

Dyke - Davidoff - Masson Syndrome : A Case Report.

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Abstract

Dyke-Davidoff-Masson Syndrome (DDMS) is the rare clinical syndrome characterised by hemiparesis, seizures with classical radiological findings of cerebral hemiatrophy, calvarial thickening and hyperpneumatization of frontal sinuses. DDMS may be congenital or acquired / secondary, occurring at prenatal period or later. Etiopathogenesis of DDMS are trauma, infection, ischemic /hemorrhagic state, in premature infants with subependymal germinal matrix or interventricular hemorrhage and congenital vascular abnormalities of the cerebral circulation. This is a report of 6 year old child with hemiparesis, focal convulsions, mental retardation and characteristic radiological findings.

Introduction

Dyke -Davidoff - Masson Syndrome (DDMS) is the rare clinical syndrome characterised by hemiparesis, seizures with classical radiological findings of cerebral hemiatrophy, calvarial thickening and hyperpneumatization of frontal sinuses. DDMS may be congenital or acquired / secondary, occurring at prenatal period or later. Etiopathogenesis of DDMS are trauma, infection, ischemic/hemorrhagic state, in premature infants with subependymal germinal matrix or interventricular hemorrhage and congenital vascular abnormalities of the cerebral circulation. This is a report of 6 year old child with hemiparesis, focal convulsions, mental retardation and characteristic radiological findings.

Case Report

A six year old boy was referred with left side hemiplegia and seizures for the last five years. There were no symptoms referable to vision or hearing. There was no past history suggestive of antenatal and perinatal complications. Non contrast cranial CT scan with 3- 6 mm axial slices with MPR reconstruction and MRI brain were done with standard sequences. CT and MRI brain showed right cerebral hemispheric atrophy with ex - vacuo dilatation of lateral ventricle, thickening of right frontal calvarium with hypertrophy of the frontal sinuses with compensatory hypertrophy of cerebellar hemisphere.

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Diffusion weighted images and MR spectroscopy confirmed vascular etiopathogenesis of DDMS.

Discussion

DDMS was described by Dyke Davidoff and Masson in 1933 [2]. Prenatal causes of DDMS are cerebral infarction, vascular malformation and infection. Congenital form of DDMS has also been reported [1] in which there is ipsilateral shift of midline structures, sulcal prominence, and replace in glial tissue will be absent which is a differentiating point for perinatal cause.

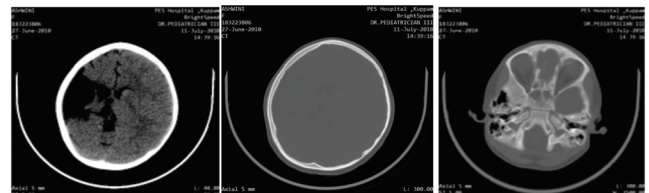


Fig. 1: Cranial CT Showing right cerebral atrophy, calvarial thickening and hyperpneumatization of bilateral mastoid air cells.

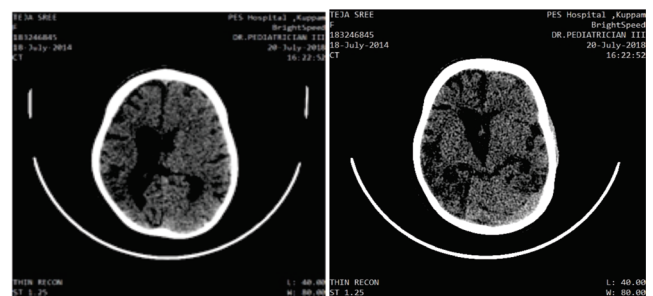


Fig. 2: Cranial CT showing right cerebral atrophy with ipsilateral ventriculomegaly, contralateral cerebral hypertrophy

Clinical presentation of DDMS depends on the degree of brain injury, seizures, fascial asymmetry, contralateral hemiplegia/ hemiparesis and mental retardation [5]. The clinical history, CT and MRI are the mainstay in arriving

at correct diagnosis of DDMS. The differential diagnoses of DDMS are Sturge Weber Syndrome, Rasmussen encephalitis, Silver Russel Syndrome, Basal ganglia germinoma, Fishman Syndrome and Linear nevus syndrome [4].

Sturge Weber syndrome (Encephalio trigeminal angiomatosis) is characterised by port wine nevus, seizures, galucoma, mental retardation and recurrent seizures due to stasis and ischemia due to cranial vascular anomaly and leptomeningial angiomatosis leading to tram track like calcification.

Rasmussen encephalitis is chronic progressive immune mediated encephalitis characterised by intractable focal epilepsy and cognitive defects and typical imaging features of cerebellar hemispheric atrophy without calvarial changes.

Silver-Russell Syndrome is characterized by the traingular face, small pointed chin, broad forehead, thin wide mouth, delayed bone age, clinodactyly, hemihypertrophy with normal intelligence.

Basal ganglia germinoma is a rare brain tumour characterised by progressive hemiparesis with cerebellar hemiatrophy and on imaging reveals cystic areas, focal haemorrhages with edema and calvarial changes.

Fishman Syndrome is a neurocutaneous syndrome with unilateral cranial lipoma and lipodermoid of the eye.

Linear Nevi Syndrome presents with facial nevus, recurrent seizures, mental retardation and unilateral ventricular dilatation.

The treatment of DDS is symptomatic. Hemispherectomy is a treatment of choice for intractable disabling seizures and hemiplegia.

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