SPECT, Magnetic Resonance and Angiographic Features in a Moyamoya Patient Before and After External-to-Internal Carotid Artery Bypass

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Brain SPECT changes in a patient with cerebral vaso-occlusive disease caused by moyamoya disease are presented. Brain SPECT after a revascularization procedure demonstrated significant improvement of brain flow which was confirmed by angiography. Clinical and magnetic resonance features are reviewed.


Moyamoya disease is a vaso-occlusive disorder involving the large vessels of the brain for which no definite therapy has been described. Diagnosis and follow-up of moyamoya patients is done with angiograms which demonstrate the characteristic appearance and dynamic nature of the disease. External carotid-to-internal carotid artery (EC-IC) anastomoses have been utilized in various vaso-occlusive disorders of the brain with varying degrees of success. This paper describes the scintigraphic and radiographic patterns of a successful EC-IC anastomosis in a patient with moyamoya disease.

CASE REPORT

Moyamoya disease was first diagnosed following a right hemispheric cerebrovascular accident when our female patient was 18 yr old. At the age of 21, she presented with progressive numbness and weakness of the left arm and left lower extremity. The patient partially recovered and over the next 3 yr she did well except for intermittent episodes of mild weakness and numbness involving the left upper and lower extremities. At the age of 24, the patient developed a more severe episode of weakness in the left arm and left leg which prompted her admission for evaluation. A magnetic resonance (MR) image of the head, an [123I]IMP brain study and cerebral angiogram were all obtained within two days (Figs. 1–5).

Brain single-photon computerized tomography (SPECT) was performed 20–25 min after intravenous injection of [123I]IMP. Radionuclide injection was performed in a quiet, dimly lit room with eyes open. A Siemens Orbiter SPECT gamma camera was used with a general-purpose, parallel-hole collimator interfaced to a MDS A® computer system. Sixty-four views were obtained at 5.6° angular increments with an acquisition time of 30 sec per view, resulting in a total acquisition time of approximately 32 min. A Gaussian pre-filter was applied to the raw data prior to reconstruction. Transaxial images were reconstructed at one pixel per slice using an attenuation correction and a Ramp filter (cutoff = 1.0). Coronal and sagittal slices were then reconstructed from this transaxial data at two pixels per slice. Two pixel transaxial slices were photographed for hardcopy along with the sagittal and coronal slices.

In view of the progression and instability of neurologic symptoms and radiographic and scintigraphic findings, a right external carotid to right middle cerebral artery bypass operation was performed 1 mo following her admission. Except for mild residual hemiparesis, the patient’s symptoms subsided. A follow-up angiogram and a brain flow study using 99mTc-HMPAO were obtained (Figs. 3, 4, 6). Technical details used for the [123I]IMP images were duplicated for the 99mTc-HMPAO images. The patient’s clinical neurologic status has remained stable and no new symptoms or neurologic deficits have developed in the succeeding 3 yr.

DISCUSSION

Moyamoya disease is a rare form of cerebrovascular disease (CVD) involving the distal internal carotid artery and/or the circle of Willis (1). Pathological findings in moyamoya disease include fibrous or fibro-cellular intimal thickening at major cranial arteries (6). Progressive narrowing and occlusion of the main cerebral arteries with development of parenchymal, leptomeningeal and transdural collaterals is demonstrated on angiography. Bilateral involvement has been suggested as a definite criterion for diagnosis of the disease (2) but evolution from unilateral to bilateral disease has been postulated in some cases (3). Pseudoangiomaticous collateral network at the base of the brain, characterized as “moyamoya” ("puff of smoke") vessels on angiography, is considered to result from a variety of vaso-occlusive conditions including diseases such as neurofibromatosis and sickle cell disease (4). How-
ever, most cases are idiopathic, as initially reported in Japanese children and young adults (5).

A wide range of presenting symptoms have been described that correlate with age (7). In patients under 21 yr old, transit ischemic events are the most common manifestation. In patients over 21 yr, intracranial hemorrhage, mostly subarachnoid hemorrhage, is common. Computed tomography (CT) and/or MR imaging are the initial diagnostic studies in practice and can confidently establish the diagnosis of moyamoya (8–10). However, angiography is usually performed to confirm the diagnosis and to plan surgical treatment (1,11).

Lipophilic radiopharmaceuticals, such as $^{[123]}$IIMP and $^{99m}$Tc-HMPAO, are capable of traversing the intact blood-

FIGURE 1. Coronal T2-weighted images. (A) Image at the frontal horns level demonstrates absence of the normal carotid terminus flow void (arrow) and sylvian branches of the right middle cerebral artery (arrow-head). Dilated moyamoya collateral vessels are represented by the heterogeneous low signal within the right coronaradiata (curved arrow). (B) Image at the atria of the lateral ventricles demonstrates a parasagittal right frontal infarct (short arrow) and focal asymmetric atrophy laterally representing effects of chronic ischemia (open arrowheads).

