ISSN : 2350-0743

www.ijramr.com

International Journal of Recent Advances in Multidisciplinary Research
Vol. 02, Issue 11, pp.0970-0972, November, 2015

CASE REPORT

DYKE DAVIDOFF MASSON SYNDROME: CASE REPORT

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ARTICLE INFO

Article History:
Received 17th, August 2015
Received in revised form
10th, September 2015
Accepted 15th, October 2015
Published online 30th, November 2015

Keywords:
Seizures,
Hemiparesis,
Hemiatrophy.

ABSTRACT

Dyke- Davidoff- Masson syndrome [DDMS] is a rare clinical entity characterized by seizures , facial asymmetry, contralateral hemiparesis and mental retardation. The radiological features are cerebral hemiatrophy with homolateral hypertrophy of skull and sinuses. We are reporting a case of DDMS in a 2 years old female child who presented with right sided weakness , occasional episodes of seizures and delayed milestones.

INTRODUCTION

Dyke Davidoff Masson or cerebral hemiatrophy is congenital, neonatal or early infantile condition characterisd by cerebral hemiatrophy, facial asymmetry, thickening or thinning of cranial vault, contralateral hemiplegia or hemiparesis, seizures, mental retardation, and behavioural changes like schizophrenia (Sharma *et al.,* 2006; Pendse *et al.,* 2004; Shetty *et al.,* 2003). These features are present in varying combinations and variable degree of severity. Mental retardation is not always present (Parker *et al.,* 1972; Sener, 1992; Zilkha, 1980) and seizures may appear months or years after the onset of hemiparesis (Zilkha, 1980). Patients may also have speech or language disorders (Parker *et al.,* 1972). Diagnosis is usually achieved by clinical examination and radiologic investigation.

Radiologically, magnetic resonance (MR) and computed tomography (CT) demonstrate the parenchymal abnormalities of unilateral loss of cerebral volume and compensatory bone alterations in the calvarium, such as thickening, hyperpneumatization of the paranasal sinuses and mastoid cells as well as elevation of the petrous ridge and greater wing of the sphenoid bone (Sener, 1992; Zilkha, 1980). First description of Dyke-Davidoff-Masson syndrome (DDMS) dates back to 1933, when Dyke, Davidoff and Masson described the plain skull radiographic and pneumatoencephalographic changes in a series of nine patients (Dyke *et al.,* 1933). Here we are reporting a case of 2 year old female child presented with clinical and radiological features suggestive of DDMS.

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CASE REPORT

A 2 year old female child presented in neurosurgery department with complaints of weakness of right upper and lower limbs since birth and occasional attacks of seizures. There was no history of antenatal or perinatal complications. However, there was history of significant delay in milestones. On clinical examination, there was right-sided spastic hemiparesis, brisk deep tendon reflexes, and extensor plantar response. There was no neurocutaneous marker or asymmetry of face or body and head circumference was normal. Vision and hearing were normal and cranial nerves were intact.

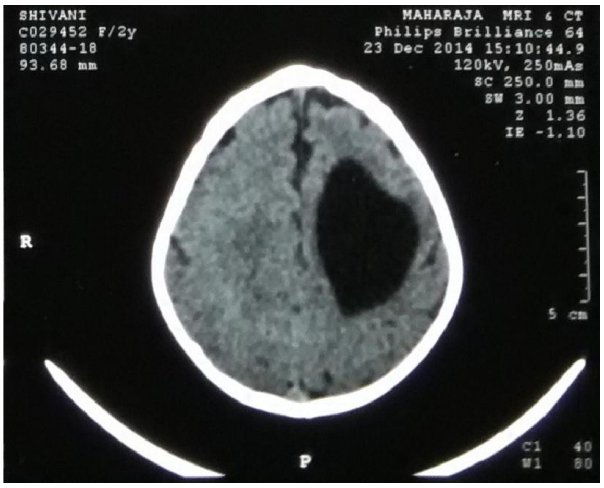

Fig. 1. CT scan of head showing hemiatrophy of left cerebral hemisphere, dilatation of left lateral ventricle and widening of ipsilateral sulci

