Not So ‘Fahr’ Off

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INTRODUCTION:

Fahr’s disease is a rare inherited or sporadic neurological disorder. It is characterized by calcification of motor regions including the basal ganglia, thalamus, dentate nucleus, cerebral cortex, cerebellum, subcortical white matter, and hippocampus (1). Here we present a case of a young gentleman who was evaluated at our primary care clinic.

CASE:

A 38-year-old gentleman with a past medical history of ulcerative colitis presented with two months of dizziness brought on by bending over. The episodes were brief, lasting a few seconds. He denied any other symptoms. There was no history of trauma or precipitating event. His physical exam, including a complete neurological exam was unremarkable. Orthostatic vital signs were negative. A CT scan of the head showed extensive bilateral symmetric calcifications involving the basal ganglia, thalamus, dentate nuclei, and subcortical white matter of the cerebrum including involvement of both frontal, temporal, parietal and occipital lobes as well as the midbrain and pons (see images). The distribution pattern of bilateral calcifications was consistent with Fahr’s disease. His calcium, parathyroid hormone, vitamin D and phosphorus levels were normal. No medication was initiated as treatment is symptomatic.

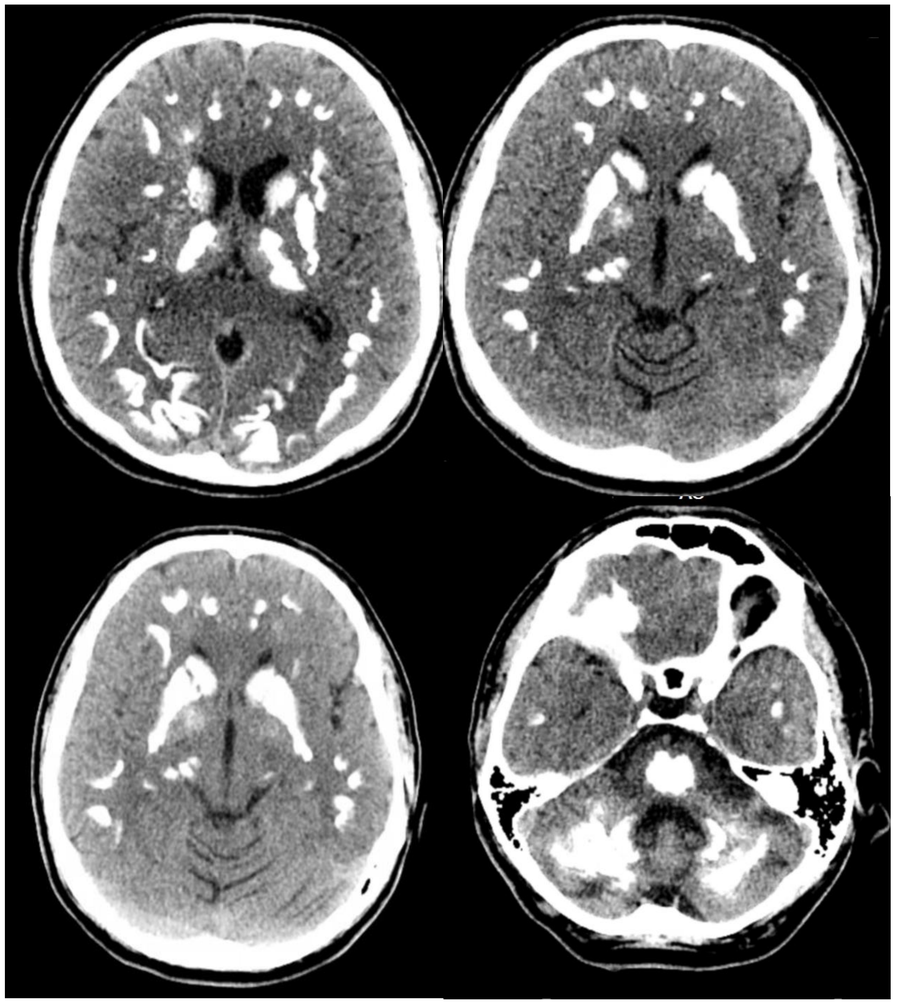
DISCUSSION:

In Fahr’s disease (FD) there is a progressive calcium deposition in the basal ganglia. It is important to distinguish Fahr’s disease from Fahr’s syndrome where certain underlying conditions such as disorders of the parathyroid gland, mitochondrial myopathies, dermatological abnormalities and infections which can result in calcification of the basal ganglia can be seen. It should also be differentiated from “radiological” basal ganglia calcification without clinical features. The frequency of asymptomatic basal ganglia calcification seen on CT is about 0.9% in the general population. Clinical manifestations of Fahr’s disease ranges from headaches, seizures, memory impairment to movement disorders including Parkinsonism, chorea and tremors. It is important to note that patients with this disease are at increased risk for neuroleptic malignant syndrome when treated with antipsychotic drugs. Another important feature is that clinical findings are localized to the central nervous system in Fahr’s disease (2). It is extremely unusual for such widespread calcification with minimal symptoms. Despite a thorough laboratory work-up of the patient, no underlying condition was found. At a two year follow up his symptoms were persistent but stable.

CONCLUSION:

Intracranial calcification can be associated with a range of abnormalities especially hypoparathyroidsim. It is important to rule out secondary causes before the diagnosis of Fahr’s Disease is made. There is currently no treatment that can halt or slow down the progression of intracranial calcification. There is no data to suggest if these individuals have a normal life span. Further studies are needed to determine the life span and potential treatment for such individuals.

IMAGES:



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