FIGURE 2. Axial proton density images. (A) Images at the basal ganglia level show heterogeneous high signal in the right external capsule caused by moyamoya collaterals with surrounding gliosis (curved arrow). There is high signal in right sylvian branches due to slow flow (arrowheads). (B) Vertex image shows a right parasagittal high signal zone caused by infarct, bridging the pre and post central gyri (arrow).

FIGURE 3. Transaxial pre- and post-EC-IC bypass images show initial marked decreased uptake in the right cerebral cortex with subsequent improvement.

FIGURE 4. Lateral view of a right common carotid angiogram. There is occlusion of the carotid terminus just distal to the posterior communicating artery (arrow). Extensive perforating collaterals are noted (arrowheads) demonstrating the typical "puff of smoke" appearance of moyamoya disease.

brain barrier and they concentrate rapidly in neurons proportionally to regional blood flow. Generally, retention in the brain parenchyma is stable for up to 24 hr with $^{[123]}$IIMP (13) and for about 8 hr with $^{99m}$Tc-HMPAO (14), which makes both suitable for SPECT. Brain SPECT with $^{[123]}$IIMP or $^{[99m}$Tc]HMPAO has been suggested as a useful study for evaluation of regional blood flow (12) and in diagnosing and managing patients with CVD, seizure disorder, dementia, brain tumor, trauma and psychiatric disorders (15,16–18). Due to the wide availability of $^{99m}$Tc and its superior counting statistics, $^{99m}$Tc-HMPAO seems to be more useful than $^{[123]}$IIMP for clinical use.

Moyamoya disease is one of the entities in which a brain SPECT study plays an important role by showing abnormal blood flow (19). Areas of hypoperfusion resulting
in our patient resulted in improved flow through surgical anastomoses, reduced flow through “moyamoya” collateral vessels and increased perfusion through the right middle cerebral artery branches, as shown on angiography (Fig. 6). This corresponded to the SPECT images which showed markedly improved perfusion in the right cerebral hemisphere after surgical intervention.

The natural history of moyamoya disease is variable. Some patients remain in a stable condition on medical therapy for an extended period of time (23). Medical treatment includes vasodilators, anticonvulsants, anticoagulants and steroids. As for surgical treatments, in addition to superficial temporal-middle cerebral artery bypass surgery, several indirect revascularization procedures have been developed (24). Encouraging results with neurological and angiographic improvements have been reported, particularly with encephalo-duro-arteriosynangiosis, in which development of spontaneous anastomoses is achieved by adhering the donor scalp arteries to the dural surface (25).

Moyamoya disease is an entity affecting the young population, in whom early bypass interventions may prevent persistent brain damage secondary to infarctions. A brain SPECT study with lipophilic agents is a promising tool to establish noninvasively the status of brain flow and to monitor patients with moyamoya before and after intervention. Long-term follow-up with neurological monitoring is necessary for a better understanding of the role of brain SPECT imaging in revascularization procedures of the brain.

REFERENCES


(continued from page 1684)

**SELF-STUDY TEST**

**Gastrointestinal Nuclear Medicine**

**ANSWERS**

Choy, et al. have reported an increase in specificity for acute cholecystitis (from 83% to 100%), compared to the use of delayed imaging without a loss of sensitivity.

Occasionally, it is difficult to distinguish pooled activity within the proximal duodenum from the gallbladder itself, or one may mistake pooled activity within the duodenum for the gallbladder. In these instances, it is useful to administer 200–300 ml of water by mouth. This will facilitate flushing of activity from the duodenum, but has no effect on the gallbladder. Since the duodenum is clearly identified in this patient, and activity within it is seen to change over time, it should not be mistaken for the gallbladder. Hence, imaging after administration to this patient would not provide further useful information.

References


**ITEMS 21–24: Factors Affecting 99mTc IDA Uptake**

**ANSWERS:** 21, T; 22, T; 23, F; 24, F

Since the development of the original IDA derivatives, work on the molecular structure of these compounds has proceeded in the direction of producing an agent with ideal biokinetics. The 99mTc IDA agents are carried in blood nonspecifically bound to plasma proteins, particularly albumin. The lipophilicity of the compounds is closely related to the level of protein binding. Substitutions of nonpolar groups on the phenyl ring of the molecule make it more lipophilic. Protein binding prevents renal excretion and promotes hepatic uptake. This is an important consideration in the jaundiced patient where these agents compete for protein binding sites with bilirubin.

References


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*SPECT, Magnetic Resonance and Angiography in Moyamoya Disease* • Ohashi et al