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Case Report

Dyke-Davidoff-Masson syndrome: A case report of an 11-year-old child managed for Erb's Palsy ☆

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ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological anomaly encompassing clinical features of seizures, contralateral hemiparesis, facial asymmetry, and intellectual dysfunction. Radiographic findings include cerebral hemiatrophy and ipsilateral calvarial thickening. We encountered an 11-year-old male who presented with new-onset seizures and a 4-year history of weakness in the abduction of his right arm, previously being managed as Erb's palsy. Brain MRI revealed atrophy of the left cerebral hemisphere with ipsilateral dilated ventricle and osseous thickening, consistent with the congenital form of DDMS. We present this case of an atypical presentation of DDMS.

Introduction

Dyke-Davidoff-Masson syndrome(DDMS) is an uncommon neurological condition characterized by convulsions, contralateral hemiplegia, and mental retardation [1]. It was initially described in 1933 [2]. Radiological findings from magnetic resonance imaging (MRI) and computerized tomography (CT) reveal cerebral hemiatrophy and bony malformations, including skull vault thickening on the affected side and hyper pneumatization of the paranasal sinuses [3]. The age of presentation and clinical features of this disorder depend upon the duration and magnitude of brain injury [4,5]. It is predominantly seen in the pediatric population, especially in males, and is thought to occur due to an insult to the developing brain in-utero or during early childhood, leading to the loss of neurons and hampering normal development [5–7]. Depending upon the age of presentation, it has 2 types: congenital (infantile) and acquired [8]. Being a rare anomaly, physicians may misdiagnose it and confuse it with other neurological disorders. Our case revolves around a patient who was initially treated for Erb's palsy with no signs of recovery and was later definitively diagnosed as a case of DDMS via radiological findings.

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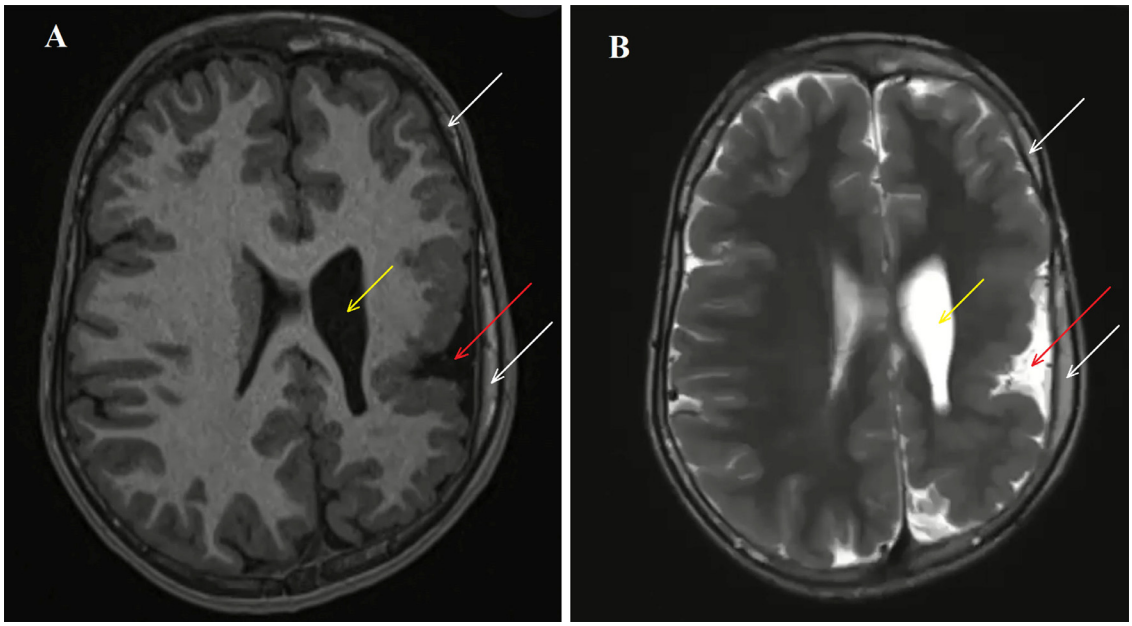

Fig. 1 – Axial MRI images: (A) T1-weighted image and (B) T2-weighted image; demonstrating left cerebral hemiatrophy (red arrows), with left ventricular dilatation (yellow arrows), and calvarial thickening on the affected side (white arrows).

