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Case Report

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 year old man.

Keywords: DDM (Dyke-Davidoff-Masson), Epilepsy, Cerebral infarction

INTRODUCTION

Health 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 characterised by refractory seizures, facial asymmetry, contralateral hemiparesis 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] Dyke-Davidoff and Masson described the plain skull radiographical 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 year old male.

CASE REPORT

A 30 year old male admitted to our hospital with features of generalised tonic clonic seizure with a history of epilepsy since his 15 years of age. Despite on being medication (phenytoin and valproate) for last 15 years his seizure frequency has been increasing for last 6 months. He had no such 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 years of age along with right sided hemiparesis and loss of speech for 2 days. He was admitted to hospital and got cured of it. CT brain at that time revealed nonenhancing hypodensity 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, paraventricular region and at supraventricular left parietal lobe associated with shrinkage of left cerebral hemisphere. MRI brain revealed hemiatrophy of left cerebral hemisphere (Figure 1). Skull X-ray revealed thickening of calvarial bones.

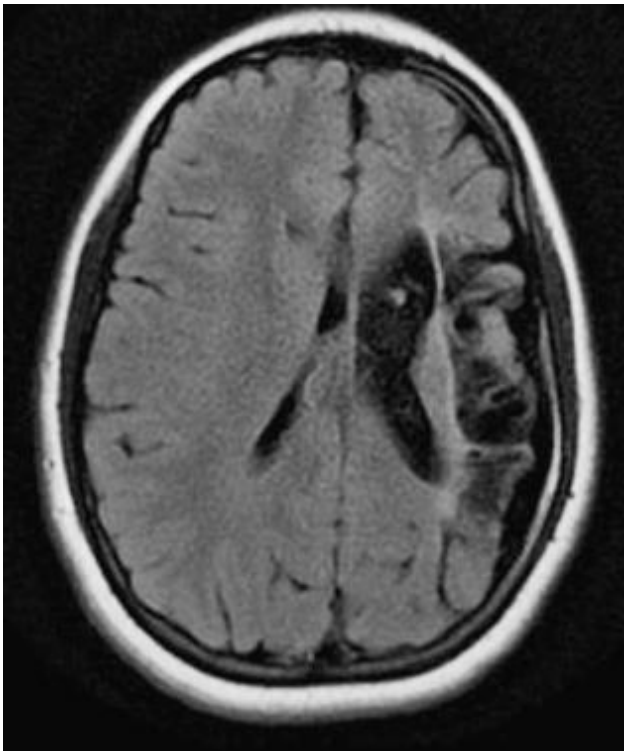

Figure 1: MRI brain revealed hemiatrophy of left cerebral hemisphere.

DISCUSSION

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

In this case we have come across same findings in this 30 year 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]

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]

Other differential diagnosis to be considered in a patient of cerebral hemiatrophy are Sturge-Weber syndrome, some brain tumours, Silver Syndrome, as well as conditions, that are associated with unilateral megaloencephaly as in the linear-nevus syndrome. A proper clinical history and CT findings provide the correct diagnosis.

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