

Case report

Moyamoya disease presenting with paroxysmal exercise-induced dyskinesia

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Received 13 February 2006; received in revised form 23 June 2006; accepted 1 July 2006

Abstract

We report a patient with moyamoya disease presenting with paroxysmal exercise-induced dyskinesia (PED). A 31-year-old lathe man developed recurrent attacks of paroxysmal hemichorea. The attacks always affected his left limbs and occurred either after several hours of working or while playing football. The duration of attacks ranged from 30 min to 4 h. Attacks were not provoked by sudden movements, consumption of coffee or alcohol, hyperventilation, emotional stress, exposure to cold or passive movement. An MRI of the brain showed no parenchymal lesions. However, ^{99m}Tc-ethylcysteine dimer SPECT study showed hypoperfusion in the right striatum. Digital subtraction angiography showed stenosis of the right internal carotid and middle cerebral artery with prominent basal collaterals, which was compatible with moyamoya disease. Imaging studies of the cerebral arteries should be done in patients with clinical features of PED in order to detect possible cases of moyamoya disease.

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Keywords: Moyamoya disease; Paroxysmal dyskinesia; Chorea

1. Introduction

Paroxysmal dyskinesias can be classified as kinesigenic dyskinesia (PKD), non-kinesigenic dyskinesia (PND), exercise-induced dyskinesia (PED) or hypnogenic dyskinesia (PHD), according to the provoking factors, duration of the paroxysms and frequency of the attacks. PED refers to rare form of paroxysmal dyskinesias that are provoked primarily by prolonged exercise (e.g., walking or running) [1].

Patients with cerebrovascular disease can present with PKD, PND, PHD, or mixed phenomenology [2,3]. However, patients having characteristic clinical features

of PED in association with cerebrovascular disease have not been reported.

Moyamoya disease is characterized by chronic, progressive obstruction of the cerebral arteries constituting the circle of Willis and the numerous collateral vessels. There are previous reports of six patients with moyamoya disease whose initial manifestations were paroxysmal dyskinesia [4]. Three of the six patients had precipitating factors, including hyperventilation, writing, walking or swimming. Only one patient showed paroxysmal chorea both after prolonged exercise and even during rest, a presentation that is atypical for PED [4].

We report a novel patient with moyamoya disease presenting with recurrent attacks of hemichorea which always occurred after a prolonged walk and affected the limbs contralateral to the side of severe vascular stenosis.

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2. Case report

A 31-year-old right-handed latheman suddenly developed heaviness in his left shoulder. A few minutes later, he noticed a forceful dorsiflexion of his left ankle and subsequent choreic movements in his left limbs lasting for about 30 min. The attack was confined to his left limbs, so that he could continue to work with his right limbs. He had been working with two automatic lathes, and had to walk around them continuously.

Approximately 20 similar attacks had occurred over the previous 10 months. The attacks lasted from 30 min to 4 h, and the frequency progressively increased, reaching 1 every 10 days. Most attacks occurred while the patient was working extra hours. Occasionally, they occurred while he was playing football or walking a long distance. Attacks were not provoked by sudden movements, consumption of coffee or alcohol, hyperventilation, emotional stress, exposure to cold or passive movements.

The patient's birth and developmental histories were normal. His family history was unremarkable, except for the fact that his father had recurrent intracerebral hemorrhages and died at the age of 55. Brain CT studies of his father showed multiple sites of intracerebral hemorrhage and previous cerebral infarction in the temporal lobe. Neither side of the middle cerebral artery (MCA) was visualized after contrast enhancement.

Neurological examinations performed during the interictal period showed that the patient's motor, sensory and cerebellar function were all normal. The deep tendon reflexes were symmetric and normoactive. Routine laboratory tests, including blood cell count, serum electrolytes, chest X-ray, ECG and urinalysis, were all normal. Serum antistreptolysin O titers and ceruloplasmin levels were within normal ranges. ANA, anti-DNA, anti-Ro and anti-La antibodies were negative. Antiphospholipid and anticardiolipin antibodies were also negative. Proteins C and S and antithrombin III levels were all within the normal range, and thyroid

