



## A Case Study of Dyke Davidoff Masson Syndrome

Authors

**Sadaf Sultana<sup>1</sup>, Gotte Priyanka<sup>1</sup>, N. Sai Kumar<sup>1</sup>,  
Dr Thatikonda Anil Kumar<sup>2</sup>, Dr K.Rohith Kumar<sup>3\*</sup>**

Vijaya Neuro Hospitals, Karimnagar, Telangana, India-505001

<sup>1</sup>Pharm D, Department of Pharmacy Practice, Sree Chaitanya Institute of Pharmaceutical Sciences

<sup>2</sup>Neurologist, Vijaya Neuro Hospitals, Karimnagar

<sup>3</sup>Asst Professor, Department of Pharmacy Practice, Sree Chaitanya institute of Pharmaceutical Sciences, Thimmapur, Karimnagar, Telangana, India-505527

\*Corresponding Author

**Dr Kadarla Rohith Kumar**

### Abstract

*A 19 years old female presented with recurrent generalized seizures, right sided hemiplegia, and had developmental delay in motor and speech domains. CT of the brain revealed characteristic features diagnostic of infantile type of cerebral hemi atrophy or Dyke-Davidoff-Masson syndrome. It is an important cause of intractable and drug-resistant seizures. It has varied clinical presentation and history with distinct neuroimaging features. Here, we describe a female patient presented with recurrent intractable convulsion, mental retardation, hemiparesis, dystonia and characteristic neuroimaging features of cerebral hemiatrophy, calvarial thickening, and ipsilateral hyperpneumatization of the frontal sinuses which is suggestive of DDMS. Early institution of neuroimaging in patients with intractable epilepsy will make early diagnosis and better outcome. DDMS also occur due to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy), which is usually due to any trauma or injury to the growing brain in early childhood. The manifestations are variable and depend on the extent of brain injury. More common symptoms are recurrent seizures, facial asymmetry, hemiplegia, mental retardation, mutism. The CT of brain mostly give impression of cerebral hemiatrophy with ipsilateral hypertrophy of the skull and sinuses.*

**Keywords:** *Dyke-Davidoff-Massion Syndrome, hemiparesis, mental retardation, hemiatrophy, hyperpneumatization.*

### Introduction

Dyke-Davidoff-Masson syndrome (DDMS) is a rare disease comprising hemiparesis, seizures, facial asymmetry, and mental retardation<sup>13</sup>. The classical findings including cerebral hemiatrophy along with calvarial thickening and hyperpneumatization of the frontal sinuses are only found if an insult to the brain occurs before 3 years of age<sup>14</sup>. The major concern of the disease

remain the intractable seizures for which drug therapy is not sufficient in most of the cases, and a surgical approach is necessary. However, if the patient presents later in life, the presentation may not be similar to that seen in childhood, and management changes accordingly. Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemi atrophy), which is usually due to an insult

to the developing brain in foetal or early childhood period<sup>1</sup>. The clinical features are variable and depend on the extent of brain injury. More commonly they present with recurrent seizures, facial asymmetry, contra lateral hemiplegia, mental retardation or learning disability, and speech and language disorders. Sensory loss and psychiatric manifestations like schizophrenia had been reported rarely<sup>2,3</sup>. The typical radiological features are cerebral hemi atrophy with ipsilateral compensatory hypertrophy of the skull and sinuses. The syndrome had been documented mainly in adolescents and adults<sup>4,5,6</sup>. However, it can also be seen in children<sup>7</sup>. The human brain reaches half of its adult size during the first year of life and three fourth of the adult size is attained by the end of 3 years. The surface of the hemisphere remains smooth and uninterrupted until early in the fourth month of gestation. By the end of the eighth month, all the important sulci can be recognized. The developing brain presses outward on the bony skull table resulting in gradual increase in head size and shape. When the brain fails to grow properly, the other structures grow inward resulting in increased width of diploic spaces, enlarged sinuses, and elevated orbital roof<sup>9</sup>. These changes can occur only when brain damage is sustained before 3 years of age; however, it may become evident as early as 9 months after the injury<sup>10</sup>.

Cerebral hemiatrophy can be of two types, infantile (congenital) and acquired<sup>11</sup>. The infantile variety results from various aetiologies such as infections, neonatal or gestational vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies, and coarctation of the mid-aortic arch<sup>11,12</sup>. The patient becomes symptomatic in the perinatal period or infancy. The main causes of acquired type are trauma, tumour, infection, ischemia, haemorrhage, and prolonged febrile seizure. Age of presentation depends on time of insult and characteristic changes may be seen only in adolescence or adult. Prognosis is better if the onset of hemiparesis is after 2 years of age and in

