Kinesigenic Choreoathetosis due to Brain Injury

J.C. Richardson*, J.L. Howes, M.J. Celinski and R.G. Allman

ABSTRACT: This uncommon paroxysmal movement disorder featuring attacks of dystonic spasm with athetoid posturing is usually of idiopathic nature, often familial and starting in childhood. There are a few reported examples due to cerebral pathology, and only four previous cases due to brain trauma. We report here a 21 year old man with left-sided motor spasms, choreoathetoid in type, which were clearly caused by a right frontal penetrating injury with contusion and haemorrhage. The attacks were relieved by phenytoin therapy. In this case, it seems that the post-traumatic paroxysmal kinesigenic choreoathetosis may be a form of reflex epilepsy.

RÉSUMÉ: Choréo-athétose kinésigénique attribuable à un traumatisme cérébral. La choréo-athétose kinésigénique est un désordre du mouvement de nature paroxystique qu'on rencontre rarement. Elle se manifeste par des accès de spasmes dystoniques accompagnés de postures dystoniques, habituellement d'origine idiopathique, souvent familiale, débutant dans l'enfance. Quelques cas attribuables à une lésion cérébrale ont été rapportés dans la littérature, mais seulement quatre cas attribuables à un traumatisme crânien ont été rapportés antérieurement. Nous rapportons ici le cas d'un jeune homme de 21 ans présentant des spasmes moteurs de type choréo-athétoïde à l'hémi-corps gauche, nettement attribuables à une lésion pénétrante avec contusion et hémorragie à la région frontale droite. Ces accès étaient soulagés par le phénitoïne. Chez ce patient, il semble que la choréo-athétose kinésigénique paroxystique post-traumatique puisse être une forme d'épilepsie réflexe.

Can. J. Neurol. Sci. 1987; 14:626-628

The term kinesigenic choreoathetosis refers to a rare condition characterized by brief choreoathetoid spasms precipitated by sudden onset of walking or other movements of the body.[1] The condition has also been referred to as "paroxysmal choreoathetosis." Opinion has been divided as to whether it represents an episodic extrapyramidal disorder or a form of reflex epilepsy.

In his book on epilepsy, Gowers[2] described a few patients with seizures induced by movement. However only one patient, a 13 year old girl, presented the features of the entity recognized more recently as paroxysmal kinesigenic choreoathetosis (PKC). In 1962, Lishman et al.[3] described seven patients with dystonic spasms associated with voluntary movement. They favored a reflex epileptic mechanism and used the term "seizures induced by movement." Subsequent reports by Plant,[4] Kinast et al.,[5] and Mayeux and Fahn[6] discuss familial and sporadic idiopathic examples with a constant pattern of childhood onset and frequent brief spasms. The movements tended to be complex, presumably extrapyramidal in origin, choreoathetoid in nature, and were either unilateral or bilateral. Control was good with phenytoin or carbamazepine with few exceptions. Przuntek and Monninger[7] emphasize the difference between the Mount and Reback type of familial paroxysmal choreoathetosis and PKC.

Most of the published sporadic and familial examples of PKC are of the idiopathic type with no demonstrable underlying systemic or cerebral pathology. However, there are a few reports of associated endocrine, metabolic or cerebral disorders including hyperthyroidism, hyperparathyroidism, hypoglycaemia, and multiple sclerosis. Watson and Scott[8] reported one PKC patient with brain stem and cerebellar atrophy demonstrated by CT scan, but it is not possible to conclude that pathology in those regions was responsible for the clinical symptoms. Gilroy[9] described a 39 year old man with typical clinical features of PKC in whom the CT scan and pneumoencephalogram suggested a right frontotemporal cerebral lesion. Although brain biopsy failed to identify the pathology, it seems likely that a focal lesion was responsible for the left sided motor spasms.

There are two previous reports of PKC secondary to brain trauma. Robin[10] described a 33 year old man whose left sided attacks commenced eight months after a moderately severe

From the Downsview Rehabilitation Centre, Toronto
Received May 21, 1987. Accepted in final form August 1, 1987
* Deceased June 15, 1986
Reprint requests to: Dr. J.L. Howes, Workers' Compensation Board, 2 Bloor Street East, Toronto, Ontario, Canada M4W 3C3

blunt head injury with post-traumatic amnesia of 18 hours. It is possible that focal traumatic cerebral pathology may have caused the paroxysmal choreoathetosis despite a negative CT scan and normal EEG. Drake et al.[11] described three cases of paroxysmal choreoathetosis following closed head injury. These patients responded well to anticonvulsants and there was no evidence of injury to the basal ganglia. They concluded that the distinction between partial seizures and involuntary movement disorders may not be clear. In the present paper, we describe a young man who suffered left sided dystonic spasms induced by sudden movement following severe unilateral frontal lobe contusion.

