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Case Report

Dyke-Davidoff-Masson Syndrome Following Head Trauma: Clinical and Radiologic Findings

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Abstract

Dyke-Davidoff-Masson syndrome (DDM) is an unusual condition characterized by facial asymmetry, hemiparesis, mental retardation, learning disabilities, sensorineural hearing loss, psychiatric disorders, and epilepsy. Some of the typical radiologic findings related to the condition are cerebral hemiatrophy, unilateral thickening of the skull, hyperaeration of frontal sinuses. In this article, we present a case of DDMS acquired after head trauma at the age of 28. Our aim is to point out some of the clinical and radiological diagnostic criteria of acquired DDMS.

Keywords: Epilepsy, hemiatrophy, hemiparesis, mental retardation, Dyke-Davidoff-Masson syndrome, acquired

INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) is a very rare condition that is congenital or acquired.[1,2] Mental retardation, hemiparesis, learning disabilities, and focal or generalized epileptic seizures, sensorineural hearing loss, and psychiatric disorders are the most remarkable clinical findings. Radiologic findings are hemicerebral atrophy or hypoplasia along with ipsilateral skull hypertrophy, facial asymmetry, hyperaeration of the mastoid cells, and frontal sinuses.[3] These classical criteria are only observed if the condition occurs before 3 years of age. Male patients were reported to be slightly the majority of cases.[4]

CASE PRESENTATION

Our patient is a 28-year-old female with post-traumatic left hemiparesis and an intellectual disability. She consulted us at our hospital emergency department with partial motor seizures on the left side, while having no loss of consciousness. The neurosurgery clinic was consulted due to the findings in her radiological examinations. In her neurological examination, the patient was evaluated as being in a post-seizure period, and no other pathology was detected except for left hemiparesis with 4/5 muscle strength and mental retardation. Following the observation period, she was discharged after being given anti-seizure treatment. There is no notable finding in the patient's anamnesis and birth history. Her parents reported that she had normal development until the age of 10. It was later revealed that she had had a head trauma after falling at the age of 10, and was treated in the intensive care unit of the hospital, where she had been admitted for 3 weeks, and then discharged with anti-seizure medication. No documentation could be obtained from this period. During outpatient check-ups, focal motor seizures were observed in the left arm twice a month without loss of consciousness. A 1.5 T magnetic resonance imaging (MRI) unit (Signa HDxt; General Electric) was used with a head coil. Based on MRI, transverse images showed right cerebral hemiatrophy with ipsilateral enlarged lateral ventricle, gliosis, and thickened diploic space (Figure 1). Informed consent was obtained from the patient's parents.

DISCUSSION

DDMS was initially described in 1933 by Dyke et al.[1] Two types of DDMS are mentioned, congenital and acquired.[1,2] Intrauterine vascular occlusion may occur in cases of the congenital type of DDMS and the symptoms begin at birth or immediately afterwards. The central nervous system damage due to trauma, ischemic and hemorrhagic conditions, and infection may be playing a role in the acquired type of DDMS.[5] The case presented is the acquired type of DDMS that developed after trauma at the age of 10.

