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 – 1, January - 2020, Page No. : 98 - 101

Dyke-Davidoff-Masson Syndrome: A Rare Case of Hemiparesis with Classical Radiological Findings

[1]Dr Ankita Singh, Resident Doctor, Department of General Medicine, SMS Medical College, Jaipur, Rajasthan, India.

[2]Dr Pradeep Mital, Senior Professor and unit head, Department of General Medicine, SMS Medical College, Jaipur, Rajasthan, India.

[3]Dr Sujata Agarwal, Associate Professor, Department of General Medicine, SMS Medical College, Jaipur, Rajasthan, India.

[4]Dr Vinay Jorwal, Assistant Professor, Department of General Medicine, SMS Medical College, Jaipur, Rajasthan, India.

[5]Dr Shashi Bhushan, Assistant Professor, Department of General Medicine, SMS Medical College, Jaipur, Rajasthan, India.

[6]Dr Jitendra Kumawat, Junior Specialist, Department of General Medicine, SMS Medical College, Jaipur, Rajasthan, India.

Corresponding Author: Dr Pradeep Mital, Senior Professor and Unit Head, Department of General Medicine, SMS Medical College, Jaipur, Rajasthan, India.

Citation this Article: Dr Ankita Singh, Dr Pradeep Mital, Dr Sujata Agarwal, Dr Vinay Jorwal, Dr Shashi Bhushan, Dr Jitendra Kumawat," Dyke-Davidoff-Masson Syndrome: A Rare Case of Hemiparesis with Classical Radiological Findings ", ijmsir- January - 2020, Vol – 5, Issue -1, P. No. 98-101.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Dyke-Davidoff-Masson Syndrome (DDMS) is an uncommon disease entity that usually manifests in childhood in a constellation of features like seizures, hemiparesis, facial asymmetry and intellectual disability. These seizures are typically drug-resistant. It results due to brain insult endured during perinatal period or early life especially birth asphyxia. Syndrome is characterised by classical neuroimaging findings that includes cerebral hemiatrophy, hyperpneumatisation of frontal sinus and thickening of calvaria.

Keywords: Seizures, hemiparesis, cerebral hemiatrophy.

Case Report

A 15-years old female presented in the emergency department of Medicine, with history of facial deviation and left-sided focal seizures with secondary generalisation followed by altered sensorium. On examination, bilateral plantars were extensor with pupil being normal sized and reactive to light bilaterally. There were no signs of meningeal irritation and thereby patient was managed with antiepileptic and intravenous fluids. Her parents gave similar history of seizure episodes since her childhood, she had first fit in her life at the age of two which was preceded by a febrile illness and then followed by left sided focal abnormal body movement for which she was

taken to local hospital and was treated with antipyretics and then taken back home after recovery. Again she had another episode of seizures after 2 weeks and she was rushed to the same hospital and was managed accordingly with advice of reference to higher centre. But family instead took the patient to local healers for magical cure out of their myths and superstitions, but she continued to have fits. So finally they took her to the hospital and was kept on antiepileptic medications after which she had improvement but few seizure episodes were there for relatively shorter duration and decreased frequency than before and with spontaneous recovery to which the parents did not paid attention. Parents also gave history of relative weakness and underdevelopment of left side. She was born at full term by normal vaginal delivery with no perinatal complications. She achieved normal milestones up to age of 2 years and thereafter she had delayed milestone. She was below average in studies and had to withdraw from school because of intellectual disability. Now since past 8 days patient was not taking medications and presented with convulsions.

On General physical examination relative thinning of left upper and lower limb compared to right was appreciated (Figure 1&2). Power was decreased on the left side. Patient's mini mental status examination (MMSE) was decreased 15/30 indicating intellectual impairment. Also on detailed examination facial asymmetry was found. (Figure 3)

Blood reports include normal renal and liver function tests, normal thyroid profile, and normal electrolytes and coagulation profile. Her chest X-Ray & ECG were also normal.

