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Case report

Restless legs may be associated with the post-polio syndrome

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Abstract

Restless legs syndrome (RLS) has been described in association with a number of conditions including iron deficiency, neuropathy and Parkinson's disease. Here we report a patient who developed RLS concurrent with the development of classic post-polio syndrome (PPS), 40 years after recovery from an episode of paralytic poliomyelitis. PPS is still frequently encountered in neurological practice, and clinicians should be aware of the possibility of associated RLS.

Keywords: Restless legs syndrome; Post-polio syndrome; Poliomyelitis; Secondary

1. Introduction

Restless legs syndrome (RLS) is a disorder characterized by an unpleasant feeling in the legs that usually occurs in the evening or at night, and is relieved by movement of the legs. RLS is typically idiopathic with an autosomal dominant pattern of inheritance, but may also be symptomatic. Secondary causes include uraemia, iron deficiency, diabetes, Parkinson's disease, peripheral neuropathies, radiculopathies, myelopathies and pregnancy [1]. However, RLS in association with post-polio syndrome (PPS) has not been described.

PPS is a condition that occurs for many years following a previous episode of poliomyelitis. The syndrome encompasses a range of neurological, musculoskeletal and systemic symptoms and signs including new limb weakness or increased limb fatigability, muscle atrophy, muscle and joint pain. Electromyography (EMG) will show denervation in the affected limb [2].

We report the case of a woman who developed classic PPS 40 years after an episode of acute poliomyelitis. With the development of PPS she had the onset of symptoms compatible with RLS, with a partial response to dopaminergic therapy.

2. Case report

This 56-year-old woman was first seen in our clinic at the age of 46. At the age of 4 she had an episode of paralytic poliomyelitis affecting both legs. She recovered completely over 2 years. Thereafter, she had no notable disability.

At the age of 45 she noticed progressive fatigability of strength in her legs. In addition she noticed twitching of the muscles in both legs, typically following exertion. At the same time she reported that when she was relaxing in the evenings, she would develop an unpleasant sensation in both lower legs. These feelings would be abolished by movement of her legs. Occasionally she would be woken by unpleasant feelings in her legs that required her to move them.

Neurological examination at the age of 46 revealed wasting of the left leg. There were fasciculations in both calves. The lower limb reflexes were brisk and plantar reflexes were flexor. Examination of the cranial nerves and sensation was normal.

Routine blood tests, including vitamin B12, folic acid, iron/ferritin serum levels, C reactive protein, ESR, and blood glucose were within normal limits, with the exception of a slight

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elevation in CPK (184, normal range 0–150), positive antinuclear antibodies (1:160), mildly positive smooth muscle antibodies but negative double stranded DNA antibodies.

Sensory and motor nerve conduction studies in both upper and lower limbs and thermal thresholds were within normal limits. EMG showed definite evidence of chronic partial denervation predominantly in the distal muscles of the lower limbs.

Electrophysiologically, the pattern of abnormality was in keeping with a post-polio syndrome. During 10 years of follow-up the electrophysiology was repeated three times with similar results. MRI scans of the lumbar spine showed no nerve root or cord compression.

Sensory symptoms in her legs and the consequent urge to move them followed a waxing and waning course. At the age of 50 these symptoms worsened and could also be present during the day. She initially tried treatment with codeine, aspirin, carbamazepine, clonazepam, buprenorphine and acupuncture with no benefit. Levodopa was temporarily effective but after a year the effect wore off, despite increasing dosage. Cabergoline was then introduced to good effect. She did, however, require increasing dose to maintain effect, and after 3 years was taking 4.5 mg daily. Because of inadequate benefit she switched to ropirinole upto 4 mg in the evening. This resulted in moderate benefit for her symptoms.

3. Discussion

This patient presented with typical symptoms of RLS in the context of a PPS. She met diagnostic criteria for both diagnoses. The RLS criteria are a desire to move the limbs associated with uncomfortable sensations in the legs, motor restlessness to gain relief from these sensations, presence or worsening of the symptoms at rest, and symptoms worsening during the evening/night [1].

Consequent with the onset of RLS she developed new loss of strength, fatigability and aching in her muscles which fulfilled clinical and neurophysiological criteria for PPS [2,3]. She had a risk factor for the development of PPS in that her recovery from the original episode of poliomyelitis was very good. Previously affected muscles are more likely than unaffected muscles to later become weak [3]. Overuse has also been proposed as a contributory factor and elevated CPK has been reported as a marker for overuse in post-polio patients [4]. Our patient led a very athletic life, and had a slightly elevated CPK level. Rarely upper motor signs can occur in PPS [3] and this can explain the presence in our patient of brisk lower limb reflexes.

There is a possibility that RLS in our patient is only a coincidental finding, but the late age at onset, the absence of family history and the incomplete response to dopaminergic therapy tend to suggest a secondary cause. None of the common secondary causes was found in our patient, and the symptoms of RLS developed at the same time as the development of the PPS. Nevertheless, we cannot exclude that the two conditions occurred together by chance.

The pathophysiology of RLS is still speculative. The success of dopaminergic therapy and the results of some imaging studies suggest that the dopaminergic system is important in the pathophysiology of RLS [5]. There is speculation that abnormal afferent input can induce dysfunction in the dopaminergic system (particularly caudal to the pons), thereby causing RLS in some patients with neuropathy [6]. Spinal cord lesions could cause RLS by abnormal sensorimotor integration at the spinal interneuronal level (perhaps mediated by dopamine) [6,7], the loss of supraspinal inhibitory influences [8], or both.

Despite the eradication of polio in developed countries, there are many patients still alive who had polio in the past. PPS is a relatively common occurrence in these patients (upto 78% of individuals with previous poliomyelitis) [4]. Therefore clinicians should be aware of the possibility that such patients may develop RLS and may respond to dopaminergic therapy.

References

[1] Allen RP, Picchietti D, Hening WA, Trenkwalder C, Walters AS, Montplaisir J. Restless legs syndrome: diagnostic criteria, special considerations, and epidemiology. A report from the restless legs syndrome diagnosis and epidemiology workshop at the National institutes of health. Sleep Med 2003;4:101–19.

[2] March of Dimes. Post-polio syndrome: identifying best practices in diagnosis and care. White Plains, NY: March of Dimes Birth Defects Foundation, www.modimes.org; 2001.

[3] Jubelt B, Cashman NR. Neurological manifestations of the post-polio syndrome. Crit Rev Neurobiol 1987;3:199–220.

[4] Trojan DA, Cashman NR. Post-poliomyelitis syndrome. Muscle Nerve 2005;31:6–19.

[5] Allen R. Dopamine and iron in the pathophysiology of restless legs syndrome. Sleep Med 2004;5:385–91.

[6] Ondo W, Jankovic J. Restless legs syndrome: clinicoetiologic correlates. Neurology 1996;47:1435–41.

[7] Lindvall O, Bjorklund A, Akagerberg G. Dopamine-containing neurons in the spinal cord: anatomy and some functional aspects. Ann Neurol 1983;14:255–60.

[8] Hemmer B, Riemann D, Glocker FX, Lucking CH, Deuschl G. Restless legs syndrome after a borrelia-induced myelitis. Mov Disord 1995;10:521–6.