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CASE REPORT OPEN ACCESS

DYKE-DAVIDOFF-MASSON SYNDROME: MYOCLONIC SEIZURES AND HEMIHYPERTROPHY IN LATE CHILDHOOD: A CASE REPORT

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ABSTRACT

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare syndrome characterized with specific clinical and radiological findings due to involvement of the developing brain with cerebral hemiatrophy of one hemisphere. The syndrome was first described from Dyke, Davidoff and Masson in 1933 in a series of nine patients. Syndrome has two forms, congenital and acquired forms and etiological factors vary due to involvement of the brain. Most common clinical symptom are focal or secondary generalized seizures. Hemiparesis, facial asymmetry, intellectual disability, mental retardation, and hemihypertrophy also seen in clinical process. In magnetic resonance imaging (MRI) DDMS has unique radiological findings. Seizures are commonly refractory to treatment and aim of the treatment is to control seizures and improve mental and intellectual capabilities. Prognosis is good when clinical findings occur after two years old.

Keywords: Dyke-Davidoff-Masson Syndrome, focal seizures, childhood

Introduction

DDMS is a rare syndrome characterized with specific clinical and radiological findings due to involvement of the developing brain with cerebral hemiatrophy or hipoplasia of one hemisphere and with or without compensatory ipsilateral calvarial changes.[1] The syndrome was first described from Dyke, Davidoff and Masson in 1933 in a series of nine patients.[2] The syndrome has clinical manifestations including contralateral hemiparesis, facial asymmetry, intellectual disability, mental retardation and epilepsy while radiological findings are cerebral hemiatrophy, ipsilateral calvarial thickening, and hyperpneumatization of the paranasal sinuses.[1] Underlying etiological causes usually cannot be revealed. However, due to middle cerebral artery (MCA) damage in prenatal or early postnatal period has considered as main pathophysiological mechanism of cerebral atrophy.[1] During the course of the syndrome epileptic seizures are usually resistant to treatment and may present with status epilepticus.[3] We presented a 12 years old male patient who has focal myoclonic seizures and diagnosed as DDMS in our clinic.

Case Report

A 12 year old male patient presented to our clinic with complaints of involuntary movements of his right arm and rigth eye for about 3 months. His prenatal and natal history was normal, with a normal delivery. Postnatal history and familial histories were normal. When he was 4 years old, he was diagnosed with right hemiparesis in the orthopedics clinic, where he was admitted with complaints of gait disturbance and limping, which was noticed by his family. His symptoms completely regressed after long term physiotherapy. In physical examination he was mild overweight (BMI=29), there was facial asymmetry, myoclonic jerks in the right arm, and distinct horizontal nystagmus in right eye. There was mild hemihypertrophy and frust weakness at the right upper and lower extremities. However, no spasticity was detected and reflex examination was normal. Mild mental retardation, tendency to self damage, and behavioural problems revealed in psychiatric examination and WISC-R test. Electroencephalogram (EEG) revealed spikes with high frequency, and low amplitude in the left frontotemporal regions, with fase reversal at F7 electrode (Figure 1).

MRI findings consistent with smaller left hemicranium, a large hemiatrophy in the left middle cerebral arter region, and thickened left calvarial bones (Figure 2).

The patient was diagnosed as DDMS with his clinical and radiological findings. He was seizure free after 4 weeks later after initializing levetiracetam treatment and no seizures seen in long term follow up. Behavioral and mental treatment in the pediatric psychiatry clinic continues with positive progress.

