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DYKE DAVIDOFF MASSON SYNDROME-AN UNUSUAL CAUSE OF REFRACTORY SEIZURE: A RARE CASE REPORT

Medicine

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ABSTRACT

BACKGROUND: Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological condition characterized by drug-resistance seizures, hemiparesis, mental retardation, facial asymmetry, and intellectual disabilities. On brain imaging, the disease is characterized by cerebral hemi atrophy with ipsilateral calvarial thickening and hyperpneumotisation of paranasal sinuses or mastoid air cells.
INTRODUCTION: we describe a rare case of Dyke Davidoff Masson syndrome with an unusual cause of refractory seizure in an adolescent.
Diagnosis: DDMS was diagnosed from its manifestations, biochemistry indexes, and imaging (computed tomography angiography, magnetic resonance venography, and so on).
Interventions: Several drugs are used to treat the disease, including valproate, carbamazepine, topiramate, and levetiracetam.
Outcomes: Under the medicine treatment of sodium valproate, levetiracetam and carbamazepine, the patient experienced generalized tonic clonic seizure seizures approximately once or twice per month that lasted 30 to 60seconds each without any complications observed during a follow-up period of.
CONCLUSIONS: It was concluded early diagnosis and pharmacotherapy are the keys to preventing intellectual decline in DDMS patients. Moreover, the combination of sodium valproate, levetiracetam and carbamazepine could significantly reduce the frequency and duration of seizures, despite not eliminating them completely24months

KEYWORDS

INTRODUCTION:

Dyke Davidoff Masson syndrome is a rare disease which is clinically characterized by hemiparesis, seizure, facial asymmetry and mental retardation.

The classical findings including cerebral hemi atrophy along with calvaria thickening and hyper pneumatization of the frontal sinuses are only found if an insult to the brain occurs before 3 years of age. DDMS is commonly caused by a perinatal insult such as trauma, intrauterine infection, and hypoxic ischemic encephalopathy. Therefore, fewer cases have been reported regarding adolescents. Sensory loss and psychiatric manifestations like schizophrenia had been reported rarely (1). Predominantly, there is no established sex predilection or involvement of a specific hemisphere but, the involvement of the left side and male gender are more common in the literature [6].

CASE REPORT:

A 19 yrs. old male presented with

1. recurrent seizure
2. Right sided hemiparesis
3. Facial asymmetry
4. Microcephaly

He has microcephaly with progressive weakness in right side of body since 3 months and not able to sit and walk by himself since 2 months. The bilateral carotid pulsations are normal with no bruit. Vision and hearing are normal, and cranial nerves are intact. Neurological examination revealed right-sided spastic hemiparesis with brisk tendon reflexes and extensor planter response, other systemic examinations being normal. Patient is mentally retarded and loss of intelligence since childhood. He has three siblings all of them are normal, had normal birth history and good in study. He has no such significant family history. He was born out of a nonconsanguineous marriage. Birth history was indicative of a full- term delivery without any antenatal and perinatal complications. However, he had a history of seizures at the age of 5 years for which he was admitted in a local hospital as a case seizure disorder. He discharged from hospital subsequently after one month on antileptic medications. However, he continued to have seizure. Besides this his parents also noted learning difficulties, slurred speech, and progressive right hemiparesis and managed conservatively by local practitioner. his seizure did not respond to several antiepileptic medications in different combinations. on examinations he did not have neurocutaneous markers also.

All routine biochemical investigations including liver and renal function were within normal limits. Random blood sugar, serum electrolytes including calcium levels were within normal range.

MRI Brain was done.

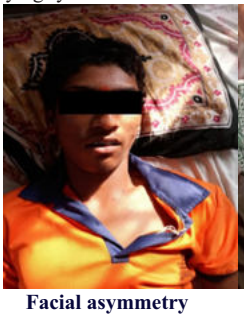
s/o:
left cerebral hemi atrophy with Wallerian degeneration of left cerebral peduncle with dilatation of left lateral ventricle seen.

Diffuse FLAIR hyperintensities seen in left fronto-parietal lobe s/o gliosis.

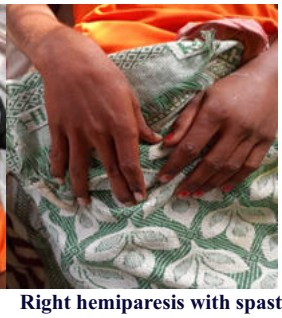
Hyper pneumatization of left frontal sinus is seen.

Opthal findings: WNL

CT Head findings:
Plain CT study of the brain shows small well defined cystic lesion in left parietal region (likely extra-axial) with surrounding gliosis and volume loss. Focal scalloping inner table of left parietal bone overlying cystic lesion.



