Evaluation of Regional Cerebral Blood Flow in Massive Intracerebral Calcifications

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Fahr's disease is histopathologically characterized by massive bilateral calcifications of the cerebral basal ganglia, the dentate nuclei of the cerebellum and both the cerebral and cerebellar cortices. We report a case of Fahr's disease in which a ^{99m}Tc-hexamethyl-propylenamine oxime (^{99m}Tc-HMPAO) brain SPECT study was used to evaluate regional cerebral blood flow to the calcified regions. There was markedly decreased perfusion to the basal ganglia bilaterally as well as decreased perfusion to the cerebral cortices that correlated well with the patient's clinical condition.

Key Words: Fahr's disease; technetium-99m-HMPAO; single-photon emission computed tomography

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Bilateral symmetrical striopallidodentate calcinosis, also known as Fahr's disease, is a rare idiopathic neurological disorder characterized by extensive symmetrical calcifications of the basal ganglia, dentate nuclei of the cerebellum and both the cerebral and cerebellar cortices. Patients have various clinical presentations, but most patients commonly display extrapyramidal and cerebellar dysfunctions, speech difficulties, dementia and neuropsychiatric symptoms. These intracranial calcifications may be detected by conventional skull radiographs and computerized tomography (CT) (*1*).

SPECT of the brain with ^{99m}Tc-hexamethylpropylenamine oxime (^{99m}Tc-HMPAO) is useful in demonstrating regional cerebral blood flow. We present a case of Fahr's disease with massive intracerebral calcifications in which the ^{99m}Tc-HMPAO SPECT study demonstrates markedly decreased blood flow to the basal ganglia as well as decreased perfusion of cerebral cortical regions.

CASE REPORT

A 43-yr-old male with a history of Fahr's disease was admitted for generalized weakness and inability to care for himself. He had finished high school and served 3 yr in the armed forces without neurologic difficulty. After discharge, however, he began developing speech problems and became a "slow thinker". Skull radiographs and CT of the head in May 1987 disclosed diffuse symmetrical intracranial calcifications in the cerebrum, cerebellum and basal ganglia. Mini-mental status examination revealed a score of 16 of 30, indicating moderate cognitive impairment. Neurological examination showed intact cranial nerves and no focal motor or sensory deficits. There was evidence of cerebellar dysfunction with ataxia, dysarthria and basal ganglia dysfunction as evidenced by bradykinesia. Serum calcium, phosphorus and parathormone levels were all within normal ranges. An EEG showed no focal abnormalities. A CT scan revealed pronounced symmetrical calcifications in the basal ganglia, thalamic nuclei, cerebral and cerebellar white matter. Moderate diffuse atrophic changes and dilatation of both the lateral and third ventricles were also reported (Fig. 1).

SPECT scintigraphy was performed 1 hr after intravenous injection of 1066 MBq (28.8 mCi) ^{99m}Tc-HMPAO. Images were obtained with a triple-head dedicated SPECT camera fitted with high-resolution parallel-hole collimators. The average sensitivity of each collimator is 3 cps/μCi. Projections (128) were obtained 45 sec per view on an 128 × 128 matrix over 360° by rotating each head 120° for a total time of 36 min.

Attenuation correction (Sorenson method) and Metz filter (based on a Gaussian line spread function with FWHM of 9 mm and order of four) were applied prior to the generation of 8-mm thick slices in the coronal, sagittal and transaxial planes. For clinical studies, the FWHM resolution is 9–10 mm. SPECT images showed moderate to severely decreased uptake of radiotracer in the mid and posterior aspects of the frontal lobes bilaterally. There was markedly decreased uptake in the basal ganglia bilaterally, especially the caudate nuclei and to a lesser extent the thalamic nuclei. Decreased uptake was also reported in the superior and posterior portion of the right parietal lobe (Fig. 2).

DISCUSSION

In 1855, Bamberger described the presence of bilateral symmetrical calcifications of the basal ganglia histologically (*2*). In 1930, Fahr described an adult case with the typical clinical and histological findings of this syndrome (*3*). Idiopathic bilateral symmetrical striopallidodentate calcinosis (Fahr's disease) is characterized histopathologically by extensive calcifications of the globus pallidus, putamen, caudate nucleus, internal capsule, the lateral parts of the thalamus and the dentate nuclei of the cerebellum. Smaller concretions are also present at the junction of

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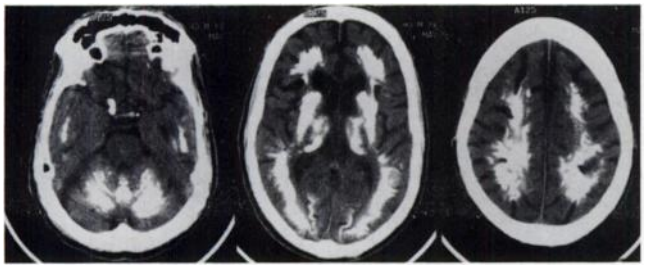

