Case Report

Dyke-Davidoff-Masson Syndrome - A Delayed Diagnosis of an Acquired Variant

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Abstract

Dyke-Davidoff-Masson syndrome (DDMS) is an important cause of intractable and drug-resistant seizures. It has varied clinical presentation and history with distinct neuroimaging features. Here, we describe a female patient presented with recurrent intractable convulsion, mental retardation, hemiparesis, and characteristic neuroimaging features of cerebral hemiatrophy, calvarial thickening, and ipsilateral hyperpneumatization of the frontal sinuses which is suggestive of DDMS. Early institution of neuroimaging in patients with intractable epilepsy will make early diagnosis and better outcome.

Keywords: Cerebral hemiatrophy, Recurrent seizures, Hemiparesis, DDMF

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Introduction

Dyke-Davidoff-Masson syndrome (DDMS) is an uncommon disease characterized by seizures, hemiparesis, facial asymmetry, and mental retardation with varied psychological manifestation with distinct features on cross-sectional radiological imaging.[1] DDMS is occasionally seen in clinical practice but one of the important causes of recurrent and refractory seizures. The clinical presentation as well as the radiological features may be variable depending on the age and extent of cerebral insult. We report a case of an acquired form of DDMS in a 20-year-old unmarried female with a history of on and off seizures with hemiparesis and presently on antiepileptic drugs.

Case Report

A 20-year-old young unmarried female born of a non-consanguineous marriage with an uneventful birth history presented with a history of generalized tonic-clonic seizures and some behavioral problem since the age of 6 years. She had a history of non-progressive weakness in the right half of his body since the age of 9 years. The behavioral problems such as disturbed sleep, irritability, anger outburst, increased psychomotor activity, suspiciousness and irrelevant talks were found. The patient did not attend school.

On examination, the patient was undernourished and had poor cognitive function. Vitals were normal. On neurological examination, she had moderate mental retardation (intelligent quotient = 50), right-sided facial palsy, and upper motor neuron type of subtle right hemiparesis involving both upper and lower extremity. No neurocutaneous markers were present. Examination of other systems was unremarkable. Various laboratory investigations were with in normal range.

She underwent various cross-sectional imaging. Axial non-contrast computed tomography (CT) of brain depicts left cerebral hemisphere atrophy with dilatation of ipsilateral lateral ventricle with a bone window showing thickening of the left hemicranium, almost twice the thickness of that on the right side in the temporal region and hyperpneumatization of the left frontal sinuses [Figure 1].

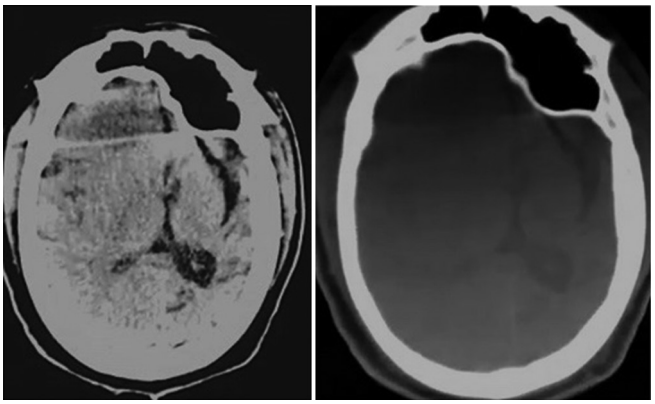

Figure 1: Axial non-contrast computed tomography head (brain and bone window). *Images showing the left cerebral hemispheric cortical atrophy dilation of lateral ventricle with leftsided calvarial thickening and the left side hyperpneumatization of the left frontal sinus

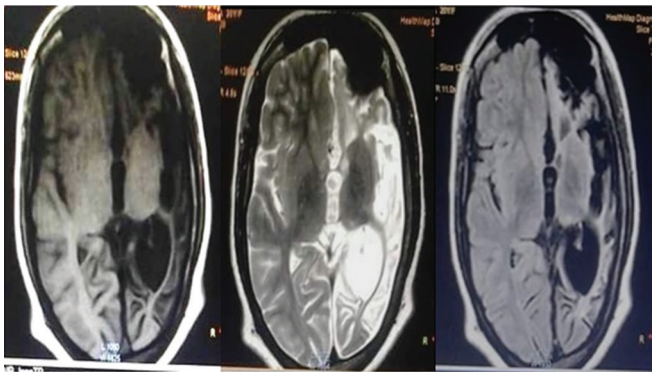

Figure 2: Axial magnetic resonance imaging brain T1, T2, T2-fluid attenuation inversion recovery (FLAIR) sequences. *Images showing left cerebral hemispheric cortical atrophy with encephalomalacia which is hypointensites on T1 and hyperintensities on T2 and T2 FLAIR with ex vacuo dilation of lateral ventricle with left-sided calvarial thickening and left side hyperpneumatization of the left frontal sinus

In addition to that magnetic resonance imaging (MRI) brain demonstrates diffuse cortical and subcortical atrophy as hyperintensities on T2, T2 fluid attenuation inversion recovery, and hypointensities in T1 sequences; however, the left basal ganglia were relatively spared [Figure 2].

