Case report

Painful leg and moving toes syndrome in secondary tethered cord syndrome

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Context: Painful leg and moving toes (PLMT) syndrome is a rare movement disorder where the patient has pain followed by movement disorder in one or both lower limbs. The exact etiology and pathogenesis is uncertain, however many cases have been related to lesions in peripheral nerve, spinal cord or radicals. Appearance of abnormal movement in PLMT soon after surgery has not, to our knowledge, been described.

Findings: We report a 40-year-old female patient who had secondary tethered cord syndrome. She had pain in left lower limb for 5 months prior to surgery. Immediately after surgery, she noticed abnormal movement in left foot. Surface electromyography suggested single motor unit discharges.

Conclusion/clinical relevance: In persons with unclear symptoms of painful leg and moving toes, PLMT syndrome should be in the differential diagnosis.

Keywords: Spinal cord injury, Tethered cord syndrome, Electromyography

Introduction

Painful leg and moving toes (PLMT) is a clinical syndrome first described in 1971.¹ The disease is characterized by pain and abnormal repetitive non-rhythmic movements of digits in lower limb.² Symptoms may be unilateral or bilateral from the onset; however in some patients they may start in one limb and involve both limbs later on. Painless variant of PLMT has also been described in literature.³ A similar type of syndrome involving fingers of upper extremities known as painful arm and moving fingers has also been described.⁴ Pathogenesis of PLMT is not clear and genesis of movement is uncertain whether they are peripheral or central origin.^{4,5} Peripheral nerve injuries leading to change in pattern of neural activation in dorsal horn, dorsal column nuclei, ventral thalamus and somatosensory cortex may generate abnormal movement.⁶ Published literature have discussed different types of lesions involving spinal cord, peripheral nerve and radicular syndrome in the pathogenesis of PLMT.³

We present a 40-year-old person who developed abnormal movement of her left foot within an hour after surgery for secondary tethered cord syndrome due to intra-spinal lipoma.

Case presentation

This 40-year-old patient had a diagnosed case of lumbosacral mass since childhood. She complained of pain and progressive thinning of the left leg for 5 months. The pain was characterized as dull in nature and did not increase with movement, coughing or sneezing. She had mild thinning of left leg, which was restricted up to the knee region only (27 cm on the left side versus 33 cm on right side measured 10 cm below tibial tuberosity). There was no history of limb weakness or bladder-bowel disturbances. On examination, she had normal motor power in all four limbs. All deep tendon reflexes were normal. She had normal sensation and bilateral plantar reflexes were flexor. On magnetic resonance imaging (MRI) of lumbosacral spine there was a low lying spinal cord up to L5 with lumbosacral lipoma with extension into intraspinal region consistent with the secondary tethered cord syndrome (Figs. 1A–D). She underwent S1 laminectomy, along with excision of lumbosacral lipoma and intraspinal lipoma. After surgery the patient and her family members noticed abnormal movement in the left foot, which was mainly a slow flexion/extension type of movement involving great toe and occasionally other toes also (see supplementary video: <http://dx.doi.org/10.1179/2045772315Y.0000000001>). On clinical examination there was no limb weakness and all modalities of

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Figure 1 Magnetic resonance imaging of spine showing low lying spinal cord with lumbosacral lipoma, which is hyper-intense on T1 W sagittal (A) and T2 W sagittal & coronal (B & D) images and hypo-intense on short tau inversion recovery (STIR) (C) sequences, confirmed by histopathology (E).

sensations were normal. Histo-pathological investigation of excised lumbosacral mass revealed normal mature adipocytes suggestive of lipoma (Fig. 1E). She was diagnosed with a case of PLMT syndrome. Surface electromyography was done, which was suggestive of single motor unit discharges (Fig. 2). She was treated with Gabapentin (600 mg twice daily) and Clonazepam (0.25 mg twice daily), which resulted in significant improvement in pain and mild decrease in toe movement at the end of 4 months follow-up.

Figure 2 Surface electromyography recorded from left great toe showing irregular single motor unit discharges.

Discussion

Our patient presented with pain followed by abnormal movement in left leg immediately after surgery, which is consistent with a diagnosis of PLMT syndrome. Pain in PLMT may be dull aching, sharp shooting, burning, crawling or tearing type and more distressing than the movement.² Occasionally, patients may only complain about sensory symptoms associated with pain such as tingling, numbness, pins needles or tight band-like sensations. Our patient complained of dull aching pain in the left lower limb associated with mild thinning of the leg. Her pain was likely related to the secondary tethered cord syndrome, which is defined as a stretched-induced (due to tissue attachment) functional disorder of the lumbosacral spinal cord.

Movements in PLMT may be flexion/extension, abduction/adduction, dystonic, twitching, fanning, clawing, waving, piano type or direction changing.^{1,2} Movement may be continuous or intermittent and may be voluntarily suppressed in some patients. In our patient, movements were mainly in the left great toe and flexion/extension type. The toe movement in our patient started following the post-operative release of tethered cord. Schott in 1991 discussed the role of trauma in the pathogenesis of PLMT in a published case series.⁸ Six patients (F:M, 4:2) had different types

of trauma (bilateral Hallux deformities in 3 patients, ankle sprain in 1 patient, and traumatic Hallux hematoma in 1 patient). Time interval between trauma and PLMT symptoms were ranging from a few weeks to 2 years. Pain followed by involuntary toe movements was present in all the patients. However, none of the patients had a history of spinal surgery. Dressler *et al.* published a series of 20 cases of PLMT, which developed in the setting of trauma to the spinal cord and cauda equina, lumbar root lesions, bony or soft tissue injuries to the feet and peripheral neuropathy.⁹ No cause could be identified in 4 patients. The interval between the event and PLMT ranged from days to 5 years. Pain preceded the movement in a majority of the patients (18/20). PLMT following discectomy at L5/S1 has also been reported where symptoms started 2 weeks after surgery.³ Several attempts of S1 nerve root block led to disappearance of pain and abnormal movements after 4 months. Spinal lesions (multiple osteopenic fractures) producing painless legs and moving toes have been reported in a patient.² Decompressive laminectomy led to disappearance of movement in this case. On the contrary, toe movement in our patient started immediately after surgery. Single cases temporally linked to epidural injection, myelography, or toe surgery have been reported.²

The surface electromyography recording looked like irregular single motor unit discharges. Due to the long-standing problem, the motor units were likely already enlarged, and post-operatively there appeared to be some increase in excitability leading to sudden onset of toe movement. In PLMT, it is more phasic bursts of activities of varying duration (50 msec to 1 second), but there is no single electrophysiological signature.^{2,10}

In published research, nerve conduction studies were normal in 33 out of 63 cases and there was evidence of peripheral neuropathy in 9 patients and chronic radiculopathy in 14 cases.² In the remaining patients there was evidence for cramps, fasciculation, fibrillation and mono-neuropathy. In our patient nerve conduction study was normal. PLMT is difficult to treat and approximately one-third patients only respond to

treatment.¹ In a study, 83% patients had persisted symptoms during the mean follow-up of 4.6 years. No predictors could be identified for good or poor response in this study. Treatment options include oral medications such as benzodiazepines, blockage of sympathetic nervous system, sympathectomy, nerve root decompression, epidural and nerve root block, and laminectomy and botulinum toxin injection.²

To conclude, PLMT is a rare movement disorder with complex pathophysiology.

Disclaimer statements

Contributors Roles of the authors: SP contributed in manuscript preparation by writing the first draft, review and critique; GS contributed in data collection and manuscript preparation including revision.

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Ethics approval None.

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