# A RARE CAUSE OF RESISTANT SEIZURES: DYKE DAVIDOFF MASSON SYNDROME

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**ABSTRACT**: Dyke – Davidoff – Masson Syndrome (DDMS), is a rare clinical condition characterized by clinical triad of seizures, contralateral spastic hemiplegia or hemiparesis, with or without mental retardation. Diagnosis requires presence of cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses on brain imaging. Here we report a case of DDMS in a 16 year old girl who presented with seizures and hemiparesis.MRI of her brain showed hemiatrophy involving the left cerebral hemisphere with enlargement of ipsilateral sinuses and ventricles.
**KEYWORD:** Cerebral hemiatrophy, Seizures, Hemiparesis

**INTRODUCTION**: Dyke Davidoff Masson Syndrome was first described by C.G. Dyke, L.M. Davidoff and C.B. Masson in 1933 where they described a series of nine patients with plain skull radiographic changes [1]. The patients typically presented with cerebral hemiatrophy, facial asymmetry, thickening or thinning of cranial vault, contralateral hemiplegia or hemiparesis, seizures, mental retardation. Diagnosis is clinico-radiological. Here we describe a 16 year old girl who presented to us with resistant seizures and hemiparesis.

**CASE REPORT:** A 16 years old girl, was admitted in the General Medicine indoor with complaints of recurrent apparently generalised seizures since 3 months of age along with left sided hemiparesis for some duration without any mental retardation or developmental delay. She had similar episodes in spite of adequate number of antiepileptic drugs in maximal doses for the above mentioned duration. There was no history of fever, headache, vomiting, trauma in head and any significant abnormal birth history. After admission, we observed that she had recurrent episodes of right sided focal onset partial seizure with secondary generalisation.

There was facial asymmetry with lesser development of left side of face. [Image 1] Anthropometric measurements revealed vertex symphysis length as 30 inches, left leg was one inch less than right with a normal arm span. The muscle bulk of left side of the body was less compared with the right. The limb circumferences of left upper and lower limbs were 3 cm less than the right. There was also left sided hemiparesis with spastic wrist flexion [Image 2]. All the deep tendon jerks of left side were exaggerated with extensor plantar response. The left side of the face showed decreased facial furrows and development. Neuropsychiatric evaluation revealed no mental retardation. Although mental retardation has been associated with this disorder it is not an essential component, and cases have been reported without mental retardation. [2]

She was on adequate doses of three antiepileptics viz phenobarbitone 120 mg/d, valproate 1000 mg/d and phenytoin 300 mg/d. There was no history suggestive of noncompliance but her seizures were not controlled and she had multiple episodes per day. Serum drug levels were measured which were phenobarbitone 24 mcg/ml (15-40 mcg/ml), valproate 55 mcg/ml(50-100

mcg/ml), Phenytoin 17 mg/ml(10-20 mcg/ml). Her electrolytes levels were : Sodium 138, Potassium 3.7, Calcium 8.6, Magnesium 1.7.mEq/L).The routine haemogram, renal and liver functions were normal. Screening for infectious agents were negative.

Electroencephalogram revealed asymmetry of amplitude and asymmetric slow wave runs on left side. Magnetic Resonance Imaging of brain revealed enlarged right temporal horn with thinning of the right temporal lobe. Right frontal, ethmoidal and right maxillary sinuses were enlarged. Right cerebral hemisphere was small in size with dilated lateral ventricle with prominent sulci and right sided calvaria was thick.[Image 3]

The patient was gradually introduced on carbamazepine and the dose was titrated with simultaneous withdrawal of valproate. The patient was maintained on carbamazepine 800 mg/d, phenobarbitone 120 mg/d, phenytoin 300 mg/d. On the above medications seizure frequency decreased significantly, after which she was discharged. The aim of therapy in such patients is maintaining the patient on the best possible seizure free state with the minimum number of antiepileptic drugs, so that side effects of these drugs don't accumulate and cause major life threatening complications. Considering carbamazepine a better alternative to valproate for focal onset seizures drug modification probably helped in better control of her seizures.

