Delayed Propriospinal Myoclonus Following Dorsal Spinal Cord Surgery

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Abstract

Cerebral disorders are known to be associated with myoclonus, but spinal pathologies have received little attention as a causative factor in movement disorders. Propriospinal myoclonus (PSM) is a rare hyperkinetic movement disorder caused by activity of a spinal pattern generator localized in a few segments of the spinal cord, spreading to other intraspinal segments via propriospinal pathways. Majority of cases of PSM are reported as functional movement disorders. Structural lesions were found in only a small number of reported cases. We present this rare case report of a patient who developed PSM 2 years following spinal surgery, done 5 years ago for D6–D7 vertebral body collapse. To the best of our knowledge, only few cases of PSM have been reported after spinal surgery and none from India.

Keywords: Functional movement disorder, myoclonus, propriospinal myoclonus, spinal surgery

Introduction

Myoclonus generated within the spinal cord can manifest clinically as focal myoclonus when localized to single spinal segments or as segmental myoclonus when several spinal segments are involved.[1] Propriospinal myoclonus (PSM) is a rare hyperkinetic movement disorder, first described by Brown *et al*. in 1991.[2] PSM presents a more complex, multisegmental activity than segmental myoclonus and is characterized by rhythmic or semi-rhythmic jerking movements, resulting in axial flexion or extension of the trunk. Jerks may be spontaneous or stimulated by acoustic startle, changes in body position, deep breathing, or striking the tendon. It is caused by activity of a spinal pattern generator localized in a few segments of the spinal cord. This spinal generator spreads to other intraspinal segments via propriospinal pathways.[3] The cause of PSM often remains obscure. Etiologically, PSM can be divided into three broad categories: idiopathic, secondary, and functional movement disorder (FMD). Idiopathic category was designated to those patients whose clinical evaluation is not indicative of a functional origin, combined with normal imaging of the spinal axis, consistent electromyography (EMG) pattern, plus an absent Bereitschaftspotential. Majority of the cases (58%) were reported as FMD and 42% as organic. Of the latter, 16% were idiopathic and 26% had a secondary etiology which included ischemic myelopathy, cervical tumors, neuromyelitis optica, and syringomyelia.[4] Few cases of PSM following spinal cord surgery have also been reported.[5]

Case Report

A 28-year old male presented with 10-year history of weakness of the lower trunk and both lower limbs following a road traffic accident. This resulted in inability to walk and get up from supine position by himself. He underwent dorsal spine fixation surgery (D5–D8) with removal of posterior elements of the corresponding vertebrae 5 years ago (details of which were not available). He started having repetitive involuntary movements for the last 3 years characterized by recurrent, rhythmic/semi-rhythmic, painful, jerky movements of the lower trunk and lower limb muscles involving both flexor and extensor groups. These abnormal movements were most prominent in supine posture with hips slightly flexed and relieved partially while sitting with legs hanging down the bed. His sensorium and cranial nerve examinations were normal. Bilateral lower-limb spasticity along with brisk deep tendon reflexes and extensor plantar response were present. The power in both upper limbs was 5/5, while in lower limbs, it was 1/5. Sensory deficit for pain, touch, joint, position, and vibration senses was present below the umbilicus. Local deformity and tenderness were present around D5–D8 spinal processes. Routine biochemical and hematological investigations including viral markers and Veneral Disease Research Laboratory were unremarkable. Electroencephalogram (EEG) was within normal limits.

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Magnetic resonance imaging (MRI) of the dorsal spine was suggestive of metallic implant involving D5–D8 vertebrae and defect in posterior element favoring postoperative status. Gibbus formation was seen at D5–D8 level. Collapse of D6–D7 vertebrae with partial fusion of posterior element of vertebral bodies was noted along with spinal cord atrophy and focal syrinx formation [Supplementary Figures 1 and 2]. Nerve conduction studies revealed nonelicitable motor and sensory responses in both common peroneal and posterior tibial nerves. EMG revealed spontaneous burst of polyphasic motor unit action potentials (MUAPs) synchronous with repeated jerky movements over gluteus medius, gluteus maximus, iliacus, adductors, biceps femoris, vastus lateralis, tibialis anterior, and rectus abdominis muscles. The duration of MUAPs ranged from 100 to 500 ms and amplitude ranged from 200 μV to 1 mV over various muscles [Figures 1 and 2]. The patient was managed conservatively with baclofen, clonazepam, and tetrabenazine, but the response was poor.

Discussion

Myoclonus is defined as a sudden, brief, involuntary movement either with muscle contraction (positive myoclonus) or inhibition of certain muscles (negative myoclonus). PSM is characterized by rhythmic or semi-rhythmic jerking movements resulting in axial flexion or extension of the trunk caused by activity of a spinal pattern generator localized in a few segments of the spinal cord. This spinal generator spreads to other intraspinal segments via propriospinal pathways.[3] In many cases, the myoclonus generator was reported at the thoracic level. Intrinsic spinal cord lesions have been implicated in the partial release of a spinal motor generator by disrupting afferent signals originating distal to propriospinal pathways.[6]

In a review of literature published in 2014,[4] re-evaluation of all the published PSM cases since 1991 was done. Of the 179 cases, 104 (58%) were reported as FMD and 75 (42%) as organic. Of the latter, 29 (16%) were idiopathic and 46 (26%) had a secondary etiology. The reported etiologies of symptomatic PSM varied. A structural lesion in the spinal cord was found in 12 cases (7% of total cases and 26% of secondary cases) including ischemic myelopathy, cervical tumors, neuromyelitis optica, and five cases of syringomyelia.[4] Although some cases of PSM after cervical trauma have been published, the same after thoracic vertebral trauma is rarely reported.[7] MRI spine of this case revealed collapsed D6–D7 vertebrae with compression over the corresponding spinal cord segment and spinal cord atrophy with evidences of previous spinal surgical intervention.