Case Report

The patient is a 21 year old man who was a grade 13 student and seasonal worker at the time of his head injury at age 18. He was examined by the authors at the Downsview Rehabilitation Centre on two occasions: 3 years and 3 ½ years after the injury. Detailed reports were available from his earlier attending surgeons, physicians, and psychologists.

In August, 1981, the patient suffered a severe head injury in a motor vehicle accident with retrograde amnesia of several weeks and post-traumatic amnesia of five weeks. He was comatose for 11 days. There was a right frontal depressed skull fracture and underlying frontal lobe contusion with a small hematoma. There was also a chest injury and subsequent emphysema which complicated and prolonged his hospital treatment. Based on available reports, the patient was considered to have made an excellent recovery by May, 1982.

The Attacks

In early reports it was noted that the patient was having episodic sensations of warmth and tingling over his left side but no objective neurological abnormalities were described. An electroencephalogram in 1982 showed a focal right temporal slow and sharp wave abnormality. He was given phenytoin which was later changed to carbamazepine.

By June, 1984 when we saw him at the Rehabilitation Centre he had recovered to a state in which he was able to give a clear detailed history. He now reported that since the injury he had been experiencing involuntary left sided movements at the onset of whole body movement. Such forced movements occurred several times a day with no preceding aura. He was able to describe and imitate them in clear detail. Passive movements would not provoke these attacks. The usual precipitating movement was quick standing and walking, particularly if there was an element of surprise or urgency of effort. The left sided movement began approximately 15-20 seconds after the commencement of activity. For example, if he was sitting in a chair and suddenly stood up and walked to answer the telephone, he would have a feeling of pulling and stiffness over the entire left side of his body. Then the face, tongue, arm, and leg would start to move. His face and tongue would twist to the left. His left arm would stiffen in a semi-flexed posture and abduct from his side with some firm rigid posturing and twisting movements of the hand. His left lower limb would become rigidly extended and pushed outward so that he would walk with a gross limp dragging forward the extended foot. The spasms and movements would subside completely after two to three minutes. On no occasion did he lose consciousness or fall. The patient's mother confirmed the above described features of the attacks.

After starting on regular medication with phenytoin and later carbamazepine, the attacks subsided almost completely. When he was reassessed in 1985 the patient recalled only four mild attacks over the preceding 18 months.

To evaluate the anticonvulsant effect, phenytoin was discontinued and four days later one of his typical attacks occurred with onset of movement. One of the authors witnessed an attack which occurred when the patient stood up quickly from a relaxed position. It consisted of flexion of the left hand and arm, and stiffening of the left leg. It lasted approximately 10 seconds and was followed by bilateral trembling of the upper limbs. Altogether he had five attacks in a period of two weeks while off medication. The phenytoin was then resumed with almost complete control of attacks.

Neuropsychological and Neurological Assessments

On examination in July, 1984 and April, 1985 he was observed to be alert, rational and somewhat euphoric. He tended to be emotionally labile and mentally fatiguable. Neuropsychological tests indicated that he was functioning within the low average range of intellectual and memory ability as assessed by the Wechsler Adult Intelligence Scale - Revised and Wechsler Memory Scale, respectively. He displayed motor slowness, concentration difficulties, verbal memory problems, and mild word finding difficulties. None of the neuropsychological tests suggested lateralized motor or sensory function deficits. His performance continued to indicate mild diffuse intellectual impairment and post-traumatic personality change (impulsivity, euphoria, social withdrawal).

An enhanced CT brain scan revealed cerebral atrophy and a hypodense area in the right frontal lobe consistent with contusion, old hematoma, and subsequent gliosis (Figure 1). An electroencephalogram in July, 1984 showed some diffuse slow wave activity and a focus of larger amplitude slow waves as well as sharp waves in the right temporal region.

Neurological examination revealed normal motor function, reflexes, and sensation. The cranial nerves were normal except for a crossed diplopia on full gaze to the left. Alternating movements of the hands did not produce a noticeable attack, but the patient reported a subjective sensation that his tongue was being pulled to the left side and his left hand was pounding.

