



Case Report

Extensive bilateral striocerebellar calcifications associated with Hashimoto's hypothyroidism

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ABSTRACT

Intracranial calcifications are not uncommon radiographic findings and usually are nonpathological. However, striocerebellar calcifications may result from metabolic derangements, one common one being hypoparathyroidism. Here, we report a case of Hashimoto's hypothyroidism where the patient experienced disturbance of consciousness and urinary incontinence and had extensive bilateral striocerebellar calcifications. After supplementation with levothyroxine, there were no more episodes of disturbed consciousness. Striocerebellar calcifications may be associated with metabolic derangement as a result of hypothyroidism, although the mechanism by which calcification occurs is not fully understood. Evaluation of thyroid function is recommended in patients with disturbed consciousness and striocerebellar calcifications.

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1. Introduction

Intracranial calcifications are occasionally seen on radiographic studies. Calcifications in the cerebellum can be detected using computed tomography (CT) and usually occur in concert with basal ganglia involvement. Traditionally, striocerebellar calcifications are considered to develop on an idiopathic basis or as the result of physiological processes [1–3]; in only a minority of patients, it has been shown that there are abnormalities in calcium and phosphorous metabolism. In addition, several conditions are known to be associated with such calcifications; these include metabolic abnormalities, such as hypoparathyroidism or hyperparathyroidism; congenital diseases, such as tuberous sclerosis and Cockayne syndrome; and infectious diseases, such as toxoplasmosis, cysticercosis, and AIDS [4]. Only limited reports have documented the association between intracranial calcifications and hypothyroidism. Burke et al [5] described a frequent association of unexplained striocerebellar calcifications and hypothyroidism over a 3-year period. In this report, we describe a case of extensive bilateral striocerebellar calcifications associated with hypothyroidism because of Hashimoto's thyroiditis.

2. Case report

A 68-year-old woman was brought in by her family because of disturbance of consciousness and urinary incontinence that had lasted for hours with spontaneous recovery 1 day before clinical evaluation. During the period when she showed dull responses, there were no limb convulsions or psychotic symptoms noted and no food or drugs were ingested. The patient's blood sugar level had been checked using a glucose meter and was not low. The patient did not remember what happened during that episode and only a residual dizziness was mentioned when she was evaluated at the neurological clinic 1 day after the event. She suffered from diabetes mellitus and had been treated with oral hyperglycemia agents for many years. The family denied previous psychomotor problems. Physiological examination revealed a thyroid goiter. No coarse facial features, hoarseness, or short stature was observed. Neurological examinations revealed only minimal impairment of her recent memory. No abnormality in pyramidal, extrapyramidal, and cerebellar system was identified. Laboratory examinations show a reduced concentration of free thyroxine (0.52 ng/dL; normal = 0.71–1.85 ng/dL) and an elevated concentration of thyroid-stimulating hormone (11.55 μIU/mL; normal = 0.4–5.5 μIU/mL). The triiodothyronine level was normal (95.45 ng/dL; normal = 70–190 ng/dL). Serum antimicrosomal antibody was found to be positive and remarkably high (1: 25,600; normal: negative).

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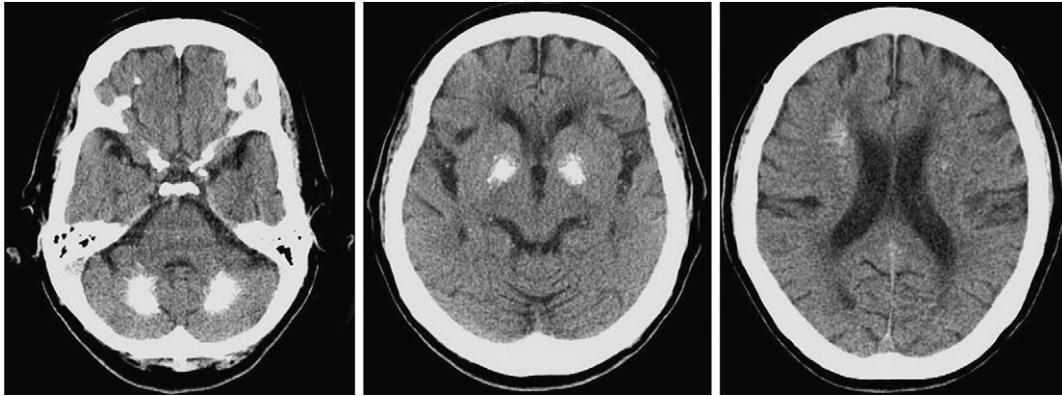


Fig. 1. Noncontrast computed tomography of the head showing extensive bilateral symmetric calcifications; these are most significant in the cerebellum and basal ganglia.