Capelle *et al*.[5] reported a series of six patients who developed movement disorders after spinal disc surgery. The movement disorders became manifested with a delay of 1 day to 12 months after surgery. Of the six patients, four underwent cervical disc surgery and two patients were operated on for lumbar disc herniation; two patients presented with paroxysmal kinesogenic segmental dystonia, one patient with focal dystonia, two with unilateral tremor, and one with bilateral tremor (Capelle *et al*.), but no patient with previous dorsal spinal surgery was reported. They hypothesized that damage to the nerve roots was the most likely cause, as there was also the persistence of pain and sensory symptoms in the corresponding segment in most instances. Time-locked EEG was not suggestive of any abnormal discharge, excluding cerebral origin of myoclonus, although more specific jerk-locked back-averaging analysis was not done.

Most of the authors suggested that the PSM depends on hyperexcitability of a specific myelomere, which, spontaneously or under various stimulations, activates itself and spreads this excitability to the other myelomere, probably through the slow propriospinal polysynaptic pathways.[8,9]

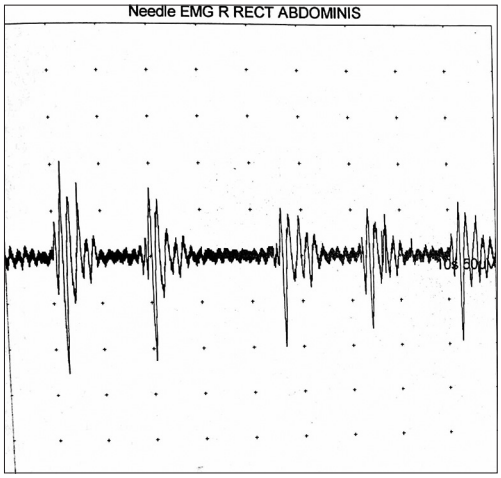

Figure 1: Electromyography of right rectus abdominis muscle suggestive of spontaneous burst of polyphasic motor unit action potentials synchronous with repeated limb movements. The duration of motor unit action potentials ranges from 100 to 500 ms and amplitude being 200 μV–1 mV in various muscles

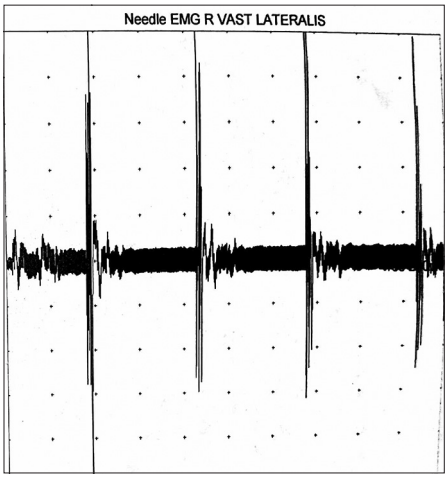

Figure 2: Electromyography of the right vastus lateralis suggestive of spontaneous burst of polyphasic motor unit action potentials synchronous with repeated limb movements. The duration of motor unit action potentials ranges from 100 to 500 ms and amplitude being 200 μV–1 mV in various muscles

EMG is considered essential for the diagnosis of PSM. However, one study found that the typical electrophysiological pattern described for PSM has also been found in patients with psychogenic axial jerks.[10] Regarding typical characteristics for PSM, rhythmicity of the jerks was never reported in FMD cases, but it was very common with secondary PSM. The present case showed EMG burst duration ranging from 100 ms in biceps femoris to 300 ms in gastrocnemius muscle.

Conclusion

We postulate postspinal surgery damage to the nerve roots and hyperexcitability of a dorsal myelomere as the most likely causes of PSM in this patient. Due to the association of movement disorders and spinal disease which is rare and relatively unknown, it may be overlooked easily. This case highlights the rare association of PSM with dorsal spinal cord surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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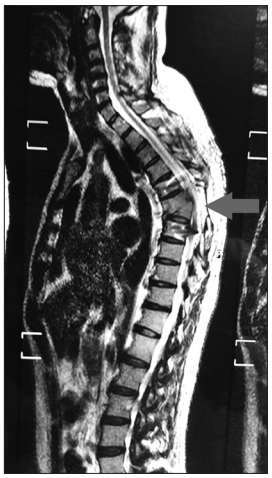
Supplementary Data



Supplementary Figure 1: T2-weighted magnetic resonance imaging sagittal view showing collapse of D6–D7 vertebrae with partial fusion of posterior element of vertebral bodies along with spinal cord atrophy (arrow)

Supplementary Figure 2: T2-weighted magnetic resonance imaging axial view showing collapse of D6–D7 vertebrae with partial fusion of posterior element of vertebral bodies along with spinal cord atrophy (arrow)