

## A 45-Year-Old Male with Fahr's Disease

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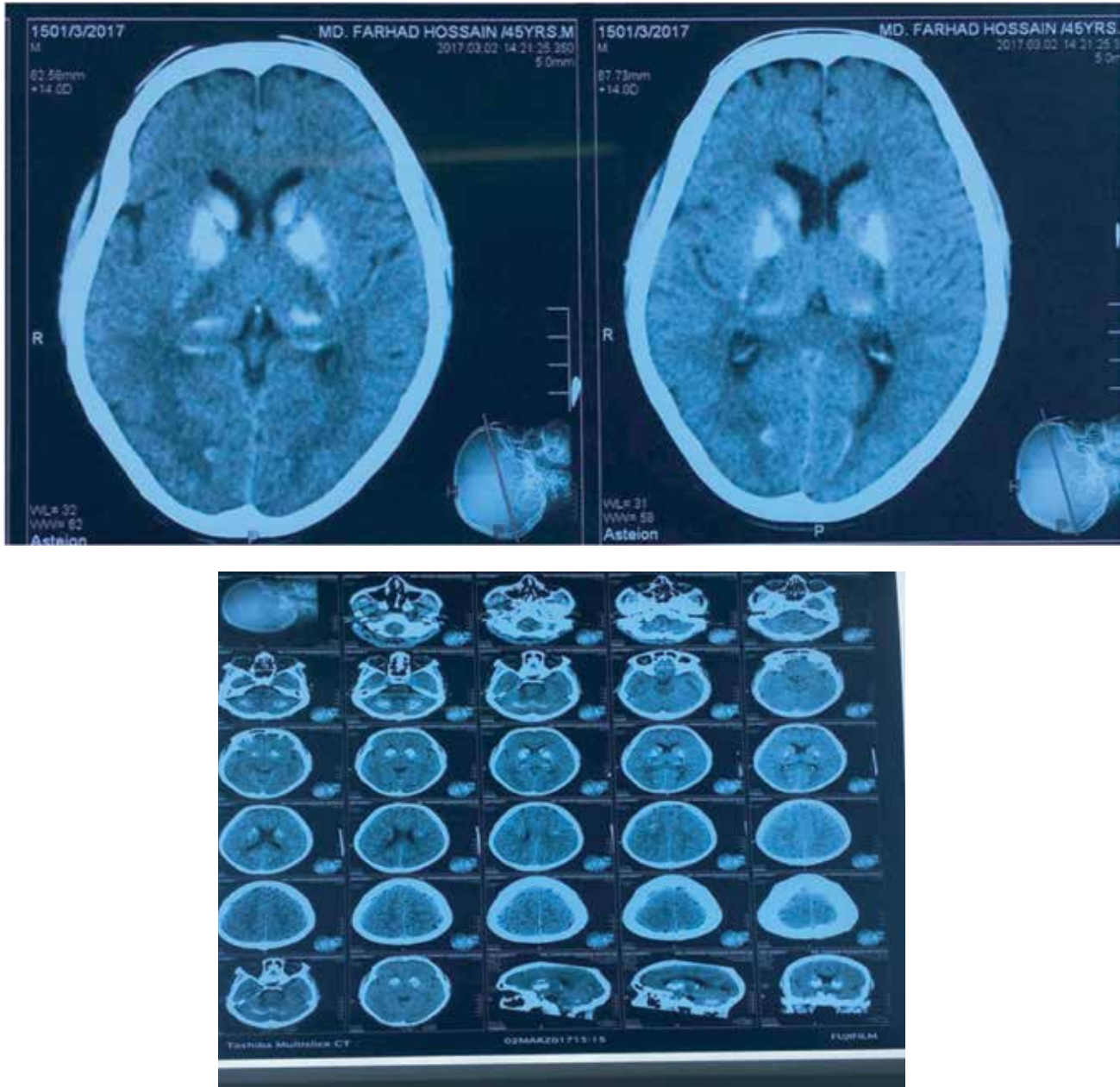


Fig 1. Symmetrical calcifications in both basal ganglia, periventricular white matter and dentate nucleus of cerebellum

A 45-year-old male was admitted in Enam Medical College Hospital with several episodes of convulsion in last three years. His general and neurological

examinations revealed no abnormalities. There was no positive family history of any neurodegenerative and movement disorders. Complete blood count,

serum creatinine, fasting blood sugar, SGPT, serum calcium and phosphate, serum thyroid and parathyroid hormone levels were within normal limits. CT scan of brain showed symmetrical calcifications in both basal ganglia, periventricular white matter and dentate nucleus of cerebellum. Fahr's disease was diagnosed based on the clinical and imaging studies. Patient was treated with anticonvulsant sodium valproate and clonazepam.

Fahr's disease is a rare neurodegenerative disorder characterized by bilateral and symmetrical intracranial calcifications. It was first described in 1930 by a German neurologist Karl Theodor Fahr.<sup>1</sup> It is an autosomal dominant and genetically heterogeneous disease. It may be sporadic or familial. Usual age of presentation is 40–60 years with no gender difference.<sup>2</sup> The exact pathophysiology of Fahr's disease is not known. It is assumed that these intracranial calcifications may be due to metastatic deposition, secondary to local disruption of blood–brain barrier (BBB), or disorder of neuronal calcium phosphorus metabolism.<sup>3</sup> The common sites of calcifications are globus pallidus, putamen, caudate nucleus, internal capsule, dentate nucleus, thalamus, and cerebral white matter.<sup>4</sup> In our patient, CT scan showed calcifications in both basal ganglia, periventricular white matter and dentate nucleus of cerebellum.

Patients of Fahr's disease may present with many neurological, psychiatric and cognitive abnormalities.<sup>5</sup> Neurological symptoms include Parkinson's disease-like movement disorder, vertigo, epilepsy, syncope, cerebellar ataxia, and dementia.<sup>6</sup> Psychiatric symptoms include lack of interest, paranoid ideation, delusions, low mood, bipolar mood disorders and auditory and visual hallucinations.<sup>7</sup>

Fahr's disease should be distinguished from Fahr's syndrome in which basal ganglia calcification occurs due to secondary causes. The differential diagnoses of pathologic basal ganglia calcification include idiopathic and secondary hypoparathyroidism, hyperparathyroidism, post-thyroidectomy, birth anoxia, cysticercosis, toxoplasmosis, calcified infarct and HIV infection. In this case all the metabolic and hormonal profiles were normal.

Fahr's disease has no definitive treatment. Patients are treated symptomatically. In some studies, a biphosphonate named disodium etidronate showed functional benefit and symptomatic improvement without reduction in the amount of calcifications.<sup>8</sup>

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