CASE REPORT

DYKE-DAVIDOFF-MASSON SYNDROME

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ABSTRACT

Dyke-Davidoff-Masson Syndrome (DDMS) is characterized by Seizures, facial asymmetry, contralateral hemiplegia and Mental Retardation. Characteristic Radiological features are Cerebral hemiatrophy with homolateral hypertrophy of skull and sinuses? We report a Case of DDMS in a 21yr old male who presented with Generalised Tonic Clonic Seizures, Hemiparesis of the left hand and leg with deformity of the left upper limb and left lower limb and also deviation of the mouth to left.

Keywords: Hemiatrophy, Hemiparesis, Seizure

INTRODUCTION

First description of Dyke-Davidoff-Masson syndrome (DDMS) dates back to 1933, when Dyke, Davidoff andMasson described the plain skull radiographic and pneumatoencephalographic changes in a series of nine patients[1]. Since then only few pediatric cases of DDMS have been reported

CASE REPORT

A 21yr old Male, born full term through normal vaginal delivery presented with recurrent Generalised Tonic Clonic Seizures from 5yrs, there was no H/o significant antenatal or perinatal complication. The present complaint started at the age of 3 yrs when the boy suddenly had vomitings and loose stools following which he developed Weakness of left upper and lower limb.

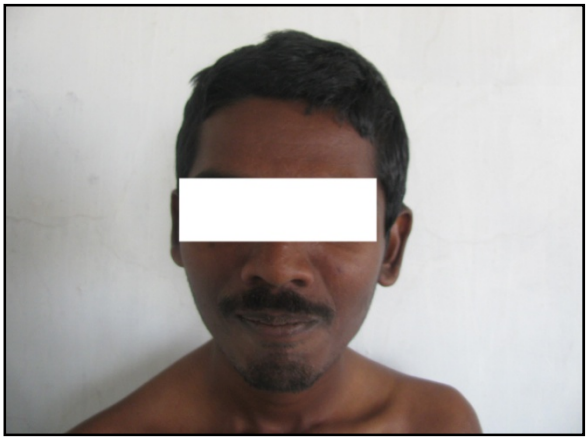

Figure 1: Facial Deviation to Left

Neurological examination

There is asymmetry of face and body and no neurocutaneous marker was seen. Cranial Nerves: Right Upper Motor Neuron facial palsy present (figure -1). Left sided spastic hemiparesis (figure - 2) All routine investigations and Hematological profile - normal. Cerebro Spinal Fluid analysis was normal.

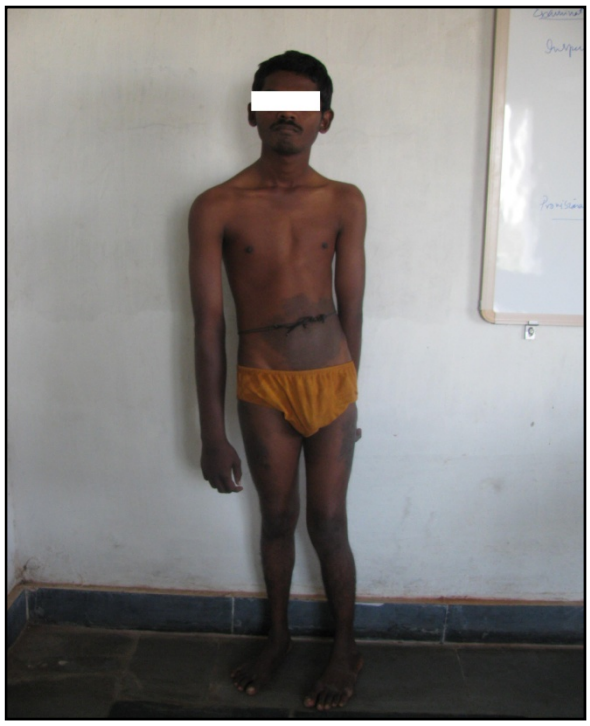

Figure 2: Left sided Hemiparesis

CT scan of Brain

Dilatation of Right lateral ventricle sinuses and right mastoid air cells (figure - 3) and excessive Pneumatization

of right sphenoid and right ethmoid and right cerebral sulcus spaces (figure - 4) was seen.

