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# A Shrunken Hemisphere, A Life time of Seizures: Dyke-Davidoff- Masson Syndrome

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**Abstract**

Dyke -Davidoff- Masson Syndrome (DDMS) is rare neurological disorder, first described in 1933, commonly affecting the children but is rarely reported in adults as well. It mostly presents with seizure, contralateral hemiparesis followed by mental retardation and facial asymmetry due to palsy of the facial nerve (CN VII). The classical radiological findings are cerebral hemiatrophy, calvarial thickening, and hyperpneumatization of the frontal sinuses. We here in report a case of 25 years old male who presented with repeated generalized tonic clonic seizures for ten years He was on regular anti convulsive therapy and failed to control seizure. General examination revealed no significant abnormality with delayed mile stones of development. Neurological examination revealed right sided spastic hemiparesis, brisk tendon reflexes and extensor planter on left side. Ultimately, he was diagnosed as DDM. This case report aims to draw the attention of health care professionals to keep DDM as a differential in a patient with drug resistant intractable epilepsy.

**Keywords:** DDM, Hemi Atrophy, Seizure, Intractable Epilepsy.

## Introduction

In the year 1993 three researchers Dyke, Davidoff, and Masson came across peculiar radiographic images of cerebral hemiatrophy, elevation of petrous ridge and compensatory hypertrophy of calvarium and frontal sinuses (also ethmoidal and mastoid air cells) in nine patients who clinically presented with seizures, facial hemiparesis, and learning/developmental disabilities - thus forming the typical presentation of this syndrome and named it as Dyke-Davidoff-Masson Syndrome (DDMS) Prenatal causes include congenital anomalies, cerebral infarction, vascular malformations and infections [1]. Insult to premature brain causing loss of neurons and impairment of growth of brain is the main pathology. Postnatal hemiatrophy can develop secondary to cerebral trauma, tumors, infections and febrile seizures. It has been reported that DDMS occur in intrauterine life when the maturation of calvarium has not been completed or due to brain damage (usually traumatic) occurring in first three years of life [2-5]. It occurs in both genders, with a predominance in males (73.5%) [6]. A diagnosis is usually made during late childhood, adolescence or adulthood [7]. The major concern is the occurrence of such convulsive episodes for which pharmacotherapy alone is insufficient in most of the cases, and where surgical management is eventually advised [8]. We are hereby describing the clinical and radiological features of this syndrome in a young adult presenting to us with refractory seizures.

## Case Report

A 25-year-old male patient presented to our emergency department with sudden onset of involuntary movements of both limbs, upward gazing of eyes, frothing of the mouth, involuntary micturition, and tongue bite. The patient's mother gave a history of 6-8 seizures since the previous night before arrival to the hospital with episodes of loss of consciousness for more than 30 minutes and post-ictal confusion for a period of 45 minutes. He had history of right-sided hemiparesis, and mental retardation since three years of his age. She was born out of a nonconsanguineous marriage. Birth history was indicative of a full-term normal delivery without any antenatal complications. But he had history of post-natal asphyxia for which he was admitted in neonatal intensive care unit and was ventilated for 6 days. He had delayed developmental milestones. From the age of five, he developed generalized seizure and started anti-epileptic drugs. Seizures were usually preceded by neck pain, nausea, and involuntary movements of the right hand, diagnosed as idiopathic generalized epilepsy by the local physician, and kept as the diagnosis without further investigations or referral to a higher center. However, he continued to have seizures which exacerbated due to poor compliance and other precipitating actors like sleep deprivation, stress, etc.

Besides, the parents also noted learning difficulties, slurred speech, facial deviation, and progressive right hemiparesis, for which he was treated conservatively by a local practitioner. His seizures did not respond to several anti-epileptic medications in different combinations. Notwithstanding, the patient's hemiparesis had worsened gradually with increased stiffness in the right extremities and poor dexterity of the right hand. There were no similar complaints in the immediate family.

The clinical examination of the central nervous system was right sided spastic hemiparesis with muscle power 3/5 in both right upper and lower limbs with right sided extensor plantar response with brisk right sided tendon reflexes. He scored poorly on the Mini-Mental Status Examination (14/30).

Blood samples were collected and sent for blood sugar levels, complete metabolic panel, and complete hemogram, in order to rule out the common causes of seizures. magnetic resonance imaging (MRI) of the brain was subsequently done, which revealed right cerebral atrophy, with gliotic and encephalomalacic changes together with compensatory thickening of the cranial vault (Figure 1). An electroencephalogram (EEG) showed abnormal EEG changes with generalized seizure discharges and diffuse background slowing (Figure 2).

