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Case Report

Dyke–Davidoff–Masson Syndrome: A Case with Electroencephalography and Lesion Discordance

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ABSTRACT

Dyke–Davidoff–Masson Syndrome (DDMS) is a rare syndrome characterized by cerebral hemiatrophy, hyperpneumatization of the paranasal sinuses, homolateral skull hyperplasia, seizures that may be associated with mental retardation, and contralateral hemiparesis. Focal onset seizures may be seen in patients. Interictal Electroencephalogram (EEG) recordings of these patients were evaluated as lower amplitude and slow background activity in the affected hemispheres. While most of the cases have lateralized epileptiform disorder compatible with the lesion, cases with continuous discordance in EEG are rarely presented in the literature. Epileptiform discordance occurs infrequently, as in our case; it is crucial to keep this in mind when finding irritative areas, particularly in patients with big lesions. We aimed to discuss a patient who had epileptiform discordance and was diagnosed with DDMS, which is an uncommon condition in the literature, and examine the literature based on this case.

INTRODUCTION

Dyke–Davidoff–Masson syndrome (DDMS) is a rare syndrome characterized by cerebral hemiatrophy, homolateral skull hyperplasia, hyperpneumatization of the paranasal sinuses, seizures that may accompany mental retardation, and contralateral hemiparesis.[1] Nine patients with cerebral hemiatrophy and seizures were reported by Dyke, Davidoff, and Masson in 1933. The frequency of DDMS is unknown.[2] The most common neuroimaging findings are cerebral hemiatrophy (100%), hemicalvarial thickening (71.4%), and hyperpneumatization of the sinuses (71.4%). The etiology of DDMS is unclear; however, it is suspected to be caused by congenital malformations, infection, and cerebrovascular events, as well as birth asphyxia and trauma.[1] Case studies revealed that male gender and left-sided involvement are common.[3] This case report aimed to analyze a patient consulted at our neurology polyclinic with epileptiform discordance, which is rarely reported in the literature, and was diagnosed with DDMS based on clinical findings and neuroradiological imaging.

CASE REPORT

A 40-year-old female patient attended our epilepsy clinic complaining of seizures. The patient's seizures began around the age of 4 years, and the seizures were nonmotor focal onset seizures with impaired awareness and sometimes evolved to bilateral tonic-clonic seizures, according to the description of the patient's relatives. The history of the patient involved preterm birth and subsequent incubator stay. She was diagnosed with congenital hypothyroidism and congenital adrenal hyperplasia. Therefore, she was receiving hydrocortisone treatment. Her seizures occurred about 2–3 times a month at the time of admission. She was prescribed valproic acid 1500 mg/day, levetiracetam 3500 mg/day, and carbamazepine 1000 mg/day. In her neurological examination, there was partial cooperation, loss of the left nasolabial fold, and left hemiparesis. Cranial MR imaging showed generalized volume loss in the right cerebral hemisphere and marked thinning in the cortex and white matter. It was observed that the right frontal sinus was widened by hyperation to posterior, and the diploe distance of the right hemispheric calvarium was thicker compared with the

