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CASE REPORT

DYKE -DAVIDOFF-MASSON SYNDROME: A RARE CASE REPORT

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ABSTRACT

Dyke-Davidoff-Masson Syndrome (DDMS)also known as cerebral hemiatrophy is a rare neurological disease characterized by Seizures, hemiparesis, cerebral hemiatrophy, facial and body asymmetry with mild intellectual disability. DDMS can be diagnosed by characterized by classical radiographical findings, that is cerebral hemiatrophy, ipsilateral dilated ventricle, ipsilateral osseus hypertrophy and excessive pneumatization of ipsilateral frontal and other sinuses. DDMS presents withearlypartial onset seizure with hemiparesis. Later in adolescence facial and body asymmetry is evident with mild cognitive dysfunction. We are presenting a case in which a23 year oldmale presented with uncontrolled partial onset seizures with secondary generalization, right hemiparesis, right sided hemiatrophy of limbs and body with characteristic radiological findings suggestive of DDMS.

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INTRODUCTION

Dyke-Davidoff-Masson Syndrome (DDMS) or Cerebral Hemiatrophy is a rare neurological condition of unknown frequency characterized by cerebral hemiatrophy/hypoplasia, hemiplegia, seizures, facial asymmetry, mild mental retardation with characterized radiological features such as cerebral hemiatrophy, ipsilateral compensatory osseus hypertrophy, hyper-pneumatization of ipsilateral frontal sinus (sometimes mastoid air cells and ethmoid sinuses also). DDMS first described by C.G. Dyke, L.M. Davidoff, and C.B. Masson in 1933 in a case series of 9 patients with hemiplegia and cranial asymmetry on plain cranial X-rays presented with seizures [(1,2)]. DDMS results from brain injury especially early in life. The etiology can be congenital (such as intra-uterine infections, neonatal or gestational vascular occlusion involving the middle cerebral artery, coarctation of mid aortic arch and unilateral cerebral arterial circulation anomalies) or acquired including trauma, tumor, prolonged febrile seizures[(3)]. DDMS is rare in occurrence, but it is an important cause of recurrent and refractory seizures. Here we report a case of DDMS in a patient who is on antiepileptic drugs and presented with uncontrolled seizures.

CASE REPORT

A 23 year old adult male came to the medical emergency with multiple seizure episodes of partial onset with secondary generalization. Patient was admitted to the Medicine ward of RNT Medical College, Udaipur for further management. On admission patient was in altered sensorium so history was given by his family members that patient had 4-5 episodes of seizures which started with abnormal movement of right upper limb and face which progressed to involve abnormalupward deviation of both eyes with impaired consciousness followed by secondary generalized tonic clonic seizures with post ictal delirium lasting for 48 hours. On further probing, it was found that patient is a known case of epilepsy with right side hemiparesis since the age of 3 years. Patient is afirst-born child with normal vaginal delivery at hospital. The Antenatal, perinatal, and postnatal period was uneventful. He cried at birth and there was no history of any hospitalization during neonatal period and infancy for any other cause. No history of febrile seizures. No significant family history was found. Patient attained all the milestones normally till the age of 3 years. Patient developed first episode of seizure at the age of three which was started from right upper limb with involvement of right half of face

with secondary generalized seizure followed by post-ictal confusion lasting for 30mins to 1 hour. The patient developed a new onset right sided weakness since then. Patient was treated symptomatically at nearby hospital and was given antiepileptics. Because of poor adherence to treatment patient had history of previous three admissions for uncontrolled/refractory status epilepticus with prolonged post-ictal confusion. Initially patient had normal mentation at onset, and he is educated up to 10th standard. But it was associated with progressive decline in intelligence with poor scholastic performance during the later grades of schooling. Patient also developed progressive hemiatrophy of right side of the body with mild facial asymmetry and contractures developed at right distal upper and lower limb. On examination, on day 3rd of admission patient had MMSE score of 8/30 and on day 5 it was 23/30 associated with reduced attention span and mild cognitive impairment otherwise patient was conscious, oriented to time, place and person. Motor examination revealed right spastic hemiparesis, brisk reflexes with right ankle clonus. It was associated with marked reduction in muscle or atrophy over right side of body.Right plantar showed extensor response. Cranial Nerve, Sensory and Cerebellum examination was within normal limits.Spinal and vertebral examination revealed no abnormality. NCCT head was done which showed left cerebral hemiatrophy with dilatation of ipsilateral lateral ventricle and hyperpneumatization of left frontal sinus and mastoid air cells. With the above clinical and radiological features, a diagnosis of Dyke Davidoff Masson syndrome (DDMS) was suspected.

