PAROXYSMAL KINESIOGENIC CHOREOATHETOSIS: A FREQUENTLY MISDIAGNOSED MOVEMENT DISORDER

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SUMMARY:
Paroxysmal kinesiogenic choreoathetosis is a rare movement disorder, which is frequently misdiagnosed as an epileptic disorder partly because of a good response to anti-epileptic drugs (Phenytoin sodium). At times these patients are mistaken for a conversion reaction. One such case is described.

INTRODUCTION:
Choreoathetosis is usually a continuous condition, but a subgroup of patients have paroxysmal attacks between which they are normal (Mount & Reback, 1940; Lance, 1977). Two varieties are recognized, both inherited as autosomal dominant traits with onset in early life. Paroxysmal kinesiogenic choreoathetosis takes the form of brief (seconds to minutes) attacks of choreoathetosis (dystonia) precipitated by sudden body movement (Plant et al, 1984). Such attacks occur many times each day. Consciousness is preserved in these episodes, which respond to treatment with anti-convulsant drugs such as phenytoin or carbamazepine (Marsden & Quinn, 1990).

Paroxysmal non-kinesiogenic choreoathetosis refers to episodes of abnormal movements without loss of consciousness, not precipitated by movement, lasting longer (many minutes to hours), occurring much less frequently, and sometimes relieved by clonazepam (Fahn & Bressman, 1983; Nair, 1988).

Though most of these diseases are familial in nature, sporadic forms are also reported (Fahn & Bressman, 1983; Nair et al, 1990). We report one sporadic case of idiopathic paroxysmal kinesiogenic choreoathetosis.

CASE REPORT
A 24 year old patient presented with a three year history of bizarre movements while performing some physical activity. Most of these movements occurred when getting up abruptly from a sitting or lying position. Each episode of abnormal movement was characterized by bilateral rapid, stereotyped choreoathetotic movements. The patient used to perform brisk walking like movements involving both the extremities, pelvis and the abdomen for about 20-30 seconds. There were several such attacks in a day. There was no loss of consciousness during the attack and the patient was also able to prevent them if he was mentally prepared for it. There was no family history of any movement disorder. Physical examination did not reveal any positive findings between the attacks. All investigations including biochemical parameters, electroencephalogram and computerized tomographic scan of head did not reveal any abnormality. The movements disappeared totally when he was treated with 300 mg of phenytoin sodium once daily at night.

DISCUSSION
Paroxysmal kinesiogenic choreoathetosis is a rare hereditary disease with autosomal dominant inheritance and with incomplete and variable penetrance. Few sporadic cases have also been reported (Nair et al, 1990). Precipitating factors include alcohol, coffee, tea, emotion, fatigue, changes in temperature, and abrupt movements (Nair, 1988). Paraesthesia of the limb to be affected is the usual prodromal symptom and some of the patients learn to avert these attacks by sensory tricks such as catching hold of the affected limb or stopping the movements by maintaining a posture (Plant et al, 1984).

These cases are often mistaken for epilepsy. Lack of alteration in consciousness, definite precipitating factors, absence of post-epileptic phenomena and at times the patient’s ability to control these episodes by ‘mentally fighting them off’ or by some sensory tricks are important differentiating features. Lack of voluntary control, stereotyped episodes, history of fall or injury, family history and normal psychiatric assessment help to differentiate this disorder from a conversion reaction (Lance, 1977). Dramatic response to anti-epileptic drugs (e.g. phenyl hydantoin) indicates against its functional origin.

Attacks may range in frequency from as many as a hundred per day or as little as one a day. Our patient used to experience 5-10 episodes each day. Similar to the form described by Lance (1977), in our patient, paroxysmal kinesiogenic choreoathetic movements were precipitated by some minor physical activity; however, he never developed it after exercise or vigorous physical activity. In paroxysmal nonkinesiogenic choreoathetosis the attacks are usually precipitated by sleep. Segawa et al (1976) described another variant of the syndrome where a diurnal variation of the dystonia was noted.

There was no positive family history in our patient, indicating its sporadic nature. The dramatic response with phenytoin sodium in usual doses (300 mg to 400 mg/day) adds to the confusion with an epileptic disorder. Our patient became completely symptom free three days after administration of phenytoin.

The frequency of paroxysms may increase during adolescence and decrease after the age of twenty. Usually, the paroxysms continue throughout life, although spontaneous remission may occur. No structural lesion has been reported; in our case also the CT scan was normal. The treatment of the condition is mandatory to avoid fall and accidents, especially while driving (Nair, 1988).
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