Evaluation of Regional Cerebral Blood Flow in Fahr Disease with Schizophrenia-Like Psychosis: A Case Report

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Summary: We report the case of a patient with Fahr disease presenting with schizophrenia-like psychosis for whom a technetium Tc99m methyl cysteinate dimmer brain study was used to evaluate regional blood flow. A brain single photon-emission CT study showed increased cerebral blood flow to the bilateral temporal lobes. Such perfusion abnormalities may reflect psychotic symptoms, including auditory hallucinations and delusions, which suggests a disruption of the cortico-subcortical neural circuits in psychosis.

Idiopathic calcification of the basal ganglia (ICBG), also known as Fahr disease, is a rare neurologic disorder of unknown etiology characterized by neuropsychiatric abnormalities, Parkinsonian or choreoathetotic-type movement disturbance, and extensive symmetrical calcification of the basal ganglia and dentate nuclei in the cerebellum. These symptoms cannot be explained by any particular disorder of the calcium phosphorus metabolism or other diseases.

Dementia is a well-recognized neuropsychiatric manifestation of ICBG that is manifested as memory disturbances, deterioration of intellectual function, and a slowing of cognitive processes. In addition, a schizophrenia-like psychosis characterized by paranoia, hallucinations, and delusions has been reported (1). The pathophysiology of psychosis in ICBG remains unknown, though previous studies have found a decreased cerebral blood flow matching the distribution of calcification or decreased perfusion in the cortex, which may reflect secondary deficits due to calcification (2–4).

In the present study, we report the case of a patient with Fahr disease presenting with a schizophrenia-like psychosis for whom a technetium Tc99m methyl cysteinate dimmer (Tc99m-ECD) brain study was used to evaluate cerebral blood flow.

Case Report

A 32-year-old, right-handed woman was referred from a local hospital for an evaluation of schizophrenia-like psychosis for whom a technetium Tc99m methyl cysteinate dimmer brain study was used to evaluate regional blood flow. A brain single photon-emission CT study showed increased cerebral blood flow to the bilateral temporal lobes. Such perfusion abnormalities may reflect psychotic symptoms, including auditory hallucinations and delusions, which suggests a disruption of the cortico-subcortical neural circuits in psychosis.

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The patient was injected with 600 MBq (16 mCi) of Tc99m-ECD 5 minutes before scanning. Scanning was performed by using a dual-head gamma camera (GE Yokokawa Millennium VG), with the acquisition parameters set at 120 projections recorded in step-and-shoot mode over a 360° rotation arch (each head acquired 60 projections). The angular step was 3° and the frame time was 17 seconds per step, whereas the acquisition matrix was $64 \times 64$ and zoom set at 2.2. All projection data were prefiltered through a Butterworth filter (cutoff at 0.5), which resulted in axial image sections sized at $64 \times 64 \times 8$ mm. SPECT images showed increased cerebral blood flow in the bilateral lateral temporal lobes and decreased cerebral blood flow in the basal ganglia (Fig 3).

About 4 weeks later, the auditory hallucinations and delusions disappeared. The patient was discharged from our hospital and treated on an outpatient basis with low-dose risperidone.

**Discussion**

We treated a patient with Fahr disease who presented with a schizophrenia-like psychosis. CT and MR imaging findings revealed a bilateral calcification of the putamen and globus pallidus consistent with Fahr disease, along with frontal brain atrophy.

Hypoparathyroidism, pseudohypoparathyroidism,
and hyperparathyroidism were denied on the basis of normal levels of serum calcium and phosphorus metabolism and lack of Albright osteodystrophy. Calcifications following congenital, infectious, traumatic, ischemic, neoplastic, vascular, and toxic brain insults also were not seen. The patient was able to acquire a driving license at 20 years of age. On the assumption that her obsessional and introverted personality was associated with the disease, calcification may have existed at an early age, which led to the development of progressive dementia.

Presenile cortical dementia with Fahr-type calcification can be diagnosed from the presence of diffuse neurofibrillary tangles with calcification (DNTC) (5). In the present case, DNTC was unlikely, because of the early-stage onset and lack of temporal lobe atrophy. Brain SPECT images were acquired when the patient was showing delusion, auditory hallucinations, and deterioration of intellectual function, and previous studies have reported decreased perfusion abnormalities in patients with dementia. Smith et al (3) reported a patient with Fahr disease who showed a perfusion deficit in the basal ganglia, whereas Uygur et al (4) described a patient with Fahr disease who had decreased blood flow in the basal ganglia and frontoparietal cortex. The latter patient was 43 years of age and showed dementia, as well as cerebellar and basal ganglia dysfunction. Further, Hempel et al (2) reported a 25-year-old patient with Fahr disease who presented with frontal lobe syndrome and dementia. In that case, reduced glucose uptake in positron-emission tomographic images was not restricted to the basal ganglia, but also involved the frontal, temporal, and parietal cortices. Others have reported cases with hypoperfusion in both thalami (6) and low metabolic changes of the corpus striatum and posterior cingulate (7). In addition, previous studies have found perfusion deficits in the basal ganglia matching the distribution of calcifications in the cortex that may reflect secondary deficits due to calcifications of the basal ganglia. To the best of our knowledge, hyperperfusion in the lateral temporal lobe cortex has not been reported in a patient with Fahr disease.

Several neuroimaging studies have detected hyperactivity in the temporal region along with auditory hallucinations in schizophrenic patients. A magnetencephalography study (8) demonstrated left superior temporal cortex activations including the auditory area, suggesting that auditory hallucinations reflect an abnormal activation of the auditory pathways. Activation, including the bilateral temporal region, was also detected in a functional MR imaging study (9), whereas, recently, Andreasen et al (10) proposed that a disruption in the cortical-thalamic-cerebellar-cortical circuit leads to a disconnection syndrome, termed “cognitive dysmetria,” and impairment in this neural circuit produces diverse symptoms of schizophrenia.

The present findings suggest that the basal ganglia impairment in ICBG led to abnormal hyperactivity in the cortex mediated by the cortico-subcortical neural network, which generated psychotic symptoms. Although we attempted to demonstrate regional cerebral blood flow (rCBF) findings associated with hallucinatory experiences or delusions, there are limitations to the design of our study. In the present patient, haloperidol was administered first for about 4 weeks, followed by risperidone for another 4 weeks. The rCBF evaluation was performed under the use of antipsychotic medications and the patient was unable to self-report auditory hallucinations; however, disorganized behavior and delusions were identified at the time of the SPECT examination. The increase in rCBF in the lateral temporal cortex may have reflected her pathologic experiences, including the auditory hallucinations.

Neuroleptics are considered to inhibit cortex activities. One SPECT study of schizophrenia patients found that improvements after risperidone treatment were associated with a decrease in frontal and temporal activities, as well as a reduction in rCBF in the entire cortical area (11). The rCBF increase in the lateral temporal cortex in the present patient could not be explained by the effects of neuroleptics, though it may have reflected hyperactivation in this area that was related to pathologic experiences, such as the delusions or auditory hallucinations.

**Conclusion**

We treated a patient with Fahr disease who presented a schizophrenia-like psychosis. A brain SPECT study showed increased blood flow in the bilateral temporal lobes. The perfusion abnormalities may have reflected hyperactivation in the cortex related to auditory hallucinations or delusions and supports the theory that a disruption of the cortico-subcortical neural circuits produces psychotic symptoms.

**References**