□ Case Report □

A case of Dyke-Davidoff-Masson syndrome in Korea

Jun Hwa Lee, M.D., Zee Ihn Lee, M.D.*
Ho Kyun Kim, M.D.† and Soon Hak Kwon, M.D.‡

Department of Pediatrics, Physical Medicine & Rehibilitation and Radiology†, Catholic University of Daegu, School of Medicine;*
Department of Pediatrics‡, Kyungpook National University School of Medicine, Daegu, Korea

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare condition characterized by asymmetry of cerebral hemispheric growth with atrophy on one side, ipsilateral compensatory osseous hypertrophy, and contralateral hemiparesis. We experienced a 17 month-old male who presented with left focal clonic or tonic-clonic seizures accompanied by left hemiparesis and developmental delay. Brain MRIs demonstrated progressive atrophy of the right cerebral hemisphere with dilatation of the lateral ventricle, expansion of the ipsilateral frontal sinus with calvarial thickening, and elevation of the petrous pyramid and orbital roof. Brain SPECT showed a decreased volume of the right hemisphere with reduced blood flow. We therefore report a case of DDMS with a review of the literature. **(Korean J Pediatr 2006;49:208-211)**

Key Words : Dyke-Davidoff-Masson Syndrome, Hemiatrophy, Hemiplegia, Seizure

Introduction

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare congenital malformation that was first described by Dyke, Davidoff, and Masson in 1933. They reported a series of 9 patients with hemiplegia who had cranial asymmetry on plain skull films[1]. The condition is characterized by cerebral hemiatrophy, facial asymmetry, contralateral hemiplegia, thickening or thinning of the cranial vault, seizures, mental retardation, and schizophrenia[2-6]. Diagnosis is made primarily by clinical features and radiological findings, but not many cases have been reported to date[1-5, 7-11]. We experienced a 17 month-old male who presented with characteristics of DDMS. We describe a case of DDMS in a Korean boy and review the literature.

Case Report

A male child was seen at 17 months with focal seizures characterized by brief, rhythmic jerks on the left with lapse of consciousness and eye deviation to the side. The onset of seizures occurred at about 11 months of age. Seizures occurred almost daily. When we examined him he had recently experienced increased frequency of seizures. The duration ranged from seconds to hours. He had had two episodes of status epilepticus, generalized convulsion in nature in the past. The pregnancy was uncomplicated and he was thought to be normal at birth. His birth weight was 3.5 kg. The family history was positive on the father's side for epilepsy. He passed through his developmental milestones slowly and stood up by himself at 15 months of age.

On physical examination, his height was 89.5 cm (98 percentile), his weight was 17 kg (99 percentile), and his head circumference was 47.5 cm (50 percentile). He jargoned and had a mild spastic hemiparesis on the left with extensor plantar response and right hand preference, but he had no obvious facial asymmetry. He was able to walk by himself, but had decreased arm swing on the left. Blood and CSF studies for stroke were unremarkable. The EEG revealed moderately suppressed background activity with lack of drug effect on the right hemisphere. Brain MRI showed a generally small right hemisphere with dilated ventricle and abnormal signals in the deep white matter (Fig. 1A). We started him on oxcarbazepine for seizures,

접수 : 2005년 8월 16일, 승인 : 2005년 10월 24일
책임저자 : 권순학, 경북대학교 의과대학 소아과학교실
Correspondence : Soon Hak Kwon, M.D.
Tel : 053)420-5704 Fax : 053)425-6683
E-mail : shkwon@knu.ac.kr

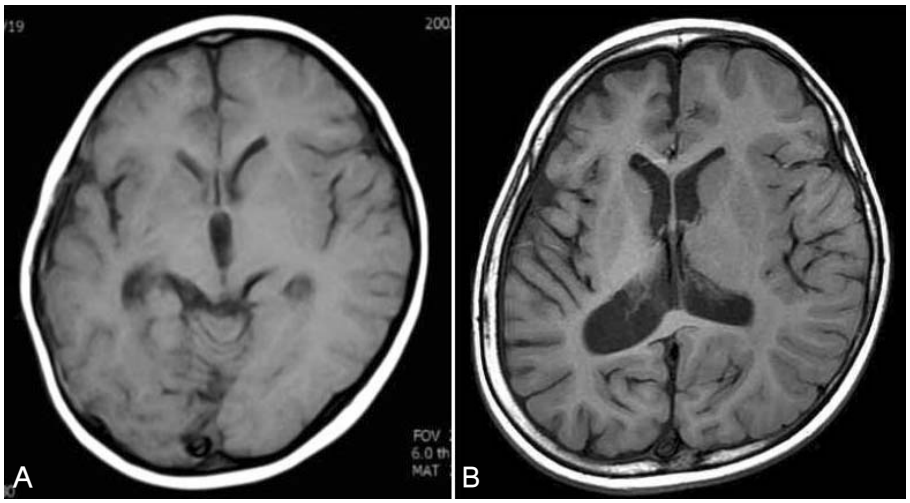

Fig. 1. MRI brain showed a relatively small right hemisphere with dilated ventricle and abnormal signals in the deep white matter **(A)** and follow up brain MRI at 34 months of age revealed progressive atrophy of the right cerebral hemispehere with ventricular dilation, prominent cortical sulci, compensatory calvarial thickening, and hypoplasia of the middle-frontal cranial fossa **(B)**.

