ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
Index Copernicus Value: 68 . 16
PubMed - National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 5, Issue –4, July - 2020, Page No. : 82 - 84

Dyke-Davidoff Masson Syndrome: A Rare Case Report

[1]Dr. Geetanjali Sethy, Associate Professor, Dept of Pediatrics, M.K.C.G Medical college, Berhampur, Ganjam,Odisha,760004, India

[2]Dr. Anjali Saji, Junior Resident, Dept of Pediatrics, M.K.C.G Medical college, Berhampur, Ganjam, Odisha,760004, India

[2]Dr.Aparna B Raj, Junior Resident, Dept of Pediatrics, M.K.C.G Medical college, Berhampur, Ganjam, Odisha,760004, India

[2]Dr. Nithya B G, Junior Resident, Dept of Pediatrics, M.K.C.G Medical college, Berhampur, Ganjam, Odisha,760004, India

Corresponding Author: Dr. Anjali Saji, Junior Resident, Dept of Pediatrics, M.K.C.G Medical college, Berhampur, Ganjam, Odisha,760004, India

Citation this Article: Dr. Geetanjali Sethy, Dr. Anjali Saji, Dr.Aparna B Raj, Dr. Nithya B G,“ Dyke-Davidoff Masson Syndrome: A Rare Case Report”, IJMSIR- July - 2020, Vol – 5, Issue - 4, P. No. 82 – 84.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Dyke-Davidoff Masson Syndrome is a rare condition of unknown frequency resulting from brain injury due to a multitude of causes especially in early life.It is characterized by cerebral hemiatrophy/hypoplasia, contralateral hemiparesis and seizures.We report a case of a 7year old female child who presented with intractable seizure, left sided hemiparesis and mental retardation.Seizures may be refractory to medical management and surgery may be required and hence timely diagnosis is required.

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

Introduction

Dyke-Davidoff Masson Syndrome (DDMS) is a rare cause of cerebral hemiatrophy in children and the diagnosis is mainly established by clinical features and positive radiological findings.It is characterized by cerebral hemiatrophy, contralateral hemiplegia or hemiparesis, thickening of cranial vault, seizures, mental retardation, facial asymmetry and behavioural changes.

Although rare, this syndrome should be considered in a case of persistent seizures especially with hemiparesis.CT and MRI are powerful imaging modalities to diagnose the characteristic imaging features associated with the disease. Knowledge of the clinical presentation, risk factors and imaging features is indispensable for appropriate management.

Case Report

A 7 year old female child presented with intractable seizures of generalized tonic type and left sided hemiparesis to the casualty. The seizure was controlled after administration of two anticonvulsants. She had a history of perinatal asphyxia and has delayed attainment of milestones. She was on sodium valproate

at 40mg/kg/day since the age of 3 years. There was history of breakthrough seizures on and off while on valproate therapy. There was no history of fever/cough/vomiting or diarrhoea.The family history was not significant and she was fully immunized.

On examination, she had microcephaly with facial asymmetry and left sided hemiparesis and exaggerated deep tendon reflexes. There was no signs of meningeal irritation and neurocutaneous markers.Other systems were found to be in normal limits.

Her routine hematological and biochemical tests including serum electrolytes, RFT, Blood sugar,serum calcium were normal. Fundoscopy was also normal. NCCT brain revealed cerebral hemiatrophy on right side (Fig 1). MRI brain showed right cerebral hemiatrophy and gliotic changes with occipital porencephalic cyst formation with exvacuo dilatation of right lateral ventricles and hyperpneumatization of right side frontal sinus.(Fig 2) These CT and MRI findings along with clinical pictures were satisfying the criteria of DDMS and hence the diagnosis was made. The patient was started on valproate and phenytoin and once when seizure was controlled, the child was discharged with oral valproate, phenytoin and nasal midazolam spray and was advised for physiotherapy. The patient is now on constant follow up.