Case report

An 11-year-old boy was referred to our medical center for evaluation and management of convulsions and weakness in his right hand. The child was born at full term via normal spontaneous vaginal delivery to nonconsanguineous parents. There was no maternal illness during pregnancy, and the postpartum period was uneventful. His family history was unremarkable. The child achieved all developmental milestones and there was no history of trauma or any childhood infections.

The child was initially brought to medical attention at age 7 when his father got concerned about weakness in his right hand. Physical and neurological examinations were unremarkable, except for weakness in right shoulder abduction. Subsequently, he started receiving regular physiotherapy. At age 11, he started experiencing focal seizures at variable times throughout the month, leading to poor scholastic performance. Upon evaluation by a pediatric neurologist, the patient was prescribed anticonvulsants and underwent blood tests and MRI brain.

Routine investigations, including total blood count, liver, and renal function tests, serum electrolytes, and blood glucose levels, were within normal limits. Electroencephalography (EEG) evaluation revealed low amplitude with slow background activity in the left cerebral hemisphere.

Brain MRI was conducted and axial (Fig. 1 and Fig. 2), sagittal (Supplementary Material Fig. S1), coronal (Supplementary

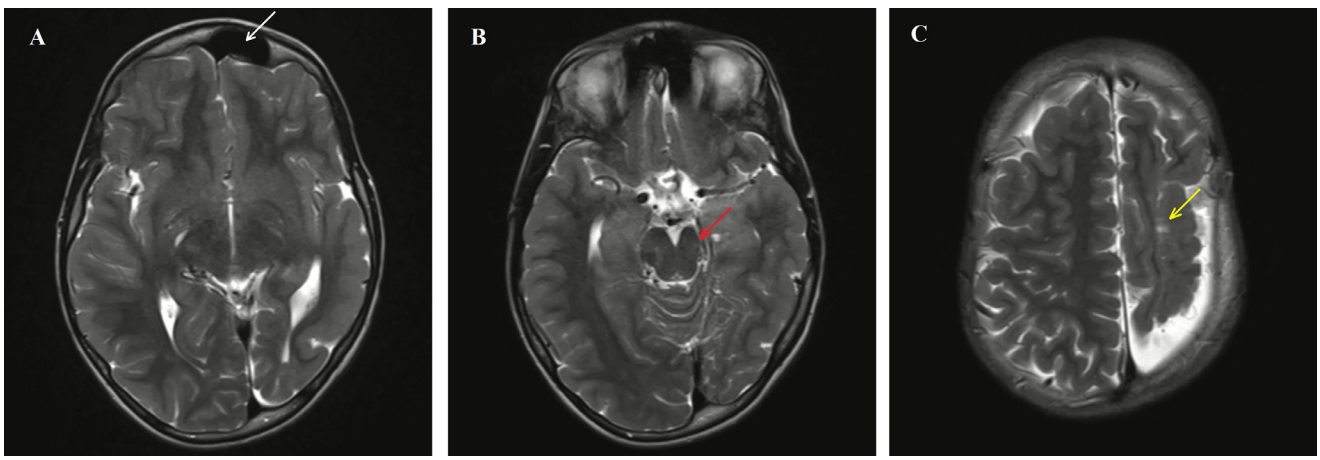

Fig. 2 – Axial T2-weighted MRI images: (A) Demonstrating hyperpneumatization of the frontal sinus on the left side (white arrow). (B) Demonstrating atrophy of the left side of the midbrain (red arrow). (C) Demonstrating abnormal signal in the left corona radiata owing to an underlying pathology (yellow arrow).

Material Fig. S2), and diffusion weighted images were obtained. MRI revealed the left cerebral hemisphere to be smaller in size compared to the right and a relative thickening of the left side of the skull vault. It demonstrated widened left extra-axial subarachnoid spaces and deepening of left cerebral sulci. The left ventricle was dilated, with undulation and irregularity of the left ventricle wall (Fig. 1). There was an enlargement of the left frontal sinus compared to the right (Fig. 2A). Left-sided basal ganglia, brainstem, and cerebellum appeared atrophied with loss of arborization (Fig. 2B). Abnormal signal was seen in the left corona radiata and centrum semiovale as hyperintensity in T2 weighted image and flair with no diffusion restriction (Fig. 2C). Mild thinning of the posterior part of corpus callosum was observed. Findings in the rest of the scan and right cerebral hemisphere were unremarkable.

