ORIGINAL RESEARCH PAPER

General Medicine

DYKE DAVID OFF MASSON SYNDROME

KEY WORDS:

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ABSTRACT

Dyke-Davidoff-Masson syndrome is a rare entity characterized by hemi cerebral atrophy/hypoplasia secondary to brain insult in fetal or early childhood period along with ipsilateral compensatory osseous hypertrophy and contralateral hemiparesis. A 25 year old male patient presented with recurrent refractory siezures and spastic hemiplegia. MRI findings revealed characteristic features of cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome.

INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) ia a rare disorder referring to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy), which is usually due to an insult to the developing brain in fetal or early childhood period. The clinical features are variable and depend on the extent of brain injury. Common presentation is with recurrent seizures, facial asymmetry, contralateral hemiplegia, mental retardation or learning disability, and speech and language disorders. Sensory loss and psychiatric manifestations like schizophrenia have also been reported rarely. Typical radiological features include cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses. The syndrome had been documented mainly in adolescents and adults. However, it can be seen in children also.

Case Report

24-year-old male patient, who is a graduate, presented to our hospital with a history of generalized tonic seizures with right sided spastic hemiparesis since 2.5 years of age. The patient was diagnosed with cerebral palsy at the age of 9 months. He was born out of a non-consanguineous marriage. Birth history was indicative of a full-term. However labor was not induced for 2 hours and hence forceps delivery was attempted but it failed and LSCS was done. Subsequent history is notable for delayed developmental milestones during infancy.The first episode of seizure occurred at the age of 2.5years in form of a stare look , raised tone and up rolling of eyes. Each episode lasted for 10-15sec followed by impaired awareness for next 10-15 seconds. He also had right sided spastic hemiparesis and slurred speech. Patient took treatment from a local hospital and was started on anti-epileptic medications along with regular speech and limb physiotherapy. However, he continued to have seizures which exacerbated at regular intervals due to poor compliance and other precipitating factors like sleep deprivation, stress, etc.

The seizures responded poorly to several antiepileptic medications in different combinations. However, the frequency and duration of seizure episodes had decreased over the past 7-8years. At the time of presentation, the patient was brought with chief complaint of involuntary tightness of all four limbs that was associated with uprolling of eyeballs, frothing from mouth that lasted for 10-15 seconds and multiple similar episodes present since 15 days. Some episodes were characteristic staring look with no response during episode. Such episodes lasted for 10-15 seconds after with the patient had no recollection of the event.

These episodes were not associated with antecedent fever, rash, joint pain, headache, nausea, vomiting, tingling, numbness or visual abnormality.

The patient was having refractory seizure and was started on multiple AEDS combination. On examination, he had relative spastic contracture of the right half of the body, high arched palate, right-sided torticollis and thoracic scoliosis with convexity toward left. Neurological examination revealed a spastic weakness of right upper and lower limbs with a spastic deformity of the right upper limb. Power was 4/5 in both right and left side, Deep tendon reflexes were brisk and right side plantar was extensor. Right sided facial deviation was present and left side facial weakness seen. Slurring of speech was also present. MMSE score was 23/25. Cell counts, liver and kidney function tests, serum electrolytes, serum homocysteine and thyroid function tests were unremarkable.

MRI of the BRAIN showed Hemiatrophy of the left cerebral hemisphere , area of gliosis and cystic encephalomalacia in left fronto-parieto-temporal lobe, left corona radiata and left basal ganglia with mild thickening of left calvaria on left side and mildly prominent left frontal sinus. The findings were consistent with Dyke Davidoff Masson syndrome. EEG brain showed theta waves which was suggestive of left hemisphere dysfunction. The patient was started on tablet levetiracetam, tablet phenytoin and tablet carbamazepine. The seizure intensity was controlled and the patient was discharged on these medications. He is currently on regular follow-up and doing well.

DISCUSSION

The differential diagnosis for the triad of hemiplegia, hemiatrophy and epilepsy include Rasmussen encephalitis, Sturge weber syndrome and hemimegalencephaly. Davidoff and Masson first described the syndrome in plain radiographic and pneumo-encephalographic changes in a series of nine patients. It is characterized by asymmetry of cerebral hemispheric growth with atrophy or hypoplasia of one side and midline shift, ipsilateral osseous hypertrophy with hyperpneumatization of sinuses mainly frontal and mastoid air cells with contralateral paresis. Other features include enlargement of ipsilateral sulci, dilatation of ipsilateral ventricle and cisternal space, decrease in size of

ipsilateral cranial fossa, and unilateral thickening of skull.

