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Case Report

Dyke-Davidoff-Masson Syndrome Presenting with Recurrent Seizures and Hemifacial Asymmetry: A Case Report from Abeokuta, Nigeria

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Abstract

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare clinical syndrome characterized by unilateral cerebral hemiatrophy, recurrent seizures which could be congenital or acquired. There are some reported cases in Nigeria. We report the first documented case in Abeokuta, a 25-year-old lady who presented to the Neurology Clinic with a major complaint of poorly controlled recurrent seizures and facial/upper limb asymmetry. Early diagnostic confirmation was possible with prompt brain magnetic resonance imaging which showed the typical radiological features of the syndrome. The patient's antiseizure medication was reviewed and commenced on multiple antiseizure drugs with subsequent good seizure control. Also, commenced on physiotherapy and occupational therapy for optimal neurorehabilitation. This case report aims to highlight the clinical features, diagnostic challenges, and multidisciplinary management of a case of Dyke-Davidoff-Masson Syndrome presenting with refractory seizures and progressive facial asymmetry. It underscores the need for heightened awareness of this rare syndrome among non-neurologists in Nigeria and the critical role of advanced neuroimaging as well as necessity of holistic care of this rare condition.

Keywords: Dyke-Davidoff-Masson Syndrome (DDMS); Nigeria

Background

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological disorder first described in 1933 by Dyke, Davidoff, and Masson, characterized by cerebral hemiatrophy with compensatory changes in the skull and sinuses, leading to a constellation of clinical features including contralateral hemiparesis, seizures, facial asymmetry, and cognitive impairments [1,2]. It typically results from an early cerebral insult (e.g., ischemia, infection, trauma) occurring in utero or during early childhood, leading to compensatory changes in the skull and brain [3,4]. These causes of injury lead to the reduction of neurotrophic factors necessary for proper brain development [1,4]. Clinically, patients often present with a spectrum of neurological deficits, including recurrent seizures, hemiparesis or hemiplegia on the contralateral side, intellectual disability, and speech disorders [5]. Facial asymmetry, though a less frequently emphasized symptom in some literature reviews, can be a subtle yet crucial clinical sign [6]. The syndrome's classical radiological hallmarks include unilateral cerebral volume loss, ipsilateral ven-

tricular enlargement (ex-vacuo hydrocephalus), prominence of ipsilateral cortical sulci, and compensatory thickening of the calvarium with elevation of the sphenoid ridge [6,7]. The diagnosis of DDMS is primarily radiological, with Magnetic Resonance Imaging (MRI) of the brain being the gold standard for its superior ability to delineate brain parenchymal changes and associated structural abnormalities[5]. Although DDMS is predominantly diagnosed in childhood, adult presentations are increasingly reported, often with delayed recognition due to variable clinical courses and limited awareness, especially in resource-limited settings [8].

However, in many low- and middle-income countries, including Nigeria, access to advanced neuroimaging like MRI remains a significant challenge, often delaying or precluding definitive diagnosis [9]. While its classic neuroimaging features are well-documented globally, the heterogeneous clinical presentation and challenges in accessing advanced diagnostics contribute to its potential underdiagnosis, particularly in low-resource settings. In Nigeria and the broader sub-Saharan African region, only a few cases of DDMS have been documented in the literature, underscoring its rarity and the likelihood of underdiagnosis [10]. This case report details the clinical and radiological features of a 25-year-old woman presenting with recurrent seizures and unilateral motor deficits at the Federal Medical Centre, Abeokuta. Our aim is to contribute to the growing body of literature on DDMS, raise clinical awareness, and highlight the importance of neuroimaging in establishing diagnosis in adult patients presenting with recurrent seizures and facial/limb asymmetry.

