Case Report

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Dyke-Davidoff-Masson Syndrome (DDMS): A rare preventable cause of refractory epilepsy

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ABSTRACT

Dyke-Davidoff-Masson Syndrome (DDMS), which is also called cerebral hemiatrophy, is a rare disease which clinically presents itself with contralateral spastic hemiplegia or hemiparesis, seizures, facial asymmetry, and mental retardation. The classical radiological findings include cerebral hemiatrophy, calvarial thickening, and hyperpneumatization of the frontal sinuses. This disease is a rare entity, and it usually appears in childhood. Adult presentation of DDMS is unusual and has not often been reported in medical literature. This case illustrates typical computed tomographic features of DDMS in an 11-year-old boy who presented with recurrent generalized seizures since childhood, left hemiparesis with mental retardation.

KEY WORDS: Cerebral hemiatrophy, Dyke-Davidoff-Masson Syndrome, epilepsy, rasmussen encephalitis, Sturge-Weber syndrome

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 *et al*., first described this case 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 to the brain; however, the characteristic clinical findings include hemiparesis, facial asymmetry, recurrent seizures, mental retardation or 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 an 11-year-old male with classic radiological findings.

CASE REPORT

An 11-year-old male patient of low socioeconomic status was presented to us. He had a history of recurrent generalized seizures from his early childhood. His first episode of seizure started at about two years of age. He was born of third order full-term normal vaginal delivery, to non-consanguineous parents. His antenatal and perinatal history was uneventful. His other two siblings are normal. He was mentally retarded (intelligent quotient = 62) and stopped schooling at 6 years of age. There was no history of any serious childhood infections or trauma. He began exhibiting abnormal behaviors, with inappropriate emotions and laughter about six years, along with left-sided weakness. He was on antiepileptic drugs since he was two years old, but his seizures were poorly controlled.

He was moderately built, conscious, and oriented. On having his neurological examination, we discovered that he had mild left extremity hemiparesis (power 4/5 in left upper and lower limb) with left plantar extensor and brisk tendon reflexes. There was no neck rigidity, sensory deficit, cranial nerve palsy, or bowel and bladder involvement. No neurocutaneous markers were present. Other systemic examinations revealed no findings. All routine blood investigations were essentially normal.

A computed tomography of brain showed right hemispheric volume loss with gliotic changes, with hypertrophy of ipsilateral skull vault, hyperpneumatisation of paranasal sinuses and mastoid cells, dilatation of ipsilateral lateral ventricle with

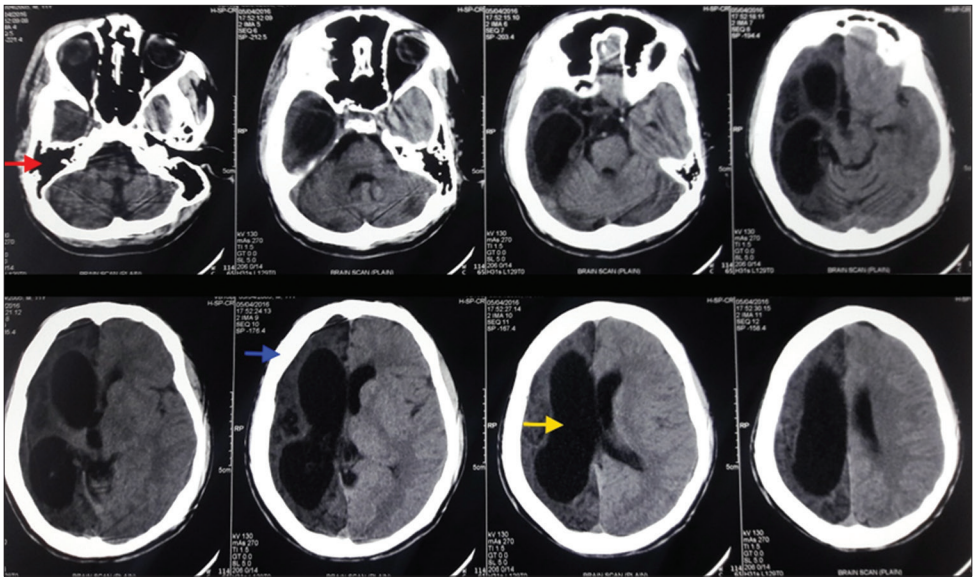

Figure 1: Computed tomography scan showing enlargement of right ethmoid sinus, mastoid air cells (red arrow), calvarial thickening (blue arrow) right hemispheric volume loss and gliosis, and right lateral ventricle dilatation (yellow arrow)

falcine shift which are characteristic findings of Dyke-Davidoff-Masson syndrome [Figure 1].

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, the brain attains three-fourth of the size of adult brain. The surface of the brain remains smooth until 3 months of gestational life. All of the important sulci begin to appear by the end of 8th 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, 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 midaortic arch [8,9]. Such patients become symptomatic in infancy or perinatal period. The main causes of acquired form include trauma, malignancy, infection, ischemia, hemorrhage, and prolonged febrile seizures. The time of presentation depends on the time of the injury.

DDMS is characterized by atrophy of cerebral hemisphere on one side leading to ipsilateral osseous hypertrophy with hyperpneumatization 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 generalized seizure. It does not have any sex or side predilection, it can occur in both sexes and any side of cerebrum; however, male gender with left cerebral involvement is more common [10].

A proper history, thorough clinical examination along with radiological findings, helps give the correct diagnosis of 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, Linear Nevus syndrome, Fishman syndrome, and Rasmussen encephalitis. [11,12].

Sturge-Weber syndrome presents itself 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 (triangular face, broad forehead, small pointed chin, and thin-wide mouth). Normal intelligence and hemihypertrophy are other distinguishing features.

Fishman syndrome (also known as encephalocraniocutaneous lipomatosis) presents itself with unilateral cranial lipoma and lipodermoid of eye along with calcified cortex and hemiatrophy.

Rasmussen encephalitis is an immune-mediated condition secondary to viral infection presenting with intractable focal epilepsy and cognitive defect in children. The main differentiating feature is hemiatrophy of brain without any calvarial changes.

Linear Nevus syndrome can be differentiated by its typical facial nevus, mental retardation, and seizures.

Treatment of this condition primarily aims at controlling seizures with anticonvulsants. Hemispherectomy gives a good result when the patient has intractable disabling seizure and hemiplegia with a success rate of 85% [13]. Physiotherapy, occupational therapy, and speech therapy are also given along with the 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 is a rare epilepsy syndrome characterized by convulsions, contralateral hemiplegia, mental retardation, and hemiatrophy of the brain. Although there are Although there are many close differential diagnoses of this case, a proper clinical history and CT findings are enough for the correct diagnosis. Since perinatal hypoxic injury is one of the causative factors, proper obstetric care is vital to prevent such conditions from occurring. It is one of the most preventable causes of refractory epilepsy.

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