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CASE REPORT

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An unusual case of Dyke–Davidoff–Masson syndrome revealed by status epilepticus in a Malian patient

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Abstract

The Duke–Davidoff–Masson syndrome (DDMS) is a rare neurological condition with unknown prevalence, globally. To date, <100 cases have been reported worldwide. We report the case of an 18-year-old patient admitted for status epilepticus seizure, and who presented a right hemiparesis, body asymmetry, joints ankylosis, and mental retardation. Brain CT-scan revealed left hemisphere atrophy, skull bone thickening, and hyperpneumatization of the frontal sinuses; all consistent with DDMS. Seizures improved remarkably on Levetiracetam and Valproate. This is the first report of an unusual DDMS in Mali, and the diagnosis delay highlights the challenges for the management of these diseases in resource-limited settings.

KEYWORDS

Africa, Dyke–Davidoff–Masson syndrome, Mali, resistant epilepsy, status epilepticus

1 | INTRODUCTION

The Duke–Davidoff–Masson syndrome (DDMS) is a rare neurological condition with unknown incidence, globally. This disease was first described by three physicians, Cornelius G. Dyke, Leo M. Davidoff, and Clement M. Masson, in 1933.[1] DDMS is a clinico-radiological recognized entity characterized by typical manifestations including drugs resistant epilepsy, intellectual disability, hemiparesis, skull bone thickening associated with cerebral hemiatrophy and hyperpneumatization of paranasal and frontal sinuses on neuroimaging.[2] Some patients may present with additional symptoms, including cerebellar and basal ganglia atrophy, ear malformations, and neuropsychiatric disorders.[3,4] DDMS etiologies are broadly categorized into congenital and acquired.[5] Congenital causes of DDMS include intrauterine vascular injury and cerebral hemispheric hypoperfusion.[6,7] Acquired causes mostly derive from birth trauma, periventricular leukomalacia, cerebral hemorrhage, cerebral infarction, cerebral infection, radiation, postictal cerebral hemiatrophy, and prolonged febrile seizures.[6,7] Almost a century after the first description, the pathogenic mechanism remains unclear and is subject to controversy. To date, <100 cases have been reported worldwide, with only four cases in the African population.[8–11] We report here the case of an

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18-year-old adolescent who was admitted to our neurology clinic for convulsive status epilepticus and in which clinical and laboratory findings were in favor of DDMS, the first case in Mali.

2 | CASE PRESENTATION

An 18-years-old male was admitted to our neurology clinic for recurring tonic–clonic seizures that started 3 days prior to his admission. Medical history was consistent with a full-term infant, born from healthy and nonconsanguineous parents after an uneventful pregnancy. However, delivery was done by cesarean section due to dystocia. My family history was unremarkable. He had mental and motor acquisition delay because he could not sit after 1 year of age and walked after age two. Their parents reported that he presented focal tonic–clonic seizures in the right half of his body at 1 year of age, at least once a day, that worsened over time. He was initially put on a progressive dose of Valproic Acid (up to 1000 mg/day) over 3 months, which did not change the disease course. Then, Phenobarbital (100 mg/day) was added, which lasted at least 1 year. On this treatment, the frequency of the seizures initially dropped with a maximum of three episodes daily before increasing gradually to more than 10 daily episodes. At the age of 14, the Phenobarbital was changed to Carbamazepine (400 mg/day) and Clobazam (10 mg/day), which poorly controlled the seizures. He also presented frequent falls and injuries due to seizures. Later, they noticed that the right side of his body was smaller and weaker than the left one. In addition, he presented a progressive intellectual disability that led him to drop out of school at the age of nine. On examination, the body temperature, blood pressure, heart and breathing rates, and oxygen saturation were normal. Neurological examination found status epilepticus with right-sided focal tonic–clonic seizures followed by secondary generalization. He had a severe cognitive decline and could not count from one to 10. Mini Mental State Examination (MMSE) was not possible. Body asymmetry and hemiparesis were confirmed in addition to ankylosis in the right wrist and elbow and a prominent forehead (Figure 1A–C). Abdominal, cardiovascular, and pulmonary examinations were unremarkable. He did not present any cutaneous lesions other than scars caused by seizure-related injuries. Blood chemistries, including total blood cell count, liver enzymes, serum creatinine, and blood ions, were normal. EEG performed 2 weeks after admission found a diffuse slow background with delta wave activities in hyperpnea and a continued focal to diffuse pseudo periodic activities (Figure 2A,B). Brain CT-scan revealed pronounced atrophy of the left cerebral hemisphere with ex vacuo of the ipsilateral ventricle. Furthermore, a bilateral abnormal thickening of the skull bone with an

