김 영 은
한림대학교 의과대학 신경과학교실

Clinical approach to myoclonus

Young-Eun Kim, M.D.
Department of Neurology, Hallym University Sacred Heart Hospital, Hallym University College of Medicine

Myoclonus presents as different clinical phenotypes according to the offending anatomical substrate. Careful history taking and neurological examination give the clue and shortcut for the pathophysiology of myoclonus and enable effective treatment. This review provides a comprehensive method to approach the myoclonic movement disorders.

Key Words: myoclonus, semiology, localization, approach

Introduction

Pathology of various movement disorders is mainly located in the basal ganglia or its connection. However, the anatomic substrate of myoclonus is not confined to basal ganglia or its connection and myoclonus can arise from cortex to peripheral nerve. Myoclonus shows different phenomenology according to the anatomical substrate of pathology. Careful examination of symptom and associated neurologic deficit can give the localization of pathology and make narrow differential diagnosis down. (1, 2)

Clinical approach to myoclonus

Myoclonus is defined sudden, brief, shock-like involuntary movement caused by muscle contractions or inhibitions. (1) Prerequisite before assessing myoclonus is that myoclonus should be distinguished from other hyperkinetic movement disorder such as tic, chorea, dystonic jerk or tremor. Although both myoclonus and tic are very brief movement, tic is distinguishably stereotypic movement with urge and can be suppressed. (2)

Overview of clinical approach to myoclonus is provided in the figure 1.

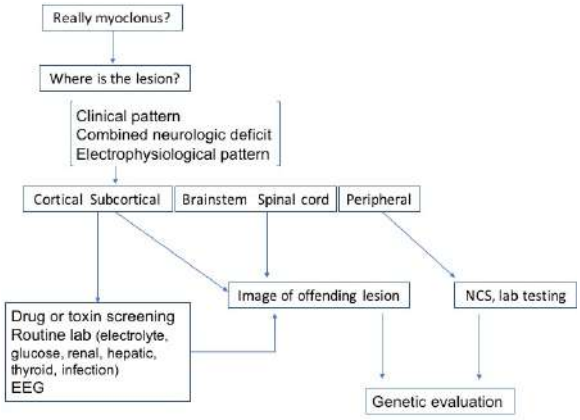

Figure 1. Overview of clinical approach to myoclonus

Young Eun Kim, MD
Department of Neurology, Hallym University Sacred Heart Hospital, Hallym University College of Medicine, Anyang, South Korea 22, Gwanpyeong-ro 170 beon-gil, Dongan-gu, Anyang-si, Gyeonggi-do, 431-070, korea
Tel +82-31-380-3740
email yekneurology@hallym.or.kr

Table 1. Different clinical and electrophysiological characteristics of myoclonus

P/Ex	Cortical	Subcortical	Brainstem (caudal brainstem)		Spinal		Peripheral
			Startle (Hyperekplexia)	Reticular reflex	Segmental	Propriospinal	
Mechanism	Sensory-motor cortex	Thalamic lesion	Fast conducting Bulbospinal pathway	Slow conducting Bulbospinal pathway	C/T cord (loss of inhibitory interneuron)	T cord generator common	a/w weakness or atrophy
Distribution	Mainly hand/face > leg Distal, flexor, Multifocal>focal, generalized	Unilateral, a/w dystonia	Excessive motor response or jump to unexpected stimuli, (Flexion of trunk/neck, abduction/flexion of arm)	Generalized axial jerk from SCM, trapezius (accessory nerve near reticular formation)	Focal or segmental (one or two spinal myotome)	Fixed pattern of generalized axial movement (one to multiple segment)	Focal, distal limbs, minipolymyoclonus
Activation profile							
Rest(spontaneous)	+		rare	+	+	+ (greatest when supine)	+
Action(intention)	+	+				-	+
Negative myoclonus	+	+	-	-	-	-	-
Reflexive	+ (noise or visual stimulus)		+ (Sudden noise or light, sensory stimuli to mantle area)	+ (greatest over the limb)	+	rare	-
Persistence during sleep	-	-	-	-	+	-	-
EMG Burst duration Back-averaging SEP	<100ms sharp before jerk Enlarged cortical		>100ms simultaneous Enlarged cortical	>100ms N/C Normal	>100ms N/C Normal	>100ms N/C Normal	<50ms N/C normal
Acquired cause of symptomatic myoclonus	Encephalopathy (Drug, toxin, anoxic, metabolic, physical, infectious), Focal structural lesion, neurodegenerative disease	Focal structural damage	Cerebral anoxia, inflammatory lesion, hemorrhage	Cerebral anoxia, toxin, metabolic, Stiffman, MS	Spinal cord lesion	Spinal cord lesion, often psychogenic	Lesion of Nerve root, plexus, peripheral nerve
Comments	Asterixis (toxic-metabolic cause, bilateral, cortical or subcortical origin)		Hereditary	[a]opsoclonus-myoclonus (brainstem origin)		Common in sleep wake transition	fasciculations/myokymia hemifacial spasm