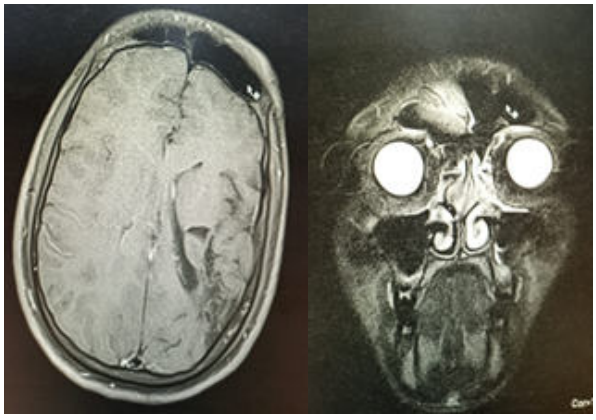
Facial asymmetry



Right hemiparesis with spastic wrist flexion

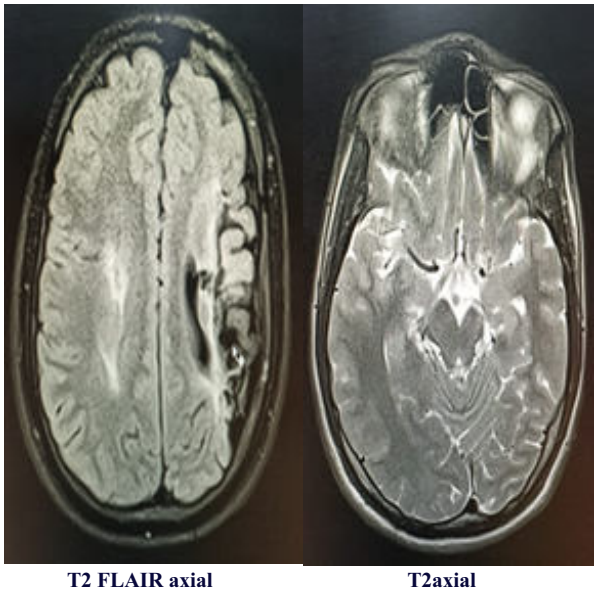
Right sided hemiparesis, microcephaly and facial asymmetry

Pneumosinus dilatans frontalis on left side



T2 FLAIR axial T2 coronal

Left cerebral hemi atrophy with Wallerian degeneration of left cerebral peduncle with dilations of left ventricles with left fronto-parietal gliosis.



T2 FLAIR axial T2axial

DISCUSSION:

Dyke Davidoff Masson syndrome is a rare disease which is clinically characterized by hemiparesis, seizure, facial asymmetry and mental retardation.

The classical findings including cerebral hemi atrophy along with calvaria thickening and hyper pneumatization of the frontal sinuses are only found if an insult to the brain occurs before 3 years of age.(2) The possible mechanism of cerebral atrophy and the related progressive neuro deficit is hypothesized to be due to several ischemic episodes resulting from these factors, which reduce the production of brain-derived neurotrophic factors, which in turn leads to cerebral atrophy.

CT and MRI are the two gold standard imaging modalities that prove to be very significant in the diagnosis of DDMS. These two imaging modalities provide very detailed cross-sectional images. The typical imaging features for DDMS include prominent cortical sulci, dilated lateral ventricles, cerebral hemi atrophy, hyper pneumatization of the frontal sinus, and compensatory hypertrophy of the skull. These imaging findings become more obvious as the patient ages. When the cerebral damage occurs during the intrauterine period or before the age of 3, compensatory calvaria involvement can be seen.

In a patient with cerebral hemi atrophy, the differential diagnosis includes Rasmussen encephalitis, Sturge-Weber syndrome, basal ganglia dysgerminoma, Fisherman syndrome, and Silver-Russell syndrome. Detailed history and examination are required along with laboratory and imaging workup to differentiate these diseases.

For the management of seizures, mono, or poly anticonvulsant medication is given. Children with refractory epilepsy and hemiplegia are potential candidates for hemispherectomy, with a success rate of 85%. Vagal stimulation is another alternative. Despite lacking any specific treatment algorithm, therapy with antiepileptics and surgery is indicated in specific cases. Long-term supportive management includes physical, language, and occupational therapy.

CONCLUSION:

Under the medicine treatment of sodium valproate, carbamazepine and levetiracetam, the patient experienced generalized tonic clinic seizure seizures approximately once or twice per month that lasted 30 to 60seconds each without any complications observed during a follow-up period of.

It was concluded early diagnosis and pharmacotherapy are the keys to preventing intellectual decline in DDMS patients. Moreover, the combination of sodium valproate and carbamazepine and levetiracetam as well could significantly reduce the frequency and duration of seizures, despite not eliminating them completely 12 months.

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