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Assessment and Determinants of Neuropsychiatric Presentation of Strio-Pallido-Dentate Calcification (Fahr's disease/syndrome)

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Abstract

Background: Strio-Pallido-Dentate calcification can be idiopathic (Fahr's disease) or secondary (Fahr's syndrome). Psychiatric disturbances were the major presenting symptoms.
Objectives: determination and assessment of psychiatric presenting symptoms of Fahr's disease/syndrome, and sociodemographic and clinical variables, and study the statistical significant of each variable.
Methods: patients attending psychiatric unit, during period of study, with Fahr's disease/syndrome were assessed by consultant psychiatrist. Sociodemographic and clinical variables were assessed. Structured clinical interview and MMSE was done. Data analysis and statistical significance and was done.
Results: total 53 patients; Idiopathic basal ganglia calcification (Fahr's disease) (FD) 75.5%, secondary calcification (Fahr's syndrome) 24.5%. The age range 35–78 years, mean 56.4±13 years, 64% married, 88% of higher education. Psychiatric presenting symptoms were; schizophrenia 32.1%, depression 18.9%, mania17.0%, bipolar disorder 17.0% and dementia 15.1%. Negative investigation was 3%, cognitive impairment 17%, seizures11.3%, movement disorders.5%, vertigo 26.4%, headache 9.4%, unsteadiness and difficulty of swallowing 56.6%. Psychiatric morbidity was significantly associated with; cognitive impairment (P<0.001), movement disorders (P=0.034), unsteadiness (P=0.019), difficulty of swallowing (P=0.019).
Conclusion: Fahr's disease/syndrome diagnosis could be challenging, due to discrepancy between clinical presentations and radio-imaging findings. Fahr's disease/syndrome should be kept in mind in any patients with late onset neuropsychiatric disorders.

Keywords: Fahr's disease; Fahr's syndrome; psychiatric presentation; schizophrenia; depression.

Introduction

Idiopathic Strio-Pallido-Dentate calcification also known as Fahr's disease or Fahr's syndrome1 is chronic, slowly progressive, neurodegenerative disorder2 characterised by extensive deposition of calcium in the basal ganglia[3], thalamus, cerebral cortex, dentate nucleus, cerebellum subcortical white matter, and hippocampus[4,5]. Within the basal ganglia, the globus pallidus is the most frequent site of the calcification but deposits may be present in the putamen, the caudate nucleus, the internal capsule,

the dentate nucleus, the thalamus, the cerebellum and the cerebral white matter[6]. Histologically, these deposits which contain proteins and polysaccharides are found in the perivascular space and in the media layer of the small vessels[7]. The pathogenesis is not known, but it may be secondary to the impairment of the blood brain barrier or to a neuronal calcium phosphoric metabolism dis¬order[3]. Basal ganglia calcification [BGC] can be idiopathic or secondary to genetic, metabolic, and endocrinological disorders8. Idiopathic BGC is known as Fahr's disease (FD) and BGC secondary to endocrinological causes is known as Fahr's syndrome2. Histological findings in the form of symmetrical brain calcifications were observed for the first time by Bomberger in 18559. Clinical manifestations of Fahr's syndrome were first described in 1930 by German neurologist Karl Theodor Fahr[4,10,11]. Neuropsychiatric, extra pyramidal and cerebellar symptoms, convulsive seizures, Parkinsonian fea¬tures, dementia and speech disorders may accompany the clini¬cal picture. This disease usually appears between the age of 40-60 years[3,12,13], with a prevalence of <1/1,000,000[14]. The onset of the disease is usually insidious and frequently is misdiagnosed as a dementia or psychiatric illness[4]. Most cases present with extra pyramidal symptoms initially. Additionally, they may present with cerebellar dysfunction, speech difficulty, dementia and neuropsychiatric symptoms[14,15]. Fahr's disease is most commonly transmitted as an Autosomal Dominant trait; but it may also be passed on as an autosomal recessive trait or it may occur sporadically[14,16]. A Locus at 14q (IBGC1) has been suggested to be involved commonly. A second locus has been identified on chromosome 8 and a third on chromosome 2[17,18]. A loss of function mutation in the gene encoding type III sodium dependent phosphate transporter 2 (SLC20A2) located on chromosome 8 has also been reported as the molecular level to form the genetic basis for the pathophysiology of this disease[19,20,21]. Congenital or early onset finding along with intellectual disability or presence of systemic involvement should alert one to the possibility of alternative diagnosis. It is differentiated from calcified angiomas, infections, enchapalitides and Addision's disease by its severity and characteristic distribution. Synonyms: Fahr's disease, Fahr's syndrome, idiopathic basal ganglia calcification, striopallidodentate calcification and calcinosis nucleorum14. Common clinical findings of the disease are character¬izing headache, movement disorders such as Parkinsonism, dystonia, chorea and ataxia, syncope, seizures and psychiatric symptoms[22].