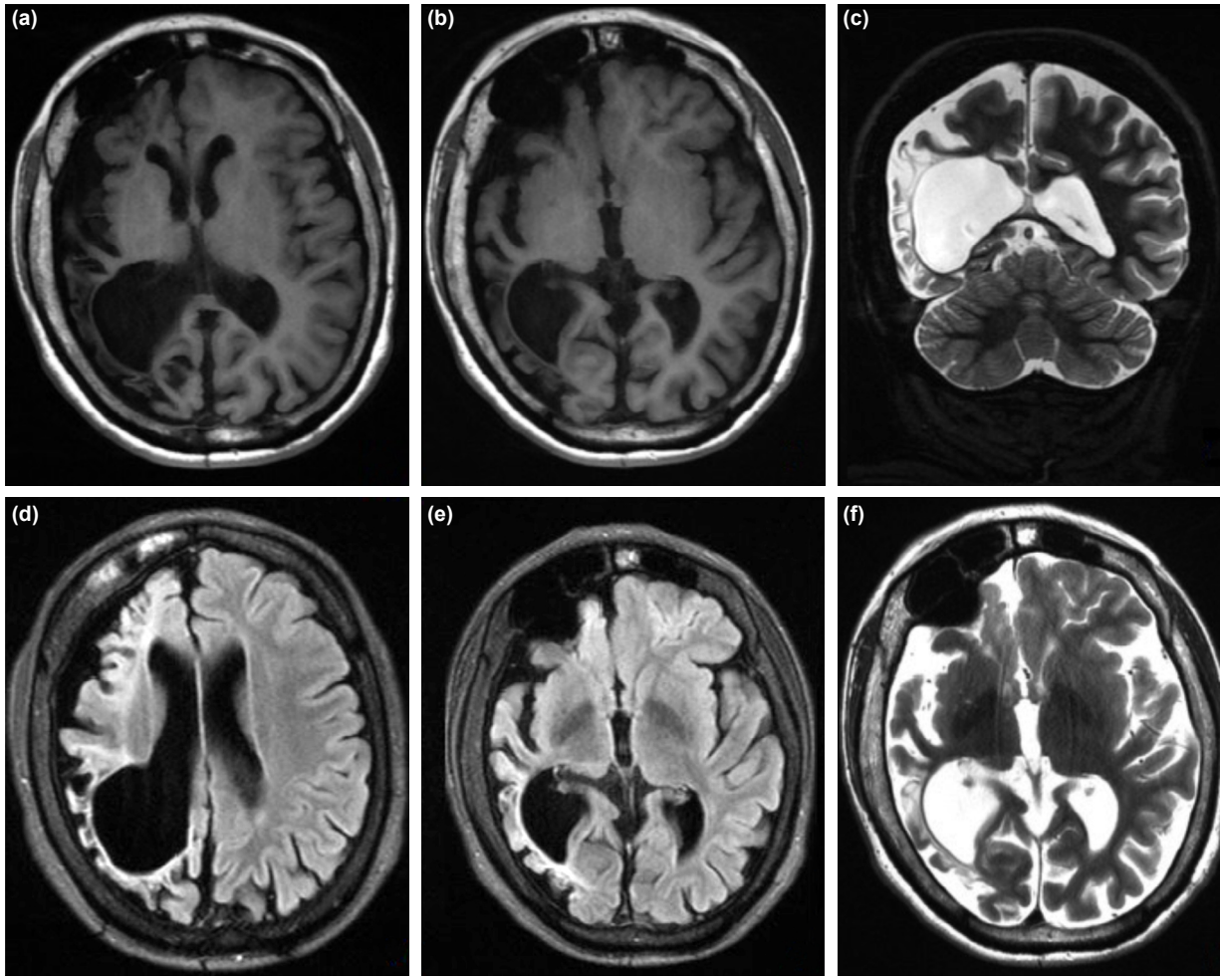

Figure 1. (a, b) Generalized volume loss in the right cerebral hemisphere, cortical thinning on the right, and volume loss in the right white matter on T1-weighted images. **(c)** Cystic enlargement of the right lateral ventricle on T2-weighted coronal image. **(d)** Thickening of the right hemicalvarium compared with the contralateral side on T2 FLAIR sequence. **(e)** Slight reduction in the volume of the basal ganglia and thalamus on the right on T2 FLAIR sequence. **(f)** Posterior enlargement of the right frontal sinus on T2-weighted imaging.

opposite side (Fig. 1). When the patient's clinical and neuroradiological findings were analyzed together, along with her medical history, it was suspected that she had DDMS. Continuous slow-wave (theta) activity in the left hemisphere and epileptiform discharges in the left hemisphere in the form of frequently recurring spike-wave activity with periodic features from time to time were observed in the interictal electroencephalogram (EEG) of the patient. This activity occasionally spreads to the right frontoparietal area (Fig. 2a and 2b). After the examination of the patient, lacosamide was added to her treatment, her seizures were controlled at a dose of 200 mg/day, and the dose of levetiracetam was adjusted to 3000 mg/day. The antiepileptic treatment that the patient is currently using is valproic acid 2×750 mg, levetiracetam 3×1000 mg, carbamazepine 2×500 mg, and lacosamide 2×100 mg. Treatment arrangement and follow-up continue in our epilepsy polyclinic.