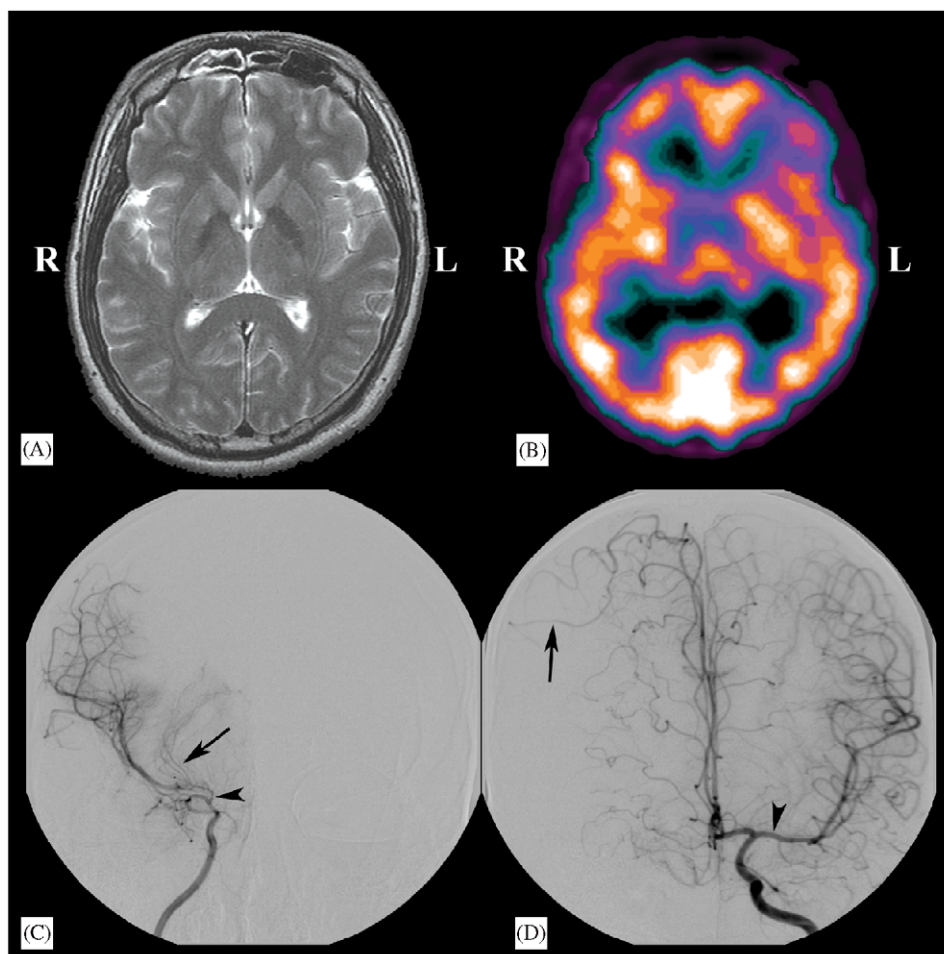


Fig. 1. (A) A T2-weighted brain MRI study shows no parenchymal lesion. (B) A ^{99m}Tc -ethylcysteine dimer brain SPECT study shows hypoperfusion in the right striatum. (C) Digital subtraction angiography shows stenosis (arrow head) in the distal part of the right internal carotid artery (ICA) and the proximal part of the right middle cerebral artery with prominent basal collaterals (arrow). (D) The left ICA angiogram shows diffuse stenosis in the M1 segment (diameter: 2 mm; arrow head) with pial collaterals (arrow) to the right hemisphere.

function tests and parathyroid hormone levels were normal. Routine cerebrospinal fluid examinations were normal, and there was no oligoclonal band. A scanning electron microscopic examination of peripheral blood smears did not show acanthocytes, and an electroencephalography study showed no abnormalities. Brain MRI studies, including T1, T2, diffusion-weighted and fluid attenuated inversion recovery (FLAIR) images, showed no abnormal parenchymal lesions (Fig. 1(A)).

A ^{99m}Tc -ethylcysteine dimer brain SPECT study showed hypoperfusion in the right striatum (Fig. 1(B)), and MR angiography showed severe stenosis of the right internal carotid artery (ICA) at the supraclinoid portion and the proximal MCA. Digital subtraction angiography confirmed MR angiography findings and showed prominent basal collaterals (Fig. 1(C)). The left ICA angiogram showed mild stenosis at the M1 segment (diameter: 2 mm) with pial collaterals to the right hemisphere (Fig. 1(D)). No transdural collaterals were found. The bilateral common carotid, subclavian and renal arteries were normal.

The patient refused to undergo the surgical treatment. Same paroxysmal dyskinesia occurred in the same situations during the several following months.

3. Discussion

PED is a rare form of paroxysmal dyskinesia. The most common phenomenology of PED is dystonia [5]. However, chorea also has been reported [6]. Attacks usually start from the leg and spread to the arm and face. They may occur unilaterally or bilaterally, and, in some patients, the same side of the body is always affected [7,8]. Attacks usually last for about 10–15 min, but may persist for up to 48 h [5,9].

In our patient, the attacks always occurred after several hours of walking and did not involve the factors known to provoke PKD or PND. Furthermore, the paroxysmal nature of the attacks, the lack of deficits on neurological examination, and the normal brain MRI made us suspect idiopathic PED as the most likely clinical diagnosis.

In moyamoya disease, hypoperfusion of the cerebral cortex [10] or of the basal ganglia [11] has been proposed as a possible mechanism for hemichorea. In our patient, blood perfusion of the striatum contralateral to the side affected by paroxysmal hemichorea was more severely decreased than that of the ipsilateral side. This finding suggests the striatal hypoperfusion is closely related to the development of chorea in this patient.

The term “unilateral moyamoya disease” has been applied to about 12.6% of patients with moyamoya disease [12]. However, careful examination may show mild stenosis of arteries on the contralateral side. In our patient, there was no focal stenosis in the left MCA, but there was a diffuse narrowing of the left proximal MCA. In epidemiological studies, up to 10% of patients with moyamoya disease have a positive family history [13]. This raises the possibility that the present patient had familial moyamoya disease.

None of the previously reported patients with PED had parenchymal lesions on brain imaging studies. However, the findings of cerebral angiography and brain perfusion study were not reported. We believe that moyamoya disease should be added to the list of possible causes of symptomatic PED.

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