International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Dyke-Davidoff-Masson Syndrome: A Rare Cause for Epilepsy - A Case Report

Dr. Ziddi Linggi

Junior Resident, Department of General Medicine, VIMSAR, Burla, Sambalpur, Pin – 768017, Odisha, India
Mob: 8837478873, 9562267398 Email: *ziddilinggi[at]gmail.com*

Abstract: ***Introduction: Dyke-Davidoff-Masson Syndrome [DDMS} refers to atrophy or hypoplasia of one cerebral hemisphere which is usually due to an insult on a developing brain in fetal or childhood period. Case Details: Here we report a case of 25 years old male presented with multiple episodes of generalized tonic clonic seizure. He was having similar episodes since last 10 years and was on antiepileptic medication irregularly. Patient had history of developmental delay and weakness of right upper and lower limb since early childhood. On physical examination patient had mild weakness in his right upper limb and lower limb and plantar reflex was extensor in right side. Laboratory data revealed no other abnormality. EEG was also normal. MRI of brain revealed left cerebral hemiatrophy and other features suggestive of DDMS. Diagnosis of Dyke-Davidoff-Masson syndrome was made and patient was started on levetiracetam and sodium valproate. Discussion: Dyke-Davidoff-Masson syndrome commonly presents with recurrent seizures, contralateral hemiplegia, facial asymmetry and mental retardation. This case highlights the importance of keeping DDMS as a differential diagnosis while evaluating a patient with seizure.***

Keywords: Dyke-Davidoff-Masson Syndrome, cerebral hemiatrophy, calvarian thickening, osseus hypertrophy, hemispherectomy

1. Introduction

Dyke-Davidoff-Masson Syndrome refers to atrophy or hypoplasia of one cerebral hemisphere which is usually due to an insult on a developing brain in foetal or early childhood period [1, 2].

2. Case Details

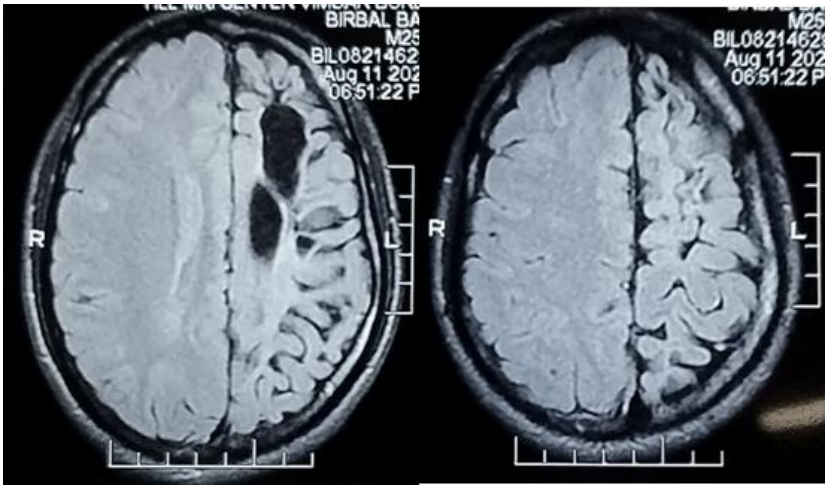
A 25 years old male presented with multiple episodes of generalised tonic-clonic seizure for last 2 days. He had history of multiple similar episodes in the past since he was 15 years old. Patient also had history of developmental delay in the form of delayed walking and speaking and during that time when patient was 2 years old his mother noticed decreased movement in his right upper and lower limb in comparison to his limbs of other side but patient was never evaluated. There was no history of prolonged fever, head injury in the past. There was a history of antenatal trauma when his mother had a blunt abdominal trauma while she was 8 months pregnant with him.

On Examination, patient was conscious and well oriented to time, place and person. His pulse rate was 90 beats per minute and blood pressure was 110/70mm of mercury in right arm supine position. Patient was having mild facial asymmetry but there were no neurocutaneous markers present in patient's body. on neurological examination, patient was left-handed, MMSE score was 20 and he was having a hemiplegic gait. Patient was having hypertonia, power of 4/5 and brisk deep tendon reflexes in his right upper limb and lower limb and there was an extensor plantar response on the right side. Cranial nerves, sensory system and other system examinations revealed no abnormality.

On investigations, all haematological parameters were found to be normal. EEG was done and it was normal.

MRI brain was done which revealed atrophy of left cerebral hemisphere with cystic encephalomalacia, atrophy of left sided brainstem, calvarian thickening on left side, hyperpneumatization of left frontal sinus.

From the above clinical and MRI findings, diagnosis of Dyke-Davidoff-Mason Syndrome was made and patient was treated with sodium valproate and levetiracetam.



