

Dyke-Davidoff-Masson Syndrome with crossed cerebellar atrophy

Rohit Kumar *DNB*¹, Deepak Kumar *MD*², Himanshu Mishra *DNB*³,
Sanjay Kumar Suman *MD*⁴, Umakant Prasad *MD*⁵

^{1,2,3,4,5} Department of Radio-diagnosis, Indira Gandhi Institute of Medical Science, Patna, India

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Abstract

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological condition characterised clinically by recurrent seizures, facial asymmetry, hemiplegia and mental retardation likely due to foetal or early childhood cerebral insult. We describe the MRI findings of DDMS in a 10-year-old male child. MRI brain revealed right cerebral atrophy, ipsilateral thickening of calvarium, right lateral ventricular dilatation, hyper-pneumatisation of frontal sinus, and contralateral cerebellar atrophy which are consistent with DDMS.

Key words: Cerebellar atrophy, Dyke-Davidoff-Masson syndrome, MRI.

Introduction

Dyke-Davidoff-Masson syndrome presents during the neonatal, infantile, or early childhood period and is diagnosed by various clinical as well as radiological findings.¹ Clinical features result likely due to cerebral insult in utero or traumatic brain damage in early life and include intractable seizures, facial asymmetry, contralateral hemiparesis or hemiplegia, mental retardation, developmental delay in the form of learning difficulties, speech and language disorders.^{2,3} Radiological manifestations include cerebral hemiatrophy, homolateral hypertrophy of calvarium and sinuses, and contralateral cerebellar hemiatrophy.^{4,5,6} Many a times, it may be misdiagnosed by majority of clinicians or radiologists. Our patient, who was initially misdiagnosed as a case of Rasmussen encephalitis, was later diagnosed as a case of DDMS on the basis of imaging findings.

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Address for correspondence:

Dr. Deepak Kumar

Associate Professor, Department of Radio-diagnosis,
Indira Gandhi Institute of Medical Science,
Sheikhpura, Patna, Bihar, India.

E-mail: 1999deepak@gmail.com

Phone: +91 9835284456

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Case Report

A 10-year-old male child presented with multiple episodes of intractable seizures and left hemiparesis for the last 5-6 months. Birth was at term and uneventful. There was no noticeable history of trauma after birth. On examination, he had microcephaly without facial asymmetry. Neurological examinations revealed reduced power in left upper and lower limbs (4/5) with brisk tendon reflexes. Vision and hearing were normal, cranial nerves were intact, but there were difficulties in speech. Systemic examinations including vitals were normal.

Complete blood picture revealed mild anaemia, but it was otherwise within normal limits. Liver and renal function tests were normal. NCCT brain done previously, reported right cerebral atrophy with prominence of cortical sulci and cisterns, dilatation of ipsilateral lateral ventricle indicative of features of Rasmussen encephalitis. Subsequent MRI brain performed at our centre showed right cerebral atrophy with features of volume loss in the form of widened cortical sulci, prominent cisterns, and ex-vacuo dilatation of right lateral ventricle associated with mild ipsilateral falcine displacement and ipsilateral calvarial thickening. Contralateral cerebellar atrophy was also noted.

Discussion

DDMS or cerebral hemiatrophy was first described by Dyke, Davidoff, and Masson in 1933 on the basis of skull radiographic findings in 9 patients who clinically presented with seizures, hemiparesis, facial asymmetry, mental retardation.⁷ They found calvarial thickening and hyperpneumatization of sinuses in the same side of the lesion. Finally, they concluded that these changes occur in brain/skull secondary to cerebral insults in the form of congenital abnormalities, vascular malformations,

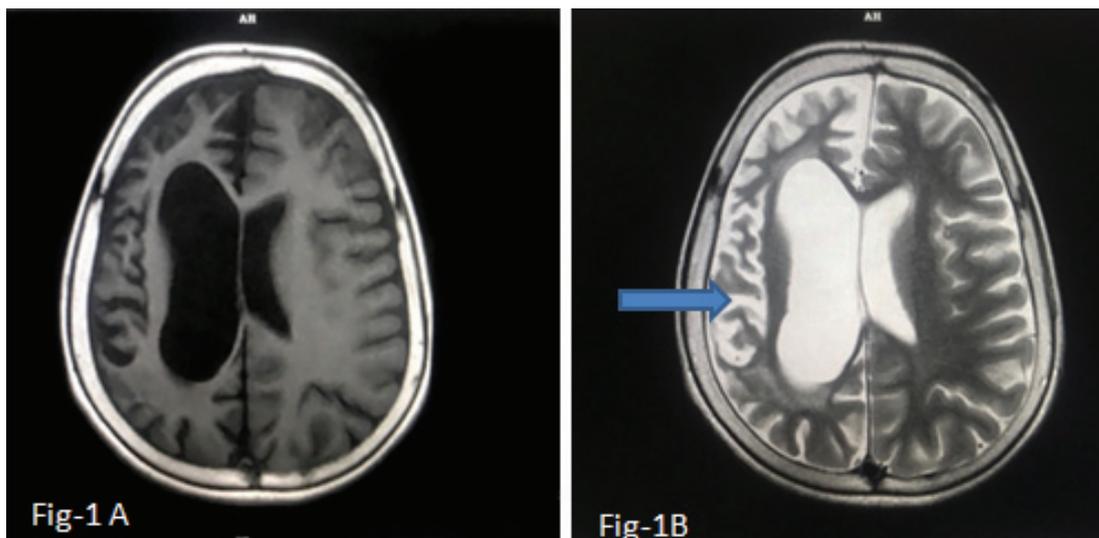


Figure 1a, 1b: (T1W) and (T2W) axial images show right cerebral atrophy with features of volume loss in the form of widened cortical sulci, cisterns and ex-vacuo dilatation of right ventricle associated with mild ipsilateral calvarial thickening.

