

Idiopathic basal ganglia calcification presenting as schizophrenia-like psychosis and obsessive-compulsive symptoms: A case report

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Abstract. Idiopathic basal ganglia calcification (IBGC) is a rare neurodegenerative disorder characterized by the deposition of calcium in the brain and variable combinations of movement disorders, gait impairment and neuropsychiatric symptoms. Few reports have described psychiatric manifestations as early symptoms of IBGC. The present study reports the case of a middle-aged man with schizophrenia-like psychosis and obsessive-compulsive symptoms as the first manifestations of IBGC. The response of the patient to olanzapine and fluoxetine suggests that low-dose olanzapine is effective and should be increased cautiously to avoid worsening parkinsonism and that fluoxetine is an effective drug for the treatment of obsessive-compulsive symptoms in IBGC.

Introduction

Idiopathic basal ganglia calcification (IBGC), also known as Fahr's syndrome, is a rare neurodegenerative disorder characterized by neuropsychiatric abnormalities, parkinsonian or choreoathetotic-type movement disturbances and extensive symmetrical calcification of the basal ganglia and dentate nuclei in the cerebellum (1). IBGC is a rare calcium metabolism disorder with high genotypic and phenotypic heterogeneity, with symptoms that cannot be explained by any particular disorder of calcium phosphorus metabolism, or other disease (2). There are no guidelines for the treatment of IBGC, and there are currently no reversible measures available to reduce the calcification. In the clinical environment, doctors often provide treatment for the symptoms, including pharma-

cologic treatment for the improvement of anxiety, depression and obsessive-compulsive behaviors. Sobrido *et al* (3) previously suggested that at-risk asymptomatic adult family members may seek genetic counseling, in order to make personal decisions regarding reproduction, financial matters, and career planning. Few reports have described psychiatric symptoms as early manifestations of IBGC. The present study reports a case of IBGC in a middle-aged male who first manifested schizophrenia-like psychosis and obsessive-compulsive symptoms without evident movement disorders. The response of the patient to treatment with olanzapine and fluoxetine is also presented. To the best of our knowledge, this is the first report of parkinsonism during treatment with olanzapine and the response to fluoxetine in IBGC. The patient provided written informed consent.

Case report

A 41-year-old, right-handed male of Han ethnicity, was admitted to the inpatient psychiatric service of the Second Affiliated Hospital of Zhejiang University School of Medicine (Hangzhou, China) with schizophrenia-like psychosis and obsessive-compulsive symptoms. For 1 month prior to admission, the patient had complained about auditory hallucinations which commented on him and commanded him to commit suicide; he also felt himself to be controlled by 'the devil'. The patient suspected that people were always discussing him and wanted to kill him. In addition, he had intrusive thoughts with repetitive behaviors. Defecation was a source of worry to the patient and he repetitively touched his anus, which he recognized to be irrational. In addition, the patient repeatedly checked whether doors were locked or lights were turned off, refused to go outside and stopped going to work.

According to the patient's father, the patient had displayed normal development. Since graduating from high school with poor grades, he had been working as a janitor. Family history was remarkable in that the patient's mother and grandmother had been diagnosed with parkinsonism. In addition, his mother's elder brother had dementia at 50 years of age and a younger brother had schizophrenia-like psychosis requiring treatment in a psychiatric hospital.

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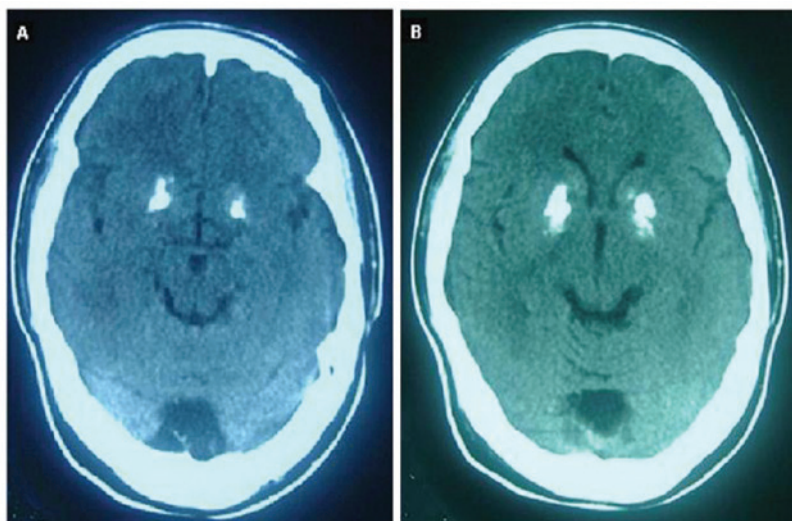


Figure 1. Brain computerized tomography scan exhibits bilateral hyperdensities in the (A) head of the caudate nucleus, and (B) the globus pallidus and putamen.

