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Case Report

DYKE-DAVIDOFF MASSON SYNDROME: A CASE REPORT

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ABSTRACT
Dyke – Davidoff – Masson Syndrome (DDMS), also called as cerebral hemiatrophy, is a rare clinical condition characterized by seizures, facial asymmetry, contralateral spastic hemiplegia or hemiparesis, with learning difficulties. It is commonly diagnosed in presence of associated radiologic findings which include cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. Here we report a case of DDMS in a 20 year old female patient who presented with seizures and mental retardation and hemiparesis. Computerized Tomography of the brain showed hemi atrophy.

Keywords: *Dyke – Davidoff – Masson Syndrome (DDMS), Hemiatrophy, Computerized Tomography*

INTRODUCTION
Dyke-Davidoff-Masson Syndrome (DDMS) is a syndrome associated with refractory epilepsy and was first described by Dyke *et al.,* (1933). They described the plain skull radiographic and pneumato-encephalographic changes in a series of nine patients characterized clinically by hemiparesis, seizures, facial asymmetry, mental retardation, learning disability and speech and language disorders (Dyke *et al.,* 1933). Sensory loss and psychiatric manifestations like schizophrenia had been reported rarely (Ono *et al.,* 2003; Amann *et al.,* 2009). The typical radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses. The syndrome had been documented mainly in adolescents and adults (Sharma *et al.,* 2006; Singh *et al.,* 2010); however, it can also be seen in children (Narain *et al.,* 2008). Since then, there were few case reports in the literature we are hereby describing the case of a 20 year old female.

CASES
Ms X a 20 year old female who presented in the outpatient section of the psychiatry department with seizure disorder and subnormal intelligence, the patient had no other symptoms as seen in other cases. There was no history of significant antenatal or perinatal complication or a family history of epilepsy. The developmental milestones were achieved at normal age however higher mental function revealed mild level of mental sub normality. The patient had seizure since past one and a half year with behavioral changes. The patient had no other symptoms apart from seizure, neurological examination revealed no abnormality. The diagnosis was established when a CT scan was done owing to seizures. The CECT head revealed intra axial, ill-defined, hypodense areas in right temporo-parieto-occipital region (Figure I) with ex-vacuuo dilation of ipsilateral lateral ventricle with ipsilateral falcine displacement (Figure II). Unilateral calvarial thickening with ipsilateral expansion of paranasal sinus and mastoid air cells. Relative elevation of petrous ridge was noted (Figure III). The patient is maintaining on antiepileptics.

DISCUSSION
DDMS is a rare condition characterized by varying degrees of facial asymmetry, seizures, contralateral hemiparesis, mental retardation and learning disabilities with behavioral abnormalities (Dyke *et al.,* 1933). Cerebral hemiatrophy can be of two types, infantile (congenital) and acquired. The infantile variety results from various etiologies such as infections, neonatal or gestational vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies and coarctation of the midaortic arch (Sener and Jinkins, 1992; Stred *et al.,* 1986). The patient becomes symptomatic in the perinatal period or infancy. The main causes of acquired type are trauma, tumor, infection, ischemia,

hemorrhage, and prolonged febrile seizure. Age of presentation depends on time of insult and characteristic changes may be seen only in adolescence or adult. The exact mechanism of cerebral atrophy is unclear in either type. The index patient also fits with the present descriptions. It is hypothesized that ischemic episodes from a variety of different causes reduce the production of brain derived neurotrophic factors, which in turn lead to cerebral atrophy (Lee *et al.*, 2006), in the index patient there was history of per natal insult. 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 (Unal *et al.*, 2004). The condition needs to be differentiated from Basal ganglia germinoma, Sturge Weber syndrome, Linear nevus syndrome, Fishman syndrome, Silver-Russel syndrome, Rasmussen encephalitis (Narain *et al.*, 2008). For DDMS cases presenting in early childhood, refractory seizures remain the usual concern. Accordingly, hemi spherectomy 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. Supportive therapy including physiotherapy, speech therapy, and occupational therapy are needed. Further longitudinal studies are required to ascertain the natural course of this syndrome especially in an adult population, which would help in planning strategies regarding the time and nature of interventions and management accordingly. The case is discussed to draw attention on a syndrome which can be managed by a holistic approach.