Discussion

This young man suffered left sided motor attacks precipitated by sudden movement. The attacks were of a choreoathetoid type typical of kinesigenic choreoathetosis and were associated with right frontal cerebral contusion, hemorrhage, and gliosis. The CT scan suggested major involvement of frontal white matter but the traumatic pathology could have involved other structures such as the caudate, putamen and thalamus.

A review of the literature provides three possible explanations. First, a non-epileptogenic mechanism based on the presence of structural damage to the basal ganglia and a disturbance of cortico-subcortical feedback mechanisms is proposed.[12] An epileptogenic mechanism is suggested by a favourable response to anticonvulsant medication in some cases. Further support for this view comes from observations that photic stimulation at low frequencies performed close to the onset of the attack

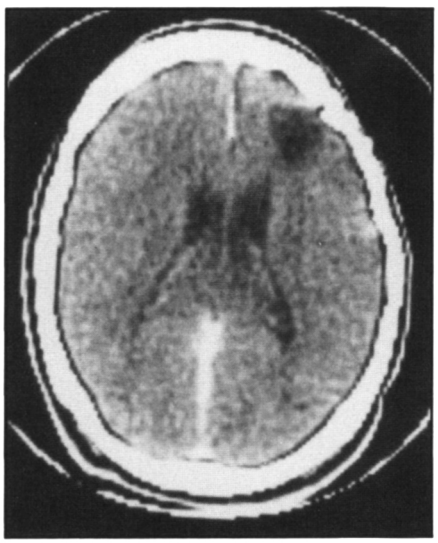

Figure 1 — CT Head Scan showing large right mid frontal low density area.

https://doi.org/10.1017/S0317167100037501 Published online by Cambridge University Press

induces paroxysmal lateralized discharges from the hemisphere contralateral to the movements.[13] However, arguments based on electrophysiological findings are not entirely convincing since EEG abnormalities are not consistently present in cases of PKC.[11] A third possibility, which does not necessarily imply an epileptic disturbance, is that PKC is associated with hypoxic brain injury. Rosen[12] reported a case of PKC associated with perinatal hypoxic encephalopathy, with EEG changes over the entire left hemisphere. The patient responded to belladonna and diphenhydramine.

In our case the positive response to anticonvulsant medication suggests the predominant mechanism for the attacks may have been a form of reflex epilepsy. The lesion on the right frontal lobe may have resulted in a disturbance of feedback mechanisms associated with cortico-subcortical connections.

References

1. Kertesz A. Paroxysmal kinesigenic choreoathetosis, Neurology 1967; 17: 680-690.
2. Gowers WR. Epilepsy and Other Convulsive Diseases Vol. 2, 1901: Philadelphia; Blakiston Co.
3. Lishman WA, Symonds CP, Whitty CWM, Willison RG. Seizures induced by movement. Brain 1962; 85: 93-109.
4. Plant G. Focal paroxysmal kinesigenic choreoathetosis. J Neurol Neurosurg Psychiatry 1983; 46: 345-348.
5. Kinast M, Erenberg G, Rothner A. Paroxysmal choreoathetosis: report of five cases and review of the literature. Pediatrics 1980; 65: 74-77.
6. Mayeux R, Fahn S. Paroxysmal dystonic choreoathetosis in a patient with familial ataxia. Neurology 1982; 32: 1184-1185.
7. Przuntek H, Monninger P. Therapeutic aspects of kinesigenic paroxysmal choreoathetosis and familial paroxysmal choreoathetosis of the Mount and Reback type. J Neurol 1983; 230: 163-169.
8. Watson R, Scott W. Paroxysmal kinesigenic choreoathetosis and brain stem atrophy. Arch Neurol 1979; 36: 522.
9. Gilroy J. Abnormal computed tomograms in paroxysmal kinesigenic choreoathetosis. Arch Neurol 1982; 39: 779-780.
10. Robin JJ. Paroxysmal choreoathetosis following head injury. Ann Neurol 1977; 2: 447-448.
11. Drake ME, Jackson RD, Miller C. Paroxysmal choreoathetosis after head injury. J Neurol Neurosurg Psychiatry 1986; 49: 837-838.
12. Rosen JA. Paroxysmal choreoathetosis associated with perinatal hypoxic encephalopathy. Arch. Neurol 1964; 11: 385-387.
13. Jacome DE, Risko M. Photic induced-driven PLEDs in paroxysmal dystonic choreoathetosis. Clinical Electroencephalography 1984; 15: 151-154.