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#### **Permalink**

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#### **Journal**

Psychosomatics, 46(6)

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#### **Publication Date**

2005-11-01

Peer reviewed

## Case Reports

# Paranoid Delusions and Cognitive Impairment Suggesting Fahr's Disease

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*The authors present the case of a 60-year-old woman with elaborate paranoid delusions and cognitive impairment found during a workup for atypical chest pain. Clinical evaluation revealed mild dementia, and radiography showed basal ganglia calcification consistent with Fahr's disease. She was treated with risperidone and transferred to a psychiatric inpatient unit for definitive care. Psychiatrists should consider Fahr's disease as a differential diagnosis in the evaluation of psychosis and cognitive impairment when neuroimaging reveals calcification of the basal ganglia.*

(Psychosomatics 2005; 46:–)

Fahr's disease, first described by Karl Theodor Fahr in 1930, refers to sporadic or familial idiopathic basal ganglia calcification that is associated with many neurological and psychiatric abnormalities.<sup>1</sup> Patients with Fahr's disease often appear with movement disorders, such as parkinsonism, paresis, dystonia, and speech impairment. Other neurological features can include stroke-like events, often combined with psychiatric conditions, such as psychosis, mood disorders, and dementia.<sup>2</sup> Although Fahr's disease appears most commonly with motor deficits, about 40% of the patients with Fahr's disease are seen with primarily cognitive and other psychiatric findings.<sup>3</sup> Fahr's disease, however, is often differentiated from Fahr's syndrome, in which basal ganglia calcification is secondary to some other disorder, such as hypoparathyroidism.<sup>2</sup> Fahr's disease should also be distinguished from incidentally found basal ganglia calcification without associated clinical neuropsychiatric features.<sup>3</sup>

The frequency of basal ganglia calcification apparent with computerized tomography (CT) is about 0.93% of 29,484 scans, but the true prevalence of Fahr's disease is unknown.<sup>3</sup> Psychiatric symptoms, as often seen in diseases of the basal ganglia, e.g., Parkinson's, Wilson's, and Huntington's diseases, are thought to be due to disruption of

cortical-subcortical circuits mediated by the basal ganglia. It has been well established that lesions in the orbitofrontal anterior cingulate system or the connecting lateral orbitofrontal circuit can produce socially inappropriate behavior, impulsivity, obsessive-compulsive and mood disorders, and personality change.<sup>4</sup> Subcortical lesions may appear with psychiatric symptoms without concurrent involvement of motor circuits that produce clinically significant motor symptoms. For example, Benke et al.<sup>5</sup> demonstrated hypometabolism of glucose in the basal ganglia and the frontal cortex of a neurologically "asymptomatic" patient with Fahr's disease characterized by basal ganglia calcification and psychiatric symptoms.

Controversy exists regarding the association of Fahr's disease with clinical symptoms. Förstl et al.<sup>6</sup> found the odds ratios for common neurological disturbances (that included dementia) in a cohort of 166 patients with basal

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ganglia calcification not to be significantly higher than that of comparison subjects and concluded that "basal ganglia calcification cannot be considered as a clinically relevant neuroradiological finding in the majority of cases and should not be used as an explanation for frequently observed neurological disturbances." However, López-Villegas et al.<sup>7</sup> studied 22 patients with basal ganglia calcification and found increased abnormal motor speed, executive function, visuospatial, and memory functions in patients with basal ganglia calcification; four had mood disorders and six others met diagnostic criteria for obsessive-compulsive disorder. López-Villegas et al. concluded that "patients with basal ganglia calcifications frequently have a subcortical pattern of neuropsychological dysfunction and behavioral changes that are known to be associated with alterations of the frontal-limbic-basal ganglia circuits. The pattern of neuropsychological impairment is consistent with basal ganglia damage."<sup>7</sup> Casanova and Araque<sup>8</sup> reviewed the literature on basal ganglia mineralization and found excessive mineral deposits to be associated with age-dependent progressive symptoms, including dementia, psychosis, and extrapyramidal symptoms. In addition to the basal ganglia, the dentate nucleus of the cerebellum is also often involved, although the extent of mineralization of these two areas has been shown to be different.<sup>9</sup>

