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DYKE-DAVIDOFF-MASSON SYNDROME: A RARE CASE REPORT

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ABSTRACT

Dyke–Davidoff–Masson syndrome (DDMS) is a rare condition, in which patient has cerebral hemiatrophy, seizures and mental retardation after some insult to the developing brain during fetal or infantile period. Diagnosis is mainly dependent upon clinical and radiological findings. Here we are presenting a case of a 14 year old girl who came to us in altered sensorium after an episode of status epilepticus with left hemiparesis and cognitive impairment since childhood. CT of brain revealed right cerebral atrophy and hyperpneumatization of right frontal and mastoid sinuses characteristic of Dyke-Davidoff-Masson syndrome[DDMS].

INTRODUCTION

DDMS is a rare syndrome of unknown frequency, mainly documented in children and adolescents but can be found in adults also.

In 1933, Dyke, Davidoff, and Masson described 9 patients with facial asymmetry and skull pneumatoencephalographic changes having clinical characteristics of hemiparesis, mental retardation and seizures. .The radiographical features of the skull were ipsilateral osseous hypertrophy of the calvarium and hyper-pneumatization of the para-nasal sinuses.

Clinical findings may be variable according to extent of the injury. Usually patient presents with recurrent seizures, hemiparesis and learning disability. CT scan of brain may reveal ipsilateral cerebral hemiatrophy withventriculomegaly, thickening of ipsilateral calvarium, hyperpneumatization of the ipsilateral paranasal sinuses mainly frontal and mastoid, midline shift to the affected side and elevation of the temporal bone. MR scan may reveal unilateral cerebral parenchyma loss, brainstem, thalamus and basal ganglia atrophy, middle fossa hypoplasia, sulcal enlargement , encephalomalacia and Wallerian degeneration of the mesencephalon .

Male gender and left side are frequently associated with DDMS .

CASE REPORT

A 14 year old girl born to non-consanguineous marriage, without any history of significant antenatal or perinatal complications, was brought to us in altered state after an episode of status epilepticus . At the age of 2 years, she had an episode of prolonged febrile seizures following which she developed weakness in left half of body, recurrent seizures and mental retardation for which she was not on any treatment. Seizures frequency was 4-5 times a month and weakness was non progressive in nature. There was no history of head injury. Patient did not have formal schooling. On examination, the bilateral carotid pulsations were normal with no bruit. Vision and hearing were normal. Cranial nerve examination was unremarkable. Neurological examination revealed 3/5 power and upper motor neuron signs in the left upper and lower limbs with mild disuse atrophy. Patient also had hemiplegia in right side of body which completely recovered within 24 hours , diagnosed as Todd's paresis. Hearing and vision were normal. Other systemic examination and vitals were normal. A plain CT of brain was done which showed atrophy of right cerebral hemisphere with hyperpneumatization of frontal and mastoid sinuses, midline shift to right side and right ventricular dilatation. There was also thickening of calvarium of right side skull. T1W and T2W MR scan of brain revealed unilateral atrophy of right cerebral hemisphere, dilatation of right lateral ventricle and thickening of right temporo-parietal bones. There was peripheral area of encephalomalacia and gliosis in right high parietal lobe. Bilateral basal ganglia, thalami, brainstem and cerebellum were normal. MR venogram was normal. Routine blood investigations including CBC, LFT, RFT, S.electrolytes and S.calcium were normal. CSF analysis was also under normal limits. By the clinical presentation and above mentioned radiological findings, the diagnosis of DDMS was confirmed.

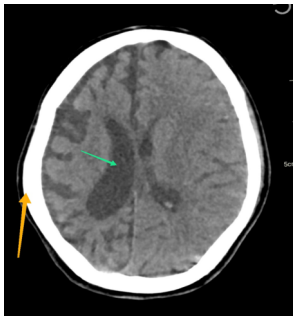

Figure 1 CT scan of brain showing right cerebral hemiatrophy, midline shift to the right, right ventricular dilatation (green arrow) and right calvarial thickening (yellow arrow)

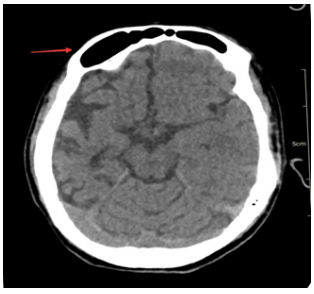

Figure 2 CT scan of brain showing hyperpneumatization of right frontal sinus (red arrow).

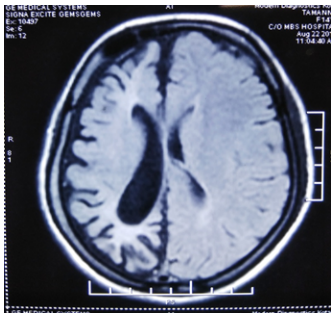

Figure 3 MR Scan of brain showing right cerebral atrophy.

Discussion

Cerebral hemiatrophy can be of two types, congenital and acquired. The congenital variety results mainly from vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies, coarctation of the midaortic arch and sometimes idiopathic . The main causes of acquired type are birth trauma, perinatal intracranial hemorrhage, Rasmussen encephalitis, prolonged febrile seizures, infections like Herpes encephalitis, TORCH syndrome, Sturge-Weber syndrome, Diaschiasis Commisuralis, Ischaemia, Neoplasia like basal ganglial germinoma and Radiation.

Characteristic radiological features are present when the insult to brain occurs before 3 years of age as the brain reaches half of its adult size during the first year of life and reaches three-fourths of that size by the end of the third year. The outward pressure of the developing brain is responsible for the gradual enlargement and general shape of the adult head. When the brain does not grow properly, the bony table tends to direct their growth inward, causing above mentioned radiological findings . In our case characteristic radiological findings suggest injury to brain before 3 years of age which is also consistent with the history.

In congenital hemiatrophy, when the insult occurs in utero, there is ipsilateral shift of midline structures, and the sulcal prominence is absent whereas the atrophied cerebral hemisphere will have prominent sulcal spaces if the insult occurs after birth or after the end of sulcation.

In our case, sulcal prominence of right side hemisphere signifies the insult to brain after the end of sulcation.

Seizures can be focal or generalized. Complex partial seizures with secondary generalization also have been reported. Fits may be refractory to treatment and combination of antiepileptic drugs may be required to control them . If the onset of hemiplegia occurs without seizures, the prognosis for future epilepsy, for intellect and behavior is good.

Differential diagnosis for DDMS are Sturge–Weber Syndrome, Basal cell germinoma, Fishman syndrome, Silver–Russell syndrome, linear nevus syndrome, and Rasmussen encephalitis.

Treatment is symptomatic and should target seizures control. Seizurs are refractory and thus sometimes multiple anticonvulsants are used. Along with drugs, speech therapy, physiotherapy and occupational rehabilitation play important role in improving quality of life of patient. Patients with intractable disabling seizures are potential candidates for functional hemispherectomy with good prognosis.

The present case exhibits classical features of the acquired type of cerebral hemiatrophy involving right cerebral hemisphere. This case has deviated from the usual presentation of male predominance and left sided DDMS. In cases of intractable seizures, physicians should consider DDMS as one among the differential diagnosis.

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