Dyke-Davidoff-Masson Syndrome

N Nand¹, S Venu², M Yadav², AR Deshmukh², R Mittal²

A 23 year man presented with focal seizures and weakness of right half of body since last 8 years. He had intractable seizures despite regular drug intake (Phenytoin 300 mg hs). Antenatal and perinatal period were uneventful. He had normal developmental milestones, but had below normal intelligence. There was no history of similar illness in family members. On examination, patient was conscious, mentally retarded (intelligence quotient of 45) and had no phakomatosis. He had facial asymmetry (Figure 1), right sided weakness with a power of 4/5, hypertonia, exaggerated reflexes and an extensor plantar reflex on the right side.

On investigations, complete hemogram, liver and renal function tests were normal. Non-contrast computerized tomography (NCCT) of head showed left sided cerebral hemisphere atrophy with ipsilateral dilatation of lateral ventricle with ipsilateral hyperpneumatization of frontal sinus and prominent sulci.

Based on characteristic radiological findings, patient was diagnosed as a case of Dyke–Davidoff–Masson syndrome (DDMS)¹ and treated with Valproic acid and Levetiracetam.

DDMS, first reported in 1933² is a rare condition and refers to atrophy or hypoplasia of one cerebral hemisphere, characterized clinically by facial asymmetry, recurrent seizures, contralateral hemiparesis, mental retardation, speech and language disorders along with various learning disabilities. The etiology is cerebral insult, which may occur in the prenatal, perinatal or post natal period. In DDMS, as brain fails to grow on one side, a vacuum is created due to which other structures tend to direct their growth inward, thus accounting for its characteristic radiological features.³ Treatment is only symptomatic, including management of seizures. Hemispherectomy is preferred for children with intractable disabling seizures and hemiplegia.

We aim to highlight this case for its rarity.

References