Eastern Green Neurosurgery Case Report

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Dyke –Davidoff – Masson Syndrome in a 28 years old pregnant woman – A rare case report.

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ABSTRACT: Dyke -Davidoff- Masson (DDM) Syndrome is rare neurological disorder commonly affecting the children but is rarely reported in adults as well. It mostly presents with seizure, hemiparesis followed by mental retardation and facial asymmetry. We here in report a case of 28 years old female (G3P2A0) with 34+ weeks of pregnancy who presented with generalized tonic clonic seizures and was incidentally diagnosed as DDM. She was managed with an External ventricular drain and antiepileptic medication. This case report aims to draw the attention of health care professionals to keep DDM as a differential in a patient with seizure.

Key words- Antiepileptic, DDM, Hemi atrophy, Seizure

Dyke- Davidoff- Masson (DDM) syndrome also known as cerebral hemi atrophy was first described in 1933. [1] It is a rare neurological entity predominantly seen in childhood which can be congenital or acquired and characterized by hemiparesis, facial asymmetry, seizures, learning disabilities and mental retardation.[2] Radiological imaging including Computed Tomography (CT) and MRI (Magnetic Resonance Imaging) are accurate tools for diagnosis. [3] We hereby report a rare case of 28 years old female with 34 + weeks of pregnancy presented with generalized tonic clonic seizures and was diagnosed as DDM incidentally.

Case Report

A 28 year old female (G3P2A0) with 34+ weeks of pregnancy who presented to the Emergency of Nobel Medical College Teaching Hospital with history of headache which was sudden in onset, severe, associated with dizziness followed by multiple episodes of vomiting. The husband also mentioned of 3 episodes of abnormal

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body movement which was sudden in onset followed by frothy discharge from mouth, twitching of legs and confused state of the patient; each episode lasting for 2 to 3 minutes. She didn't have any previous history of seizure or delayed milestones of development. There was no family history of seizure. Her previous pregnancy was eventful with her first issue was preterm with oligohydramnios and born by a Caesarean Section. Her second pregnancy was uneventful. The baby was healthy and delivered at term. She was intubated in the ER in a view of low Glasgow Coma Scale of E1V1M5. Per abdominal examination revealed a uterus of 34 weeks size, cephalic presentation, fetal heart rate was regular with a baseline 100bpm. On per vaginal examination, os was closed and uneffaced. Neurological status couldn't be assessed as the patient was unresponsive. Routine investigations were sent and all of them were within the normal limits. An ultrasound scan showed single live intrauterine fetus of about 35 weeks of gestation with fetal weight of 2000gms and the amniotic fluid was adequate. CT scan revealed intraventricular hemorrhage with mild hydrocephalus with midline shift with mass effect and cerebral edema. (Figure 1) There were hypodense areas in bilateral periventricular white matter suggestive of chronic ischemic changes. (Figure 1) She was managed with injection levetiracetam 1 gm, 3% NaCl 100ml, paracetamol 1gm and ondansetron 4mg. On the ground of multidisciplinary approach, an external ventricular drain (EVD) was placed by the team of Neurosurgery. (Figure 2) Intraoperatively the pressure of cerebrospinal fluid was high, and it was blood mixed, following which emergency lower uterine caesarean section (LUCS) was performed. There was no Intraoperative complication. A male child weighing 2000gm was delivered with Apgar score of 3/10 at 1st minute of birth and 4/10 at the 5th minute. The child was intubated and advised for Neonatal Intensive care unit admission which was denied by the father and later the baby was shifted to mother side. The patient was shifted to Neurosurgery ICU. The EVD drain was 120ml over 1st 24hrs and her GCS was E4VTM5. A MR Imaging with magnetic resonance angiogram (MRA) with magnetic resonance venogram (MRV) was done on the 2nd day which showed marked atrophy of the left fronto-temporo-parieto-occipital lobe with relative thickening of the skull vault compared to the right side. (Figure 3) No diffuse restriction or blooming or abnormal enhancement was seen. There was ex – vacuo dilatation of the left lateral ventricle suggestive of Dyke- Davidoff Masson syndrome. She was extubated on the 3rd post operative day and the plan is to clamp the EVD on the 5th day followed by a repeat scan.

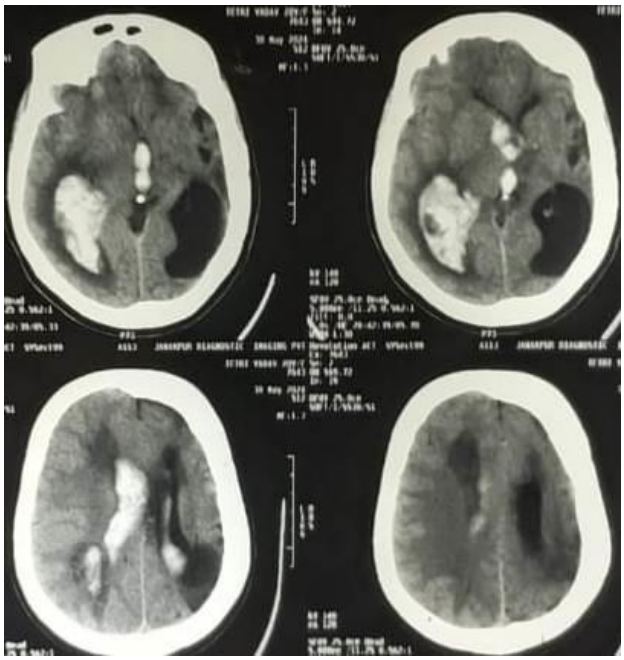

Figure 1: non contrast CT head showing intraventricular hemorrhage with mild hydrocephalus with midline shift with mass effect and cerebral oedema with hypodense areas in bilateral periventricular white matter suggestive of chronic ischemic changes.

