

**Case Report****Dyke-Davidoff-Masson Syndrome**

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ABSTRACT

Dyke –Davidoff-masson syndrome (DDMS) is characterized by seizures, hemiparesis and mental retardation with radiological features are cerebral hemiatrophy with homolateral dilation of ventricles with hyperpneumatization of sinues.

We report a case of DDMS in 30 years old patient who presented with complaint of fever ,high grade ,with chills and rigors,which associated with headache and cough from last 4 days with left sided body weakness.

Introduction

Dyke –Davidoff-masson syndrome is disease which is clinically characterized by hemiparesis, seizures, facial asymmetry, and mental retardation. This disease is a rare entity, and it mainly presents in childhood. Adult presentation of DDMS is unusual, it should be kept in mind as a possibility in an adult who presents with long duration of progressive hemiparesis with seizures and mental retardation and has been rarely reported in the medical literature.

Case Report

A 30 years male who residing in delhi, born with full term normal vaginal delivery, without any birth trauma, having history of recurrent seizures taking treatment for left parietal motor seizures.

There was no H/O significant antenatal and perinatal complications.

at the age of 9 month patient develop high grade fever with chills and rigors with 1st episode of seizures and left sided hemiparesis that time patient admitted in some other hospital and treated for? tubercular meningitis.

Patient put on anti-epileptics treatment. However patient continued to have seizures for which patient taking several antiepileptics in different combinations.patient at the time of presentation was on 2 medications. His hemiparesis gradually worsened with increased stiffness in left side of body.

Having history of delayed mile stones. start walking and talking at age of 7-8 yrs. He was not able to perform on MISIC .on VSMS he was found to have SA of 6 yrs 9 months corresponding to SQ in range of 43-47.IQ test in range of 38-42.

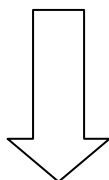


Neurological Examination

Patient is conscious
 No sign of meningeal irritation.
 Cranial examination-,no ptosis ,no facial deviation
 Reflexes –bilateral planters –extensor
 Left side knee jerk-brisk
 Bilateral ankle clonus present
 Nystagmus present –horizontal and vertical gaze towards left side
 Atrophy of left side body upper and lower limb muscles.

Hematoloical profile

Hb 5.6 →6.6→8.2→10.1
 Tlc -26000→31800→16000→11800
 DLC –N78% L14.4%M4.2%E1.5%
 PLT 30000→25000→50000→62000

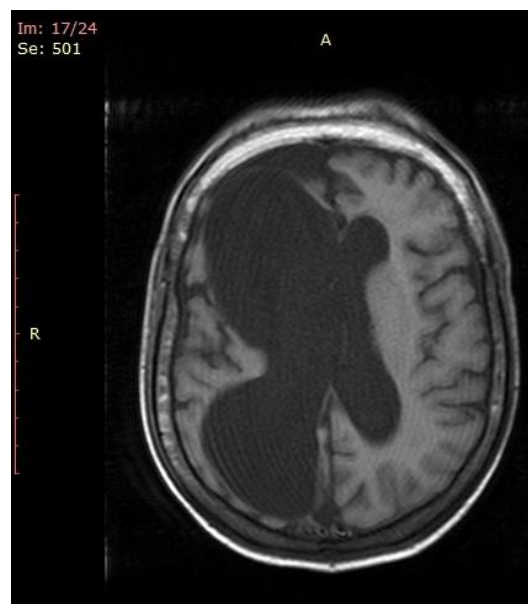
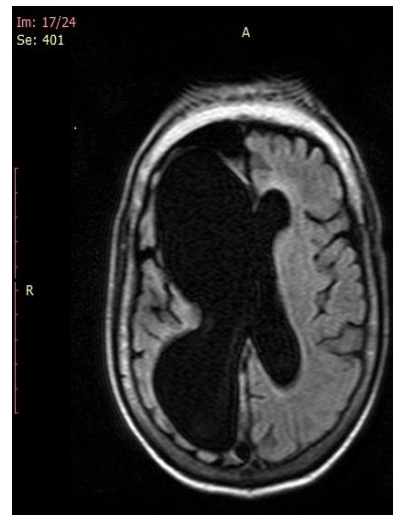
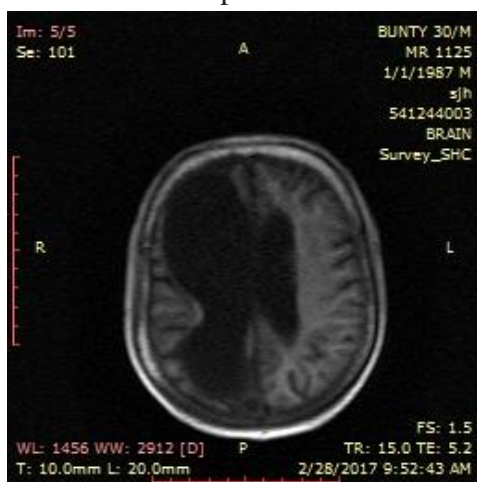


all are in improving trend

Serum electrolyte normal in range
 LFT AND KFT within normal limits
 Blood cultures →no growth
 2D ECHO → normal study
 VEP study →noraml study
 Fundus Examination → bilateral optic atrophy present

Radiological

MRI brain →right sided hemiatrophy with T2W/FLAIR hyperintensities with compensatory exvacuodilation of ipsilateral ventricle and hyperpnematisation of ipsilateral sinuses.



CECT CHEST →multiple nodules with cavitation predominantly supleural in location diffusely distributed in bilateral lung parenchyma s/o likely septic emboli.

Discussion

This is a rare condition which derives its names from the researchers Dyke Davidoff and Masson who first reported this condition in 1993 they took 9 patients who presented with seizures facial asymmetry and mental retardation and noticed plain skull radiographic changes^[1] these findings are due to cerebral injury that may occur early in life or in utero insult to immature brain cells lead to neuronal loss and impaired brain growth. Etiological factor due to trauma inflammation of vascular malformation or

occlusions. When the insult occurs in-utero, it could be due to gestational vascular occlusion, primarily involving the middle cerebral vascular territory.

The radiological features are unilateral loss of cerebral volume and associated compensatory bone alterations in the calvarium, such as thickening, hyperpneumatization of the paranasal sinuses and mastoid cells and elevation of the petrous ridge.

These radiological findings develop early in life (before 3 years of age) like hemiatrophy of one cerebral hemisphere which compensatory cranial changes but these changes are not frequently encountered in clinical practice^[2]

The classical clinical presentation includes seizures, facial asymmetry contralateral hemiplegia or hemiparesis and mental retardation. With history of treatment for meningoencephalitis. However mental retardation was not always present and seizures may appear months or years after the onset of hemiparesis and can have history of delayed milestones^[3,5].

The clinical findings mainly depends on degree of the extent of the brain Injury occur during in utero period or perinatal period or secondary to trauma, inflammation or vascular malformations. Imaging studies show unilateral loss of volume of brain, ventricular dilatation and enlargement of sulci^[4]. A possible etiological relation of cerebral hemiatrophy and seizures has been reported by different studies in India^[6,7].

Dyke Davidoff Masson Syndrome should be differentiated from Rasmussen encephalitis, Basal cell germinoma, Sturge Weber syndrome, Fishman syndrome, Linear Nevus syndrome, Silver-Russell syndrome and^[8]

For the diagnosis of condition a proper clinical history and CT/MRI findings provide the correct diagnosis. treatment mainly according to symptoms.

Prognosis is better if there is absence of prolonged seizures and hemiparesis and hemiplegia occurs after 2yrs of age.

Reference

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