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Congenital Bilateral Perisylvian Syndrome : Analysis of the First Four Reported Korean Patients

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The advent of MRI technique has enabled the diagnosis of neuronal migration disorders(NMD) and made it possible to make "in vivo" diagnosis. Congenital bilateral perisylvian syndrome(CBPS) is a recently described disease identity characterized by pseudobulbar palsy, epilepsy, mental retardation, and migration disorders in the bilateral perisylvian area. We have identified four CBPS patients based on neuroimaging and dysarthria patterns among the candidates for epilepsy surgery. All the patients had orofacial diplegia and variable degrees of mental retardation. In the spectrographic analysis of dysarthria, the loss of specific characteristics of formants of vowels and increment of noise in the high frequency formants were observed. Epilepsy was present in all, but only one patient showed intractable seizure requiring surgical intervention. MRI was most helpful in identifying NMD and polymicrogyria in both centroparietal areas in this context. Great alertness is needed to identify this disorder to determine the etiology of epilepsy and dysarthria of uncertain origin.

Key Words : *Congenital bilateral perisylvian syndrome, Neuronal migration disorder, Epilepsy, Dysarthria.*

INTRODUCTION

Congenital bilateral perisylvian syndrome(CBPS) is a recently described entity diagnosed on the bases of neuroimaging and speech abnormalities(Kuzniecky et al., 1989 ; Palmini et al., 1991). This syndrome consists of orofacial diplegia, markedly limited movement of the tongue and lips, difficult swallowing, variable degrees of mental retardation, and epilepsy. More than 85% of cases have epilepsy which is frequently intractable(Kuzniecky et al., 1990 ; Palmini et al., 1991).

This syndrome was originally described in identical twins by Graff-Radford et al.(1986). Around forty patients have been reported in the literature, mostly from American and European countries. We herein present the analysis of four Korean patients with CBPS.

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PATIENTS AND METHODS

We could detect four CBPS patients out of 86 intractable epileptic patients investigated for surgical candidacy in Chonbuk National University Hospital from September, 1992 to September, 1993. Sixteen channel standard/sphenoidal EEGs were carried out and seizure was recorded with Telefactor video-EEG telemetry. All the patients had MRI studies. Multimodal evoked potential studies were performed using Nicolet Viking Ile EP/EMG system. We evaluated the acoustic characteristics of dysarthria with a

digital computerized spectrograph, Visi-Pich® (Kay Elemetrics Corp., Model 6087 AT) and a Nasometer ® (Kay Elemetrics Corp., Model 6202-2). Neuropsychological tests were performed according to the Korean-Wechsler Adult Intelligence Scale(KWALS) in all the patients.

Three patients were treated with antiepileptic drugs. One patient who had medically intractable epilepsy was treated surgically. Anterior temporal resection with amygdalohippocampectomy was performed under general endotracheal anesthesia. Dissection was carried out using a cavitron ultrasonic suction aspirator under a surgical microscope.

RESULTS

Clinical features

Clinical features of the 4 patients are summarized in Table 1. The patients consisted of 2 males and 2 females. All the patients had a past history of delayed developmental milestones in their infancy and childhood : All walked and talked after 24 months of age. They went to school but were not able to keep up with others. K-WAIS revealed trainable level of mental retardation in 3 patients and a educable level in 1 patient. Their social life was dependent in case 1 and 3.

Physical examination showed varying degrees of pseudobulbar symptoms. Swallowing difficulties were noted in all. Protrusion and movement of the tongue were impossible in case 1, moderately impaired in case 3, and slightly impaired in the two other cases(Fig. 1). Drooling was accompanied in cases 1 and 3. In addition, facial expression was relatively set in all.