Patients and Methods

Data Source:

A cross-sectional study with analytic component was conducted at Psychiatry Unit, Imamain Kadhimain Medical City, Baghdad, Iraq. The data collection was done with cooperation of neurology and radiology units, during the period from September 1st, 2013 to November 1st, 2017.

Patient Selection:

All patients with Strio-Pallido-Dentate calcification, both genders who have consulted during the study period and given their consent were included. The study was approved by psychiatric department. A questionnaire was prepared and administered to each patient consulted during the study period.

Patients' Background Characteristic:

All patients attended psychiatric clinic with suspicion of Fahr's disease/syndrome were send for brain radio-imaging and blood investigations to confirm the diagnosis. A questionnaire filled out by consultant psychiatrist, which included on the one part the collection of clinical and sociodemographic data (age, gender, marital status, level of education, occupation, investigation, movement disorders, headache, vertigo, seizures) and on the other part the mini-mental state examination (MMSE) was done to assess the cognitive impairment for each patient. Cutoffs points were: 27–30 is normal, 21–26 is mild impairment, 11–20 is moderate impairment and ≤10 is severe impairment[23]. All patients were selected for administration of the Structured Clinical Interview for DSM-IV-TR (SCID)[24] by consultant psychiatrist to assess the psychiatric morbidity. All patients sent for investigations to assess the endocrine state. Parathyroid hormone, calcium and phosphate serum levels were assessed. Other clinical symptoms were collected through the same interview.

Definition of variables:

The independent variables evaluated to explain psychiatric morbidity were socio-demographic and clinical variables. Sociodemographic includes; age, gender, marital status, level of education, occupation. Clinical variables include the associated features like; investigation, movement disorders, headache, vertigo, seizures.

Statistical analysis:

Data processing and analysis was conducted using a Statistical Package for Social Sciences (SPSS) version 19. Results are expressed as percentages for

qualitative variables. Chi-square test was used for finding association between two qualitative variables. P values were calculated to determine associations between sociodemographic factors and mental illness. P≤0.05 was taken as statistically significant.

Ethical issues:

Informed consent was obtained from the patients after clarifying the objectives of the study. Names were kept anonymous and interviews were conducted with full privacy.

Results

Total 57 patients with Strio-Pallido-Dentate calcification were included in this study. Five of them were withdrawn from the study due to incomplete interview. Data analysis was done for 53 patients; Idiopathic basal ganglia calcification (Fahr's disease) (FD) 75.5%, secondary calcification (Fahr's syndrome) 24.5%. The sociodemographic characteristics were; age range was 35–78 years, mean 56.4±13 years, half of cases fall into the age group 40 - 60 years, three forth of the sample was male, 64% married, 88% of higher education, only one forth still working. MMSE was done to assess the cognitive function results in 17% of patients with Strio-Pallido-Dentate calcification of poor cognitive function. Administration of the Structured Clinical Interview for DSM-IV-TR (SCID) by consultant psychiatrist was done for every patient. Clinical interview found; psychosis and schizophrenia 32.1%, depression 18.9%, mania17.0%, bipolar disorder 17.0% and dementia 15.1%. Almost 94.3% negative investigation results regarding parathyroid hormone, calcium and phosphate. 11.3% have history of seizures. Abnormal movements Parkinson like symptoms; tremor, rigidity, was found among 24.5% of patients. Vertigo present in 26.4% of patients. Headache present in 9.4% of patients. Unsteadiness and difficulty of swallowing found among 56.6% of patients. (Table 1) Correlation of age with sociodemographic and clinical variables was done. The age of patients with strio-pallido-dentate calcification was significantly associated with; occupation (P=0.001), psychiatric morbidity (P<0.001), investigation (P=0.033), cognitive impairment (P=0.003), movement disorders (P=0.009), unsteadiness (P<0.001) and difficulty of swallowing (P<0.001) (Table 2). Correlation of psychiatric morbidity, which obtained from Structured Clinical Interview, with sociodemographic and clinical variables, was done. Psychiatric morbidity of patients with Strio-Pallido-Dentate calcification was significantly associated with; cognitive impairment (P<0.001), movement disorders (P=0.034), unsteadiness (P=0.019), difficulty of swallowing (P=0.019) (Table 3). Correlation of cognitive impairments, which obtained from application of MMSE, with sociodemographic and clinical variables, was done. Cognitive impairment of patients with strio-pallido-dentate calcification was significantly associated with; occupation (P=0.023), psychiatric morbidity (P<0.000), unsteadiness (P=0.032), difficulty of swallowing (P=0.032) (Table 4). Correlation of types of Strio-Pallido-Dentate calcification, Fahr's Disease and Fahr's syndrome, with sociodemographic and clinical variables was done. Strio-Pallido-Dentate calcification, idiopathic and secondary, was significantly associated with; age of the patients (P=0.014), psychiatric morbidity (P<0.001), investigation (P=0.002), cognitive impairment (P<0.001) (Table 5).

