

Paroxysmal pruritus as the first relapsing symptom of neuromyelitis optica

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Abstract

Itching is a rare symptom in neurological disease. Paroxysmal symptom is uncommon in neuromyelitis optica (NMO) or multiple sclerosis. We describe a 45-year-old woman who presented with paroxysmal pruritus in association with relapse of NMO on two separate occasions. Cervical MRI showed a lesion at a level corresponding to the dermatomal site of the itching. Paroxysmal pruritus could be a predictive symptom of relapse in NMO unless there are other comorbid causes such as systemic diseases, allergy, drug reaction, or emotional stress responsible for the itching.

INTRODUCTION

Paroxysmal symptoms in neuromyelitis optica (NMO) or multiple sclerosis (MS) are defined as those that last for seconds or minutes. This is in contrast with exacerbations where the symptoms exist for at least 1 or 2 days. Paroxysmal itching in MS patients has been rarely reported,¹⁻⁴ but it was not described in NMO. Here, we describe a NMO patient with paroxysmal pruritus that was a predictive symptom of relapse. The dermatomal location of itching was well-correlated with the spinal cord lesion on MRI.

CASE REPORT

A 45-year-old woman was referred due to paroxysmal itching. Two months previously, she developed acute paraparesis, followed by a tingling sensation below the lower trunk. Deep tendon reflexes were brisk in the lower limb and the right plantar response was extensor. Lhermitte's sign was negative. Spinal cord MRI revealed diffuse swelling and high signal intensities with inhomogenous gadolinium enhancement in T4-T9 segment cord on T2 weighted images (Figure 1A). Brain MRI showed high signal intensities in the right temporal periventricular white matter and left posterior medulla on fluid-attenuated inversion recovery and T2-weighted images. CSF IgG index was 1.03 and oligoclonal band was negative. Visual evoked potential study showed no abnormalities. The paraparesis was much improved after methylprednisolone infusion and she could walk without aid.

Paroxysmal itching occurred in the C3 segmental areas at the back of the neck. The attacks started and ended abruptly. They lasted two to five minutes. She had 10 to 20 attacks a day. Paroxysmal itching developed spontaneously, but it was usually provoked by tactile stimulation or movement. The itching sensation was so intense that she could not resist the urge to scratch although this gave no relief. Three days after the paroxysmal itching, she developed quadriparesis. Superficial and deep sensations were moderately decreased below the C4 segment. Spinal cord MRI revealed high signal intensities in the cord of C2-C5 and C7-T5 segment on T2 weighted images (Figure 1B). She was given gabapentin 1,800 mg a day. The itching resolved over 2 weeks. Six weeks after the first relapse, she complained of the same paroxysmal itching on the left arm. Ten days later, the weakness in her left upper and lower limbs worsened and she became completely quadriplegic. Cervical cord MRI revealed strong gadolinium enhancement in the previous cervical cord lesions. The itching had resolved over a month. After 6 months, she developed right optic neuropathy.

DISCUSSION

Itching is a rare symptom in neurological disease. An itch can be cutaneous, neuropathic, neurogenic, mixed or psychogenic. A neuropathic itch can originate at any point along the afferent pathway as a result of damage to the nervous system. Localized pruritus has been reported with peripheral nerve

