

**CASE REPORT**

Paroxysmal Kinesigenic Dyskinesia as the Presenting and Only Manifestation of Multiple Sclerosis after Eighteen Months of Follow-Up

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Other than tremor, movement disorders are uncommon in multiple sclerosis. Among these uncommon clinical manifestations, paroxysmal kinesigenic dyskinesia is the most frequently reported. It is characterized by episodic attacks of involuntary movements that are induced by repetitive or sudden movements, startling noise or hyperventilation. The diagnosis is essentially clinical and based on a good observation of the attacks. It is very easy to misdiagnose it. We describe the case of a young female patient who presented paroxysmal kinesigenic dyskinesia as the first and only clinical manifestation of multiple sclerosis, with no recurrence of attacks nor any other neurologic symptom after eighteen months of follow-up.

Key Words Movement disorders; paroxysmal kinesigenic dyskinesia; multiple sclerosis; MRI lesions.

Paroxysmal dyskinesias (PxD) are a rare heterogeneous group of neurological conditions that are characterized by intermittent and involuntary movements manifested by dystonia, chorea, athetosis, ballism, or any combination of these hyperkinetic disorders.^{1,2} There are four types of PxD according to the classification of Demirkiran and Jankovic:³ paroxysmal kinesigenic dyskinesia (PKD) is induced by sudden movement, paroxysmal non-kinesigenic dyskinesia occurs spontaneously, paroxysmal exertion-induced dyskinesia appears after prolonged exercise, and paroxysmal hypnogenic dyskinesia occurs during sleep.^{3,4} Most cases of PxD are primary and are categorized as either familial or idiopathic.¹ However, some symptomatic forms, secondary to a specific cause, have been reported.⁵ PKD is the most common type of PxD.⁶ We describe the case of a young female patient who was admitted for diagnostic work-up: epileptic seizures versus psychogenic movement disorders. We established a diagnosis of multiple sclerosis (MS) with PKD as the first and only clinical manifestation. After eighteen months

of follow-up with beta-1a interferon (Avonex[®], Biogen Idec Limited, Berkshire, United Kingdom) therapy, there was no recurrence of attacks or any other neurologic symptom.

CASE REPORT

A 22-year-old female patient with a medical history of infectious mononucleosis at age 16 and without any personal or familial history of neurological diseases was admitted with a new-onset movement disorder. She presented with brief attacks of involuntary, sudden and painful hyperkinetic movements of the left limbs (hyperextension and external rotation of the lower limb and extension-pronation of the upper limb) and a left rotation of the head. She had twenty to thirty attacks daily for two days, lasting twenty to forty-five seconds. There was neither loss of consciousness nor postictal confusion or amnesia. The attacks occurred during position changes or sudden movements or even in cases of emotional stress. She was able to predict the

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occurrence of the attacks and even trigger them with specific movements of her left leg. Her relatives and her general practitioner thought it might be psychogenic movement disorders or epileptic seizures. The neurological examination was unremarkable between the attacks.

Blood tests were normal. The electroencephalogram (EEG) showed neither asymmetric nor epileptiform activities. Her brain magnetic resonance imagery (MRI) revealed more than twenty hyperintense lesions in the supratentorial white matter in T2 and fluid attenuated inversion recovery sequences (Figure 1A, B, and D). The largest (14 mm) was located in the left semi-oval center. Others were located in the right parietal, left cingular and left frontal areas. One left frontal lesion was enhanced after contrast (Gadovist®, Bayer Pharma AG, Leverkusen, Germany) injection (Figure 1E). Others were within and at the border of the corpus callosum. There were also periventricular lesions with radial disposition (Figure 1A). The spinal cord MRI showed several T2 lesions in the medulla oblongata and the cervical and thoracic spinal cords (Figure 1C and 1F), with a discrete enhancement of a lesion facing the third and fourth cervical vertebrae.

The 2010 McDonald diagnostic criteria of MS were fulfilled, so we did not perform a cerebrospinal fluid analysis. We prescribed intravenous methylprednisolone (1 g per day during five days) and clonazepam (0.3 mg three times a day during 3 days) as symptomatic treatment for the attacks. Twenty-four hours after starting these medications, there was a complete cessation of the abnormal movements.

Two weeks later, the patient began treatment with beta-1a interferon. Eight months later, the control MRI of the brain and spinal cord showed neither additional nor enhancing lesions but did show a reduc-

tion in the size of the lesion in the medulla oblongata (Figure 1F). After eighteen months of follow-up, the patient did not present a recurrence of attacks, and the neurologic examination remained unremarkable.