The thyroid was asymmetric in thickness with diffuse hypoechogenicity when examined by ultrasonography. The serum levels of calcium, phosphate, parathyroid hormone, and blood sugar were normal. Electroencephalography revealed occasional diffuse slow waves. CT of the head without contrast showed extensive bilateral symmetrical calcifications; these were most significant in the cerebellum, basal ganglia, and periventricular regions (Fig. 1). Levothyroxine was prescribed to alleviate her hypothyroidism caused by Hashimoto's thyroiditis. The patient indicated that she had had no further memory loss, disturbances of consciousness, or urinary incontinence during the last 6 months when assessed at follow-up.

3. Discussion

We report here a case of Hashimoto's hypothyroidism where the patient experienced disturbance of consciousness and had extensive bilateral striocerebellar calcifications. The possibility of hypothyroidism-related disturbance of consciousness was highly suspected in our patient, although other causes, such as a seizure or hypoglycemia, could not be totally ruled out. However, the level of blood sugar had been checked during the event and was normal. There were no limb convulsions, whereas the patient had disturbed consciousness; and electroencephalography did not reveal significant evidence of epileptiform discharges. Striocerebellar calcifications are often associated with metabolic derangements, usually as a result of hypoparathyroidism or hyperparathyroidism. However, when the serum levels of calcium, phosphate, and parathyroid hormone were checked for abnormalities in calcium metabolism that might cause the intracranial calcifications, they were found to be normal. Overall, there was no other identifiable factor responsible for the extensive bilateral striocerebellar calcifications seen in our patient except for the hypothyroidism. The correlation between striocerebellar calcifications and the neurological symptoms in our patient remains unverified and controversial.

Intracranial calcifications develop much more frequently in the basal ganglia than in the cerebellum and have been shown to be present at frequencies ranging from 0.02% to 0.64% [3]; cerebellar calcifications usually occur in concert with basal ganglia involvement. The correlation between striocerebellar calcifications and neurological symptoms is controversial based on previous studies; nonetheless, asymptomatic patients and patients with nonspecific symptoms, such as dementia, have been mentioned in cases with striocerebellar calcifications [5–7]. The clinical significance of bilateral striocerebellar calcifications therefore remains elusive and most cases probably develop from various unidentified physiological processes. A minority of patients have been shown to have abnormalities in calcium and phosphorous metabolism. A number

of diseases have been reported to be associated with intracranial symmetric calcifications. These include hypoparathyroidism and other metabolic diseases, genetic diseases, and various sporadic condition and these patients do not have any abnormalities of calcium metabolism. The association of intracranial calcifications with hypothyroidism has been demonstrated in a limited number of reports [5–8]. Halpern et al [8] reported a frequency of 30% (15 of 50 subjects) for brain CT-detectable intracranial calcifications during two studies of endemic cretinism. Burke et al [5] described an association (4 of 6 patients) between unexplained striocerebellar calcifications and hypothyroidism over a 3-year period. The pattern of intracranial calcification seen in our patient exhibits a predilection toward the basal ganglia and dentate nuclei; however, involvement of thalamus, cerebral and cerebellar cortices, and centrum semiovale also can be observed. The contents of the deposits included mucopolysaccharide colloid material; hydroxyapatite; and trace amounts of iron, zinc, magnesium, and manganese. The microscopic localization of calcifications is mainly in and around the small cerebral blood vessels [9,10]. However, the cause of striocerebellar calcifications is still not clear, and hypoparathyroidism is only one of several metabolic diseases that cause the deposition of colloid material in and around the small cerebral arteries. The presence of local levels of alkaline phosphatase located in endothelial cells has been proposed as an important factor that may be responsible for the preferential positioning of the calcifications in the arteriolar and capillary walls [2]. Manyam et al [11] reported that the level of homocarnosine, a central nervous system-specific peptide, in cerebrospinal fluid was increased twofold in patients with autosomal dominant bilateral striopallidodentate calcinosis; nonetheless, in sporadic cases, there has been no detectable increase in homocarnosine found and a decrease in the level of histidine was also noted.

Striocerebellar calcifications may be associated with metabolic derangement as a result of hypothyroidism, although the mechanism by which calcification occurs is not fully understood. Evaluation of thyroid function is recommended in patients with disturbed consciousness and striocerebellar calcifications, particularly in cases where there is no identifiable abnormality in calcium metabolism. CT-detected symmetric intracranial calcifications should not be considered to be just "physiological" before there has been a thorough clinical investigation. The relationship between the intracranial calcifications and hypothyroidism clearly needs further investigation.

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