Research Paper Medical Science

Dyke-Davidoff-Masson syndrome- A Case report

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ABSTRACT

Dyke-Davidoff-Mason syndrome was diagnosed in a 2 year child who presented with history of generalised seizures since the age of 5 months. Significant birth history included delayed cry and a stay of 30 days in intensive care unit. The milestones were delayed. On examination the vitals were stable. The head circumference measured 44 cm (< -3 SD for the age). There were no neurocutaneous markers. The child had left hemiparesis with brisk tendon reflexes and extensor plantar reflex. CT brain revealed hemiatrophy on the right side with ipsilateral shift of falx . The sulcal spaces and ventricular system on the same side were prominent. The overlying calvarium was thickened.

INTRODUCTION

Dyke-Davidoff-Masson syndrome is a rare epilepsy syndrome with predominant manifestations of seizures, facial asymmetry, hemiatrophy, contralateral hemiparesis, skull vault thickening and mental retardation.

KEYWORDS Cerebral hemiatrophy; Dyke-Davidoff-Mason syndrome; seizures

CASE REPORT

A 2 year old male child, 1st by birth order and born of non-consanguinous marriage, presented to our services with history of generalised seizures since the age of 5 months. Significant birth history included delayed cry and a stay of 30 days in intensive care unit. The milestones were delayed. On examination the vitals were stable. The head circumference measured 44 cm (< 3 SD for the age). There were no neurocutaneous markers. The child had left hemiparesis with brisk tendon reflexes and extensor plantar reflex. The cranial nerves and CSF examination were normal. CT brain revealed hemiatrophy on the right side with ipsilateral shift of falx (arrow in figure 1). The sulcal spaces and ventricular system on the same side were prominent. The overlying calvarium was thickened(arrow in figure 2). Based on above findings, a diagnosis of Dyke-Davidoff-Mason syndrome was made and the child was started on anti-convulsant therapy.

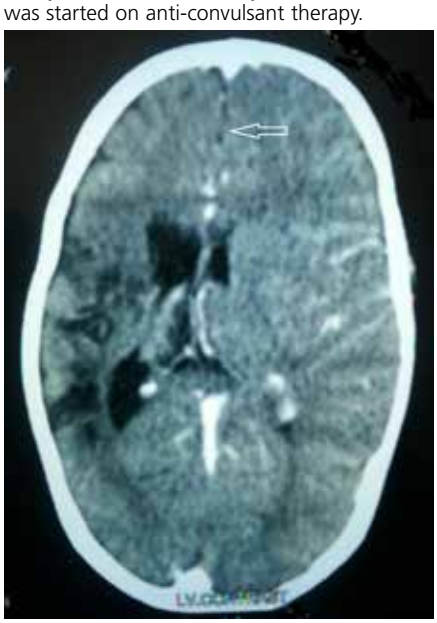

Figure 1

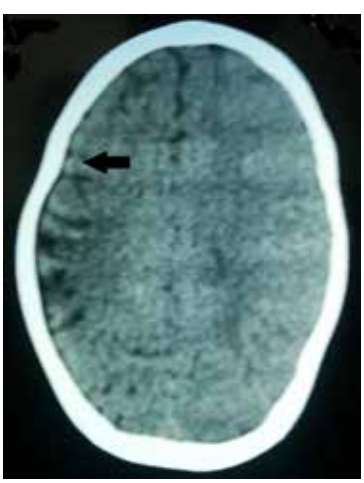

Figure 2

Figure 3

DISCUSSION

Dyke-Davidoff-Mason syndrome, first described in 1933 (1), is characterised by unilateral cerebral atrophy with compensatory hypertrophy of the overlying skull and hyperpneumatisation of the ipsilateral sinuses. The atrophic cortex displays a prominence of ventricular system and sulcal spaces (2). There is a slight male preponderance(1). The patient presents with convulsions, contralateral hemiparesis and mental retardation(1). Seizures can be focal or generalised. Other complaints include facial asymmetry, learning disability, psychiatric complaints and impaired speech(2).

The cerebral hemiatrophy may be a consequence of occlusion of middle cerebral artery due to infections or developmental anomalies(-the congenital form)(1). The acquired variety results from trauma, tumor, infection, ischemia, haemorrhage and prolonged febrile seizure(1, 3). It is the time of onset of symptoms that differentiates the two. Besides, there is presence of unilateral calvarial thickening in the congenital variety. This widening of diploic space is believed to be due to compensatory bone-growth as a result of vacuum created by atrophic neuroparenchyma(1, 3).

Differentials include: a)Sturge-Weber syndrome (cerebral atrophy associated with leptomengeal angioma and presence of port-wine facial nevus, intracranial tramtrack calcification, and the absence of midline shift) and b)Rasmussen encephalitis (unilateral hemispheric atrophy without any calvarial changes with intractable epilepsy). Other less common differentials are linear nevus syndrome, Fishman syndrome and basal ganglia germinoma(1, 4). A proper history, thorough clinical examination and imaging findings clinch the diagnosis(2).

Management approach to a child with Dyke-Davidoff-Mason syndrome includes anti-epileptic medications, physiotherapy and speech rehabilitation(4). Hemisperectomy (1)is performed in cases with intractable cases. Prognosis is favourable if there is absence of recurrent seizures and the onset of hemiparesis is after 2 years of age.

REFERENCES

[1] Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. J Neurosci | Rural Pract. 2012 Sep;3(3):411-3. doi: 10.4103/0976-3147.102646. PubMed PMID: | 23189018; PubMed Central PMCID: PMC3505357. | | [2] Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson syndrome. Indian Pediatr. | 2008 Nov;45(11):927-8. PubMed PMID: 19029569. | | [3] Paramdeep Singh, Kavita Saggar, and Archana Ahluwalia. Dyke–Davidoff–Masson syndrome: Classical imaging findings. J Pediatr Neurosci. 2010 Jul-Dec; 5(2): 124–125. doi: 10.4103/1817-1745.76108 | [4] S SHARMA, D GOYAL, A NEGI, RG SOOD, A JHOBTA, M SURYA. Dyke-Davidoff Masson Syndrome. Ind J Radiol Imag. 2006;16:2:165-166. |