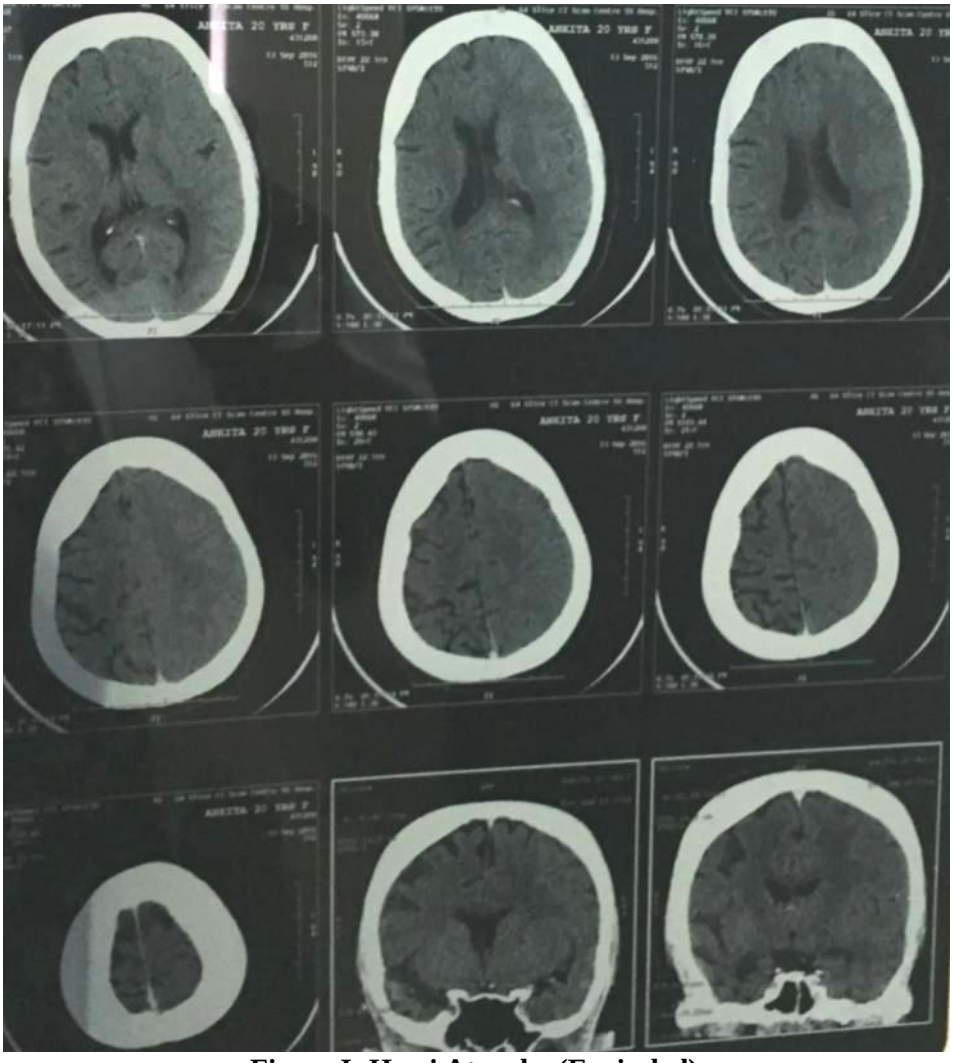

Figure I: Hemi Atrophy (Encircled)

