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Letter to the editor

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Etiologic Insights from Magnetic Resonance Angiography in Dyke–Davidoff–Masson Syndrome Presenting with Aggressive Behavior

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Dyke–Davidoff–Masson syndrome (DDMS) is a rare neurological disorder characterized by cerebral hemiatrophy and associated with diverse clinical manifestations, including recurrent seizures, hemiplegia or hemiparesis, facial asymmetry, and cognitive impairment [1]. Although these manifestations are well described, behavioral and psychiatric features have received less attention in the pediatric literature. In addition, although vascular abnormalities have been implicated in DDMS, the role of vascular imaging in clarifying its etiology remains underexplored.

We report the case of an 11-year-old boy with DDMS who presented to our pediatric neurology clinic with intractable focal epilepsy and aggressive behavioral problems. He was the second child of healthy, nonconsanguineous parents. His medical history was notable for an episode of status epilepticus caused by an untreated intracranial infection at 20 months of age. After this episode, his parents observed right-sided facial asymmetry and a mild hemiparetic gait. Since then, he had experienced recurrent focal seizures characterized by staring spells and left-sided limb jerking. Physical examination revealed a spastic hemiparetic gait, left-sided hyperreflexia, and a positive Babinski sign. Mild left facial weakness was also noted. The seizure semiology was consistent with focal impaired awareness seizures accompanied by left-sided clonic movements.

In addition to his seizure disorder, the patient had substantial behavioral disturbances. His parents reported frequent episodes of anger and irritability, recurrent arguments with adults, rapid loss of temper, and heightened sensitivity to perceived provocation. During these episodes, he sometimes slammed objects and physically hit others when his demands were not met. During the clinic visit, he displayed aggressive behavior, refused to cooperate with the physical examination, and threw objects when asked to perform specific tasks. At 10 years of age, he sustained a fracture of the left olecranon during an aggressive outburst after forcefully striking a door. Oppositional and aggressive behaviors had been present since approximately 7 years of age. A comprehensive psychological and behavioral evaluation was performed by a clinical child psychologist in collaboration with a pediatric consultant in growth and developmental-behavioral pediatrics. Behav-

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ioral assessment included the Conners rating scale, the strengths and difficulties questionnaire, and the oppositional defiant disorder rating scale. Scores on these standardized instruments were within the clinical range for oppositional and conduct-related behaviors. Based on the clinical evaluation and assessment results, the patient met the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5) criteria for oppositional defiant disorder, fulfilling five of the eight criteria; these symptoms had persisted for more than 6 months and were observed during interactions with individuals other than siblings [2]. Academically, the patient had substantial difficulties, frequently performed below grade level, and repeated the sixth grade. Cognitive testing with the Wechsler Intelligence Scale for Children showed a total intelligence quotient (IQ) of 54, verbal IQ of 55, and performance IQ of 61, consistent with moderate intellectual disability.

Brain magnetic resonance imaging (MRI) revealed right cerebral hemiatrophy with ipsilateral ventriculomegaly, calvarial thickening, and hyperpneumatization of the right frontal sinus. In addition, a sharply marginated, amorphous pathological lesion was observed in the right parieto-temporo-occipital region. The lesion was hypointense on T1-weighted and fluid-attenuated inversion recovery images, hyperintense on T2-weighted images, and showed no diffusion restriction, suggesting a chronic infarct. Brain magnetic resonance angiography (MRA) demonstrated hypoplasia of the right middle cerebral artery (MCA) branches, with otherwise normal intracranial arteries. Electroencephalography showed frequent right frontal epileptiform discharges. Management included three antiseizure medications at maximum doses to control seizures, as well as behavioral and occupational therapy for aggressive behavior and intellectual disability. However, seizures persisted, occurring approximately 15 times per month (Fig. 1).

DDMS has traditionally been classified as congenital or acquired according to the timing of cerebral injury. However, distinguishing between these subtypes in clinical practice is often challenging. Congenital forms are usually attributed to prenatal or perinatal vascular insults, whereas acquired cases develop after postnatal brain injury caused by infection, trauma, hypoxia, hemorrhage, or ischemia [1,3]. In the present case, the onset of neurological deficits after an intracranial infection in early childhood suggests an acquired form. However, several neuroimaging findings complicate this classification. The presence of compensatory calvarial thickening and hyperpneumatization of the frontal sinus indicates that hemispheric injury likely occurred during an early developmental period, when skull remodeling remained possible [4,5]. Previous studies have reported that such compensatory changes are most prominent when cerebral injury occurs early in life, before cranial maturation is complete [4,6]. This observation underscores the importance of considering the timing of injury rather than relying solely on a strict congenital–acquired distinction.

A notable imaging finding in this case was unilateral hypoplasia of the right MCA branches on MRA. Vascular abnormalities have occasionally been described in DDMS, although angiographic findings are not routinely reported in many case descriptions. The presence of MCA hypoplasia may indicate early vascular compromise affecting the development of the involved hemisphere. However, the precise relationship between this vascular abnormality and the observed parenchymal injury cannot be determined from a single case. Early vascular disruption may have contributed to hemispheric underdevelopment, although secondary vascular remodeling after cerebral injury cannot be excluded.

