Dyke Davidoff Masson Syndrome – A Case Report

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Abstract:

A 10 year old male child reported to paediatric neurology OPD of BIRDEM General Hospital with the complaints of weakness of left side of body, seizure and facial asymmetry for 2 years. He was on regular anti convulsive therapy and failed to control seizure. General examination revealed no significant abnormality neither delayed mile stones of development happened. Neurological examination revealed left sided spastic hemiparesis, brisk tendon reflexes and extensor planter on left side. With detailed history and examination he was diagnosed as a case of infantile seizure and undergone CT scan of brain followed by MRI scan. Both the reports revealed severe atrophy of right cerebral hemisphere, thinning of cortical gyri, widening of sulci and dilatation of right lateral ventricle with ipsilateral midline shift and was concluded as hemiatrophy of right cerebral hemisphere with suspicion of Dyke Davidoff Masson Syndrome (DDMS), Hemimegalencephaly and Sturge-Weber syndrome. Dyke-Davidoff-Masson syndrome (DDMS) is a rare condition characterized by asymmetric cerebral hemispheric growth with unilateral atrophy, ipsilateral compensatory osseous hypertrophy, hyperpneumatization of the paranasal sinuses and mastoid cells, and contralateral paresis. Varying degrees of hemiparesis, hemiplegia, seizures, mental retardation, and facial asymmetry can be associated with DDMS. Considering clinical history and imaging findings, final Diagnosis was Dyke Davidoff Masson Syndrome.

Key words: ***Dyke Davidoff Masson Syndrome, CT scan, MRI scan, Hemi atrophy of cerebral hemisphere.***

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Introduction:

Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemi atrophy) which is secondary to brain insult in fetal or early childhood period.[1-3] It is characterized by thickening of the skull vault (compensatory), enlargement of the frontal sinus (also ethmoidal and mastoid air-cells), elevation of the petrous ridge and ipsilateral falcine displacement. In 1993, Dyke, Davidoff, and Masson described the plain skull radiographic and pneumatoencephalographic changes in their series of 9 patients whose clinical characteristics included hemiparesis, seizures, facial-asymmetry and mental retardation.[4] Prenatal causes include congenital anomalies, cerebral infarction, vascular malformations and infections. Insult to premature brain causing loss of neurons and impairment of growth of brain is the main pathology. Postnatal hemiatrophy can develop secondary to cerebral trauma, tumors, infections and febrile seizures. It has been reported that DDMS occur in intrauterine life when the maturation of calvarium has not been completed or due to brain damage (usually traumatic) occurring in first three years of life.[5-8] 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 ipsilateral hypertrophy of the skull and sinuses occur. The compensatory cranial changes occur to take up the relative vacuum created by the hypoplastic cerebrum.Mental retardation was not always present and seizures may appear months or years

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after the onset of hemiparesis.[9-11] The clinical findings may be of variable degree according to the extent of the brain injury. Imaging studies show unilateral loss of volume of brain and calvarial changes, finding of cerebral atrophy, ventricular dilatation and enlargement of sulci.[3, 5,6-11]

Case Report:

A 10-year-old male child hailing from Kustia was admitted to our hospital few months back with the complaints of weakness of left side of body, convulsion and facial asymmetry for 2 years. He was on regular anticonvulsant therapy and failed to control the seizure. He had no feature of mental retardation. There was no history of significant perinatal and antenatal complications. Neurological examination revealed left sided spastic hemiparesis with brisk tendon reflexes and extensor planter response on left side. There was no history of delayed mile stones. With detailed history and examination he was diagnosed as a case of infantile seizure. Then the patient was referred to department of radiology and imaging, BIRDEM for CT scan. Atrophy of the right cerebral hemisphere with sparing of the basal ganglia was revealed in CT scan. Midline structures were shifted by the intact cerebral hemisphere and skull vault thickening and prominent frontal sinus were noted (Fig.-2). CT scan concluded as Hemi atrophy of right cerebral hemisphere. Then the patient reported to Paediatric OPD with CT report and was advised to do MRI scan of brain for better evaluation. MRI of the patient showed severe atrophy of cerebral white matter on right side, thinning of cortical gyri and ex vacuo dilatation of right lateral ventricle (Fig.-1). Atrophy of the right cerebral peduncle and thickening of right sided calvarium was also noted. MRI report concluded as Hemiatrophy of right cerebral hemisphere. In the end considering clinical history and imaging findings this case was diagnosed as a case of Dyke Davidoff Masson Syndrome. Hemimegalencephaly and Sturge-Weber syndrome were considered as other possibilities. In Hemimegalencephaly (Figure no. 3) the involved hemisphere is larger than normal. Addition to be larger, it is also profoundly dysfunctional as the brain area is not developed properly. Sturge-Weber syndrome (Figure no. 4) is characterized by ipsilateral cortical atrophy of cerebral cortex with dystrophic calcifications. The unaffected side has normal cortical development but is abnormally small. As there were dissimilar radiological changes on the affected side of brain and no typical clinical features of Hemimegalencephaly and Sturge-Weber syndrome were observed, the final diagnosis was of Dyke Davidoff Masson Syndrome.

