Case Report

Dyke – Davidoff – Masson Syndrome

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INTRODUCTION

There are a variety of causes that lead to cerebral hemi atrophy with resultant complications like poor physical growth , hormonal abnormalities , intellectual insufficiencies, seizures , hemiplegia etc. [1-4] Dyke,Davidoff and Masson published a series of nine cases in 1933 presenting with hemi paresis , seizures , facial asymmetry. This condition is called as Dyke-Davidoff-Masson Syndrome.(DDMS).[6] Brain insult from a variety of causes from foetal stage to early infancy can result in cerebral hemi atrophy. The causes are either congenital, idiopathic (Primary) and intrauterine vascular injury or acquired, i.e. perinatal intracranial hemorrhage, infection (Encephalitis), trauma, vascular abnormalities (Sturge-Weber's Syndrome), ischemia, hypoxia, etc. Magnetic Resonance Imaging (MRI) reveals changes in the brain parenchyma with thinning of grey matter, reduced volume of white matter, enlarged lateral ventricle, reduced size of cerebellar hemisphere.

We report a case of recurring seizures in a young 18 year-old female who presented to psychiatry outpatient in a tertiary care hospital and was diagnosed with cerebral asymmetry that could be traced to a cerebral insult in infancy.

CASE PRESENTATION

A 18-year-old female was brought to the Psychiatric OPD by her father with the complaint of behavioral problems, recurring seizures, and weakness of right upper and lower limbs, Detailed history revealed that the patient was born at full term through vaginal delivery without any trauma/birth asphyxia. Birth weight recorded was 2100g. At 5 months of age, the child suffered high-grade fever, had convulsions, and was admitted to the hospital for a month. She was suspected to be a case of Encephalitis. Although the child recovered from fever and febrile convulsions, the milestones were delayed and speech was affected. Later, an IQ assessment showed moderate mental retardation (IQ 30-49). As she grew up, seizures returned at regular intervals and she was put on anticonvulsants by the neurophysician. She started showing behavioral changes in 2013 (at 13 years of age) in the form of irritability, violence and withdrawn behaviour with not doing any household work while on anticonvulsants Tab Oxcarbazepine 150 mg two times a day. She was put on low doses of atypical antipsychotic tab. Risperidone 1 mg once a day and small dose of benzodiazepine (Clonazepam 0.25 mg/day) and multivitamins. Behaviour therapy and counselling of family members helped in better outcome of the case. She remained symptom free. Frequency of seizures reduced considerably to once in 5 to 6 months. General physical examination was normal. Her gait was hemi paretic type with motor power grade III in right upper and lower limbs.

All hematological and hormonal (T3, T4, TSH, FSH, LH) investigations revealed no abnormality. NCCT brain done on 08/08/2019 revealed no thickened skull vault with thickened diploic spaces. Mild prominence of the sylvian fissure and sulcal spaces on the

left side indicated atrophy of the brain. Mild prominence of left lateral ventricular system was noted. Midline structures were maintained. Cerebellum and brain appeared normal (Figure 1).

C.T. Brain Images of the Patient:

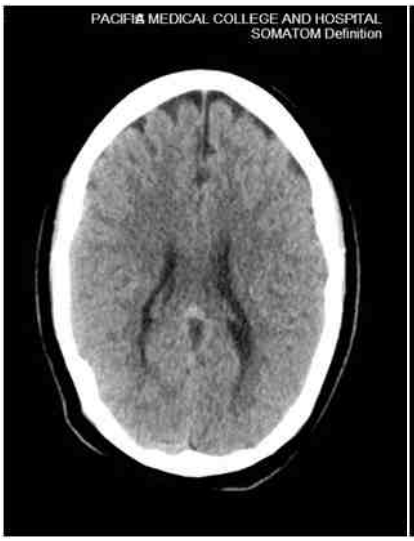

Figure – 1

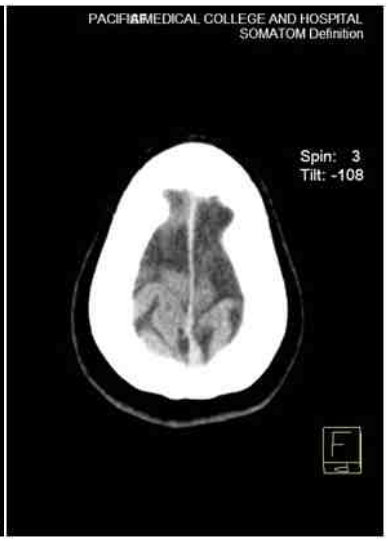

Figure – 2

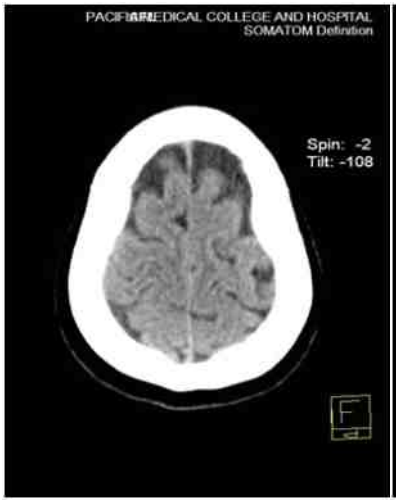

Figure – 3

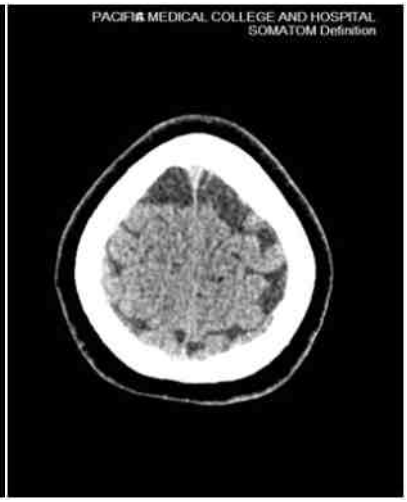

Figure – 4

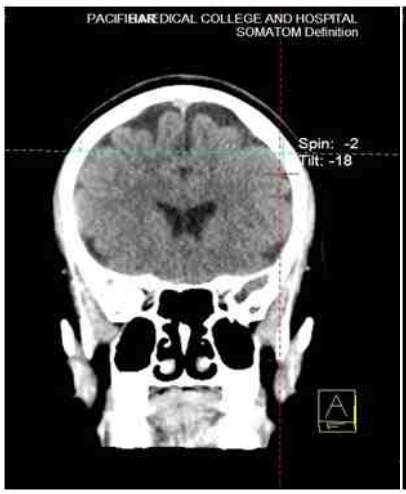

Figure – 5

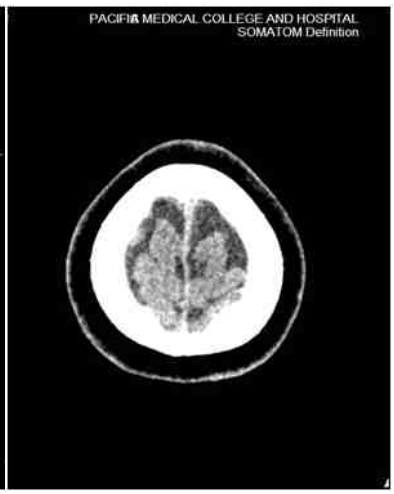

Figure – 6

DISCUSSION

From the foregoing, it is apparent that the patient suffered a febrile illness, which was labeled as Encephalitis at a very vulnerable age of 5 months and affected the growth of the brain. The NCCT Brain findings (figure 1 to 3) of temporal and frontal lobe atrophy together with seizures and mentally challenged state resembled the condition described by Dyke, Davidoff, and Masson's series of 9 cases in 1933. The appearance in these cases showed thickening of calvarium, dilatation of frontal, and ethmoid sinuses, and elevation of greater wing of sphenoid and petrous ridge. The CT Brain findings in the above case almost conform to this condition. It is a fact that the brain grows to half of its adult size by the first year and reaches 75% of the adult size by three years. Any insult during these years will result in stunted growth of the brain and the skull vault grows inwards resulting in thickening of the vault, enlargement of frontal sinuses and increases width of diploic spaces and greater wing of sphenoid and petrous ridge on the affected side. This is possible when the brain damage is sustained in the first few months of infancy, as in this case. This shows that DDMS although not a very common condition, it can present in varying degree of severity. What is important is to document a proper clinical history, a detailed clinical examination and investigations (CT, MRI, and Hormonal) to confirm the diagnosis of DDMS. Cerebral insult very early in infancy due to febrile illness (Encephalitis) seems likely to be the cause in this case.

CONCLUSION

Dyke-Davidoff Masson Syndrome is a rare condition resulting from brain injury in early age due to a multitude of causes. Characteristics include cerebral hemiatropy/ hypoplasia, contralateral hemi paresis, seizures and compensatory osseous hypertrophy.

This 18 yrs female presented with right sided weakness , recurring seizures , poor intellectual functioning and behavioural problems. History of the Encephalitis at the age of 5 months led to development of above presentation. Milestones were delayed and the IQ was between 30 to 45.

Proper detailed history and relevant investigations , with suspicion of the DDMS diagnosis helped in confirming the diagnosis .Prognosis of the case remained guarded.

Declaration of the Patient Consent:

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient and relatives understand that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be assured .

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Nil

Conflicts of Interest :

There are no conflicts of interest.

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