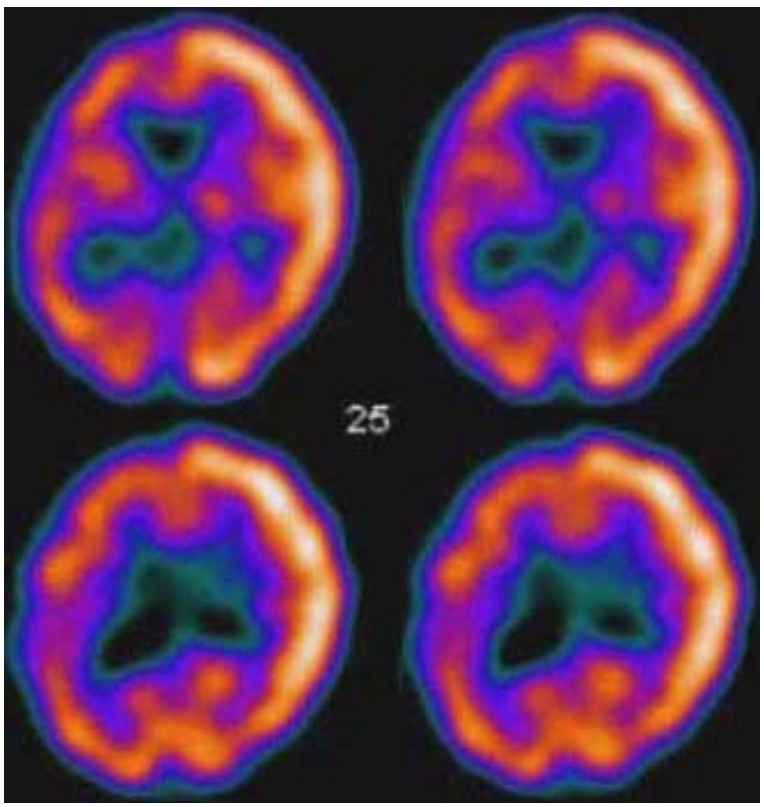

Fig. 2. Brain SPECT using ^{99m}Tc-ECD showed decreased volume of the right hemisphere with reduced blood flow.

but had to put him on additional antiepileptic drugs such as valproic acid, lamotrigine, and clonazepam. Despite multiple medications, he continued to have breakthrough seizures on occasions. Physical therapy was also started for left-sided weakness and dystonic posture. Follow-up brain MRI at 34 months of age revealed progressive atrophy of

the right cerebral hemispehere with ventricular dilation, prominent cortical sulci, and thalamic hypoplasia. There was also compensatory calvarial thickening and hypoplasia of the middle-frontal cranial fossa (Fig. 1B). At that time, brain SPECT showed decreased volume of the right hemisphere with reduced blood flow (Fig. 2).

He is currently receiving valproic acid (35 mg/kg) and topiramate (6 mg/kg). The frequency of seizures has been markedly reduced to near the seizure-free state. He has been receiving physical therapy and speech therapy since the first visit. However, no obvious changes have been found on his left hemiparesis with right hand preference and speech and language delay.

Discussion

DDMS is characterized by variable degrees of unilateral loss of cerebral volume, contralateral hemiplegia, and compensatory changes of the calvarium. The etiology of DDMS may be roughly divided into two categories, either congenital or acquired. In the congenital type, the cerebral insult is believed to be vascular in origin and likely occur during intrauterine life[3]. In the acquired type, trauma, infection, vascular abnormalities, or intracranial hemorrhage in the perinatal period or shortly thereafter may be responsible for the condition[3, 4, 7, 11]. In our case, T2-weighted images showed diffuse right hemisphere atrophy with dilated ventricle and abnormal signals in the posterior region of the deep white matter, suggesting secondary demyelinating changes after ischemic insult. In addition, cerebral cortical sulci and gyri were prominent. Some cases with similar radiological findings have been previously reported[7, 12, 13]. The current findings indicate that the brain insult occurred after the late perinatal period or right after birth, after completion of sulci formation. Considering the additional finding of compensatory skull changes, brain insult likely occurred in early life after birth. The mechanism of cerebral atrophy is still unclear, but it is hypothesized that ischemic episodes from a variety of different causes reduce the production of brain derived neurotrophic factors, which in turn lead to cerebral atrophy[7].

Because of limited experience in pediatrics, treatment guidelines for DDMS have not been clearly developed yet. Our case was moderately to severely affected and required a great deal of medical care and rehabilitation. The frequency of his seizures has been markedly reduced with aggressive medical treatment and the motor ability of his left hand has been somewhat improved by physical and occupational therapies, although he has had a persistent, moderate hemiparesis. Despite speech therapy, his speech has not developed like other cases of DDMS. Because of the progressive nature, other conditions such as Rasmussen syndrome or hemiconvulsion-hemiplegia-epilepsy (HHE) were once considered during the follow-up period[14-19], but immunosuppressive or immunomodulatory treatments have not been applied.