**DISCUSSION:** Dyke Davidoff Masson Syndrome is characterized clinically by variable degrees of facial asymmetry, hemiparesis, seizures, learning disabilities and mental retardation with behavioural abnormalities.The radiological findings include cerebral hemiatrophy, ipsilateral osseous hypertrophy and hyperpneumatization of sinuses. Although either sex may be affected Unal et al., in a retrospective study of 26 patients showed that it was more frequent in the males with left cerebral hemisphere involvement [3].This syndrome is generally classified into Congenital (Primary) and Acquired(secondary). Congenital variety is mainly caused as a result of vascular occlusions in-utero or in the postnatal period. Hageman et al., propounded the terms cerebral hemi-hypoplasia or unilateral cerebral hypoplasia for the primary form since there is lack of cerebral growth rather than atrophy [4]. Vascular occlusions in the perinatal period in the middle cerebral vascular territory, unilateral cerebral arterial circulation anomalies, coarctation of the middle aortic arch, mesencephalon hypoplasia and Wallerian degeneration are proposed as some of the causes for the Congenital variety [5]. Primary form can be differentiated radiologically by careful observation of compensatory brain changes. Development of the brain reaches 50% of adult size during first year of life and 75% by the end of third year. When it enlarges, it presses the bony tables outwards which gradually results in general shape of the adult head. However, failure of the brain to grow causes other structures in the calvaria to grow inwards, accounting for hyper pneumatization of ipsilateral sinuses, increased width of the diploic space,elevations of the greater wing of the sphenoid as well as the petrous ridge, shift of midline structures towards ipsilateral side and the sulcal prominence is absent. Differential diagnosis to be considered in a patient of cerebral hemiatrophy are Sturge-Weber Syndrome ,Linear nevus syndrome, Silver Russel Syndrome and Rassmussen encephalitis. Polypharmacy is usually needed to control seizures, but judicious use of antiepileptic drugs is required to maximize outcome with minimal side effects. Outcome is better if hemiparesis occurs after 2 years and in absence of prolonged or recurrent seizures. Children with recurrent and disabling seizures and hemiplegia can undergo hemispherectomy with a success rate of 85% and immunotherapy can occasionally be undertaken in selected cases.[5].

**CONCLUSION:** This case reminds us that structural abnormalities can cause resistant seizures and aggressive injudicious polytherapy can expose the patient to unnecessary adverse complications of therapy. It is best in these cases to keep the patient on minimum and appropriate type and dosage of antiepileptic medications even with persisting non disabling seizure spells.

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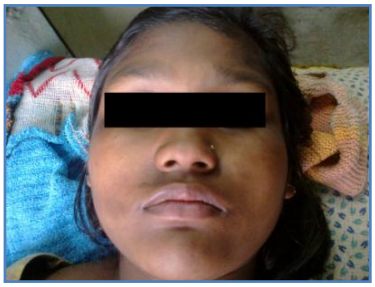

Image 1 : Facial asymmetry in Dyke Davidoff Masson Syndrome

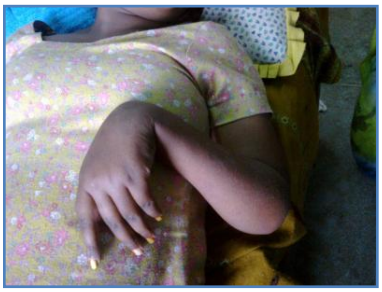

Image 2 : Left sided spastic wrist flection.

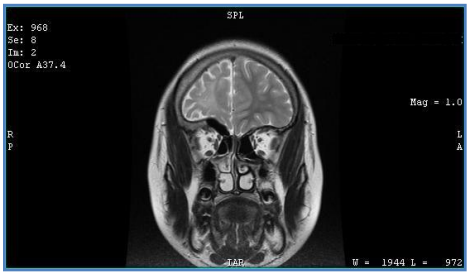


Image 3 : Unilateral hemiatrophy with thickened calvaria of right side.

# CASE REPORT

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