Case Report

Dyke-Davidoff Masson Syndrome: A Rare Case Report with Delayed Diagnosis

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ABSTRACT

Dyke-Davidoff Masson syndrome is a rare case characterized by cerebral hemi-atrophy secondary to in-utero or early childhood cerebral insult, such as infarct, trauma or infection. It usually presents in infancy or childhood with unilateral hemiplegia or hemiparesis, seizures, facial asymmetry and mental retardation. Here we are reporting a rare case of Dyke-Davidoff Masson syndrome in an 18-year female patient who came for routine evaluation to medicine OPD and was diagnosed with Dyke-Davidoff Masson syndrome on MRI scan.

Key words: Cerebral hemi-atrophy, childhood, MRI.

INTRODUCTION

Dyke-Davidoff Masson syndrome (DDMS) is an uncommon disease presenting in infancy or childhood with unilateral hemiplegia or hemiparesis, seizures, facial asymmetry and mental retardation. However, the clinical presentation as well as the radiological features may be variable depending upon the age and extent of cerebral insult. We report a case of DDMS in an 18-year female patient who came for routine evaluation to medicine OPD and was diagnosed with Dyke-Davidoff Masson syndrome on MRI scan.

CASE REPORT

An 18 yr. old unmarried female born of a non-consanguineous marriage with an uneventful birth history presented with no significant active complains and history of minimal non-progressive weakness of left upper and lower limbs at the age of 6 months. She had no history of seizure and no past history or any documents supporting the etiology. There was no history of any behavioral problems. On neurological examination, cognitive functions were normal. Cranial nerve examination was unremarkable. Left upper and lower limbs were spastic with power 4/5 and brisk deep tendon reflexes. No neurocutaneous markers were present. Various laboratory investigations were within normal limits.

MRI brain was performed in Siemens 3T Magnetom skyra MRI machine, images follows – Fig 1 to 4.

With the above findings, diagnosis of Dyke-Davidoff Masson syndrome was made. The patient was treated conservatively.

DISCUSSION

DDMS refers to hemi-atrophy of one cerebral hemisphere secondary to brain insult in fetal or early childhood period. Typical clinical features of hemiparesis, facial asymmetry, seizures and mental retardation are described in literature; though atypical presentation is possible. Predominantly there is no sex predilection as well as any particular cerebral hemispheric involvement. However, involvement of left cerebral hemisphere and male gender have shown to be more common in literature.

Depending upon the age at presentation, it is divided into two types: Infantile (congenital)- Results from in-utero infection, neonatal or gestational vascular occlusion involving middle cerebral artery and coarctation of mid aortic arch. Acquired- Main causes are early childhood cerebral insults like trauma, tumor, infection, ischemia, hemorrhage or prolonged febrile seizures.

To know the pathogenesis, we have to go through the development of brain precisely. The brain sulci formation occurs between the fourth to end of eighth months of fetal life. However, the maximum growth of a child's head reaches half of its adult size at the end of first year and three fourths of the adult size by the end of three years; outward pressure of the enlarging human brain on the bony skull is the cause behind rapid growth of head circumference. Hence, whenever brain damage is sustained before three years of age, bony skull overlying the brain grows inward resulting in an increased width of the diploic spaces, para nasal sinuses, and elevation of petrous ridge and orbital roof, which are tell-tale fea

tures of this disorder.

The plain skull radiograph illustrates thickening of 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.

