Paroxysmal kinesigenic choreoathetosis in hyperthyroidism

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Summary: Paroxysmal kinesigenic choreoathetosis is an unusual movement disorder often triggered by attempts to use the limbs, and has sometimes been associated with diffuse or focal brain injury. We report its occurrence in hyperthyroidism, with which choreoathetosis has rarely been described in the past without known cause. Choreoathetosis has also occurred with other metabolic and toxic disorders, and the mechanism is uncertain. The development of involuntary movements activated by limb motion during hyperthyroidism suggests an influence by thyroid hormone or metabolic state on motor neurone excitability or motor system organization.

Introduction

Paroxysmal kinesigenic choreoathetosis is an unusual dyskinesia often precipitated by voluntary movement and sometimes mistaken for seizures.¹ It is often familial, but sporadic cases are increasingly recognized and generally have no neuropathological findings.² Sporadic cases have been associated with birth injury,³ thalamic infarction,⁴ and closed head injury.⁵ We recently observed choreoathetosis activated by voluntary movement in a hyperthyroid patient, and suggest it is a variant of the choreoathetosis occasionally reported in thyrotoxicosis.^{6–8}

Case report

A 57 year old woman developed paroxysms of involuntary twisting and jerking of the limbs. These initially involved the right arm, and later came to affect the right arm and leg, and less frequently the trunk and both upper extremities, with intact consciousness but an intermittent prodromal tingling sensation in the involved limbs and frequent soreness and stiffness after an attack. They were generally precipitated by shoulder elevation, internal or external rotation of the arm or leg, extension of the arm, or flexion or extension of the leg. They occurred when standing or rising from the supine position, but happened infrequently when seated. She was sometimes able to diminish the attacks by grasping the affected limb, or by holding it rigid. The movements occurred several times daily, and lasted for one or two minutes.

She also complained of nervousness, weakness, increased perspiration, and heat intolerance. She had otherwise been healthy but had been given desiccated thyroid some years previously, when she had complained of fatigue and weight gain and had had abnormal thyroid tests. She had not had medical follow-up since that time, but had continued to take thyroid for purposes of weight loss and rejuvenation, in a dosage schedule of her own devising.

After the onset of her attacks, she had had a normal computed tomographic (CT) brain scan and electroencephalogram (EEG), and had been given phenobarbital and phenytoin without effect.

Physical examination showed blood pressure 150/80 mmHg, pulse 92 beats per minute, and normal temperature and respiration. Her skin was smooth and her hair fine, but she had no ocular abnormalities. The thyroid gland was soft and within normal limits in size, without nodularity or bruits. Neurological examination was normal except for fine tremor in the outstretched arms, with brisk and symmetrical deep tendon reflexes and flexor plantar responses. Involuntary movements were not precipitated by vibration or elicitation of tendon reflexes. Laboratory results were within normal limits except for thyroid function tests. Serum thyroxine was 167 nmol/l (upper limit of normal, 141), T₃ uptake 0.96 (normal), free thyroxine index 12.5 (upper limit of normal, 11), and thyroid stimulating hormone less than 0.1 mU/l. ¹³¹Iodine thyroid scan showed no focal abnormality of uptake.

Because of the question of seizures, anti-epileptic medications were discontinued and she had a prolonged sleep-deprived EEG. The latter was normal, but she was asked to produce an episode, which she did by raising the right arm above her head. This produced spasms of dystonic movement in the right upper

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extremity, which spread to involve the right leg and proximal left arm, and on which were superimposed rapid choreiform movements of the fingers. She remained conscious and attempted to abort the attack by clenching and releasing her fists, but was unsuccessful. The EEG remained normal throughout and after the attack. Her thyroid supplementation was discontinued, and the attacks subsided. Her other symptoms gradually diminished, and thyroid function tests were normal two months later, at which time she seemed clinically euthyroid.

Discussion

The neurological, ophthalmic, and psychiatric manifestations of hyperthyroidism are well-described.⁹⁻¹⁰ Chorea was described in a Graves' disease patient in 1903,⁶ and reported in a thyrotoxic patient with laboratory confirmation of hyperthyroidism in 1971.⁷ Subsequent cases have been infrequent and sometimes mistaken for psychogenic signs and symptoms.⁸ The mechanism of choreoathetosis was not known in these cases, although a correlation with thyroid hyperactivity was established in recent patients.^{7,8} Our patient also showed a relationship between involuntary movements and thyroid function: her abnormal movements occurred while hyperthyroid and subsided when euthyroid, and her thyroid function studies are consistent with excessive exogenous thyroid supplementation, as suggested by her history. It is unclear why the movements developed after a long period of thyroid ingestion, but it is most likely that she did not become iatrogenically hyperthyroid until she began to

manipulate her own thyroid dose. Her thyroid function studies were mildly elevated, while chorea has previously occurred with marked elevation of thyroid tests and clinically-evident thyrotoxicosis.⁷ She is thus different from the classical thyrotoxic chorea metabolically as well as clinically, but chorea is not always associated with obvious thyrotoxicosis.⁸

Our patient also differs from those described previously in manifesting paroxysmal choreoathetosis, involuntary movements triggered by attempts at voluntary limb use, and resembling a partial seizure but with no evidence of epilepsy, no EEG correlate to the clinical paroxysm, and some degree of voluntary alteration of the movement by effort.^{1,2} This occurs as a familial entity, and is often responsive to anti-epileptic drugs,² but has been reported sporadically³ and ascribed to a disturbance in motor integration, related in at least one instance to a lesion of the ventral posterolateral thalamus.⁴ Its sporadic occurrence after head trauma has also suggested aberrant reorganization of motor pathways, inasmuch as a characteristic latency is seen in the development of kinesigenic involuntary movements after recovery from initial unconsciousness and neurological deficit.⁵ The mechanism of such aberrant reorganization is not known, nor is the mechanism of its apparent precipitation by hyperthyroidism in our patient.

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