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CASE REPORT

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Dyke-Davidoff-Masson syndrome—A rare cause of recurrent seizures in adulthood

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Key Clinical Message

It is important to consider DDMS as a differential diagnosis in any patient with early childhood onset of epilepsy. Early diagnosis and optimal management are key to reducing the disabling effect of DDMS.

KEYWORDS

adulthood, Dyke-Davidoff-Masson syndrome, hemiparesis, recurrent, seizures

1 | INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological condition which may be congenital or acquired during childhood as a sequelae of foetal or early childhood brain insult.[1,2] It is one of the uncommon causes of intractable seizures.[3] The syndrome is characterized by contralateral hemiparesis, seizures, facial asymmetry, and mental retardation of varying degrees.[4] Individuals with this syndrome present with different combinations and severities of the characteristic clinical features. It occurs in both genders with a male preponderance (73.5%).[5] Findings on radiological imaging include cerebral hemiatrophy, calvarial thickening, ventriculomegaly and hyperpneumatization of the frontal sinuses.[2,6,7] We report a case of a 22-year-old female with an acquired form of DDMS who presented with recurrent seizures.

2 | CASE PRESENTATION

A 22-year-old female with a known history of seizure disorder starting at the age of 8 years who defaulted medications due to financial constraints was admitted to the Emergency unit with a 1-week history of left-sided weakness and two episodes of generalized tonic-clonic seizures which self-aborted after 45 seconds. These were associated with deviation of the mouth to the right and difficulty swallowing. She was delivered at term with no significant history of antenatal or perinatal complications. She had delayed milestones of development in motor and language. There was also a history of poor performance in school and mild socially disinhibited behavior. On presentation, she was afebrile, with a pulse of 109 beats per minute (bpm), blood pressure of 140/91 mmHg, oxygen saturation (SPO_2) of 99% on room air and random blood glucose of 8.5 mmol/L. She had a body mass index (BMI) of 22 kg/m^2. On neurological

examination, the Glasgow coma scale was 15/15, there was forehead sparing and mouth deviation to the right. For both right upper and lower limbs, power was 5/5 and reflexes (biceps, triceps, and knee jerk) were normal and for both left upper and lower limbs, power was 3/5 with brisk reflexes. Vision and hearing were normal.

2.1 | Differential diagnosis, investigations, and treatment

An initial impression of acute untyped stroke with left hemiparesis and Seizure disorder was made. The patient was started on tablet Levetiracetam 500 mg 12 hourly. A Magnetic Resonance Imaging (MRI) of the Brain was requested, and the patient was put on a seizure monitoring chart. Laboratory showed Hemoglobin (Hb) of 12.9 g/dL, mean corpuscular volume (MCV) of 95.9 fL, Platelets (PLT) of 350×10^3, white blood cell (WBC) of 7.53×10^3/uL, Urea of 3.70 mmol/L, and Creatinine of 53 umol/l. The MRI of the brain showed a reduction in the volume of the right cerebral hemisphere with contiguous abnormal cortical and subcortical T2-weighted-Fluid Attenuated Inversion Recovery (T2w/FLAIR) hyper-intensities noted at the right cerebral hemisphere. Also seen was hyperpneumatization of the right frontal sinus and mastoid air cells and associated calvaria thickening. Additionally, atrophy of the basal ganglia was seen. There was a normal appearance of the brainstem and cerebellum. The falx was centred in the midline. Ventricles appeared normal. No abnormal contrast enhancement was seen. Also, no abnormalities were seen in the internal capsule, corpus callosum, thalamus, brainstem and cerebellum. There were normal cerebellopontine angles, sella, pituitary gland, and parasellar structures and vessels. There was mucosal thickening in the right sphenoid sinus. The other paranasal sinuses and mastoid air cells showed normal development and pneumatization Figure 1.

These features were consistent with DDMS and thus the patient was diagnosed as such. By day 4 on admission, seizures were still persistent but only involved the left side of the face associated with twitching of the mouth lasting for about 15–20 s and seem to be provoked whenever patient started to speak. The seizures self-aborted and no acute interventions were put in place. Tablet Carbamazepine 200 mg × 12 h was added to the levetiracetam. Carbamazepine dose was increased to 400 mg × 12 h on the 7th day of admission after which seizures were controlled.

2.2 | Outcome and follow-up

He was started on physiotherapy with improved motor response and was discharged after 12 days of admission. The physiotherapy was continued on outpatient. She was seizure free for about 8 months but then unfortunately lost to follow-up.

