Case Report

Dyke-Davidoff-Masson syndrome

N Vinay Kumar, TS Gugapriya, Arun T Guru[1], SD Nalina Kumari

Departments of Anatomy and [1]Radiodiagnosis, Chennai Medical College Hospital and Research Centre, Trichy, Tamil Nadu, India

Abstract

Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere, due to an insult to the developing brain in fetal or early childhood period. Age of presentation depends on the time of neurologic insult, and characteristic changes may be seen only in adolescence. Male gender and left hemisphere are more frequently involved.
A 17-year-old female adolescent with a history of recurrent refractory seizures, hemiplegia and mental retardation reported to Department of Radiology for computed tomography (CT) assessment of brain. On examination, she had facial asymmetry, delayed milestones, and spastic hemiplegia. The CT brain showed right cortical atrophy with ventricular dilatation, prominent sulci, and shifting of falx to the right side. Bone window image showed asymmetry in skull vault thickness, the width of diploic space, the size of paranasal air sinuses and inclination of the petrous ridge between the affected and normal sides. As the above case deviates from the usual presentation of male left sided DDMS, hence the report.

Key words: Cerebral atrophy, paranasal air sinuses, petrous ridge, prominent sulci

Submission: 19-12-2014 **Accepted:** 03-09-2015

Introduction

Dyke-Davidoff-Masson syndrome (DDMS), reported first in 1933[1] refers to atrophy or hypoplasia of one cerebral hemisphere (hemi atrophy), which is usually due to an insult to the developing brain in fetal or early childhood period. Congenital and acquired presentations of DDMS are recognized.[1,2] Frequently male gender and left hemisphere involvement is seen. Age of presentation depends on the time of neurologic insult, and characteristic changes may be seen only in adolescence.[3]

The clinical features of this syndrome are variable and depend on the extent of brain injury.[2] It is characterized by cerebral hemiatrophy (CH) with prominent sulci, contralateral hemiplegia/hemiparesis with homolateral hypertrophy of the skull and paranasal air sinuses, elevation of the sphenoid wing and petrous ridge, facial asymmetry, seizures, mental retardation, and delayed developmental milestones.[1,4]

Case Report

A 17-year-old female adolescent with a history of recurrent refractory seizures, hemiplegia and mental retardation reported to Department of Radiology for computed tomography (CT) assessment of brain. On examination, the patient had facial asymmetry, delayed developmental milestones and spastic hemiplegia. The CT brain showed right CH, lateral ventricular dilatation (LVD), prominent sulci, and displacement of falx cerebri attachment with midline shift toward the atrophic side [Figure 1]. Bone window image showed thick skull vault with widened diplopic space on the right when compared to left

Address for correspondence: Dr. N Vinay Kumar, Department of Anatomy, Chennai Medical College Hospital and Research Centre, Irungalur, Trichy - 621 105, Tamil Nadu, India.
E-mail: vinaydr1981@gmail.com

Access this article online

Quick Response Code:

Website: www.ijabmr.org

DOI: 10.4103/2229-516X.174016

How to cite this article: Kumar NV, Gugapriya TS, Guru AT, Nalina Kumari SD. Dyke-Davidoff-Masson syndrome. Int J App Basic Med Res 2016;6:57-9.

side [Figure 2]. There was the elevation of the petrous ridge and upward tilting of planum – sphenoidale [Figure 3] along with narrowed right anterior and middle cranial fossae. The mastoid air cells on the affected side appeared larger than the other side [Figure 3] along with larger frontal, ethmoidal, and sphenoidal air sinuses.

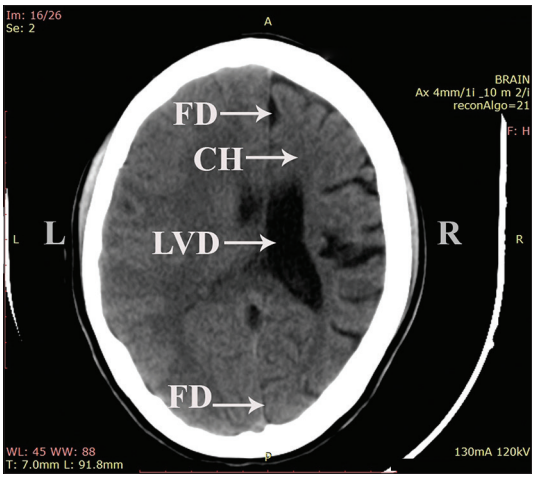

Figure 1: Right cerebral hemiatrophy (CH) with prominent sulci, lateral ventricular dilatation (LVD), falx displacement (FD)

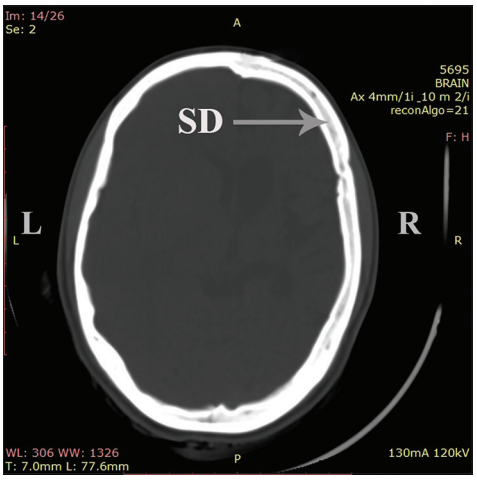

Figure 2: Thick skull vault with widened diploic space (SD)