Neurological examination was essentially normal with the exception of a slight bilateral tremor in the hands and a slightly expressionless face. On mental status examination, the patient was oriented and cooperative but nervous. He had auditory hallucinations, delusions of reference and persecution, obsessive ideas, repetitive behaviors and difficulty in concentration and long-term recall. The patient was anxious and mildly depressed, but denied homicidal ideation or suicidal behavior. Psychological testing revealed a positive and negative syndrome scale (PANSS) score of 82, a Yale-Brown obsessive-compulsive scale symptom checklist (YBOCS) score of 25, and a Wechsler Adult Intelligence Scale-Chinese Revision IQ score of 55, including a verbal IQ of 54 and a performance IQ of 63.

Blood tests including hemocytology, liver and kidney function, glucose, calcium, phosphorus, parathyroid hormone, ceruloplasmin, antinuclear antibodies, thyroid hormone, vitamin B₁₂, folate and ammonia were all within the normal range. The results of the electrocardiogram, chest computerized tomography (CT), electroencephalogram, abdominal and thyroid ultrasonography were all normal. The brain CT scan revealed symmetrical calcification in the basal ganglia (Fig. 1).

The patient was therefore diagnosed with IBGC and was prescribed 20 mg fluoxetine and 5 mg olanzapine daily. The olanzapine dose was increased to 10 mg after 2 days. Two weeks later, the patient's obsessions were unchanged but the psychotic symptoms were absent; however, the patient showed a parkinsonian gait, hypomimia, muscle rigidity and evident tremors with a positive finger-to-nose test. His Unified Parkinson's Disease Rating Scale (UPDRS) motor score was 25, while his PANSS score was 42 and his YBOCS score was 24. The fluoxetine dose was then increased to 40 mg and the olanzapine dose was decreased to 5 mg. Two weeks later, parkinsonism and obsessions improved with a UPDRS score of 13 and a YBOCS score of 15. Treatment with the two drugs was continued for 3 months. The patient then had no psychosis but continued to have mild obsessions with a YBOCS score of 6 and parkinsonian symptoms. After the olanzapine dose was decreased to 2.5 mg, the parkinsonism improved to a

UPDRS score of 5. The neuropsychiatric condition of the patient remained stable during 3 months of follow up.

In summary, the patient with IBGC exhibited schizophrenia-like psychosis and obsessive-compulsive symptoms as the first manifestations that required hospitalization, and also showed mild dementia and parkinsonism. Brain CT confirmed IBGC. Metabolic diseases including hypoparathyroidism, pseudohypoparathyroidism and hyperparathyroidism were excluded on the basis of normal levels of serum calcium and phosphorus. No other significant symptoms or history suggested calcifications following a systemic disorder such as mitochondrial encephalopathy, tuberous sclerosis, Sturge-Weber syndrome and idiopathic hemochromatosis (4).

Discussion

The major clinical features of IBGC are movement disorders, which may be associated with cognitive deficits and psychiatric disturbances. There have been few reports of obsessive-compulsive symptoms in IBGC or psychiatric symptoms as the initial manifestation of IBGC. The patient in the current study initially presented with a schizophrenia-like psychosis and obsessive-compulsive symptoms. The brain abnormalities found in schizophrenia and obsessive-compulsive disorder (OCD) reveal the involvement of similar brain regions, including the frontal lobe, basal ganglia and thalamus (5). We considered both psychiatric symptoms to be shown simultaneously and linked to the basal ganglia pathology in this patient.

There are no guidelines for the treatment of IBGC, and drug therapy aims only to alleviate the symptoms. Few data on appropriate psychotropic treatment have ever been reported, and the use of neuroleptics is complicated by the risk of worsening the underlying movement disorders. Rosenblatt and Leroi suggested that high-potency neuroleptics or atypical antipsychotics could be used in IBGC (6). Shoyama *et al* reported that an IBGC patient with schizophrenia-like psychosis became well when treated with low-dose risperidone without worsening parkinsonism (1). Mishra *et al* used olanzapine 30 mg to treat an patient with IBGC and Capgras syndrome (7). The

patient in the current study clearly had parkinsonism after being treated with an olanzapine dose of 10 mg but improved when the dose was decreased to 2.5 mg, suggesting that the tolerance to antipsychotics of patients with IBGC may vary. The management of OCD in basal ganglia disorders follows the treatment for idiopathic OCD. Fluoxetine was the first selective serotonin reuptake inhibitor antidepressant and the effective dose range is 40-60 mg in OCD therapy (8). The current case was treated with fluoxetine 40 mg, which showed definite efficacy and was well tolerated.

The present case demonstrates that patients with IBGC may exhibit schizophrenia-like psychosis and obsessive-compulsive symptoms as the first manifestations of the condition. IBGC should be considered as part of the differential diagnosis in the evaluation of initial psychiatric symptoms, and brain CT is a useful adjuvant examination. Low-dose olanzapine is effective and should be increased cautiously to avoid worsening parkinsonism. Fluoxetine is an effective drug to treat obsessive-compulsive symptoms in IBGC.

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