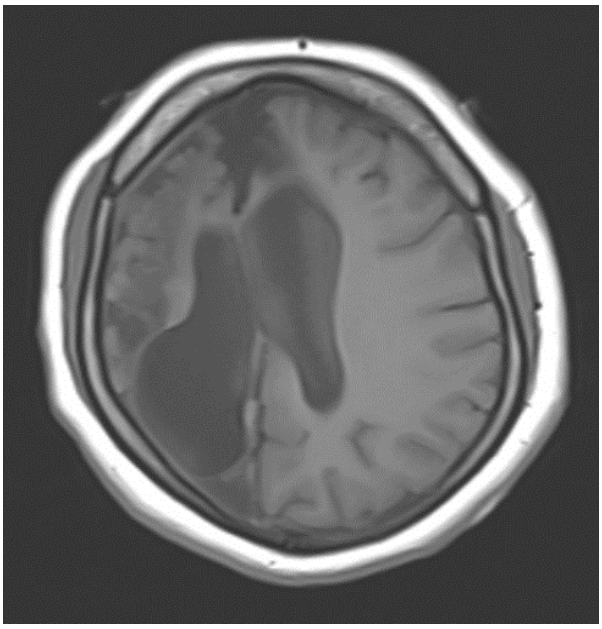

Figure 1: Axial T1WI shows right cerebral hemiatrophy with enlarged right lateral ventricle, off midline falx and inter- hemispheric fissure and thickened calvaria.

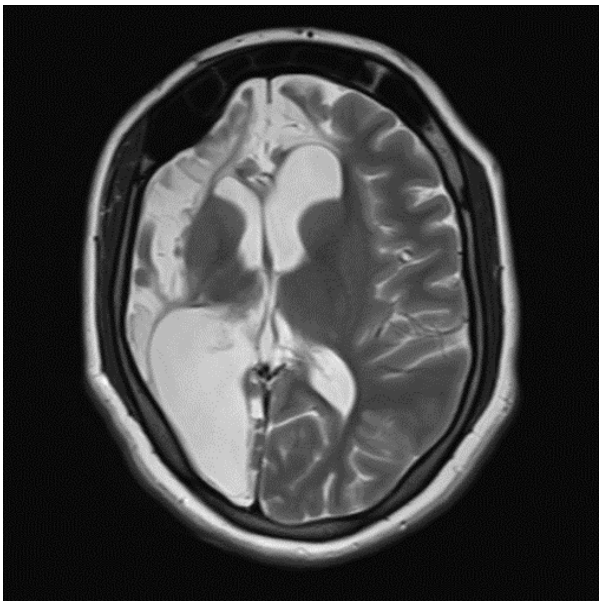

Figure 2: Axial T2WI in the same patient shows the enlarged sulcal spaces with atrophic right hemisphere and enlarged frontal sinuses.

It is associated with compensatory calvarium thickening, hyper-pneumatization of paranasal sinuses and mastoid cells and elevation of petrous ridge. In congenital hemi-atrophy, when the insult occurs in utero, there is a shift of midline structures towards the disease side, but there is absence of sulcal prominence replacing the gliotic tissue. This is the salient feature differentiating congenital from acquired form.

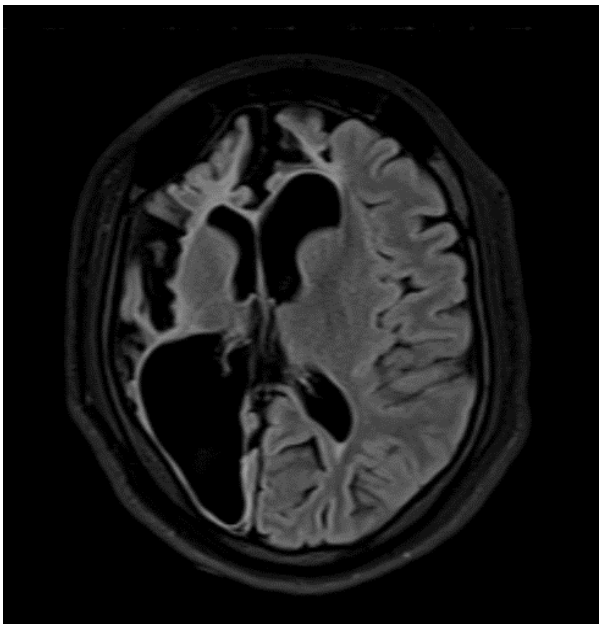

Figure 3: Axial FLAIR images in the same patient shows cortical atrophy with extensive white matter gliosis & hyperintensity and shrunken basal ganglia.