Table 1. Clinical characteristics of CBPS* Patients

Case	Age/Sex	Type of Seizure	Mental Retardation	Dysarthria	Tongue Protrusion
1	19/M	Partial motor Generalized	Trainable	Severe	Impossible
2	23/M	Partial motor	Trainable	Moderate	Slightly impaired
3	17/M	Partial motor Generalized	Trainable	Moderate	Moderately impaired
4	22/F	Partial motor	Educable	Moderate	Slightly impaired

*Congenital Bilateral Perisylvian Syndrome

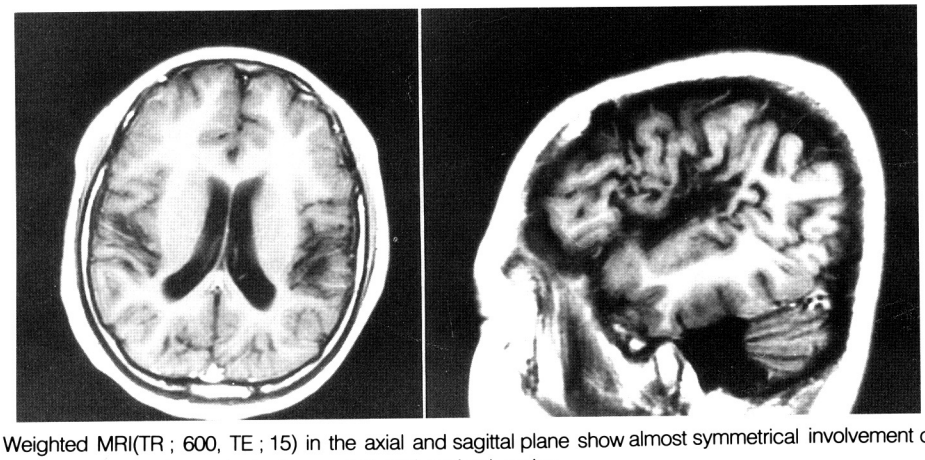

Fig. 1. T1 Weighted MRI(TR ; 600, TE ; 15) in the axial and sagittal plane show almost symmetrical involvement of the cortex, insular exposure, and polymicrogyria in the central and parietal region.

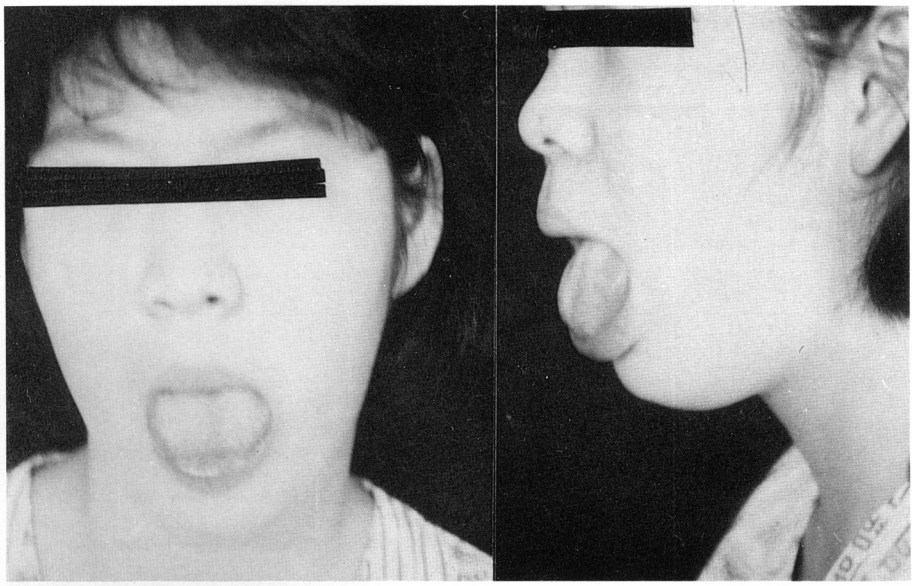

Fig. 2. Limited movement and protrusion of the tongue outside the mouth.

Dysarthria

Dysarthria was the most prominent clinical feature. Articulation of lingual, dental, and alveolar consonants showed a peculiar pattern of impairments. In the spectrographic analysis, loss of specific characteristics of formants of vowels and increment of noise in the high frequency formants were observed(Fig. 2). The voice onset time was prolonged in the bilabial and alveoloar stop consonants. Total duration of meaningless two syllables were increased as well. In the Visi-Pich® analysis, monopitch and monoloudness were characterized and diadochokinetic rate was decreased. The degree of nasalance was markedly increased in CBPS patients in comparison with normal subjects.