Table 1: Frequency and percentages of sociodemographic and clinical variables for patients with Strio-Pallido Dentate calcification

Sociodemographic and Clinical variables		**No.**	**%**
Age Group	31-40 years	6	11.3
	41-50 years	11	20.8
	51-60 years	16	30.2
	61-70 years	9	17.0
	71-80 years	11	20.8
Gender	Male	41	77.4
	Female	12	22.6
Marital Status	Single	5	9.4
	Married	34	64.2
	Widow	14	26.4
Education	Intermediate	19	35.8
	Secondary	28	52.8
	College	6	11.3
Occupation	Unemployed	32	60.4
	Employed	13	24.5
	Retired	8	15.1
Psychiatric Morbidity	Psychosis	17	32.1
	Depression	10	18.9
	Mania	9	17.0
	Bipolar Disorder	9	17.0
	Dementia	8	15.1
Investigation	No	50	94.3
	Yes	3	5.7
Cognitive Impairment	No	44	83.0
	Yes	9	17.0
Seizure	No	47	88.7
	Yes	6	11.3
Movement	No	40	75.5
	Yes	13	24.5
Vertigo	No	39	73.6
	Yes	14	26.4
Headache	No	48	90.6
	Yes	5	9.4
Unsteadiness	No	23	43.4
	Yes	30	56.6
Difficulty Swallowing	No	23	43.4
	Yes	30	56.6

Table 2: Correlation of age groups with sociodemographic and clinical variables of patients with Strio-Pallido Dentate calcification

sociodemographic and Clinical Characteristics of Patients		Age Groups					P value
		31-40 years	41-50 years	51-60 years	61-70 years	71-80 years	
Gender	Male	5	8	12	7	9	0.980
	Female	1	3	4	2	2	
Marital Status	Single	2	2	1	0	0	0.054
	Married	4	9	8	5	8	
	Widow	0	0	7	4	3	
Education	Intermediate	1	7	4	2	5	0.204
	Secondary	5	4	8	6	5	
	College	0	0	4	1	1	
Occupation	Unemployed	4	8	8	7	5	0.001
	Employed	2	3	8	0	0	
	Retired	0	0	0	2	6	
Psychiatric Morbidity	Psychosis	2	1	3	9	2	0.000
	Depression	2	4	3	0	1	
	Mania	2	5	0	0	2	
	Bipolar Disorder	0	1	7	0	1	
	Dementia	0	0	3	0	5	
Investigation	No	4	11	15	9	11	0.033
	Yes	2	0	1	0	0	
Cognitive Impairment	No	6	11	13	9	5	0.003
	Yes	0	0	3	0	6	
Seizure	No	6	10	13	9	9	0.499
	Yes	0	1	3	0	2	
Movement	No	6	7	16	4	7	0.009
	Yes	0	4	0	5	4	
Vertigo	No	5	7	10	7	10	0.456
	Yes	1	4	6	2	1	
Headache	No	6	10	14	8	10	0.934
	Yes	0	1	2	1	1	
Unsteadiness	No	6	11	4	1	1	0.000
	Yes	0	0	12	8	10	
Difficulty Swallowing	No	6	11	4	1	1	0.000
	Yes	0	0	12	8	10	

Table 3: Correlation of psychiatric presentations with sociodemographic and clinical variables of patients with Strio-Pallido Dentate calcification

