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Dyke-Davidoff-Masson syndrome: A case report

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ABSTRACT

A 7 years girl presented with seizures, intellectual disability, right sided spastic cerebral palsy, and strabismus of right eye. Magnetic resonance imaging (MRI) brain was suggestive of severe volume loss of brain parenchyma with dilatation of ipsilateral lateral ventricle, and other MRI features diagnostic of infantile type Dyke-Davidoff-Masson syndrome also termed as cerebral hemiatrophy. Seizures can be refractory to medical treatment and surgical treatment may become necessary. Greater awareness about the disease is required to diagnose the condition timely and refer to neurosurgery in the case of refractory seizures as hemispherectomy is the treatment of choice with high success rate.

Key words: *Calvarium thickening, Cerebral hemi atrophy, Dyke-Davidoff-Masson syndrome, Infantile, Intellectual disability, Refractory seizures*

Cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome (DDMS) is atrophy or hypoplasia of one cerebral hemisphere with homolateral hypertrophy of the skull and sinuses and is usually due to an insult to the developing brain in fetal or neonatal or early childhood period [1]. The clinical features vary depending on the extent of brain injury, characterized by seizures, facial asymmetry, contralateral hemiplegia or hemiparesis, and intellectual disability. Intellectual disability is not always present and seizures many times refractory may appear months or years after the onset of hemiparesis. Learning disabilities, impaired speech and behavioral abnormalities can be seen [2]. Either side of the brain may be affected without any sex predilection. Although in a case series left cerebral hemisphere and male dominance was seen [3]. The disease, supposed to be a rare entity once, is increasingly diagnosed and reported [2-5] recently may be because of increased survival rates in critical newborn babies and availability of better diagnostic facilities in the urban area and poor obstetric and newborn care in the rural area. Therefore, we report such a case being managed as spastic cerebral palsy and later found to have DDMS.

CASE REPORT

A 7-year-old girl presented with multiple types of seizures (tonic, myoclonic), intellectual disability, right sided hemiparesis, facial asymmetry for past 4 years. The patient had a history of severe perinatal asphyxia and delayed developmental milestones. She was on valproate and diazepam for past 2 years with very poor compliance and continued to have 3-4 episodes of seizures per day. On examination, the patient had typical spastic hemiplegic gait and posture. She had mild hemiatrophy on the right side and strabismus of right eye. Her IQ score was 40 according to Stanford-Binet test.

Her routine hematological and biochemical investigations including serum electrolytes, blood sugar, calcium, LFTs, and RFT were normal. Her serum valproate level was <1 mcg/ml which is markedly below minimum effective concentration. A plain magnetic resonance imaging (MRI) of the brain was done which revealed extensive areas of cystic encephalomalacia and gliosis with nearly complete loss of parenchyma in the left cerebral hemisphere with ex vacuo dilatation of ipsilateral lateral ventricle (Fig. 1) on axial plane images. Pulling of interhemespheric fissure on the left side was also seen. Furthermore, enlargement of left sided frontal and mastoid sinuses (Fig. 2) with mild thickening of skull vault (Fig. 3) was seen in T1W images.

These MRI findings along with the clinical picture were fulfilling the criteria for DDMS, and hence, the diagnosis of DDMS (infantile form) was made. The patient was started on oral valproate and increased up to a dose of 25 mg/kg/day to achieve therapeutic levels, but seizures continued and levetiracetam added and dosing increased up to 30 mg/kg/day. The patient is kept in follow-up and might need an increment of doses as she was not completely seizure free before the discharge again in favor of developing refractory seizures as in the case of DDMS.

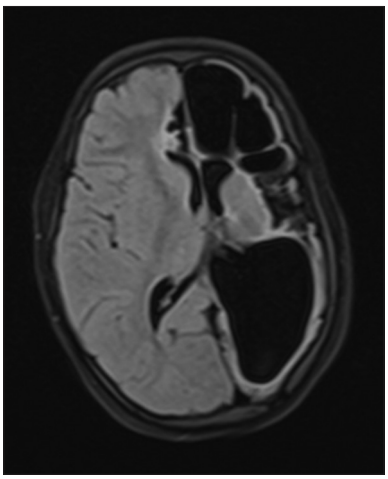

Figure 1: Cystic encephalomalacia and gliosis with parenchymal loss & ex vauco dilatation of ipsilateral lateral ventricle with pulling of interhespheric fissure

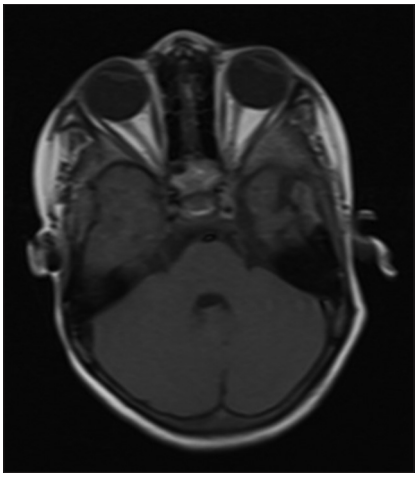

Figure 2: T1W MRI images showing left hypertrophy of frontal and mastoid air sinuses