The psychiatric manifestations of Fahr's disease are varied. The most common are mood disorders occurring in one-fifth to one-third of patients.<sup>3</sup> Anxiety disorders are also prominent, with one-third of the patients with Fahr's disease seen with obsessive-compulsive disorders.<sup>3</sup> Other manifestations include cognitive disorders, appearing usually as progressive subcortical dementia and psychoses with atypical features, such as perceptual distortions.<sup>3</sup> There is little literature on psychiatric symptom profiles and their treatment in Fahr's disease.<sup>3</sup> Given the paucity of reports, we present the case of a woman with Fahr's disease and her response to treatment.

### *Case Report*

Ms. A, a 62-year-old married Caucasian woman, came to the hospital with vague reports of chest pain, which she described as "electric shocks." She also reported multiple paranoid delusions. She was sure that people were controlling her by manipulating electrical dials. She was so certain that there were microphones in her home and that her activities were being recorded that she made several

service calls to the public utility company to complain. She explained that "electricity in the air and flickering lights" were responsible for both her "muscle spasms" and chest pain. She reported that unknown other people were "listening through the walls" of her home. She had taken to sleeping on the floor because of fears that her bed was "magnetized." Her paranoia had increased since the terrorist attacks of Sept. 11, 2001, when her daughter was going to work as a flight attendant. Furthermore, Ms. A's worsening paranoia was associated with increasingly reclusive behavior. Notably, about the time of the terrorist attacks, her spouse was hospitalized after a motor vehicle accident.

In the emergency department, Ms. A became agitated, removed her intravenous line, and attempted to leave against medical advice. After an exhaustive workup, including a negative ECG, cardiac enzyme tests, and an exercise tolerance test, a medical cause of her chest pain was ruled out. An incidental urinary tract infection was detected and appropriately treated with a course of antibiotics. Her neurological examination at admission was unremarkable; in particular, no tremor, ataxia, weakness, abnormal reflexes, or lack of coordination were noted. A formal neurological consultation was not obtained. The results of laboratory studies included normal serum chemistry levels, a mildly elevated glucose level (she had a history of type II diabetes mellitus), and normal levels of thyroid-stimulating hormone, B<sub>12</sub>, folate, and phosphate. Her serum calcium was not measured.

Ms. A's past psychiatric history was remarkable for an episode 2 years before, when she came to an inpatient psychiatric hospital accompanied by a close family member because of ideas of reference, paranoid delusions, auditory hallucinations, and occasional suicidal ideation, which had reportedly begun 6 months before. At that time, collateral history from a family member revealed that Ms. A had been admitted to a psychiatric hospital more than 20 years ago, but further details were not available from collateral sources, and Ms. A would not elaborate on this earlier admission.

A mental status examination revealed the following: paranoid delusions of being controlled by others and electricity threatening her, mild lability, a nontearful affect, mild psychomotor agitation, and a tangential thought process. Cognitive impairment was detected with a Mini-Mental State Examination (MMSE) score of 20 of 30, with notable deficits in recall and attention. Ms. A had no suicidal or homicidal ideation and no hallucinations. She was given a low dose of oral risperidone, 0.5 mg b.i.d., for her psychotic symptoms. Because of her cognitive impairment,

a CT scan of her brain was done, with a finding of bilateral symmetric basal ganglia calcifications (Figure 1). Because Ms. A's delusions interfered with her ability to take care of herself, she was admitted to an inpatient psychiatric facility after medical clearance.

