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CASE REPORT

Dyke-Davidoff-Masson Syndrome -A Case report

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Abstract:

Dyke-Davidoff-Masson Syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemi atrophy) which is secondary to brain insult in fetal or early childhood period. Hemi atrophy is not frequently encountered in clinical practice. This clinical condition manifests with seizures, facial asymmetry, contra lateral spastic hemiplegia or hemiparesis and rarely mental retardation. The characteristic radiologic features are cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. We report here a case of a 6 year old boy with features suggestive of DDMS.

Keywords: Dyke-Davidoff-Masson Syndrome, cerebral hemiatrophy, seizures

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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. DDMS was first described by Dyke, Davidoff and Masson in a series of nine patients with plain skull radiographic and pneumatoencephalographic changes in 1933 [1]. The condition is usually characterized by seizures, contralateral hemiplegia or hemiparesis, sometimes mental retardation and characteristic radiological features are cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses(2-4). We report here a case of a 6 year old boy with seizures, left sided hemiparesis and the right cerebral hemiatrophy which is suggestive of DDMS.

Case Report

A 6 yr old boy presented with H/O repeated episodes of focal seizures affecting left side of the body and weakness of left upper and lower limbs since 3 years of age. He was on carbamazepine medication for the same. Patient's birth history was uneventful. Child attained mile stones at appropriate age. There was no family history of seizures. On examination there were no

neurocutaneous markers and there was atrophy of left upper & lower limbs. Neurological examination revealed left sided spastic hemiparesis with brisk tendon reflexes and extensor plantars.[Fig.1] His blood counts and CSF analysis were normal.

A plain and contrast CT head was done which revealed atrophy of the right cerebral hemisphere with dilatation of the ipsilateral lateral ventricle, widening of right sided Sylvian fissure and ipsilateral sulci [Fig.2,3]. With these features, a diagnosis of DDMS was made.

Discussion

Dyke, Davidoff, and Masson reported series of 9 patients whose clinical characteristics included hemiparesis, seizures, facial-asymmetry, and mental retardation and described the plain skull radiographic and pneumato encephalo graphic changes in 1993[1]. 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[2]The disease is generally classified into primary (congenital) and secondary(Acquired) types. In the primary (congenital)

type, the entire cerebral hemisphere is characteristically hypoplastic. The secondary type results from a cerebrovascular lesion, inflammatory process, or cranial trauma[2,3]. 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. This feature differentiates it from cerebral hemiatrophy which occurs in early life.



Fig.1 showing left upper limb and lower limb atrophy

The etiological factors for DDMS include 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[2-4]. A possible etiological relation of cerebral hemiatrophy and seizures has been reported by different studies in India [5, 6].

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 usual clinical presentation includes seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation.[7] However mental retardation may not present always and seizures may appear months or years after the onset of hemiparesis[8] .

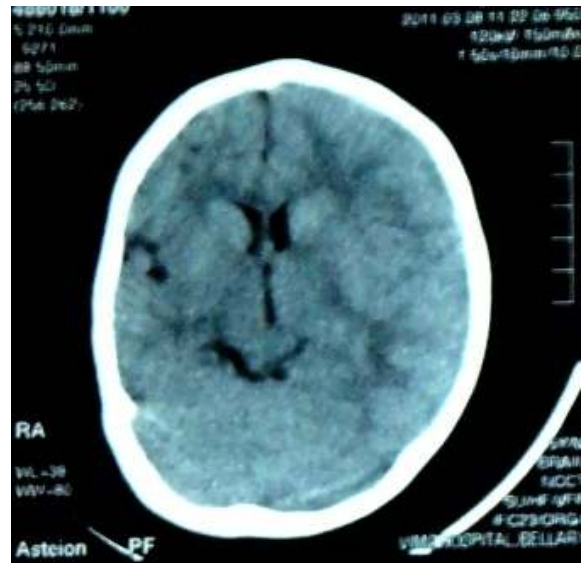


Fig.2- Axial section of CT shows prominent right sylvian cistern and dilated frontal horn of lateral ventricle.



Fig.3- Axial section of CT shows right cerebral hemiatrophy with illdefined hypodensity in right posterior parietal lobe-s/o chronic infarct with gliosis.

The clinical findings may vary 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 [2]. In the present case, the findings of dilated cortical sulci and widening of ipsilateral diploei with illdefined hypodensity in right posterior parietal lobe suggestive of chronic infarct with gliosis reflect a late onset of brain insult probably of vascular origin.

DDMS should be differentiated from Basal cell germinoma, Sturge Weber syndrome, Linear Nevus syndrome, Fishman syndrome, Silver- Russell syndrome and Rasmussen encephalitis. A proper clinical history and CT findings provide the correct diagnosis [9,10].

The treatment is symptomatic, and includes management of convulsions, hemiplegia, hemiparesis. If hemiparesis occurs after the age of 2 yrs and in absence of prolonged or recurrent seizures, the prognosis is better. Children with intractable disabling seizures and hemiplegia are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases[9].

References

- 1- Dyke CG, Davidoff LM, Masson LB. Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinus. *Surg Gynecol Obstet* 1933; 57:588-600.
- 2- Sharma S, Goyal D, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff Masson syndrome. *Indian J Radiol Imaging* 2006;16:165-166.
- 3- Pendse NA, Bapna P, Menghani V, Diwan A. Dyke – Davidoff – Masson Syndrome (DDMS). *Ind J Paed.* 2004; 71: 943.
- 4- Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson syndrome: Classical imaging findings. *J Pediatr Neurosci* 2010;5:124.
- 5- Nair KP, Jayakumar PN, Taly AB, Arunodaya GR, Swamy HS, Shanmugam V. CT in simple partial seizures in children: a clinical and computed tomography study. *Acta Neurol Scand* 1997; 95:197-200.
- 6- Garg RK, Karak B. Cerebral hemiatrophy: a possible etiological relation with febrile seizures. *Indian Pediatr* 1998; 35: 79-81.
- 7- Shetty DS, Lakhkar BN, John JR. Dyke-Davidoff-Masson syndrome . *Neurol India* 2003;51:136.
- 8- Zilkha.A. CT of cerebral hemiatrophy. *AJR* 1980; 135:259-262.
- 9- Narain NP, Kumar R, Narain B. Dyke – Davidoff – Masson Syndrome. *Indian Pediatrics.* 2008; 45: 927-928.5.
- 10- Rao KCVG. Degenerative diseases and hydrocephalus. *In: Lee SH, Rao KCVG, Zimmerman RA, editors. Cranial MRI and CT. Fourth edition. New York: McGraw-Hill; 1999. p.212-214.*