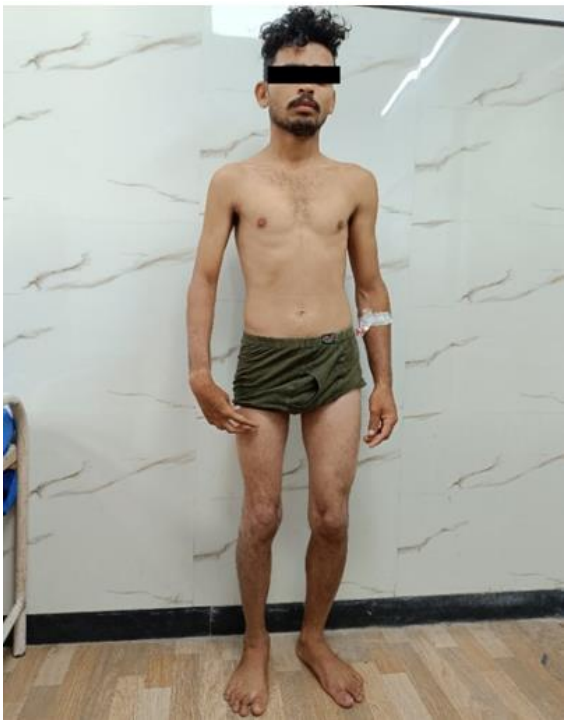

Figure 1. Showing hemiatrophy of right half of bodyand contracture of right wrist

Then EEG and MRI Brain was planned. EEG showed normal study with no abnormality on hyperventilation and photic stimulation. MRI brain was planned which revealed FLAIR/T1W hypointense and T2W hyperintense with peripheral hyperintensity areas in left fronto-parietal-temporal regions with ex-vacuo dilatation of ipsilateral lateral ventricle.

