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Case Report

Dyke–Davidoff–Masson syndrome in a Nigerian

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ABSTRACT

Dyke–Davidoff–Masson syndrome (DDMS) is a rare, but important cause of drug-resistant seizures. Dyke–Davidoff–Masson syndrome is a constellation of clinical features that consists of hemiparesis, seizure, facial asymmetry, and intellectual disability with distinct neuroimaging features. A 27-year-old lady presented to us with drug-resistant epilepsy, hemiparesis, and intellectual disability that necessitated her withdrawal from school. Her brain magnetic resonance imaging (MRI) showed cerebral hemiatrophy, calvarial thickening, and hyperpneumatization of the frontal sinuses consistent with DDMS. We discuss the diagnostic and therapeutic implications of DDMS and advocate early referral and evaluation of people with epilepsy in sub-Saharan African settings.

1. Introduction

Although a rare, but an important cause of drug resistant seizures, the Dyke–Davidoff–Masson syndrome (DDMS) is not a difficult diagnosis to make in the era when neuroimaging has profoundly improved epilepsy care. Acquired or congenital (infantile) cerebral hemiatrophy, also referred to as DDMS, is a diagnostic constellation made up of hemiparesis, facial asymmetry, intellectual disability, and epilepsy. On brain imaging, the findings of cerebral hemiatrophy, calvarial thickening, and hyperpneumatization of the frontal sinuses provides radiographic support for the clinical diagnosis [1]. The significance of an early and accurate diagnosis is the prospect of improving the patient's prognosis and improving their quality of life. This case illustrates a unique epilepsy syndrome and the need for a heightened awareness among epilepsy care providers. Furthermore, it emphasizes the importance of early evaluation of patients with epilepsy in sub-Saharan Africa where the diagnostic gap is still close to 40% [2]. We report a case of DDMS in a Nigerian with childhood-onset epilepsy and review the literature.

2. Case report

A 27-year-old female presented to our clinic with her father because of recurrent seizures and poor scholastic performance. There was no history of maternal illness during pregnancy. Her delivery was normal, and she cried immediately after birth. She had normal development until she was 2 years when she had a febrile seizure which was described as focal clonic involving the left side of the body. The seizure evolved to a bilateral convulsion. She then became unconscious for three days. On discharge, she had residual left hemiparesis.

About 15 years prior to presentation, she developed recurrent generalized tonic-clonic seizures. She demonstrated impaired cognition and poor scholastic performance which prevented her from completing her secondary education. She had been on phenobarbital, carbamazepine, and phenytoin sodium at different times. She presented with poor seizure control and gingival hyperplasia. Her general physical examination revealed a young female who was small for her age, with a weight of 43.7 kg and a height of 1.56 m (body mass index, BMI = 17.9 kg/m^2). Her score on Ravens Standard Progressive Matrices (RSPM) was 11/60, indicating intellectual impairment (below the 5th percentile for her age). Neurologically, she had left facial hemiatrophy, left hemibody atrophy, and spastic hemiparesis (power of 4/5 in both left upper and lower limbs with a left extensor plantar response. The rest of the examination was normal. On clinical grounds, we suspected Rasmussen encephalitis and requested a cranial magnetic resonance imaging (MRI) and an electroencephalogram (EEG). Her brain MRI (See Fig. 1.) revealed right cerebral hemiatrophy with features of encephalomalacia surrounded by gliosis (Figure). Furthermore, on MRI, she had right calvarium thickening and hyperpneumatization of the frontal sinus. Her EEG revealed a low amplitude background over the right hemisphere with intermittent bursts of generalized sharp-and-slow wave complexes. These features place her in the ILAE category of epilepsy with structural–metabolic causes [3]. She is presently on valproic acid 400-mg twice daily. Her seizure frequency has been