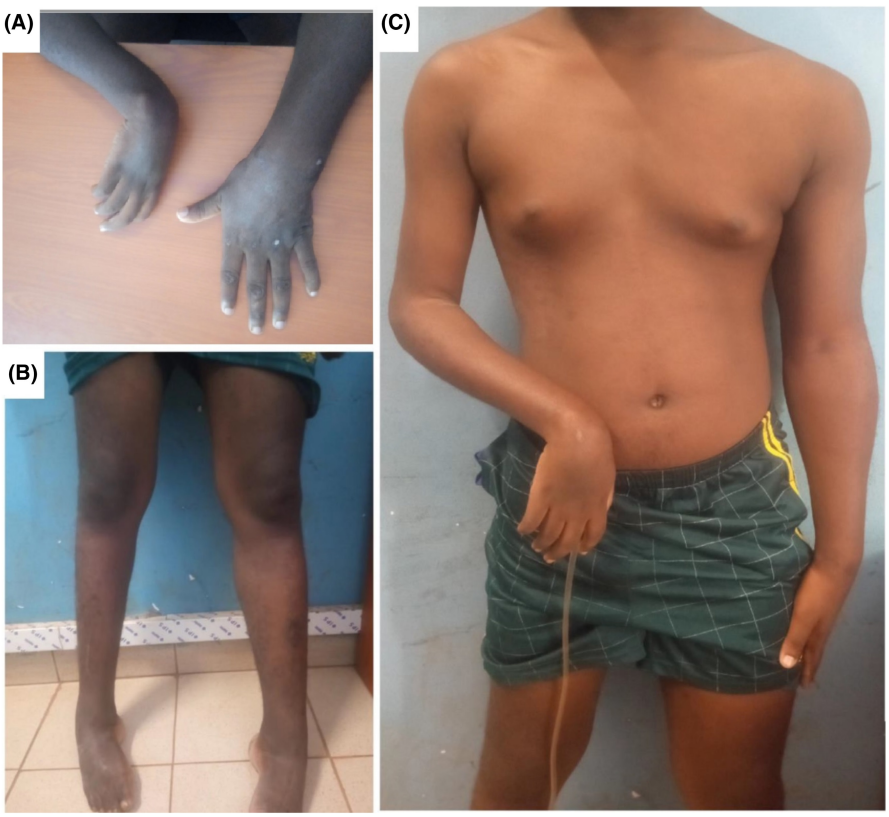

FIGURE 1 Clinical findings in patient with the Duke–Davidoff–Masson syndrome. (A) Images of the patient showing right side hemi atrophy of the body more pronounced in the upper limb. (B, C) Ankylosis of the right wrist and elbow.

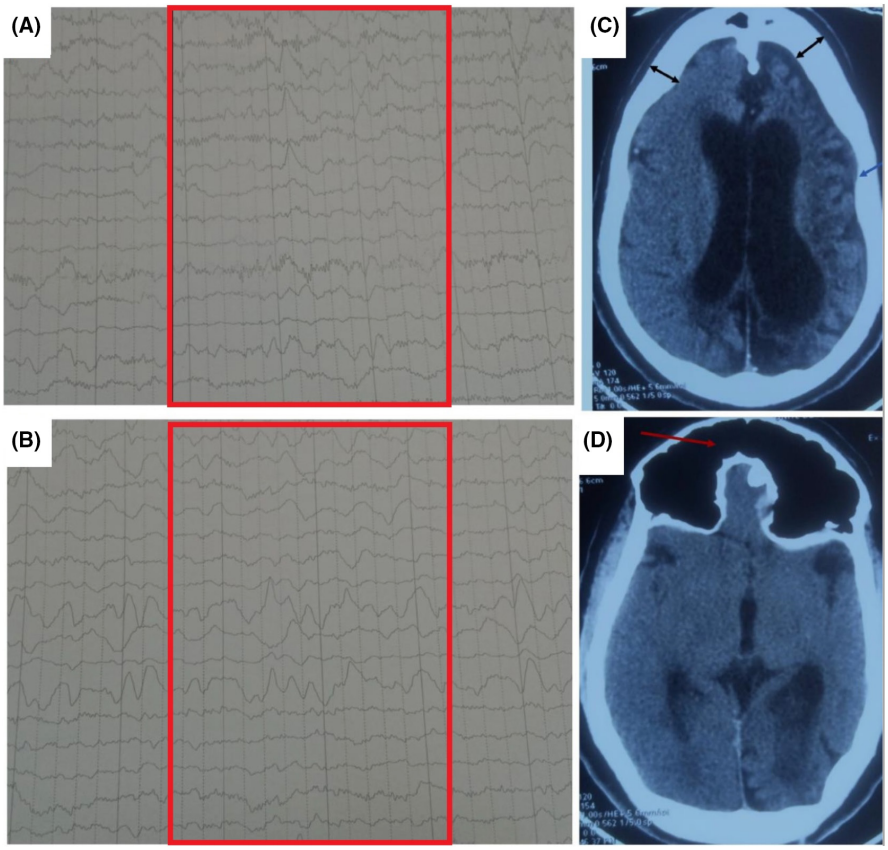

FIGURE 2 Laboratory findings in patient with the Duke–Davidoff–Masson syndrome. (A, B) EEG showing slowed background with delta wave activities in hyperpnea and a continued focal to diffuses pseudo periodic activities (red squares). (C, D) Brain CT-scan in axial cut showing a left hemisphere atrophy (blue arrow), bilateral calvarial thickening (black arrow), and hyperpneumatization of the frontal sinuses (red arrow).

