

## CASE REPORT

# Psychosis, Idiopathic Hypoparathyroidism and Basal Ganglia Calcification: A Case Report of Fahr's Syndrome

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### ABSTRACT

Making a medical diagnosis in an elderly person presenting with first time psychiatric symptoms can be challenging. A 61 year old lady presented with 4 years history of tactile hallucinations in her legs and delusions of persecution, and 2 weeks history of depression and mild cognitive impairment. There were no other significant physical findings, except for dysarthria and fine bilateral hand tremors. As patient had no prior psychiatric history, she was investigated for differential diagnoses of late-onset schizophrenia, psychotic depression and early dementia. A thorough investigation for concomitant medical illnesses was done which revealed low serum calcium, high serum phosphate and relatively low serum parathyroid hormone levels. A diagnosis of Fahr's syndrome (FS) was made based on history, hematological findings of idiopathic hypoparathyroidism and bilateral basal ganglia calcifications (BGC) on neuroimaging. Treatment of FS is non-specific and mainly symptomatic. This patient improved with treatment comprising antidepressant, antipsychotic and calcium supplement.

**Keywords:** Basal ganglia calcification (BGC), Fahr's syndrome (FS), Idiopathic hypoparathyroidism (IH)

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### INTRODUCTION

This case report highlights Fahr's syndrome, an uncommon medical condition occurring in an elderly person presenting with psychiatric symptoms. Presence of first onset psychiatric symptoms in an elderly person will always warrant an intensive patient assessment, comprising thorough psychiatric history and organic workout. Fahr's syndrome (FS) is a term referred to the presence of Basal Ganglia Calcification (BGC) and neuropsychiatric symptoms, with a low prevalence of 1<1,000,000 (1). Aetiology of FS includes adult-onset neurodegenerative disorder, inherited disorder, infectious and inflammatory disease, toxins or radiation exposure, but the most common cause are endocrine disorders, particularly parathyroid disturbances. Idiopathic hypoparathyroidism (IH) is an uncommon condition in the elderly, which is characterized by low serum levels of parathyroid hormone (PTH), hypocalcemia and hyperphosphatemia, due to either absence, fatty replacement or atrophy of the parathyroid glands (1).

### CASE REPORT

A 61 year old Malay lady, with a 4 year history of experiencing tactile hallucinations of squirming skin sensations over both lower limbs and persecutory delusions of being harmed by other people. Two weeks prior to her admission, she developed depressive symptoms of low mood, lethargy, insomnia and anhedonia. There were no other hallucinatory experiences, suicidal tendencies or past history of manic symptoms in this patient. Neither was there any history of significant memory impairment, seizures or chest pain. Neurological examination revealed presence of dysarthria and fine tremors of bilateral hands, without other extra pyramidal signs. Other systemic examinations, including the neck region were normal. Chvostek's and Trousseau's signs were also negative. She scored 19/30 on Mini Mental State Examination (MMSE) and 10/15 for Geriatric depression scale (GDS). Investigations revealed low serum calcium:1.76 mmol/L (normal: 2.14-2.58 mmol/L), high serum phosphate:1.84mmol/L (normal:0.71-1.36) and relatively low serum parathyroid hormone levels: 2.15pmol/L (normal: 1.3-7.6). Other blood tests such as full blood count, renal profile, serum iron, serum magnesium, vitamin B12, folate, lipid profile, fasting blood sugar, thyroid function test, 24 hour urinary calcium and protein levels were normal. Antinuclear antibody, rheumatoid factor, serum protein electrophoresis to look for monoclonal band and urine protein electrophoresis to look for Bence-Jones protein,

all were negative. Erythrocyte sedimentation rate was raised: 55 mm/hr (normal:1-20 mm/hr) but infectious disease screening for tuberculosis, hepatitis B and C, VDRL and HIV were all negative. Nerve conduction study revealed bilateral S1 radiculopathy but no evidence of myopathy and sensory problems on electromyography. Brain CT showed presence of bilateral calcifications of the basal ganglia comprising corona radiata, heads of caudate nuclei, bilateral lentiform nucleus and dentate nucleosus of the cerebellum (figure 1). Her psychiatric symptoms and cognitive functions improved, with MMSE score of 23/30 and GDS score 8/15 following symptomatic treatment with psychotropic medications and calcium supplements.

## DISCUSSION

Late onset psychosis in the elderly is relatively uncommon and possibility of organic aetiology has to be ruled out first. In this patient, the tactile hallucinations of squirming sensation in the limbs could also be attributed to the long standing state of hypocalcaemia which usually presents as muscle twitching, numbness or cramps. She was clinically quite stable without any manifestations of hyperpigmentation, seizure, cardiac or respiratory problems related to hypocalcaemia. Blood investigations failed to identify the definite causes of hypocalcaemia in this patient, suggesting idiopathic hypoparathyroidism as a significant finding.

Diagnostic criteria for FS include the presence of bilateral BGC on neuroimaging, progressive neurological

**Illustration 1.** Brain Computerized Tomography (CT) scan of the patient, image shows presence of bilateral calcification of the basal ganglia (marked as X).



dysfunction, onset of illness usually between fourth to fifth decades, family history, no biochemical abnormalities and somatic features suggestive of a mitochondrial or metabolic disease or other systemic disorder, and no infectious, toxic, or traumatic causes (1). In this case diagnosis of FS was made based on psychiatric history, presence of IH and bilateral BGC. Family history of similar illness was unknown as the patient was an adopted child since birth.

In the elderly, about 0.3%-1.5% of physiological and asymptomatic BGC in brain CT could be an incidental finding (2). However, pathological BGC may present as a result of metabolic disorders, infectious or genetic diseases, the most common causes were hypoparathyroidism and pseudohypoparathyroidism (3). Thus, symmetrical BGC is a consequence of the hypoparathyroidism, as reported by Eaton in 1939 who first found BGC associated with chronic hypoparathyroidism. The natural history of IH and related complications are not well understood due to lack of large long-term prospective studies (4).

About 50% of patients with extensive calcification had psychiatric findings as compared to 34.5% patient with limited calcification(1). Seizures, parkinsonism, psychosis and dementia may occur due to pathology in the limbic system and basal ganglia (2, 4). BGC has been observed in mentally healthy patients, but is 8 times more common among non-demented elderly individuals with hallucinations or delusions, thus signifying possibilities of psychoses due to disturbance in the basal ganglia dopaminergic system (2). BGC might have been the eventual results of neuronal loss, hypometabolism, hypoperfusion and impairment in the neurochemical activity of dopamine, glutamate and acetylcholine (5).

This patient's differential diagnosis of probable Alzheimer disease with behavioural and psychological symptoms of dementia was unlikely due to the lack of history of memory impairment, deterioration in social functioning, intact independent living and reversible improvement in cognitive functions with treatment.

Currently, there is no definitive treatment for FS, except symptomatic relief. This patient responded well to daily oral calcium supplements, antidepressant (sertraline) and atypical antipsychotic (risperidone).

## CONCLUSIONS

In conclusion, FS should be suspected in the presence of psychiatric symptoms, endocrine disorders and BGC in the elderly. Patients tend to respond well to symptomatic treatment with psychotropic medications and calcium supplements.

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