DISCUSSION

In the literature, EEG usually shows epileptiform discharges compatible with a lesion; however, some patients may have a discordant and widespread presentation. Lateralization variations can be explained by the extent of hemispheric damage and calvarial thickness.[4] Due to severe neuron damage, there may not be enough neurons left on the ipsilateral side to reveal epileptogenic activity that can be obtained by recording over the scalp. Likewise, if the skull thickness is severely increased, epileptogenic activity may not be recorded due to the increased barrier in recording over the scalp. In a study by Sammaritano et al.,[5] epileptogenic discharges were observed on the contralateral side of the cerebral lesion in the scalp EEG recordings of 3 patients with gross focal cerebral lesions acquired in the early stages of life. In epileptic patients with gross focal lesions, extracranial EEG data should be interpreted with caution. In such cases, invasive monitoring should be considered if required to clarify the lateralization of seizure onset. In a study conducted between 1993 and 2008, all patients followed up in the epilepsy polyclinic with a diagnosis of clinically and neuroradiologically confirmed right hemiatrophy (DDMS) were evaluated

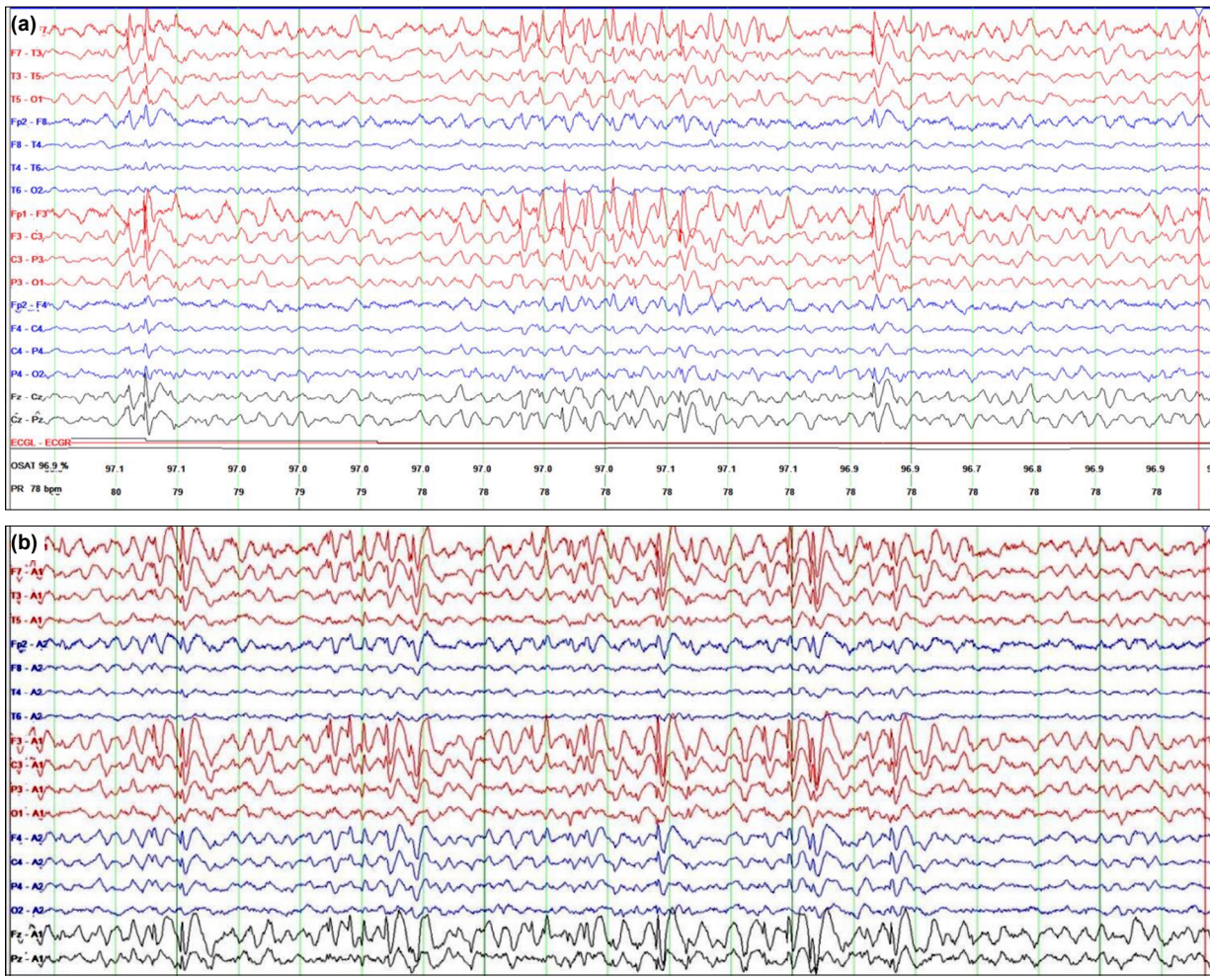

Figure 2. (a) Interictal continuous slow-wave (theta) activity in the left hemisphere and epileptiform discharges in the left hemisphere (longitudinal bipolar montage). **(b)** Interictal epileptiform discharges spreading to the right frontoparietal area (ear reference montage).

as retrospective and 5 patients (4 women) were identified. Interictal EEG recordings of these patients were evaluated as lower amplitude and slow background activity in the affected hemispheres (atrophic hemisphere).[6] According to the current classification, the main seizure types are focal motor in which awareness is not affected, focal seizures in which awareness is affected, and seizures that change from focal to bilateral tonic-clonic.[7]