DISCUSSION

Other than tremor, movement disorders are uncommon in MS.⁷⁻⁹ Their prevalence is less than 2%.⁷ Among these uncommon clinical manifestations of MS, PKD is the most frequently reported. PKD is characterized by episodic attacks of involuntary movements that are induced by repetitive or sudden movements, startling noises or hyperventilation.²⁻⁶ There can be up to a hundred attacks daily, and each attack can last a few seconds to less than five minutes. They are usually unilateral⁷⁻⁹ but may alternate or even be bilateral.^{4,5} They are very often painful.^{7,8} Many patients experience aura-like symptoms manifested as tingling or parasthesia in the affected limb.^{4,5,8,9}

The diagnosis of PKD is essentially clinical and is based on a good observation of the attacks. It is very easy to misdiagnose it, particularly when it occurs in the absence of other neurological symptoms. Patients are often suspected of having epileptic seizures or psychogenic disorders,⁴ similar to our patient. Blood tests are often normal. EEG or video-EEG monitoring are almost always normal even during attacks, excluding epileptic seizures.^{2,3,6}

Neuroimaging is useful for differential diagnosis. It is normal in primary cases of PKD, but the results are variable in symptomatic forms. PKD has been described in few MS patients. Various lesion sites have been reported, including cervical spinal cord, brainstem, cerebellar or cerebral peduncles, cerebellum, subthalamic nucleus, internal capsule, thal-

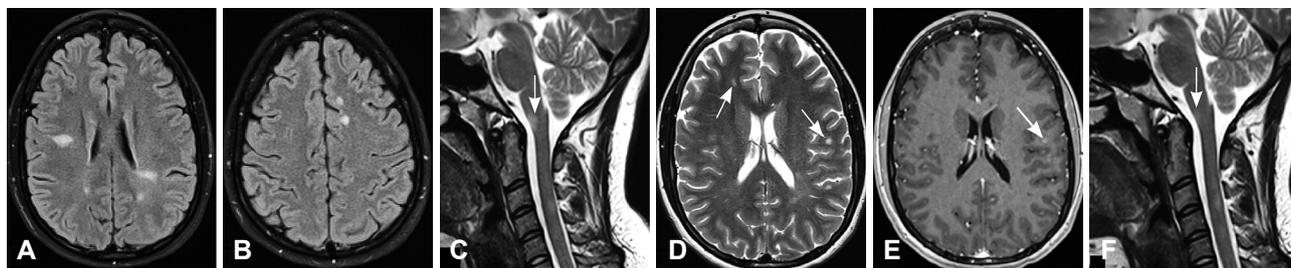


Figure 1. Axial FLAIR slices show several hyperintense subcortical lesions. A and B: Notice the ovoid form of some of them and the perpendicular disposition of periventricular ones. C: T2-weighted sagittal spinal MR shows one large brainstem (arrow) and several cervical spinal cord hyperintense lesions. D: T2-weighted axial brain MR image shows sub-cortical lesions (arrows). E: T1-weighted image after gadolinium injection shows an enhancement of one left frontal lesion (arrow). F: T2-weighted sagittal spinal MR control 8 months later showing a reduction of the brainstem lesion (arrow). FLAIR: fluid attenuated inversion recovery.

amus, and basal ganglia.^{7,8} Due to this wide dispersion of lesions in the central nervous system, the anatomical correlation between the location of plaques and the PKD remains unclear. It is uncertain whether the lesions seen on MRI are always responsible for the attacks.⁷⁻⁹ The pathophysiology of PKD in MS is still poorly understood. The most widely accepted hypothesis is that PKD is a result of the transversely spreading ephaptic activation of axons due to a demyelinating lesion at any level of the motor pathways.⁷⁻¹⁰

The unique feature in our patient was the uncommon locations of some of the plaques. To our knowledge, this is the first time that the corpus callosum or cingular sub-cortical demyelinating lesions have been described in PKD secondary to MS. Furthermore, PKD related to parietal subcortical lesions has been reported only in one case of MS and in another case with arteriovenous malformation.⁹

Although our patient had several brain and spinal cord lesions, the only clinical manifestation was PKD, without any history of previous neurological symptoms. PKD usually appears after the diagnosis of MS has been made, particularly during paroxysms.^{8,9} Even when it is reported as the presenting symptom, it is often accompanied by other neurological issues. Without the MRI, we could not have established a diagnosis of MS by solely considering the clinical presentation.

In conclusion, PKD is very uncommon as the presenting clinical manifestation of MS. Clinicians should be aware of this possibility. Otherwise, PKD can easily be misdiagnosed, leading to the use of an unnecessary, inappropriate or even dangerous medication. In the presence of paroxysmal movement dis-

orders, particularly in young individuals, neuroimaging and electrophysiological tests should be realized before establishing the diagnosis of epileptic or psychogenic disorders.

Conflicts of Interest

The authors have no financial conflicts of interest.

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