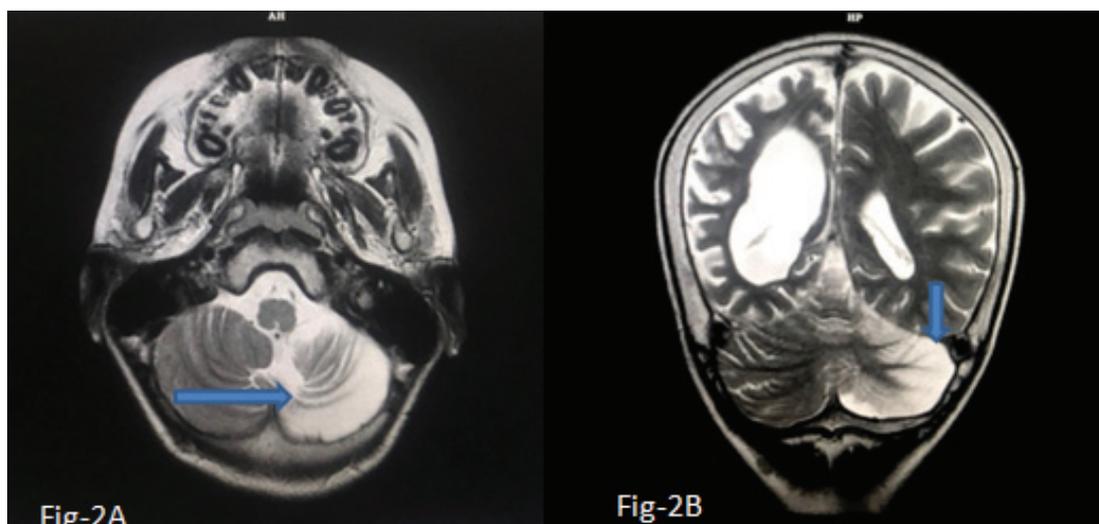


Figure 2a, 2b: T2WI axial and T2WI coronal images show left cerebellar atrophy.

and vascular occlusions leading to cerebral infarction and infections in the prenatal period, birth trauma in the perinatal period as well as infection, tumour, trauma, hypoxia, haemorrhage in the postnatal period.⁷

Cross sectional imaging reveals elevation of orbital roof, sphenoid wing, and petrous ridge ipsilateral to the lesion as skull manifestations as well as cerebral hemiatrophy, prominent cortical sulci and cisterns, ex-vacuo dilatation of ipsilateral ventricular system, and pachygyria as brain parenchymal changes. In addition to these findings, contralateral cerebellar atrophy is also occasionally seen in DDMS which was present in our case and is infrequently reported in literature.⁸

Development of the nervous system starts at 3 weeks of gestation and ¾th of adult size is reached by the age of

3 years. Cerebral insults in the perinatal period hampers the normal development of brain which is compensated by secondary changes in the form of increased width of diploic spaces, i.e., calvarial thickening, ipsilateral hyperaeration of frontal, ethmoid, and mastoid air cells, and elevation of orbital roof.⁹ These changes begin to manifest when cerebral insults occur before the age of 3 years and may become evident as early as 9 months after the cerebral injury.¹⁰ Vascular occlusion involving the middle cerebral artery and decrease in carotid artery flow due to coarctation of aorta are the main vascular causes of DDMS.¹¹ Our patient had a history of prolonged febrile seizures at the age of 3 years and was treated as a case of encephalitis. He came to our department at the age of 10 years with the complaints of intractable seizures for 5

months. MRI findings comprised of right cerebral atrophy with features of volume loss in the form of widened cortical sulci, prominent cisterns, and ex-vacuo dilatation of right lateral ventricle associated with mild ipsilateral calvarial thickening and mild ipsilateral falx displacement in addition to contralateral cerebellar atrophy.

Differential diagnosis for cerebral hemiatrophy includes Rasmussen encephalitis, hemimegalencephaly, and Sturge Weber syndrome. Rasmussen encephalitis is characterised by focal lobar or cerebral hemispheric atrophy (volume loss) without calvarial thickening.¹² Sturge Weber syndrome is characterised by leptomeningeal angiomatosis, cerebral venous malformation, intracranial tram track calcification, and cortical laminar necrosis.¹³ Hemimegalencephaly is characterised by enlarged dysplastic unilateral cerebral hemisphere or a portion of hemisphere, so that contralateral normal cerebral hemisphere may be mistaken for atrophy. Enlarged ipsilateral lateral ventricle, thickened calvarial vault, and contralateral displacement of the posterior falx are also noted in hemimegalencephaly.¹⁴ Treatment of DDMS depends on symptoms; it includes anticonvulsant therapy, physiotherapy, occupational therapy, and speech therapy. Surgery in the form of hemispherectomy can be performed for intractable seizures with approximately 85 % success rate.¹⁵ Prognosis is better if the onset of hemiparesis occurs after the age of 2 years without intractable seizures.¹⁶

Conclusion

Dyke-Davidoff-Masson syndrome is a rare cause of cerebral hemiatrophy in children and may sometimes be misdiagnosed. Apart from clinical presentations and risk factors, imaging findings play an important role in making accurate diagnosis as well as help to decide appropriate management of patients for the clinicians.

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Conflict of Interest: None

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