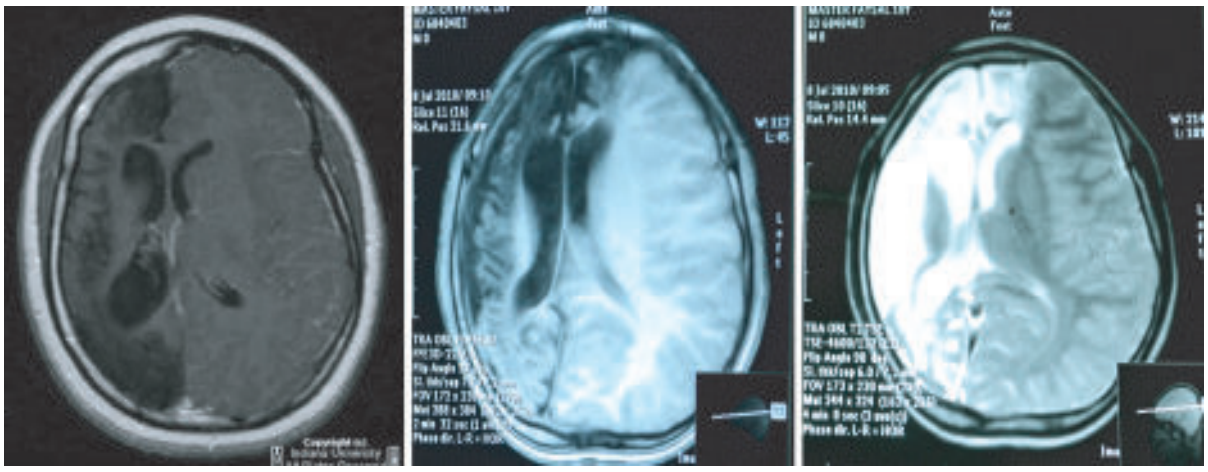

Fig.-**1:** *Multiple sequences of MRI (axial) image severe atrophy of cerebral white matter on right side, thinning of cortical gyri and ex vacuo dilatation of right lateral ventricle.*

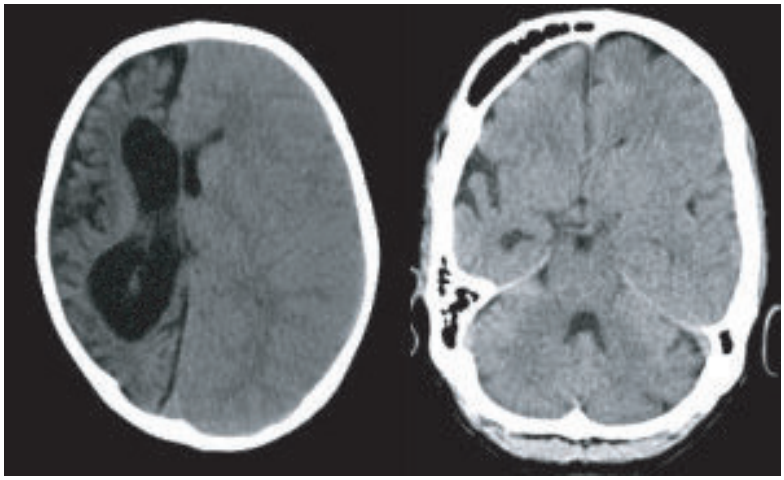

Fig.-2: CT scan of brain showing atrophy of the right cerebral hemisphere.

