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Paroxysmal Dyskinesia

A Case Responsive to Benztropine Mesylate

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Paroxysmal dyskinesias provoked by sudden muscular activity, shock, or emotional upset have been variously called a form of reflex epilepsy (Whitty, Lishman, and FitzGibbon, 1964), dystonic seizures (Lance, 1963), and, in the American journals, paroxysmal choreoathetosis (Mount and Reback, 1940; Pryles, Livingston, and Ford, 1952; Rosen, 1964; Williams and Stevens, 1963). Speculations concerning the site and nature of the responsible lesions have been based on the clinical analyses of a relatively few cases and have not had pathological correlation.

In a descriptive study of seven cases and a review of the published reports, Lishman, Symonds, Whitty, and Willison (1962) characterized the clinical syndrome and suggested a common, though unspecified, aetiology. In a subsequent paper, Whitty *et al.* (1964) reported an additional five cases, proposing that movement-induced seizures are a form of idiopathic epilepsy.

Although dyskinesias have been a symptom of a variety of neurological diseases, this paroxysmal form has only recently been so recorded. Lance (1963) pointed out that dystonic seizures may be either an accompaniment of overt neurological disease, idiopathic or familial. For the latter cases he suggested a dominant mode of inheritance. The cases of Mount and Reback (1940) and Pryles *et al.* (1952) were presumably familial.

The dyskinesias have been reported to begin as early as 6 months and as late as 40 years. They are usually precipitated by either sudden movement or shock. Occasionally they may be heralded by a variety of sensory phenomena or muscle spasms in the involved parts. Emotion enhances the frequency of attacks. They have infrequently been reported with an apparent absence of precipitating factors. Voluntary movement is interrupted in any combination of extremities or trunk by a composite of dyskinesias which include dystonia, athetosis, ballism, or chorea. Bulbar function is often disrupted: this may be manifest by athetotic grimacing, tongue writhing, inability to speak or swallow, and tonic gaze phenomena.

Consciousness is always maintained and frequently, if there is forewarning, incipient attacks can be more or less effectively aborted by the avoidance of sudden movement. The precipitating movement and subsequent dyskinesia are usually stereotyped in a particular individual.

Bladder or bowel incontinence does not occur, and following an attack, which may last a few seconds or several hours, there is seldom clouding of consciousness or other post-ictal abnormality. The major dyskinesia may occur infrequently or serially, however. Minor interictal abnormal movements, such as athetosis, are mentioned.

Concomitant epilepsy has been reported infrequently. Slow wave dysrhythmias represent the most common electroencephalographic finding, though an occasional patient will have spike wave bursts not temporally associated with the dyskinesia.

Favourable response to diphenylhydantoin, phenobarbitone, or primidine has been reported (Lishman *et al.*, 1962; Whitty *et al.*, 1964; Williams and Stevens, 1963; Pryles *et al.*, 1952). However, in many cases there was no improvement with these anticonvulsant drugs. Mount and Reback (1940) noted reduction in the frequency of familial paroxysmal choreoathetosis with scopalamine hydrobromide. Rosen (1964) was able to abort similar attacks in his patient with tincture of belladonna or diphenhydramine. He drew attention to the similarity of the dyskinesia to those seen in phenothiazine toxicity and the favourable response to antiparkinsonian drugs.

The present report describes a case of paroxysmal dyskinesia illustrating a variety of abnormal movements, and responsive to anticholinergic drugs.

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Case Report

A 9-year-old boy was admitted to the Neurology Service of the University of Virginia Hospital in October 1964 because of frequent seizures. He was the product of an uncomplicated 36-week gestation. The birth was assisted with forceps. Immediately thereafter he was seen by the father who described severe moulding of the head. Three days later he seemed normal except for ecchymoses in the area of forceps' marks. He developed normally until at 6 months, during a febrile illness, he exhibited what were described as 'running movements of the legs and jerking of the arms'. Dancing movements of the eyes were also noted. Although not documented, the illness was thought to be an encephalitis. Subsequently he began to have slight constant writhing movements of the fingers and frequent attacks consisting of tonic flexion of the arms and hyperextension of the legs, associated with groaning without loss of consciousness. The abnormal movements continued until the age of 11 months and then gradually ceased. During this illness he had considerable motor regression. Subsequently he began to crawl, and at 3 years of age, with support, took a few steps. However, shortly thereafter he became spontaneously unable to do this and walked only with the help of a brace and harness apparatus. By the age of $3\frac{1}{2}$ years he was able to say a few words, but never developed syntactical speech. He was described as alert and active. At 5 he had several tonic-clonic convulsions with unconsciousness. These continued in serial fashion until controlled by phenytoin sodium and phenobarbitone. After one year, administration of anticonvulsants was stopped and he remained free from seizures. However, he resumed slight writhing movements of the fingers, which worsened when he was startled or pushed. One month before admission, after attending a funeral, he began to have what was described as serial *grand mal* convulsions, which responded poorly to diphenylhydantoin and phenobarbitone.

The father had a history of several post-traumatic *grand mal* seizures. Otherwise there was no family history of relevant neurological disorder. The past medical history revealed only a recent ascaris infection and numerous lacerations. Nose-bleeds occurred during the violent seizures.

