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Case Report

Dyke-Davidoff-Masson Syndrome: A Case Report of an Adolescent Boy at a Tertiary Hospital in Adamawa, North-Eastern Nigeria

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Received: 24-Nov-2021;
Revision: 17-Feb-2022;
Accepted: 06-Apr-2022;
Published: 22-Sep-2022

Abstract

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare clinical condition in which atrophy or hypoplasia of one cerebral hemisphere occurs secondary to brain insult during fetal or early childhood, which results in variable clinical manifestations like hemiparesis, seizures, expressive aphasia, and mental retardation. This rare entity mainly presents in childhood and is unusual in adults. DDMS is a rare cause of epilepsy and should be considered and excluded in cases of refractory seizures. Few cases have been reported from a developing nation like Nigeria but not from the north-eastern part of Nigeria to the best of our knowledge. Though fewer specialists exist in Adamawa State, efforts to train more specialists and education of medical officers to manage this rare case need to be strengthened. Herein is a case of an adolescent boy with recurrent generalized tonic-clonic convulsions complicated by left-sided hemiparesis, expressive aphasia, and mental age equivalent of a six-year-old by the Goodenough draw-a-person test. Found to be obese with Body Mass Index (BMI) of 29 kg/m^2 (Z-score >2 Standard deviation SD), microcephaly, Occipito-frontal Circumference (OFC) of 45 cm (Z-score > –3 SD), spastic left-sided hemiplegia and hemiplegic gait. Magnetic resonance imaging (MRI) of the brain showed hyper-intensity in the right cerebral hemisphere, extensive atrophy of the right cerebral hemisphere involving the ipsilateral fronto-temporoparietal lobes, cerebral peduncle, and a contralateral megalencephaly, ipsilateral lateral ventricular dilatation, hypertrophic calvarium, hyperpneumatization of sphenoidal sinuses and midline shift due to loss of volume on the right. A diagnosis of Dyke-Davidoff-Masson syndrome was made; the patient did well on carbamazepine and physiotherapy. Caregivers were counseled, and the patient was discharged home and is currently on a follow-up visit.

Keywords: *Adamawa, atrophy, cerebral hemisphere, Dyke-Davidoff-Masson syndrome, epilepsy, hypoplasia*

Introduction

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare clinical condition in which atrophy or hypoplasia of one cerebral hemisphere (hemi-atrophy) occurs secondary to brain insult during the fetal or early childhood period.[1] Its clinical presentation is variable and largely depends on the extent of brain injury.[2] It presents with seizures for which drug therapy may be insufficient and in some cases, requires a surgical intervention, which has become the major concern in the management of this syndrome.[3] As hemispherectomy is hard to find even in many urban tertiary care centers, it is vital for a pediatrician or neurologist to diagnose this ailment timely by means of suitable neuroimaging in order to institute timely treatment.[4] This syndrome is a cause of epilepsy; although rare, it has to be considered and excluded, especially in cases of refractory seizures.

Access this article online

Quick Response Code:

Website: www.njcponline.com

DOI: 10.4103/njcp.njcp_1955_21

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How to cite this article: Usman AB, Emmanuel P, Namuya NP, Manchan DB, C Dahiru AM, Raheem N, *et al.* Dyke-davidoff-masson syndrome: A case report of an adolescent boy at a tertiary hospital in Adamawa, North-Eastern Nigeria. Niger J Clin Pract 2022;25:1604-7.

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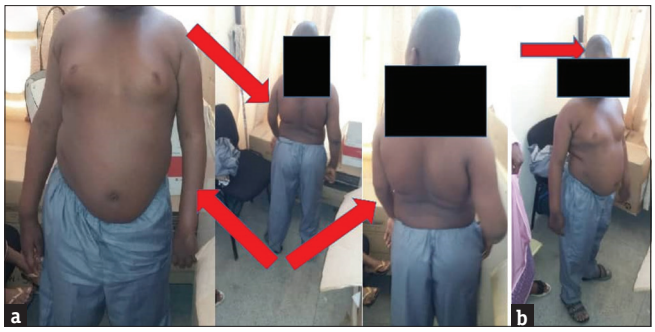

Figure 1: Picture showing hemiatrophy of the left side ipsilaterally (a) and Microcepahaly (b) respectively

Because it is a rare condition, few cases have been reported especially in developing nations. None, to the best of our knowledge, has been published in the north-eastern part of Nigeria, in the Adamawa state, where the current case is being reported. Most doctors here are medical officers with few specialists, and there is a need for medical officers handling cases of epilepsy to be aware of this condition to enhance their productivity. Herein is a case report of epilepsy associated with left hemiparesis, microcephaly, and mental retardation in a 12-year-old boy diagnosed with DDMS following neuroimaging combined with clinical features.