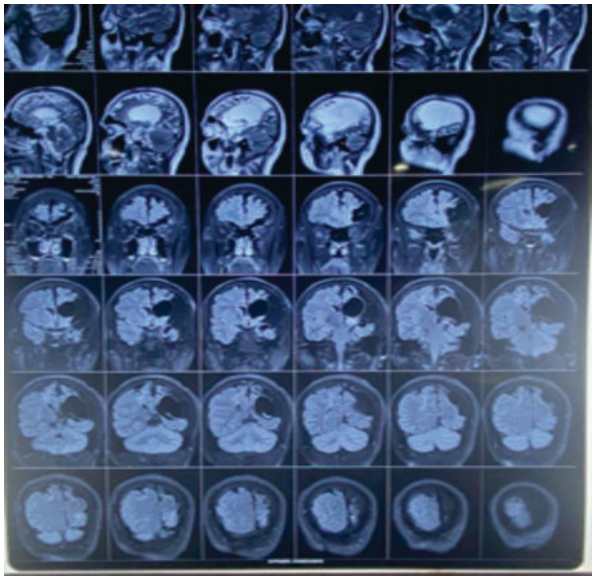
Clinical presentations include variable degree of facial asymmetry, seizures, contralateral hemiparesis, mental retardation, learning disabilities, impaired speech, etc. Seizures can be focal or generalized. Complex partial seizure with secondary generalization have also been reported. Both sexes and any of the hemisphere may be affected, but male gender and left side involvement are more common.

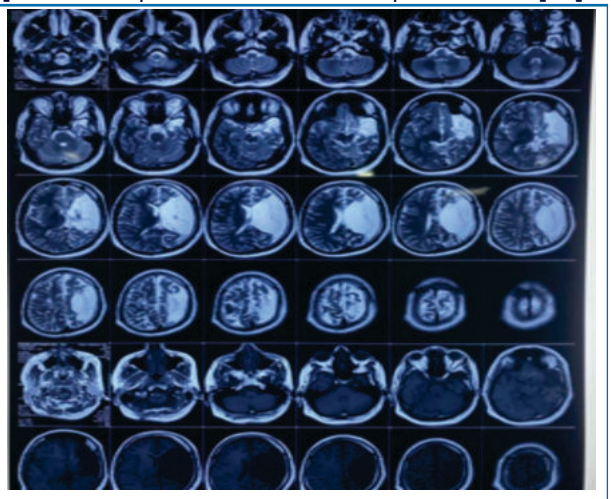
Our patient, therefore, had non-progressive infantile spastic hemiplegia, slurred speech and multiple episodes of refractory seizures. He had history of normal full term delivery with failed instrumental trial. He has delayed developmental milestones. All these clinical features are consistent with the diagnosis. However there is no mental retardation in this patient – which is suggestive of an atypical manifestation of this condition.

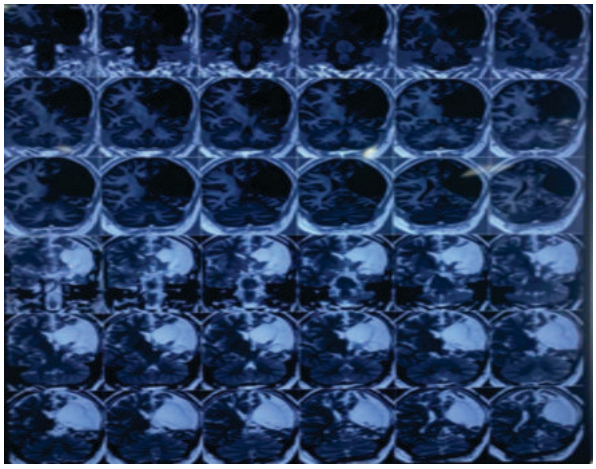
However the progressive loss in dexterity as seen in our case can be explained on the basis of hemispherectomy models, which show that hand movements are more under the control of corticospinal pathways than leg movements. Hemispherectomy has also been traditionally used as a therapeutic principle for intractable seizures in the early stage of this disease. In a way, near-complete hemiatrophy would lead to a similar therapeutic effect, which may explain the decrease in the seizure frequency in our case. In summary, chronic hemiatrophy of the brain in our case perhaps resulted in a 'functional hemispherectomy' leading to this clinical picture.

CONCLUSION

We report a rare case of Dyke Davidoff Masson Syndrome. In cases presenting in early childhood, refractory seizures remain the usual concern. Accordingly, hemispherectomy is the treatment of choice with a success rate of 85% in selected cases. However, if the presentation is late as in our case and if seizures are under control, the patient can be kept on antiepileptic medications, instead of surgery along with supportive therapy including physiotherapy, speech therapy, and occupational therapy. Due to the rarity of this syndrome, it may be easily missed. CT and MRI are powerful imaging modalities to diagnose the pertinent imaging features associated with this syndrome. Knowledge of the clinical presentation, risk factors, and imaging features is therefore indispensable for appropriate patient management. Further longitudinal studies are required to ascertain the natural course of this syndrome especially in an adult population, which would help in planning strategies regarding the time and nature of interventions and management accordingly.







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