Correlation of Psychiatric Presentations with Sociodemographic and Clinical Variables		Psychiatric Presentations					P value
		Psychosis	Depression	Mania	Bipolar Disorder	Dementia	
Age Group	30-40 years	2	2	2	0	0	0.000
	40-50 years	1	4	5	1	0	
	50-60 years	3	3	0	7	3	
	60-70 years	9	0	0	0	0	
	70-80 years	2	1	2	1	5	
Gender	Male	12	7	9	7	6	0.491
	Female	5	3	0	2	2	
Marital Status	Single	2	2	0	1	0	0.411
	Married	10	5	9	5	5	
	Widow	5	3	0	3	3	
Education	Intermediate	5	4	2	5	3	0.532
	Secondary	9	4	7	3	5	
	College	3	2	0	1	0	
Occupation	Unemployed	11	7	6	5	3	0.511
	Employed	3	2	2	4	2	
	Retired	3	1	1	0	3	
Investigation	No	14	10	9	9	8	0.151
	Yes	3	0	0	0	0	
Cognitive impairment	No	17	9	9	9	0	0.000
	Yes	0	1	0	0	8	
Seizure	No	14	10	9	8	6	0.331
	Yes	3	0	0	1	2	
Movement	No	11	9	4	9	7	0.034
	Yes	6	1	5	0	1	
Vertigo	No	12	7	7	7	6	0.989
	Yes	5	3	2	2	2	
Headache	No	15	10	8	7	8	0.443
	Yes	2	0	1	2	0	
Unsteadiness	No	4	6	7	5	1	0.019
	Yes	13	4	2	4	7	
Difficulty Swallowing	No	4	6	7	5	1	0.019
	Yes	13	4	2	4	7	

Table 4: Correlation of cognitive impairment with sociodemographic and clinical variables of patients with Strio-Pallido Dentate calcification

Correlation of Cognitive Impairment with Sociodemographic and Clinical variables		Cognitive Impairment		P value
		No	Yes	
Age Group	30-40 years	6	0	0.003
	40-50 years	11	0	
	50-60 years	13	3	
	60-70 years	9	0	
	70-80 years	5	6	
Gender	Male	34	7	0.974
	Female	10	2	
Marital Status	Single	5	0	0.289
	Married	29	5	
	Widow	10	4	
Education	Intermediate	16	3	0.983
	Secondary	23	5	
Occupation	Unemployed	29	3	0.023
	Employed	11	2	
	Retired	4	4	
Psychiatric Morbidity	Psychosis	17	0	0.000
	Depression	9	1	
	Mania	9	0	
	Bipolar Disorder	9	0	
	Dementia	0	8	
Investigation	No	41	9	0.420
	Yes	3	0	
Seizure	No	40	7	0.257
	Yes	4	2	
Movement	No	32	8	0.305
	Yes	12	1	
Vertigo	No	33	6	0.605
	Yes	11	3	
Headache	No	39	9	0.288
	Yes	5	0	
Unsteadiness	No	22	1	0.032
	Yes	22	8	
Difficulty Swallowing	No	22	1	0.032
	Yes	22	8	

Table 5: Correlation of Fahr's disease/syndrome with sociodemographic and clinical variables of patients with Strio-Pallido Dentate calcification

Correlation of Fahr's disease/syndrome with Sociodemographic and Clinical variables		Strio-Pallido-Dentate Calcification				P value
		Idiopathic (Fahr's Disease) (40)		Secondary (Fahr's Syndrome) (13)		
		No.	%	No.	%	
Age Group	30-40 years	4	10.0	2	15.4	0.014
	40-50 years	11	27.5	0	0	
	50-60 years	11	27.5	5	38.5	
	60-70 years	9	22.5	0	0	
	70-80 years	5	12.5	6	46.1	
Gender	Male	32	80.0	9	69.2	0.420
	Female	8	20.0	4	30.8	
Marital Status	Single	3	7.5	2	15.4	0.292
	Married	28	70.0	6	46.1	
	Widow	9	22.5	5	38.5	
Education	Intermediate	16	40.0	3	23.1	0.394
	Secondary	19	47.5	9	69.2	
	College	5	12.5	1	7.7	
Occupation	Unemployed	27	67.5	5	38.4	0.108
	Employed	9	22.5	4	30.8	
	Retired	4	10.0	4	30.8	
Psychiatric Morbidity	Psychosis	14	35.0	3	23.1	0.000
	Depression	8	20.0	2	15.4	
	Mania	9	22.5	0	0	
	Bipolar Disorder	9	22.5	0	0	
	Dementia	0	0	8	61.5	
Investigation	No	40	100	10	76.9	0.002
	Yes	0	0	3	23.1	
Cognitive Impairment	No	40	100	4	30.8	0.000
	Yes	0	0	9	69.2	
Seizure	No	36	90.0	11	84.6	0.595
	Yes	4	10	2	15.4	
Movement	No	28	70.0	12	92.3	0.104
	Yes	12	30.0	1	7.7	
Vertigo	No	31	77.5	8	61.5	0.257
	Yes	9	22.5	5	38.4	
Headache	No	35	87.5	13	100	0.180
	Yes	5	12.5	0	0	
Unsteadiness	No	20	50.0	3	23.1	0.089
	Yes	20	50.0	10	76.9	
Difficulty swallowing	No	20	50.0	3	23.1	0.089
	Yes	20	50.0	10	76.9	