hyperpneumatization of the frontal sinuses was seen (Figure 2C,D). His clinical and imaging features were consistent with the Dyke–Davidoff–Masson syndrome. He was initially put on intravenous Levetiracetam (1500 mg/day), and continuous infusion of Clonazepam (2 mg/day). The disease course remarkably changed to fewer seizures (down to 10 a day). Then, we added oral Valproic Acid (500 mg twice a day) which drastically improved the seizures to a maximum of two seizures daily, and the patient was discharged on this treatment.

3 | DISCUSSION

Dyke, Davidoff, and Masson recognized and reported the first cases of this syndrome in 1933.[1] Almost a century later, <100 cases have been reported worldwide, but its global incidence is still unknown. DDMS falls in the category of epilepsy with brain structural abnormalities of the International League Against Epilepsy classification (ILAE).[12] The clinical manifestations encompass focal and/or generalized drugs-resistant epilepsy, hemiparesis or hemiplegia, facial or body asymmetry with atrophy, and mental retardation as seen in our patient.[2,3] Besides these manifestations, rare cases may include cerebellar atrophy, neuropsychiatric disorders, and ear malformations.[3] However, the disease phenotype can vary from one patient to another, and some may not present epilepsy, mental retardation, or body asymmetry.[13] The patient was referred for convulsive status epilepticus, which led to the diagnosis as seen in previous reports.[14] DDMS is usually diagnosed in childhood, mostly in the first decade, but cases of late diagnosis or adult cases were also reported.[2] Although, the syndrome is easily recognizable by clinical and brain imaging findings, the diagnosis delay was 18 years in our patient. This is likely due to the limited access to specialists who could further investigate with brain imaging, as this was not performed before we saw the patient. A similar case with a long diagnosis odyssey was reported in a patient from Nigeria.[11] Brain CT-scan or MRI have contributed to facilitate the diagnosis by typically showing hemi cerebral atrophy, ipsilateral thickening of the skull bone, and ipsilateral hyper pneumatization of the frontal and paranasal sinuses.[15] In our case, the hyper pneumatization of the frontal and paranasal sinuses and the thickening of the skull bone were bilateral. To the best of our knowledge, these brain imaging findings have not been previously reported. This case could be another phenotypic variant of DDMS that might be due to some specific genetic factors as suggested by the previous studies[2] or to recurring skull injuries due to seizure-related falls. Although, our patient fulfilled the diagnosis criteria of DDMS, other diseases such as Rasmussen's encephalitis, hemiplegia-hemi convulsion epilepsy (HHE)

syndrome, and Sturge Weber syndrome are also possible.[15,16] However, in RE and HHE, there is no skull bone thickening nor a hyper pneumatization of the sinuses that are pathognomonic radiological findings in DDMS, as seen in the patient presented here. In addition, in HHE syndrome, the seizures are in the hemiplegia/hemiparesis side without secondary generalization. The Sturge Weber syndrome is a neurocutaneous syndrome characterized by a facial birthmark, also called port-wine birthmark, and cerebral vascular calcifications, which were absent in our patient. The management of DDMS is challenging due to the resistance to several antiepileptic drugs (AED). The treatment is based on the use of a combination of AEDs, and surgery with hemispherectomy is an option in the case of refractory epilepsy.[14]

4 | CONCLUSION

This is the first report of the Dyke–Davidoff–Masson syndrome is seen in a Malian patient. It highlights the challenges of an early diagnosis and the treatment of this rare condition. We suggest that in the presence of pharmacoresistant epilepsy in childhood and body deformities, further investigations should be undertaken to establish an evidence-based diagnosis for an informed disease management strategy, particularly in low-resource settings where access to the surgery of epilepsy is highly limited or unavailable.

AUTHOR CONTRIBUTIONS

SOD, management, data acquisition, and interpretation; AY, analysis and interpretation, and drafted the first version of the manuscript; AK, HS, PK, CAG, clinical management, and data acquisition; ABM and ASS, critical revision of the manuscript, GL editing of the manuscript and supervision of the study.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

All the data included in this study are available from the corresponding author upon reasonable request.

ETHICAL APPROVAL

None.

CONSENT

The written informed consent was obtained from the parent to publish this report in accordance with the journal's patient consent policy.

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