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RESEARCH ARTICLE

Dyke-Davidoff-Masson Syndrome: A Rare Cause of Refractory Epilepsy with Cerebral Infarction.

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Abstract

Refractory epilepsy is not very uncommon in recent time. Dyke-Davidoff-Mason syndrome is one of the rare causes of refractory epilepsy. Here is a report of this rare case we have come across during treatment of refractory epilepsy in a 30 yr old man who presented with Generalised Tonic Clonic Seizures, Hemiparesis of the left hand and leg with deformity of the right upper limb and right lower limb and also deviation of the mouth to left. The **Dyke-Davidoff-Masson syndrome (DDMS)** was initially described as changes in the skull seen on skull X-ray in patients with cerebral hemi-atrophy, but is now applied more broadly to cross-sectional imaging also

INTRODUCTION

The **Dyke-Davidoff-Masson syndrome (DDMS)** was initially described as changes in the skull seen on skull X-ray in patients with cerebral hemi-atrophy, but is now applied more broadly to cross-sectional imaging also. It is characterised by thickening of the skull vault (compensatory), enlargement of the frontal sinus (also ethmoidal and mastoid air-cells), elevation of the petrous ridge , ipsilateral falcine displacement, Capillary malformations(are a novel finding for children with Dyke-Davidoff-Masson syndrome)[7]. In some sources it is equated to hemispheric infarction, whereas in other sources any cause of cerebral hemi-atrophy are included. It is characterised by refractory seizures, facial asymmetry, contra-lateral hemi-paresis and sometimes mental retardation. It is also associated with learning disability, speech and language disorders. Sensory loss and psychiatric manifestations like schizophrenia has been reported rarely [1] [2]. Differential diagnosis: General imaging differential considerations include-hemimegalencephaly, Sturge-Weber syndrome - can also be an association, Rasmussen encephalitis - tends not to have calvarial changes. Dyke-Davidoff and Masson described the plain skull radio-graphical features of Dyke-Davidoff-Masson Syndrome (DDMS) in 1933 [3] . Since then many cases have been reported in child adolescent and adult age group. Here we are describing a case of this rare syndrome in a 30 yr old male.

CASE REPORT: A 30 yr old male admitted to our hospital with features of generalised tonic-clonic seizure with a history of epilepsy since his 15 yrs of age. Despite on being medication (phenytoin and valproate) for last 15 yrs his seizure frequency has been increasing for last 6 months. He had no recent epileptogenic factors in recent past. On examination patient was found to be of subnormal intelligence. Plantar reflex on right side was extensor and knee and ankle jerk was brisk. Motor power was 3/5 in both upper and lower right limb and no sensory deficit. His routine blood and biochemistry report was within normal limits. He had a history of seizure during birth but developmental milestones achieved in time. He experienced first attack of seizure at 15 yrs of age along with right

sided hemi-paresis and loss of speech for 2 days. He was admitted to hospital and got cured of it. CT brain at that time revealed non-enhancing hypo density in left fronto parietal white matter suggestive of post ictal cerebral oedema. Since then he has been experiencing seizure episodes. His recent NCCT brain revealed old infarct with gliotic reaction seen in left basal ganglia, Para-ventricular region and at supra-ventricular left parietal lobe associated with shrinkage of left cerebral hemisphere. MRI brain revealed hemi-atrophy of left cerebral hemisphere (Fig 1) .Skull x ray revealed thickening of calvarial bones.

Consent from the patient was taken for publication of the case. I am the submitting author of this case report.

DISCUSSION:

In 1933, Dyke, Davidoff, and Masson described the plain skull radiographic and pneumato-encephalographic changes in a series of nine patients characterized clinically by hemi-paresis, seizures, facial-asymmetry, and mental retardation.

Cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome is a condition characterized by seizures, facial asymmetry, contralateral hemiplegia or hemiparesis, and learning difficulties. These findings are due to cerebral injury that may occur early in life or in utero. Insult to immature brain results in neuronal loss and impaired brain growth. In congenital hemiatrophy, when the insult occurs in utero, there is shift of midline structures towards the side of the disease and the sulcal prominence replacing the gliotic tissue is absent [14]. This feature differentiates it from cerebral hemiatrophy which occurs in early life. The etiological factor for Dyke-Davidoff-Masson syndrome has been postulated as trauma, inflammation or vascular malformations and occlusions. When the insult occurs in-utero, it could be due to gestational vascular occlusion, primarily involving the middle cerebral vascular territory. The causes in the perinatal period birth trauma, anoxia, hypoxia and intracranial haemorrhage. Postnatal causes are trauma, tumor, infection and prolonged febrile seizures.The radiological features are unilateral loss of cerebral volume and associated compensatory bone alterations in the calvarium, such as thickening, hyperpneumatization of the paranasal sinuses and mastoid cells and elevation of the petrous ridge.