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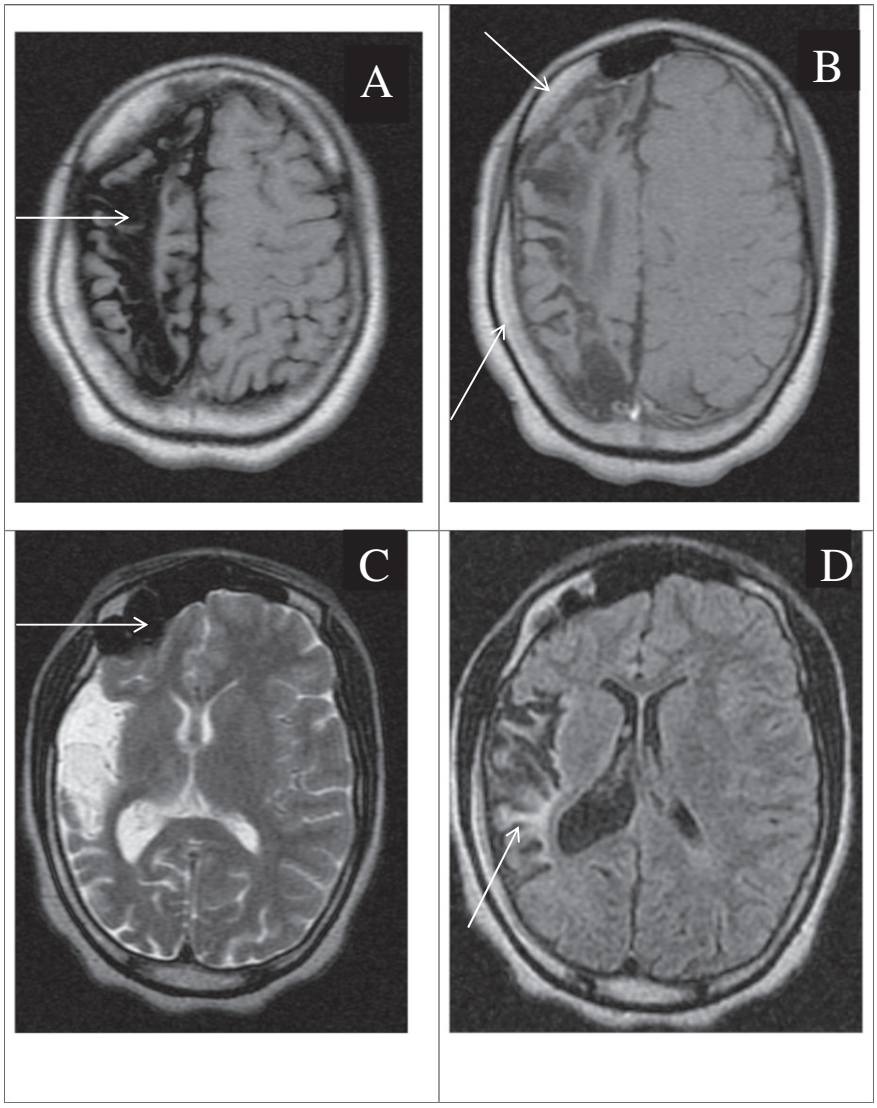

Figure. Axial sequences of brain MRI in DDMS of the patient. A. Axial T1-weighted MRI showing right cerebral hemiatrophy with prominent sulci and encephalomalacia. B. Axial T1-weighted MRI showing thickened calvarium on the right. C. Axial T2-weighted MRI showing right frontal sinus enlargement and hyperpneumatization. D. Axial T2 FLAIR showing abnormal signal intensity consistent with gliosis.

reduced from "at least once a week" to "less than weekly but at least once a month". She is also currently undergoing regular physical and cognitive rehabilitation.

3. Discussion

The findings of cerebral hemiatrophy, calvarial thickening, and hyperpneumatization of the frontal sinuses on MRI in the setting of drug-resistant seizure, intellectual impairment, and hemibody atrophy are the diagnostic criteria for DDMS in this case. For epilepsy care givers, our case illustrates the fact that seizures associated with hemibody atrophy have a differential diagnosis beyond Rasmussen's syndrome. Acute focal weakness following a seizure in a child has many serious and treatable causes. Although benign etiologies such as Todd's paralysis and metabolic etiologies such as glucose, sodium, potassium, calcium, and magnesium abnormalities are easily identifiable, structural lesions such as neoplasms, intracerebral abscesses, acute disseminated encephalomyelitis, developmental brain malformations, and signs of trauma and vascular disorders such as ischemic and hemorrhagic infarctions should be excluded [4]. Hemiconvulsion-hemiplegia-epilepsy (HHE) syndrome should also be entertained. In HHE syndrome, neuroradiological studies may show unilateral edematous swelling of the ipsilateral hemisphere at the time of initial status epilepticus (SE) [4]. Although neuroimaging studies were not conducted at the time of the initial insult in the presented case, the generalized tonic-clonic seizures after hemiconvulsions, which characterized this patient's presentation, are less typical of HHE syndrome, which is characterized by hemiconvulsions alone. This is in addition to the long latency (10 years) between the acute encephalopathy and the onset of epilepsy (typically 2 months to 2 years in HHE) [5], though the two syndromes have been recognized synonymously [6].

