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Fahr´s disease: idiopathic abnormal basal ganglia calcifications in the brain

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Image in medicine

A 56-year-old diabetic man complains for isolated chronic headaches for many years. He denied any seizure, involuntary limb movements or blurring of vision. Physical examination was unremarkable for any focal neurological deficits. Cranial computed tomography (CT) scan showed extensive symmetrical calcifications of the bilateral basal ganglia and over the lateral periventricular areas (figure). Laboratory studies including serum calcium, phosphate, parathormone, thyroid hormones and vitamins were within normal range. There was no other obvious etiology or family history of similar illness. Based on these findings, the patient was diagnosed with Fahr´s disease and was managed conservatively. Fahr´s disease (FAD),

also known as “familial idiopathic basal ganglia calcification”, is a rare neurological disorder characterized by abnormal calcified deposits in the basal ganglia, cerebellum and subcortical white matter. FAD differs from Fahr´s syndrome in which there is an underlying cause for brain calcifications. Neurological and psychiatric symptoms are variable and may include deterioration of motor function, Parkinsonism, seizures, headache, dysarthria, spasticity, visual disturbances, dementia, psychosis, and affective disorders. FAD could be easily confused with other more common neurological and psychiatric disorders. It is particularly important to not miss the treatable etiologies of Fahr´s syndrome. The diagnosis is based on the CT scan because it is difficult to identify calcifications by routine magnetic resonance imaging. Treatment is essentially symptomatic.

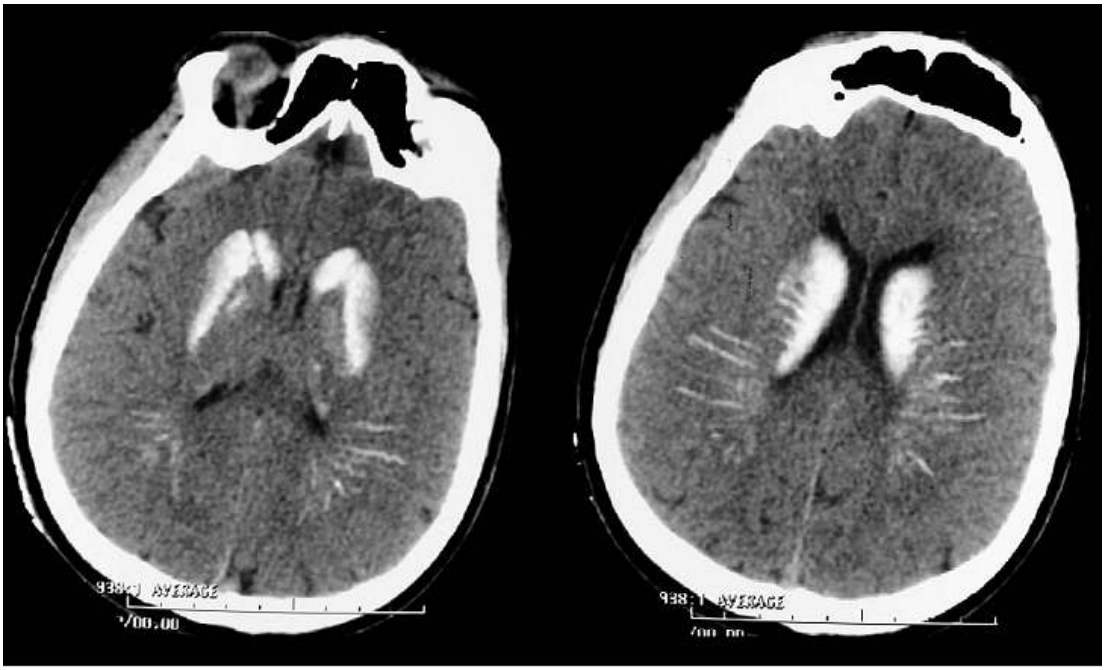

Figure 1: Fahr´s disease: idiopathic abnormal basal ganglia calcifications in the brain