Case Presentation

We present the case of a 25yr old lady who presented to the neurology outpatient clinic on account of recurrent seizures, facial and limb asymmetry. Her past medical history was notable for prolonged febrile convulsion in early childhood (around 2 years of age), which resolved spontaneously and was not associated with any apparent neurological sequelae at that time. She remained healthy until the age of 19 years when she experienced the onset of recurrent focal seizures, with secondary generalization. Initially, these seizures were characterized by brief episodes of staring spells, lip-smacking, and unresponsiveness, lasting less than a minute. They occurred infrequently (approximately once every 2-3 months) and initially responded partially to prescribed antiepileptic drugs (AEDs), though the specific agents used at that time could not be precisely recalled by the patient or her family.

Over the past two years, her seizure frequency had significantly escalated to multiple episodes per week, sometimes occurring daily, and becoming less responsive to her current AED regimen. These recent seizures were described as focal to bilateral tonic-clonic type, with initial stiffness and jerking of the right upper limb, followed by generalization, associated with tongue biting and postictal confusion. This escalation in seizure activity prompted her presentation to our tertiary center.

Concurrently with the worsening seizures, she noticed a gradual and progressive asymmetry of her face, which had become more noticeable over the past few years. There was no history of head trauma, central nervous system infection, or stroke. Her developmental milestones were reportedly as essentially normal, though her academic performance had been consistently about average and presently in the higher institution.

On neurological examination, the patient was conscious, alert, and well oriented. Higher mental functions revealed normal findings. Cranial nerve examination revealed a mild right-sided hemifacial atrophy, characterized by flattening of the nasolabial fold on the left side. There was no apparent deviation of the tongue or uvula. Her voice quality was assessed and noted to be normal; the earlier reported "hoarseness" was not clinically prominent at the time of presentation. Motor examination revealed mild weakness (Medical Research Council (MRC) grade 4/5) with reduced bulk in the right upper limb, with no significant weakness in the lower limbs. Deep tendon reflexes were brisk on the right side compared to the left, with an extensor plantar response on the right. Sensory examination was unremarkable.

Investigations done include Electroencephalography (EEG) performed on presentation showed generalized background slowing with frequent focal epileptiform discharges localized to the left cerebral hemisphere, consistent with a left-sided epileptogenic focus and diffuse cortical dysfunction. Brain Magnetic Resonance Imaging (MRI) was performed, which was crucial for establishing the diagnosis (Figure 1A, 1B). The MRI sequences (T1-weighted, T2-weighted, and FLAIR) revealed features of left cerebral hemiatrophy. Specifically, there was significant volume loss of the right cerebral hemisphere, including the frontal, parietal and temporal lobes. This was accompanied by marked ipsilateral ex-vacuo dilatation of

the right lateral ventricle and prominence of the cortical sulci and sylvian fissure on the right side. Compensatory changes included diffuse thickening of the right calvarium and elevation of the right sphenoid ridge. No mass lesions, acute infarcts, or overt vascular malformations were identified. These findings were unequivocally consistent with a diagnosis of Dyke-Davidoff-Masson Syndrome. Following the definitive diagnosis, the patient's antiepileptic drug regimen was optimized with polytherapy regimen comprising Carbamazepine and Lamotrigine, titrated gradually to achieve seizure control. She also commenced occupational therapy and physiotherapy sessions tailored to improve her right upper limb function and overall motor coordination by improving activities of daily and strengthening exercises/gait training respectively.

Over a 6-month follow-up period, the patient showed significant improvement. Her seizure frequency reduced dramatically from multiple times per week to approximately one seizure every 1-2 months. Her quality of life improved notably, with enhanced independence in daily activities. She reported feeling much better and was more engaged socially. She continues on her optimized dual AED regimen and ongoing rehabilitation.

