Case Report

Paroxysmal kinesigenic dyskinesia manifestation of hyperthyroidism

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Sporadic paroxysmal kinesigenic dyskinesia (PKD) secondary to thyrotoxicosis is an extremely rare entity. A 36-year-old female presented with the features of PKD. Her investigations revealed thyrotoxicosis. Her dyskinesia did not respond to carbamazepine but remitted with the anti-thyroid drug, neomercazole. Perhaps hyperthyroidism-related PKD is a result of a metabolic disturbance of the basal ganglia circuits rather than a permanent and irreversible change.

Key Words: Paroxysmal kinesigenic dyskinesia, Thyrotoxicosis, Thyrotoxicosis-related dyskinesia.

Introduction

Paroxysmal kinesigenic dyskinesia (PKD) is an unusual dyskinesia often precipitated by voluntary movement and characterized by brief unilateral or bilateral chorea, choreothetosis, ballism, or dystonic postures. It is often familial, but sporadic cases have also been reported. PKD may be secondary to diverse etiologies, including multiple sclerosis, ischemic stroke, birth injury, head injury, drug abuse, diabetes mellitus, or hypoparathyroidism, and the sites of lesion can involve the cerebral cortex, basal ganglia, thalamus, medulla, and the spinal cord. There are only three reports describing PKD related to hyperthyroidism, in the literature.

We report a case of PKD in a hyperthyroid patient in whom the involuntary movements disappeared as the patient became euthyroid.

Case Report

A 36-year-old female presented with cramping sensations in the left hand, followed by uncontrollable jerking, predominantly in the left limbs of two years duration. Typically, the attack started with dystonic posturing of either the left hand and wrist or of the left foot and ankle, spreading proximally. Sometimes the attack became choreic and on other occasions there was ballistic movement around either the shoulder or the hip joint. Infrequently, the involuntary movements were confined to the right side only. On rare occasions the attack occurred bilaterally and made her fall. Her consciousness remained clear throughout the attack. The abnormal movements were transient in nature, lasting for 20-30 seconds, mostly triggered by sudden voluntary activities like jumping out of bed, arising quickly from the chair, running to open the door, or sudden turning to look back. These attacks would occur several times in a day with variable inter-attack interval. Sometimes she did not experience any attack for 10-15 days. The attacks never occurred during sleep, nor could they be provoked after hyperventilation, startle, touch, fatigue, excitement or exercise. She denied any of her family members having similar or any other involuntary movements. She never experienced heat intolerance, palpitation, diarrhea, weight loss, or tremors.

Physical examination was non-contributory. Detailed neurological examination was essentially normal. There was no KF ring on slit-lamp examination. Both fundi were normal. Two of these attacks involving the left side could be observed. In one of these episodes she had flexion of the left wrist, hyperpronation of the left hand and abduction of the shoulder; and in another there was abduction of the hip with the foot assuming the attitude of equinovarus deformity. The episodes were stereotyped. There was no axial muscle involvement, nor any oromandibular dystonia or facial grimacing.

Her hemogram, including red cell morphology, blood chemistry for blood sugar, blood urea, serum creatine, serum electrolytes, profile for liver function, lipid, serum copper, ceruloplasmin, serum calcium, phosphorus, and parathormone level were normal. Collagen profile for rheumatoid factor, dsDNA, antinuclear factor were normal. Serum free triiodothyronine was 6.7 pg/ml (normal range 2.3-4.2 pg/ml), free thyroxine 3.8 ng/dl (normal range 0.89-1.8 ng/dl), thyroid-stimulating hormone was 0.15 IU/ml (normal range 0.35-5.5 IU/ml). MR brain was normal. EEG did not show any epileptiform discharge.

She was put on carbamazepine 300 mg in three divided doses, gradually increased to 1200 mgm/day along with neomercazole 30 mg in divided doses. The patient did not show any response initially. However, she responded three weeks after the initiation of the medi-
cation and became euthyroid after about three months. The carbamazepine was gradually withdrawn and she never experienced the attacks after its withdrawal. The attacks, however, recurred four months after the withdrawal of neomercazole. Neomercazole was restarted and she has been asymptomatic on 20 mg/day of neomercazole for the last one year.

Discussion

Neurological manifestations of thyrotoxicosis are not uncommon. However, chorea or choreoathetosis occur rarely with thyrotoxicosis. The movement disorder usually resolves completely with the use of anti-thyroid medications and recurs if hyperthyroidism redevelops. The present case had the features of PKD which was unresponsive to the treatment with carbamazepine but disappeared when the levels of thyroid hormones returned to normal, and recurred after the withdrawal of neomercazole. The involuntary movements in the present case as well as in the previous reports were characterized by prominent dystonic posturing of the limb at the beginning of the attacks and were induced by voluntary movements.

Acquired choreoathetosis has been described in a number of diseases, including hypocalcemia, systemic lupus erythematosus, Schönlein-Henoch purpura, polycythemia, carbon monoxide poisoning, thromboembolism of the posterior cerebral artery and post viral encephalitis.

The clinical profile of the present case was not consistent with any of these disorders.

Besides the induction of PKD, several reports have mentioned the association of hyperthyroidism with the occurrence of chorea. All the previous three reports of PKD with hyperthyroidism had obvious thyrotoxicosis. It has been reported by Drake that PKD may occur when the patient has only mildly elevated levels of thyroid hormones. The present case did not have clinically evident thyrotoxicosis. Chorea is not always associated with obvious thyrotoxicosis. Evolution of choreoathetosis to chorea has been suggested to be associated with idiopathic basal ganglia calcifications. Hyperthyroidism-related PKD or chorea may also share a similar mechanism.

The exact mechanism of how thyroid hormones induce paroxysmal dyskinesia is not clear. Functional hypersensitivity of dopaminergic receptors in the striatum may be responsible for hyperthyroid chorea. Positron emission tomographic studies are usually normal in patients with idiopathic PKD. However, decreased striatal glucose metabolism was observed in most patients with symptomatic chorea, suggesting a pathogenetic mechanism different from that of idiopathic PKD. The present case had no structural lesion on neuroimaging as also in the other three reports. It can be postulated that hyperthyroidism-related PKD is more likely the result of a metabolic disturbance of the basal ganglia circuits rather than a permanent and irreversible change.

Thus, the present case is unique with PKD as the presenting manifestation of sub-clinical thyrotoxicosis.

References


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