Behavioral disturbances in DDMS are less frequently emphasized in the literature and are often overshadowed by more prominent motor deficits and epilepsy. Previous reports have described various psychiatric manifestations, including aggression, irritability, mood disturbances, and psychotic symptoms [6,7]. In this patient, aggressive and oppositional behaviors were persistent and caused substantial functional impairment. A multidisciplinary behavioral assessment by a clinical psychologist and a pediatric consultant in growth and developmental-behavioral pediatrics, together with standardized behavioral instruments, supported the diagnosis of oppositional defiant disorder according to DSM-5 criteria [2]. Although structural abnormalities involving frontal networks may plausibly contribute to behavioral dysregulation, the relationship between hemispheric structural abnormalities and the behavioral phenotype cannot be established from a single case.

The timing of cerebral injury may also influence the clinical manifestations of DDMS. Brain insults that occur before completion of hemispheric and skull maturation, generally within the first 2 years of life, can substantially affect neural network development. Injuries during this period may produce not only focal structural abnormalities but also broader functional reorganization, potentially contributing to long-term cognitive and behavioral sequelae that appear disproportionate to the degree of structural damage observed on imaging [8]. In the present case, unilateral hypoplasia of the right MCA branches was identified on MRA, suggesting a possible vascular contribution to hemispheric development. Although MRA does not provide direct perfusion data, reduced arterial caliber may reflect either primary developmental hypoplasia or secondary vascular remodeling after early cerebral injury. Therefore, the relationship between the vascular abnormality and the observed parenchymal changes cannot be determined from a single case.

Intractable epilepsy represents another major challenge in DDMS and may further exacerbate cognitive and behavioral dys-

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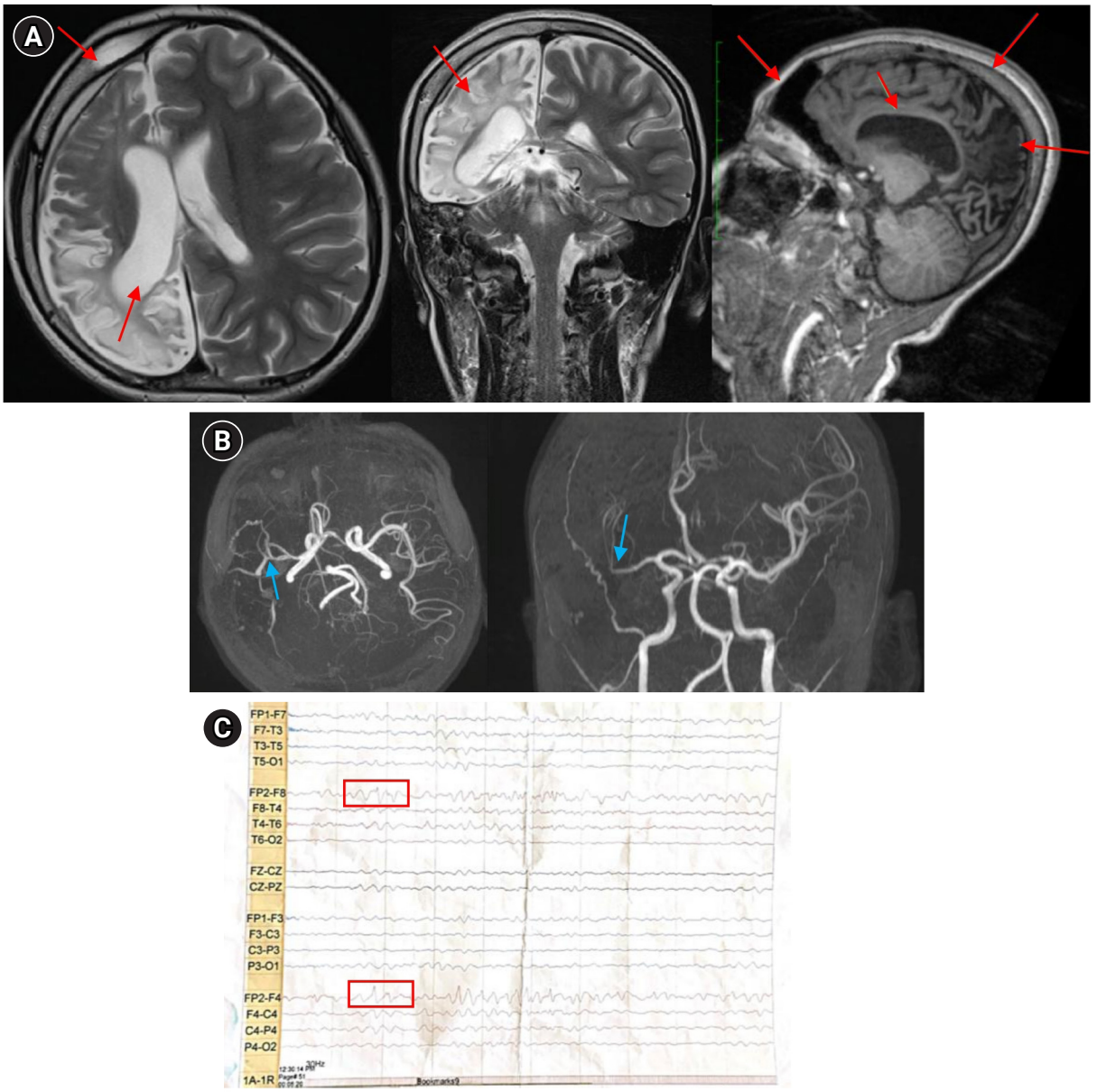