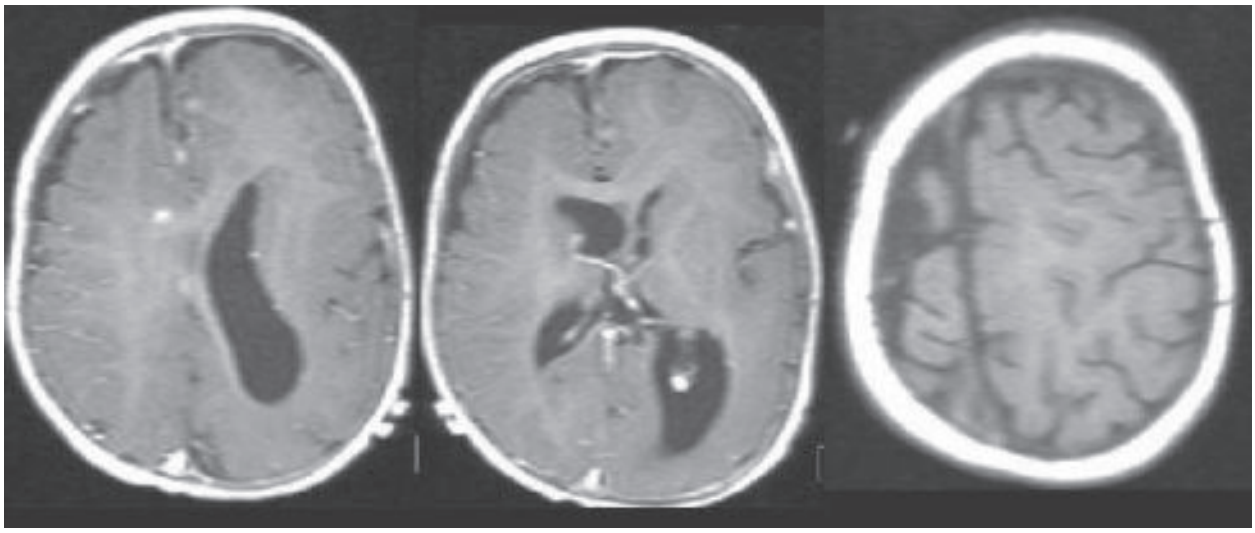

Fig.-3: *T1WI MRI images of a case of Hemimegalencephaly.*

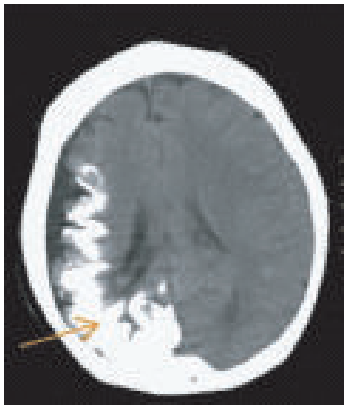

Fig.-4 : *Non contrast CT scan of a case of Sturge-Weber syndrome with intra cranial calcifications.*

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. It is a rare disease which is characterized by thickening of the skull vault (compensatory), enlargement of the frontal sinus (also ethmoidal and mastoid air-cells), elevation of the petrous ridge and ipsilateral falcine displacement.[1-4] Some authors reported as a rare clinical syndrome consisting of hemiatrophy of cerebral hemisphere, enlargement of ventricles and prominent pneumatization of petrous pyramid of the temporal bone and its clinical

manifestations are hemiparesis, several types of seizure, mental retardation and other neurologic deficits. In our study subject, there was seizure, no mental retardation, left sided upper motor neuron type spastic hemiparesis. These findings is very much comparable with the previous reported cases.[2, 4-6] Dyke-Davidoff-Masson syndrome associated with crossed cerebellar atrophy (CCA) has been previously described [7,8] but this was not observed in our case. Imaging findings of both CT scan and MRI were similar to the previous reported cases of DDMS[4-11] which was evident by severe atrophy of right cerebral hemisphere, thinning of cortical gyri, widened sulci and dilated right lateral ventricle with ipsilateral midline shift.

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