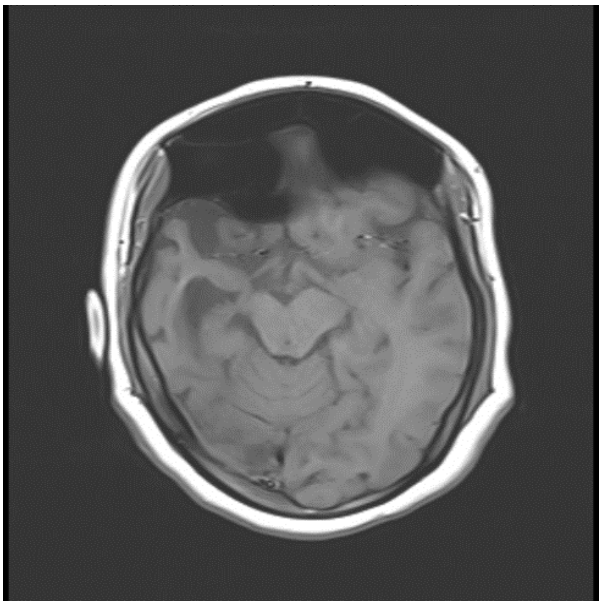

Figure 4: More caudal axial T1WI show mild shows mild atrophy of ipsilateral cerebral peduncle and enlarged paranasal sinuses.

In a patient with cerebral hemi-atrophy, Rasmussen encephalitis, Sturge-Weber syndrome, Silver-Russell

syndrome are common differential diagnoses to be considered. These three can be differentiated by thorough clinical examination and cross-sectional neuroimaging. Rasmussen encephalitis does not show calvarial changes with almost similar clinical history and Sturge-Weber syndrome additionally shows enhancing pial angiomas and cortical calcifications and facial port-wine nevus. Silver-Russell syndrome has a classical phenotype, clinodactyly, delayed bone age with normal intelligence and normal head circumference.

The treatment of DDMS is symptomatic, and must be oriented to treat convulsion, hemiplegia, hemiparesis and learning difficulties. Prognosis is better if hemiparesis occurs after the age of 2 years and without prolonged or repetitive seizures. Children with intractable seizures are potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases. Hence, early diagnosis makes early decision making and intervention.

CONCLUSION

DDMS are a rare preventable cause of hemiplegia, refractory epilepsy and mental retardation in childhood. Adult presentation of DDMS is unusual and has not been reported in medical literature. A thorough history and imaging can give an early diagnosis and differentiate from other close differentials. Treatment is largely supportive and aims at controlling seizures along with physiotherapy, occupational therapy and speech therapy.

REFERENCES

1. Osborn's Brain Imaging, Pathology and Anatomy by Anne G. Osborn
2. Radiopaedia.org article on Dyke-Davidoff Masson syndrome by Assoc Prof Frank Gaillard and Dr MaiLan Ho
3. Grossman RI, Yousem DM. Neuroradiology, the requisites.
4. Vosskamper M, Schachenmayr W. Cerebral Hemiatrophy: A clinicopathological report of two cases with a contribution to pathogenesis and diferential diagnosis.
5. Dix JE, Call WS. Cerebral hemiatrophy: classification on the basis on MR imaging findings of mesial temporal sclerosis and childhood febrile seizures.
6. Kirts DR, Griscom NT. Practical pediatric imaging, diagnostic radiology of infants and children. Lippincott Williams & Wilkins.
7. Dyke CG, Davidoff LM, Masson CB: Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. Surg Gynecol Obstet.
8. Singh P, Saggar K, Ahluwalia A. Dyke- Davidoff- Masson syndrome: Classical imaging findings.
9. Thakkar PA, Dave RH. Dyke-Davidoff- Masson syndrome: A rare cause of cerebral hemiatrophy in children. (2016) Journal of pediatric neurosciences.
10. Gokce E, Beyhan M, Sade R. Radiological imaging findings of Dyke-Davidoff- Masson syndrome. (2017) Acta neurologica Belgica.