CASE REPORT

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Dyke-Davidoff-Masson Syndrome (DDMS) in an Adult

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Abstract

Dyke-Davidoff-Masson Syndrome (DDMS), also called cerebral hemiatrophy, is a rare disease which clinically presents as hemiparesis, seizure, facial asymmetry and mild mental retardation. The classical radiological findings are cerebral hemiatrophy, calvarial thickening, and hyperpneumatisation of the frontal sinuses. This disease is a rare entity and it mainly presents in childhood. Adult presentation of DDMS is unusual and has been rarely reported in the medical literature. However, it should be kept in mind as a diagnostic possibility in an adult who presents with a long duration of progressive hemiparesis with seizures. We are presenting a 22-year-old patient who presented clinically with generalised tonic clonic seizures, right hemiparesis, right-sided limb deformity, facial asymmetry with a typical magnetic resonance image finding suggestive of DDMS.

Key words: Dyke-Davidoff-Masson Syndrome (DDMS), cerebral hemiatrophy, hemiparesis.

Introduction

Dyke-Davidoff-Masson Syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere which is usually due to an injury to the developing brain in infancy or early childhood[1]. Dyke *at el* first described this in a series of nine patients with plain skull radiographic and pneumatoencephalographic changes, in the year 1933[2,3]. The features of this disease vary and depend upon the severity of the assault on the brain. However, the characteristic clinical findings include contralateral hemiparesis, facial asymmetry, recurrent seizures, mental retardation, learning disabilities, speech, and language disorders. Rarely, it may present with psychiatric manifestations such as schizophrenia[4]. Radiological findings include cerebral hemiatrophy with compensatory thickening of the skull vault, enlargement of frontal sinuses (may also include ethmoid and maxillary sinus), elevation of petrous ridge, ipsilateral falcine displacement, and capillary malformations. Herein, we are reporting a case of DDMS in a 22-year-old male with classic clinical and radiological findings.

Case report

A 22-year-old male patient of low socio-economic status presented in MB Government Hospital and RNT Medical College, Udaipur, (Rajasthan) with a history of right-sided hemiparesis and deformity of the right upper and lower limbs with facial asymmetry since 20 years (2 years of age) and from last 2 years he had recurrent episodes of tonic-clonic seizures, at least 4 to 5 times in a month. He was born of a non-consanguinous marriage. Birth history was indicative of a full-term normal delivery without any antenatal, perinatal and post-natal complications. He had normal developmental milestones during infancy and up to the second year of life. He was moderately built, conscious and oriented (Fig. 1). On neurological examination, we discovered that he had right hemiparesis (power 4/5 in right upper and lower limbs at all joints) with right plantar extensor and brisk tendon reflexes on the right side. On mental function examination he was mildly mentally retarded (IQ = 60). All other neurological examination including cranial nerve, sensory system, bowel and bladder involvement and signs of meningeal irritation were normal. No neurocutaneous markers were present. All routine blood investigations and electroencephalography were normal. A magnetic resonance imaging (MRI) of the brain was subsequently done which revealed an area of gliosis in left cerebral hemisphere, predominantly in the temporal and parietal lobes with cystic encephalomalacia and *ex-vacuo* dilatation of the left lateral ventricle (Fig. 2). There was also thickening of the skull vault on left side with over-pneumatisation of the frontal sinus (Fig. 3). On clinical and radiological findings, we kept a diagnosis DDMS and managed him conservatively with anti-epileptics, muscle relaxants and physiotherapy. Subsequently, his symptoms improved.

Discussion

During development, the human brain reaches half of its adult size during the 1st year of life and by the end of 3 years, brain attains three-fourths of the size of adult brain. All of the important sulci begin to appear by the end of 8th

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Fig. 1: *Right hemiparesis with deformities in the right extremities.*

gestational month[5]. The developing brain presses outward on the encasing bony skull resulting in gradual increase in size and shape of head. When the brain stops growing,

