Dyke-Davidoff-Masson Syndrome

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Dyke-Davidoff-Masson syndrome is a rare cause of refractory epilepsy was detected in 18 years old male patient who presented with complaints of generalised tonic-clonic seizures, since the age of 2-3 months, inability to move left half of the body since 5-6 months of age, history of mental retardation and cerebral palsy along with speech problem. MRI brain imaging showed diffuse atrophic changes in right cerebral hemisphere with ex-vacuo dilatation of lateral ventricle (Figure 1).1

Patients all routine investigations were normal. EEG record showed localised seizure discharge from left frontal region. Hyperventilation and photic provocation produced no added information.

DDMS is a rare condition with variable degree of facial asymmetry, contralateral hemiparesis/hemiplegia, seizure, mental retardation, radiological finding of asymmetry of cerebral hemispheric growth with atrophy on one side, ipsilateral osseous hypertrophy and hyperpneumatization of sinuses (Figure 2).

The treatment of DDMS is symptomatic and should target convulsions, hemiparesis/hemiplegia and learning difficulties. For treatment multiple anti-epileptics is the best option. Children with intractable disabling seizures and hemiplegia are the potential candidates for hemispherectomy with success rate of 85% in the selected cases. Prognosis is better if hemiparesis occurs after the age of 2 years and in the absence of prolonged or recurrent seizures.1,2

References