

# Bilateral striopallidentate calcinosis in a patient with myasthenia gravis

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## Abstract

Bilateral symmetric calcification involving striatum, pallidum with or without deposit in dentate nucleus is reported from asymptomatic individuals to a variety of neurological conditions. Clinically it may present with an array of movement disorders, dementia, cerebellar impairment and speech disorder. We report a myasthenia gravis patient with dysarthria which did not respond to pyridostigmin. Brain MRI showed bilateral striopallidentate calcinosis (BSPDC). To our knowledge, there is no reported case having anti-acetylcholine receptor antibody and massive intracranial calcifications. This case indicates that BSPDC may develop in diverse neurological disorders. (*J Med Life Sci* 2015;12(2):37–38)

**Key Words :** Anti-acetylcholine Receptor Antibody, Calcification, Myasthenia Gravis, MRI

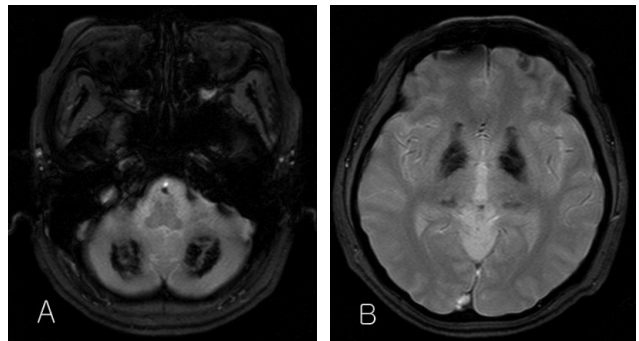
## Introduction

Fahr's disease refers to a sporadic or familial idiopathic calcification of the basal ganglia that may lead to neurological, psychiatric, and cognitive abnormalities. According to the recently proposed classification of bilateral cerebral calcification, the present patient may be regarded as bilateral striopallidentate calcinosis (BSPDC)<sup>1)</sup>. Overlap of neurologic manifestations such as hypokinetic movement disorder associated with cognitive impairment and cerebellar signs were often present. Calcification of basal ganglia and other subcortical nuclei has resulted in a considerable dilemma as to its significance and relationship to varieties of neurological disorders. We describe a myasthenia gravis patient with BSPDC.

## Case Report

A 48-year-old woman presented with a 10 month history of ptosis and dysarthria. Her medical history was remarkable for hypertension. She had no history of alcohol or drug abuse. The family history was unremarkable. Bilateral ptosis and a soft nasal voice were present, but the

physical examination was otherwise normal. Repetitive nerve stimulation test showed decremental responses in orbicularis oculi and trapezius muscles. The titer of anti-acetylcholine receptor antibody was high at 18 nmol/L. She received pyridostigmine and ptosis was relieved, but dysarthria did not improve. Brain MRI showed extensive bilateral calcifications in the dentate nuclei of the cerebellum, basal ganglia and centrum semiovale (Fig. 1).



**Figure 1.** MRI findings in patient. Brain MRI shows bilateral symmetrical calcification in the dentate nuclei (A) and basal ganglia (B).

Laboratory and endocrinological investigations were all normal. In particular, screening for hypoparathyroidism, iron or copper deficiencies, mitochondrial encephalopathy and encephalitis were all negative. No thyroid disease or vitamin deficiency could be found. Although she had no family history, we investigated the other families including the

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patient's younger brother, sister, and daughter and son. All the families were negative on brain CT imaging. Based on the MRI findings, normal parathyroid function and the patient's clinical symptom, we made a diagnosis of BSPDC.

## Discussion

The main aspect of this report is that massive intracranial calcifications were detected in a patient with myasthenia gravis. It is uncertain whether dysarthria is caused by myasthenia or basal ganglia calcification. Combination of BSPDC with other neurological conditions have been reported, among them corticobasal degeneration<sup>2)</sup>, mitochondrial encephalopathy, central nervous system lupus, motor neuron disease, or Alzheimer disease<sup>3)</sup>. To our knowledge, there is no reported case having anti-acetylcholine receptor antibody and massive intracranial calcifications.

BSPDC is a rare clinical entity characterized by non-atherosclerotic calcification of the striopallidodentate region bilaterally. A degree of calcification in the globus pallidus and dentate nucleus without specific symptoms can be detected in 40–70% of routine autopsies<sup>4)</sup>. The features of BSPDC can be varied and the diagnosis is established by obtaining a CT or MRI scan of the head and ruling out abnormalities of known calcium metabolism and developmental defects. The major differential diagnosis is hypoparathyroidism.

The mechanism of ectopic calcification and its pathogenetic significance are yet to be clarified fully. It has been suggested that the hyperintense T2-weighted MRI may reflect a slowly progressive metabolic or inflammatory process in the brain, which subsequently calcifies and is probably responsible for the neurologic deficits observed<sup>5)</sup>. Considering that calcification of basal ganglia is also commonly observed in cases of carbon monoxide poisoning, abnormal oxygen metabolism might be one of the common mechanisms of this ectopic calcification in brain tissue. According to previous studies, vascular membrane abnormalities may be responsible for the leakage of

plasma-derived fluid, and this, in turn, may damage the neurophil and result in mineral accumulation in the vessel walls<sup>6)</sup>. While the exact reason why the basal ganglia is vulnerable for calcium deposits has not been established, it appears that the basal ganglia is a target for many other deposits in addition to various minerals.

This case indicates that BSPDC may develop in diverse neurological disorders. Although the association between myasthenia gravis and intracranial calcifications is unclear, if myasthenic patient has unexplained symptoms, brain imaging is recommended.

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