]

Conclusion:

Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy), which is usually due to an insult to the developing brain in fetal or early childhood period. MRI is diagnostic .showing left frontal, temporal, occipital and parietal lobe atrophy with hyperpneumatised left frontal sinuses. As hemispherectomy is not available even in many urban tertiary care centers, it is very important for a rural neurologist or paediatrician to diagnose the condition early by means of suitable imaging (CT) and the treatment should focus on optimum control of seizures, revision of drug doses from time to time, and domiciliary physiotherapy.

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