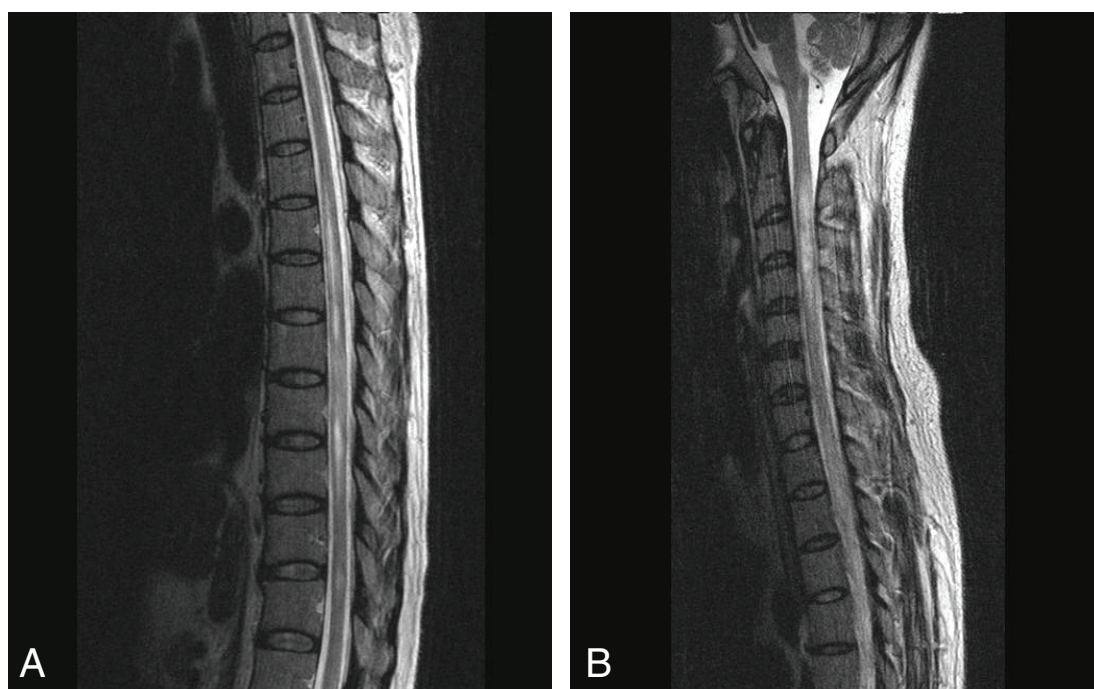


Figure 1. Spinal cord imaging of first attack and relapse. (A) T2-weighted spinal cord MRI revealed diffuse swelling and high signal intensities in T4-T9 segment cord. (B) Spinal cord MRI showed high signal intensities in the cord of C2-C5 segment on T2 weighted images at the first relapse.

lesions in post-herpetic neuralgia, sensory nerve entrapment syndrome, and spinal cord tumor. Paroxysmal itching in MS was first reported in 1 patient out of 22 patients with paroxysmal symptoms from 235 MS patients.¹ In other series of 377 MS patients, 17 had experienced itching, and 1 as an isolated initial symptom.³ There has been one case of paroxysmal pruritus in MS that showed a cervical cord lesion at the level corresponding to the dermatomal location by MRI.⁴ However, the paroxysmal itching as the first or predictive symptom of relapse in MS or NMO was not described.

The characteristics of paroxysmal itching in MS are as follows.^{2,4} The itching starts and ends abruptly. It is intense and the duration ranges from several seconds to several minutes. The frequency is variable, but the attacks may occur up to 20 times a day. The distribution involves any region of the body. Superficial sensory disturbances are present in the affected area, which may persist after the itching has disappeared. The itching usually starts spontaneously, but it may also be provoked by movement.

The pathophysiology of paroxysmal itching is poorly understood, but it was proposed that the paroxysmal itching is caused by transversely spreading ephaptic activation of axons lying in a partially demyelinated lesion in fiber tracts in

the CNS.¹ This hypothesis is compatible with the observation that many instances of paroxysmal itching seem to be provoked by sensory stimuli or movement. It was also suggested that itching is a subthreshold pain sensation. The sensations produced by stimuli inducing intense afferent C fiber activity were reported as burning or delayed pain, whereas stimuli eliciting low frequency activity often were reported as itching.⁵ These observations suggest that pain and itching sensations travel along the same conducting fibers, the frequency of the axonal activity being different. However, Tuckett contested the idea that the same sensory neurons transmit both pain and pruritus impulses, and provided evidence to suggest that they may be carried along different populations of primary sensory neurons.⁶ Considering the transitory nature of the pruritus, whatever the cause, paroxysmal itching seems to result from a demyelinating lesion not sufficient to result in permanent deficit, but rendering the surrounding axons hypersensitive to minor irritation.

It is likely that the frequency of paroxysmal itching in MS or NMO has been underestimated. When a NMO patient complains of itching, which is localized and paroxysmal, we might consider it as one of several symptoms or a specific predictive symptom of relapse unless there are other comorbid causes such as systemic diseases, allergy,

drug reaction, or emotional stress responsible for the itching.

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