With the above findings, a diagnosis of DDMS was finally made. The patient was treated with three antiepileptic drugs and was followed up for 5 months. However, the seizures were poorly controlled. She was referred to higher center for better interventions.

Discussion

Dyke, Davidoff, and Masson in a series of nine patients with clinical features of hemiparesis, seizures, facial asymmetry, and mental retardation described this specific syndrome called DDMS in 1933.[1] This syndrome refers to atrophy or hemiatrophy of one cerebral hemisphere which is secondary to brain insult in the fetal or early childhood period. Predominantly, there is no sex predilection as well as any particular cerebral hemisphere involvement. However, involvement of the left side and male gender has been shown to be more common in literature.[2]

DDMS has varied form of clinical presentation depending on the extent of the brain injury starting from facial asymmetry, seizures, unilateral hemiplegia, or hemiparesis to mental retardation along with learning disabilities and rarely, patients can have neuropsychiatric symptoms.[3] In our patient, seizures for months or years are the cause behind mental retardation, and similar finding is also noted by Sharma *et al*.[3,4]

DDMS broadly categorized into two distinct forms, the infantile or congenital form which becomes symptomatic in the infancy or perinatal period and results from fetal vascular occlusion involving unilateral cerebral arterial circulation specifically middle cerebral artery territory anomalies, coarctation of the midaortic arch culminating in mesencephalon hypoplasia, and Wallerian degeneration.[5] In acquired DDMS, the proposed etiologic factors are trauma, hemorrhage, ischemia, infection, birth asphyxia, prolonged febrile seizures, and tumor. It is mainly due to several ischemic episodes resulting from variable causes, which, in turn, reduces brain-derived neurotrophic factors production, which finally ends in cerebral atrophy.[3,6]

In an illustrative way to know the pathogenesis, we have to go through the development of the brain precisely. The brain sulci formation occurs between the fourth to end of the 8 months of fetal life.[7] However, the maximum growth of a child's head reaches half of its adult size at the end of 1st year and three-fourths of the adult size by the end of 3 years, outward pressure of the enlarging human brain on the bony skull is cause behind rapid growth of head circumference.[3] Hence, whenever brain damage is sustained before 3 years of age, bony skull overlying the brain grows inward resulting in an increased width of the diploic spaces, paranasal sinuses, and elevation of petrous ridge and orbital roof, which are telltale features of this disorder.[8]

The plain skull radiograph illustrates thickening of the calvarium and dilatation of the ipsilateral frontal and ethmoid sinuses. CT and MRI show unilateral atrophy of the cerebral hemisphere with an ipsilateral shift of the ventricle, widening of sulcal spaces on the involved side. It is associated with compensatory calvarium thickening, hyperpneumatization of the paranasal sinuses and mastoid cells, and elevation of the petrous ridge.[9] In congenital hemiatrophy, when the insult occurs *in utero*, there is a shift of midline structures toward the disease side, but there is the absence of sulcal prominence replacing the gliotic tissue. This is the salient feature differentiating congenital from acquired form.[6]

To differentiate congenital from acquired type of DDMS, a thorough clinical history from the parents and features of CT or MRI is two keys to unlock the diagnosis. In a patient with cerebral hemiatrophy, Rasmussen encephalitis, Sturge-Weber syndrome, Silver-Russell syndrome are common differential diagnoses to be considered. These three can be differentiated by thorough clinical examination and cross-sectional neuroimaging. Rasmussen encephalitis does not show calvarial changes with almost similar clinical history, and Sturge-Weber syndrome additionally shows enhancing pial angiomas and cortical calcifications and facial port-wine nevus.[10,11] Silver-Russell syndrome has the classical facial phenotype, clinodactyly, delayed bone age with normal intelligence, and normal head circumference.[12]

Refractory seizures remain the usual concern in DDMS patients.[7] The treatment is symptomatic and must oriented to treat convulsion, hemiplegia, hemiparesis, and learning difficulties. Prognosis is better if hemiparesis occurs after the age of 2 years and without prolonged or repetitive seizures. Children with intractable seizures are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases.[13] Hence, early diagnosis makes early decision-making and intervention. In our setting, the perinatal hypoxic injury is one of the causative factors of DDMS, hence, the role of proper obstetric care in preventing such conditions.

Conclusion

DDMS is a rare preventable cause of refractory epilepsy. Adult presentation of DDMS is unusual and has not often been reported in medical literature. A thorough history and imaging can give an early diagnosis and differentiate from other close differentials. Treatment is largely supportive and aims at controlling seizures along with physiotherapy, occupational therapy, and speech therapy.

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