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ADULT PRESENTATION OF DYKE-DAVIDOFF-MASSON SYNDROME: RARE CAUSE OF REFRACTORY EPILEPSY- A CASE REPORT WITH MRI FINDINGS.

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ABSTRACT Dyke Davidoff Masson Syndrome (DDMS) is a rare neurological syndrome which is associated with refractory seizures, contralateral hemiparesis, mental retardation, behavioural and learning disabilities. It is characterised according to congenital and acquired causes. Although it is more prevalent in males with left hemispheric predominance, either of the hemispheres and sex groups can be affected. Our case report describes a 35 year old male patient of Dyke Davidoff Masson syndrome with right hemispheric involvement. The patient's clinical history and radiological findings of DDMS are discussed.

KEYWORDS : Seizures, Refractory epilepsy, Calvarial thickening, Cerebral hemi-atrophy, Dyke Davidoff Masson Syndrome

Introduction:

Dyke Davidoff Masson Syndrome refers to the cerebral hemiatrophy associated with constellations of the neurological symptoms. Probable cause is the ischemic insult to the developing brain in the late antenatal, perinatal, postnatal or early childhood period. The syndrome usually presents with recurrent and refractory seizures, contralateral hemiparesis, mental retardation, learning disability, delayed milestones, speech and language disorders.

Case report: A 35 year old male patient with a history of long standing epilepsy was referred to our department for MRI evaluation of the brain to rule out the cause of the seizures. MR imaging was done on a 3 Tesla - MR scanner (signa Hdxt, General Electrics Medical System, Milwaukee, USA) with standard 12 channel head coil. Axial T2 (propeller), T1 W, T2 Flair, diffusion weighted imaging with b value of 1000 m2/sec, BRAVO and post contrast T 1 sequences were performed using appropriate parameters. MR imaging revealed presence of right cerebral hemiatrophy with ipsilateral sulcal widening, right lateral ventricular dilatation, and enlargement of the right sided paranasal sinuses and petromastoid air cells along with thickening of the calvarium on the right side. No evidence of any abnormal white matter signals noted. The features were consistent with Dyke Davidoff Masson Syndrome (DDMS).

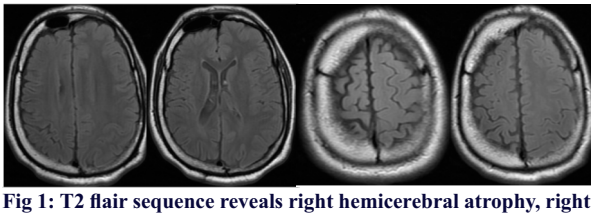

Fig 1: T2 flair sequence reveals right hemicerebral atrophy, right sided calvarial thickening, ipsilateral ventricular and right frontal sinus dilatation. No white matter signal abnormalities noted.

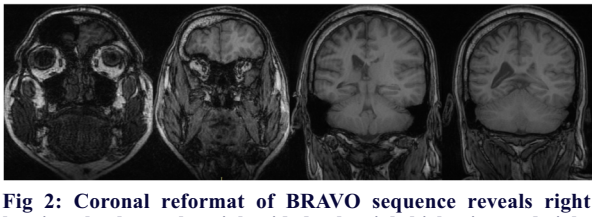

Fig 2: Coronal reformat of BRAVO sequence reveals right hemicerebral atrophy, right sided calvarial thickening and right lateral ventricular dilatation. Right frontal sinus is prominent as compared to the left side. Cerebellar hemispheres are normal.

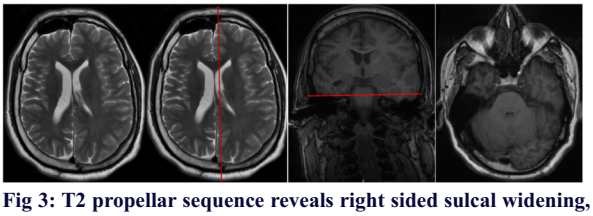

Fig 3: T2 propellar sequence reveals right sided sulcal widening, right lateral ventricular dilatation and mild ipsilateral falcine shift. Coronal and axial T1 weighted images revealed right petrous ridge elevation with prominence of the right sided petromastoid air cells

DISCUSSION:

Dyke Davidoff Masson Syndrome (DDMS) is usually characterised by cerebral hemi atrophy, refractory seizures, contralateral hemiplegia, facial asymmetry, and mental retardation [1]. The etiology of DDMS could be traced to two groups: congenital or primary type and acquired or secondary type. In the congenital type, vascular insult to the cerebral parenchyma occurs during intrauterine life and symptoms appear at birth or postnatal period. In the acquired or secondary type, cerebral insult occurs during the perinatal period or later, etiologic factors being infection, trauma, cerebral vascular abnormalities, ischaemic and haemorrhagic causes, and in premature infants, subependymal germinal matrix and intraventricular haemorrhage [2,3].

The development of the cerebral hemiatrophy could be explained by the looking into the embryological development of the brain. The sulci formation starts around the fourth month of gestational period and usually gets completed by the eighth month. The maximum growth of a child's head usually occurs in the early postnatal life due to outward pressure of the growing human brain on the bony calvarium. Human brain reaches half of its adult size at the end of first year and three fourths of the adult size by the end of 3 years [4]. If brain damage occurs before 3 years of age, structures overlying the brain start growing inward, with resultant increased width of the diploic spaces, enlarged sinuses, and an elevated orbital roof, which are characteristic of this disorder [5]. The classical MRI changes of this disease are seen only if there is a brain insult due to the various causes listed above occurs before 3 years of age [5]. In congenital insult there is shift of midline structure towards the side of disease and absence of the sulcal prominence, but the atrophied cerebral hemisphere will have prominent sulcal spaces if the insult occurs at the end of sulcation or after birth [6]. Several ischemic episodes resulting from different causes are supposed to reduce the production of brain-derived neurotrophic factors, resulting into cerebral atrophy [7].

MRI findings in our case were consistent with DDMS as has been described in literature [8, 9]. Our patient has been kept on empirical antiepileptics and follow up in neurology OPD. There are only few case reports of adulthood presentation of DDMS described in medical literature [10]. The differential diagnosis of this cerebral hemiatrophy includes Sturge-Weber syndrome, Rasmussen encephalitis, Silver-Russell syndrome, basal ganglia germinoma, Fishman syndrome, and linear nevus syndrome which can be differentiated by performing a proper history taking, clinical examination and by characteristic neuroimaging features [11, 12].

DDMS usually present as refractory seizures in early childhood. Hemispherectomy has been mentioned as a treatment of choice with a success rate of 85% in selected cases [13]. However, in late adulthood presentation as in our case, if seizures are under control, the patient can

be kept on antiepileptic medications instead of surgery, along with supportive physiotherapy, speech therapy, and occupational therapy. Further longitudinal studies are required to document the natural course of this syndrome especially in an adult population, which could help in planning treatment strategies, whether conservative or intervention, in these patients accordingly.

Conclusion:

Radiological findings of our patient are consistent with DDMS which involved the right cerebral hemisphere. MRI played an important role in diagnosis of this patient to delineate the cerebral changes along with changes in calvarium and sinuses. This patient was kept on follow up antiepileptic medication under neuromedicine department and is currently symptom free.

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