Dyke-Davidoff-Masson Syndrome: A Case Report

Vidyashree Musti, Jayashree R.G., Rudresh S. Halawar, Tukaram Rathod

Department of Radiodiagnosis, S. N. Medical College & HSK Hospital, Bagalkot, Karnataka, India

Abstract

Dyke–Davidoff–Masson syndrome (DDMS) is a rare congenital, neonatal or early infantile condition characterized clinically by hemiparesis or hemiplegia, variable degrees of facial weakness, mental retardation and seizures. This condition results from atrophy or hypoplasia of one of the cerebral hemisphere. The hallmark radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses and crossed cerebellar atrophy. A 39 year old woman was brought to the casualty with history of status epilepticus. Seizure semiology was generalized tonic clonic variety. She had significant past history in the form of left hemiparesis since childhood, mild mental retardation and seizures which were poorly controlled. She was treated with antiepileptics and general anaesthetics. Patient underwent imaging of the brain. Computed Tomography (CT) scan of the brain showed atrophy of right cerebral hemisphere with dilation of ipsilateral lateral ventricle, dilatation of the sylvian fissure and sulci. There was thickening and compensatory hypertrophy of the calvarium on right side with hyperaeration ofright frontal sinus. There was also contralateral cerebellar atrophy seen. In essence, due to rarity of this syndrome, it may be easily misdiagnosed by the untrained eye. CT and MRI are powerful imaging modalities to diagnose the pertinent imaging features associated with this syndrome.

Key words: Dyke-Davidoff-Masson Syndrome, Computed tomography, brain.

Introduction

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare congenital, neonatal or early infantile condition characterized clinically by hemiparesis or hemiplegia, variable degrees of facial weakness, mental retardation and seizures. This condition results from atrophy or hypoplasia of one of the cerebral hemisphere. The hallmark radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses and crossed cerebellar atrophy. The major concern of the disease remain the intractable seizures for which drug therapy is not sufficient in most of the cases, and a surgical approach is necessary.

Case Report

A 39 year old woman was brought to the casualty with history of status epilepticus. Seizure semiology was generalized tonic clonic variety. She had significant past history in the form of left hemiparesis since childhood, mild mental retardation and seizures which were poorly controlled. She was treated with antiepileptics and general anaesthetics. Patient underwent imaging of the brain. Computed Tomography (CT) scan of the brain showed atrophy of right cerebral hemisphere with dilation of ipsilateral lateral ventricle, dilatation of the sylvian fissure and sulci (Figure 1). There was thickening and compensatory hypertrophy of the calvarium on right side with hyperaeration ofright frontal sinus (Figure 2 and 3). There was also contralateral cerebellar atrophy seen (Figure 3 and 4)

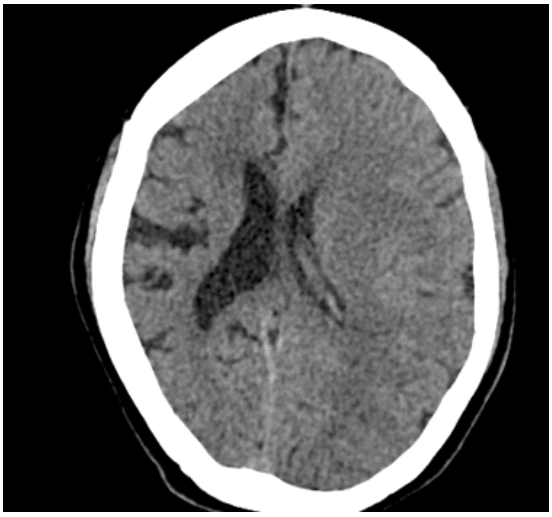

Figure 1: CT Scan of Brain showing right cerebral hemiatrophy with dilation of ipsilateral lateral ventricle

Address for Correspondence:

Dr. Vidyashree M.