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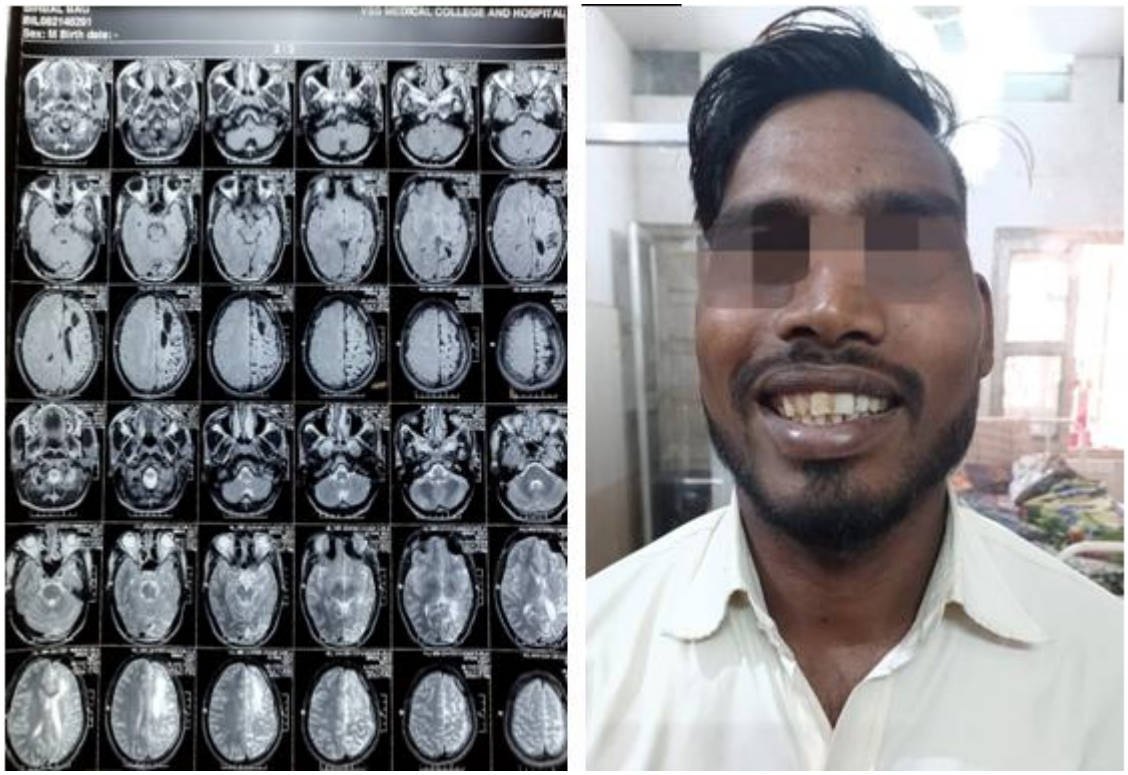

III IV

Figure 1: Image I, II and III is plain MRI brain of the patient showing cerebral hemiatrophy with dilatation of ventricles, prominent sulci. Image IV showing facial asymmetry

3. Discussion

Dyke-Davidoff-Masson Syndrome is a rare entity, which was first described by Dyke, Davidoff and Masson in 1933 in aseries of 9 patients [3]. It is characterised by atrophy of cerebral hemisphere on one side and ipsilateral osseus hypertrophy and hyperpneumatization of frontal and mastoid air cells [4, 5, 6]. The osseus hypertrophy is secondary to brain damage and volume loss. As human brain grows in size in first few years of life it presses the overlying bone outward resulting in increase in head size. Since there is atrophy of cerebral hemisphere in this disease the skull bone gets space to grow inward. The clinical feature of the disease is varied depending upon the extent of brain damage. It includes facial asymmetry, learning disability, contralateral hemiparesis, seizures: both focal and generalised.

It is divided into two types [7]

1) Infantile type: due to neonatal infection, vascular occlusion. Patient develop symptoms in early infancy.
2) Acquired type: due to trauma, tumour, ischemia

Diagnosis is usually obtained by proper clinical history, clinical examination and imaging. Both CT scan and MRI are useful. Imaging findings includes varying degree of cerebral hemiatrophy with dilatation of ipsilateral lateral ventricle with homolateral hypertrophy of skull and sinuses. Petrous ridge elevation and ipsilateral falcine displacement may be present. Atrophy of brainstem and calvarian thickening on the affected side may also be present.

Differential diagnosis includes Sturge weber Syndrome, Silver-Russel Syndrome, Fishman Syndrome and Rasmussen Syndrome [8].

Treatment includes control of seizure with antiseizure medications, physiotherapy, speech therapy. Hemispherectomy is an option for children having irretractable seizures.

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