RADIONUCLIDE STUDIES IN VASCULAR INFANTILE HEMIPLEGIA

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Two cases of acute infantile hemiplegia are presented. The first case is a patient with moyamoya disease with excellent correlation between the brain scan findings and her clinical signs and symptoms and the carotid arteriogram. The second patient had occlusive cerebrovascular disease secondary to thromboemboli with excellent correlation between the brain scan findings and his clinical state and the neuropathologic examination. In both cases the static brain scans and the radionuclide angiogram were comparable to those reported in adults with cerebrovascular disease both in appearance and in temporal sequence. Radionuclide studies can be an important and reliable diagnostic tool in the evaluation of acute infantile hemiplegia.

Although radionuclide studies are an important diagnostic tool in the evaluation and diagnosis of occlusive cerebrovascular disease in the adult patient, their application and interpretation in infantile cerebrovascular disease has received only scanty attention, probably because of the rarity of these conditions in the pediatric age group (1). The following two cases of acute infantile hemiplegia demonstrate findings similar to those that would be expected in adults and show an excellent correlation between the scintigraphic findings and the clinical state, the arteriographic studies, and neuropathologic findings.

CASE REPORTS

Case 1. The patient was born on Jan. 15, 1971 and first admitted to Strong Memorial Hospital (S.M.H.) on July 3, 1972 for evaluation of right hemiparesis of 8-days duration.

Seventeen days before admission she developed persistent rhinorrhea and later became irritable and febrile. On June 25, she did not use her right upper extremity. She was found to have a right hemiparesis with right facial weakness.

Upon admission to S.M.H. the patient was afebrile. Examination revealed right visual field neglect, slight flattening of the right nasolabial fold, a mild right hemiparesis, and right hyperreflexia.

She had a hypochromic, microcytic anemia, elevation of SGOT and LDH enzymes, and white cells in her urine. Cerebrospinal fluid examination was normal. An EEG revealed poor formation of sleep spindles and absent drug induced fast activity over the left hemisphere. A brain scan on July 6 (Fig. 1) was interpreted as showing cerebral infarction of the orbitofrontal and rolandic branches of the left middle cerebral artery. A left carotid arteriogram the same day revealed marked stenosis of the terminal internal carotid artery and the proximal portions of the anterior and middle cerebral arteries with hypertrophy of the lenticulostriate, ophthalmic, and middle meningeal arteries with tortuous collateral channels over the left hemisphere.

A second brain scan on Aug. 19 showed residual uptake in the distribution of the left middle cerebral artery.

She was readmitted to S.M.H. on Sept. 25, 1972 following the onset of a left hemiparesis. Examination on admission was normal except for a temperature of 100.4°, a mild left hemiparesis, left hyperreflexia, and a left Babinski reflex.

An EEG was unchanged from the previous tracing. A brain scan on Sept. 27 was similar to that of Aug. 19.

Upon discharge, on Sept. 27, the left hemiparesis was improved with less posturing and much recovery of fine distal movements.

A final brain scan performed Oct. 10 again showed a faint, residual uptake over the left hemisphere and no abnormality over the right hemisphere.

Case 2. The patient was born on Mar. 6, 1971.
He was admitted to S.M.H. on July 21, 1972 with a right hemiparesis of 1-day duration.

Prior history included a moderate hyperbilirubinemia following birth. Subsequently he was found to have a simian crease, hypochromic, microcytic anemia, congenital heart disease of unknown configuration with a right-to-left shunt and congestive heart failure, and congenital abnormalities of the ribs and spine. Neurologic examinations were without abnormality.

Two days before admission he developed a left otitis media. One day before admission his mother noted that he was not moving his right upper extremity as well as usual and the morning of admission she noted weakness of his right face.

On examination he had a left otitis media, a right hemianopsia, a flaccid right hemiplegia, and bilateral Babinski reflexes. The deep tendon reflexes were normal and pain sensation was intact. The anterior fontanelle was flat.

A brain scan on July 21 was normal. An attempted radionuclide angiogram was unsuccessful. The EEG demonstrated slowing on the left, greatest in the parietal region. Cerebrospinal fluid examination was normal. A repeat brain scan on July 28 (Fig. 2A) showed an area of increased uptake deep within the posterior left frontal lobe.

A third brain scan on Aug. 4 showed residual uptake deep within the left frontal lobe. A fourth brain scan on Aug. 18 was normal.

He was readmitted on Sept. 26 following a generalized seizure. He had had an upper respiratory infection for the previous 3 days. Examination revealed an unconscious patient with occasional right-sided seizures and deviation of the eyes to the right and decerebrate posturing of both upper extremities. The right pupil was smaller than the left and did not react to light. There was right ptosis, a left hemiparesis, bilateral clonus, and right-sided hyperreflexia.