Department of Radiodiagnosis, S.N. Medical college, Bagalkot, Karnataka, India

Email: sweetvidya27@gmail.com

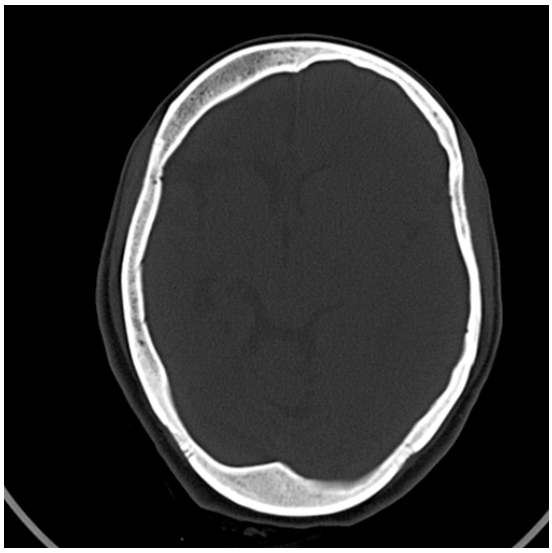

Figure 2: Thickening and compensatory hypertrophy of the calvarium on right side

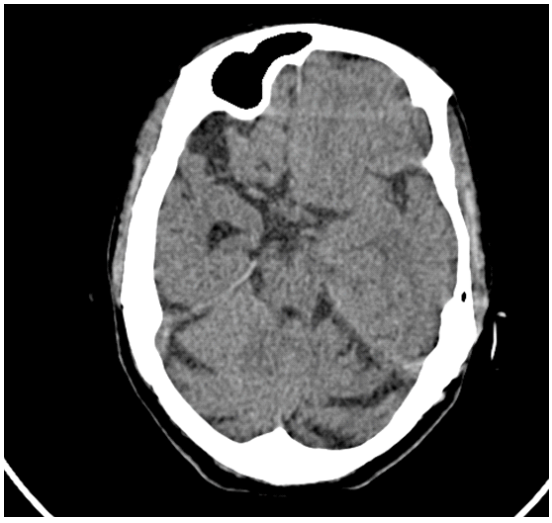

Figure 3: CT scan of Brain showing hyperaeration of right frontal sinus

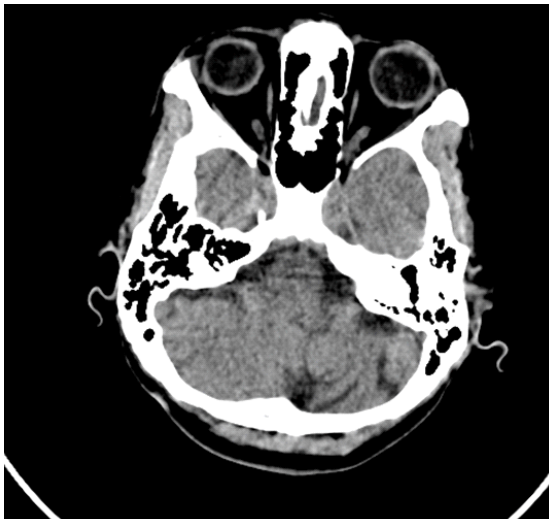

Figure 4: CT Scan of brain showing left cerebellar diaschisis

Discussion

Dyke-Davidoff-Masson syndrome is a syndrome characterized by cerebral hemiatrophy and a contralateral hemiplegia, epilepsy and mental retardation with characteristic skull changes. In 1933, Dyke, Davidoff, and Masson first described skull radiographic and pneumatoencephalographic changes in a series of nine patients characterized by hemiparesis, seizures, facial-asymmetry.1

Brain imaging may additionally reveal prominent cortical sulci, dilated lateral ventricles and cisternal space, calvarial thickening, ipsilateral osseous hypertrophy with hyperpneumatization of the sinuses (mainly frontal and mastoid air cells), and an elevated temporal bone.[2]

The clinical features include contralateral hemiparesis with an upper motor neuron type of facial palsy, focal or generalized seizures, and mental retardation along with learning disabilities.[3] There is no sex predilection, and any side of the brain can be involved, although involvement of the left side and male gender have been shown to be more common in one study.[2]

There are two varieties of cerebral hemiatrophy-infantile and acquired. The infantile or congenital variety results from neonatal or gestational vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies, coarctation of the mid-aortic arch or infections and patients become symptomatic in the perinatal period or infancy. The acquired type results from various causes like birth asphyxia, prolonged febrile seizures, trauma, tumor, infection, ischemia, and hemorrhage[4,5].

When the cerebral hemiatrophy develops in utero or during first two years of life, it is associated with certain cranial changes like ipsilateral hypertrophy of the skull and sinuses as a compensatory change to take up the relative vacuum created by the hypoplastic cerebrum.[6] As the brain enlarges, the brain presses outward on the bony tables which gradually results in the general shape of the adult head. However, failure of the cerebrum to grow causes other structures to direct their growth inward, accounting for ipsilateral hyperpneumatization of the sinuses.

The differential diagnoses of DDMS are Rasmussen's encephalitis, Sturge Weber syndrome and Hemimegalencephaly. Rasmussen encephalitis is characterized by its progressive course of hemiplegia with worsening documented clinically as well as on MRI on separate time occasions and is associated

with intractable epilepsy.[7] Sturge–Weber syndrome is characterized by the classical port wine stain and the cerebral venous malformations and tram track calcification.[8] Hemimegalencephaly is a congenital condition characterized by the defective cellular organization and neuronal migration resulting in hamartomatous overgrowth of the brain.

Our patient had presented with status epilepticus on the background of left hemiparesis and seizures since childhood. CT scan features were consistent with that of typical cases of DDMS. The manifestations of DDMS may be so subtle as to be overlooked on plain radiographs; however, CT is the diagnostic modality of choice.

Conclusion: In essence, due to rarity of this syndrome, it may be easily misdiagnosed by the untrained eye. CT and MRI are powerful imaging modalities to diagnose the pertinent imaging features associated with this syndrome

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