After her psychiatric admission, she maintained that electricity was "controlling her" and was fearful that "electromagnetic fields" in her apartment were dangerous to her. She also described persecutory delusions of neighbors bothering her in "subtle ways," and she felt threatened by the psychiatric hospital staff. She reported no history suggestive of mania or depression. She reported no drug or alcohol use in both the remote and recent past. Upon a repeat mental status examination, Ms. A appeared mildly unkempt and disheveled. She was partially cooperative after a short period of distrust of the hospital staff. Her mood was mildly irritable and anxious; her affect was restricted and congruent with her mood. Her speech was rapid but not pressured, her thought process tangential, her insight poor, and her judgment moderately impaired. Her cognition was reassessed with a repeat of the MMSE, on which she scored 22 of 30, with notable deficits in recall, attention, and copying.

**FIGURE 1.** A Computerized Tomography Scan Showing Hyperdensities Suggestive of Bilateral Symmetric Calcifications in the Globus Pallidus of a Woman With Possible Fahr's Disease



Ms. A stayed at the inpatient psychiatric hospital for 3 days to stabilize her psychosis and delusions. Although her delusions improved mildly, she was still paranoid regarding the hospital staff and continued to be concerned about the electricity in her apartment. Her risperidone dosage was increased to 3 mg at bedtime, and she was discharged with this dosage. Upon discharge, she was referred to outpatient psychiatry and neurology for further evaluation of her mild dementia and CT findings.

### Discussion

Although we did not rule out unlikely causes for the patient's basal ganglia calcifications, such as toxoplasmosis, mitochondrial encephalopathy, tuberous sclerosis, idiopathic hemochromatosis, myotonic muscular dystrophy, and disorders of calcium metabolism, she had no other significant symptoms or history warranting such a workup. Although Fahr's syndrome could not be ruled out, it is likely that the patient did indeed suffer from Fahr's disease. The patient has the most common distribution of calcification in Fahr's disease, which is limited to the globus pallidus.<sup>3</sup> She did not have calcification of the dentate nucleus of the cerebellum. Psychiatric symptoms in Fahr's syndrome and Fahr's disease (as well as other disorders of the basal ganglia) are most likely mediated by disruption of functioning of the basal ganglia rather than other effects of the primary disorder,<sup>2</sup> thus the distinction may be irrelevant.

Psychotic symptoms seen in Fahr's disease include auditory and visual hallucinations, complex perceptual distortions, delusions, and fugue states.<sup>3</sup> They were manifest in this patient as paranoia and visual and tactile hallucinations. It is likely that the psychosis in both Fahr's disease and schizophrenia share a similar pathology, although the particular lesions causing schizophrenia are ill defined. Positive psychotic symptoms, hallucinations, and paranoia are not necessarily generated by the classical hypothesis of dopamine-mediated attachment of salience to internally generated stimuli.<sup>10</sup> Still, there is some evidence that disruption of the cortex involved in the pathophysiology of schizophrenia is also seen in Fahr's disease, particularly in areas of the limbic system.<sup>2,5</sup>

There are few data to guide clinicians on appropriate psychotropic treatment of psychiatric symptoms in Fahr's disease.<sup>3</sup> This patient was exposed to antipsychotic treatment for 4 days and was noted to improve mildly. Too little

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time had passed to assess the effectiveness of her response to risperidone, and she has since been lost to follow-up. This patient showed no evidence of extrapyramidal side effects to antipsychotic treatment, although there is limited evidence in the literature that patients with Fahr's disease are at greater risk for extrapyramidal side effects. In one case series, four of seven patients with basal ganglia calcification were prone to extrapyramidal side effects with treatment with typical antipsychotics.<sup>11</sup> Moreover, extrapyramidal movement disorders occur frequently in patients with Fahr's disease, as many as 56% of 213 cases in one review.<sup>12</sup> Although more studies need to be done, clinicians

should be advised of this risk and preferentially use atypical antipsychotics for psychotic symptoms and delirious or psychotic agitation.

Clearly, future studies should be performed because the prevalence of bilateral basal ganglia calcifications in patients with new-onset psychosis could be estimated with data obtained with routine neuroimaging of new cases of psychotic and dementing illness. This case, along with others in the literature, further emphasizes the importance of the role of neuroimaging in any patient with psychosis and/or dementia to rule out structural causes of neuropsychiatric phenomena.

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