 Persistent Hemichorea Associated with Thyrotoxicosis
Masayuki Baba, Akinori Terada*, Ryohei Hishida*, Muneco Matsunaga, Yasunori Kawabe** and Kazuo Takebe*

We describe a case with unilateral chorea associated with thyrotoxicosis. A 23-year-old female with no family history of neurological diseases acutely developed choreic movements of the left extremities during gross thyrotoxicosis. CT scan and MRI study demonstrated no abnormality. Single-photon emission CT with technetium Tc 99m-labeled hexamethylpropyleneamino oxime revealed normal cerebral perfusion. Although the choreic movements were partially improved by dopamine antagonist, they persisted for two months until successful treatment of the thyrotoxicosis finally abolished these movements. Increased sensitivity of dopamine receptors may be responsible for persistent choreic movements in thyrotoxicosis. (Internal Medicine 31: 1144–1146, 1992)

Key words: choreoathetosis, hyperthyroidism, dopamine antagonist, MRI, SPECT

Introduction

Various neurological symptoms may accompany hyperthyroidism. Among them choreoathetosis is a rare but recognized manifestation of thyrotoxicosis (1). More than ten cases have been reported from the Caucasian population in the past 20 years; only one case has been published in the Japanese literature during the same period (2). We recently treated a 23-year-old thyrotoxic female with acute onset of hemichoreic movements. She was the only patient with choreic movements among over 2000 patients evaluated for hyperthyroidism at the endocrinology unit of the Third Department of Medicine, University Hospital of Hirosaki, during the 20-year period from 1971.

Case Report

A 23-year-old Japanese woman visited us because of severe involuntary movements in the left extremities. She had no family history of neurological disease. Since the age 16 she had noticed occasional palpitation and night sweating. Her family noticed that she had exophthalmic eyes at the age of 18. One night in June 1990, she felt dullness in the left arm. She then noticed that her left hand trembled. Two days later violent involuntary movements of the left arm developed acutely. Her left arm was easily injured as it hit anything on her left side and she had to hold the left hand with the other hand. Because the involuntary movements of the arm worsen and expanded to the left leg and the neck, she was referred to the neurological clinic.

Her height was 157 cm and body weight was 55 kg. She was alert and well oriented, but a little irritable. Blood pressure was 120/68 mmHg and pulse rate was 84/min. Her thyroid gland was enlarged and a diffuse soft goiter was palpable. Bruit was audible over the goiter. Both eyes were exophthalmic. The skin was moist and excess sweating was noted over the whole body.

She had violent choreic movements of her left arm. They were jerky, purposeless, intermittent, irregular movements, dominantly in the distal joints. The left leg was also involved distally in similar involuntary movements, but they were mild enough for her to stand without support. She also showed a quick neck rotation and protruded tongue. No involuntary movements were seen in her face and right side of the body. There was a generalized but left-side dominant hypotonia. Fine positional tremor was noted on the fingers of both sides. Muscle strength was normal and tendon reflexes were symmetric and slightly brisk. Vibratory and skin sensation, and proprioception were all intact. There was no ataxia.

The hematological investigation, serum electrolytes, renal and liver function tests, copper studies were all normal, except for a low cholesterol level of 103 mg/dl.

From the Department of Neurology, Institute of Neurological Diseases, and * the Third Department of Medicine, Hirosaki University School of Medicine, Hirosaki, and ** Mutsu General Hospital, Mutsu
Received for publication January 10, 1992; Accepted for publication July 6, 1992
Reprint requests should be addressed to Dr. Masayuki Baba, the Department of Neurology, Institute of Neurological Diseases, Hirosaki University School of Medicine, 5 Zaifucho, Hirosaki 036, Japan

Internal Medicine Vol. 31, No. 9 (September 1992)
Thyrotoxic Chorea

Fig. 1. Sequence of abnormal choreic movements of the left arm (No.1 to No.7).
Note quick, jerky, irregular movements of the whole arm. Some writhing features are seen in the distal segment.

There was no acanthocytosis. CRP, ASLO and RA were negative. Laboratory tests for thyroid function were as follows; BMR: +89%, serum T3: 8.3 ng/ml, T4: 28.4 μg/dl, free T3: 13.6 pg/ml, free T4: 6.7 ng/dl, TSH: <0.1 μIU/ml, thyroglobulin: 243 ng/ml, thyroid test: negative, and microsome test: negative. Other auto-antibodies including anti-cardiolipin antibody were all negative. Daily urinary excretion of norepinephrine, epinephrine, DOPA, metanephrine, normetanephrine, VMA and HVA were all within normal limits. Electro-encephalogram showed no paroxysmal discharges. No abnormalities were detected by electrocardiogram or echocardiogram. The CT scan performed with contrast enhancement on the fifth day showed no abnormalities. Axial and coronal MRI performed on a 1.5-Tesla system on the 17th day demonstrated no abnormalities in any of T1-weighted, T2-weighted or proton images. A single-photon emission CT (SPECT) performed by GCA-70AS gamma-camera (Toshiba) with technetium Tc99m-labeled hexamethyl propyleneamine oxime showed no abnormal perfusion on the 20th day.

