SHORT REPORT

Increased ictal perfusion of the thalamus in paroxysmal kinesigenic dyskinesia

S Shirane, M Sasaki, D Kogure, H Matsuda, T Hashimoto

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.

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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. Paroxysmal kinesigenic dyskinesia is associated with neither EEG abnormalities during or after attacks nor impairment of consciousness. 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 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 father had had Parkinson's disease from 70 years. He was a 6 year old boy. His grandparents and parents 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. Carbamazepine was effective. Seven months later, the second SPECT was performed; 123I-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. Some investigators reported that ictal SPECT showed increased perfusion in the thalamus ipsilateral to the cortical focus in patients with partial seizures. 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, right frontotemporal region, globus pallidus, dorsal medulla oblongata, cervical spinal cord, or thalamus. Sunohara et al reported a necropsied case in which the patient had shown exercise induced dystonia of the left limbs. 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. 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
substantial new information on the neural responsible for our finding. Recent method-
we conclude that voluntary movement is not
side to the attacks. The di
unilateral attacks, showed a noticeable increase
mal kinesigenic dyskinesia; but their SPECT
for seven normal subjects, and decreased with
them, the ictal perfusion of the basal ganglia was
hypoactive cortex areas
also decreased. The hypoactive cortex areas
in the insular cortex, primary motor area,
supplementary motor area, medial primary sen-
sorimotor area, striatum, cerebellar vermis,
and visual cortex. Our patient had not moved his
upper limbs voluntarily during the loading of
13-T-IMP in the examination. From these facts,
we conclude that voluntary movement is not
responsible for our finding. Recent method-
ological advances in neuroscience have provided
substantial new information on the neural
circuit of the basal ganglia.25–27 Chorea and
ballism appear as a consequence of functional
inactivation or a lesion of the subthalamic
nucleus leading to reduced neuronal activity in
both the globus pallidum pars externa and pars
internara.28,29 Decreased CBF has been reviewed as a consequence of
the release of premotor cortical activity from
thalamic control, which is supported by recent
positron emission tomographic studies.29 In
the present case, as judged from the results of
SPECT, neuronal activity in the posteriotal
part of the left thalamus showed primary or sec-
ondary increases during the attacks. If the inhi-
bition of the globus pallidum pars interna/
substantia nigra pars reticulata is insufficient,
when these thalamic neurons activate the corti-

cal motor area in the frontal lobes, paroxysmal
dyskinesia could occur.

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