International Journal Dental and Medical Sciences Research
Volume 6, Issue 4, July. – Aug. 2024 pp 195-198 www.ijdmsrjournal.com ISSN: 2582-6018

A Rare Case Report on Dyke-Davidoff-Masson Syndrome Variant

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Date of Submission: 20-07-2024 Date of Acceptance: 30-07-2024

ABSTRACT
Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological disorder that results from brain injury in intrauterine or early years of life. Prominent cortical sulci, dilated lateral ventricles, cerebral hemiatrophy, hyperpneumatization of the frontal sinus, and compensatory hypertrophy of the skull are the characteristic findings. Here we describe a female patient who presented with generalized tonic-clonic seizure and left-sided body weakness and facial asymmetry and inability to close jaw and mouth with drooling of saliva from angle of mouth and neuroimaging findings of cerebral hemiatrophy, dilatation of right lateral ventricle, right frontal sinus hyperpneumatization, and asymmetric calvarial thickening. Knowledge of its features on imaging enables timely and accurate diagnosis, allowing appropriate management.

I. INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological condition that was first described by Dyke, Davidoff, and Masson in a series of nine patients with hemiplegia and plain skull Xray changes [1,2]. It is characterized by cerebral hemiatrophy/hypoplasia, facial asymmetry, refractory seizures, and contralateral hemiplegia [1-3], speech and language disorders. These clinical features can present with diverse combinations and severity. Sensory loss and psychiatric manifestations like schizophrenia had been reported rarely (Ono et al., 2003; Amann et al., 2009). Imaging studies are utilized to make a diagnosis in correlation with clinical features. Specific imaging findings include unilateral brain volume loss, ventriculomegaly, and compensatory bone hypertrophy resulting in cerebral hemiatrophy. In addition, calvarial thickening and hyperpneumatization of frontal sinuses may occur [3,4]. As it is a rare disorder, it may be misdiagnosed and consequently mismanaged by the majority of physicians.

II. CASE PRESENTATION

Here we report a case of 29 year old unmarried female first born child of three children, home maker in profession with education qualification upto 4th standard. Patient presented in outpatient department of medicine with complaints of recurrent seizures, drooling of saliva from mouth with inability to close her jaw completely, difficulty in speech and left sided hemiparesis. There was no history of significant antenatal or perinatal complication as she had a normal term delivery. There was no family history in of epilepsy, stroke or any genetic abnormality including the siblings. The developmental milestones were achieved at normal age however higher mental function revealed mild level of mental sub normality. The left sided weakness /hemiparesis was acute in onset non progressive which occurred after a sudden collapse while playing in the ground at the age of 6 years associated with with facial weakness and inability to close her jaw completely and difficulty in speech ever since . Patient also complained of repeated generalized tonic clonic seizures 2 days post the incident and has been having similar episodes since then at a variable interval upto 2-5 episodes per week . Patient did not undergo any treatment or radiological imaging at that time and has not taken any medications for the same but underwent physiotherapy for 3 years after which she could walk without support.

On general physical examination, there was emaciation with decreased cognitive functioning; 15/30 on the Mini-Mental State Examination. On neurological examination, the power was 2/5 on the left upper limb and 3/5 left lower limb of the body with decreased sensations, hypertonia, spasticity and brisk reflexes with positive babinski on left side. We also observed mild left-sided facial angle deviation with inability to close mouth completely with drooling of saliva from angle of mouth with widely spaced teeth and microcephaly with mild left cervical dystonia. Patient showed hemiplegic gait disturbance .The rest of the neurological examination showed no signs of meningeal

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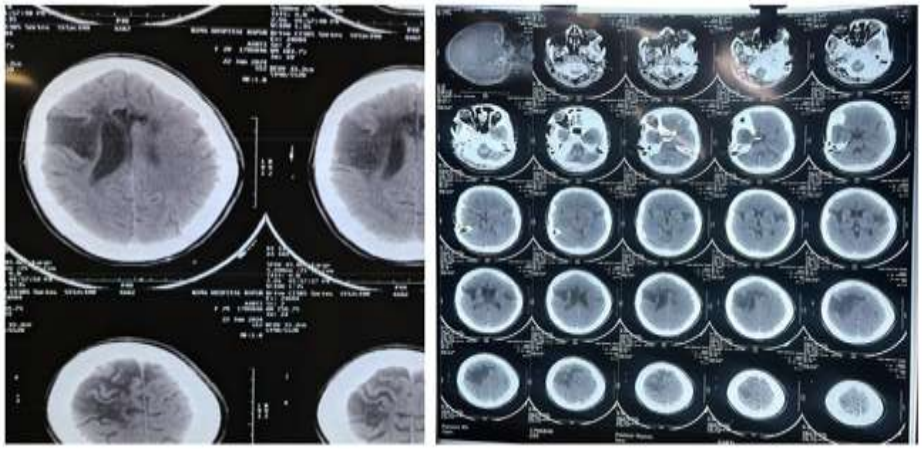
irritation,no cerebellar signs or neurocutaneous markers. Other systemic examination was unremarkable with no significant Respiratory, Cardiac or Abdominal findings. Laboratory workup included baseline investigations and a thorough workup for young stroke along with an autoimmune profile and Cerebrospinal fluid examination. All laboratory investigations were within a normal range. A Fundoscopy was done which revealed no abnormality.

