PubMed

ISSN - 0972-2327

Volume 17, Issue 4, October 2014

Annals of Indian Academy of Neurology

www.annalsofian.org

Official Publication of Indian Academy of Neurology

Wolters Kluwer Health | Medknow

Indexed with EMBASE and
Science Citation Index (E)

Pure psychiatric presentation in a patient with Fahr's disease

Rosaria Renna, Francesca Plantone[1], Domenico Plantone

Department of Neurology, Catholic University of Sacred Heart, Rome, [1]Department of Surgical Sciences, Section of General Surgical Clinics and Surgical Therapy, Parma University Medical School, Parma, Italy

For correspondence:
Dr. Rosaria Renna, Department of Neurosciences, Institute of Neurology, Catholic University of Sacred Heart, Largo Gemelli 8,00168 Rome, Italy.
E-mail: rosrenna@yahoo.it

Ann Indian Acad Neurol 2014;17:466-7

A 69-year-old Caucasian man presented with a 2-month history of anxiety, irritability, apathy, sadness, visual hallucination, suicidal ideation, and paranoid delusion. He suffered from hypertension and denied any previous psychiatric or neurological symptoms and any recreational drug use. He smoked about 20 cigarettes per day and consumed alcohol sparingly. The patient had no family history of neurological or psychiatric disease. He did not report any infectious disease, exposure to toxic substances or other significantly traumatic events in his life. Neurological examination revealed normal orientation to place and time with normal level of consciousness. There were no dysarthria, dysdiadochokinesia, ataxia, spasticity, and no evidence of any extrapyramidal signs. Brain CT scan revealed bilateral and symmetric cerebral calcifications involving the caudate nucleus, lentiform nucleus, thalamus, and cerebellar dentate nucleus [Figure 1]. Blood levels of calcium, phosphorus, glucose, iron, ferritin, lactate, vitamin D, calcitonin and parathormone, phosphaturia, and calciuria were all within normal limits. Biochemical and somatic features did not support a diagnosis of metabolic or mitochondrial diseases, or other systemic disorders. The brain CT scan findings of bilateral calcifications of basal ganglia, thalamic, and cerebellar nuclei in a clinical context of progressive psychotic symptoms (visual hallucination, lability of mood, irritability, and paranoid delusion) suggested the diagnosis of Idiopathic Basal Ganglia Calcification (IBGC).

Bilateral and symmetric basal ganglia calcification has been described in a variety of diseases, such as primary hypoparathyroidism, mitochondrial encephalopathies, Alzheimer's disease, myotonic muscular dystrophy, and lupus.[1,2] If the cause is not identified, the condition is termed IBGC, also known as Fahr's disease. Blood calcium and parathormone levels, in addition to the other routine blood tests, are useful to distinguish IBGC, characterized by unremarkable laboratory test results, from secondary forms. IBGC was first described by German neurologist Karl Theodor Fahr in 1930 and represents a rare neurodegenerative disease. The usual age of onset is 4th–5th decade, albeit it can present also during childhood or adolescence. Patients with IBGC may present with multiple neurological and psychiatric symptoms, such as rigidity, hypokinesia, tremor, choreoathetosis, ataxia, cognitive impairment, seizures, psychosis, or mood disorder.[3] In some cases, IBGC may remain asymptomatic for the entire life. Psychiatric symptoms are present in 20-30% of all patients;[3] however, a pure psychiatric presentation in adulthood is rare, but possible.[4] Familial and non-familial cases have been described with predominance of the autosomal dominant inheritance in the familial ones.[5]

Interestingly, neuropsychological deficits in a patient with Fahr's disease, frontal lobe type dementia and hyperkinetic-

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	DOI: 10.4103/0972-2327.144041

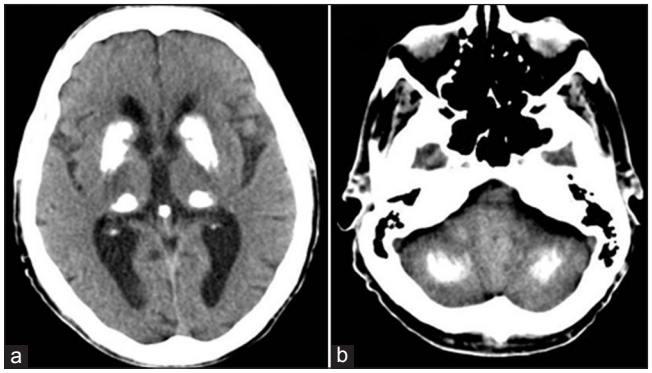

Figure 1: Brain CT scan: Brain non-contrast transaxial CT scan demonstrating dramatic bilateral and symmetric calcifications in caudate nucleus, lentiform nucleus, thalamus (a), and cerebellar dentate nucleus (b)

hypotone syndrome have been correlated with positron emission tomography findings of reduced glucose uptake not only confined to the basal ganglia, but extending also to the frontal, temporal and parietal cortices, suggesting that these functional changes may underlie neuropsychiatric symptoms.[6]

The diagnosis of IBGC should be always taken into account in patients presenting with neuropsychiatric disturbances and, in the meanwhile, all patients with incidentally detected basal ganglia calcifications should undergo neuropsychiatric and biochemical evaluation.

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How to cite this article: Renna R, Plantone F, Plantone D. Pure psychiatric presentation in a patient with Fahr's disease. Ann Indian Acad Neurol 2014;17:466-7.
Received: 13-04-14, **Revised:** 08-05-14 **Accepted:** 09-05-14

Source of Support: Nil, **Conflict of Interest:** None declared