Involuntary Jerking of Lower Half of the Body (Spinal Myoclonus)

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Abstract
A 55 years old, hypertensive, diabetic lady presented with sudden onset jerky movement of lower trunk and legs. It was present both in awake and sleep and got aggravated by mental stress as well as sensory stimulation. Examination revealed rhythmic jerks affecting muscles of lower abdomen and legs. The lower limbs had normal muscle bulk and power, increased tone, exaggerated deep tendon reflexes, bilateral flexor plantar response with normal sensory autonomic and cerebellar function. Investigations including CSF study, MRI of dorsal spine and NCV were normal. A combination therapy with tizanidine, baclofen and clonazepam induced gradual improvement within 6 weeks. ©

INTRODUCTION
Myoclonus is best defined as sudden, brief, shock-like involuntary movement due to contraction of a group of muscle fibers, triggered by an event within the central nervous system. Based on its source of origin, it may be classified as cortical, subcortical and spinal. Spinal myoclonus may be segmental or propiospinal. In spinal segmental myoclonus, jerks are confined to the muscles innervated by a few adjacent spinal motor roots, whereas in propiospinal myoclonus extensive jerks are produced by many segments. The prevalence of myoclonus is 8.6 cases per 100,000 per year. Spinal myoclonus is infrequent in clinical practice in contrast to cortical and subcortical one.

While bedside diagnosis of myoclonus is often easy, it can easily be confused with psychogenic movement disorder in inexperienced hands. Often detailed neurophysiological assessment is necessary to establish the nature of the disorder and many of the cases of spinal myoclonus may have a structural lesions which are potentially treatable.

Here we are presenting a case of primary spinal myoclonus because of rarity and potentiality of therapeutic scope.

CASE REPORT
A 55 years hypertensive and diabetic lady presented with sudden onset involuntary movement of both lower limbs and lower abdomen for ten days. There were paroxysmal rhythmic thrusting movements in the pelvis, along with shock like jerks of the legs, which were occurring synchronously in both the legs. She had these involuntary movements at irregular interval both during awake state and sleep, without any history of associated pain in the lower limbs, loss of consciousness or sphincteric disturbances.

On examination, general survey was unremarkable. Higher mental functions and cranial nerve examination were within normal limit. The jerky movements were rhythmic, electric shock like affecting all muscle groups of lower abdomen and both lower limbs. The amplitude and frequency of jerks increased by a variety of stimuli such as mental arithmetic, sudden loud noise, tapping a tendon or standing. At times involuntary jerks were vigorous enough to propel the patient from her bed or wheelchair. The jerks have been video-recorded. Tone was normal in both upper limbs and increased in lower limbs, the nature being rigidity. Muscle power was normal in all four limbs. Deep tendon reflexes were normal in upper limbs and exaggerated in both lower limbs with bilateral flexor plantar response. Examination of sensory, autonomic, and cerebellar system did not reveal any abnormalities.

Investigation showed normal findings in complete haemogram, routine urine examination, serum urea, creatinine, liver function tests, serum Na/K. Immunological tests like ANF and rheumatoid factor were negative. Blood sugar was well controlled. CSF studies showed clear fluid, normal pressure, with a cell count of 02/cmm, sugar-50mg%, protein-54%. CSF was negative for gram stain, AFB, India ink preparation and oligoclonal band. EEG, CT scan of brain, MRI of dorsal spine were normal.

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spine and NCV study were normal. The patient was treated with tizanidine 6mg/day, clonazepam 3mg/day, baclofen 30mg/day in addition to amlodipine for control of blood pressure and insulin for blood sugar. Her jerky movements diminished significantly within next two weeks and it almost disappeared after one month. At 6th month follow up, she can walk independently but hypertonia and exaggerated deep tendon reflexes are persisting till now.