Seizure and EEG findings

All the patients showed partial motor seizures, two patients had additional generalized tonic clonic seizures. Antiepileptic drugs could prevent seizures in cases 2, 3, and 4. Case 1 showed as intractability to medication and surgical treatment was needed.

Interictal epileptiform abnormalities consisted of bilateral spike discharges recorded from parietal areas without predominance in case 2 and 3, unilateral spikes from the inferomesial temporal area and the centrotemporal area in case 1 and 4, respectively. Seizure recording showed rhythmic activitives of 5-7 Hz in the right temporal region in case 1.

Evoked potentials

Three channel somatosensory evoked potential study of bilateral median and posterior tibial nerves showed prolonged N19 and p 37 latencies and decreased p37 N45 amplitude in the left side in case 1. Two channel brain stem auditory evoked potential study showed normal Ⅰ, Ⅱ, Ⅲ, Ⅳ, Ⅴ latencies and Ⅰ-Ⅲ, Ⅲ-Ⅴ, Ⅰ-Ⅴ interpeak latencies in all. In flash visual evoked potential study, slightly prolonged N1 latencies were noted bilaterally and in the left side in cases 1 and 3 respectively. The worse clinical symptoms were more correlated with findings of abnormal evoked potentials.

MRI Findings

MRI revealed bilateral perisylvian migration disorders around the sylvian-rolandic region. Polymicrogyria and opening of sylvian fissure were noted in all (Fig. 3). The involvements were symmetrical in case 1 and asymmetrical in the others. In addition,

