

Clinical Image

Fahr Disease

Debasish Kumar Saha¹, Madhurima Saha², Mohammad Omar Faruq³, ASM Areef Ahsan⁴, Kaniz Fatema⁵, Fatema Ahmed⁶

A 60-year old lady, diabetic, normotensive, known CKD (baseline creatinine-2 mg/dl) admitted into BIRDEM General Hospital under Surgery in-patient unit with gangrene of left foot for 1 month. She also had abnormal movements of head and rolling of eyeballs for years. Following admission left sided below knee amputation was done. Postoperatively she was shifted to intensive care unit for altered level of consciousness and hypotension. On examination, she was found moderately anemic, dehydrated, febrile, tachycardic, tachypnoeic with hypotension (BP 80/50 mm Hg). She was grade II unconscious. Abnormal rolling of both eyeballs in all directions with abnormal movements of head and both upper limbs were noted. She had no signs of meningeal irritation and had no signs of cerebellar dysfunction. Her reflexes were normal with bilateral equivocal plantar responses. She was provisionally diagnosed as uremic encephalopathy due to AKI on CKD, with septic & hypovolaemic shock. Laboratory investigations revealed mild anaemia with neutrophilic leucocytosis. Urea-212 mg/dl, S creatinine-3.3 mg/dl, S calcium-7.4 mg/dl (corrected calcium 9.0 mg/dl), phosphate-4.2 mg/dl, iPTH-8.77pg/ml were noted. Blood C/S showed growth of *Acinetobacter* in two consecutive samples. The organism was sensitive to colistin in both samples. CT scan of head showed extensive bilateral calcifications suggestive of Fahr disease (see Fig). Other causes of brain calcifications were excluded. Her family history was negative for Fahr disease. She was managed with blood transfusion, adequate fluid and appropriate antibiotics. Symptomatic treatment with oral Procyclidine (anticholinergic) given for her dyskinetic movements. Subsequently she became afebrile, shock resolved and renal functions improved. Abnormal movements

were significantly improved. Later she was shifted to High Dependency Unit for step down care.

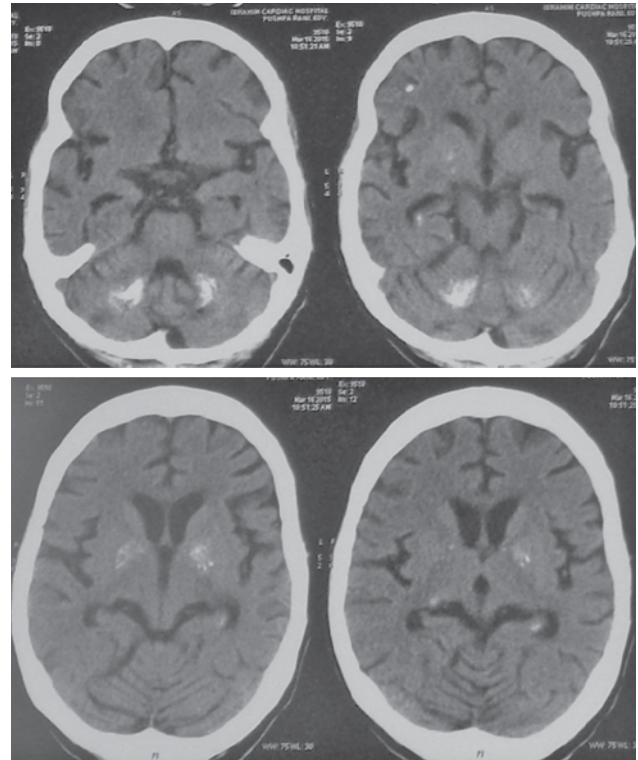


Figure: CT Scan of head showing bilateral symmetrical calcifications in corona radiata, both basal ganglia, thalamus and cerebellum suggestive of Fahr disease.

Discussion:

Fahr disease, also known as familial idiopathic basal ganglia calcification is a rare, genetically dominant, neurological disorder characterized by abnormal deposits of calcium in the areas of the brain that control movement. The disease was first noted by German Neurologist Karl Theodor Fahr in 1930.¹

The disease usually manifests itself in the third to fifth decade of life but may appear in childhood or later in life.²

Symptoms include deterioration of motor functions and speech, seizures, and other involuntary movements. Significant other symptoms are headache, dementia, and visual impairment. Characteristics of Parkinson's Disease are also found in Fahr disease.³ Neuropsychiatric symptoms, which may be the first or the most prominent manifestations, range from mild difficulty with concentration and memory to change in personality and /or behavior, to psychosis and dementia.⁴

1. Dr. Debasish Kumar Saha, FCPS (Medicine), Registrar, Dept of Critical Care Medicine, BIRDEM.
2. Dr. Madhurima Saha, FCPS (Medicine), Registrar, Dept of Critical Care Medicine, BIRDEM
3. Dr. Mohammad Omar Faruq, MD, FACP, FACEP, FCCM, FCPS, Professor, Dept of Critical Care Medicine, BIRDEM.
4. Dr. A.S.M. Areef Ahsan, FCPS (Medicine), MD (Chest), MD (Critical Care Medicine), Associate Professor and Head, Dept of Critical Care Medicine, BIRDEM.
5. Dr. Kaniz Fatema, FCPS (Medicine), MD (Critical Care Medicine), Assistant Professor, Dept of Critical Care Medicine, BIRDEM.
6. Dr. Fatema Ahmed, FCPS (Medicine), MD (Critical Care Medicine), Junior Consultant, Dept of Critical Care Medicine, BIRDEM.

Corresponding Author:

Dr. Debasish Kumar Saha,
FCPS (Medicine), Registrar, Dept of Critical Care Medicine, BIRDEM.
Email: debasish81dmc@yahoo.com

There is currently no cure and no standardized course of treatment for Fahr disease. The available treatment is directed at symptomatic control.

There is no reliable correlation between age, extent of calcium deposits in brain, and neurological deficit. Progressive neurological deterioration generally results in disability and death.

References:

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