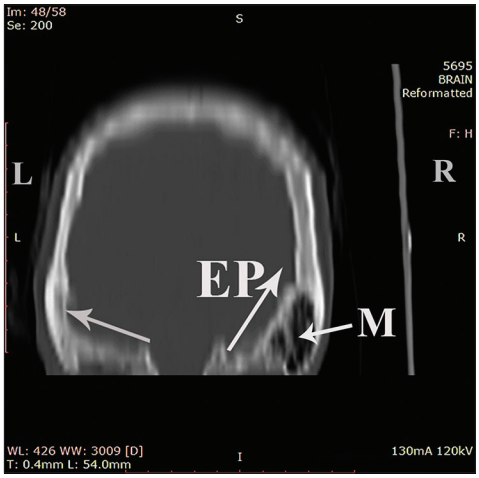

Figure 3: Elevated petrous ridge (EP) and larger mastoid air cells (M)

Discussion

DDMS may be classified as congenital or primary and acquired or secondary.[1,2] In congenital type, there is usually no apparent etiologic factor but most likely it may be due to intrauterine vascular occlusion involving middle cerebral artery territory[5] and the symptoms are present at birth or shortly thereafter. In acquired type, the symptoms are related to central nervous system damage that occurs in the perinatal period or later. The etiologic factors may be trauma, ischemic and hemorrhagic conditions, infection – cerebral malaria[6] and encephalitis,[7] vascular abnormality such as coarctation of the midaortic arch, internal carotid hypoplasia or agenesia, reduced or absent middle cerebral artery.[1,8]

CT or magnetic resonance imaging is the gold standard for the diagnosis of DDMS. The nature and extent of the underlying pathologic processes vary widely. The largest series of DDMS in the literature concluded male sex dominance (73.5%) with left hemisphere involvement (69.2%) among 26 patients with a mean age 11.[3] Studies have reported delayed appearance of mental retardation and early appearance of seizures associated with cerebral atrophy after the onset of hemiparesis.[8,9]

The finding of right CH with LVD and prominent sulci and displacement of falx attachment with midline shift toward the atrophic side reflect a late onset of brain insult due to any of the acquired causes resulting in abnormal neuronal and glial proliferation or apoptosis during cortical development. Prominent sulci will be absent when the brain insult occurs during embryogenesis, where the formation of sulci and gyri are incomplete, but a shift of midline structures towards the disease side will be seen.[2,8]

The findings of thick skull vault with widened diplopic space, hyperpneumatisation of paranasal air sinuses and mastoid air cells on the affected side, elevation of petrous ridge and upward tilting of planum – sphenoidale with a small anterior and middle cranial fossae may be due to insult to brain occurring during first 18 months to 2–3 years of life and compensatory adaptation to unilateral decrease of brain substance.[1,4,8]

Management is mainly symptomatic. Patient with intractable hemiplegia/hemiparesis and seizures are the candidates for functional hemispherectomy with a success rate of 85% in carefully selected cases. Prognosis is good when hemiparesis occurs after the age of 2 years, and there is the absence of prolonged or recurrent seizures.[10] Conditions that are associated with CH such as Rasmussen encephalitis, Silver's

syndrome, linear nevus sebaceous syndrome, progressive multifocal leukoencephalopathy, Sturge–Weber syndrome, and Fishman syndrome are to be differentiated from DDMS.

The present case exhibits classical features of the acquired type of CH involving right cerebral hemisphere. This case has deviated from the usual presentation of male predominance and left sided DDMS. In cases of intractable seizures, physicians should consider DDMS as one among the differential diagnosis.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Dyke CG, Davidoff LM, Masson CB. Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. Surg Gynecol Obstet 1933;57:588-600.
2. Aguiar PH, Liu CW, Leitão H, Issa F, Lepski G, Figueiredo EG, *et al*. MR and CT imaging in the Dyke-Davidoff-Masson syndrome. Report of three cases and contribution to pathogenesis and differential diagnosis. Arq Neuropsiquiatr 1998;56:803-7.
3. Unal O, Tombul T, Cirak B, Anlar O, Incesu L, Kayan M. Left hemisphere and male sex dominance of cerebral hemiatrophy (Dyke-Davidoff-Masson Syndrome). Clin Imaging 2004;28:163-5.
4. Shrestha B. Acquired cerebral hemiatrophy: Dyke-Davidoff-Masson Syndrome – A case report. Turk Neurosurg 2013;23:117-21.
5. Shen WC, Chen CC, Lee SK, Ho YJ, Lee KR. Magnetic resonance imaging of cerebral hemiatrophy. J Formos Med Assoc 1993;92:995-1000.
6. Karuppiah S, Rodgman C, Lombard J. Dyke-Davidoff-Masson syndrome in postcerebral malaria. J Child Neurol 2009;24:487-90.
7. Atalar MH, Icagasioglu D, Tas F. Cerebral hemiatrophy (Dyke-Davidoff-Masson syndrome) in childhood: Clinicoradiological analysis of 19 cases. Pediatr Int 2007;49:70-5.
8. Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy. Clin Imaging 1992;16:93-7.
9. Sharma S, Goyal D, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff Masson syndrome. Indian J Radiol Imaging 2006;16:165-6.
10. Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson syndrome. Indian Pediatr 2008;45:927-8.