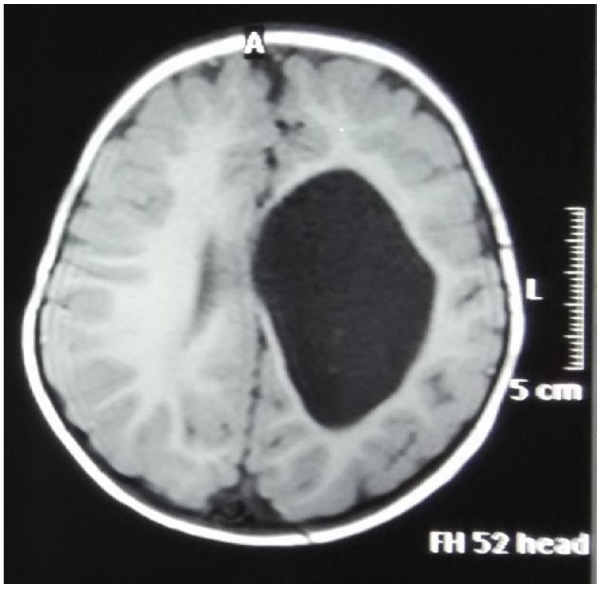

Fig. 2. MRI brain of the same patient showing asymmetrical dilatation of left lateral ventricle with mild atrophy of left cerebral hemisphere, Sulci in left frontoparietal region are prominent

Hematological profile and cerebrospinal fluid examination were normal. Computed Tomography (CT) scan of head (Fig. 1.) revealed hemiatrophy of left cerebral hemisphere, dilatation of the left lateral ventricle, and widening of ipsilateral sulci . MRI of brain (Fig. 2.) also showed similar findings and thus diagnosis of Dyke Davidoff Masson Syndrome was made. Thereafter, cystoperitoneal shunt surgery was performed. Postoperative CT head showed significant reduction in size of left lateral cyst.

DISCUSSION

DDMS is a rare condition characterized clinically by varying degrees of facial asymmetry, seizures, contralateral hemiparesis, mental retardation and learning disabilities with behavioural abnormalities (Shetty *et al.,* 2003; Narain *et al.,* 2008). The radiological findings include cerebral hemiatrophy, ipsilateral osseous hypertrophy and hyperpneumatization of sinuses (Narain *et al.,* 2008). Either sex may be affected with involvement of any cerebral hemisphere, however the male sex and left hemispherical involvement is seen more frequently (Unal *et al.,* 2004).

Clinical features vary depending on the extent of brain injury. A detailed history, meticulous clinical examination with radiologic findings provide clue to the diagnosis. The disease is generally classified into Infantile (congenital) and Acquired variety (Pendse *et al.,* 2004). Congenital 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 midaortic arch, mesencephalon hypoplasia and Wallerian degeneration have been propounded as some of the etiologies for the congenital variety (Sharma *et al.,* 2006; Pendse *et al.,* 2004; Goyal *et al.,* 2009). When the cerebral hemiatrophy develops in-utero or during first two years of life, it is associated with certain cranial changes like ipsilateral hypertrophy of the skull and sinuses as a compensatory change to take up the relative vacuum created by the hypoplastic cerebrum (Goyal *et al.,* 2009). The brain reaches half the adult size during first year of life and three-fourths by the end of third year. As it enlarges, the brain presses outward on the bony tables which gradually results in general shape of the adult head. But, failure of the cerebrum to grow causes other structures to direct their growth inward, accounting for ipsilateral hyperpneumatization of the sinuses, increased width of the diploic space and elevations of the greater wing of the sphenoid and petrous ridge (Sharma *et al.,* 2006).

The other change in infantile type is a shift of midline structures towards the side of the disease and the sulcal prominence replacing the gliotic tissue is absent. This feature differentiates it from cerebral hemiatrophy that occurs in later life. Acquired variety may be due to infections, trauma, ischemia and hemorrhage. Age of presentation depends on the time of occurrence of the brain insult and often clinical features may not be evident till adolescence. The exact mechanism of cerebral atrophy is still unclear in either type.

It is hypothesized that ischemic episodes from a variety of different causes reduce the production of brain derived neurotrophic factors, which in turn lead to cerebral atrophy (Lee *et al.,* 2006). Atalar *et al.,* 2007 in their clinico-radiologic analysis of 19 patients concluded that, Computed tomography and, in particular, magnetic resonance imaging is the procedures of choice with respect to assessment of the etiology and extent of cerebral parenchymal involvement in cerebral hemiatrophy[(Atalar *et al.,* 2007). The condition needs to be differentiated from Basal ganglia germinoma, Sturge Weber syndrome, Linear nevus syndrome, Fishman syndrome, Silver-Russell syndrome and Rasmussen Encephalitis (Sener *et al.,* 1992; Narain *et al.,* 2008).

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