Dyke-Davidoff-Masson Syndrome: A Case Report

Dyke-Davidoff-Masson Sendromu: Bir Olgu Sunumu

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ABSTRACT

Dyke–Davidoff–Masson Syndrome (DDMS) is a rare clinical condition characterized by seizures, facial asymmetry, contralateral hemiplegia or hemiparesis, mental retardation, sensorineural hearing loss and behavioral changes. The clinical features are variable and depend on the extent of brain injury in fetal or early childhood period. The typical radiological features are cerebral hemiatrophy with ipsilateral compensatory thickening of the skull and hypertrophy of paranasal sinuses. 22-year old woman admitted to physical medicine and rehabilitation polyclinic due to weakness in her left side of the body. She had history of seizures and was taking medications regularly. She had fascial asymmetry. Neurological evaluation revealed Brunnstrom stages for upper extremity, hand and lower extremity as 5, 3, 5 respectively. There was increased tone in flexors of wrist and phalanx showing Modified Ashworth Scale (MAS) grade 2 spasticity. EEG showed generalized spike-wave activity. IQ test revealed that she had borderline intellectual functioning. Cranial magnetic resonance imaging revealed hemiatrophy of right cerebral hemisphere, dilatation of the right lateral ventricle and widening of ipsilateral sulci. The patient was diagnosed with DDMS based on typical clinical and imaging features. The syndrome was first described in 1933 by Dyke et al. Both congenital and acquired factors are involved in the etiology. Although it is a rare syndrome, we presented the case report to emphasize the importance of recognizing DDMS in differential diagnosis of hemiparesis and epilepsy.

Keywords: Hemiparesis, hemiatrophy, epilepsy, fascial asymmetry, rehabilitation

ÖZET

Dyke–Davidoff–Masson Sendromu (DDMS) nöbetler, fasiyal asimetri, kontralateral hemipleji veya hemiparezi, mental retardasyon, sensorinöral işitme kaybı ve davranış değişiklikleri ile karakterize nadir görülen bir klinik durumdur. Klinik özellikler değişkendir ve fetal veya erken çocukluk döneminde gelişen beyin hasarının derecesine bağlı olarak değişir. Tipik radyolojik özellikleri serebral hemiatrofi ile birlikte ipsilateral kompansatuvar kafatası kalınlaşması ve paranazal sinüslerin hipertrofisidir. 22 yaşındaki bayan hasta vücut sol yarısında güçsüzlük nedeniyle fiziksel tıp ve rehabilitasyon polikliniğine başvurdu. Nöbet hikayesi mevcuttu ve ilaçlarını düzenli olarak almaktaydı. Fasiyal asimetrisi vardı. Nörolojik değerlendirmesinde üst ekstremite, el ve alt ekstremite Brunnstrom evreleri sırasıyla 5,3 ve 5 idi. El bilek ve falanks fleksörlerinde Modifiye Ashworth Skalasına (MAS) göre evre 2 spastisiteyi gösteren artan tonus mevcuttu. EEG'de generalize diken dalga aktivitesi görülmekteydi. IQ testi sınırda mental kapasite ile uyumlu bulundu. Kranial manyetik rezonans görüntülemede sağ serebral hemisferde atrofi, sağ lateral ventrikülde dilatasyon ve ipsilateral sulkuslarda genişleme saptandı. Hastada tipik klinik ve görüntüleme özelliklerine dayanılarak DDMS tanısı kondu. Bu hastalık tablosu ilk defa Dyke ve arkadaşları tarafından 1933 yılında tanımlanmıştır. Etiyolojide hem doğumsal ve edinsel faktörler rol alır. Her ne kadar nadir görülen bir sendrom olsa da hemiparezi ve epilepsi ayırıcı tanısında DDMS tanımanın önemini vurgulamak için bu olgu sunumunu yaptık.

