AN INTERESTING CASE OF PSYCHOSIS WITH BILATERAL BASAL GANGLIA CALCIFICATION

Sivaraman Subramanian[1], Ronald Roy Kingsly[2], Deepak Kumar[3], Prabakaran Govindaswamy[4]

[1]Associate Professor, Department of Psychiatry, Chennai Medical College Hospital and Research Centre, Irungalur, Trichy.
[2]Senior Resident, Department of Psychiatry, Chennai Medical College Hospital and Research Centre, Irungalur, Trichy.
[3]Senior Resident, Department of Psychiatry, Chennai Medical College Hospital and Research Centre, Irungalur, Trichy.
[4]Professor, Department of Psychiatry, Chennai Medical College Hospital and Research Centre, Irungalur, Trichy

ABSTRACT

BACKGROUND

A 31-year-old unmarried male presented with 3 months' duration of acute onset psychosis mimicking schizophrenia with few atypical features. On evaluation, we found an interesting finding of bilateral basal ganglia calcification in imaging study. Our literature review revealed the clinical features of the patient correlated with Fahr's disease (Idiopathic basal ganglia calcification). This case in turn sparked the importance of evaluation of organicity especially in patients presenting with atypical features of psychosis and the role of interdisciplinary approach in management of such cases.

KEYWORDS

Basal Ganglia Calcification, Psychosis, Fahr's Disease.

HOW TO CITE THIS ARTICLE: Subramanian S, Kingsly RR, Kumar D, et al. An interesting case of psychosis with bilateral basal ganglia calcification. Journal of Research in Psychiatry and Behavioural Sciences 2016; Vol. 2, Issue 2, July-December 2016; Page:12-15.

BACKGROUND

The term psychosis has historically received a number of different definitions, none of which achieved universal acceptance. The narrowest definition of psychosis is restricted to delusions or prominent hallucinations, with distorted reality in the absence of Insight.(1) Psychosis is not pathognomonic of primary psychiatric illness. It is simply a nonspecific cluster of signs and symptoms that may possibly occur in a broad array of medical, neurologic and surgical disorders or as a consequence of substance abuse, withdrawal of drugs or pharmacologic treatment.**(2)** So it is imperative that Psychiatrists and Primary care medicine physicians ought to differentiate psychotic symptoms caused by general medical or neurological conditions from psychosis caused by a primary psychiatric illness.

Psychoses are classified as organic or functional mental disorders. Psychosis presenting with organic conditions do not follow a definite pattern and are usually indistinct and nonconforming.(2) This makes the role of the clinician even more tedious and may cause a diagnostic dilemma which in further might impact the prognosis. Furthermore, the need for an interdisciplinary approach crops up in the form of general physicians, neurologists and psychiatrists uniting in liaison to evaluate and manage these kinds of conditions more effectively. As a fact, organic evaluation for psychotic symptoms is under estimated in clinical practice and a possibility of overlooking the symptoms due to a unidimensional approach might even cost the patient their lives.

Illustrating such complex clinical situations we have come across an interesting case of psychosis which we will be discussing in detail.

Financial or Other, Competing Interest: None.
Submission 25-08-2016, Peer Review 10-11-2016,
Acceptance 17-11-2016, Published 26-12-2016.
Corresponding Author:
Dr. Sivaraman Subramanian,
#1-1355 D, First Floor,
Vasan Nagar 15th Cross,
Vayalur Road, Trichy-620020,
Tamilnadu.
E-mail: sivaramanaiims@gmail.com

Case Vignette

A 31-year-old unmarried male, presented to outpatient department of tertiary care centre (Chennai Medical College Hospital & Research Centre) with acute onset complaints of muttering & Smiling to self, disturbed sleep, reduced appetite and food intake, suspiciousness & withdrawn behaviour. He was preoccupied with the thought that his mother, along with her relatives, was responsible in hindering his marriage plans and believed that all problems will be solved only if he gets married. He firmly believed his mother and relatives were plotting against him and were trying to harm him indirectly. His self-care deteriorated gradually, and he refused to interact with others. He complained of frequent headaches and multiple non-specific somatic symptoms. Later, he was noted laughing to self occasionally and was frequently hostile towards his mother. He became extremely suspicious and locked himself inside alone, never allowing anyone to come inside the house. Subsequently, he lost significant weight, talked irrelevantly, was inattentive and had memory disturbances to recent events. His symptoms were continuous, progressive and happened over the span of 3 months.

According to the patient's mother, he was her only son and had displayed normal early development. Since graduating from high school with average grades, he had been working as a labourer. He had a dysfunctional family with his father separated from them and lived alone. He was an occasional alcohol user. There was no family history of any psychiatric disorders.