The history, clinical features, and radiological findings were highly suggestive of DDMS. Since then, the patients' seizures have been well-controlled with anticonvulsants. He has been advised to undergo regular neurological follow-up and physiotherapy.

Discussion

DDMS was first reported by C G Dyke, L M Davidoff, and C B Masson in 1933, with findings of seizures, hemiparesis, facial asymmetry, and mental retardation in a series of nine patients [2]. In addition to these features, radiological findings comprising of an atrophied cerebral hemisphere, dilated ventricle, and calvarial thickening on the affected side are highly suggestive of this atypical neurological abnormality [7]. Other features of this disease include mental retardation and speech and language disorders [8]. Our case presented with the most typical clinical and radiographic features of DDMS including seizures, cerebral hemiatrophy, and thickening of the skull vault.

DDMS can be classified into 2 types: congenital and acquired [9]. The congenital variant occurs as a result of various etiologies like vascular occlusion involving the middle cerebral artery in-utero, infections, or coarctation of the middle aortic arch [9,10]. The symptoms appear at birth or later in life [9,10]. Literature review has also linked it with genetic defects [11]. In acquired form, the main causative factors include trauma, tumor, infection, hemorrhage, ischemia, and prolonged febrile seizures [8]. Congenital type accompanies midline shift towards the affected side, while the sulci prominence replacing the gliotic tissue is not seen, thus differentiating it from the acquired type [8,11]. Singh et al concluded that the ventricular dilation, skull vault thickening, and enlarged frontal sinus of the affected side were highly suggestive of the congenital form of cerebral hemiatrophy [7]. These features corroborate our findings from the MRI. Studies also highlight that the calvarium involvement resulting in bone thickening, reflects cerebral damage during the intrauterine period or before the age of three [12,13]. These changes occur in the bone as a protective mechanism to occupy the space created by the atrophied cerebral hemisphere [11]. The findings in our case were suggestive of previous neonatal or early childhood brain insult, pointing towards the congenital form of the disease. However, our case was atypical as the patient had no significant history pointing towards DDMS, thus proving difficulty in diagnosis. Only later in childhood did the symptoms start appearing, which pointed towards this rare anomaly and discovering its findings through MRI.

According to studies, most of the patients affected belong to the pediatric age group, especially the male gender, with left cerebral hemisphere involvement and a mean age of 11 years; however, it has also been observed in the adult population [14,15]. Rasmussen encephalitis, Sturge-Weber syndrome, Basal ganglia germinoma, Linear Nevus syndrome, Silver-Russell syndrome, and Fishman syndrome are important differential diagnoses of this disease [16,17]. Hence proper history, clinical examination, and radiological investigations can help rule out the differentials.

Magnetic Resonance Imaging(MRI) or Computerized Tomography(CT) of the brain is the gold standard for diagnosing DDMS. MRI is highly valuable in differentiating congenital and acquired forms of DDMS, as it can detect changes in both cerebral hemispheres and highlight bony structures [7]. According to Shetty et al, MRI helps demonstrate gray-white matter loss along with hyperintensities in T2 weighted images and atrophy of basal ganglia of the affected side, which are constant with our study [18]. Other investigations of the brain including, electroencephalography (EEG), Positron Emission Tomography (PET), single-photon emission computed tomography (SPECT), functional magnetic resonance imaging (fMRI) and baseline investigations can help in diagnosis and management.

Patients with DDMS suffer from seizures and hemiparesis. Consequently, the management should focus on anticonvulsants, regular physiotherapy, and speech therapy.

Prognosis depends upon the age, as studies have shown it to be favorable if the onset is after 2 years and seizures are well controlled [19]. For children suffering from recurrent and severe seizures, hemispherectomy is the treatment of choice as it has been shown to have an 85% success rate [20].

Nonetheless, proper detailed history, examination, and appropriate investigations can help diagnose this rare anomaly. Timely diagnosis leads to early management and thus improves the well-being and prognosis of the patient. Prompt use of imaging modality like MRI avoids misdiagnosis of this relatively rare condition.