On examination he was a well-developed, thin child with healed scars over the face, trunk, and extremities. The nose was distorted and collapsed from previous fractures. He was able to say a few monosyllabic words, and during interictal periods was alert and responsive. He was incontinent. The fundi and optic discs were normal. Visual acuity seemed normal. Slit-lamp examination did not reveal pericorneal pigmentation. The other cranial nerves were intact. There were constant athetoid movements of the fingers, and horizontal, coarse, irregular, asynchronous nystagmoid movements of both eyes, with mild alternating strabismus. Despite seemingly adequate lower extremity strength, he would not walk, but sat and crawled without difficulty. The upper extremities were strong and co-ordinated. Tone seemed normal. Sensation to pain and temperature was intact. The reflexes were equally hypoactive. The plantar responses were flexor.

Shortly after admission he began to have stereotyped attacks of involuntary movement. These usually occurred if he was startled or had emotional upset, but on occasion they could be provoked by sudden movement of his trunk or an extremity. Auditory, visual, or tactile stimuli did not provoke them. The episodes initially occurred 4 to 5 times each day, lasting 5 to 30 minutes. They consisted of severe athetoid and dystonic movements with associated grimacing, tongue writhing and protrusion, and tonic alternating conjugate deviation of the eyes. The attitude of his trunk was often opisthotonic, with the upper extremities adducted and flexed in pronation at the elbow, then suddenly flung upward (Fig.). The legs slowly flexed, then extended in an unsynchronized tonic fashion, with plantar flexion

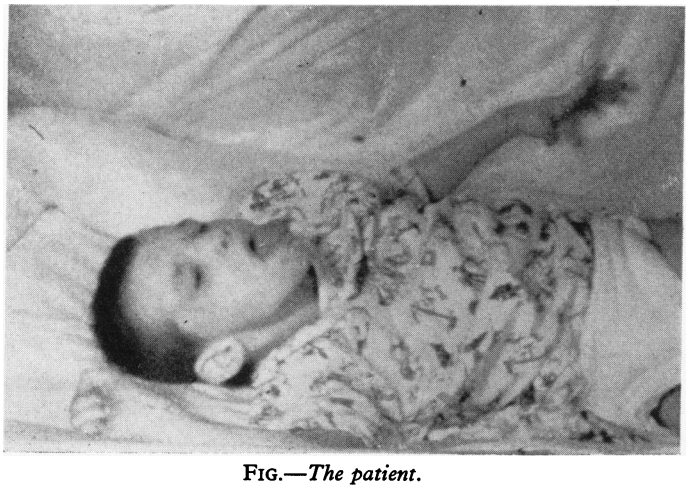

FIG.—*The patient.*

and inversion of the feet. Consciousness was maintained and no incontinence occurred during the attacks. During the post-ictal period he cried and was moderately irritable, but not sleepy. Reflexes were quite brisk at this time.

The attacks failed to respond to either oral or parenterally administered diphenylhydantoin or phenobarbitone, but seemed to lessen in frequency and intensity following administration of tincture of belladonna. Intravenous administration of 1·0 mg. benztropine mesylate aborted the fully-developed attack within a few seconds. The drug was continued in oral maintenance dose, and the attacks stopped. Attempts to induce them were unsuccessful. He appeared more alert, became able to use a wheelchair, and with instructions began to use additional words and short sentences. Although athetoid finger movements continued, they were less severe, as were the nystagmoid eye movements.

Because of an episode of vomiting, the drug was withdrawn. Eight hours later he had another dyskinetic episode which promptly responded to 1·0 mg. intravenous benztropine mesylate. A saline placebo was ineffectual. This was followed by a generalized tonic-clonic seizure with unconsciousness and incontinence, controlled by intravenous phenobarbitone.

While at a rehabilitation centre he continued to make improvement in speech, began to walk short distances without aid, and became toilet trained. However, he exhibited aggressive behaviour toward other children. At that time he was having only an occasional dyskinetic attack.

When seen in a seizure clinic six months after the initial period in hospital, he was unchanged and taking 4·0 mg. benztropine and 200 mg. diphenylhydantoin per day. A subsequent evaluation six months later disclosed only mild rigidity of the trunk, with a tendency toward gait festination. At that time the mother had discontinued all drugs and he had not had an attack for several months.

Laboratory examinations revealed a fasting blood sugar of 89 mg./100 ml.; normal serum Na, K, Cl, CO_2, Ca, P, urea; caeruloplasmin 606 units; serum Cu 104 mg./100 ml.; uric acid 3·3 mg./100 ml.; normal liver function studies; cerebrospinal fluid—total protein 27 mg./100 ml., glucose 92 mg./100 ml. without cells. Skull *x*-ray examination and a pneumoencephalogram were normal. Numerous electroencephalograms revealed a diffuse, non-specific slow wave abnormality in both theta and delta range. No seizure activity was seen. A recording made immediately after an attack of dyskinesia was identical to the interictal recordings.

Discussion

Although these episodic dyskinesias have a similar clinical pattern, it seems certain that they, like cortical epilepsy, are symptomatic of diverse aetiology and not a single nosological entity.

In addition to preservation of consciousness, they can be distinguished from cortical epilepsy by the gross nature of the movements and in some instances by the response to anticholinergic drugs. The latter seems similar to the effect of these compounds in extrapyramidal dyskinesias commonly seen with some of the phenothiazine and Rauwolfia derivatives.

Summary

A case of paroxysmal dyskinesia, responsive to benztropine and belladonna, is presented. The similarity of previously reported cases is reviewed.

A trial of anticholinergic drugs, particularly benztropine, seems warranted in these cases. Further studies to elucidate the neurophysiological and neuropharmacological relationships in this and similar cases are intended.

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