In a study conducted on 28 DDMS patients, interictal epileptiform discharges were observed in 27 patients. While 26 of the cases were lateralized and compatible with the lesion, continuous discordance was observed in the EEG in 1 patient.[4]

False lateralization of the ictal onset by scalp EEG has also been reported in patients with severe hippocampal sclerosis associated with hemispheric lesions or atrophy.[8] False lateralization of the ictal onset with scalp EEG has been reported in patients with severe hippocampal sclerosis. This recalled that the hippocampus was severely damaged ("burnt-out hippocampus") and not enough neocortical neurons to generate the visible discharge on the scalp EEG.[9] An example of rare false lateralization is that ictal and interictal EEG abnormalities showed paradoxical lateralization to the false hemisphere or bilateral abnormalities in 6 children with cerebral palsy and unilateral hemispheric encephaloclastic lesions evaluated for epilepsy surgery.[10] The lack of further evaluation such as SPECT and PET-CT to show the detailed neuronal loss in our patient is the limitation of this case report.

CONCLUSION

Epileptiform discordance, or false lateralization, occurs infrequently, as in our case; it is crucial to keep this in mind when finding irritative areas and analyzing EEG recordings, particularly in patients with big lesions.

Informed Consent

Written informed consent was obtained from the patient parents for the publication of the case report and the accompanying images.

Peer-review

Internally peer-reviewed.

Authorship Contributions

Concept: N.H.Y., B.Ç., G.K.; Design: N.H.Y., B.Ç., G.K.; Supervision: G.K.; Fundings: N.H.Y., B.Ç., G.K.; Materials: N.H.Y., B.Ç., G.K.; Data: N.H.Y., B.Ç., G.K.; Analysis: N.H.Y., B.Ç., G.K.; Literature search: N.H.Y., B.Ç., G.K.; Writing: N.H.Y., B.Ç., G.K.; Critical revision: N.H.Y., B.Ç., G.K.

Conflict of Interest

None declared.

