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Adult Presentation Of Dyke Davidoff Masson Syndrome At Age Of 35- An Atypical Presentation

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Abstract

Dyke-Davidoff-Masson syndrome (DDMS) is a rare disease which is clinically characterized by hemiparesis, seizures, facial asymmetry, and mental retardation. The classical radiological findings are cerebral hemi-atrophy, calvarial thickening, and hyper-pneumatization of the frontal sinuses. Dyke-Davidoff-Masson Syndrome (DDMS) is a rare condition usually diagnosed in pediatric patients. Adult presentation of DDMS is unusual and has been rarely reported. Here we report a case of 35-year old male who presented to our hospital with generalized tonic clonic seizures and was diagnosed with dyke Davidoff masson syndrome after MRI Brain.

Keywords: NIL

Introduction

Dyke Davidoff Masson syndrome (DDMS) is a rare disease and was first described by C.G. Dyke, L.M. Davidoff, and C.B. Masson in 1933.[1] Dyke-Davidoff-Masson syndrome (DDMS) comprise hemiparesis, seizures, facial asymmetry, and mental retardation. The classical findings including cerebral hemi-atrophy along with calvarial thickening and hyper-pneumatization of the frontal sinuses are only found if an insult to the brain occurs before 3 years of age.[2] Dyke-Davidoff Masson syndrome (DDMS) mainly present in infancy or childhood. However, the clinical presentation as well as the radiological features may be variable depending upon the age and extent of cerebral insult.[3] We report a case of DDMS in a 35-year old male patient who came to our emergency with GTCS and was diagnosed with Dyke-Davidoff Masson syndrome on MRI scan.

Case Report:

A 35-year old male presented to emergency department of our hospital with active generalized tonic clonic seizures (GTCS). Patient was having 6th episode of GTCS in previous 5 hours at time of presentation to hospital. Patient was having history of GTCS from previous 2 years. Patient was on antiepileptic medication from previous 2 years and was not taking medication regularly. At time of presentation patient was having GTCS which stopped after giving midazolam. He was given loading dose of antiepileptic medication. Patient Blood pressure was 126/88 mmhg, pulse rate was 118 beats/minute, temperature was 98.6degree Fahrenheit, SPO2 was 92% on room air, RBS was 128 mg/dl. Patient was non-alcoholic, non-smoker, vegetarian by diet. Patient was married from previous 10 years and was having 2 children (7and 5 years old). Patient was not having any history of allergy. After half an hour patient became conscious and oriented to time place

and person. Patient was not having any weakness in any body part. Power of all limbs was normal and all the reflexes were normal. Patient was illiterate but was not having any mental retardation. Patient was having MMSE score of 25. Cranial nerve examination was normal. Blood and C.S.F studies were normal. Patient was having history of normal antenatal period and also history of ICU admission for 20 days after birth. After that patient was having normal development and all milestones development was at normal time. Figure 3,4 and 5 showing normal development of hands, legs and chest. MRI Brain showed hemi-atrophy of left cerebral hemisphere associated with homo lateral hypertrophy of the calvaria and enlarged ipsilateral frontal and ethmoid sinuses- findings suggestive of dyke Davidoff masson syndrome. Radiological findings are provided in figure 1 and 2. Patient was started on antiepileptic medication. Patient's course during hospital was uneventful. Patient didn't have any further seizure episode during hospital course. Patient was discharged on antiepileptic medication with stable vitals.

Figure 1 and 2 showing- Hemi-atrophy of left cerebral hemisphere associated with homo lateral hypertrophy of the calvaria and enlarged ipsilateral frontal and ethmoid sinuses.

