

Dyke Davidoff Masson syndrome in Children

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Citation

J Goyal, V Shah, S Rao, N Jindal. *Dyke Davidoff Masson syndrome in Children*. The Internet Journal of Pediatrics and Neonatology. 2008 Volume 10 Number 2.

Abstract

Cerebral hemiatrophy is not frequently encountered in clinical practice in children. We present here a case of 4 years old female child, who presented with seizures and weakness of left upper and lower limb and on MRI diagnosed to have Dyke Davidoff Masson Syndrome (DDMS).

INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy) which is secondary to brain insult in fetal or early childhood period. It is not frequently encountered in clinical practice in children. We present here a case of a 4 year old female child, who presented with seizures and weakness of left upper and lower limb and on MRI was diagnosed to have DDMS.

CASE REPORT

A 4 years old female child was admitted to our hospital with complaints of weakness of left side of body for 2 years. Weakness was preceded by fever (indicating inflammatory etiology) and left sided recurrent tonic seizures which were relieved by anticonvulsant (carbamazepine). Since then the child had the same type of seizures even on regular medication. There was no history of significant perinatal and antenatal complications. Neurological examination revealed left sided spastic hemiparesis, brisk tendon reflexes and extensor planter on left side. There was no history of delayed mile stones. There were no neurocutaneous markers or asymmetry of face. Head circumference was 44 cm (<3rd percentile). Vision and hearing were normal and cranial nerves were intact. Hematological and CSF examination was normal. MRI of the patient showed severe atrophy of cerebral white matter on right side, thinning of cortical gyri and ex vacuo dilatation of right lateral ventricle. Atrophy of the right cerebral peduncle and thickening of right sided calvarium was also noted (Fig 1 & 2).

Figure 1

Fig 1: FLAIR coronal section of MRI

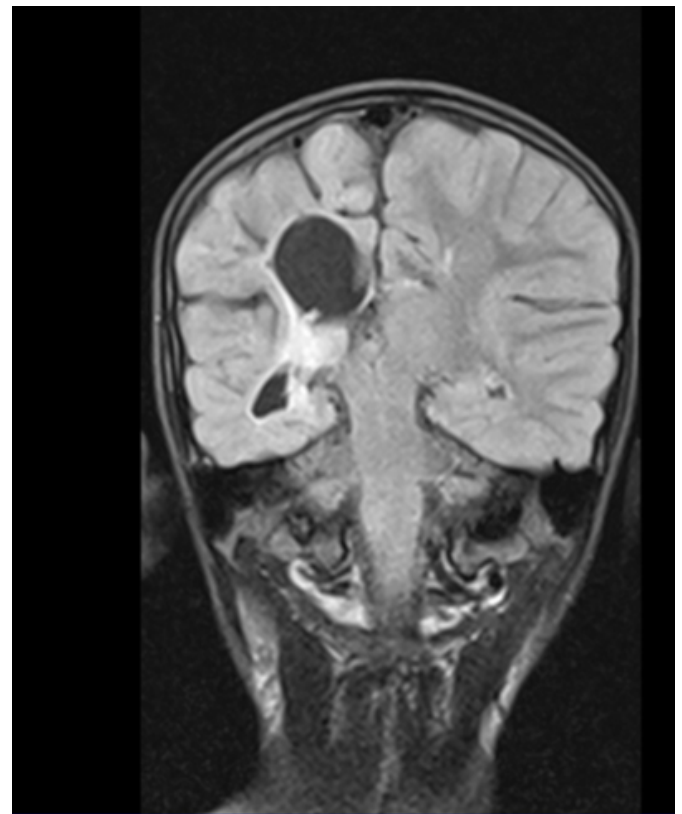
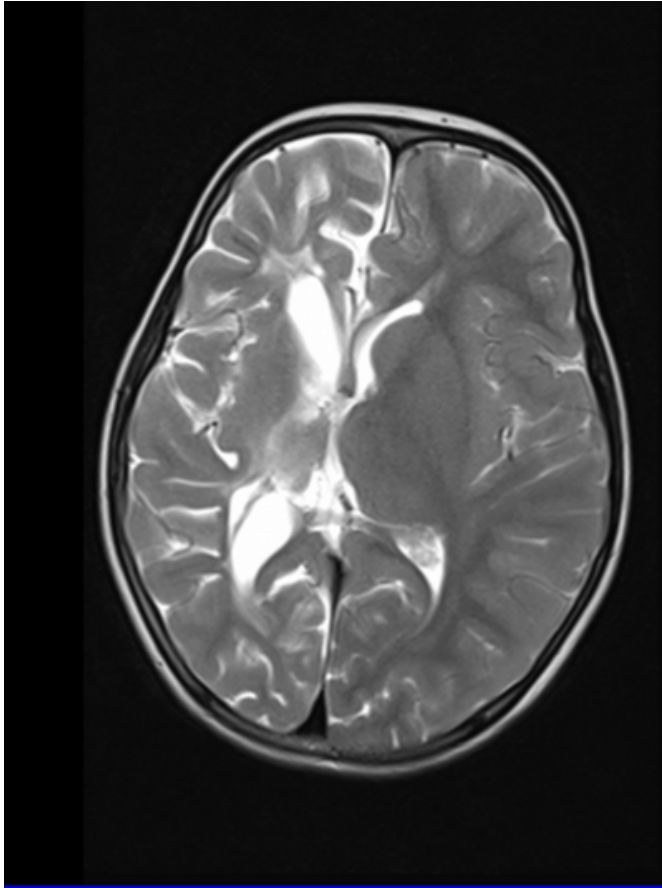


Figure 2

Fig 2: T2 axial section of MRI



DISCUSSION

In 1993, Dyke, Davidoff, and Masson described the plain skull radiographic and pneumatoencephalographic changes in their series of 9 patients whose clinical characteristics included hemiparesis, seizures, facial-asymmetry, and mental retardation (1). It has been reported that DDMS 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 hypoplastic 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 dilatation 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 studies in India (6, 7).

DDMS 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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