Other differential diagnoses should include Sturge–Weber syndrome, basal ganglia germinoma, Silver–Russel syndrome, Fishman syndrome, and linear nevus sebaceous syndrome. In Rasmussen's syndrome, the calvarial changes are not found. Basal ganglia germinoma presents with similar clinical features, but brain imaging reveals cystic areas, focal hemorrhages, and mild surrounding edema along with calvarial changes [7]. In Sturge–Weber syndrome, brain imaging shows enhancing pial angiomas and cortical calcifications in addition to the clinical features of the phacomatosis. Silver–Russell syndrome is characterized by the classical facial phenotype (triangular face; small, pointed chin; broad forehead; and thin, wide mouth), poor growth with delayed bone age, clinodactyly, hemihypertrophy with normal

head circumference, and normal intelligence [7]. Fishman syndrome or encephalocraniocutaneous lipomatosis is a rare neurocutaneous syndrome including unilateral cranial lipoma with lipodermoid of the eye, which usually presents with seizures. Neuroimaging, however, shows a calcified cortex and hemiatrophy [7]. Linear nevus sebaceous syndrome (a.k.a. Schimmelpenning-Feuerstein–Mims syndrome) is characterized by a typical facial nevus, mental retardation, recurrent seizures, and hemimegaencephaly [8].

The etiology of DDMS is due to a cerebral insult that may occur in utero or early in life. There are two forms of DDMS; acquired and congenital (infantile) forms. The congenital form presents during the perinatal period or in early infancy. The acquired type depends on the time of the insult and may be seen in adolescent or adults [1]. In the congenital type, there is a midline shift towards the affected hemisphere, and the sulcal prominence replacing the gliotic tissue is absent [9]. Three patterns of DDMS are recognized on MRI. Pattern I corresponds to diffuse cortical and subcortical atrophy. Pattern II corresponds to diffuse cortical atrophy coupled with a porencephalic cyst, while pattern III corresponds to previous infarction with gliosis in the middle cerebral artery (MCA) territory [10]. The clinical profile and imaging features in our patient are in keeping with acquired DDMS pattern III.

The mechanism of cerebral atrophy and the related progressive neurological deficit are posited to be due to ischemic episodes which reduce the production of brain-derived neurotrophic factors, which, in turn, lead to cerebral atrophy [11]. In some cases, DDMS is accompanied by crossed cerebro-cerebellar atrophy, which is thought to be a continuum of cerebro-cerebellar diaschisis. It has been postulated that frequent and excessive excitatory input during seizures via glutaminergic corticopontine–cerebellar pathways induces cerebellar atrophy in this group of patients [12]. The etiology of DDMS in our case was due to protracted febrile seizures. The 3 days of unconsciousness was due to either non-convulsive status epilepticus, a prolonged postictal state, or a coma of unknown etiology. The possible etiological association between DDMS, prolonged febrile seizures, and middle cerebral artery stroke has been described [9]. Because of the high incidence (11–18%) [13] of febrile seizures in our environment, with nearly 4% of patients going on to develop epilepsy [13], inappropriate management of febrile seizure should be highly discouraged to forestall untoward outcomes.