Patient consent

The authors declare, that the patient's father gave informed consent for his child's participation in this report. The images are anonymized and do not contain any information about the concerned patient.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2024.02.025.

REFERENCES

[1] Ono K, Komai K, Ikeda T. Dyke–Davidoff–Masson syndrome manifested by seizure in late childhood: a case report. J Clin Neurosci 2003;10(3):367–71. doi:10.1016/S0967-5868(03)00011-0.

[2] Dyke CG, Davidoff LM, Masson CB. Cerebral hemiatrophy and homolateral hypertrophy of the skull and sinuses. Surg Gynecol Obstet 1933;57:588–600.

[3] Adebayo PB, Bakare A, Bello MM, Olaewe OD, Wahab KW. Dyke-Davidoff-Masson syndrome in a Nigerian. Epilepsy Behav Case Rep 2016;7:10–12. doi:10.1016/j.ebcr.2016.09.003.

[4] Aguiar PH, Liu CW, Leitão H, Issa F, Lepski G, Figueiredo EG, et al. MR and CT imaging in the Dyke-Davidoff-Masson syndrome. Report of three cases and contribution to pathogenesis and differential diagnosis. Arq Neuropsiquiatr 1998;56(4):803–7. doi:10.1590/s0004-282x1998000500016.

[5] Kumar NV, Gugapriya TS, Guru AT, Kumari SN. Dyke-Davidoff-Masson syndrome. Int J Appl Basic Med Res 2016;6(1):57–9. doi:10.4103/2229-516X.174016.

[6] Abdul Rashid AM, Md Noh MSF. Dyke-Davidoff-Masson syndrome: a case report. BMC Neurol 2018;18(1):76. doi:10.1186/s12883-018-1079-3.

[7] Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson syndrome: classical imaging findings. J Pediatr Neurosci 2010;5(2):124–5. doi:10.4103/1817-1745.76108.

[8] Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. J Neurosci Rural Pract 2012;3(3):411–13. doi:10.4103/0976-3147.102646.

[9] Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy. Clin Imaging 1992;16(2):93–7. doi:10.1016/0899-7071(92)90119-t.

[10] Stred SE, Byrum CJ, Bove EL, Oliphant M. Coarctation of the midaortic arch presenting with monoparesis. Ann Thorac Surg 1986;42(2):210–12. doi:10.1016/s0003-4975(10)60522-x.

[11] Atalar MH, Icagasioglu D, Tas F. Cerebral hemiatrophy (Dyke-Davidoff-Masson syndrome) in childhood: clinicoradiological analysis of 19 cases. Pediatr Int 2007;49(1):70–5. doi:10.1111/j.1442-200X.2007.02299.x.

[12] Arora R, Rani JY. Dyke-Davidoff-Masson syndrome: imaging features with illustration of two cases. Quant Imaging Med Surg 2015;5(3):469–71. doi:10.3978/j.issn.2223-4292.2014.11.17.

[13] Uduma FU, Emejulu JK, Motah M, Okere PC, Ongolo PC, Muna W. Differential diagnoses of cerebral hemiatrophy in childhood: a review of literature with illustrative report of two cases. Glob J Health Sci 2013;5(3):195–207. doi:10.5539/gjhs.v5n3p195.

[14] 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. doi:10.1016/S0899-7071(03)00158-X.

[15] Roy U, Panwar A, Mukherjee A, Biswas D. Adult Presentation of Dyke-Davidoff-Masson syndrome: a case report. Case Rep Neurol 2016;8(1):20–6. doi:10.1159/000443521.

[16] Rao KC. Degenerative diseases and hydrocephalus. In: Lee SH, Rao KC, Zimmerman RA, editors. Cranial MRI and cT. New York: McGraw-Hill; 1999. p. 212–14.

[17] Zilkha A. CT of cerebral hemiatrophy. AJR Am J Roentgenol 1980;135(2):259–62. doi:10.2214/ajr.135.2.259.

[18] Shetty DS, Lakhkar BN, John JR. Dyke-Davidoff-Masson syndrome. Neurol India 2003;51:136.

[19] Pendse NA, Bapna P, Menghani V, Diwan A. Dyke-Davidoff-Masson syndrome. Indian J Pediatr 2004;71:943. doi:10.1007/BF02830843.

[20] Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson syndrome. Indian Pediatr 2008;45(11):927–8.