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Dyke Davidoff Masson Syndrome- A Rare Case Report

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Abstract:
Dyke-Davidoff-Masson Syndrome (DDMS) is a rare disease in Indian subcontinent. It is more common in western countries. DDMS is characterized by seizures, facial asymmetry, contralateral hemiplegia and mental retardation. Characteristic radiological features are cerebral hemiatrophy with homolateral hypertrophy of skull and sinuses. We report a case of DDMS in a 19 year old girl who presented with seizures, Developmental delay in motor and speech domains, hemiparesis of left upper and lower limb and MRI revealed characteristic features diagnostic of DDMS.

Introduction:
Hemiatrophy of one cerebral hemisphere is not frequently encountered in clinical practice. When this develops early in life (during the first two years), certain cranial changes like homolateral hypertrophy of the skull and sinuses occur. The compensatory cranial changes occur to take up the relative vacuum created by the hypoplastic cerebrum. The classical clinical presentation includes seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation. The clinical findings may be of variable degree according to the extent of the brain injury. Imaging studies show unilateral atrophy of the cerebral hemisphere with ipsilateral shift of the ventricles.[1] The diagnosis of DDMS is based on the correlation between clinical and neuroimaging features. Classic imaging findings are cerebral hypoplasia, ventriculomegaly, paranasal sinus hyper-pneumatization, and compensatory osseous enlargement.[6] The sulci on the involved side are wide and often replaced by gliotic brain tissue. We present here a case of 19-year old girl who presented with seizures and left sided upper motor neuron type of hemiparesis and on magnetic resonance imaging (MRI) scanning was diagnosed to have Dyke Davidoff- Masson Syndrome (DDMS).

Case Report:
A 19 year old female came to emergency in Bhagat phool singh medical college Khanpur kalan, Sonepat, Haryana with complaint of Unconsciousness from 1 day after 2 episodes of abnormal body movements. Patient was asymptomatic 1 day back then she had 1st episode of abnormal body movements. Spasm of hands and legs was present during episode. Froathing was present. Clenching of teeth was present. Tongue bite was present. Spontaneous micturition was present. Patient was unconscious after episode. Patient was taken to some nearby hospital where she had 2nd episode of generalized tonic clonic seizure. Patient was given midazolam to stop GTCS. Patient was given loading dose with antiepileptic drug Levetriacetam 1.5

gm. Then patient was referred to our center for further management. On presentation patient was unconscious. Patient was having history of similar episodes of GTCS from previous 15 years for which she was taking anti-epileptic medications. Patient was non-compliant on medications. She only took medications for 5-6 days after episodes. She was having seizure episodes in every 3-4 months. Frequency of occurance of seizure episodes was increasing from previous 1 year. Patient was mentally retarded from childhood. All the milestones were delayed during childhood. Patient was having slurring of speech, spastic hemiparesis of left side of body, decreased vision from childhood. Patient's antenatal and prenatal period was uneventful. Patient's family history was non-contributory. Patient was having left spastic hemiparesis, brisk tendon reflexes, decreased power with extensor plantar response and right hand preference. But there was no facial asymmetry. She has hemiplegic gait. She was able to walk by herself. Cranial nerve examination was normal. Blood and C.S.F studies were normal. Patient's antenatal and prenatal period was uneventful. MRI Brain showed diffuse hemi-atrophy of right cerebral hemisphere with gross dilatation of ipsilateral lateral ventricle and dilatation of ipsilateral cerebral sulci. Atrophy of right basal ganglia, right thalamus, right hemi-midbrain and right hemi-pons with right crus cerebri atrophy and minimal ipsilateral falcine deviation with atrophy of right half of corpus callosum. Compensatory ipsilateral diffuse clavarial thickening with enlargement of right frontal sinus. Mild atrophy of left cerebellar hemisphere.

Patient was started on antiepileptic medication. Patient's course during hospital was uneventful. Patient was given physiotherapy and speech therapy. Patient didn't have any seizure episode during hospital course. Patient was discharged in stable condition on antiepileptic medication.

