

Idiopathic Basal Ganglia Calcification with Bipolar Mood Disorder Presentation

Paria Azari MD *

Fahr's disease is a progressive and idiopathic basal ganglia calcification with normal metabolism of calcium and phosphorus with motor and psychiatric signs and symptoms. Dementia, chorea athetosis, psychosis and depression due to Fahr's disease are frequently reported, but Fahr's disease with bipolar mood disorder manifestation is very rare and we found only 3 cases in review of literature from 1955 to 2005. In this case report, a 21-year-old girl is presented who was admitted to Sari-Zare psychiatric hospital for aggression, restlessness and insomnia. After mental status examination and Para clinical investigation, bipolar disorder due to Fahr's disease was detected. To date no specific treatment was found for this disease. This point is important that the patients with Fahr's disease are sensitive to neuroleptic malignant syndrome.

Iranian Journal of Psychiatry and Behavioral Sciences (IJPBS), Volume 1, Number 1, Spring and Summer 2007: 36-38.

Keywords: Basal Ganglia Diseases • Bipolar disorder • Fahr's disease

Introduction

Fahr's disease, first described by Karl Theodore Fahr in 1930, refers to sporadic or familial idiopathic basal ganglia calcification that is associated with many neurological and psychiatric abnormalities (1). 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 (2). 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 (3). Organic mood disorder has been reported less often, and mania secondary to idiopathic basal ganglia calcification has been noted rarely (4). The definition requires bilateral calcifications with neuropsychiatry and extra pyramidal disorders attended by normal calcium and phosphorus metabolism. CT-Scan is an easy

exam and has greater diagnostic specificity for basal ganglia calcification. About 40% of patients with BGC present initially with psychiatric features (5). Cognitive, psychotic, and mood disorders are common. Clinical features are important because BGC may be viewed as an incidental finding. Headache, vertigo, movement disorders, paresis, stroke like events, cognitive impairment, psychiatric disorders, pyramidal signals and seizures are the most common manifestations (5,6). Symptomatic features may change over time (7,8).

Case presentation

A 21-year-old girl was admitted in Zare-psychiatric hospital with 2-month history of insomnia, irritability, restlessness and ritualistic behavior. In previous psychiatric history, she had two episodes of illness, the first, in 3 years ago she suffered from depressed mood, insomnia, decreased appetite that was changed to the second episode with manic presentation and she was admitted in a psychiatric hospital. During the mental status examination, psychomotor agitation, pressure of speech, irritable mood and appropriate affect, poor concentration and obsessive thinking were detected. In laboratory investigation, serum concentration of calcium and phosphorus, are within normal limits. Routine hematological and biochemical investigations, as well as workup for metabolic,

Author's affiliation : * Psychiatry and Behavioral Sciences Research Center and Department of Psychiatry, Mazandaran University of Medical Sciences and Health Services, Sari, Iran

• **Corresponding author** : Paria Azari MD, Psychiatrist and researcher of Psychiatry and Behavioral Sciences Research Center and Department of Psychiatry, Mazandaran University of Medical Sciences and Health Services, Sari, Iran
Tel : +98 151 3283886
Fax : +98 151 3285109
E-mail: pariaazari@yahoo.com

inflammatory, and infectious conditions, do not disclose specific abnormalities. In computed tomographic scan, bilateral calcification of basal ganglia was detected (fig 1).

Discussion

Mood disorders include mania and depression are the most common behavioral changes in BGC, initially present in one-fifth, eventually occurring in about two-thirds, and occurring more commonly than in neuropsychiatry control subjects (9). Of 58 patients, mania eventually occurred in up to 31% (10). Perhaps up to one-half of patients develop depression at some point over the illness course. One study found DSM-III depressive disorders in 37% (5), and another found DSM-III-R minor depressive symptomatology in 16.7% (10). More extensive calcification and subarachnoid space dilatation correlate with the presence of psychiatric manifestations (5,10,11), but calcify distribution and etiology does not (5). Both the symptoms of mood disorders and biological research findings support the hypothesis that mood

disorders involve pathology of the limbic system, the basal ganglia, and the hypothalamus. The limbic system and the basal ganglia are intimately connected, and the limbic system may well play a major role in the production of emotion. Structural brain imaging studies support the notion that mood disorders are associated with regional structural brain abnormalities; in particular regions involved in mood regulation, such as the limbic portion of the basal ganglia and brainstem structures (12).

References

1. Fahr T. [Idiopathic calcifying of the cerebral vessels] Idiopathische verkalkung der hirngefäÙe. Centralblatt für allgemeine Pathologie und pathologische Anatomie 1930; 50:129-33. German.
2. Ring HA, Serra-Mestres J. europsychiatry of the basal ganglia. J Neurol Neurosurg Psychiatry 2002; 72: 12-21.



Fig 1 : Bilateral calcification of basal ganglia in the CT Scan

3. Lauterbach EC, Cummings JL, Duffy J, Coffey CE, Kaufer D, Lovell M, et al. Neuropsychiatric correlates and treatment of lenticulostriatal diseases: A review of the literature and overview of research opportunities in Huntington's, Wilson's, and Fahr's diseases. *J Neuropsychiatry Clin Neurosci* 1998; 10:249-66.
4. Trautner RJ, Cummings JL, Read SL, Benson DF. Idiopathic basal ganglia calcification and organic mood disorder. *Am J Psychiatry* 1988; 145: 350-3.
5. Konig P. Psychopathological alterations in cases of symmetrical basal ganglia sclerosis. *Biol Psychiatry* 1989; 25: 459-68.
6. Levy G. Basal ganglia calcification (striopallidodentate calcification) and cognitive impairment. *Arq Neuropsiquiatr* 1999; 57: 148.
7. Lauterbach EC, Spears TE, Prewett MJ, Price ST, Jackson JG, Kirsh AD. Neuropsychiatric disorders, myoclonus, and dystonia in calcification of basal ganglia pathways. *Biol psychiatry* 1994; 35: 335-44.
8. Rosenberg DR, Neylan TC, EL-Alwar M, Peters J, Van Kammen DP. Neuropsychiatric symptoms associated with idiopathic calcification of the basal ganglia. *J Nerv Ment Dis* 1991; 179(1): 48-9.
9. Forstl H, Krumm B, Eden S, Kohlmeyer K. What is the psychiatric significance of bilateral basal ganglia mineralization? *Biol Psychiatry* 1991; 29: 827-33.
10. López-Villegas D, Kulisevsky J, Deus J, Junqué C, Pujol J, Guardia E, et al. Neuropsychological alterations in patients with computed tomography-detected basal ganglia calcification. *Arch Neurol* 1996; 53(3): 251-6.
11. Taxer F, Haller R, Konig P. Clinical early symptoms and CT findings in Fahr syndrome. *Nervenarzt* 1986; 57:583-8.
12. Sadock BJ, Sadock VA editors. Mood disorders. In: *Synopsis of psychiatry*. New York: Lippincott williams & wilkins; 2003. p. 540.

Archive of SID