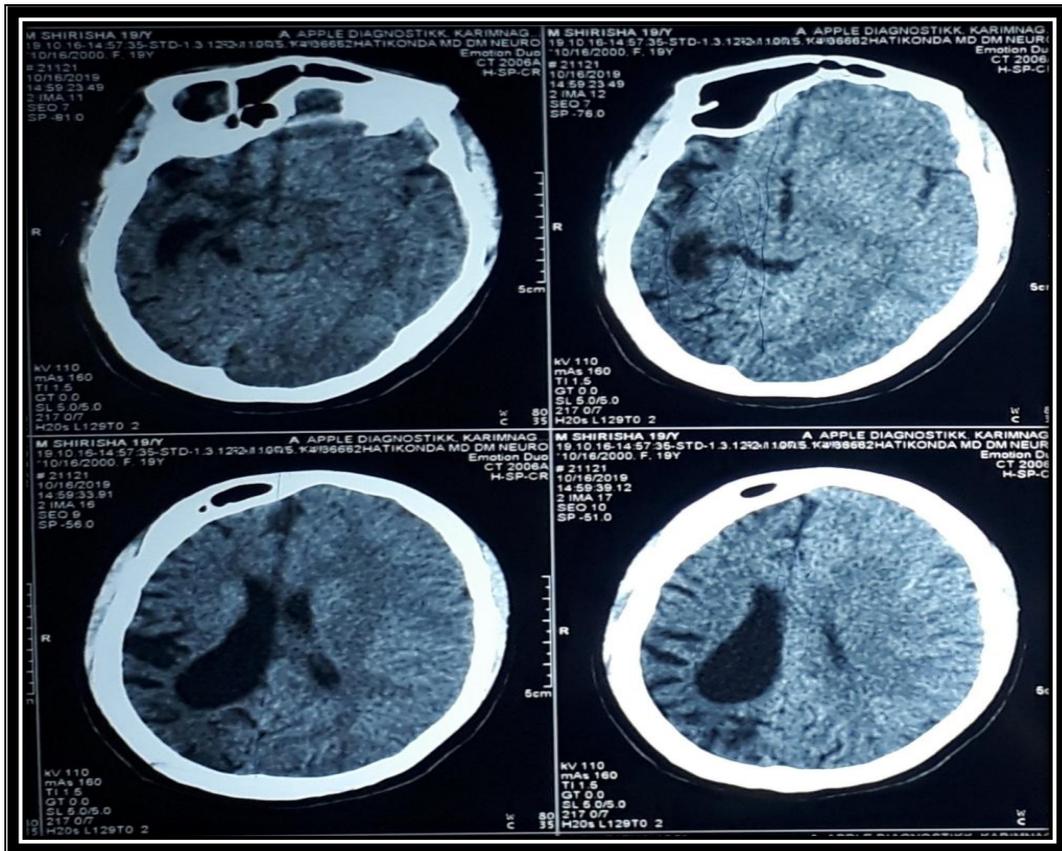
absence of prolonged or recurrent seizure<sup>16</sup>. Hemispherectomy is the treatment of choice for children with intractable disabling seizures and hemiplegia with a success rate of 85% in selected cases<sup>7</sup>.