Figure 1,2,3 and 4 showing MRI Brain findings- diffuse hemi-atrophy of right cerebral hemisphere with gross dilatation of ipsilateral lateral ventricle and dilatation of ipsilateral cerebral sulci. Atrophy of right basal ganglia, right thalamus, right hemi-midbrain and right hemi-pons with right crus cerebri atrophy and minimal ipsilateral falcine deviation with atrophy of right half of corpus callosum. Compensatory ipsilateral diffuse clavarial thickening with enlargement of right frontal sinus. Mild atrophy of left cerebellar hemisphere.

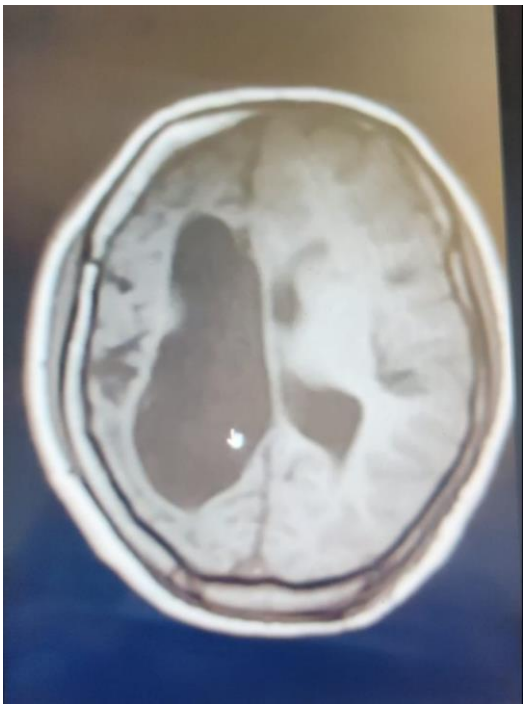

Figure 1

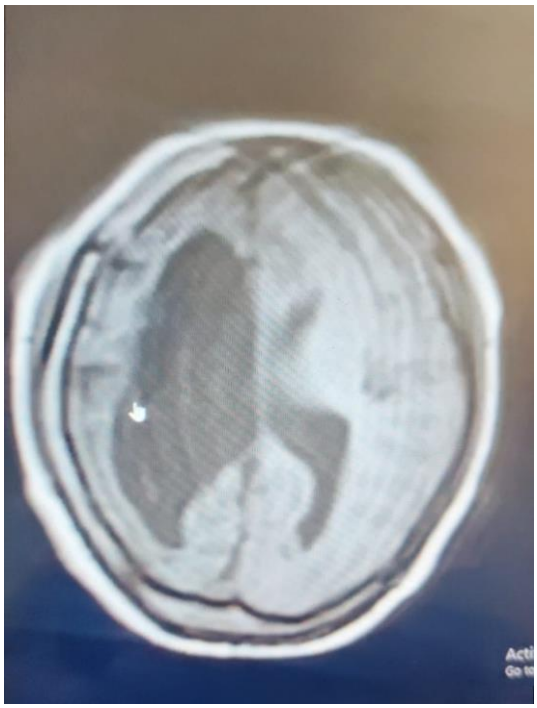

Figure 2

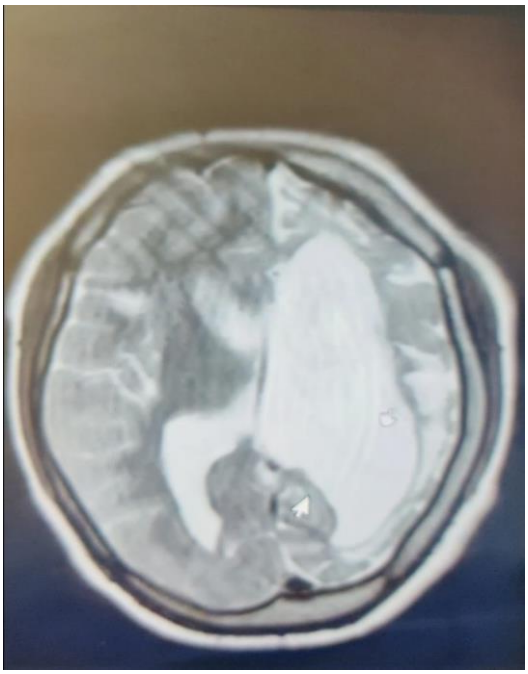

Figure 3

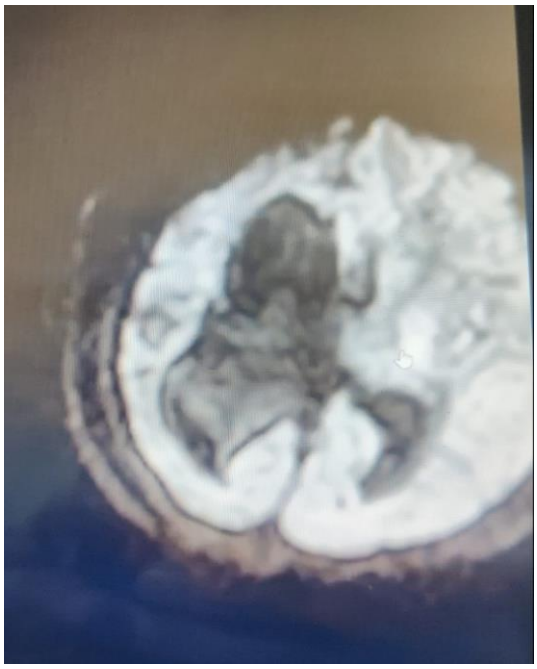

Figure 4

Discussion:

The Dyke-Davidoff-Masson syndrome (DDMS) was initially described as changes in the skull seen on skull X-ray in patients with cerebral hemiatrophy, but is now applied more broadly to cross-sectional imaging also.[2] It is characterized by seizures, hemiplegia or hemiparesis and mental retardation. The condition may be congenital or acquired. In the congenital form, there are usually no evident etiological factors and symptoms present at birth or shortly after. Cerebral damage is likely due to an intrauterine vascular occlusion during the prenatal period. In the acquired form, symptoms are related to a damage of the central nervous system occurring around or after the perinatal period. Some of the etiological factors include trauma, infection, vascular anomalies, hemorrhagic and ischemic conditions, amniotic bands, and an intraventricular and subependymal germinal matrix. The mechanism of cerebral atrophy remains unknown[3]. It is hypothesised that ischemic episode from a variety of different causes reduce the production of brain derived neurotropic factors, which in turn lead to cerebral atrophy. When hemiatrophy of one cerebral