Figure 1

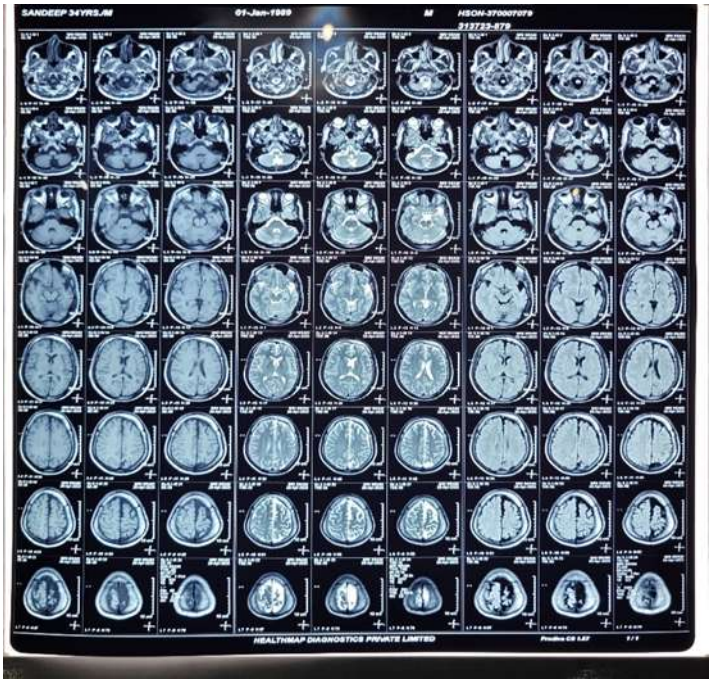

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Figure 2

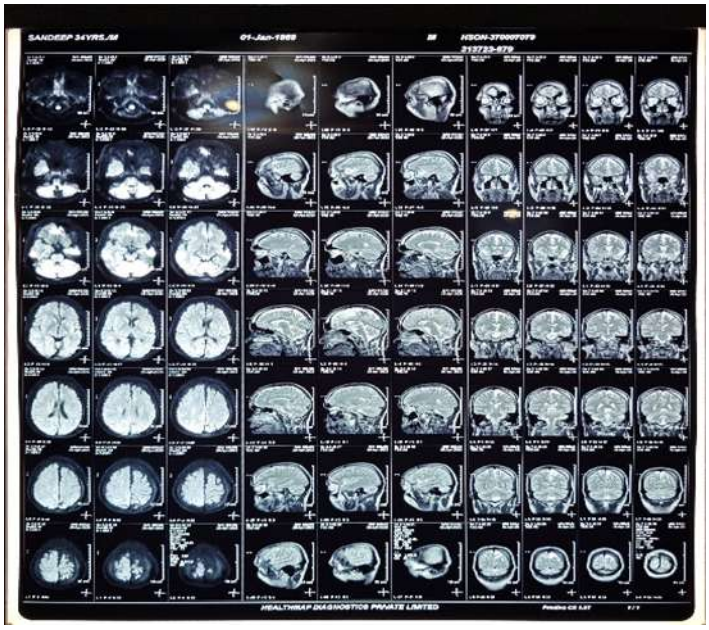

Figure 3 and 4 showing- Normal development of all 4 limbs, chest and abdomen

Figure 3

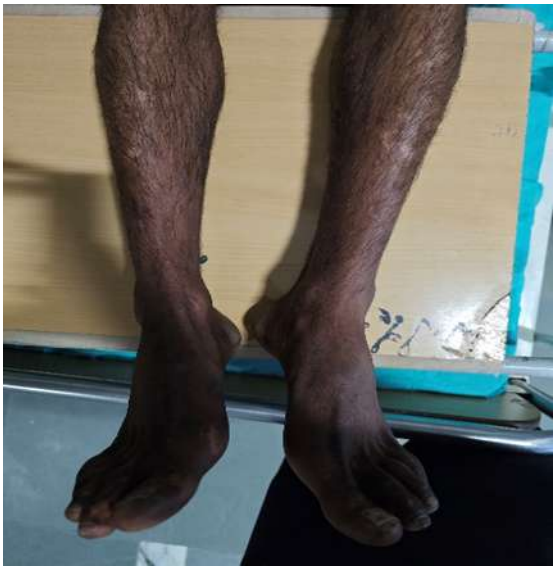

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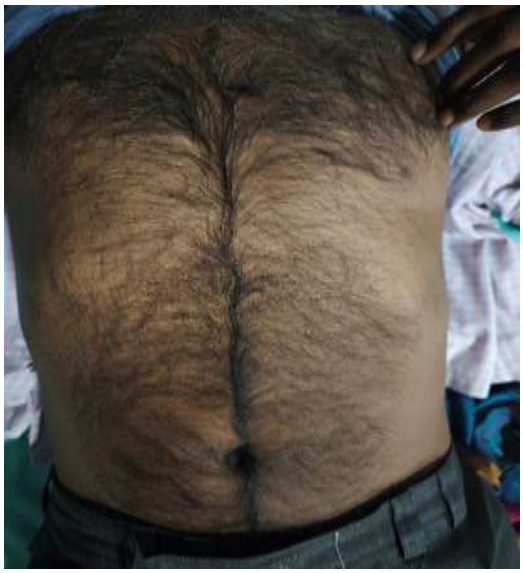
Figure 4

Figure 5



Discussion:

In 1933, C.G. Dyke, L.M. Davidoff, and C.B. Masson reported a series of nine patients with DDMS and showed the following radiological findings: contralateral cerebral atrophy in the form of widening of the sulci and dilatation of the ipsilateral lateral ventricle, thickening of the adjacent calvarium, and hypertrophy of the frontal and ethmoid sinuses.[1] An early study on pediatric hemiplegia showed that imaging findings consistent with DDMS were only apparent in children with weakness manifesting prior to 3 years of age. Thus, pathologies such as cerebrovascular insults occurring during the gestational period up to 3 years of age would most likely have the highest propensity to result in DDMS. Prolonged ischemia during this period can decrease the production of brain-derived neurotrophic factors, resulting in brain atrophy. Subsequently, structures such as the calvarium and the sinuses tend to grow inward to compensate.[4] DDMS presents clinically with a variety of possible features

that may differ in severity depending on the degree of injury. Seizures (both focal and generalized), facial asymmetry, contralateral hemiparesis, and

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intellectual disability are some of the more prominent features. Associated neuropsychiatric symptoms and psychiatric disorders have been described as well.