She was started on thiamazole 30 mg/day and metoprolol 60 mg/day, but this had a very slight effect on choreic movement. When chlorpromazine 0.5 to 1.0 mg/day was given in place of metoprolol, the involuntary movements started to be suppressed. Thiamazole was then increased to 60 mg/day. One month later, when serum T3 and T4 was decreased to 3.4 ng/ml and 21.5 μg/dl, respectively, the choreic movements dramatically subsided, and were nearly abolished two months later. For over a year, she has been euthyroid and has had no return of choreic movements.

Discussion

Several etiologies may cause choreic movement in young people, among which Huntington’s disease, Sydenham’s chorea, neuroleptic-induced hyperkinesia and chorea gravidarum are well known condition (1). Some other causes which damage the striatum may be found in choreic patients: brain tumors, slight bleeding and small lacunar lesions are most frequent. All of these possibilities were ruled out in the present case by

Fig. 2. Samples of the patient’s writing ability before (left) and two weeks after (right) the commencement of chlorpromazine administration. Note some improvement of irregular writing traces following treatment. The choreic movements were further subdued by successful treatment of hyperthyroidism.
laboratory tests and brain imagings.

Thyrotoxic chorea has been known since 1888, when Sir William Gowers referred to it in the section of exophthalmic goiter of his famous textbook of neurology (3). He wrote that true chorea is thought to exist in a few cases in childhood or youth. Early in this century, 20 years after the first report by Gowers thyrotoxicosis was already regarded as one of the important causes of choreic movements, because some other reports (4–5) had confirmed the relationship between thyrotoxicosis and chorea.

Although hyperthyroidism is not a rare disease, only one case has been reported in the Japanese literature since 1970 (2). On the other hand, 14 cases were published in the European and North American literature (5–16). Although the precise incidence rate of chorea in hyperthyroidism is obscure, the rarity of Japanese case reports possibly suggests a lower incidence in Japanese than in Caucasian patients. Gowers pointed out the possible transformation of tremor to choreic movements in thyrotoxicosis (3). A similar possibility was recently raised again by Marks and colleagues (10), because carbimazole was effective in one of their cases. It was thus suggested that the tremor of mild thyrotoxicosis was one end of a spectrum and choreoathetosis—represented the most severe form of incoordination in thyrotoxicosis. However, the effects of beta-blocking agents are never consistent. As there was no effects reported in many cases with thyrotoxic chorea (5, 12, 16), recently, the sympathetic hypothesis is, thus, a minority opinion.

The question of the efficacy of dopamine antagonists has been recently raised (14–15). In fact, in the present case the choreic movements were partially suppressed by chlorpromazine, and finally subsided with normalization of the thyroid function. Since a similar clinical course was observed in some of the recent cases with thyrotoxic chorea (9, 14), hypersensitivity of dopaminergic receptors to dopamine due to a thyotoxic state is strongly suspected as the cause of chorea (14–17). Since the concentration of homovanillic acid is decreased in the cerebrospinal fluid in hyperthyroidism (17), the production and turnover of dopamine by negative feedback may be decreased. Some investigators (18–19) reported a diminished prolactin response to TRH in hyperthyroidism, suggesting enhanced central dopaminergic tone.

However, it is an enigma why unilateral manifestation can develop on the basis of metabolic derangement. Among 16 reported cases including the present case, three showed hemichorea and 13 cases were bilaterally involved. This is the reason why morphological change in unilateral striatum has been suspected elsewhere (16). The present patient is probably the first case studied using MRI and SPECT, in which the possibility of focal morphological damage of striatum has clearly been ruled out. SPECT revealed no changes in cerebral perfusion. There might be a functional laterality in the striatum, and their critical metabolic levels are possibly different. On this line of thought, one of the 13 bilateral cases is particularly interesting, because the chorea was initially unilateral and later came to affect the other side of the body (13). Further study by PET scan may be useful to achieve a detailed approach to the possible metabolic, functional laterality of the striatum in thyrotoxic chorea.

References