On admission, the patient was emaciated with poor self-care and decreased psychomotor activity. He was seen muttering to himself and was distracted evidently. On establishing rapport, his conversations dwelled on topics regarding his marriage, inability to go to work and betrayal from mother & relatives. His affect was constricted, and his thought process was tangential. He had delusions of persecution and reference. There were no consistent hallucinations but had hallucinatory behaviours in the form of muttering. He was alert and oriented. Sustained attention was impaired along with recent memory impairment, but his other executive function tests were normal. His abstract thinking

was impaired and had absent insight. Except being thin & ill built, his physical, including neurologic examination, was unremarkable.

He was clinically diagnosed initially as a case of Psychosis unspecified according to ICD 10 diagnostic criteria provisionally and was prescribed Risperidone and clonazepam. His initial score on BPRS was 56.

On Initial Evaluation, complete blood count, basic metabolic panel, Urine routine screening, liver function tests done were normal. A general physician opinion was obtained in view of emaciation, recurrent headaches and recent memory impairment. On his advice, Thyroid function tests, CRP, ESR, Mantoux test, HIV and Hep-B screening and CT Brain were done with patient's consent. All these investigations were within normal limits except for Computed tomography (CT) scan of the brain which showed symmetrical bilateral basal ganglia calcifications.

Neurologist opinion was obtained where he stated that there were no focal neurological deficits or any extrapyramidal motor symptoms. On his advice, Peripheral smear, Serum calcium, Phosphate, Sr. ALP and PTH were evaluated but were found to be within normal limits.

With gradual titration of antipsychotics over a period of 3 weeks, patient had shown improvement with BPRS score dropping to 14. The diagnosis was revised as a case of Organic Psychosis and on literature review we considered the possibility of Fahr's disease (Idiopathic basal ganglia calcification) presenting with psychotic symptoms.

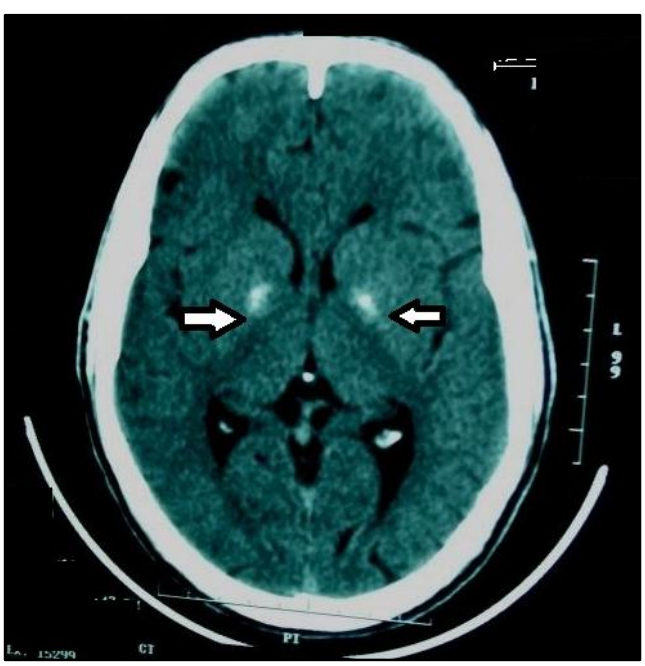

Figure 1

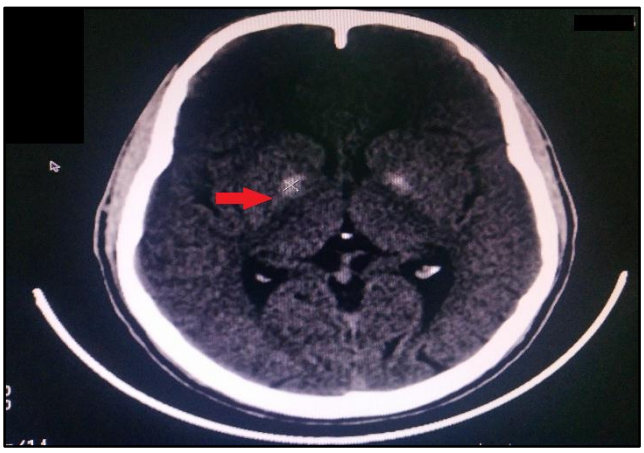

Figure 2

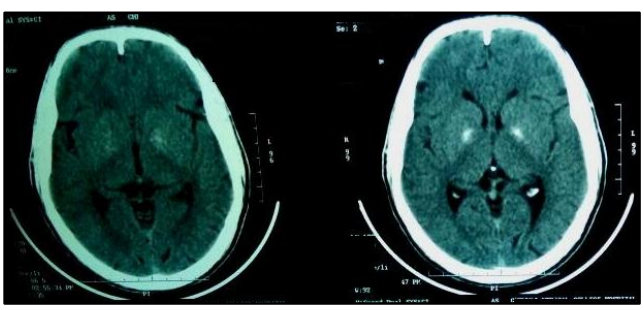

Figure 3

Figure 1, 2, 3. (CT Brain image) Arrows in the figures indicates Bilateral Basal Ganglia Calcification