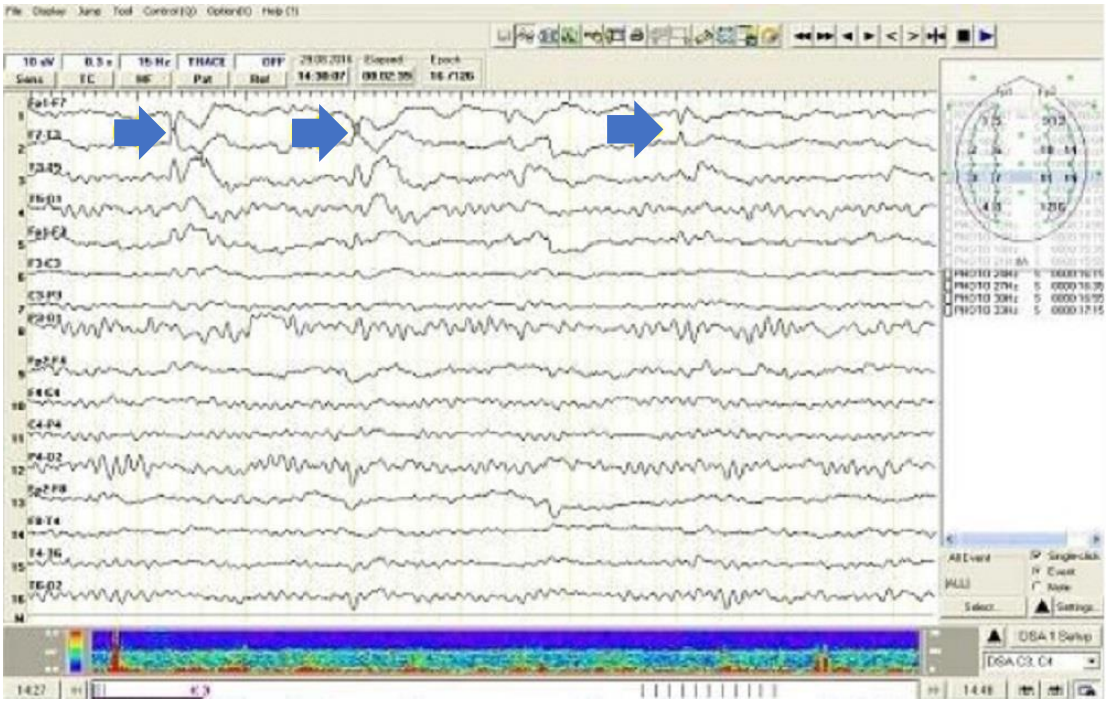

Figure 1. Left fronto-temporal spikes with fase reversal at F7 electrode (blue arrows).