Magnetic resonance imaging of brain was done which shows: Cranial MRI of the patient revealed diffuse right cerebral parenchymal atrophy leading to enlargement of the ipsilateral lateral ventricle with prominent sulci (Figure 4), frontal sinus of the same side is markedly enlarged (Figure 5). Other interesting finding was thickened skull vault on the side of cerebral hemiatrophy probably indicating the compensatory changes (Figure 6).

Based on history, clinical presentation and typical neuroimaging findings, our patient was diagnosed as a case of Dyke-Davidoff-Masson syndrome.

Therefore, Patient was put on Antiepileptic carbamazepine in an appropriate dose and physiotherapy was taught on her discharge and she is currently in our regular follow up and is clinically well now.

Discussion

Dyke-Davidoff-Masson syndrome is a rare neurological disorder that mostly manifests in childhood and very rarely in adults. It was originally described by three US neuroradiologists Cornelius Gysbert Dyke, Leo Max Davidoff and CB Masson in 1933 by studying the plain skull radiographic changes in nine patients who presented clinically with seizures, hemiparesis, facial asymmetry and intellectual impairment. [1]

It is typically characterised by hypoplasia /atrophy of any one cerebral hemisphere which is mainly due to brain insult that has occurred in utero or during early childhood resulting in hemiparesis, convulsions, facial asymmetry, mental retardation and compensatory thickening of calvaria and hyperpneumatisation of frontal sinus [2]. Both sexes can be involved with occurrence found more in males. Also any of the cerebral hemisphere could be involved but left hemisphere involvement is seen commonly [3].

The DDMS has been categorised into two forms: congenital (infantile) and acquired. Congenital type presents in the early infancy or childhood while acquired one presents later in life in adolescence or adulthood depending on the time of cerebral insult [3]. There are

three recognised patterns of cerebral hemiatrophy on MRI given by Shen et al [4]. Pattern I denotes to diffuse cortical and subcortical atrophy. Pattern II is diffuse cortical atrophy coupled with a porencephalic cyst, while pattern III corresponds to previous infarction with gliosis in the middle cerebral artery (MCA) territory [5].

Other differential diagnosis to be considered in such patient include Sturge- Weber syndrome, Rasmussen encephalitis, Basal ganglia germinoma, Fishman syndrome [7], Silver Russell syndrome and Linear nevus sebaceous syndrome [6].

Management of DDMS is mainly symptomatic that includes antiepileptic drugs, surgical intervention like hemispherectomy is required in case of refractory seizures that guarantees 85% chance of success [7]. Other supportive treatment measures include physical rehabilitation and occupational therapy.

References

1. CG Dyke, LM Davidoff, CB Masson Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses In: Surgery, Gynecology and Obstetrics, Chicago , 1933, Vol. 57, pp. 588-600.
2. Arora R, Rani JY. Dyke–Davidoff–Masson syndrome: imaging features with illustration of two cases. Quant Imaging Med Surg 2015;5(3):469–71.
3. Sharma B, et al. BMJ Case Rep 2014. doi:10.1136/bcr-2014-204679
4. Shen WC, Chen CC, Lee SK, et al. Magnetic resonance imaging of cerebral hemiatrophy. J Formos Med Assoc 1993;92:995–1000.
5. Singh P, Saggar K, Ahluwalia A. Dyke–Davidoff–Masson syndrome: classical imaging findings. J Pediatr Neurosci 2010;5(2):124–5.
6. Herman T.E., Siegel M.J. Hemimegalencephaly and linear nevus sebaceous syndrome. J Perinatol. 2001;21(5):336–338.
7. Roy U, Panwar A,Mukherjee A, Biswas D. Adult presentation of Dyke–Davidoff–Masson syndrome: a case report. Case Rep Neurol 2016;8(1):20–6.

Legends Figure

Figure 1: relative thin left forearm and hand compared to right showing prominent knucles of left hand.

