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CASE STUDY

DYKE-DAVIDOFF-MASSON SYNDROME: RADIOLOGICAL IMAGING FINDINGS

***Dr. Holebasu, Dr. Bhushita Lakhkar and Dr. Karthik Adiraju**

Shri B M Patil Medical College, Vijayapur, India

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ABSTRACT

Dyke-Davidoff-Masson Syndrome (DDMS) is seldom seen in clinical practice. It is characterized by cerebral hemi atrophy with ipsilateral hypertrophy of skull and sinuses. We report two cases of DDMS, A 11yr old male who presented with generalised tonic clonic seizures and mental retardation. Another case of 17 yr old male who presented with seizures, hemiparesis of the right hand and leg with deformity of the right upper limb and right lower limb.

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INTRODUCTION

The Dyke-Davidoff-Masson Syndrome (DDMS) is defined/ characterized by/consists of as the atrophy of one cerebral hemisphere secondary to brain insult such as infarct, trauma or infection in utero or early childhood period. Due to lack of brain growth the calvaria and diploic space are thickened while paranasal sinuses and mastoids become enlarged and hyperaerated.

Case Report

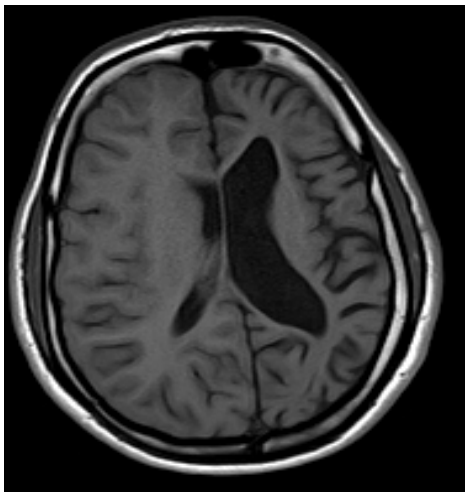
A 11-year-old boy was referred for CT-scan with history of generalised tonic clonic seizures since 5 yrs and mental retardation. There was no clear history of perinatal and antenatal complications. A 17 yr old male presented with seizures, hemiparesis and deformity of the right upper & lower limb referred for MRI-Brain. Prominent cerebral sulci, sylvian fissure, ventricles and cerebellar folliae are noted in left cerebral hemisphere predominantly in left parieto-occipital lobes with focal cortical atrophy in left parietal lobe. Adjoining skull vault was normal in thickness with no evidence of falcine herniation, gyral calcification and obvious vascular anomalies. Hyper-Pneumatization of the left frontal sinus was noted.

DISCUSSION

DDMS is a rare condition characterized by varying degree of facial asymmetry, seizures, contralateral hemiparesis, mental retardation and learning disabilities with behavioural abnormalities (Goyal, 2009). Clinical features vary depending on the extent of brain injury. A detailed history, diligent clinical examination with radiologic findings provide clue to the diagnosis. The disease is generally classified into primary (congenital) and secondary (acquired) variety (Kumar, 2011). Primary variety is mainly caused due to vascular occlusions or malformations in-utero or in the neonatal period. Neonatal or gestational vascular occlusion involving the middle cerebral vascular territory, unilateral cerebral arterial circulation anomalies, coarctation of the aortic arch, midbrain hypoplasia and Wallerian degeneration have been considered as some of the causes. Congenital type of DDMS, in contrast to adult (acquired) DDMS, shows enlargement of calvarium, diploic space and paranasal sinuses. These compensatory cranial changes occur to take up the relative vacuum created by the atrophied cerebral hemisphere (Atalar, 2007 and Kochar, 2001). Acquired variety may be due to infections, trauma, ischemia and haemorrhage. Age of presentation depends on the time of occurrence of the brain insult and often clinical features may not be evident till adolescence (Lin, 2010). The characteristic calvarial changes may or may not be present depending upon the time of injury. Further, these insults occur in post natal period, after completion of sulci formation, however, mental retardation remains unexplained in the acquired variety. The exact mechanism of cerebral atrophy is still unclear in both the types. It is hypothesized that ischemic episodes from a variety of different causes reduce the production of brain derived neurotrophic factors, which results in cerebral atrophy.

****Corresponding author: Dr. Holebasu,***
Shri B M Patil Medical College, Vijayapur, India

CT-SCAN AND MRI BRAIN RESULTS



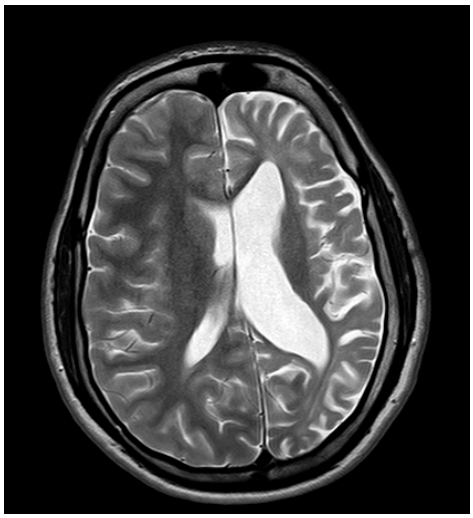


Fig 1 T1W image and Fig 2 T2W image of axial section MRI brain shows left cerebral hemiatrophy with dilatation of the left lateral ventricle

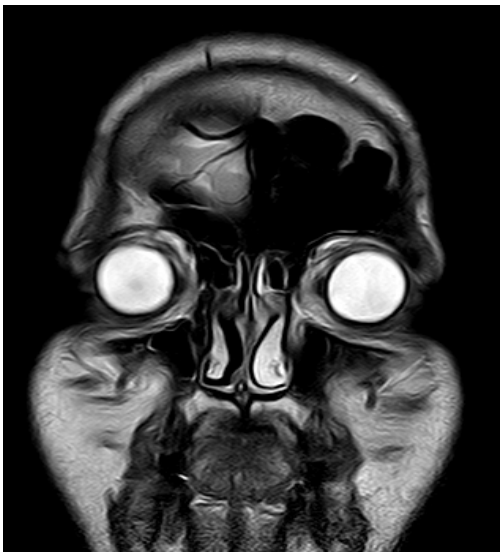


Fig. 3. T2W coronal image showing hyperpneumatization of left frontal sinus

Shen *et al*. (1993) depicted three MR imaging patterns of cerebral hemiatrophy:

MR imaging pattern I corresponds to diffuse cortical and subcortical atrophy; pattern II corresponds to diffuse cortical atrophy coupled with porencephalic cysts; and pattern III corresponds to previous infarction with gliosis in the middle cerebral artery (MCA) territory. In our case, pattern I was present. The atrophied cerebral hemisphere will have prominent sulcal spaces if the vascular insult occurs after birth or after end of sulcation. However, if ischemia occurs during embryogenesis when the formation of gyri and sulci is deficient, prominent sulcal spaces will be absent (Zilkha, 1980). The condition needs to be differentiated from Sturge Weber syndrome, basal ganglia germinoma, Linear nevus syndrome, Fishman syndrome, Silver-Russell syndrome and Rasmussen encephalitis (Rao, 1999). The treatment is symptomatic, includes management of convulsion, hemiparesis learning difficulties, physiotherapy and rehabilitation. 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 (Naraian, 2008).

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