FIGURE 1. CT scan shows symmetrical calcifications in the basal ganglia, thalamic nuclei, cerebral and cerebellar white matter. Diffuse atrophic changes and ventricular dilatation also are present.

the cortex and white matter at the bases of the cerebral sulci, and in parts of the cerebellar cortex (*4*). The onset of clinical symptoms usually occurs in the fourth and sixth decades of life, although it may also be evident in childhood. Clinical signs and symptoms may be quite variable and include: parkinsonism, mental deterioration, speech impairment and rarely seizures. This disorder is thought to be inherited, although all cases do not exhibit this pattern (*5*).

CT is considerably more sensitive than skull x-rays to detect intracerebral calcifications. MRI provides better anatomical detail than CT, but is less sensitive in detecting calcification (*6*). Small calcifications in the basal ganglia and, less commonly, the dentate nucleus of the cerebellum, without associated symptoms are frequently seen in elderly patients. Calcium deposition in these patients are thought to be related to vascular changes associated with aging (*7*). Numerous other conditions are also associated with radiologically identified basal ganglia and dentate nucleus calcifications. The most frequent association is with hypoparathyroidism of spontaneous origin; they are rarely seen following parathyroidectomy. The calcifications in pseudohypoparathyroidism are radiographically indistinguishable. Diseases associated with scattered intracerebral calcifications such as tuberous sclerosis, toxoplasmosis and, rarely, cranial irradiation, birth anoxia, lead and carbon monoxide poisoning, congenital neurological disorders such as Cockayne's syndrome (a rare form of truncal dwarfism with retinal atrophy) are among the causes of radiologically identified basal ganglia and dentate nucleus calcifications (*8*).

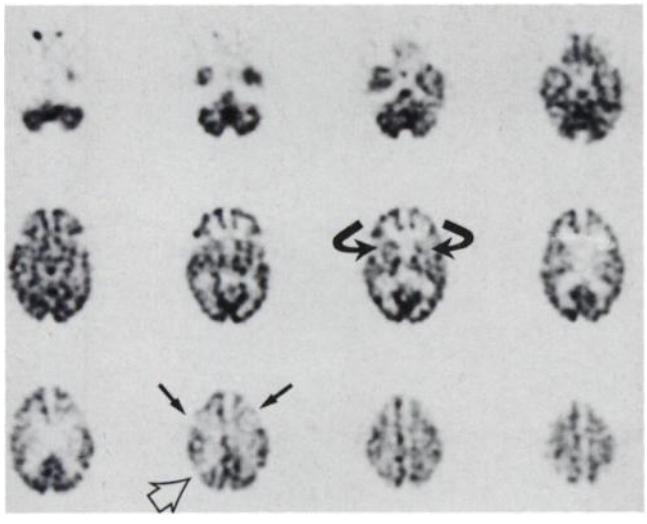

FIGURE 2. Technetium-99m-HMPAO brain SPECT shows moderate to severely decreased radiotracer uptake in the mid and posterior aspects of the frontal lobes (straight arrows). There is markedly decreased uptake in the basal ganglia especially in the caudate nuclei bilaterally (curved arrows). Decreased uptake is also noted in the superior and posterior portion of the right parietal lobe (open arrow).

The functional nature of brain SPECT imaging complements anatomical imaging studies such as CT and MRI. Brain SPECT abnormalities are often present earlier than the abnormalities seen on anatomical imaging studies. The detection of perfusion abnormalities with brain SPECT imaging can potentially lead to an early diagnosis and assist with clinical management. Perfusion deficits involving the basal ganglia and related structures on brain SPECT scans were reported in movement disorders such as Huntington's disease, Parkinson's disease, progressive supranuclear palsy, Wilson's disease, spasmotic torticollis and hemiballismus (*9*). Smith et al. reported a patient with Fahr's disease who showed prominently decreased basal ganglia HMPAO uptake matching the distribution of calcifications observed on CT. They did not, however, report cortical perfusion abnormalities (*10*). In our patient, the brain SPECT study showed decreased blood flow to the frontal lobes and the right parietal lobe besides the basal ganglia. These cortical perfusion changes may reflect interruption of pathways between the basal ganglia and the cortex as a result of calcification in the white matter. Cortical perfusion changes in patients with basal ganglia and white matter calcifications may better explain the patients' altered cognitive and motor functions.

In conclusion, SPECT imaging of the brain with ^{99m}Tc-HMPAO can be a useful tool in demonstrating regional cerebral blood flow and function in patients with conditions in which there is basal ganglia calcification.

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