Figure 2: Atrophied left leg

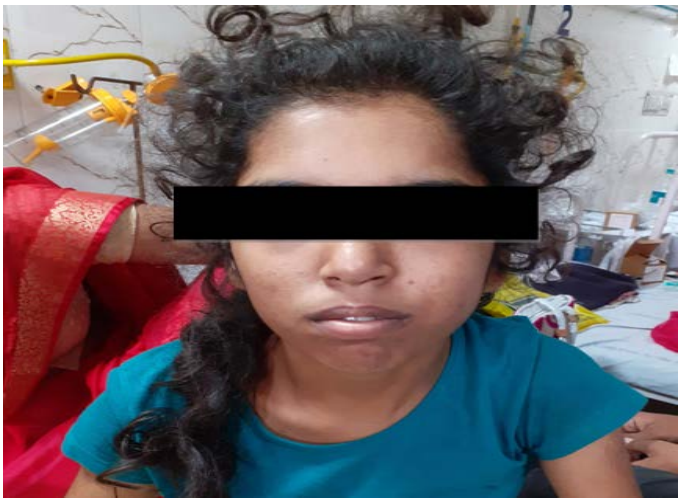

Figure 3: Facial asymmetry

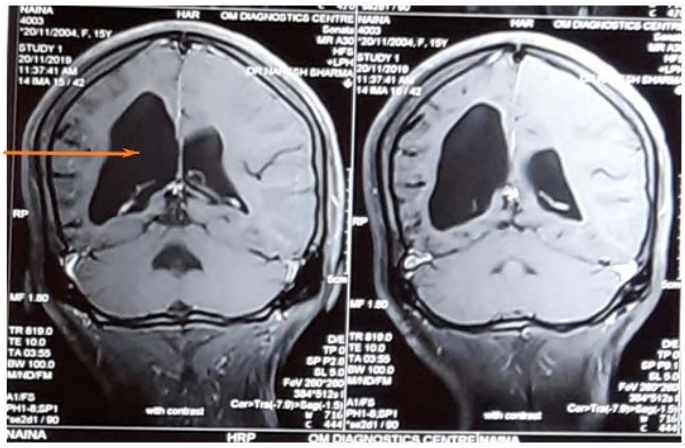

Figure 4: Coronal section T2 flair showing right ventriculomegaly – lateral ventricle (red arrow)

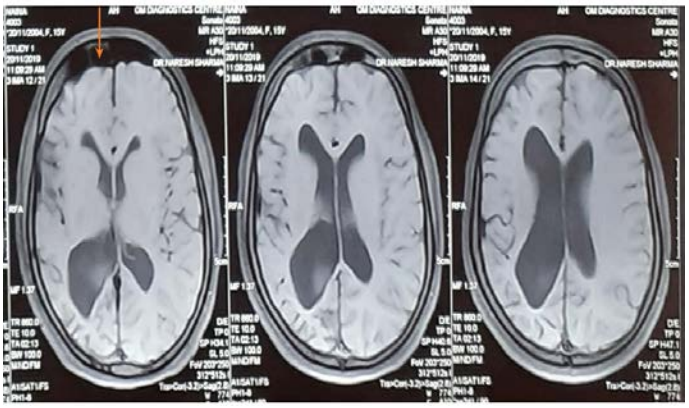

Figure 5: Axial section T1 Weighted image showing hyperpneumatisation of frontal sinus (red arrow).

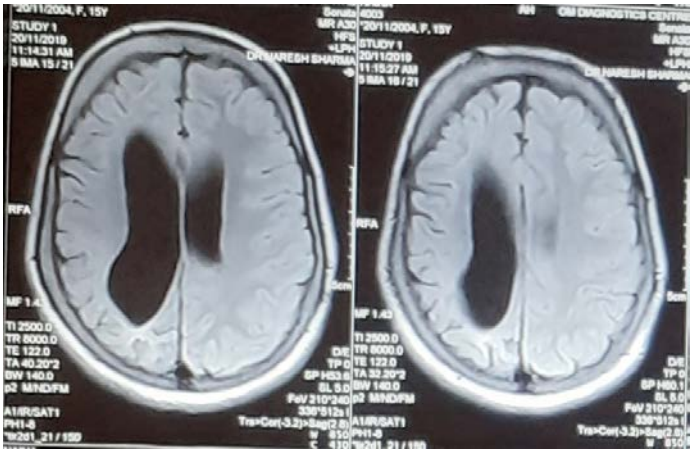

Figure 6: T1W axial section showing right cerebral atrophy and thickening of calvaria.