Anahtar sözcükler: Hemiparezi, hemiatrofi, epilepsi, fasiyal asimetri, rehabilitasyon

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Introduction

Dyke–Davidoff–Masson Syndrome (DDMS) is a rare clinical condition characterized by seizures, facial asymmetry, contralateral hemiplegia or hemiparesis, mental retardation, sensorineural hearing loss and behavioral changes (1). The clinical features are variable and depend on the extent of brain injury in fetal or early childhood period. The typical radiological features are cerebral hemiatrophy with ipsilateral compensatory thickening of the skull, dilatation of the lateral ventricle, sulci and cisternal spaces, and hypertrophy of paranasal sinuses (2). Hereby we present a case of DDMS with typical clinical and imaging features and briefly discuss the literature.

Case Report

22-year old woman admitted to physical medicine and rehabilitation polyclinic due to weakness in her left side of the body. She had history of seizures and was taking medications regularly. She could not achieve the developmental milestones as compared to healthy child of her age. She had fascial asymmetry. Neurological evaluation revealed Brunnstrom stages for upper extremity, hand and lower extremity as 5, 3, 5 respectively. There was increased tone in flexors of wrist and phalanx showing Modified Ashworth Scale (MAS) grade 2 spasticity. Vision and hearing were normal and cranial nerves were intact. Electroencephalography (EEG) showed right fronto-centro-temporal spike and sharp wave discharge activity which shows intermittently generalized sharp-wave epileptiform discharge. IQ test revealed that she had borderline intellectual functioning. Cranial magnetic resonance imaging revealed hemiatrophy of right cerebral hemisphere, dilatation of the right lateral ventricle, widening of ipsilateral cortical sulci, and Sylvian fissure (Figure 1). There was unilateral thickening of calvarium and asymmetric widening of right frontal sinus. The patient was diagnosed with DDMS based on typical clinical and imaging features. Range of motion exercises, progressive-resistive exercises of the upper and lower limbs, balance, and gait training have been shown to improve the functional capacity and quality of life of the patient.

Discussion

In 1933, Dyke, Davidoff, and Masson described the plain skull radiographic and pneumatoencephalographic findings in a series of nine patients who presented with hemiparesis, facial-asymmetry, seizures, and mental retardation (1). The plain skull radiographic changes were thickening of calvarium, dilatation of ipsilateral frontal and ethmoid sinuses, and elevation of the greater wing of sphenoid and petrous ridge. Both sexes and any of the hemisphere may be affected, but male gender and left side involvement are more common (3).

Congenital and acquired factors are involved in the etiology. The congenital type results from various etiologies such as infections, vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies, and coarctation of the midaortic arch. The main causes of acquired type are trauma, tumor, infection, ischemia, hemorrhage, and prolonged febrile seizure (2,4,5). Age of presentation depends on time of insult. In our case, the findings of right cerebral hemiatrophy with enlarged cortical sulci reflect an onset of brain insult after the completion of sulci formation.

Differential diagnoses to be considered in a patient with cerebral hemiatrophy are, Sturge Weber syndrome, some brain tumors, Silver-Russel syndrome, linear nevus syndrome, and Rasmussen encephalitis. A proper history, clinical examination, and radiologic findings provide the correct diagnosis. In the present case report, the clinical features, radiologic and electroencephalographic characteristics of the patient were compatible with DDMS.

Treatment of DDMS is symptomatic. Even though a specific program for management of the disease is lacking, there is the indication for therapy including anticonvulsant drugs and surgery in specific cases (6). DDMS rehabilitation protocol is based on motor recovery therapies that facilitate neural plasticity to compensate for functional loss. Clinical approach to hemiparesis is the same given to those recovering from stroke or brain injury. Treatment is focused on improving motor abilities, allowing the patient to better manage their activities of daily living. Physical therapy, occupational therapy and speech therapy play significant role in the long term management of the patients.

Conclusion

Although DDMS is a rare syndrome, we presented the case report to emphasize the importance of recognizing DDMS in differential diagnosis of hemiparesis and epilepsy.