DISCUSSION

Psychotic symptoms-delusions, hallucinations, disorganised speech and behaviour are observed in a wide range of organic or nonorganic medical or neurological conditions, and are, therefore, diagnostically nonspecific. Organic disorders with psychosis are caused by structural defects or physiologic dysfunction of the brain.[2]

Especially in times when psychobiological points of view in psychiatry seem the main focus of attention, one must not forget the primary organic root of many psychotic disturbances. If such cases are not promptly diagnosed, there is a danger that appropriate therapy will be neglected. Since every possible type of psychotic symptoms can sometimes be organically caused, the organic point of view still is of enormous practical value. In addition, this aspect is of great academic importance, as it illuminates the dynamic process not only of the symptomatic psychosis but also of the alleged endogenous psychosis.

The point of interest lies wherein, clinical presentation with ambiguous non-specific psychotic symptoms raising a suspicion of an organic aetiology warrants neurological and radiological evaluation of the brain.

Psychosis as part of Schizophrenia typically presents between ages 15 and 35.[3] Because of our patient's age, atypical presentation of symptoms, work-up for an underlying neurologic disorder ensued. Because most patients with Psychosis do not have Basal Ganglia Calcification,[4] and also because many patients with Basal Ganglia Calcification are asymptomatic, we wondered about the association between

our patient's symptoms and his evident Basal Ganglia Calcification.

Basal Ganglia Calcification is a relatively uncommon finding, and bilateral Basal Ganglia Calcification is even rarer. CT imaging is the radiologic modality of choice to identify calcium deposits. A literature review reported the incidence of Basal Ganglia Calcification on a sample of 29,484 CT scans to be 0.93% for all clinical indications.[5]

Basal Ganglia Calcification appears to be more common in patients with neuropsychiatric manifestations-it has been found in 9% of CT scans for neurologic or psychiatric indications.[6] Psychological disorders affecting the basal ganglia can be difficult to diagnose, whether they are concomitant with the neurological disease or part of the Comorbidity or its consequence.

Fahr's disease is a rare, degenerative, neurological condition characterised by idiopathic calcification of the basal ganglia. The clinical manifestations of Fahr's disease vary. One definition proposed by Trautner et al requires bilateral calcifications with neuropsychiatric and extrapyramidal motor disorders with normal calcium and phosphorus metabolism.[7]

Our patient has presented with an indistinct pattern of psychotic symptoms and lacked extrapyramidal symptoms or a metabolic disorder and had normal neurological examination whose CT brain findings showed bilateral basal ganglia calcification. Kotan et al had reported similar cases wherein the presentation of patient in early adulthood with recent onset of progressive first episode psychosis with schizophrenia like symptomatology and without any neurological symptoms leading to misdiagnosis of schizophrenia.[8]

Flint and Goldstein et al opined that radiologists may view basal ganglia calcification as an incidental finding, so clinical findings associated with Fahr's disease should be considered important.[9] According to Rasmussen et al, before age 50, incidental discovery of Basal Ganglia Calcification merits diagnostic investigation.[10] The course of Fahr's disease is progressive as reported by Nishiyama et al.[11] In adult-onset Fahr's disease, calcium deposition generally begins in the third decade of life, with neurological deterioration two decades later as reported by Manyam et al.[12] All these literature reviews concur with the variable presentation of psychosis in our patient.

Konig et al postulated that about 40% of patients with Fahr's disease are found to present initially with psychiatric features similar to our case where cognitive, psychotic, and mood disorders are common.[13] According to Cummings et al, paranoid and psychotic features often present between the ages of 20 and 40 in Fahr's disease relating to the age (31 years) of our patient.[14]

Two patterns of psychotic presentation in Fahr's disease are known, including early onset (Mean age 30.7 years) with minimal movement disorder and late onset (mean age 49.4 years) attended by dementia and movement disorder (Cummings et al) Our patient fits into early onset pattern of presentation with psychotic symptoms and no extra pyramidal involvement.