hemisphere appears early in life (during first two years of life) certain changes like homolateral hypertrophy of the skull and sinuses occur. The compensatory cranial changes occur to take up the relative vacuum created by the hypoplastic cerebrum. The classical clinical features of DDMS are seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation. However mental retardation was not always present and seizures may appear months or years after the onset of hemiparesis[4]. Differential diagnosis of cerebral hemiatrophy includes Hemiconvulsion-hemiplegia-epilepsy (HHE) syndrome, Rasmussen's encephalitis, Sturge-Weber syndrome, Silver-Russell syndrome, linear nevus sebaceous syndrome, progressive multifocal leukoencephalopathy, and Fishman syndrome. HHE syndrome is characterized by hemiconvulsion seizures, hemiplegia, and epilepsy syndrome in sequence. Hemiplegia can be transient or permanent, and epilepsy can develop after a variable period of 1-3 years, and neuroimaging shows unilateral edematous swelling of the contralateral hemisphere at the time of initial status epilepticus followed by characteristic cerebral hemiatrophy later. Detailed history, thorough clinical examination and appropriate investigation including MRI findings are the key to the diagnosis. The main concern in DDMS is refractory seizures unresponsive to medical therapy; these patients are

candidates for cerebral hemispherectomy, which is helpful in eradicating or significantly reducing seizures in 85% of the patient.[5]

Conclusion:

Seizures in DDMS are often refractory to the medical treatment and surgical treatment may become necessary in these cases. Greater awareness about the disease is required to diagnose the condition timely and refer to neurosurgery in the case of refractory seizures as hemispherectomy is the treatment of choice with high success rate.

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