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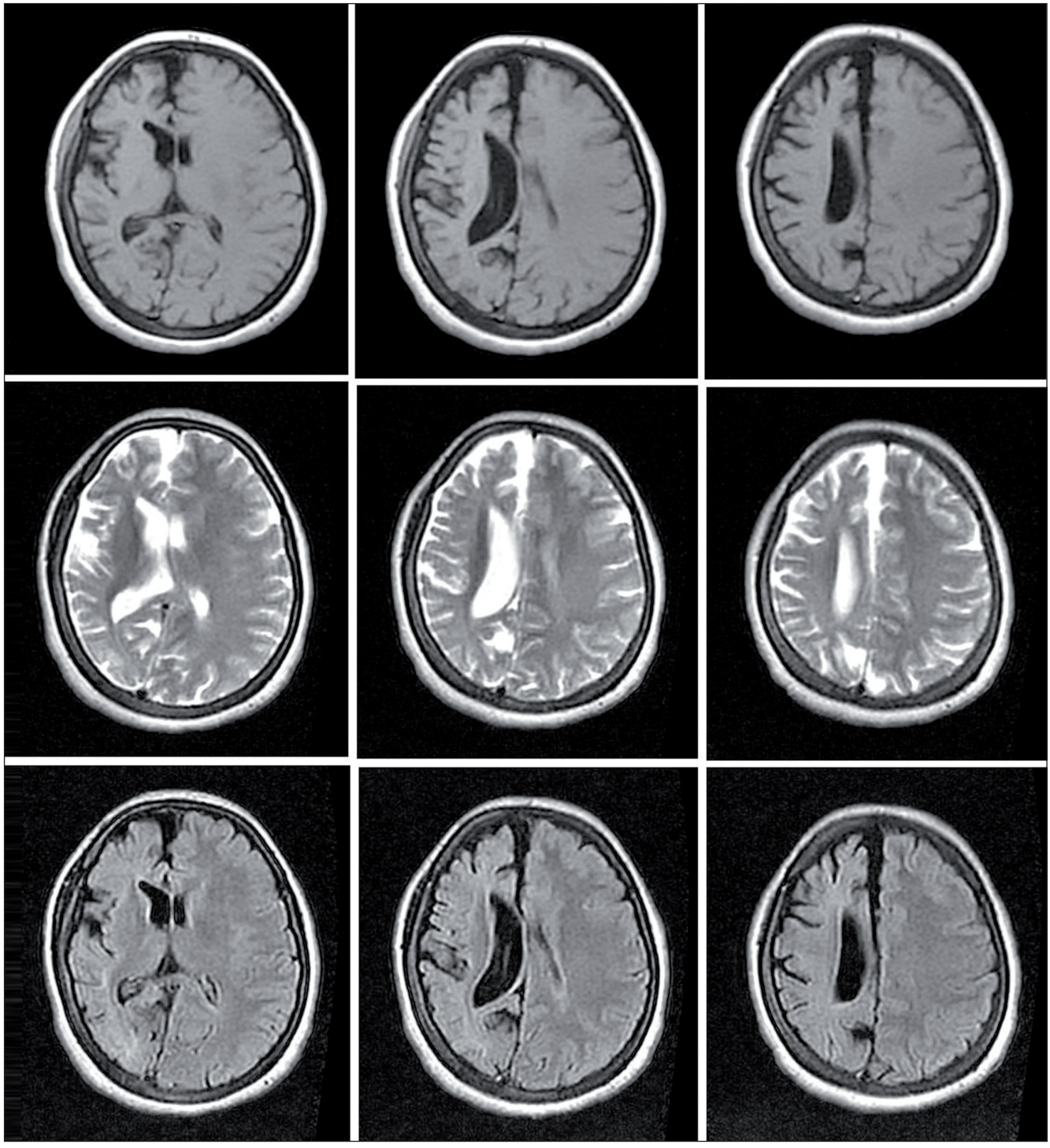

Figure 1. **T1-weighted axial, T2-weighted axial and axial FLAIR sequences show hemiatrophy of right cerebral hemisphere, dilatation of the right lateral ventricle, widening of ipsilateral cortical sulci, and unilateral thickening of calvarium.**

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