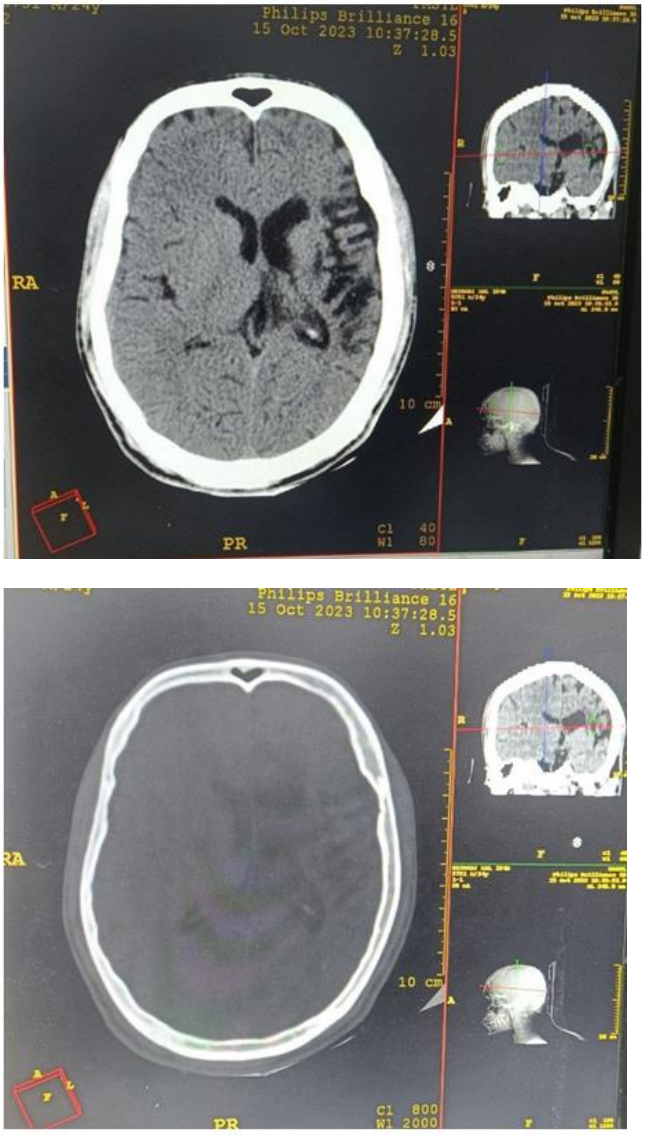

Figure 2. NCCT Head showing left cerebral hemiatrophy with dilatation of ipsilateral lateral ventricle (Left) and ipsilateral compensatory calvarial hypertrophy (Right)

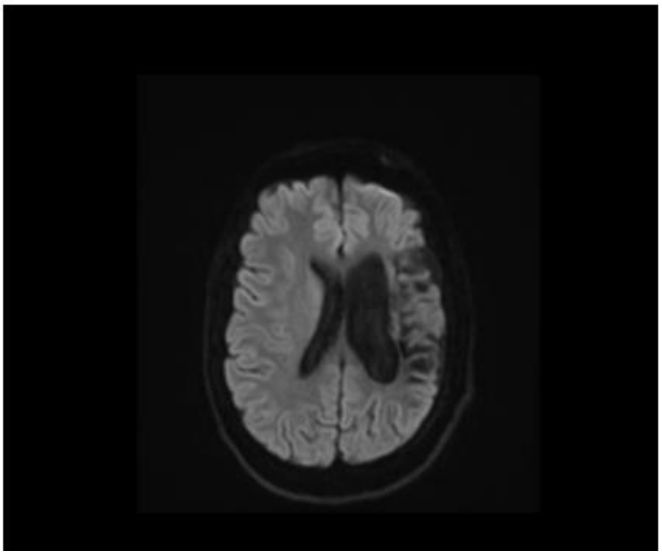

Figure 3.

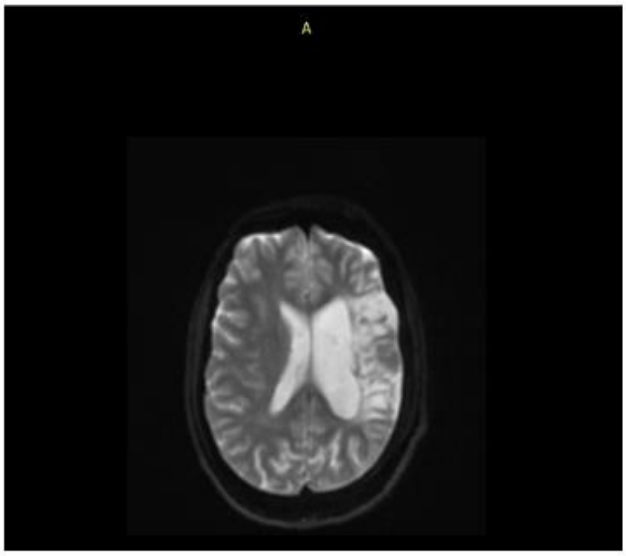

Figure 4. MRI Brain showing left cerebral hemiatrophy with ex-vacuo dilatation of ipsilateral lateral ventricle on T2 FLAIR (Left) and T2 (Right)

A diagnosis of DDMS was confirmed. Patient was taking sodium valproate, but seizures are not controlled. So, Levetiracetam and carbamazepine also started. The patient was discharged in stable condition and advised to continue Anti-epileptic as prescribed with monthly follow-up in Medicine OPD.