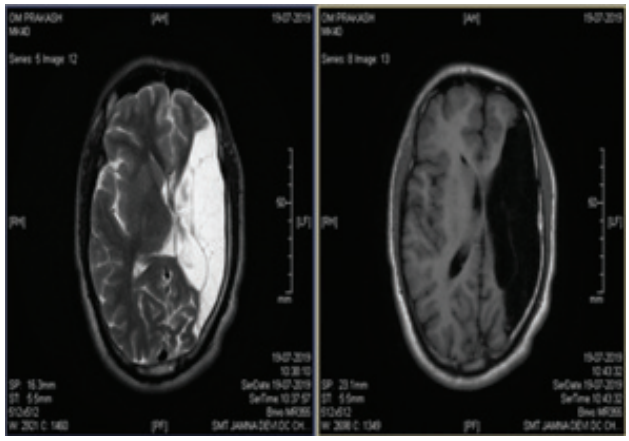

Fig. 2: *MRI brain showing an area of gliosis of left cerebral hemisphere in T1W and T2W images.*

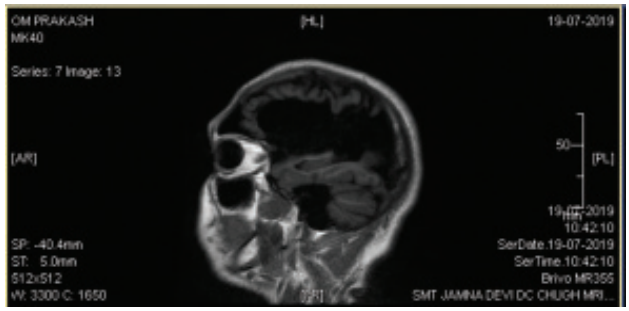

Fig. 3: *MRI brain showing hyperpneumatisation of frontal and maxillary sinuses.*

other surrounding structures grow inward resulting in increased width of diplopic space, enlarged sinuses and elevated orbital roof[6]. These changes are prominent when trauma is inflicted to brain before the age of 3 years. However, the changes become evident generally after 9 months of trauma[7].

Cerebral hemiatrophy can be of two types- Congenital/ Infantile and acquired. The causes of congenital cerebral atrophy are infections, neonatal or gestational vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial anomalies and coarctation of mid-aortic arch[8,9]. Such patients become symptomatic in infancy or perinatal period. The main causes of the acquired form include trauma, malignancy, infection, ischaemia, haemorrhage and prolonged febrile seizures. The time of presentation depends on the time of the injury.

DDMS is characterised by atrophy of the cerebral hemisphere on one side leading to ipsilateral osseous hypertrophy with hyperpneumatisation of sinuses, mainly frontal and mastoid air sinuses with contralateral hemiparesis, enlargement of ipsilateral sulci and dilated

ipsilateral ventricle due to atrophy of the brain parenchyma. Clinically, patient presents with hemiparesis, mental retardation and focal or generalised seizure. It does not have any gender or side predilection. However, male gender with left cerebral involvement is more common[10].

A proper history and thorough clinical examination along with radiological findings helps to give the correct diagnosis in this condition. However, there are other conditions which may mimic the findings of DDMS and may cause error in diagnosis of this syndrome. Hence, this should be differentiated from clinical conditions such as basal ganglia germinoma, Sturge-Weber Syndrome, Silver–Russel syndrome, etc.[11,12].

Sturge-Weber syndrome presents with cerebral atrophy along with leptomeningeal angioma. The differentiating features are port-wine facial nevus, tram-track cortical and subcortical calcification and absence of midline shift.

Silver-Russel syndrome is differentiated by its characteristic facial appearance like triangular face, broad forehead, small pointed chin and thin-wide mouth. Normal intelligence and hemihypertrophy are other distinguishing features.

Treatment of DDMS primarily aims at controlling seizures with anticonvulsants. Hemispherectomy gives a good result when the patient has intractable disabling seizures and hemiplegia with a success rate of 85%[13]. Physiotherapy, occupational therapy, and speech therapy are also given, along with drugs, on a long-term basis. Prognosis of this condition is better when the disease starts after 2 years of age or in the absence of intractable seizures.

Conclusion

The Dyke-Davidoff-Masson Syndrome (DDMS) is a rare epilepsy syndrome characterised by convulsions, contralateral hemiplegia, mental retardation and hemiatrophy of the brain. Although there are many close differential diagnoses of this case, a proper clinical history and MRI findings are enough for the correct diagnosis. It is one of the most preventable causes of refractory epilepsy. 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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