Patient and Case Report

A 12-year-old boy, a product of full-term uncomplicated normal vaginal delivery, presented with recurrent generalized tonic-clonic convulsions since early childhood, complicated by left-sided hemiparesis, expressive aphasia, and a mental age equivalent of a six-year-old by the Goodenough draw-a-person test. Examination revealed an obese child with a Body Mass Index (BMI) of 29 kg/m^2 (Z-score >2 Standard deviation SD), microcephaly Occipito-frontal Circumference (OFC) of 45 cm (Z-score >–3 SD), spastic left-sided hemiplegia as well as hemiplegic gait [Figure 1a and 1b]. Other systemic examinations were unremarkable. Magnetic resonance imaging (MRI) of the brain showed hyper-intensity in the right cerebral hemisphere, denoting an extensive atrophy of the right cerebral hemisphere involving the ipsilateral fronto-temporoparietal lobes and cerebral peduncle and a contralateral megalencephaly [Figure 2a and 2b]. There was associated ipsilateral lateral ventricular dilatation, hypertrophic calvarium, hyperpneumatization of sphenoidal sinuses, and midline shift due to loss of volume on the right [See Figure 2b-d]. A diagnosis of Dyke-Davidoff-Masson syndrome was made. He was placed on carbamazepine, counseled, and commenced physiotherapy. He responded to treatment, was discharged, and is currently on a follow-up visit as an outpatient.

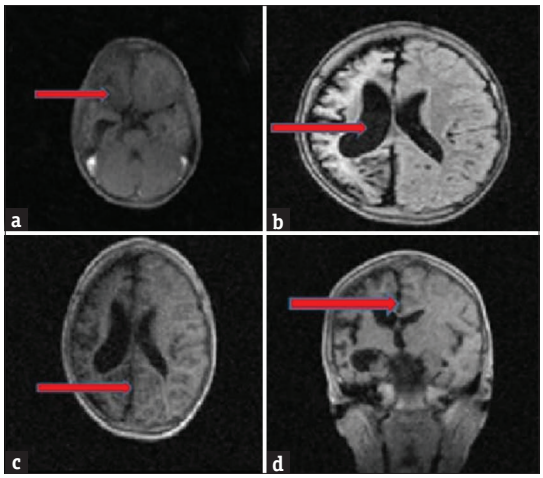

Figure 2: MRI showing right cerebral hemiatrophy and encephalomalacia (a) dilated lateral ventricle, mass effects, cerebral hemiatrophy and hypertrophic calvarium (b and c), and hyperpneumatisation of sinuses (d) respectively

Discussion

Dyke–Davidoff–Masson Syndrome comprises clinical features that range from hemiparesis, seizure, facial asymmetry, intellectual disability, and aphasia, depending on the degree of insult to the brain.[5] All of the above, except facial asymmetry, were seen in the index case. He is still not enrolled in school because of his intellectual disability and expressive aphasia.

Dyke–Davidoff–Masson Syndrome results from cerebral insult which occurred either in utero or early in life. Two forms of DDMS exist. In the congenital form, insult occurs during the perinatal period or in early infancy. In the acquired form, which is usually seen in adolescents or adults, the features of the ailment depend on the time of insult.[6] The cause of the congenital form is often from vascular insult during intrauterine life, which culminates in hypoplasia of a cerebral hemisphere and loss of brain sulci. The acquired form usually results from trauma, infection, vascular abnormalities, and intracranial hemorrhage in the perinatal period or shortly thereafter.[7] The present case most likely was the acquired form, because clinical features were picked in early childhood, MRI showed encephalomalacia, and the brain sulci were present, even though history pointing to cerebral insult from aforementioned causes was difficult to verify. Hypertrophic calvarium in the patient further indicates that brain injury was probably in infancy or early in toddlerhood. This is on the premise that portrays such changes occurring in patients who were exposed to brain injury early enough.[8] The pathogenesis of cerebral atrophy and the associated progressive neurological