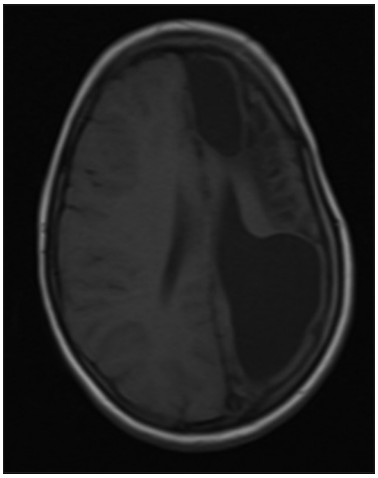

Figure 3: T1W MRI image showing calvarium thickening

DISCUSSION

This rare entity was first described by Dyke, Davidoff, and Masson in 1933. In a series of 9 patients, they described characteristics clinical features including hemiparesis, seizures, facial-asymmetry, mental retardation and with characteristic plain skull radiographic, and pneumatoencephalographic changes [1]. The radiological findings include cerebral hemiatrophy, ipsilateral osseous hypertrophy causing enlargement of calvarium, diploic spaces and hyperpneumatization of sinuses, and elevated temporal bone. These compensatory cranial changes occur to take up the relative vacuum created by the atrophied cerebral hemisphere [5].

It has been reported that DDMS is caused by the cerebral insult that may occur in utero when the maturation of calvarium has not been completed, or during early life due to brain damage. Prenatal causes include congenital abnormalities, cerebral infarction, vascular malformations, and infections. Perinatal causes include birth trauma, hypoxia, and intracranial hemorrhage. Finally, cerebral hemiatrophy can develop secondary to cerebral trauma, tumors, infections, and prolonged febrile seizures after birth. Bony changes can be better visualized on X-ray and computed tomography than in MRI. It is classified in congenital or infantile and acquired variety [6]. Shen et al. [7] described three patterns of MRI showing cerebral hemiatrophy; pattern I) corresponds to diffuse cortical and subcortical atrophy; pattern II) corresponds to diffuse cortical atrophy coupled with porencephalic cysts and pattern III) corresponds to the previous infarction with gliosis in middle cerebral artery territory. Our patient had a history of perinatal insult with developmental delay and infantile hemiplegia; she developed seizures at the age of 3 years requiring multiple antiepileptic drugs and MRI findings were diagnostic of DDMS.

Differential diagnosis of cerebral hemiatrophy includes Hemiconvulsion-hemiplegia-epilepsy (HHE) syndrome, Rasmussen's encephalitis, Sturge-Weber syndrome, Silver-Russell syndrome, linear nevus sebaceous syndrome, progressive multifocal leukoencephalopathy, and Fishman syndrome. HHE syndrome is characterized by hemiconvulsion seizures, hemiplegia, and epilepsy syndrome in sequence. Hemiplegia can be transient or permanent, and epilepsy can develop after a variable period of 1-3 years, and neuroimaging shows unilateral edematous swelling of the contralateral hemisphere at the time of initial status epilepticus followed by characteristic cerebral hemiatrophy later [8]. Detailed history, thorough clinical examination and appropriate investigation including MRI findings are the key to the diagnosis. The main concern in DDMS is refractory seizures unresponsive to medical therapy; these patients are candidates for cerebral hemispherectomy, which is helpful in eradicating or significantly reducing seizures in 85% of the patient [9].

CONCLUSION

Seizures in DDMS are often refractory to the medical treatment and surgical treatment may become necessary in these cases. Greater awareness about the disease is required to diagnose the condition timely and refer to neurosurgery in the case of refractory seizures as hemispherectomy is the treatment of choice with high success rate.

REFERENCES

1. Dyke CG, Davidoff LM, Masson CB. Cerebral hemiatrophy and homolateral hypertrophy of the skull and sinuses. Surg Gynecol Obstet. 1933;57:588-600.
2. Ono K, Komai K, Ikeda T. Dyke-Davidoff-Masson syndrome manifested by seizure in late childhood: A case report. J Clin Neurosci. 2003;10(2):367-71.
3. Unal O, Tombul T, Cirak B, Anlar O, Incesu L, Kayan M. Left hemisphere and male sex dominance of cerebral hemiatrophy (Dyke-Davidoff-Masson syndrome). Clin Imaging. 2004;28(3):163-5.
4. Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson syndrome: Classical imaging findings. J Pediatr Neurosci. 2010;5(2):124-5.
5. Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. J Neurosci Rural Pract. 2012;3(3):411-3.
6. Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy. Clin Imaging. 1992;16(2):93-7.
7. Shen WC, Chen CC, Lee SK, Ho YJ, Lee KR. Magnetic resonance imaging of cerebral hemiatrophy. J Formos Med Assoc. 1993;92(11):995-1000.
8. Bhat RY, Kakkar S, Prakashini K. Hemiconvulsion-hemiplegia-epilepsy syndrome: Clinical course and neuroradiological features in a 20-month-old girl. BMJ Case Rep. 2014;2014. Pii: bcr2013203482.
9. Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson syndrome. Indian Pediatr. 2008;45(11):927-8.

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