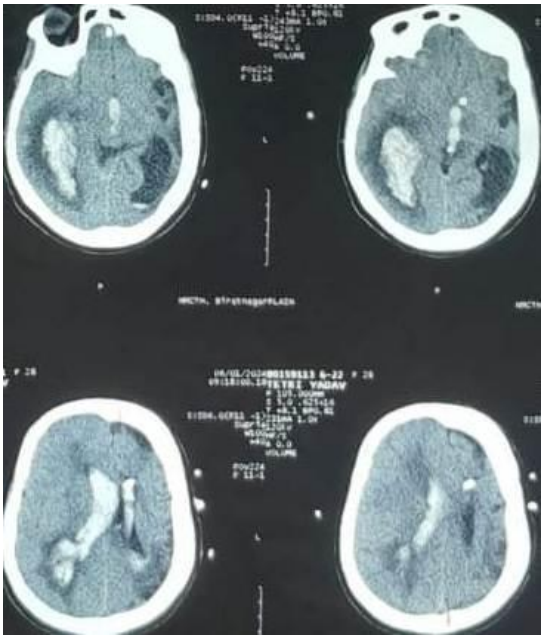

Figure 2: An external ventricular drain (EVD) was placed via left Kocher's point

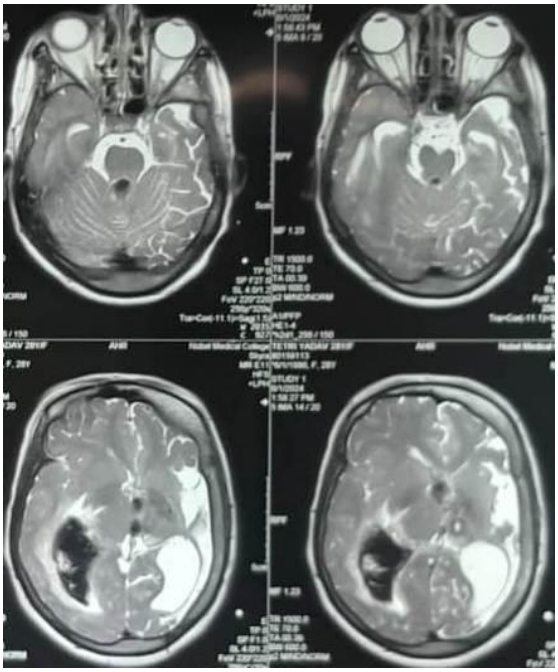

Figure 3: MRI showing marked atrophy of the left fronto-temporo-parieto-occipital lobe with ex – vacuo dilatation of the left lateral ventricle suggestive of Dyke- Davidoff Masson syndrome.

Discussion:

DDM Syndrome is a rare neurological disorder first described by Dyke, Davidoff and Masson in 1933. [1,2] The etiology can be congenital or acquired .[2] It is also known as cerebral hemi atrophy owing to the varying degree of atrophy in cerebral hemispheres accompanied by the compensatory change in calvarium. [3] There are two types of DDM (congenital or acquired. Congenital type presents in early infancy secondary to previous intrauterine brain insults like vascular occlusion or anomaly of the middle cerebral artery. The acquired type occurs later in the childhood secondary to variable causes affecting brain perfusion like infection, prolonged febrile seizure , trauma, hemorrhage and ischemia.[4] Clinical manifestations range from hemiplegia, seizure, facial asymmetry and intellectual disability Seizures can be focal or generalized. However, generalized tonic clonic seizure is the most common presenting feature. [1-3] A precise diagnosis and implementation of suitable management are made possible by imaging via CT and MRI, which is of great importance. Cerebral hemi atrophy, hypoplasia, hyper pneumatization of paranasal sinus and compensatory osseous hypertrophy are relevant imaging characteristics of DDM. [5] Sonali et al reported a similar case of DDM in a pregnant woman who presented with generalized tonic clonic Seizures.[6] There is no definitive guideline for the management of seizure in DDM. [7] Our patient was started on single antiepileptic and she responded well without requiring any dosage adjustment or other additional therapy. For patients especially children with hemiplegia and recurrent Seizures not responding to pharmacotherapy, hemispherectomy is the treatment of the choice.[3,7] In general if patient's diagnosis is incidental or late as in our case and the seizures are well controlled; it is better to avoid surgery. Quality of life can be further improved

with physiotherapy, speech therapy and occupational therapy. [3,7]

Because of its clinical uniqueness and wide range of presentation the prognosis is unclear and depends on how early the disease in diagnosed. [7]

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