### Case Report

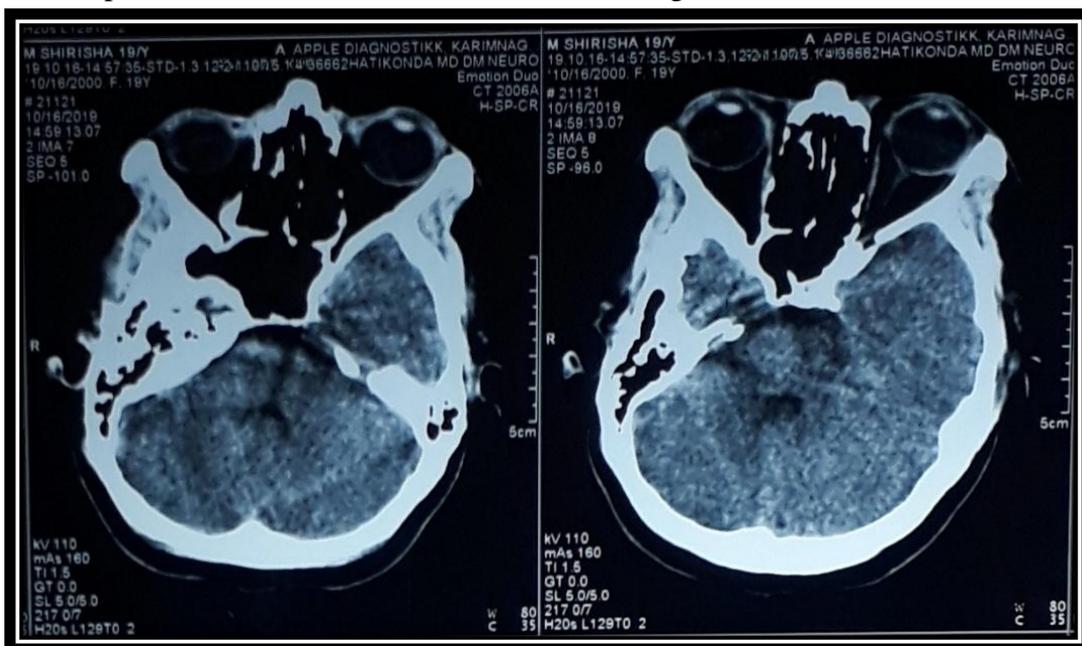
A 19-year-old female presented with multiple types of seizures (tonic, myoclonic), intellectual disability, right sided hemiparesis, facial asymmetry, dystonia for past 17 years. The patient had a history of severe perinatal asphyxia and delayed developmental milestones. She was on valproate and diazepam for past 3 years with very poor compliance and continued to have 3-4 episodes of seizures per day. On examination, the patient had typical spastic hemiplegic gait and posture. She had mild hemiatrophy on the right side and Her IQ score was 40 according to Stanford-Binet test. Her routine hematological and biochemical investigations including serum electrolytes, blood sugar, LFTs, and RFT were normal but her calcium levels were abnormal (6.5). Her serum valproate level was <1 mcg/ml which is markedly below minimum effective concentration. A CT SCAN of the brain was done which revealed sulcal spaces and cisternal spaces are prominent in right cerebral hemisphere. Right lateral ventricle is dilated. Diploic space is thickened in right frontal, parietal, temporal bones. Right cerebral atrophy and with prominent significant impression which revealed as DYKE DAVIDOFF MASSION SYNDROME and gliosis with nearly complete loss of parenchyma in the left cerebral hemisphere with ex vacuo dilatation of ipsilateral lateral ventricle (Fig. 1) on axial plane images. Pulling of interhemispheric fissure on the left side was also seen. Furthermore, enlargement of left sided frontal and mastoid sinuses (Fig. 2). These CT SCAN findings along with the clinical picture were fulfilling the criteria for DDMS, and hence, the diagnosis of DDMS (infantile form) was made. The patient was started on oral valproate and increased up to a dose of 25 mg/kg/day to achieve therapeutic levels, but

seizures continued and levetiracetam added and dosing increased up to 30 mg/kg/day. The patient is kept in follow-up and might need an increment

of doses as she was not completely seizure free before the discharge again in favour of developing refractory seizures as in the case of DDMS.



**Figure 1:** Sulcal spaces and cisternal spaces are prominent in right cerebral hemisphere. Right lateral ventricle is dilated. diploic space is thickened in right frontal, parietal, temporal bones. Right cerebral atrophy and with prominent significant impression which revealed as DYKE DAVIDOFF MASSION SYNDROME and gliosis with nearly complete loss of parenchyma in the left cerebral hemisphere with ex vacuo dilatation of ipsilateral lateral ventricle calvarial thickening



**Figure 2:** Enlargement of left sided frontal and mastoid sinuses

### Discussion

This rare condition derives its name from the researchers Dyke Davidoff and Masson who first reported the condition way back in 1933. Brain imaging reveals right cerebral atrophy which may occasionally be noted in the cerebral peduncles and the thalamic, pontine, crossed cerebellar, and Parahippocampal regions. Brain imaging may additionally reveal prominent sulcal spaces and cisternal spaces are prominent in right cerebral hemisphere. Right lateral ventricle is dilated. Diploic space is thickened in right frontal, parietal, temporal bones. Right cerebral atrophy and with prominent significant impression which revealed as DYKE DAVIDOFF MASSON SYNDROME and gliosis with nearly complete loss of parenchyma in the left cerebral hemisphere with ex vacuo dilatation of ipsilateral lateral ventricle. calvarial thickening, ipsilateral osseous hypertrophy with hyperpneumatization of the sinuses (mainly frontal and mastoid air cells), and an elevated temporal bone. The clinical features include contralateral hemiparesis with an upper motor neuron type of facial palsy, focal or generalized seizures, and mental retardation along with learning disabilities<sup>15</sup>. In our case, the findings of left cerebral hemiatrophy with enlarged cortical sulci, and presentation at the age of 2 years reflect an onset of brain insult after the completion of sulci formation, probably of vascular origin involving left middle cerebral artery.

### Conclusion

The prominent clinical manifestation in DDMS is refractory seizures which should be controlled over a period of time by treating with anticonvulsants. The seizures can be treated either by single or multiple anticonvulsants. Along with the pharmacological therapy, non-pharmacological therapy like physiotherapy, speech therapy plays a significant role. Prognosis is better if the therapy of anticonvulsants continued for 3 years and the onset of hemiparesis is continued with the presence or absence of

seizure. In Children with intractable disabling seizures and hemiplegic, Hemispherectomy is treatment of choice. By the advance CT scan or MRI scan the neurologist can diagnose the condition and the treatment can be focused on control of seizures, and adherence of drugs with the accurate doses from time to time, and domiciliary physiotherapy.

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