Original Article

Dyke-davidoff-Masson syndrome

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Abstract A 27 years old male presented with recurrent generalized seizures, left hemiparesis and had developmental delay in motor and speech domains.MRI of the brain revealed characteristic features diagnostic of cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome (DDMS).
Keywords: Dyke-davidoff-Masson

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INTRODUCTION

DDMS, also known as cerebral hemiatrophy, is typically caused by an in utero or early childhood cerebral insult such as infarct, trauma or (less commonly) infection. Lack of ipsilateral brain growth causes the calvaria and diploic space to thicken, while the paranasal sinuses and mastoids become enlarged and hyperaereated. The clinical features are variable and depend on the extent of brain injury. More commonly they present with recurrent seizures, facial asymmetry, contralateral hemiplegia, mental retardation or learning disability, and speech and language disorders. Sensory loss and psychiatric manifestations like schizophrenia had been reported rarely. The syndrome had been documented mainly in adolescents and adults. However, it can also be seen in children. We present here a 27 year old male with typical imaging and clinical features of DDMS.

CASE REPORT

A 27 years old male presented with recurrent generalized seizures, left hemiparesis and had developmental delay in motor and speech domains since childhood. Birth history, developmental milestones till 5 years of age were normal according to history narrated by the mother. Vision and hearing were normal, and cranial nerves were intact. Neurological examination revealed left sided spastic hemiparesis with brisk tendon reflexes and extensor planter response, other systemic examinations being normal .Blood counts and serological status was unremarkable. MRI Brain of the same patient was performed. Following differentials are to be considered and ruled out by appropriate clinical history, examination and cross sectional modalities – Sturge Weber syndrome, Rasmussens encephalitis, hemimegaloencephaly and large MCA territorial infarcts

IMAGING FINDINGS

There is severe right hemispheric volume loss, prominent sulci and cisterns, T2 and FLAIR demonstrating encephalomalacia with shrunken hyperintense gyri and subcortical white matter .The ipsilateral cerebral peduncle is atrophic with atrophy of contralateral cerebellum. There is thickening of ipsilateral calvarium and diploic space, enlarged hyperaereated right mastoid and ipsilateral paranasal sinuses. Gross ipsilateral dilatation of right lateral ventricle. Left cerebral hemisphere and right cerebellar hemisphere are unremarkable**.** It neither enhances on T1 C+ nor demonstrates restricted diffusion.

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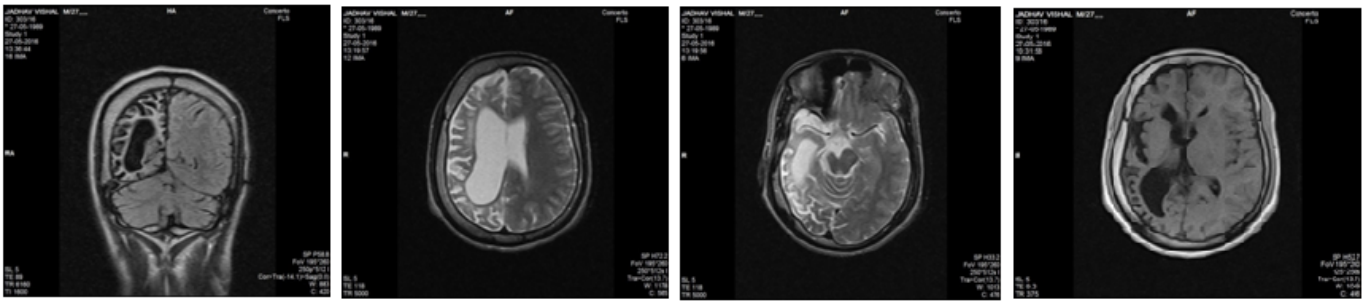

Figure 1: Coronal FLAIR reveals dilated right lateral ventricle, right hemispheric volume loss and atrophy of contralteral cerebellum
Figure 2: Axial T2WI demonstrates right hemispheric volumes loss with prominent sulci and cisterns
Figure 3: Axial T2WI reveals small ipsilateral cerebral peduncle
Figure 4: Axial T1WI demonstrates right calvarial and diploic space thickening

DISCUSSION

In 1933, Dyke, Davidoff and Masson first described the syndrome in plain radiographic and pneumoencephalographic changes in a series of nine patients.[1] 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.[2] Other features are enlargement of ipsilateral sulci, dilatation of ipsilateral ventricle and cisternal space, decrease in size of ipsilateral cranial fossa, and 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.[3] Both sexes and any of the hemisphere may be affected, but male gender and left side involvement are more common.[4]. When the brain fails to grow properly, the other structures grow inward resulting in increased width of diploic spaces, enlarged sinuses, and elevated orbital roof.[5] Cerebral hemiatrophy congenital(infantile) or acquired. Causes of infantile type include infections, neonatal or gestational vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies, and coarctation of the midaortic arch.[6,7]. Causes of acquired include trauma, tumor, infection, ischemia, hemorrhage, and prolonged febrile seizure. Age of presentation varies accordingly depending upon the time of insult. Treatment is mainly seizure control using anticonvulsants. Supportive therapy is needed in the form of speech therapy, occupational therapy and physiotherapy. Hemispherectomy is the definitive treatment for recurrent generalized seizures. As hemispherectomy is not available even in many urban tertiary care centers, it is very important for a neurologist or pediatrician to diagnose the condition early by means of appropriate cross sectional imaging and the treatment should focus on optimum control of seizures, revision of drug doses from time to time, and domiciliary physiotherapy. Following differentials are to be considered and ruled out by careful history, clinical examination and appropriate cross sectional imaging- Sturge Weber syndrome, Ramussens encephalitis, hemimegaloencephaly and large MCA territorial infarcts.

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