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Case Report

Schizencephaly Type I : Magnetic Resonance Imaging and Single Photon Emission Computed Tomography Features

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SUMMARY

We report a case of type I schizencephaly diagnosed by magnetic resonance imaging in a 2 - year - old boy with delayed speech and hemiplegia. Cerebral single photon emission computed tomography showed an abundant cerebral blood flow at the site of the cleft lesion. Single photon emission computed tomography thus may provide useful cerebral blood flow images in schizencephaly patients.

Key Words : Schizencephaly, Delayed speech, Hemiplegia, Magnetic resonance imaging, Single photon emission computed tomography, Inversion recovery

INTRODUCTION

Schizencephaly is a congenital brain anomaly resulting from impaired migration of nerve cells beginning at a fetal age of about seven weeks. This pathological entity was initially described by Yakovlev and Wadsworth in 1946 [1,2]. Clinical signs of schizencephaly include delayed psychomotor development, hemiplegia, quadriplegia and intractable epilepsy. We report a case of unilateral (left) schizencephaly diagnosed by magnetic resonance imaging (MRI) of the brain in a male child presenting with delayed speech and right hemiplegia. In addition, single photon emission computed tomography (SPECT) provided significance findings.

CASE REPORT

The patient was 2 years and 7 months old. He was born during the 39th week of pregnancy by spontaneous cephalic delivery. Birth weight was 2,580 g. His parents were not consanguineous. During pregnancy, his mother had no infections and took no medication. With respect to motor development, the patient could hold his head steady at the age of 4 months and could stand with support at 1 year and 2 months. At 1 year and 9 months, he walked alone and right hemiparesis was developed. Speech development was delayed and he was able to speak only several words at 2 years. There was no history of convulsions. He was referred to our hospital for delayed speech and right hemiplegia.

Physical examination at the time of referral showed a left - handed patient of average physique with no visible congenital anomalies. Biochemical blood examination and urine analyses were normal. His neurological examination was remarkable only for an acceleration of the deep tendon reflexes of the right upper and lower limbs. Electroencephalography revealed no definitive laterality or paroxysmal waves. MRI of the head (Fig. 1) showed a cleft extending from the cortex of the brain surface to the left lateral ventricle and covered by a thick layer of abnormal gray matter. The transparent septum was defective, and dimples were observed in the lateral wall of the left

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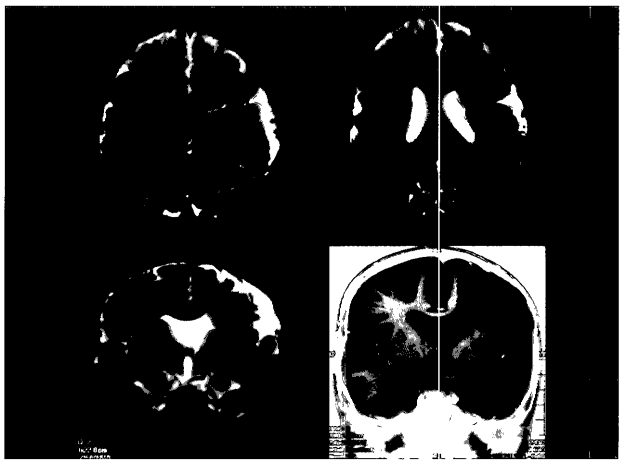

Fig. 1 T2-weighted (spin echo TR = 3,500 msec TE = 100 msec) and inversion recovery magnetic resonance images of the lower right side.

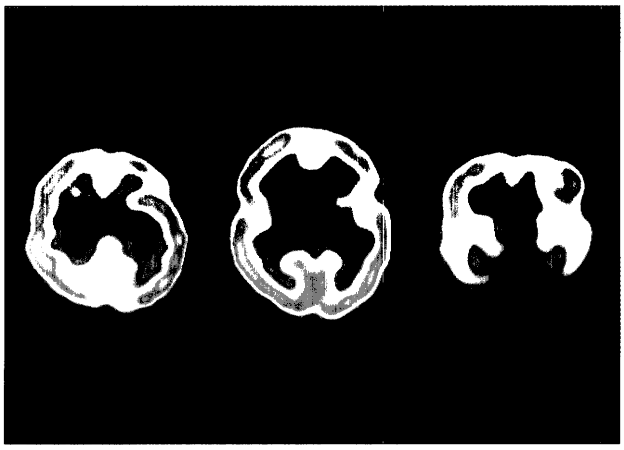

Fig. 2 Single photon emission computed tomography study with ^{99m}Tc-ECD.

lateral ventricle. Diagnosis of type I schizencephaly (closed lip type) was made on the basis of these MRI findings. SPECT with ^{99m}Tc-ECD (Fig. 2) revealed increased regional blood flow at the site of the cleft lesion. At present, the patient is receiving rehabilitation and undergoing orthosis for his right lower limb position.

DISCUSSION

Schizencephaly is morphologically classified into two subtypes : closed lip (type I) and open lip (type II). Type I schizencephaly is characterized by a narrow cleft, fusion of the cerebral walls surrounding the cleft, and an absence of communication between the subarachnoid space and lateral ventricle. Type II is characterized by a wide cleft, separated cerebral walls, the presence of communication between the subarachnoid space and lateral ventricle, and hydrocephalus[3~6]. The more widespread use of MRI over the past few years has facilitated the diagnosis of this disorder, resulting in increasing numbers of reports of clinically mild cases, and the recognition that the incidence of this disorder is higher than what was previously reported.

In our case, the inversion recovery (IR) method (Fig. 1) provided MRI images with good contrast between gray matter and white matter which proved useful for identifying gray matter covering the cleft. Single photon emission computed tomography (SPECT) (Fig. 2) showed an increased blood flow at the site of the schizencephaly cleft, suggesting that cerebral blood flow in the cleft lesion was maintained. Our SPECT findings indicate that SPECT imaging of schizencephaly patients may provide useful information regarding cerebral blood flow in the abnormal cortex and the degree of cortical morphological abnormality.

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