Fig. 1. (A) Brain magnetic resonance imaging revealed right cerebral hemiatrophy with ipsilateral ventriculomegaly, calvarial thickening, hyperpneumatization of the right frontal sinus, and a parenchymal lesion in the right parieto-temporo-occipital region, consistent with chronic infarction. The arrows highlight features of right cerebral hemiatrophy, including ipsilateral ventriculomegaly, associated calvarial/frontal sinus changes, and the right parieto-temporo-occipital parenchymal lesion consistent with chronic infarction. (B) Brain magnetic resonance angiography demonstrated hypoplasia of the right middle cerebral artery branches, with smooth vessel walls and no visible thrombus. The arrows indicate hypoplasia of the right middle cerebral artery branches on magnetic resonance angiography, with no visible thrombus. (C) Electroencephalography showed frequent sharp epileptiform discharges originating from the right frontal region.

function. Although seizures are common, not all patients develop pharmacoresistant epilepsy. Factors associated with intractability include younger age at the time of brain injury, extensive hemispheric involvement, and disruption of epileptic networks [9,10]. In this patient, seizures persisted despite combination therapy, highlighting the potential severity of epileptic manifestations in DDMS. Management of DDMS therefore requires a comprehensive, multidisciplinary approach. Although antiseizure medications remain the cornerstone of epilepsy treatment, additional options, such as the ketogenic diet, neuromodulation, or surgical intervention, may be considered in selected cases. Recognition and treatment of behavioral disturbances through behavior-focused therapy, caregiver education, and psychosocial support may reduce long-term morbidity and family burden.

In summary, this case illustrates the coexistence of aggressive behavioral disturbance, refractory epilepsy, and unilateral MCA hypoplasia in a patient with DDMS. The combined use of MRI and MRA may provide additional insight into vascular and structural abnormalities associated with cerebral hemiatrophy. Recognition of behavioral manifestations may facilitate more comprehensive clinical evaluation and multidisciplinary management of patients with DDMS. Written informed consent by the patients was waived due to a retrospective nature of our study.

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Conflicts of interest

No potential conflict of interest relevant to this article was reported.

Author contribution

Conceptualization: APN and ESH. Formal analysis: APN and DKN. Project administration: APN, DKN, and AT. Visualization: BWI, DKN, AT, and ESH. Writing-original draft: APN and ESH. Writing-review & editing: BWI, DKN, and AT.

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References

1. Rondao MB, Hsu BR, Centeno RS, de Aguiar PH. Dyke-Davidoff-Masson syndrome: main clinical and radiological findings- systematic literature review. Seizure 2023;110:58-68.
2. American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders. 5th ed. Arlington: American Psychiatric Publishing; 2013.
3. Aguiar PH, Liu CW, Leitao 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;56:803-7.
4. Atalar MH, Icagasioglu D, Tas F. Cerebral hemiatrophy (Dyke-Davidoff-Masson syndrome) in childhood: clinicoradiological analysis of 19 cases. Pediatr Int 2007;49:70-5.
5. Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson syndrome: classical imaging findings. J Pediatr Neurosci 2010;5:124-5.
6. Younas A, Saim M, Maqsood H, Younus S, Hassan Raza M. Dyke-Davidoff-Masson syndrome: a case report and review of literature. Cureus 2020;12:e11919.
7. White JH, Rust JB. Davidoff-Dyke-Masson syndrome presenting as childhood schizophrenia. J Autism Dev Disord 1979;9:37-40.
8. Benjamin R, Galuska MA. Middle cerebral artery stroke. In: StatPearls. Treasure Island: StatPearls Publishing; 2026 [cited 2026 May 28]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK556132.
9. Malik P, Garg R, Gulia AK, Kario J. Dyke-Davidoff-Masson syndrome: a rare cause of refractory epilepsy. Iran J Psychiatry 2014;9:42-4.
10. Dutta A, Bose S, Sen K, Pandit N, Sharma S. Refractory seizure in childhood: Dyke-Davidoff-Masson syndrome revisited. Oman Med J 2016;31:304-8.

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