Rosenberg et al found that Symptoms develop when the deposits accumulate in the basal ganglia, with progressive deterioration of mental function including disorganised behaviour, suspiciousness, inattention and memory impairment which have been associated with Fahr's disease which our patient also exhibited.[6]

Overt psychotic symptoms, such as the delusions experienced by our patient, have been reported with Basal Ganglia Calcification disorders such as Huntington's disease.[1] Given the overlap between the psychotic symptoms present in basal ganglia disorders such as Huntington's disease and the symptoms seen in schizophrenia, similar mechanisms have been proposed. Moreover, altered dopaminergic transmission in the basal ganglia has been suggested as a mechanism sustaining positive psychotic symptoms in schizophrenia and Fahr's disease, a disorder of bilateral Basal Ganglia Calcification.[15]

Fronto-subcortical circuits provide a framework for the interpretation of psychiatric symptoms in patients with basal ganglia dysfunction.[16] One proposed mechanism includes basal ganglia neuronal degeneration and mineralisation that disrupts these pathways, generating psychiatric symptoms.

CONCLUSION

There have been many major advances in our understanding of organic Psychosis, especially in terms of aetiology, the breadth of the clinical presentation and accurate diagnostic methods for evaluation.

There is no simple solution to this dilemma and uncertainty. While considering early accurate diagnosis and better prognosis of patients with organic psychosis, it is crucial for a more integrative approach involving the Physician, Neurologist and Psychiatrist for efficient management.

Secondly, evaluation for such organic psychotic disorders should be more systematic and organised keeping in mind the complexity of such disorders and the organic evaluation should never be underestimated.

Thirdly, research aiming at better understanding of Idiopathic Basal Ganglia Calcification (Fahr's disease) with its variable presentations might throw some light on organic psychotic disorders as a whole.

REFERENCES

1. American Psychiatric Association. Diagnostic and statistical manual of mental disorders. 4th edn. Washington DC 2000.
2. Dilsaver SC. Differentiating organic from functional psychosis. Am Fam Physician 1992;45(3):1173-80.
3. Kessler RC, Berglund P, Demler O, et al. Lifetime prevalence and age-of-onset distributions of DSM-IV disorders in the national comorbidity survey replication. Arch Gen Psychiatry 2005;62(6):593–602.
4. Wider C, Dickson DW, Schweitzer KJ, et al. Familial idiopathic basal ganglia calcification: a challenging clinical-pathological correlation. J Neurol 2009;256(5):839-42.
5. Lauterbach EC, Cummings JL, Duffy J, 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: a report of the ANPA committee on research. J Neuropsychiatry Clin Neurosci 1998;10(3):249–66.

6. Rosenberg DR, Neylan TC, el-Alwar M, et al. Neuropsychiatric symptoms associated with idiopathic calcification of the basal ganglia. J Nerv Ment Dis 1991;179(1):48–9.
7. Trautner RJ, Cummings JL, Read SL, et al. Idiopathic basal ganglia calcification and organic mood disorder. Am J Psychiatry 1988;145(3):350–3.
8. Kotan D, Aygul R. Familial Fahr disease in a Turkish family. South Med J 2009;102(1):85-6.
9. Flint J, Goldstein LH. Familial calcification of the basal ganglia: a case report and review of the literature. Psychol Med 1992;22(3):581–95.
10. Rasmussen MJ, Pilo L, Nielsen HR. Basal ganglia calcifications demonstrated by CT. Is further investigation necessary when this is found incidentally? Ugeskr Laeger 1991;153(29):2051–3.
11. Nishiyama K, Honda E, Mizuno T, et al. A case of idiopathic, symmetrical non-arteriosclerotic, intracerebral calcification (Fahr's disease) associated with M-proteinemia, followed by multiple myeloma. Rinsho Shinkeigaku 1991;31(7):781–4.
12. Manyam BV, Bhatt MH, Moore WD, et al. Bilateral striopallidodentate calcinosis: cerebrospinal fluid, imaging, and electrophysiological studies. Ann Neurol 1992;31(4):379–84.
13. Konig P. Psychopathological alterations in cases of symmetrical basal ganglia sclerosis. Biol Psychiatry 1989;25(4):459–68.
14. Cummings JL, Gosenfeld LF, Houlihan JP, et al. Neuropsychiatric disturbances associated with idiopathic calcification of the basal ganglia. Biol Psychiatry 1983;18(5):591–601.
15. Savoldi F, Nappi G, Martignoni E, et al. Biochemical evidence of dopaminergic involvement in Fahr's syndrome. Acta Neurol (Napoli) 1980;2(4):231-9.
16. Tekin S, Cummings JL. Frontal-subcortical neuronal circuits and clinical neuropsychiatry: an update. J Psychosom Res 2002;53(2):647–54.