Discussion

Idiopathic Strio-Pallido Dentate calcification (Fahr's disease) represent the majority of this study (75.5%), mostly 80% of them were male, 70% married, 60% higher education level, more than two third were unemployed. Clinical interview found that; schizophrenia 35%, bipolar disorders 22.5%, mania 22.5%, depression 20%. These findings were statistically significant (P<0.001). Around 40% of patients with basal ganglia calcifications presented with psychiatric symptoms[25]. The criteria for the diagnosis of Fahr's disease include[26]: bilateral basal ganglia calcification; progressive neuropsychiatric symptoms; late onset in the fourth or fifth decade (earlier onset can also occur). Fahr's disease clinical symptoms are reported in the literature either as individual case reports or as family reports due to the clinical rarity of the disease[27], so there were no exact figures to compare the results of this study with it, but some sporadic case reports. Psychiatric manifestation of Fahr's disease might be the end product of the interaction of basal ganglia and cerebellum. Fahr's disease patients of this study showed no any positive results of investigations, which is statistically significant (P=0.002). MMSE results were no cognitive impairment found in patients with Fahr's disease P<0.001). Other symptoms associated with Fahr's disease were; seizures 10%, movement disorders 30%, vertigo 22.5%, headache 12.5%, unsteadiness 50% and difficulty of swallowing 50%. These symptoms have no significant statistical association with Fahr's disease. Movement disorders account for 55% of the total symptomatic Fahr's disease patients[3]. Secondary basal ganglion calcification (Fahr's syndrome) majority presented with dementia 601.5%, psychosis and schizophrenia 23.1% and depression 15.4%, which is statistically significant (P<0.001). Fahr's syndrome showed 23.1% positive investigation, with statistically significant association (P=0.002). Almost 70% of Fahr's syndrome with poor cognitive function which of significant association (P<0.001). Unsteadiness and difficulty of swallowing associated with 77% of cases of Fahr's syndrome. There was no exact figure to make comparison with it, although there was some sporadic case report. The functions of the basal ganglia and the cerebellum may be integrated across motor and non-motor domains. Both the motor and non-motor domains of the dentate nucleus of the cerebellum provide disynaptic inputs to the basal ganglia[25]. Also, the motor and non-motor domains of the substantia nigra provide disynaptic inputs to the cerebellar cortex. These interactions may have clinical implications for neuropsychiatric disorders such as schizophrenia[28]. Basal ganglia have numerous neurotransmitters[29]. The basal ganglia are thought to be involved in several functions including motor learning, sequencing, movements, attentional allocation, working memory, and implicit memory[30]. These operations may have roles in the acquisition of automatically-performed behaviors as well as in enhancing the efficiency of higher order processors like those involved in working memory, and reward processes[31]. Clinically, interactions between dopamine and acetylcholine are applied in Parkinson's disease. The basal ganglia may be involved in generating neuropsychiatric symptoms in major psychiatric disorders such as schizophrenia and depression[29]. Several basal ganglia disorders can have mental and cognitive manifestations[25]. The common psychiatric features are cognitive deterioration, psychotic symptoms, and mood disorders[32]. Organic affective symptoms were found more common in chronic cases of basal calcification than those with initial presentation, with depression being the most commonly reported mood disorder[25]. Calcifications sites, clinical features and relevant investigations are important particularly upon incidental cases presenting with disorganised behavior, as there may be no other significant clinical features. Calcifications should be evaluated with clinical pictures for proper diagnosis[4]. In conclusion, Fahr's disease/syndrome diagnosis could be challenging, due to discrepancy between clinical presentations and radio-imaging findings. Clinical presentation includes neurological and psychiatric symptoms. Neurological symptoms include Parkinson's-like movement, vertigo, seizures, and dementia. Psychiatric symptoms include schizophrenia, psychosis, bipolar disorder, mania and depression. Fahr's disease/syndrome should be kept in mind in any patients with neuropsychiatric disorders, especially of late onset and suspected metabolic and endocrine abnormalities.

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