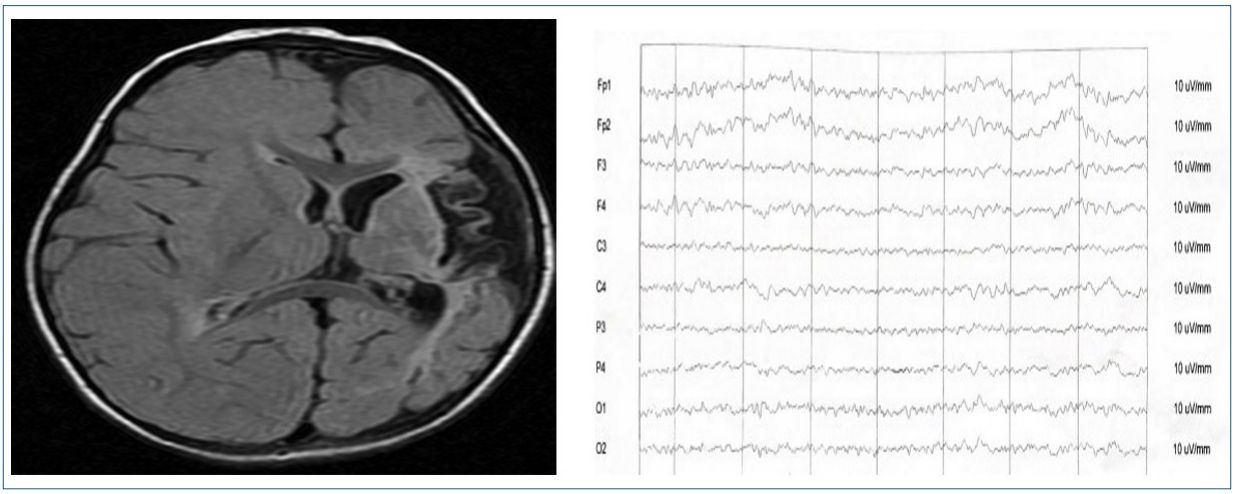


**Figure 1 and Figure 2:** MRI of brain showing left cerebral atrophy, with gliotic and encephalomalacic changes together with compensatory thickening of the cranial vault and EEG showing generalized seizure discharges and diffuse background slowing.

We accordingly kept a diagnosis of DDMS, managed him conservatively with combination of levetiracetam and lamotrigine, and referred him to a comprehensive center for further management upon the patient's attendees' request. The patient's attendees refused surgical intervention due to financial constraints, and are continuing the anticonvulsants. Informed consent for publication was obtained from the patient's representative.

**Discussion**

DDMS, which is a rare but important condition commonly associated with refractory seizures, was first documented by Dyke, Davidoff, and Masson in 1933 when they noted radiographic images in a series of 9 patients with similar presentations [1]. They described plain skull radiographic changes in 9 patients who presented with seizures, facial asymmetry, hemiparesis, and mental retardation [1]. Subtotal or diffuse cerebral hemiatrophy is a classical imaging finding. However, unilateral focal atrophy may occasionally be noted in the cerebral peduncles and the thalamic, pontine, crossed cerebellar, and parahippocampal regions. Brain imaging may additionally reveal prominent cortical sulci, dilated lateral ventricles and cisternal space, calvarial thickening, ipsilateral osseous hypertrophy with hyperpneumatization of the sinuses (mainly frontal and mastoid air cells), and an elevated temporal bone [9].

The etiology can be congenital or acquired [10]. Congenital type presents in early infancy secondary to previous intrauterine brain insults like vascular occlusion or anomaly of the middle cerebral artery. The acquired type occurs later in the childhood secondary to variable causes affecting brain perfusion like infection, prolonged febrile seizure, trauma, hemorrhage and ischemia [11].

The clinical features of this syndrome are hemiparesis on the same side as hemiatrophy, with an upper motor neuron type palsy of the facial nerve (CN VIII), focal or generalized convulsions, and poor intellect with a delay in the achievement of milestones either occurring alone or in combination based on the side of hemiatrophy [12]. However, generalized tonic clonic seizure is the most common presenting feature [10,13]. There is no sex predilection, and any side of the brain can be involved, although involvement of the left side and male gender have been shown to be more common in one study [9].