Medical treatment of DDMS includes rational use of antiseizure drugs. If seizures are drug-resistant, they can be treated surgically with hemispherectomy, which has an 85% chance of rendering the patient seizure free [7]. Long-term management also includes physiotherapy, occupational therapy, and speech therapy. We could not offer hemispherectomy to our patient because epilepsy surgery and other tertiary care epilepsy management, including vagus nerve stimulation is still unavailable in our region of Nigeria [14]. Currently, management of epilepsy is still limited to rational use of the antiseizure drugs. The relatively high cost of ASDs, on the background of high unemployment, poor remuneration, and out-of pocket payment for treatment, facilitates poor adherence to medications and widening of the treatment gap [15]. Efforts at rapid resolution of acute symptomatic seizures should be advanced to prevent untoward outcome. Early diagnosis by an appropriate imaging study, optimization of medical management of the seizures, and enhancement of neurorehabilitation are equally important.

4. Conclusion

To the best of our knowledge, this is the first case of DDMS described in our environment in Nigeria, although it has been described in other lower-middle income countries like Iran, India, Tunisia, and Turkey. The paucity of neuroimaging facilities in our region may deter early recognition, and, therefore, a high clinical index of suspicion is required. We advocate prompt referral of patients with suspected DDMS and refractory epilepsy to regional specialty care facilities in underserved countries to facilitate an early diagnosis and to provide optimal regional treatment.

Conflict of interest

The authors declare no conflicts of interest.

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References

[1] Arora R, Rani JY. Dyke–Davidoff–Masson syndrome: imaging features with illustration of two cases. Quant Imaging Med Surg 2015;5(3):469–71.
[2] Nwani PO, Nwosu MC, Enwereji KO, Asomugha AL, Arinzechi EO, Ogunniyi AO. Epilepsy treatment gap: prevalence and associated factors in Southeast Nigeria. Acta Neurol Scand 2013;128(2):83–90.
[3] Berg AT, Berkovic SF, Brodie MJ, Buchhalter J, Cross JH, van Emde BW, et al. Revised terminology and concepts for organization of seizures and epilepsies: report of the ILAE Commission on Classification and Terminology, 2005–2009. Epilepsia 2010; 51(4):676–85.
[4] Tenney JR, Schapiro MB. Child neurology: hemiconvulsion–hemiplegia–epilepsy syndrome. Neurology 2012;79(1):e1–4.
[5] Saito T, Saito Y, Sugai K, Nakagawa E, Komaki H, Okazaki T, et al. Late-onset epilepsy in children with acute febrile encephalopathy with prolonged convulsions: a clinical and encephalographic study. Brain Dev 2013;35(6):531–9.
[6] Chand G, Goel R, Kapur R. Dyke–Davidoff–Masson syndrome. Arch Neurol 2010; 67(8):1026 [Internet].
[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.
[8] Herman TE, Siegel MJ. Hemimegalencephaly and linear nevus sebaceous syndrome. J Perinatol 2001;21(5):336–8.
[9] Zawar I, Khan AA, Sultan T, Rathore AW. Dyke–Davidoff–Masson syndrome. An unusual cause of status epilepticus. Neurosciences (Riyadh) 2015;20(4):385–7.
[10] Singh P, Saggar K, Ahluwalia A. Dyke–Davidoff–Masson syndrome: classical imaging findings. J Pediatr Neurosci 2010;5(2):124–5.
[11] Ono K, Komai K, Ikeda T. Dyke–Davidoff–Masson syndrome manifested by seizure in late childhood: a case report. J Clin Neurosci 2003;10(3):367–71.
[12] Algahtani HA, Aldarmahi AA, Al-Rabia MW, Young GB. Crossed cerebro-cerebellar atrophy with Dyke Davidoff Masson syndrome. Neurosciences (Riyadh) 2014; 19(1):52–5.
[13] Eseigbe EE, Adama SJ, Eseigbe P. Febrile seizures in Kaduna, north western Nigeria. Niger Med J 2012;53(3):140–4.
[14] Adebayo PB, Akinyemi RO, Ogun SA, Ogunniyi A. Seizure severity and health-related quality of life of adult Nigerian patients with epilepsy. Acta Neurol Scand 2014; 129(2):102–8.
[15] Ezeala-Adikaibe B, Onwuekwe I, Ekenze S, Achor J, Onwukwe J. Estimated direct cost of treating epilepsy per month in Southeast Nigeria. Internet J Neurol Internet Sci Publ 2009;13(1).