Paroxysmal Kinesigenic Choreoathetosis Associated with Frontotemporal Arachnoid Cyst
—Case Report—

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Abstract
A 17-year-old male presented with paroxysmal kinesigenic choreoathetosis (PKC) associated with frontotemporal arachnoid cyst. Xenon-133 single photon emission computed tomography detected a slight but equivocal decrease in regional cerebral blood flow in the vicinity of basal ganglia associated with the PKC episodes. PKC continued after surgical removal of the cyst but was well controlled by oral administration of carbamazepine. Whether the pathogenesis of symptomatic PKC was associated with the cortical lesion could not be determined in the present case.

Key words: paroxysmal kinesigenic choreoathetosis, arachnoid cyst

Introduction
Paroxysmal kinesigenic choreoathetosis (PKC) is a benign disease, characterized by attacks of uni- or bilateral choreoathetosis that usually last up to a minute and are precipitated by sudden or fast movements. PKC has been variously called reflex epilepsy, movement-induced seizures, subcortical epilepsy, reflex tonic epilepsy, kinesthetic reflex epilepsy, and painful tonic spasm. Whether PKC is a form of epilepsy remains controversial, but PKC was recently classified as a type of paroxysmal dystonia/choreoathetosis under the category of paroxysmal dyskinesia. PKC is clinically distinct from non-kinesigenic paroxysmal dystonic choreoathetosis (PDC) and intermediate forms, hypnogenic paroxysmal dystonia (HPD), and benign paroxysmal dystonia/torticollis in infancy. PKC may be caused by cortical or subcortical dysfunctions but this remains unclear. Most patients with PKC have no underlying or associated disease, but symptomatic patients have shown various lesions in the brainstem, basal ganglia, and cerebral cortex, as well as diffuse lesions in the central nervous system. Symptomatic PKC is sometimes associated with multiple sclerosis or head trauma, but the exact site of the cause of PKC is often difficult to determine in cases associated with multiple or diffuse brain lesions. Detailed analysis of cases associated with clearly localized lesions is required to clarify the pathophysiological mechanism of this disease. We describe a rare case of PKC associated with a frontotemporal arachnoid cyst.

Case Report
A 17-year-old male had a 6-year history of episodes characterized by sudden tonic contraction of the muscles of the left side of the face, left arm, and left leg that occurred five to six times a day after vigorous activity such as running or sudden rising from a chair. The contractions lasted about 10–30 seconds and were not associated with consciousness disturbance, premonitory symptoms, or sequelae. During the height of the episodes, the left side of his face was contorted in a grimace. His left arm was flexed at the elbow and his left leg at the knee, and there were occasional choreoathetotic movements of the left side of the face, left arm, and left leg. He complained of headaches which gradually increased from the age of 14 years. His birth had been uneven but there was a past history of convulsive seizures from the age of 3 to 10 months which then spontaneously disappeared. No particular medical
problems were found in his family history.

Neurological examination on admission to our hospital showed a low intelligence quotient, 66 on the Wechsler Intelligence Scale for Children-Revised, and heavy head sensation in the right temporoparietal region. Skull radiography detected thinning of the right frontotemporal bone. Computed tomography (CT) and magnetic resonance imaging showed an arachnoid cyst lateral to the right of the sylvian fissure, which had mildly compressed the right frontotemporal lobes (Fig. 1). CT cisternography showed delayed filling and delayed clearance of the cyst. Repeated electroencephalography (EEG) examinations showed occasional spike waves in the right hemisphere only during the early sleep stage, but no focal abnormality. Video-EEG monitoring detected the onset of episodes when he began to pedal a bicycle ergometer, but there was no epileptic discharge before or after these episodes. The diagnosis was PKC, and no medication including phenytoin or carbamazepine was given before.

### Table 1

<table>
<thead>
<tr>
<th></th>
<th>Cerebral hemisphere</th>
<th>Basal ganglia (perforator area)</th>
<th>ACA area</th>
<th>Anterior part of MCA area</th>
<th>Posterior part of MCA area</th>
<th>PCA area</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-episode</td>
<td>49 ± 14</td>
<td>54 ± 12</td>
<td>51 ± 10</td>
<td>51 ± 16</td>
<td>46 ± 10</td>
<td>39 ± 13</td>
</tr>
<tr>
<td>Post-episode</td>
<td>44 ± 13</td>
<td>50 ± 9</td>
<td>53 ± 12</td>
<td>50 ± 9</td>
<td>45 ± 10</td>
<td>40 ± 12</td>
</tr>
</tbody>
</table>

Values are in ml/100 g/min. ACA: anterior cerebral artery, MCA: middle cerebral artery, PCA: posterior cerebral artery.
the surgery.