Discussion

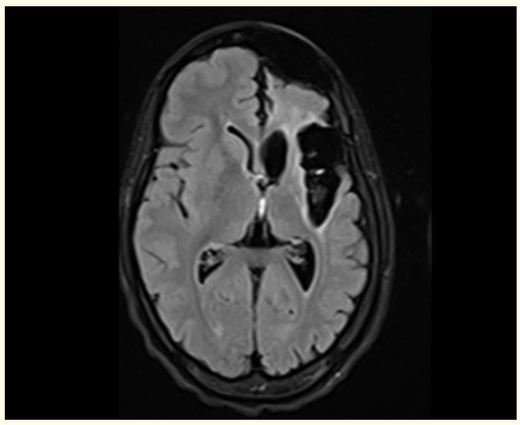

Figure 1A

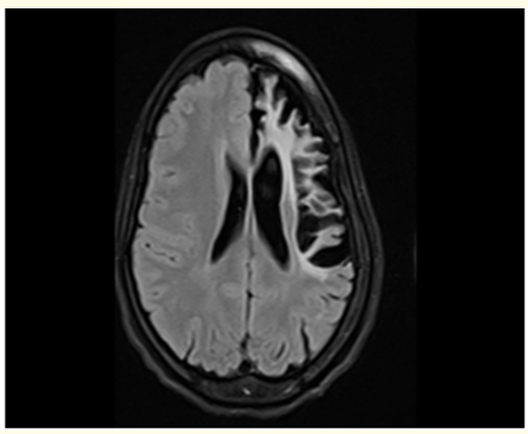

Figure 1B

Dyke-Davidoff-Mason syndrome, is an uncommon neurological disorder characterized by cerebral hemiatrophy accompanied by compensatory hypertrophy of the skull bones and hyperpneumatization of the frontal sinuses [1]. It was first described in a nine-case series following which the syndrome was named[1]. The epidemiology of this rare neurological syndrome is not well documented but several studies have shown that prevalence is higher among the pediatric age group. There is no precise global incidence or prevalence figure because it's largely reported through individual case reports and small case series, with less than 100 overall cases reported few years ago since the syndrome was discovered [11]. Santos., *et al*. reported in a recent literature review that the mean age of diagnosis is 22.9 ± 3.2 years, but the age of diagnosis is variable with the majority of cases only being diagnosed at adulthood though the reasons for delay in diagnosis were not specified [14]. The international league against epilepsy classified DDMS into the category of epilepsy with brain structural abnormalities [12]. Varying etiologies have been reported, including ischemia, trauma, infection, and hemorrhage, to name a few, mainly occurring in utero or after birth [1,4]. The pathogenesis of DDMS involves an insult to the developing brain either in utero or during early childhood, resulting in cerebral hemiatrophy and secondary compensatory changes such as calvarial thickening and sinus enlargement [18,19]. Two forms of DDMS have been described: congenital, where the insult occurs prenatally or perinatally, and acquired, which results from postnatal brain injury due to trauma, infection, ischemia, or prolonged seizures [8,15]. These causes of injury lead to the reduction of neurotrophic factors necessary for proper brain development. The history of prolonged febrile convulsions has been reported as a causative factor in DDMS[15]. This reflect an underlying neurological vulnerability or even be an early manifestation of the cerebral insult that subsequently led to hemiatrophy. It is postulated that ischemia leads to reduction of neurotrophins important for neural development and plasticity [16]. The clinical manifestation includes recurrent seizures, facial asymmetry, contralateral hemiplegia, cognitive changes or learning disability, and speech and language disorders [3,7]. Seizures are the most common presenting symptom in DDMS, often starting in childhood or adolescence and tending to be refractory to treatment due to the underlying structural brain damage [1,13]. The presence of facial asymmetry as a prominent feature is a key learning point from this case. While hemiparesis is frequently reported, facial involvement is sometimes underemphasized in DDMS literature, though it is a direct consequence of the unilateral cerebral pathology affecting facial nerve pathways [1,2,17]. Furthermore, the reported change in voice, described as becoming hoarser, offers another intriguing, albeit less commonly

emphasized, manifestation. While voice changes are not typical hallmark features, isolated reports do exist, potentially linked to subtle involvement of brainstem pathways or supratentorial control of laryngeal function, emphasizing the broad and varied neurological impact of DDMS [6].