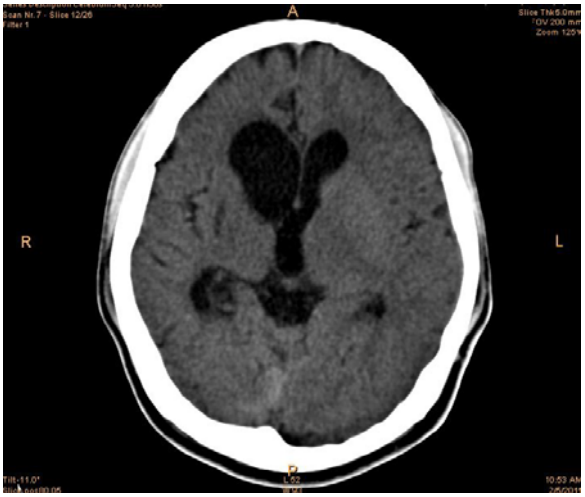

Figure 3: CT scan of Brain: Dilatation of right lateral ventricle sinuses and right mastoid air cells

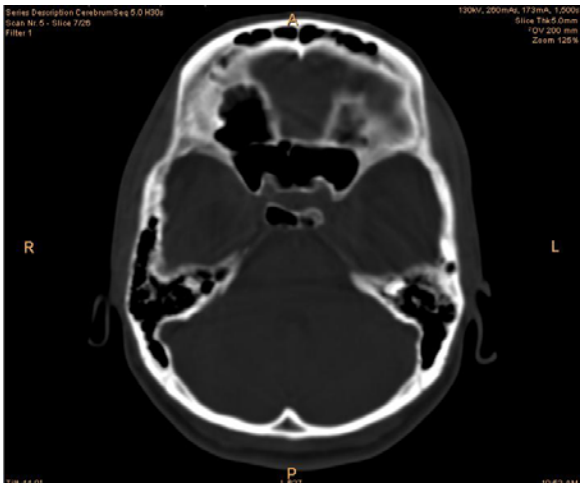

Figure 4: Excessive Pneumatization of right sphenoid, right ethmoid and right cerebral sulcus spaces

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, facial-asymmetry, and mental retardation1. 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 first 3 years of life 2.

Hemiatrophy of one cerebral hemisphere is not frequently encountered in clinical practice. When this develops early in life (during the first two years) certain cranial changes like homolateral hypertrophy of the skull and sinuses occur. The compensatory cranial changes occur to take up the relative vacuum created by the hyppoplastic cerebrum. The classical clinical presentation includes seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation. However mental retardation was not always present and seizures may appear months or years after the onset of hemiparesis[3].The clinical findings may be of variable degree according to the extent of the brain injury. Imaging studies show unilateral loss of volume of brain and calvarial changes, finding of cerebral atrophy, ventricular dilatatition and enlargement of sulci [4].

In congenital hemiatrophy, when the insult occurs in utero, there is shift of midline structures towards the side of the disease and the sulcal prominence replacing the gliotic tissue is absent [5]. This feature differentiates it from cerebral hemiatrophy which occurs in early life. The etiological factor for Dyke-Davidoff-Masson syndrome has been postulated as trauma, inflammation or vascular malformations and occlusions. When the insult occurs in-utero, it could be due to gestational vascular occlusion, primarily involving the middle cerebral vascular territory. A possible etiological relation of cerebral hemiatrophy and seizures has been reported by different studiesin India [6, 7].

Dyke Davidoff Masson Syndrome should be differentiated from Basal cell germinoma, Sturge Weber syndrome, Linear Nevus syndrome, Fishman syndrome, Silver-Russell syndrome and Rasmussen encephalitis [8]. A proper clinical history and CT/MRI findings provide the correct diagnosis

The treatment is symptomatic, and should target convulsion, hemiplegia, hemiparesis and learning difficulties. Prognosis is better if hemiparesis occurs after the age of 2 years and in absence of prolonged or recurrent seizures. Children with intractable disabling and hemiplegia are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases [9].

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