Case Report

Medicine Section

Dyke-Davidoff-Masson Syndrome: An Uncommon Cause of Refractory Epilepsy Identifyied by Neuro-Imaging

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ABSTRACT

Refractory epilepsy is common in practice of neurology. Dyke-Davidoff-Masson Syndrome (DDMS) is one among the syndromes associated with refractory epilepsy. We have come across a case of Dyke-Davidoff-Masson syndrome while investigating a case of refractory epilepsy. When cerebral hemi atrophy is associated with the radiological features of osseous hypertrophy of calvarium, hyper-pneumatisation of sinuses dilatation, DDMS is to be considered.

Key Words: Dyke-Davidoff-Masson syndrome, Refractory epilepsy, DDMS, Cerebral hemi-atrophy

INTRODUCTION

Refractory epilepsy is common in practice of neurology. Dyke-Davidoff-Masson Syndrome (DDMS) is one among the syndromes which are associated with refractory epilepsy. Dyke-Davidoff and Masson described the plain skull radiographical features of Dyke-Davidoff-Masson Syndrome (DDMS) in 1933 [1]. This was described in a series of 9 patients with hemiparesis, seizures, facial asymmetry and mental retardation. The radiographical features of the skull were asymmetry, ipsilateral osseous hypertrophy of the calvarium and hyper-pneumatization of the sinuses. There were many adult and paediatric case reports in the literature since then. We are describing here, the clinical and radiological features of DDMS in a patient with refractory epilepsy.

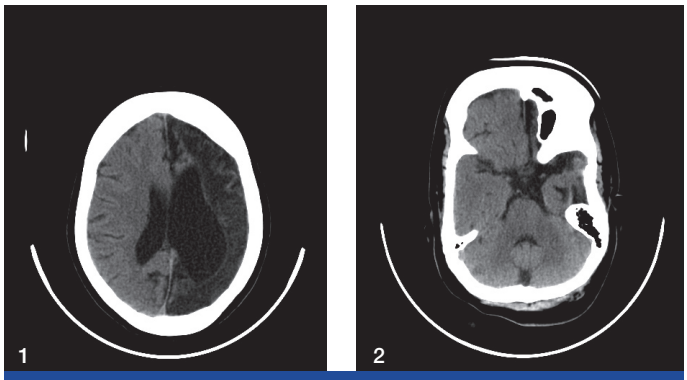
CASE REPORT

A 26 year old mentally retarded woman with a history of epilepsy from her eighth month of life, presented to us with increasing numbers of generalized tonic – clonic seizures of about three weeks of duration. She was on oral carbamazepine and phenobarbitone at escalating doses since childhood. Of late, she was started on magnesium valproate for the control of the seizures. She had no new precipitating epileptogenic factors in the recent past. She had paucity of movements in her right sided limbs since birth and had been having recurrent attacks of sinusitis since childhood. In her antenatal history, it was reported that her mother had febrile illness in the first trimester of gestation. On examination, she was found to be conscious and mentally retarded and had no phakomatosis or signs of meningial irritation. She had facial asymmetry, right sided hypertonia, exaggerated reflexes and an extensor plantar reflex on the right side. Her routine haematological and biochemical investigations were within normal limits. The non-enhanced CT images of her brain showed left cerebral hemispheric atrophy with left lateral ventricular dilatation and diffuse white matter hypodensities with a midline falx. Enlargement of the sinuses and thickening of the calvarial bones and sphenoid were also seen on the left side [Table/Fig-1 & 2]. These features are suggestive of DDMS.

DISCUSSION

There is a wide range of anti-epileptics which are available for clinical use. Most of these drugs are known for their adversities. These drugs are often too expensive for an economically backward patient of the sub continent. Thus, the control of refractory epilepsy at large is a challenge in the sub continent and in other developing countries. Identification of the cause for epilepsy is essential in planning management. There are various tests which have been described to investigate epilepsy. Neuro-imaging is one of the main tools used in the investigation of epilepsy. There are many syndromes which are associated with refractory epilepsies and some of them are identified by their imaging characteristics [1].

We came across this case of Dyke-Davidoff-Masson syndrome while investigating the patient for refractory epilepsy. She had generalised tonic-clonic seizures since childhood, which were refractory to the titrating doses of anti-epileptics. She had unilateral pyramidal signs and mental retardation on examination. The CT images showed the features of cerebral hemi atrophy, calvarial thickening ipsilateral sinus enlargement and either ipsilateral ventricular enlargement and sulcal prominence, which were characteristic of the Dyke-Davidoff-Masson syndrome [2].



[Table/Fig-1 & 2]: Non-enhanced CT images of the brain show left cerebral hemispheric atrophy with left lateral ventricular dilatation and diffuse white matter hypo densities. The falx is seen in the midline and there is thickening of calvarium and sphenoid bone.

Hemiatrophy of one cerebral hemisphere is not frequently encountered in clinical practice. The classic presentation of cerebral hemiatrophy was found with refractory seizures, mental retardation, contralateral hemiplegia or hemiparesis and facial asymmetry [2].

When the clinical features of cerebral hemiatrophy are associated with the radiological features of cerebral atrophy, osseous hypertrophy of the calvarium and hyper-pneumatisation of the sinuses dilatation, DDMS has to be considered [3, 4]. The pathogenesis of DDMS is thought to originate from a childhood cerebral insult due to trauma, inflammation or vascular malformations/occlusions of the middle cerebral artery [4, 5]. In this case, the patient's mother had a febrile episode in the first trimester of gestation. This insults occurring early in the first two years, the compensatory cranial changes like calvarial thickening and sinus enlargement occurs due to a relative vacuum created by the hypo plastic brain [2, 6]. In 2008, Karuppiah S has reported a case of DDMS as a complication of cerebral malaria in an 18 year old lady [7].

Cerebral hemiatrophy with refractory seizures could be due to the Sturge-Weber Syndrome, brain tumours, Rasmussen's encephalitis or the Silver Syndrome. Assessment of the complete clinical history and examination along with radiological features can only provide the diagnosis of the Dyke-Davidoff-Masson Syndrome. The treatment of DDMS with multiple anti-epileptics is the best option. If the seizures are refractory, hemi-spherectomy is the best treatment option.

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