Xenon-133 enhanced single photon emission computed tomography (SPECT) showed a slight decrease in regional cerebral blood flow (rCBF) in the right hemisphere in the resting state (Fig. 2 left, Table 1). Serial SPECT study 20 minutes later of the induced episodes during pedaling of the bicycle ergometer showed a slight or equivocal decrease in the rCBF of the right basal ganglia and adjacent area (Fig. 2 right, Table 1). A slight increase in the rCBF of the anterior cerebral artery territory was also noted. The latter observation was interpreted as activation of the cortical foot area by the pedaling movement.

The arachnoid cyst was removed by right frontotemporoparietal craniotomy to treat his headaches. An intraoperative cortical EEG showed no epileptic discharge. Subsequent to surgery, his headaches disappeared, and the arachnoid cyst did not reappear. However, the episodes remained unchanged, but became well controlled after he began taking 200 mg of carbamazepine per day.

**Discussion**

Patients with symptomatic PKC have shown neuroimaging evidence of various lesions: Multiple sclerosis plaque in the cerebral peduncle, thalamus, basal ganglia, and paraventricular; traumatic injury of the frontal white matter, caudate, putamen, and thalamus; small cerebral infarction in the putamen and thalamus; and cerebral arteriovenous malformation (AVM) in the basal ganglia. Such findings strongly suggest that subcortical structures, especially those in the vicinity of the basal ganglia, are the sites responsible for PKC. Symptomatic PKC has also been described in cases of hypoxic encephalopathy, encephalitis, hemiatrophy syndrome, diffuse brainstem atrophy, and associated with metabolic disorders such as hypoparathyroidism without or with calcifications of the bilateral basal ganglia and cerebellar hemispheres, hyperglycemia with AVM in the basal ganglia, and choreoathetosis disease are rare. PET using [18F]deoxyglucose (FDG) detected hypometabolic foci of the frontal or frontotemporal areas in four patients with HPD. HPD is a focal epilepsy involving the internal frontal regions, rather than a pathology of movement. Another patient had painful bilateral tonic spasms as well as perfusion defects in the left frontoparietal and right parietooccipital regions based on technetium-99m d,l-hexamethyl-propyleneamine oxime SPECT findings. A patient with PKC and PDC caused by hypoparathyroidism also had associated calcification of the basal ganglia, thalamus, and the dentate nucleus of the cerebellum. PET study with FDG showed decreased glucose metabolism in the basal ganglia and thalamus. The first four of these cases are disorders categorized differently to PKC, although they may be related to PKC by some common pathomechanism. All three reported SPECT and PET studies were done in the resting stage and in the absence of episodes. In contrast, the SPECT study was done just after the PKC episodes in our patient. The decrease in the rCBF in the vicinity of the basal ganglia after the PKC episodes may be equivocal because the change was very small. Nevertheless, this finding may be correlated to the PKC episodes because the dynamic change in the rCBF is expected to be small as the duration of the PKC episode is very short. Our study produced no evidence as to how to change the rCBF during a PKC episode.

The present case suggests two possible causes for...
PKC. The arachnoid cyst may only be incidental with no direct relationship to the PKC symptoms, and so a transient functional disorder caused by changes in metabolism or blood flow in the basal ganglia is responsible for PKC. The supratentorial arachnoid cyst may affect the ipsilateral corticosubcortical connections because it is located on the side contralateral to the PKC episodes in the extremities. The latter possibility agrees with the hypothesis that any disturbance in the cortical control of the neostriatum and its thalamic connection can induce PKC. Further study is required to clarify whether PKC can be caused by a solitary cortical lesion and whether the symptoms will disappear when the lesion is treated.

