CEREBELLAR ATAXIA AS A PRESENTING FEATURE OF HYPOTHYROIDISM

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Abstract

Context. Besides its typical features, hypothyroidism comes to notice sometimes with neurologic features like reversible cerebellar ataxia, dementia, peripheral neuropathy, coma, etc. Therefore hypothyroidism should be suspected in all cases of cerebellar ataxia, as it is easily treatable.

Objective. Here we illustrate a case of hypothyroidism initially reported with cerebellar ataxia.

Case report. A 40 year-old male presented with history of gait-ataxia. His investigations revealed frank primary hypothyroidism with positive anti-TPO antibody. The patient was put on thyroxine and he improved completely within eight weeks.

Conclusions. This case report emphasizes that hypothyroidism can present with ataxia as one of the initial features. Therefore, hypothyroidism should be considered in all cases of cerebellar ataxia as it is a reversible cause of ataxia.

Key words: hypothyroidism, cerebellar ataxia, thyroxine.

INTRODUCTION

Hypothyroidism is a common medical condition in the general population. Its common systemic manifestations include: fatigue, constipation, cold intolerance, weight gain, hair loss, dry skin, and hoarseness. A variety of central and peripheral nervous system manifestations are common in patients with hypothyroidism. In many cases, the neurologic manifestations occur in conjunction with the systemic features of the disease and may be noted only incidentally. However, symptoms and signs of neurologic dysfunction may develop in some patients and can contribute to significant disability. Most of these complications are partially or fully responsive to thyroxine replacement (1). Hypothyroidism causing ataxia is often an overlooked cause. Here we describe a case of hypothyroidism with cerebellar ataxia.

CASE REPORT

A 40-year-old male was reported to us with history of instability of gait from 3 weeks which was acute at onset and then progressive. There was no history of weakness in any part of the body, headache, vomiting, the convulsions or alteration of sensorium. There was no history of trauma to the head, fever or drug intake. On examination his vitals were normal. He has puffiness in the face. Cognitive functions were normal. Neurological examination showed gait ataxia, dysarthria and dysmetria on finger-nose and heel-to-knee tests. There was horizontal nystagmus. The gait was wide-based and there was a tendency to fall to the right side. His fundus was normal. His power was normal but had hung up reflexes. Sensory system was normal. His hemogram was normal. Serum electrolytes, blood sugar, renal and liver function tests were normal. Serum CPK (creatinephosphokinase): 1220U/L. Total cholesterol: 225 mg/dL, Triglycerides: 275mg/dL, HDL cholesterol: 36 mg/dL, LDL cholesterol: 150mg/dL, VLDLcholesterol: 35mg/dL.

T3: 60 (normal range 80-200ng/dL) T4: 2.5ug/dL (normal range 4.5-12.5), TSH: 88 mU/L (0.27-5.5). The serum anti-TPO antibody was elevated (500IU/L; reference: < 34). His chest X-ray was normal. MRI brain was normal (Fig. 1). He was started on 75 µg of thyroxine. Thyroid function tests after 2 months of therapy were normal. His lipid profile also normalized (Table 1). He was not having ataxia when last seen.

DISCUSSION

Hypothyroidism is one of the causes of acute onset ataxia. Stroke, viral encephalitis and drugs can also cause acute cerebellar ataxia. Mass lesions in the posterior fossa, infections such as HIV, and vitamin deficiencies like B1 and B12, alcohol and paraneoplastic syndromes are causes of sub acute onset cerebellar ataxia in an adult. Hypothyroidism has been

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Figure 1. T1 Weighted MRI showing normal cerebellum.

recognized as a cause of gait ataxia (2).

In early case series of patients with adult-onset hypothyroidism, a wide-based gait ataxia was a prominent feature in a considerable number, around 10 to 30% of cases (1, 3). In many cases, this was the presenting feature, and variably included clumsiness, intention tremor, dysmetria and dysarthria (4). Treatment of the hypothyroid state led to improvement or resolution of the cerebellar features.

Cerebellar dysfunction is also a feature of congenital hypothyroidism. Up to 40 percent of children and adolescents with congenital hypothyroidism have been shown to demonstrate mild findings of cerebellar dysfunction, such as intention tremor, difficulty with tandem gait, clumsy running, fine motor dysfunction, and unsteady stance (5). Animal studies have shown that hypothyroidism interferes with cerebellar development (6). The cause of cerebellar ataxia in association with hypothyroidism remains unidentified. TRH (thyrotropin releasing hormone) besides its role in energy homeostasis has been studied for its function in cerebellar ataxia (7). In animal models, TRH has been associated with improvement in ataxic gait. TRH analogs are now being developed not only for their anti-ataxic effect but also for the arousal effects and treatment of depression (8). No typical morphological changes in the cerebellum have been described. It is assumed that a thyrogenic, specific metabolic factor is responsible which aggravates the already existing non

Table 1. Thyroid functions and lipid profile before and after thyroxine treatment

Parameter	Before treatment	After treatment
T3(ng/dL)	60	120
T4(ug/dL)	2.5	8.6
TSH (mU/L)	88	2.8
Total cholesterol(mg/dL)	225	180
Triglycerides(mg/dL)	275	150
HDL(mg/dL) ^{\$}	36	55
LDL(mg/dL) [#]	150	95
VLDL(mg/dL) [*]	35	30

HDL=High density lipoprotein\$, LDL=Low density lipoprotein#, VLDL=Very low density lipoprotein*.

specific cerebellar changes. The prognosis is very good. On the other hand, cerebellar ataxia resulting from congenital hypothyroidism has typical histological cerebellar changes and prognosis is very poor unless thyroxine treatment is started soon after birth (9).

Cerebellar degeneration in patients with raised antithyroid antibodies may be immune mediated. Some cases of hypothyroidism associated cerebellar ataxia do not reverse with normalization of their thyroid state with thyroid hormone replacement and may be harboring undiagnosed Hashimoto's thyroiditis (10). Dinkar *et al.* have reported a case of Hashimoto's encephalopathy presenting purely with subacute cerebellar syndrome (11). A similar case has been reported by Nakagawa *et al.* (12). Necropsies of patients with hypothyroidism and cerebellar ataxia have been reported. Barnard *et al.* described degenerative changes in the cerebellum, particularly in the anterosuperior portion of the vermis, together with atrophy of ventral portion of the pons, transverse pontine fibres, and middle and superior peduncles in a patient with hypothyroidism and cerebellar ataxia (2). A case of gait instability has been described in a patient with subclinical hypothyroidism with complete resolution after thyroxine treatment (13).

In conclusion, this case report highlights that hypothyroidism should be considered in all cases of cerebellar ataxia, as it is a reversible cause of ataxia.

Conflict of interest

The authors declare that they have no conflict of interest concerning this article.

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