In this case we have come across same findings in this 30 yr old male presented to us with status epilepticus. He had generalized tonic-clonic seizures since childhood, which were refractory to the titrating doses of anti-epileptics. He also had unilateral pyramidal signs and mental retardation on examination. The CT images and MRI images showed the features of cerebral hemi atrophy, calvarial thickening ipsilateral sinus enlargement both ventricular enlargement. and sulcal prominence, which is characteristic of the Dyke-Davidoff-Masson syndrome [4] Although computed tomography (CT) and MR are complimentary, it is felt that MR represents the imaging procedure of choice with respect to the assessment of the aetiology and extent of cerebral parenchymal involvement in patients presenting with a clinical combination of congenital or early onset of seizures, hemiparesis/plegia, and/or craniofacial asymmetry.

In 1939, Alpers and Dear defined two types of cerebral hemiatrophy [5]. In the primary (congenital) type, the entire cerebral hemisphere is characteristically hypoplastic. The secondary type results from a cerebrovascular lesion, inflammatory process, or cranial trauma. Twenty-two cases of primary variety were collected from the literature until 1939. A clinical triad of hemiplegia, seizures and mental retardation was defined. However mental retardation was not always present and seizures may appear months or years after the onset of hemiparesis. [6]

Dyke Davidoff Masson Syndrome should be differentiated from Basal cell germinoma, Sturge Weber syndrome, Linear Nevus syndrome, Fishman syndrome, Silver- Russell syndrome and Rasmussen encephalitis [15]. A proper clinical history and CT/MRI findings provide the correct diagnosis

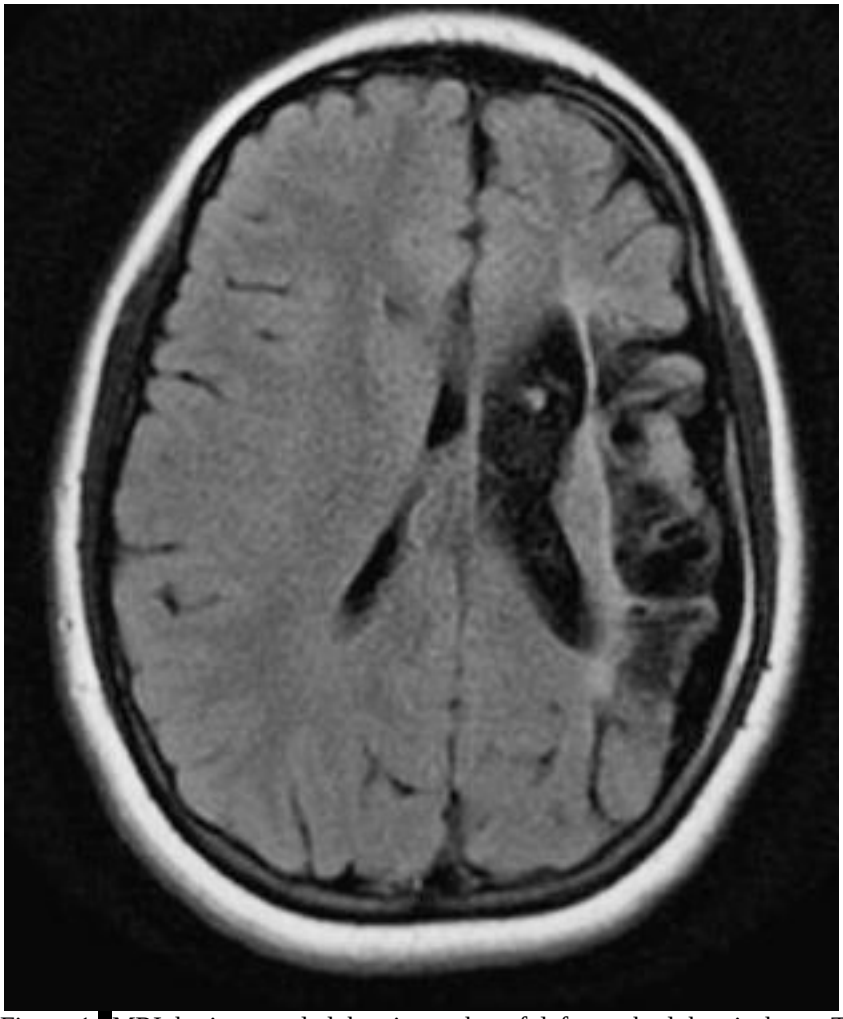

Figure-1. MRI brain revealed hemi-atrophy of left cerebral hemisphere. There is obvious atrophy of the left hemisphere, in the temporal and parietal lobes with enlargement of the adjacent subarachnoid spaces, frontal, temporal horns of the left lateral ventricle

The treatment is symptomatic, and should target convulsion, hemiplegia, hemiparesis and learning difficulties. Prognosis is better if hemiparesis occurs after the age of 2 years and in absence of prolonged or recurrent seizures. Children with intractable disabling and hemiplegia are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases [16].

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