Non contrast CT brain and paranasal sinus revealed hemiatrophy and gliotic changes of the right cerebral hemisphere, with the prominence of extra-axial cerebrospinal fluid (CSF) spaces and dilatation of the ipsilateral right lateral ventricle with pneumatization of right frontal and right maxillary sinusitis with mild right calavarial thickening . An EEG of the patient showed focal as well as generalized slowing of waves suggestive of seizure disorder . These CT findings along with clinical pictures were satisfying the criteria of Dyke-Davidoff-Masson Syndrome (DDMS) and hence the diagnosis was made. The patient was managed conservatively with anticonvulsants levetiracetam and clobazam and once when seizure was controlled, the patient was discharged on levetiracetam and clobazam for seizures glycopyrrolate for continuous drooling of saliva and baclofen for symptomatic relief of spastic hemiplegia. Patient was advised for continuous physiotherapy , speech therapy and occupational therapy for the long run . Patient is now on constant follow up



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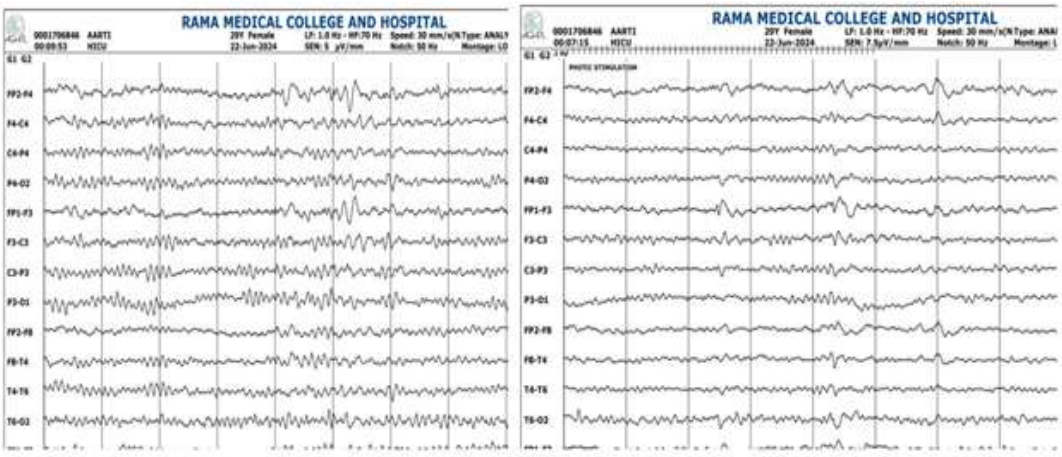

Abnormal awakened stage 1 sleep eeg record suggestive of generalized epilepsy

III. DISCUSSION

In 1933, Dyke, Davidoff, and Masson described the plain skull radiographic and pneumatoencephalo - graphic changes in their series of 9 patients whose clinical characteristics included hemiparesis, seizure s, facialasymmetry, and mental

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retardation(5). It has been reported that Dyke David of Masson Syndrome occur in intrauterine life when the maturation of calvarium has not been completed, or due to brain damage (usually traumatic) occurring in early childhood (6).

The clinical presentation of DDMS include seizures, contralateral hemiparesis of upper motor neuron type disease, facial asymmetry, and cognitive disabilities. DDMS can be classified into two forms depending upon its etiology. The congenital subtype, which becomes symptomatic in infancy, and its pathogenesis include fetal vascular occlusion. The other is the acquired subtype, which presents in childhood. Its etiological factors include perinatal hypoxia, intracranial hemorrhage, infections, cranial trauma, and cerebrovascular lesion [7]. The possible mechanism of cerebral atrophy and the related progressive neuro deficit is hypothesized to be due to several ischemic episodes resulting from these factors, which reduce the production of brain-derived neurotrophic factors, which in turn leads to cerebral atrophy [8]. CT and MRI are the two gold standard imaging modalities that prove to be very significant in the diagnosis of DDMS. These two imaging modalities provide very detailed cross-sectional images. The typical imaging features for DDMS include prominent cortical sulci, dilated lateral ventricles, cerebral hemiatrophy, hyperpneumatization of the frontal sinus, and compensatory hypertrophy of the skull. These imaging findings become more obvious as the patient ages [9]. When the cerebral damage occurs during the intrauterine period or before the age of 3, compensatory calvarial involvement can be seen [10,11]. In a patient with cerebral hemiatrophy, the differential diagnosis includes Rasmussen encephalitis, SturgeWeber syndrome, basal ganglia dysgerminoma, Fisherman syndrome, and Silver-Russell syndrome. Detailed history and examination are required along with laboratory and imaging workup to differentiate these diseases. For the management of seizures, mono, or poly anticonvulsant medication is given. Children with refractory epilepsy and hemiplegia are potential candidates for hemispherectomy, with a success rate of 85%. Vagal stimulation is another alternative. Despite lacking any specific treatment algorithm, therapy with antiepileptics and surgery is indicated in specific cases. Supportive therapy including physiotherapy, speech therapy, and occupational therapy are needed. 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. The case is discussed to draw attention on a syndrome which can be managed by a holistic approach.

IV. CONCLUSION

The disease, was supposed to be rare once, is increasingly reported now, may be because of the better neonatal resuscitation facilities and better diagnostic modalities. Patients may present with variations in clicical presentation like in this patient with major facial asymmetry. A Detailed history and examination with imaging modalities help to diagnose it. Physicians should be aware of signs and symptoms, risk factors, and diagnostic features of DDMS so that the patients could be diagnosed early and managed properly. Timely referal to a neurosurgeon in refractory seizures for hemispherectomy has better success rates.

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