In conclusion, DDMS should be considered in any case that a child has hemiplegia, facial asymmetry, seizures, developmental delay, or mental retardation. The diagnosis may be made with the help of magnetic resonance (MR) of the brain and other images in such cases.

한 글 요 약

국소적 경련과 편마비를 동반한 Dyke-Davidoff-Masson 증후군 1례

대구가톨릭대학교 의과대학 소아과학교실,
재활의학교실*, 방사선과학교실†,
경북대학교 의과대학 소아과학교실‡

이준화 · 이지인* · 김호균† · 권순학‡

Dyke-Davidoff-Masson 증후군은 일측성 대뇌 반구의 위축과 동측의 대상성 골비대, 반대측의 편마비 등을 특징으로 하는 드문 신경질환이다. 본 증례는 17개월 남아가 좌측 국소적 경련 외에 좌측 편마비 및 기타 발달지연을 동반한 경우이다. 당시 시행한 뇌 자기공명영상에서 우측 대뇌 반구의 진행성 위축과 뇌실 확장, 두꺼워진 두개관과 동측 전두부 부비동의 확장, 추체 접형골의 거상을 보였고 뇌 SPECT 촬영상에 우측 대뇌 반구의 위축 및 혈류감소를 보였다. 이 같은 소견들은 Dyke-Davidoff-Masson 증후군에 합당하여 이에 저자들은 본 증례를 문헌 고찰과 함께 보고한다.

References

1) Dyke CG DL, Masson CB. Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. Surg Gyn Obstet 1933;57:588-600.
2) Parker CE, Harris N, Mavalwala J. Dyke-Davidoff-Masson syndrome. Five case studies and deductions from dermatoglyphics. Clin Pediatr (Phila) 1972;11:288-92.
3) Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy. Clin Imaging 1992;16:93-7.
4) Zilkha A. CT of cerebral hemiatrophy. Am J Roentgenol 1980;135:259-62.

5) Tasdemir HA, Incesu L, Yazicioglu AK, Belet U, Gungor L. Dyke-Davidoff-Masson syndrome. Clin Imaging 2002;26: 13-7.
6) White JH, Rust JB. Davidoff-Dyke-Masson syndrome presenting as childhood schizophrenia. J Autism Dev Disord 1979;9:37-40.
7) Ono K, Komai K, Ikeda T. Dyke-Davidoff-Masson syndrome manifested by seizure in late childhood : a case report. J Clin Neurosci 2003;10:367-71.
8) El Bahri-Ben Mrad F, Mrabet H, Ben Sghaier R, Mrabet A. Dyke-Davidoff-Masson syndrome : a report of two cases. J Neuroradiol 2005;32:50-3.
9) Yamazaki K, Hirata K. Dyke-Davidoff-Masson syndrome. Ryoikibetsu Shokogun Shirizu 2000:177-8.
10) Sener RN. Growing skull fracture in a patient with cerebral hemiatrophy. Pediatr Radiol 1995;25:64-5.
11) 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.
12) Lee BC, Lipper E, Nass R, Ehrlich ME, de Ciccio-Bloom E, Auld PA. MRI of the central nervous system in neonates and young children. Am J Neuroradiol 1986;7:605-16.
13) Dix JE, Cail WS. Cerebral hemiatrophy : classification on the basis of MR imaging findings of mesial temporal sclerosis and childhood febrile seizures. Radiology 1997;203:269-74.
14) Maeda Y, Oguni H, Saitou Y, Mutoh A, Imai K, Osawa M, et al. Rasmussen syndrome: multifocal spread of inflammation suggested from MRI and PET findings. Epilepsia 2003;44:1118-21.
15) Hart Y. Rasmussen's encephalitis. Epileptic Disord 2004;6: 133-44.
16) Granata T, Gobbi G, Spreafico R, Vigevano F, Capovilla G, Ragona F, et al. Rasmussen's encephalitis : early characteristics allow diagnosis. Neurology 2003;60:422-5.
17) Salih MA, Kabiraj M, Al-Jarallah AS, El Desouki M, Othman S, Palkar VA. Hemiconvulsion-hemiplegia-epilepsy syndrome. A clinical, electroencephalographic and neuroradiological study. Childs Nerv Syst 1997;13:257-63.
18) Freeman JL, Coleman LT, Smith LJ, Shield LK. Hemiconvulsion-hemiplegia-epilepsy syndrome : characteristic early magnetic resonance imaging findings. J Child Neurol 2002; 17:10-6.
19) Herbst F, Heckmann M, Reiss I, Hugens-Penzel M, Gortner L, Neubauer B. Hemiconvulsion-Hemiplegia-Epilepsy-Syndrome (HHE). Klin Padiatr 2002;214:126-7.