To understand the genesis of the brain pathology in this syndrome one needs to look in-to the development of the brain minutely. The formation of the brain sulci starts around the fourth month of gestation and gets completed by the end of the eighth month. Overall, the maximum growth of a child's head occurs in the early years due to outward pressure of the enlarging human brain on the bony skull table which reaches half of its adult size at the end of infancy and three fourths of the adult size by the end of 3 years [14]. Therefore, only when brain damage is sustained before 3 years of age, other structures overlying the brain grow inward, thus resulting in an increased width of the diploic spaces, enlarged sinuses, and an elevated orbital roof, which are characteristic of this disorder [15]. The plausible mechanism of cerebral atrophy and the related progressive neurodeficit is hypothesized to be due to several ischemic episodes resulting from different causes, which reduce the production of brain-derived neurotrophic factors, which in turn leads to cerebral atrophy [16].

Refractory epilepsy has many etiologies. These are commonly associated with failure of adherence to antiepileptic drugs, and include seizures that are non-epileptic, misdiagnosed, or inappropriate use of medications such as inadequate dosing,

drug-to-drug interactions, and lifestyle choices such as alcohol & drug abuse, stress, and sleep deprivation [17]. Identification of the causative etiology is essential in planning its management, since refractory seizures are associated with high rates of morbidity and mortality. Out of the variety of tests available to investigate epilepsy, neuroimaging is the main tool used in its investigation. We came across this rare case of Dyke-Davidoff-Masson syndrome presenting as refractory seizures alone without the other typical features mentioned above.

The differential diagnosis of this presentation seen in our case includes Sturge-Weber syndrome and Rasmussen encephalitis. Also, certain syndromes like Fishman syndrome, Silver-Russell syndrome, and linear nevus syndrome have to be kept in the picture as rare but possible causes. These syndromes are recognized through neuroimaging and clinical correlation [18,19].

A precise diagnosis and implementation of suitable management are made possible by imaging via CT and MRI, which is of great importance. The plain skull radiograph illustrates thickening of the calvarium and dilatation of the ipsilateral frontal and ethmoid sinuses. CT and MRI show unilateral atrophy of the cerebral hemisphere with an ipsilateral shift of the ventricle, widening of sulcal spaces on the involved side. It is associated with compensatory calvarium thickening, hyperpneumatization of the paranasal sinuses and mastoid cells, and elevation of the petrous ridge [20]. In congenital hemiatrophy, when the insult occurs in utero, there is a shift of midline structures toward the disease side, but there is the absence of sulcal prominence replacing the gliotic tissue. This is the salient feature differentiating congenital from acquired form [21].

With the clinical features of cerebral hemiatrophy along with supportive radiological evidence of cerebral hemiatrophy, osseous hypertrophy of the skull, and compensatory hyper-pneumatization of the sinuses, DDMS has to be considered as the cause [16]. Even though our patient had just refractory seizures and learning difficulties as the clinical features, radiographic assistance is the one that aided in the prompt diagnosis of this syndrome.

Refractory seizures remain the usual concern in DDMS patients [22]. There is no definitive guideline for the management of seizure in DDM [23]. The treatment is symptomatic and must oriented to treat convulsion, hemiplegia, hemiparesis, and learning difficulties. Long-term management also includes adjunctive usage of physiotherapy, occupational and speech therapy. At present, management of epilepsy is still limited to monotherapy or adjunct usage of antiseizure drugs as the first-line management. Prognosis is better if hemiparesis occurs after the age of 2 years and without prolonged or repetitive seizures. Children with intractable seizures are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases [24]. Hence, early diagnosis makes early decision-making and intervention. In our setting, the perinatal hypoxic injury is one of the causative factors of DDMS, hence, the role of proper obstetric care in preventing such conditions.

## Conclusion

DDMS is a rare preventable cause of refractory epilepsy. DDMS usually presents in early childhood or adolescents requiring lifelong pharmacotherapy with anticonvulsants. CT and MRI are powerful imaging modalities to diagnose the pertinent imaging features associated with this syndrome. Accordingly, hemispherectomy is the treatment of choice with a success rate of 85% in selected cases. However, if the presentation is late as in our case and if seizures are under control, the patient can be kept on antiepileptic medications in spite of surgery, along with supportive therapy including physiotherapy, speech therapy, and occupational therapy.

**Conflict of Interest:** None declared

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