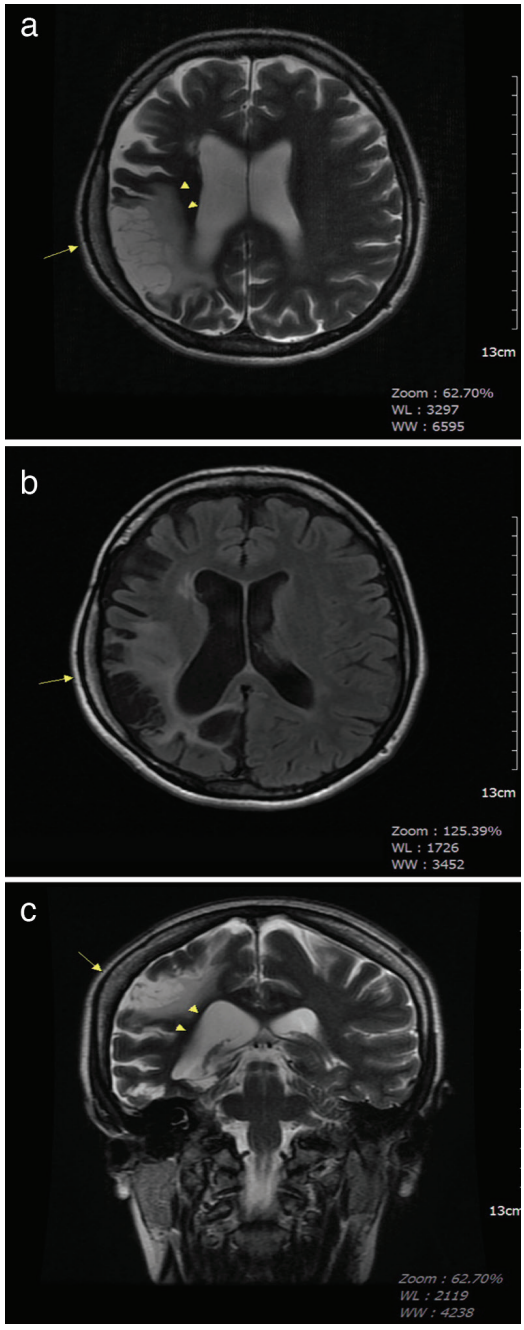

Figure 1. a) T2-W FSE and b) FLAIR supraventricular transverse images show that right cerebral hemiatrophy with enlarged sulcal space, right lateral ventricle, gliosis (arrow head), and also ipsilateral thickening of the calvarium (long arrow). c) Coronal T2W image shows right cerebral hemiatrophy with enlarged sulcal space and right lateral ventricle (arrow head), and also thickening of the calvarium (long arrow)

MAIN POINTS

- Dyke-Davidoff-Masson syndrome is a rare entity characterized by hemi cerebral atrophy/hypoplasia secondary to brain insult in fetal or early childhood period.
- Classical radiological findings and hyperpneumatization of the frontal sinuses are not always seen if an insult to the brain occurs older ages.
- We believe that calvarial thickening and hyperpneumatization of the sinuses replace brain parenchyma.

The clinical features include seizures, facial deformity, contralateral hemiparesis, and intellectual disability. The radiological findings typically demonstrate fascial deformity, enlargement of frontal sinuses, and mastoid cells on craniography. Brain computed tomography and MRI showed cerebral hemiatrophy, ipsilateral dilated lateral ventricles, thickening of the skull vault, elevation of the petrous back, and ipsilateral falcine displacement.[3] The clinical features of the case included seizures, contralateral hemiparesis, and intellectual disability. Radiological examinations revealed right cerebral hemiatrophy with ipsilateral, enlarged lateral ventricle, gliosis, and thickened diploic space. There were no facial deformities, enlargement of the frontal sinuses, and the mastoid cells in the classic triad. However, with its clinical and radiological findings, the case we present can be defined as acquired type DDMS.

The differential diagnosis includes hemimegalencephaly, Sturge-Weber syndrome, and Rasmussen encephalitis (RE). Sturge-Weber syndrome can also be associated with DDMS. RE tends not to have hyperosteosis. The clinical features of the case included seizures, contralateral hemiparesis and mental retardation. Radiological examinations revealed right cerebral hemiatrophy with ipsilateral enlarged lateral ventricle, gliosis, and thickened diploic space. There were no facial deformities, enlargement of frontal sinuses and mastoid cells in the classic triad. However, the case we present can be defined as acquired type DDMS with clinical and radiological differential diagnosis findings such as seizures, contralateral hemiparesis, intellectual disability, right cerebral hemiatrophy with ipsilateral enlarged lateral ventricle, and thickened diploic space.

The treatment consists of controlling seizures, physiotherapy, and speech therapy. Prognosis is better in cases when hemiparesis onset occurs after 2 years of age. Furthermore, the frequency of prolonged or recurrent seizures decreases. Hemispherectomy is a treatment option to alleviate treatment-resistant seizures in selected cases.[6]

CONCLUSION

DDMS can be taken into account in the differential diagnosis of seizure syndrome cases with clinical and radiological findings typical of the syndrome. The case we present includes many of the typical findings that would suggest the presence of DDMS. Also, it is important to remember that DDMS can occur after trauma.

Ethics

Informed Consent: Informed consent was obtained from the patient's parents.

Note: We presented in Turkish Neurosurgical Society 30. Scientific Congress, e-poster (EPS-158), 8-12 April 2016 Belek, Antalya, Türkiye.

Footnotes

Authorship Contributions

Surgical and Medical Practices: M.E.A., Concept: E.E.E., Design: M.E.A., Data Collection or Processing: M.E.A., Analysis or Interpretation: M.E.A., Literature Search: E.E.E., Writing: M.E.A.

Conflict of Interest: No conflict of interest was declared by the authors.

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