Increased ictal perfusion of the thalamus in paroxysmal kinesigenic dyskinesia

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Abstract
The ictal and interictal cerebral blood flow (CBF) were evaluated in a patient with right unilateral short lasting paroxysmal kinesigenic dyskinesia, by means of single photon emission computed tomography (SPECT). The patient was a 6 year old boy with no family history. During an attack, increased CBF was seen in the left thalamus. Subtraction of interictal CBF from ictal CBF disclosed a prominent increase in CBF in the left posterolateral part of the thalamus. This finding suggests that abnormal hyperactivity of thalamic neurons could be responsible for the pathophysiology of paroxysmal kinesigenic dyskinesia in this patient.

Keywords: paroxysmal kinesigenic dyskinesia; SPECT; thalamus

Paroxysmal kinesigenic dyskinesia is a disease characterised by episodes of dyskinesia triggered by quick voluntary movement. The dyskinetic episodes consist of any combination of sudden dystonic posturing, choreoathetosis, and ballismus.1 Paroxysmal kinesigenic dyskinesia is associated with neither EEG abnormalities during or after attacks nor impairment of consciousness.2 The pathophysiological basis and lesion in this disease remain unclear.

To obtain information on these aspects, ictal and interictal cerebral blood flow (CBF) were studied by single photon emission computed tomography (SPECT), in a patient with paroxysmal kinesigenic dyskinesia.

Case report
The patient was a 6 year old boy. His grandfather had had Parkinson’s disease from 70 years of age. The prenatal and perinatal histories were normal, but he only started to utter phrases at the age of 3 years. His mother noticed that he sometimes grimaced from the age of 8 months. From the age of 1 year, he experienced paroxysmal dyskinesic attacks on the right side of his body. The attacks occurred on initiation of rapid movement, usually in the morning. During the attacks, there was no loss of consciousness. The episodes lasted less than 1 minute and occurred up to 50 times a day. The result of neurological, general physical, and laboratory examinations, and radiological studies were normal in the interictal period. He was diagnosed as having short lasting, sporadic paroxysmal kinesigenic dyskinesia, and thus was treated with carbamazepine, phenobarbital, valproate, phenytoin, or haloperidol, without improvement of the episodic dyskinesia. He was referred to our hospital for treatment at the age of 6. He looked restless and his speech was slightly unclear. He had mild mental retardation. In the ictal period, he exhibited paroxysmal choreoathetotic movements involving the right side of his face and right limbs, and a dystonic posture involving the right side of the trunk and right limbs. The attacks were induced by sudden voluntary movement such as starting to speak, walk, or run. There was no loss of consciousness. The attacks occurred 10 to 30 times a day and lasted for less than 1 minute. General and neurological examinations disclosed no remarkable findings. In the interictal period, there was no laterality or abnormality of his movement, posture, or muscle tone. Neither pyramidal signs nor cerebellar signs were found. The results of laboratory examinations, cerebral MRI, and interictal EEG were normal. Ictal EEG showed no paroxysmal discharges.

Measurement of cerebral blood flow (CBF) by SPECT was performed twice (in the ictal and interictal periods). The SPECT measurements were made with a rotating gamma camera equipped with a high-resolution collimator (Siemens, MULTISPECT 3). Before the first SPECT, all medications (phenytoin and haloperidol) were tapered off. The attacks were easily provoked by sudden voluntary movement. Just after an attack started, $^{123}$I-IMP was injected. Twenty minutes later, sleep was induced with a pentobarbiturate and SPECT was carried out. After the first SPECT, he was treated again with carbamazepine. The frequency of attacks was remarkably reduced. Carbamazepine was effective. Seven months later, the second SPECT was performed; $^{123}$I-IMP was injected while he was awake and voluntarily moving both his arms. Twenty minutes later, sleep was again induced with a pentobarbiturate. No attacks occurred during the study. To allow accurate CBF subtraction, we performed MRI to match the level of the section on SPECT. Brain MRI was performed with a 3-D turbo FLASH (Siemens Magnetom Impact Expert).

Results
Ictal SPECT showed increased CBF in the left thalamus. Interictal SPECT showed increased
CBF in the left medial thalamus. Subtraction of interictal CBF from ictal CBF showed a prominent increase of CBF in the left posterolateral part of thalamus (fig 1).

Discussion
The pathophysiological and anatomical bases of paroxysmal kinesigenic dyskinesia remain uncertain. There is discussion about the pathophysiological basis concerning reflex epilepsy versus dysfunction of basal ganglia. Although the surface EEG recording did not show any epileptic activity, we cannot completely rule out the possibility that the dyskinetic phenomenon in this patient was epileptic. Recent studies have shown that the thalamus may play an important part in the initiation or propagation of seizures in several types of epileptic disorders.3–5 Some investigators reported that ictal SPECT showed increased perfusion in the thalamus ipsilateral to the cortical focus in patients with partial seizures.3–5 Our patient does not correspond to these because no cortical hyperperfusion or electroencephalographical abnormality were seen.

There have been several reports that paroxysmal kinesigenic dyskinesia is associated with specific lesions of the putamen,7 right frontotemporal region,9 globus pallidus,9 dorsal medulla oblongata,10 cervical spinal cord,11 or thalamus.12–13 Sunohara et al reported a necropsied case in which the patient had shown exercise induced dystonia of the left limbs.13 They found necrotic lesions in the posterolateral ventral part of the right thalamus and a part of the right internal capsule. Camac et al described a patient who developed paroxysmal kinesigenic dystonic choreoathetosis after a thalamic infarct.12 Their patient exhibited a large area of increased signal intensity in the right thalamus, including the ventral posterolateral, lateral posterior, and ventral lateral nuclei on T2 weighted MRI. Although these reports suggest that a dysfunction of the posterolateral part of the thalamus may play an important part in paroxysmal kinesigenic dyskinesia, they did not indicate whether or not there was in vivo abnormal neuronal activity in this region during the attacks. Our patient showed no remarkable lesion on MRI, but he showed a prominent increase in CBF in the left posterolateral part of the thalamus during an attack while on SPECT. From this finding, we conclude that abnormal hyperactivity of thalamic neurons contributed to the pathophysiology of paroxysmal kinesigenic dyskinesia.
dyskinesia in this patient. There have been a few reports of evaluation of the lesions responsible for paroxysmal kinesigenic dyskinesia by means of SPECT. Humano et al described a 15-year-old boy with a large porencephalic cyst in the left parietotemporo-occipital region who had right sided dystonic spasms induced by running.16 A cerebral blood flow study involving SPECT showed hyperfusion of the lenticular nucleus involving the putamen and regions corresponding to the atrophy and the porencephalic cyst, but it was an interictal study only. Hayashi et al reported the results of measurement of postictal regional cerebral blood flow (rCBF) in three patients with paroxysmal kinesigenic choreoathetosis.17 Two of the three patients, who had unilateral attacks, showed a noticeable increase in rCBF in the basal ganglia on the contralateral side to the attacks. The difference in rCBF between the right and left basal ganglia in these two patients was over the mean value plus 2SD for seven normal subjects, and decreased with phenytoin treatment. From these results, they concluded that the neural activity of the basal ganglia is raised during the attacks in paroxysmal kinesigenic dyskinesia; but their SPECT data were for a postictal state and they did not mention which part of the basal ganglia was responsible for the attacks. Kuki et al described two patients with exercise induced paroxysmal kinesigenic dyskinesia. Our findings also support this suggestion.

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