FAHR'S SYNDROME AND ACUTE ISCHEMIC STROKE: A CASE REPORT

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INTRODUCTION

Familial idiopathic basal ganglia calcification (Fahr's disease) is a rare neurodegenerative disorder characterized by symmetrical and bilateral calcification of the basal ganglia. We describe a rare case of Fahr's disease secondary to hypoparathyroidism presenting with acute ischemic stroke.

OBJECTIVE

To report an atypical case of acute ischemic stroke associated with Fahr's disease and to emphasize this disease as a possible etiology in similar cases.

CASE REPORT

A 55-year-old female was admitted to our emergency department with a 1-day history of a sudden stabbing headache, followed by numbness and weakness on left hemibody. She was previously diagnosed with hip osteoarthritis and tension-type headache. Family history was unremarkable.

Her neurologic examination showed complete disproportionate left hemiparesis, with Babinski sign. NIHSS scored 3 points. Head CT disclosed diffuse calcifications in basal ganglia, cerebellum and periventricular white matter (Figure 1).

ECG had sinus rhythm, carotid Doppler ultrasound and transthoracic echocardiogram resulted normal.

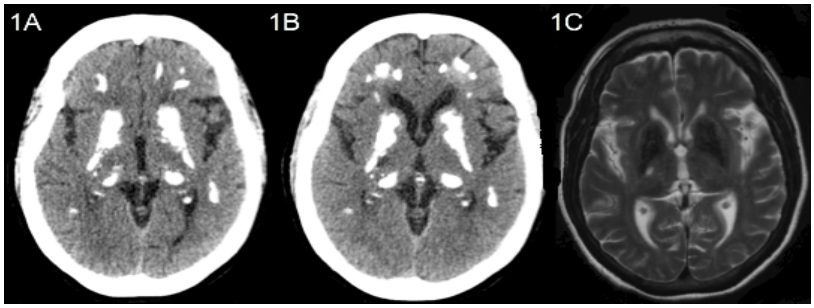

Figure 1. Serial CT and MRI. First head CT (A) revealed diffuse calcifications in basal ganglia, cerebellum and periventricular white matter. Following CT (B) showed hypodensity on the right thalamus. Subsequent T2 weighted MRI (C) demonstrated lacunar hypointensity on the right thalamus.

Twenty-four hours later head CT was repeated, showing a lacunar hypodensity on the right thalamus. PTH levels were 5 pg/mL.

Subsequently, a brain MRI (Figure 1C) confirmed an acute thalamic infarction on DWI. The low levels of calcium and PTH were compatible with the diagnosis of primary hypoparathyroidism.

Treatment was started with aspirin, atorvastatin, vitamin D3, calcium and calcitriol, with adequate control of calcium levels and no recurrence in one year.

DISCUSSION

The association between young-onset ischemic stroke and Fahr's disease has yet to be further studied.

Transient ischemic attack-like episodes had been linked to Fahr's disease in two case reports.

There is only one report demonstrating acute infarction in Fahr's disease with positive MRI findings.

The proposed underlying pathogenic process is the presence of extensive calcium and mineral deposits in affected vessels predisposing to ischemia.

Previous pathologic studies showed calcium and other mineral deposits in the walls of capillaries, arterioles, small veins and in the perivascular spaces.

CONCLUSION

Although extremely rare, it is important to account for the association between Fahr`s disease and acute ischemic stroke, given its potentially treatable nature, with control of calcium and phosphate levels.

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