DISCUSSION

DDMS is a rare neurological disease first described in 1933, clinically characterized by hemiplegia, recurrent seizures, hemiatrophy, facial asymmetry and varying degree of intellectual disability. DDMS defined bycharacteristic radiographic features including cerebral hemiatrophy, dilated ipsilateral ventricle, compensatory ipsilateral calvarial hypertrophy and enlarged ipsilateral frontal sinus, mastoid air cells and ethmoidal sinuses[4,5]. DDMS results in cerebral hemiatrophy, more commonly involving the left hemisphere.DDMS can occur in both sexes, no gender preponderance is seen yet. DDMS is a disease of childhood which presents with partial onset seizures with or without secondary generalization and hemiparesis. Hemiparesis usually occur years after seizure presentation or sometimes they can present at the same time (as in our case) and less commonly, hemiparesis precedes the seizure onset. Although DDMS presents early in life with seizures, but it is easily missed and most cases in India diagnosed in late childhood and early adulthood. DDMS is associated with mild Intellectual disability which is progressive.Patients with DDMS attain normal milestones in their early life and had no cognitive dysfunction at the disease onset. As in our case, this patient is educated up totenth standard with poor scholastic performance in the last 2 years of schooling which compelled him to discontinue schooling.As the patient grows older, the seizures become more refractory,and the patient needs multiple anti-epileptic drugs to control them. It shows the progressive nature of the disease and ongoing insult to the brain. Thus, the characteristic changes of DDMS may be only seen in adolescence and adults.Differential diagnosesof DDMS include Rasmussen Encephalitis, Sturge-Weber syndrome, Silver-Russell Syndrome and Fishman Syndrome[5,6,7]. Rasmussen Encephalitis[8,9]is a chronic progressive disorder of unknown etiology presented with partial onset seizures, may evolve to complex partial seizures and 50% of them have epilepsiapartialis continua. Other features are progressive hemiatrophy of brain, hemiplegia, and mild intellectual disability. In some cases, possible autoimmune etiology has been described that is antibody against Glutamate Receptor-3(Anti-GluR3 Ab). It can be differentiated from DDMS as CT/MRI does not show the compensatory calvarial hypertrophy and hyper-pneumatization of frontal and other sinuses. Sturge-Weber Syndrome[10,11]is a neurocutaneous disorder characterized by angiomas involving face, choroid and leptomeninges. Seizures are usually the first neurological manifestation. Other features include facial port wine stain in the distribution of ophthalmic and maxillary division of trigeminal nerve. CT head reveals calcification classically describes as “tram track calcifications”, leptomeningeal enhancement and cortical atrophy. Silver-Russell Syndrome [12] characterized by severe intra-uterine growth retardation with a preserved head circumference. Facial dysmorphism and asymmetry are typical features such as broad forehead tapering to small jaw giving triangular face appearance. Other features are clinodactyly of fingers, limb and body asymmetry and low birth weight baby. Fishman Syndrome[13]also known as Encephalocraniocutaneous Lipomatosis (ECCL) is a rare congenital Neurocutaneous disorder characterized by early onset seizures, intellectual disability, lipomas of skull, eyes and heart with cerebral malformations such as hemiatrophy, intracranial cysts and calcifications. Treatment of DDMS is mainly aims around controlling seizures with anti-epileptic drugs. In cases of intractable disabling seizures, Hemispherectomy can be done with a success rate of 85%. Occupational rehabilitation therapy and physiotherapy should also be advised to patients.

CONCLUSION

DDMS is a rare neurological disabling diseasewhose etiology is not well understood. Treatment largely includes anticonvulsant therapy to control seizures. Because of its rarity it can be easily missed but one should suspect this if a child presents with recurrent and refractory seizures with other clinical features of DDMS. Patients are advised to adhere to long term follow-up and anticonvulsant therapy.

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