Our patient, a 25-year-old woman presenting with recurrent seizures, unilateral motor deficits and some hoarseness of voice following pronged febrile seizure in childhood, exemplifies the classical clinical and radiological features of DDMS. Facial and Limb asymmetry further underscore the chronicity and severity of cerebral damage. Importantly, the diagnosis in adults can be delayed due to variable clinical courses and limited awareness, as seen in Nigeria where few cases have been documented highlighting the challenges in diagnosis, evaluation and management in adult patients [8].

Neuroimaging remains the cornerstone of diagnosis. Computed tomography (CT) and magnetic resonance imaging (MRI) typically reveal unilateral cerebral atrophy, ventricular enlargement, calvarial thickening, and hyperpneumatization of the frontal sinuses ipsilateral to the affected hemisphere [19]. The pivotal role of the brain magnetic resonance imaging (MRI) in this case cannot be overstated. Despite its limited availability in many parts of Nigeria, the successful acquisition of an MRI at Federal Medical Centre (FMC), Abeokuta was instrumental in confirming the diagnosis. While Computed Tomography (CT) can suggest DDMS by showing gross hemiatrophy and skull changes, MRI offers superior anatomical detail, better visualization of parenchymal changes (e.g., gliosis, areas of encephalomalacia), and can rule out other underlying pathologies more definitively [20]. This case highlights the critical disparity in diagnostic capabilities in different healthcare settings and underscores the urgent need for expanded access to advanced neuroimaging in developing countries to ensure timely and accurate diagnosis of complex neurological disorders [9]. Without MRI, this patient's diagnosis might have remained elusive or significantly delayed, impacting her management and long-term prognosis.

The long-term management of DDMS is largely symptomatic, focusing on seizure control, physical rehabilitation, and supportive care aimed at improving functional outcomes and quality of life [5,9]. This long-term management requires a multidisciplinary approach, as demonstrated by the improved morbidity and quality of life observed in our patient. Beyond optimizing antiepileptic drug therapy, which is crucial for seizure control, comprehensive care involves rehabilitation services such as physiotherapy for managing hemiparesis and occupational therapy to improve functional independence in daily activities [21]. Neuropsychological assessment and support are often vital to address cognitive deficits and learning difficulties, which are common in DDMS patients [22]. Psychological counseling can also benefit patients and families coping with the chronic nature of the illness. Although not explicitly detailed in this report for brevity, speech therapy may be required for associated dysarthria or dysphagia, and social support services can help integrate patients into educational and community settings. The improvement noted in our patient, particularly the reduction in seizure burden and enhanced daily functioning, highlights the effectiveness of even partial implementation of such a comprehensive care model in a challenging environment.

In summary, this case adds to the sparse but growing literature on DDMS from sub-Saharan Africa, contributing to a better understanding of its presentation and diagnostic pathways in this region. It also serves as a powerful advocacy for investment in advanced medical infrastructure and specialized neurological care in developing nations like Nigeria

Conclusion

Dyke-Davidoff-Masson Syndrome, though uncommon, should be strongly considered in the differential diagnosis of children and young adults presenting with refractory seizures and progressive neurological deficits, especially when accompanied by subtle but progressive facial asymmetry. This case from FMC, Abeokuta underscores the critical role of accessibility to advanced neuroimaging, specifically Brain MRI, in confirming this complex diagnosis. Furthermore, the case emphasizes the undeniable necessity of a comprehensive, multidisciplinary care approach, encompassing pharmacological management and rehabilitation, to significantly improve patient outcomes and quality of life, even in settings with evolving healthcare infrastructure. This case reinforces the global need for equitable access to diagnostic tools and integrated care for rare neurological conditions.

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Consent for Publication

All authors consented.

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Conflicts of Interest

The authors declare no conflicts of interest.

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