

Case Report

Dyke Davidoff Masson Syndrome: A Rare Disease Presented without Motor Weakness

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Abstract

Medical science is full of surprises. We faced one such surprise while treating a 47-year-old male patient with difficulty in walking for 6 months duration along with occasional seizures since childhood. After thorough clinical examination and investigations, patient was diagnosed as a case of Dyke Davidoff Masson syndrome (DDMS). It is a rare syndrome due to hemiatrophy of cerebral hemisphere and associated with hemiplegia or hemiparesis. But in this case, patient had only ankle clonus, causing difficulty in walking without any other motor deficit. Alcohol neurolysis of gastrosoleus muscles was done followed by gait training and improvement of gait was noticed over time.

Key words: Syndrome, hemiatrophy, hemiplegia, clonus, neurolysis, hemispherectomy.

Introduction:

Dyke Davidoff Masson syndrome is characterised by hemiatrophy or hypoplasia of brain as a result of insult of brain during foetal period or in early childhood¹. Dyke, Davidoff and Masson first described the features of this syndrome in skull radiograph in 1933¹. It is manifested with hemiparesis or hemiplegia, seizures, mental retardation, facial asymmetry¹. Radiological features of this syndrome includes hemiatrophy of cerebral hemisphere, ipsilateral compensatory hypertrophy of skull and sinuses^{1,2}. Here we are presenting a case of Dyke Davidoff Masson syndrome without motor weakness.

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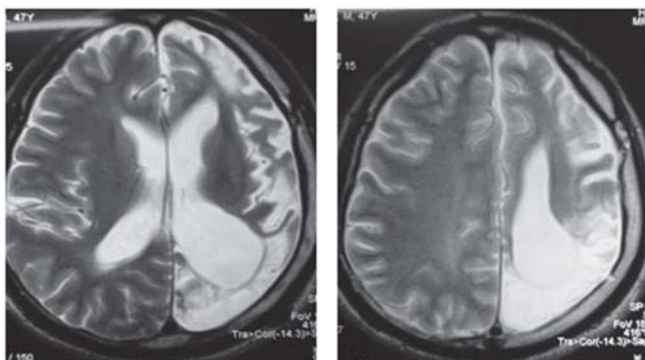
Case Report:

A 47-year-old married male presented with difficulty in walking for 6 months duration. He had also history of recurrent seizures since childhood and he was on carbamazepine. Patient was walking with short stepping gait, bouncing of feet from midstance through preswing phase. Patient was examined thoroughly and no motor weakness was found. Deep tendon reflexes were increased on both sides in upper and lower limbs. Hoffmann's sign was positive on both sides. There was bilateral ankle and right sided patellar clonus. Sensory level was intact. Along with these features, facial asymmetry was also noticed. All routine investigations were found to be within normal limits. MRI of brain (Figs 1-4) showed left cerebral atrophy with dilatation of ipsilateral lateral ventricles and thickening of overlying calvarium. Patient was diagnosed as a case of Dyke Davidoff Masson syndrome. He was treated conservatively and alcohol neurolysis of bilateral gastrosoleus muscles were done to reduce the clonus causing difficulty in walking. Gait training was given to the patient and improvement of gait was noticed over time.

Discussion:

Dyke, Davidoff and Masson first described this syndrome in the year 1933^{1,2}. They described the syndrome through plain radiographic and pneumoencephalographic changes in a series of nine patients¹. Clinically this

syndrome is characterised by hemiparesis, seizures, facial asymmetry, mental retardation, etc. CT scan or MRI of brain is gold standard investigation for the



Figs 1 & 2- Axial T_2 MRI Images Showing Left Cerebral Atrophy with Ipsilateral Dilated Lateral Ventricle and Thickened Calvarium

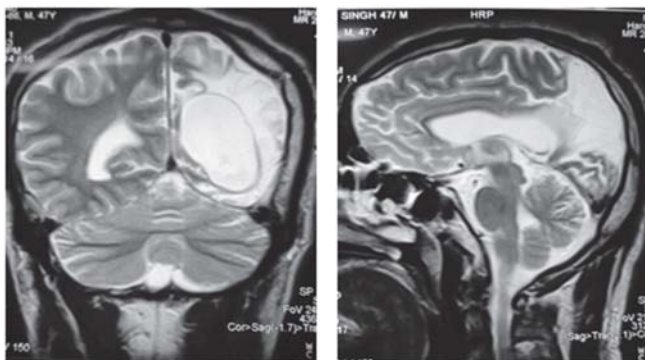


Fig 3- Coronal T_2 MRI Shows Left Cerebral Atrophy with Ipsilateral Ventricular Dilatation **Fig 4-** Sagittal T_2 MRI Shows Ventricular Dilatation and Myelomalacic Occipital Lobe

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^{2,3}. Other features are unilateral thickening of skull, dilatation of ventricle and cisternal space, enlargement of ipsilateral sulci, dilatation of ipsilateral frontal and ethmoid sinuses^{1,4}.

Aetiology of cerebral hemiatrophy can be divided into congenital or primary and acquired^{5,6}. Congenital causes can be idiopathic or intrauterine cerebral vascular injury. Acquired causes are birth trauma, perinatal intracranial haemorrhage, Rasmussen encephalitis, infection, Sturge-

Weber syndrome, etc⁵. Dyke Davidoff Masson syndrome, thus can be congenital or acquired and it is commonly due to vascular insult involving middle cerebral artery^{5,7}.

Brain reaches half of adult size by the age of 1 year and three-fourths of the adult size by 3 years. Therefore, if vascular insult occurs before 3 years of age, there will be compensatory thickening of calvarium and enlargement of sinuses to fill the vacuum created by hypoplastic brain. If the insult occurs in intrauterine life, there will be shifting of midline structures towards the affected side and prominence of sulci replacing absent gliotic tissues. These features can differentiate affection of brain in utero from early life². During diagnosis, other causes of cerebral hemiatrophy should be excluded.

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. Good prognostic indicators are onset of hemiparesis after 2 years of age and absence of recurrent or prolonged seizure².

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