[5] The interesting finding, in this case, was the delay in presentation until a late age, and with no permanent neurological sequelae. To the best of our knowledge, this is one of rare case of DDMS identified in a late adult-aged man with only generalized tonic clonic seizures which started at age of 33 years and with no mental retardation and any kind of weakness. In most cases, DDMS has been identified at a young age. Therefore, it is possible that DDMS can occur even at a late adult age. The differential diagnoses are Sturge–Weber syndrome, basal ganglia germinoma, Fishman syndrome, Silver–Russell syndrome, linear nevus syndrome, and Rasmussen's encephalitis, which can be differentiated by performing a proper history taking, clinical examination and by characteristic neuroimaging features. DDMS usually present as refractory seizures in early childhood. Hemispherectomy has been mentioned as a treatment of choice with a success rate of 85% in selected cases. However, in late adulthood presentation as in our case, if seizures are under control, the patient can be kept on antiepileptic medications instead of surgery, along with supportive physiotherapy, speech therapy, and occupational therapy. Further longitudinal studies are required to document the natural course of this syndrome especially in an adult population, which could help in planning treatment strategies, whether conservative or intervention, in these patients accordingly.[6]

Conclusion:

DDMS are a rare preventable cause of hemiplegia, refractory epilepsy and mental retardation in childhood. Adult presentation of DDMS is unusual and has rarely been reported in medical literature. This study reports a confirmed case of DDMS in the late adult age without permanent neurological sequelae. Radiological findings of our patient are consistent with DDMS which involved the left cerebral hemisphere. MRI played an important role in diagnosis of this patient to delineate the cerebral changes along with changes in clavaria and sinuses. This patient was kept on follow up antiepileptic medication under medicine department and is currently symptom free.

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