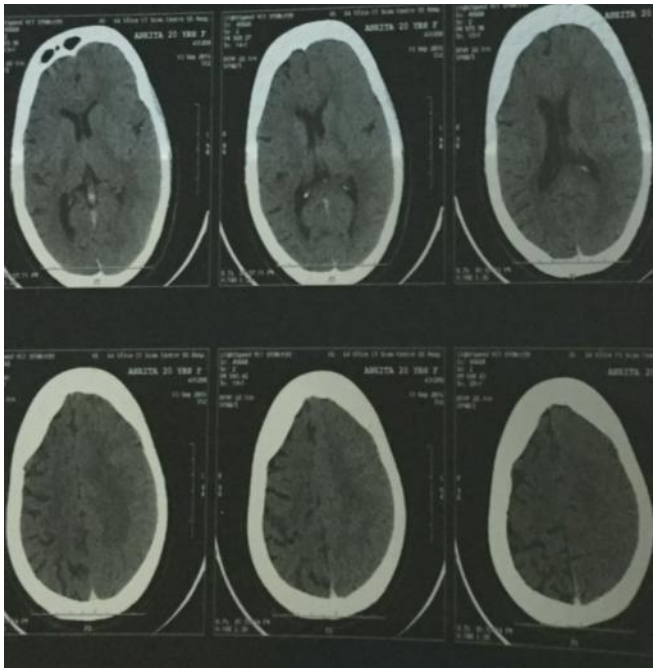

Figure II: Atrophic Side having Dilated Ventricles

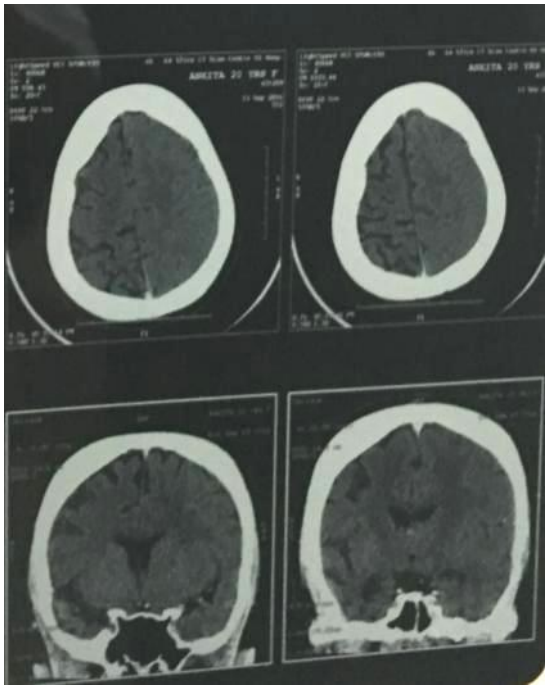

Figure III: Petrous Ridge

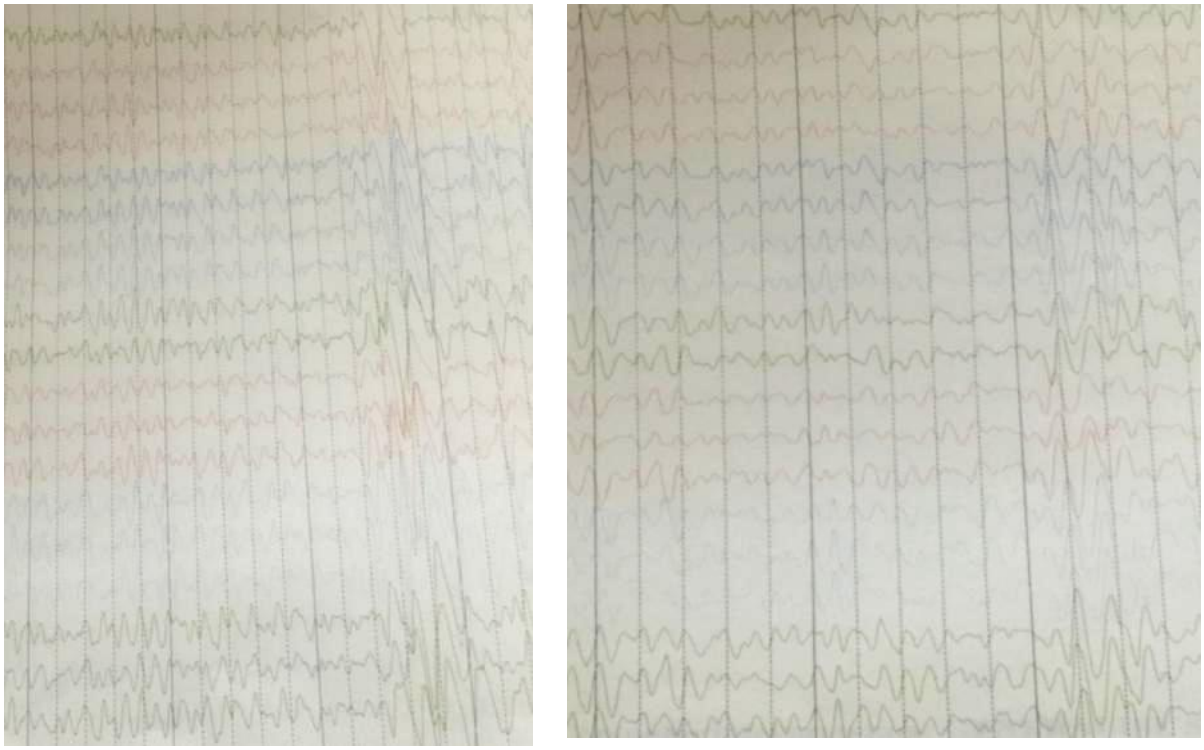

Figure IV: Seizure Discharges

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