3 | DISCUSSION

The human brain is a very complex structure and arguably the most complex of all biologic systems. Averagely, the adult human brain has a volume of about 1350 cm^3, a total surface area of 1820 cm^2, an average cortical thickness of 2.7 mm and contains approximately 100 billion neurons, of which 20 billion are located in the cerebral cortex.[8,9] The development of the human brain begins in the 5th week of embryonic life and continues until adulthood at around 25 years through a series of fine phases which are highly regulated. Gyri and sulci formation begin as early as the 12–16th week of gestation and continue through the third trimester. 70% of the adult brain weight is achieved at 18 months, 80% at 3 years, 90% at 5–8 years and approximately 95% by the 10th year.[10] This presupposes that three fourth of the adult brain size is achieved by the end of the third year of life. Therefore, when a neurologic insult is sustained congenitally or acquired most especially during the first 3 years of life, compensatory skull changes occur due to reduced brain growth on the side of the insult. The cerebral hemiatrophy is associated with ipsilateral calvarial thickening, elevated orbital roof, hyper pneumatization of the ipsilateral paranasal sinuses, and enlargement of mastoid cells which are characteristic features seen in Dyke-Davidoff-Masson-Syndrome.[3] The congenital pathogenesis of DDMS include fetal vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies, coarctation of the mid-aortic arch, mesencephalon hypoplasia, wullerian degeneration or infection.[3] Acquired causes include perinatal hypoxia, intracranial hemorrhage, infections, cranial trauma, and cerebrovascular lesions.

The characteristics of DDMS may be variable and largely dependent on the time of onset and the extent of brain injury. If vascular ischemia occurs during embryogenesis or fetal life, there would be marked cerebral atrophy and no prominent sulci. However, there will be prominent sulci if the injury occurs after birth or after sulcation is completed in the third trimester.[11] More prominent features such as ipsilateral dilatation of lateral ventricles, midline shift, thickening of the calvarium, hyperpneumatization of the sinuses and hypoplasia of the cranial fossa are generally seen if the insult occurs before the age of 3 years.[3,11] The features are less prominent if the insult occurs after 3 years of age. Other brain parenchymal features that may also be seen include hypoplasia of mesencephalon, lentiform nucleus and thalamus, diffuse