A radionuclide angiogram on Oct. 30 showed delayed perfusion and delayed clearing over the right hemisphere. The static brain scan showed increased activity peripherally over the right hemisphere in the distribution of the right middle cerebral artery (Fig. 2B).

He remained intermittently decerebrate, had frequent right-sided seizures with deviation of the eyes to the right, and developed a left hemiparesis with bilateral Babinski reflexes. He died on Nov. 4, 1972.

At autopsy, the child was found to have congenital heart disease with multiple defects. The brain weighed 830 gm and appeared swollen and congested. There was thromboembolus formation in a branch of the left middle cerebral artery and an old cavitory lesion of the left corona radiata and putamen (Fig. 3, thick arrow). There was a second thromboembolus at the trifurcation of the right middle cerebral artery, and the branches immediately distal to this with recent encephalomalacia of the lateral aspect of the right frontoparietal and right frontotemporal regions that extended medially to involve the right extreme capsule, the claustrum, the external capsule, and part of the right putamen (Fig. 3, thin arrows).

Microscopically, there were recent, minimally organized thromboemboli with intact red blood cells within the right middle cerebral artery, one of its sylvian branches, and a branch of the left middle cerebral artery. There was recent, organizing coagulation necrosis of the right fronto-parieto-temporal region with eosinophilic neuronal cytoplasm, neovascularization, nuclear swelling, and increase in cytoplasm of the astrocytes and perivascular macrophages. The infarction of the left putamen was older (estimated 3½ months) and was becoming a cavity.

FIG. 1. Case 1. Scan obtained 11 days after acute onset of right hemiparesis.

FIG. 2. Case 2. (A) Scan obtained 8 days after acute onset of right hemiparesis. (B) Scan obtained 4 days after onset of seizures (second admission). Note clearing of previous abnormality on left.
that contained a decreasing number of macrophages within its center and large number of Nissl plump astrocytes and neovascularization around its periphery. Bacterial and fungal stains were negative.

DISCUSSION

The causes of acute, infantile hemiplegia are many and varied and include degenerative disease, infections, neoplasms, seizures, trauma, and vascular diseases. It is of interest that in many of the vascular cases there is a period of illness, often febrile, and irritability before the onset of the neurologic deficit as was seen in these two cases (2,3).

Our first patient with infantile hemiplegia is felt to represent a case of "moyamoya" or multiple progressive intracranial arterial occlusive disease (3). The narrowing of the distal internal carotid artery and of the proximal portions of both the anterior and middle cerebral arteries, the "moyamoya" (Japanese, meaning "something hazy") appearance of the lenticulostrate arteries, and the external-internal carotid artery collateral circulation seen on her left carotid arteriogram are characteristic of an early stage of this disease. The appearance of the first brain scan (Fig. 1) correlated with the arteriographic and clinical findings.

The etiology of "moyamoya" cerebrovascular disease is unknown even though some authors mention that many of their patients had preceding infections of the head or neck (4). It is a disease primarily of infants and young adults and may be more common in Japanese (3,4). The disease is progressive and may ultimately be bilateral, both clinically and arteriographically, and involve not only both internal carotid arteries distally and both anterior and middle cerebral arteries proximally, but also the basilar artery and the posterior cerebral arteries (3,4). However, as in this patient, it is not uncommon initially to have unilateral signs and symptoms and arteriographic evidence of "moyamoya" vascular disease; then several weeks or months later, contralateral manifestations become evident (3,4).

The second case of infantile hemiplegia, also vascular in origin, was secondary to embolization of thrombotic material which probably originated from the malformed heart. The thromboemboli and the right-sided infarction are felt to be 7–10 days old. The brain scan findings of Oct. 30, 1972 (Fig. 2B) are thus explained on the basis of the right middle cerebral artery thromboembolus and the resultant recent infarction in its vascular distribution. The infarction of the left putamen and corona radiata correlates well with the prior right hemiparesis of July 21 and the positive brain scan of July 28 (Fig. 2A). This infarction was the result of a thromboembolus within the lenticulostrate arteries. The clinical history (except for the age) and brain scan are remarkably similar to a case described by De-Land (5,6).

Radionuclide studies, both static imaging and radionuclide angiography, are ideally suited for the evaluation of neurologic problems in infants and children. Application of the same criteria used in the diagnosis of adult cerebral vascular disease can give equally reliable and rewarding results in the diagnosis of similar problems in the younger age group. Both the flow study and scan findings and the temporal sequence of changes in these findings in the infants reported here are similar to those previously described in adults. The use of these techniques should be encouraged in the evaluation of pediatric neurologic disease.

REFERENCES