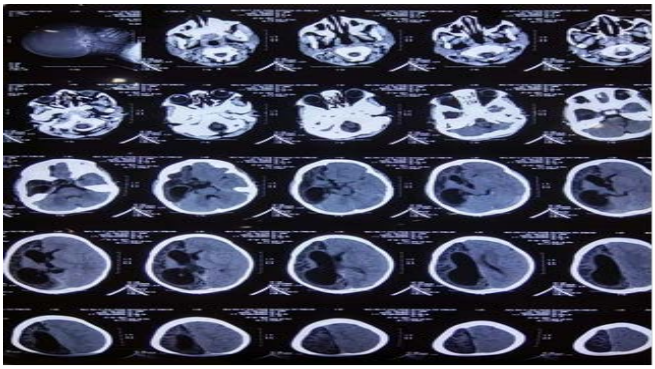

Fig 1: NCCT Brain : cerebral Hemiatrophy of right side

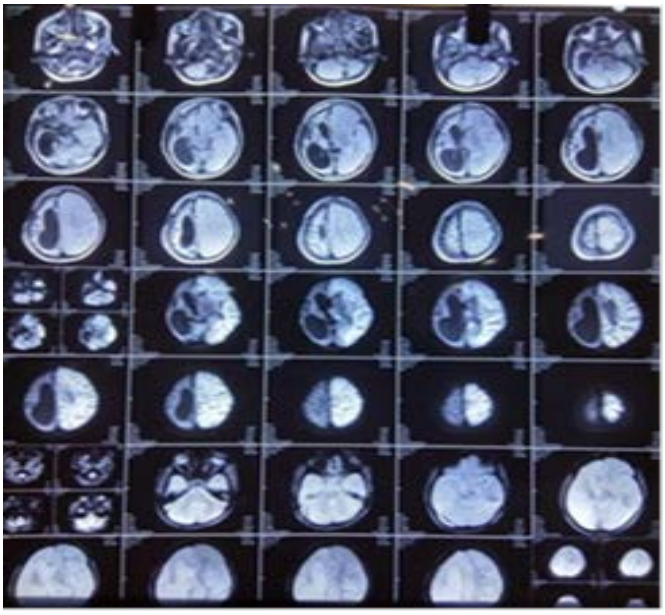

Fig 2: MRI Brain

Discussion

Dyke-Davidoff Masson Syndrome is a rare neurological condition of unknown frequency, usually affecting paediatric population[(1)]. It was first reported in 1933,by Dyke, Davidoff and Masson in a case series of 9 patients with clinical features of hemiparesis, facial asymmetry, seizures and mental retardation.They were noted to have pneumatoencephographic changes on skull radiograph[(2)]. CT and MRI findings of this entity include cerebral hemiatrophy, ipsilateral ventriculomegaly, hyperpneumatization of sinuses on affected side and compensatory calvarial thickening[(3)].

The condition is hypothesized to be caused by unknown brain injury, due to congenital abnormalies, perinatal hypoxia, intra cranial hemorrhage and infections[(4)].

Hageman et al proposed the term cerebral hypoplasia or unilateral hemiatrophy because there is a lack of cerebral development[(5)].When this occurs within 2 years of life, it is associated with compensatory changes in skull to take up the relative vaccum created by hypoplastic cerebrum.

Clinically, it will present with seizures, mental retardation, contralateral hemiparesis and facial

asymmetry. Imaging through CT and MRI proves to be of value, with features being cerebral hemiatrophy/hypoplasia, hyperpneumatization of para nasal sinus and compensatory osseous hypertrophy.

The condition needs to be differentiated from basal ganglia germinoma, Sturge Weber syndrome, Rasmussen encephalitis[6], Silver Russell syndrome, Hemiconvulsion hemiplegia epilepsy(HHE).

Treatment includes symptomatic management with management of convulsions, hemiplegia,hemiparesis, and learning difficulties. Hemispherectomy has been tried in refractory cases and it significantly reduces seizures in 85% of them[6].

Thus appropriate diagnosis of the condition by combined clinical and radiological investigation and control of seizures and physiotherapy helps in improving the child.

Conclusion

The disease, was supposed to be rare once,is increasingly reported now, may be because of the better neonatal resuscitation facilities and better diagnostic modalities. Awareness about the disease and management of seizures is important in the management. Timely referal to a neurosurgeon in refractory seizures for hemispherectomy has better success rates.

Abbreviations

DDMS – Dyke Davidoff Masson Syndrome

CT- Computed Tomography

MRI- Magnetic Resonance Imaging

NCCT – Non Contrast Computed Tomography

RFT- Renal Function Tests

HHE- Hemiconvulsion Hemiplegia Epilepsy

References

1. Anna Misyail,Abdul Rashid et al. Dyke Davidoff Masson syndrome; BMC Neurology case reports 2018;18:76.
2. Dyke CG, Davidoff LM ,Masson CB. Cerebral hemiatrophy and homolateral hypertrophy of skull and sinuses. Surg Gynecol Obstet 1933;57:588-600.
3. Roy U, Panwar A et al. Dyke Davidoff Masson Adult presentation: A case report.Case Rep Neurol.2016;8:20-26.
4. Verma R, Sahu R. Dyke Davidoff Masson syndrome. BMJ Case Reports.2012. http://doi.org/10.1136/bcr-2012-006729.
5. Hageman G, Gooskens RH, Willemse J.A cerebral cause of Arthrogryposis:unilateral cerebral hypoplasia.Clin Neurol Neurosurg 1985;87:119-22.
6. Narain NP, Kumar R, Narain B. Dyke Davidoff Masson syndrome. Indian Pediatr 2008;45:927-8.