References

1) Barbas G, Tucker SM: Idiopathic hypoparathyrodi
dism and paroxysmal dystonic choreoathetosis. Ann
Neurol 24: 585, 1988
2) Berger JR, Sheremata WA, Eldad M: Paroxysmal
dystonia as the initial manifestation of multiple sclero
3) Bortolotti P, Schoenhuber R: Paroxysmal kinesigenic
4) Buchman AS, Goetz CG, Klawans HL: Hemiparkin
sonism with hemiatrophy. Neurology 38: 527-530,
1988
5) Burguera JA, Catald I, Casanova B: Thalamic demye
lination and paroxysmal dystonia in multiple scleros
6) Busard HLSM, Renier WO, Gabreels FJM, Vos AJM,
genic dystonic choreoathetosis associated with a thalamic infarct. Mov Disord 5: 235-238, 1990
8) Drake ME Jr, Jackson RD, Miller CA: Paroxysmal
colloid after head injury. J Neurol Neurosurg Psychi
atry 49: 837-838, 1986
9) Falconer MA, Driver MV, Serafetinides EA: Seizures
induced by movement: report of a case relieved by
operation. J Neurol Neurosurg Psychiatry 26: 300-
307, 1963
10) Fishbeck KH, Layzer RB: Paroxysmal choreoatheto
sis associated with thyrotoxicosis. Ann Neurol 6:
453-454, 1979
tonic spasms: an interesting phenomenon in cerebral
12) George MS, Pickett JB, Kohli H, Allison MA,
Pritchard P: Paroxysmal dystonic reflex choreoatheto
sis after minor closed head injury. Lancet 336:
1134-1135, 1990
13) Gilroy J: Abnormal computed tomograms in parox
ysmal kinesigenic choreoathetosis. Arch Neurol 39:
779-780, 1982
14) Kato H, Kobayashi K, Kohari S, Okita N, lijima K:
Paroxysmal kinesigenic choreoathetosis and paroxys
mal dystonic choreoathetosis in a patient with familial idiopathic hypoparathyroidism. Tohoku J
An entity within the paroxysmal choreoathetosis syn
drome. Description of 10 cases, including 1 aut
16) Kinast M, Erenberg G, Rothner D: Paroxysmal
choreoathetosis: report of five cases and review of the
17) Lance JW: Familial paroxysmal dystonic choreoatheto
sis and its differentiation from related syndromes.
Ann Neurol 2: 285-293, 1977
18) Matthews WB: Tonic seizures in disseminated sclero
19) Merchant MP, Brumlil J: Painful tonic spasms caused
by putaminal infarction. Stroke 17: 1319-1321, 1986
20) Mushet GR, Mosylate B: Paroxysmal dyskinesia. A
case responsive to benztpipine mesylate. Arch Dis
Child 42: 654-656, 1967
21) Ohya T: [Paroxysmal choreoathetosis]. Shinkei
Shinp 39: 471-478, 1975 [jpn, with Eng abstract]
22) Richardson JC, Howes JL, Celinski MJ, Allman RG:
Kinesigenic choreoathetosis due to brain injury. Can
J Neurol Sci 14: 626-628, 1987
23) Robin JI: Paroxysmal choreoathetosis following head
mal kinesigenic choreoathetosis as presenting
symptom of multiple sclerosis. J Neurol Neurosurg
25) Rosen JA: Paroxysmal choreoathetosis associated with
perinatal hypoxic encephalopathy. Arch Neurol 11:
385-387, 1974
26) Sellal F, Hirsch E, Maquet P, Salmon E, Franck G,
Collard M, Kurtz D, Marescaux C: Postures et mou
vements anormaux paroxytiques au cours du som
meill: dystonie paroxystique hypnogénique ou épilepsie partielle? Rev Neurol (Paris) 147: 121-128,
1991
27) Sethi KD, Hess D, Huffnagle VH, Adams RJ:
Acetazolamide treatment of paroxysmal dystonia in
central demyelinating disease. Neurology 42: 919-
921, 1992
28) Shintani S, Shirozawa Z, Tsunoda S: [A simultaneous
appearance of paroxysmal kinesigenic choreoatheto
sis (PKC) and paroxysmal dystonic choreoathetosis
(PDC) in a case with arterio-venous malformation of
parietal lobes]. Rinsho Shinkei 26: 1182-1189, 1986
(jpn, with Eng abstract)
29) Soffer D, Licht A, Yaar I, Abramsky O: Paroxysmal
choreoathetosis as a presenting symptom in idiopathic hypoparathyroidism. J Neurol Neurosurg
Psychiatry 40: 692-694, 1977
30) Stevens N: Paroxysmal choreoathetosis: a form of

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