REFERENCES

1. Diestro JDB, Dorotan MKC, Camacho AC, Perez-Gosiengfiao KT, Cabral-Lim LI. Clinical spectrum of Dyke-Davidoff-Masson syndrome in the adult: an atypical presentation and review of literature. BMJ Case Rep 2018;2018:bcr2018224170. [CrossRef]
2. Abdul Rashid AM, Md Noh MSF. Dyke-Davidoff-Masson syndrome: a case report. BMC Neurol 2018;18:76. [CrossRef]
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;28:163–5.
4. Bhushan B, Kasundra G, Shubhakaran K, Guruprashad Sp, Basavaraj B, Bhargava A. Dyke-Davidoff-Masson syndrome: A study of clinicoradiological variability in hemiplegia, hemiatrophy and epilepsy patients. CHRISMED Journal of Health and Research 2014;1:187–93. [CrossRef]
5. Sammaritano M, de Lotbinière A, Andermann F, Olivier A, Gloor P, Quesney LF. False lateralization by surface EEG of seizure onset in patients with temporal lobe epilepsy and gross focal cerebral lesions. Ann Neurol 1987;21:361–9. [CrossRef]
6. Demirtas-Tatlidede A, Yalcin AD, Uysal E, Forta H. Right cerebral hemiatrophy: neurocognitive and electroclinical features. Epilepsy Behav 2010;17:536–40. [CrossRef]
7. Fisher RS, Cross JH, French JA, Higurashi N, Hirsch E, Jansen FE, et al. Operational classification of seizure types by the International League Against Epilepsy: Position Paper of the ILAE Commission for Classification and Terminology. Epilepsia 2017;58:522–30.
8. Adamolekun B, Afra P, Boop FA. False lateralization of seizure onset by scalp EEG in neocortical temporal lobe epilepsy. Seizure 2011;20:494–9. [CrossRef]
9. Mintzer S, Cendes F, Soss J, Andermann F, Engel J, Dubeau F, et al. Unilateral hippocampal sclerosis with contralateral temporal scalp ictal onset. Epilepsia 2004;45:792–802. [CrossRef]
10. Nagarajan L, Ghosh S, Palumbo L, Lee S, Shipman P, Dyke J. Discordant electroencephalogram epileptiform activity and hemispherectomy in children with refractory epilepsy and encephaloclastic lesions: a case series. Dev Med Child Neurol 2022;64:387–94. [CrossRef]

Dyke-Davidoff-Masson Sendromu: Elektroensefalografi ve Lezyon Diskordansı İzlenen Olgu

Dyke-Davidoff-Masson sendromu (DDMS) serebral hemiatrofi, paranazal sinüslerin hiperpnömatizasyonu, homolateral kafatası hiperplazisi, mental retardasyonla birliktelik gösterebilen nöbetler ve kontralateral hemiparezi ile karakterize nadir bir sendromdur. Hastalarda fokal başlangıçlı nöbetler görülebilmektedir. Bu hastaların interiktal EEG kayıtları, etkilenen hemisferlerde daha düşük amplitüd ve yavaş zemin aktivitesi şeklinde değerlendirilmiştir. Olguların çoğunda lateralize, lezyonla uyumlu epileptiform bozukluk görülürken litetatürde nadir olarak EEG'de sürekli diskordans izlenen olgular sunulmuştur. Epileptiform diskordans veya diğer bir deyişle yanlış lateralizasyon bizim olgumuzda da olduğu gibi nadir de olsa görülebilir; irritatif alan tespiti yaparken özellikle geniş lezyonu olabilecek hastalarda bu durumunun akılda tutulması önem teşkil eder. Dyke-Davidoff-Masson sendromu tanısı alan ve literatürde nadir olarak bildirilen epileptiform diskordansı olan bir hastayı tartışmayı ve bu olgudan yola çıkarak litetatürü gözden geçirmeyi amaçladık.

Anahtar Sözcükler: Epileptiform diskordans; Dyke-Davidoff-Masson sendromu; yanlış lateralizasyon.