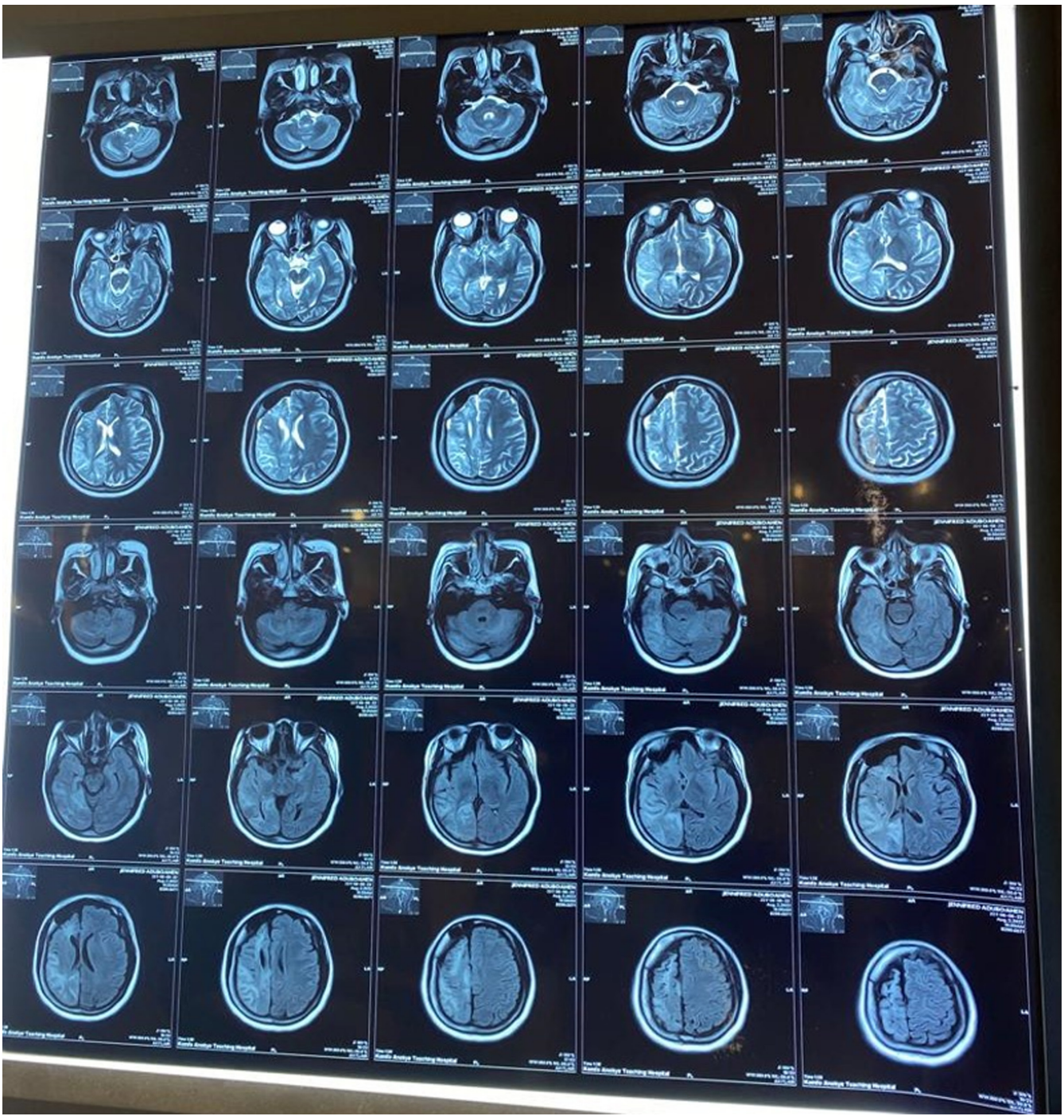

FIGURE 1 The MRI of the brain showing a reduction in the volume of the right cerebral hemisphere. There is right calvarial thickening and atrophy of the right basal ganglia.

cerebellar atrophy, and ipsilateral atrophy of the cerebral peduncle.[11] Our patient had a significant right cerebral hemiatrophy, atrophy of right basal ganglia with a compensatory ipsilateral calvarial thickening and hyperpneumatization of the right frontal sinus and mastoid air cells. However, she had normal appearing lateral ventricles, brainstem, and cerebellum. The radiologic features of this patient most likely presuppose that her brain injury occurred before the age of 3 years.

Seizures are the most common clinical manifestation of DDMS occurring in about 60%–85% of the cases.[11,12] The seizures usually start early in life and may present as either generalized seizures or focal onset seizures or focal seizures with secondary generalization. These seizures are recurrent and unprovoked and most of these patients may be managed for epilepsy. It is therefore important to consider DDMS as a differential diagnosis in any patient who presents with early onset epilepsy. Other important

clinical manifestations of DDMS include, contralateral hemiplegia/hemiparesis of upper motor neuron lesion, speech and language disorders, mental retardation, facial asymmetry, and psychiatric manifestations.[3,11,12] Our patient, a 22-year-old, was diagnosed with epilepsy from childhood and put on anticonvulsants. However, no neuroimaging was done to ascertain the possible cause. This was probably due to financial constraints or lack of MRI or Computed Tomography (CT) imaging facilities at the centre where she was diagnosed. It is important to note that in a resource-poor facility, certain imaging modalities such as a simple skull X-ray which is less expensive and readily available can be very important in diagnosing DDMS. Moreover, the first description of DDMS by Dyke, Davidoff and Masson was done using a plain skull X-ray in 1933. Early diagnosis ensures appropriate and optimal treatment of patients.

Differential diagnosis of DDMS mainly include the disorders that cause cerebral hemiatrophy or midline structural shift such as Sturge–Weber syndrome, Rasmussen encephalitis, Silver-Russel syndrome, and basal ganglia germinoma. However, Sturge–Weber syndrome typically has port-wine facial nevus, associated with leptomeningeal angioma and intracranial tram track calcification, whereas in Rasmussen encephalitis, an immune-mediated brain disorder characterized by unilateral hemispheric atrophy, intractable focal epilepsy, and progressive neurological dysfunction, there are no calvarial changes.[13] Other differentials diagnosis is hemiconvulsion-hemiplegia-epilepsy (HHE) syndrome. Differentiating this syndrome involves a time frame of 2 months to 2 years between the onset of encephalopathy and seizures, as well as unilateral edema and swelling in the same cerebral hemisphere during seizures.[14] The management of DDMS is usually symptomatic and involves a multidisciplinary approach including the neurologist, physiotherapist, speech, and occupational therapist and psychiatrist. Anticonvulsants are essential in managing recurrent seizures, a speech and occupation therapist helps with language and learning disabilities and a psychiatrist to manage neuropsychiatry disorders such as affective disorders and behavioral disorders.[12] Prognosis is better for patients who develop hemiparesis after the age of 2 years in the absence of prolonged or recurrent seizures.[11] Hemispherectomy is the treatment of choice for patients with intractable disabling seizures and hemiplegia with a success rate of 85% in selected cases.[15]

AUTHOR CONTRIBUTIONS

Patricia Afrim: Conceptualization; investigation; supervision; writing – review and editing. **Emmanuel Ofori:** Conceptualization; investigation; resources; writing – original draft; writing – review and editing. **Nana Akosua Owusu-Danso:** Conceptualization; resources; writing – original draft; writing – review and editing. **Denise Akua Owusua:** Investigation; validation; writing – original draft; writing – review and editing. **Rexford Adu Gyamfi:** Investigation; writing – original draft; writing – review and editing. **Solomon Gyabaah:** Conceptualization; investigation; resources; writing – original draft; writing – review and editing. **Fred Stephen Sarfo:** Conceptualization; investigation; resources; writing – original draft; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

Data is available upon reasonable request to the corresponding Author.

CONSENT

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

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