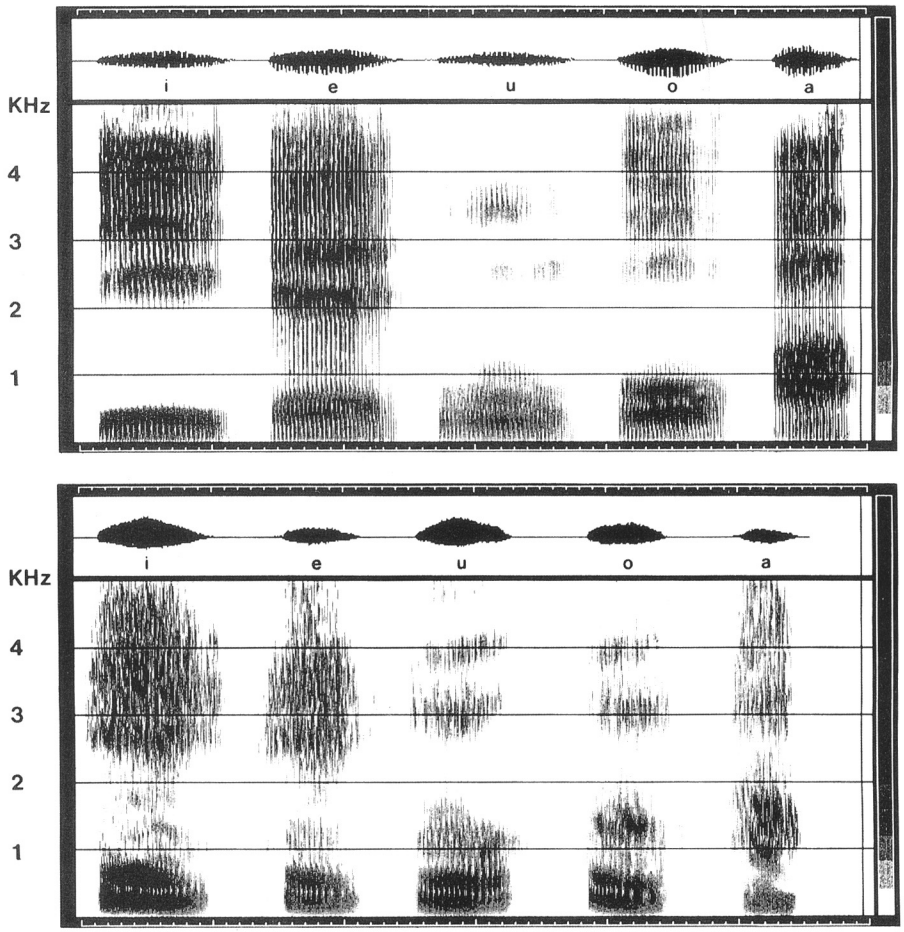

Fig. 3. Acoustic characteristics of vowel formants measured with a spectrogram in CBPS(C-2) and normal subject(C-1). Note the loss of specific characteristics of formants of vowels and increment of noise in the high frequency in CBPS.

case 1 showed hippocampal sclerosis.

Follow-up after treatment

Eight months of follow-up after anterior temporal resection with amgdalohippocampectomy showed complete seizure relief in case 1. The seizures of the other 3 patients were satisfactorily controlled with anticonvulsants.

DISCUSSION

The search for an etiology in patients with severe epilepsy is imperative. The etiological definition allows (1) adequate selection of antiepieptic drugs, (2) delineation of prognosis and (3) of possibilities for surgical treatment, and (4) determination of future planning for comprehensive management of patients. When severe epilepsy is associated with other disabilities such as mental retardation and speech abnormalities, detailed etiological explanation is very informative to the parents as well as the personnel involved in the rehabilitation program.

CBPS has a peculiar pattern of neuronal migration defect. Some evidence postulated genetic etiology to explain this peculiar pattern, based on the presence of CBPS in one pair of twins and siblings(Graff-Radford et al., 1986 ; Christie et al., 1989). The genetic etiolgy explains that either a restricted

and selective aberration of neuronal migration occurring between the third and fifth gestational months or predisposing a vulnerability to a superimposed insult occurring within this period can determine the pattern of neuronal migration disorder in CBPS. However, another hypothesis, based on experimental studies, was proposed. This hypothesis suggests that vascular accident in utero is responsible for postmigration, neuronal loss, and glial replacement, thus causing the focal polymicrogyria(Dobyns and McClugage, 1986 ; Dvorak and Feit, 1977 ; Dvmakiorak et al., 1978 ; Richman et al., 1974 ; Williams and Caviness, 1976).

The value of MRI in the recognition of CBPS cannot be overemphasized. The advent of MRI enabled recognition of this syndrome and made it possible to make "in vivo" diagnosis. Especially, MRI has a high resolution to differentiate the gray matter from the white matter, which allows precise definition of the cortical gyral pattern(Barkovich et al., 1988 ; Kuzniecky et al., 1988 ; Kuzniecky et al., 1990 ; Palmini et al., 1991). This enables us to easily detect the cortical abnormality. Otherwise, neuronal migration disorders(NMD) remain unrevealed and are described as cryptogenic or idiopathic. Some authors used CT to diagnose NMD but many artifacts were observed(Dobyns and McClugage, 1985 ; Ohno et al., 1979 ; Zimmernan et al., 1983). In our institute, MRI is recommended first if there is a suspicion of NMD.

The characteristic clinical features are made by bilateral involvement of the orofacial area in the motor cortex. This bilateral involvement in the brain abolishes the compensatory mechanism contralateral to the lesion site. Among various clinical features, dysarthria is the most prominent symptom. We tried to quantify the dysarthria using computerized spectrographs, Visi-Pich®, and Nasometer®. We could conclude that CBPS patients had a unique pattern of dysarthria and the study of acoustic characteristics in these patients reflects well the impaired movement of bilateral lips, tongue, and velopharynx-(Kim et al., 1993).

There have been forty cases of CBPS reported in the literature(Kuzniecky et al., 1993). This entity seems to be much more common than is usually appreciated. We were able to identify four CBPS patients out of 86 intractable epileptic patients. The clinical picture is sufficiently similar to suggest the existence of this syndrome, and MRI imaging supports this contention. Therefore a high index of alertness is needed to identify this syndrome.

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