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DYKE DAVIDOFF MASSON SYNDROME

General Medicine

Dr Mital Rathod 2nd Year Medicine Resident, P.D.U Civil Hospital Rajkot
Dr Praful Dudharejiya Professor & Head of unit – Dept. of General Medicine

ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological disorder that results from brain injury in intrauterine or early years of life. Prominent cortical sulci, dilated lateral ventricles, cerebral hemiatrophy, hyperpneumatization of the frontal sinus, and compensatory hypertrophy of the skull are the characteristic findings. We describe a male patient who presented with generalized tonic- clonic seizure and left-sided body weakness and neuroimaging findings of cerebral hemiatrophy, dilatation of right lateral ventricle, right frontal sinus hyper pneumatization, and asymmetric calvarial thickening. Knowledge of its features on imaging enables timely and accurate diagnosis, allowing appropriate management.

KEYWORDS

An 21 year old male child presented with recurrent focal (left-sided) seizures from 26 months of age. He had history of sudden onset left-sided complete hemiplegia since a week without any history of fever or altered sensorium with behaviour changes he had delayed developmental milestones (motor and language).

Patient initially had focal seizures, tonic-clonic in type but later on they become focal to bilateral tonic-clonic with impaired awareness.

Patient was on anti-epileptic treatment (Valproate and Phenobarbitone), compliance was poor, seizures were recurrent.

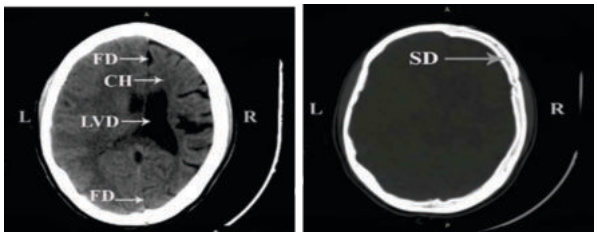
Neurological examination:
- Higher mental functions: conscious & non-oriented,
- Decrease cognitive functions
- Complete left-sided hemiparesis
- Exaggerated reflexes on left side
- Plantar extensor reflex on left side

Investigations
- **Haematological Profile**

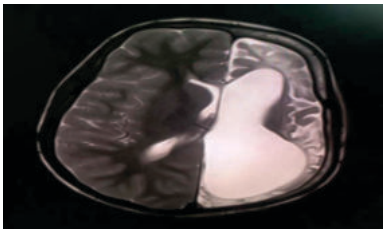
Hb: 12.0, TLC: 12,800, DLC: P75 L21 M03 E01 B00
PCV: 38.0%, Platelets: 2.8lacs/cumm
- **Cerebrospinal fluid examination**

Colour: Clear, Total Cells: 18 cells/cumm, Only lymphocytes seen
Sugar: 87 mg/dl, Protein: 51 mg/dl
- **Biochemistry**

Na: 151.79 meq/l, K: 4.86 meq/l, Ca: 8.9 mg/dl
- **EEG** S/O generalized seizure activities



CT BRAIN: Atrophy of Right cerebral hemisphere with dilatation of the ipsilateral ventricle, widening of sulci and sylvian fissure on the same side. There was also shift of midline to right and thickening of calvarium on the left side.



MRI Brain: Showed loss of right cerebral lobe

DIFFERENTIAL DIAGNOSIS
- Rasmussen encephalitis
- Sturge Weber syndrome
- Silver Russel syndrome
- Fishman syndrome
- Basal Ganglia Germinoma

DISCUSSION

In 1933, Dyke, Davidoff and Masson first described the syndrome in plain radiographic and pneumoencephalographic changes in a series of nine patients. It is characterized by asymmetry of cerebral hemispheric growth with atrophy or hypoplasia of one side and midline shift, ipsilateral osseous hypertrophy with hyperpneumatisation of sinuses mainly frontal and mastoid air cells with contralateral paresis. Other features are enlargement of ipsilateral sulci, dilatation of ipsilateral ventricle and cisternal space, decrease in size of ipsilateral cranial fossa & unilateral thickening of skull.

Clinical presentations include variable degree of facial asymmetry, seizures, contralateral hemiparesis, mental retardation, learning disabilities, impaired speech, etc. Seizures can be focal or generalized. Complex partial seizure with secondary generalization also had been reported. Both sexes and any of the hemisphere may be affected, but male gender and left side involvement are more common.

Cerebral hemiatrophy can be of two types: infantile (congenital) and acquired. The ***infantile variety*** results from various etiologies such as infections, neonatal or gestational vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies, and coarctation of the midaortic arch. The patient becomes symptomatic in the perinatal period or infancy. The main causes of ***acquired type*** are trauma, tumor, infection, ischemia, hemorrhage, and prolonged febrile seizure. Age of presentation depends on time of insult and characteristic changes may be seen only in adolescence or adult. In our case, the findings of Right cerebral hemiatrophy with enlarged cortical sulci, microcephaly, and presentation at the age of 11 months reflect an onset of brain insult after the completion of sulci formation, probably of vascular origin involving right middle cerebral artery.

TREATMENT

Patients with DDMS usually present with refractory seizures and the treatment should focus on control of the seizures with suitable anticonvulsants. Along with drugs, physiotherapy, occupational therapy, and speech therapy play a significant role in long-term management of the child. Prognosis is better if the onset of hemiparesis is after 2 years of age and in absence of prolonged or recurrent seizure. Hemispherectomy is the treatment of choice for children with intractable disabling seizures and hemiplegia with a success rate of 85% in selected cases.