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deficit results from ischemic episodes, which decrease the production of brain-derived neurotrophic factors with consequent cerebral atrophy.[9] Dyke–Davidoff–Masson Syndrome is sometimes accompanied by crossed cerebro-cerebellar atrophy, which is reasoned by some to be a continuum of cerebro-cerebellar diaschisis. Therefore, it may not be surprising for the recent case to present in the future with cerebro-cerebellar diaschisis due to the continuum nature of the condition. Frequent and excessive excitatory input during seizures via glutaminergic corticopontine–cerebellar pathways has been postulated to induce cerebellar atrophy in this disorder.[10] Three different patterns of DDMS are visualized on MRI. In the first pattern (Pattern I), a diffused cortical and subcortical atrophy is seen. Pattern II is characterized by diffused cortical atrophy coupled with a porencephalic cyst, while in pattern III, a previous infarction with gliosis in the middle cerebral artery (MCA) territory is seen.[5] Our patient presented with conditions corresponding to the pattern found in pattern I.

A series 19 DDMS recently revealed that all the patients experienced seizures with ipsilateral ventricle dilatation and cortical sulcus enlargement as the most common MRI findings. It occurred more in males like index cases and was thought to be linked to the presence of circulating androgens in the developing male brain. This generates a hyperplastic condition resulting in more extensive neuronal remodeling after injury than in the female brain.[11] Our patient had atrophy of the right cerebral hemisphere, including ipsilateral fronto-temporoparietal lobes and cerebral peduncle. Additionally, he had associated right lateral ventricular dilatation, midline shift, hypertrophic calvaria, hyperpneumotization of sphenoidal sinuses, and contralateral megalencephaly. The later findings indicated compensatory changes taking place in the brain, diploic spaces of the calvarium, and air cells of the sinuses.[1]

Differential diagnosis of DDMS includes Sturge Weber syndrome, which will show cerebral calcifications (tram track sign or railroad sign). Linear-Nevus syndrome having mental retardation, epilepsy, and hemimegaloencephaly. Fishman syndrome will show cranial lipoma with lipodermoid of the eye in addition to calcified cortex and hemiatrophy on neuroimaging. Others are Silver-Russell syndrome having a typical triangular face, prominent chin, broad forehead, wide mouth, short stature, delayed bone age, clinodactyly, and hemihypertrophy. Rasmussen encephalitis will demonstrate cerebral hypoplasia on neuroimaging; however, it does not affect the calvarium.[12] Remote differentials include basal ganglia germinoma and progressive multifocal leukoencephalopathy. Neuroimaging of the former shows cystic and edematous areas, and the latter will exhibit encephalomalacia.[5] After careful clinical scrutiny of these differentials, the argument in favor of DDMS in the current case cannot be overemphasized.

Management involves the use of appropriate anticonvulsants to control seizures, as most patients with DDMS present with refractory seizures.[3] Gladly, his response to carbamazepine was unprecedented. In addition, physiotherapy, occupational therapy, and speech therapy are an important part of the management of these patients. In patients with hemiplegia and intractable disabling seizures, hemispherectomy is indicated. Hemispherectomy has been successful in 85% of the cases.[6] In settings such as ours, it is hard to find a different specialist to provide such care.

Poor prognosis is noticed in patients with prolonged or recurrent seizures and in a situation where hemiparesis occurs prior to age two years. Therefore, it is necessary that pediatricians, neurologists, and radiologists be familiar with DDMS to promptly diagnose and initiate treatment.[6]

Conclusion

Dyke–Davidoff–Masson Syndrome as a cause of epilepsy, although rare, should be considered and excluded in cases of refractory seizures. Few cases have been reported from a developing nation like Nigeria and not from Adamawa state in the north-eastern part of Nigeria to the best of our knowledge. Dyke–Davidoff–Masson Syndrome should be suspected in children presenting with generalized convulsions complicated by one-sided hemiparesis, expressive aphasia, and mental retardation. Most doctors in Adamawa State are medical officers, with only a handful of specialists rendering recognition and management of such cases a huge challenge. There is thus a need to train more specialists and encourage medical officers handling cases of epilepsy to increase their index of suspicion and equip themselves with its diagnosis and treatment.

Ethics approval and consent to participate

Every aspect of the management of this patient, including using her case for educational purposes and teaching other health workers, has been consented by the patient and her relatives.

Consent for publication

All authors consented.

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Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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