**Discussion**

Segmental myoclonus is a descriptive term that indicates that the distribution of muscle jerking is limited to muscles innervated by one to two contiguous spinal segments. As such it could be used to describe muscle jerks occurring in some cases of epilepsy partialis continua or after lesions of peripheral nerve. To obviate confusion, it is best to use the term ‘spinal segmental myoclonus’. Friedreich in 1881, first suggested that myoclonus could originate in the spinal cord. Lhermitte in 1919 established myoclonus of spinal origin as a clinical entity when he reported a case of traumatic transection of the spinal cord complicated by myoclonus below the level of the lesion. There are several possible causes of spinal myoclonus including AV malformation, intradural tumor, cyst or spondylosis, trauma, multiple sclerosis, amyotrophic lateral sclerosis and viral infection.1 Spinal segmental myoclonus arising from late-delayed sequelae of spinal cord irradiation and following spinal anaesthesia with bupivacaine has recently been reported.2,3 Review of literature has revealed development of atypical propriospinal myoclonus following interferon alpha 2a therapy in a patient of kidney cancer.4 The possible role of interferon was highlighted as focal lesion or paraneoplastic pathology were excluded. Campos et al has reported a case of primary spinal myoclonus the aetiopathology of which is unknown.5 Reflex-sensitive spinal segmental myoclonus associated with vitamin B12 deficiency has recently been described by Tsao JW et al.6 Besides this, a unique form of propriospinal myoclonus has been recognized where enteropathogenic toxin was found to be the possible aetiopathogenic factor.7

Various possible mechanisms have been suggested as the cause of spinal myoclonus. These are loss of inhibitory function of local dorsal horn interneuron, abnormal hyperactivity of local anterior horn cell, aberrant local axons re-excitation and loss of inhibition from suprasegmental descending pathways. Most clinical studies suggest that motor neuronal involvement is rare since weakness and denervation potentials are usually absent.

Histological study of spinal cord in 3 cases of presumed viral infection showed that the disease has caused preferential loss of small to medium size neurons in spinal grey matter.8 Large motor neurons are relatively spared. Disorder of activity in spinal interneuron is associated not only with myoclonus but also with sustained muscle contraction. Spinal interneuronitis is more usually associated with rigidity than myoclonus, although some cases are rigid as well as jerky.

The abnormal movements seen in our patient, as limited to both legs and trunk with preserved sensorium may be due to spinal myoclonus, myoclonus like abnormal movement due to peripheral nerve lesion, epilepsy partialis continua or psychogenic movement disorder. Psychiatric consultation and detailed psychometric evaluation reasonably ruled out the possibility of psychogenic myoclonus. Normal clinical examination and nerve conduction study exclude the possibility of peripheral nerve lesion accounting for myoclonus. Normal EEG and neuroimaging speaks against the possibility of epilepsy partialis continua. Though reflex-sensitive spinal segmental myoclonus associated with vitamin B12 deficiency has recently been reported, we did not consider it in view of normal findings in sensory examination, complete haemogram and NCV study. Thus, the condition strongly favours the diagnosis of spinal segmental myoclonus. The MRI of dorsal spine and normal CSF study exclude the secondary structural causes of myoclonus.

Our patient had a history of undergoing general anaesthesia two weeks prior to the onset of illness but it is unlikely to produce this segmental type of myoclonus. This is because the patient made an uneventful recovery following anaesthesia and was well for subsequent 2 weeks. Post-hypoxic myoclonus following general anaesthesia and surgery usually starts during the period of anaesthesia and manifests as action myoclonus as the patient emerges from is anaesthesia. It is usually multifocal or generalized and there history of postural lapses (negative myoclonus). Majority of patients of posthypoxic myoclonus manifest persistent seizure disorder and they are mostly resistant to treatment.

So the only remaining possibility is viral neuronitis involving spinal interneuron or motor neurons. Our patient had both rigidity and myoclonus favoring the etiology of spinal interneuronitis. It is thought that injury or infection of motor neurons could cause changes in the distribution of ion channels in the membrane leading to repetitive discharge.9 The atypical feature of spinal myoclonus in our patient is that it is affected by supra spinal influences such as mental arithmetic. However, the final diagnosis of spinal interneuronitis can only be proved by histological study of spinal cord.

The patient made a substantial recovery following combination therapy with tizanidine, baclofen and clonazepam.

**References**


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