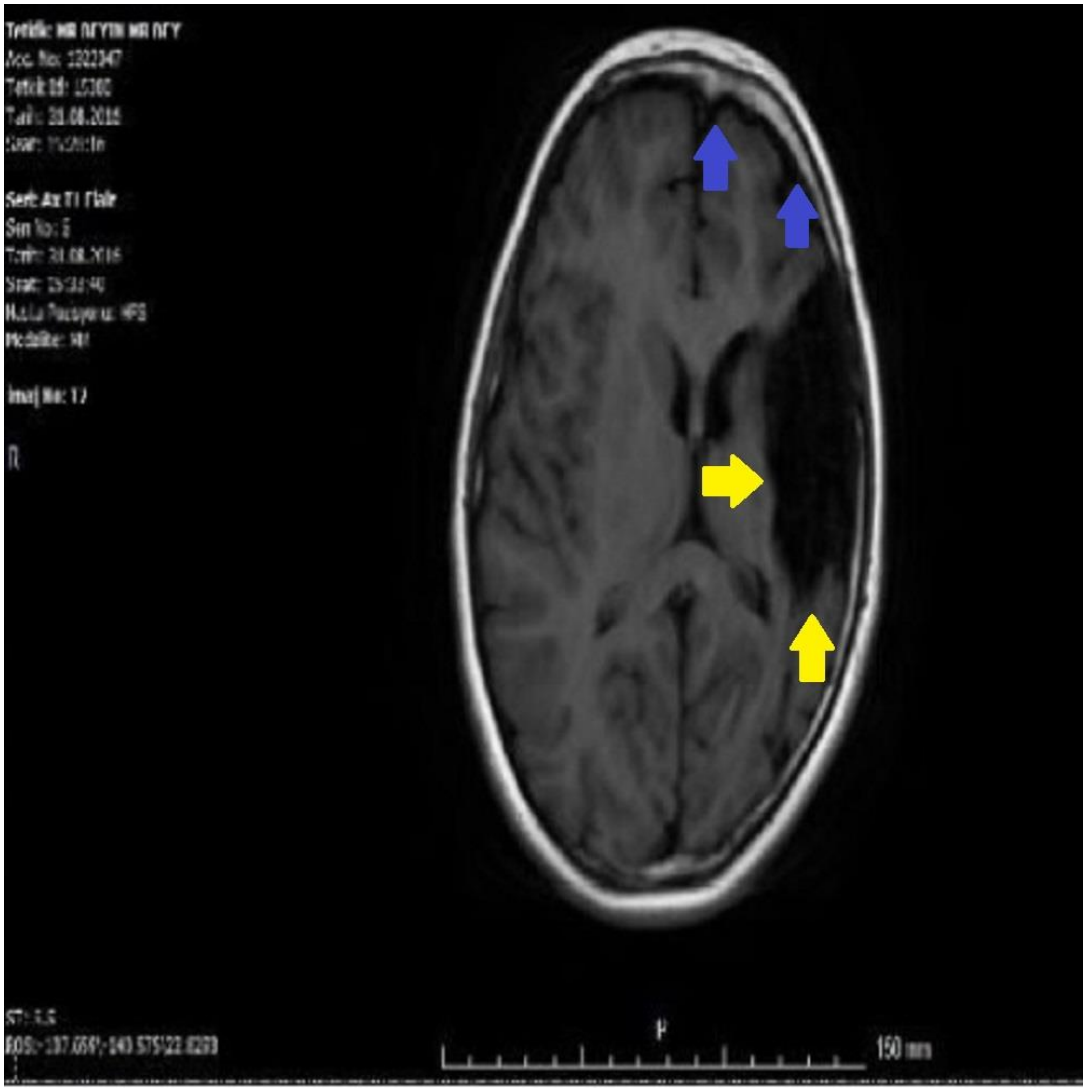

Figure 2. Left cranial hemiatrophy (yellow arrows) and left calvarial thickness (blue arrows) at T1 weightened axial MRI.

Discussion

DDMS is characterized with cerebral hemiatrophy or hipoplasia of one hemisphere with compensatory changes such as increased ipsilateral calvarial thickness and enlargement of frontal or/and ethmoidal sinuses.[4] Cause cerebral atrophy frequently have seen in the MCA region, mainly underlying pathophysiology is is thought to be secondary to MCA injury.[1] Depending on the time of involvement of the brain, there are two types of syndrome: congenital and acquired forms. Intrauterine or early postpartum cranial infections, perinatal hypoxia and congenital vascular malformations are the most common etiologies of congenital form. While head trauma, intracranial hemorrhage, tumor formations, vasculer anomalies, and cranial infections plays role in etiology of acquired form.[5] There are rare conditions such as DDMS seconder to malarial infection.[6] No genetic mutation has been demonstrated for DDMS. Our patient's prenatal and delivery history was normal. there was no history of hypoxia or trauma. No clues to common congenital or acquired etiologies were detected in laboratory and imaging examinations.

The severity of clinical and radiologic findings of the syndrome vary according to the time of brain involvement.[1] Depending on the early involvement of the cranial structures in congenital form compensatory sulcus erosion, thickening of the ipsilateral calvarial bones, expansion of ipsilateral paranasal sinuses, and petrous ringe increasing occurs usually while prominent sulcus, and compensatory bone structure changes are less common in acquired form.[5,7] Considering the radiological and clinical findings such as facial asymmetry and hemihipertrophy of our patient, we thought he was compatible with congenital form. Since there is not a natal or postnatal known factor to explain the syndrome we predict that onset of involvement began in prenatal period with unknown etiology.

Although the syndrome affects both male and female and both side of the brain, male gender and left hemisphere involvement is more common.[7] The most common clinical symptom of DDMS are focal or secondary generalized seizures. Seizures are usually resistant to treatment and may present with status epilepticus.[3] But prognosis is good when the hemiparesis and seizures occur after 2 years old.[4] Other clinical manifestations includes facial and limb asymmetry, hemiparesis, hemiplegia, speech and mental retardation.[3] Our patient's seizures started at a late childhood and response to treatment was excellent. We thougt that parents of could not recognize hemiparesis and behaviour problems of patient in early childhood. And we could not reach the imaging findings of hemiparesis attack period.

Gold standart for diagnosis is cranial MRI and clinical findings. In differantial diagnosis conditions with cerebral atrophy and/or hemihypertrophy such as Sturge Weber Syndrome, Silwer Russel Syndrome, Rasmussen Encephalitis, Linear Nevus Syndrome, Hemiconvulsion-Hemiplegia-Epilepsy (HHE) can be considered, but differential diagnosis can be ease by both clinic and radiological findings.[1]

Treatment is symptomatic. The aim of the treatment is prevent seizures, managing hemiparesis and hemiplegia. Mental retardation and speech disorders must have supported by rehabilitation. Surgical treatment approaches, like hemispherectomy for treatment of drug resistance epilepsy cases may be considered.[8]

Conclusion

DDMS is a rare condition with clinical manifestasions such as seizures, hemiparesis and mental retardation due to cerebral hemiatrophy. Etiological factors vary in a large range of spectrum but radiological findings unique and easy to diagnose. DDMS should be kept in mind in patients which have focal seizures, focal neurological deficits and mental retardation.

Acknowledgement

None.

Conflict of Interest

The authors declare no conflict of interest.

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