P/Ex, physical examination; C/T cord, cervical/thoracic spinal cord; A/W, associated with; N/C, no correlate; SCM, sternocleidomastoid muscle;
[a]details of opsoclonus myoclonus syndrome is described in the text.

First step for the assessment of myoclonus is full history. Mode of myoclonus onset (acute, subacute, chronic), presence of other neurological problems (ataxia, parkinsonism, dystonia, dementia), history of seizure, drug or toxin history, past or current medical problems, and family history need to be evaluated carefully. (1)

Second, the most important step for the assessment of myoclonus is pattern recognition of movement. The distribution, temporal profile and activation profile of myoclonus should be evaluated. Distribution of myoclonus is divided into focal, segmental, multifocal, hemi, generalized regarding involved body parts and synchronization. Temporal profile means whether myoclonus is continuous or intermittent (sporadic or trains). Activation profiles can be an important clue: Resting/action myoclonus; positive/negative myoclonus; stimulus sensitivity (for touch, light, sound or muscle stretch); persistency during sleep. Although little is known about the sensitivity or specificity of these clinical features or with electrophysiological study of myoclonus subtype, it helps to recognize anatomical substrate of myoclonus such as cortical, subcortical, spinal, and peripheral form. (1, 2) Different clinical and electrophysiological characteristics of myoclonus according to the anatomical location are provided in table 1. (1-3)

Opsoclonus myoclonus syndrome is distinct myoclonus syndrome originated from brainstem. Opsoclonus is rapid conjugate ocular saccades without intersaccadic interval in all directions that are irregular in amplitude and frequency. Myoclonus is triggered by muscle contraction and axial and limbs muscles are involved. Ataxia can be combined. In adults, opsoclonus myoclonus syndrome is associated with paraneoplastic syndrome such as breast cancer or small cell lung cancer but is idiopathic in more than 50%. Several other disorders including stroke, multiple sclerosis, infection and toxin may be associated with this syndrome as well. (4)

For determining the etiology of myoclonus, next step is categorizing major clinical syndrome. Physiological myoclonus occurs in healthy individuals such as hypnic jerk. In essential myoclonus(11%), myoclonus is the most prominent or isolated clinical finding, sometime hereditary. Epileptic myoclonus(17%) is the presence of myoclonus in the setting of epilepsy as one component of a seizure. Symptomatic myoclonus manifests in the setting of an identifiable underlying disease which comprise 72% of myoclonus. (1) If symptomatic myoclonus, possible causes should be prioritized according to the offending anatomic substrate. If no symptomatic cause was found by drug screening, laboratory test and image, genetic evaluation can be considered according to the associated neurologic manifestation.

Conclusion

Approach to myoclonus is different with other movement disorders because the anatomical substrate offending myoclonus varies from cortex to peripheral nerve. Localization of myoclonus is important step for efficient approach of myoclonus

References

1. Caviness JN, Brown P. Myoclonus: current concepts and recent advances. Lancet Neurol. 2004;3(10):598-607.
2. Zutt R, van Egmond ME, Elting JW, van Laar PJ, Brouwer OF, Sival DA, et al. A novel diagnostic approach to patients with myoclonus. Nat Rev Neurol. 2015;11(12):687-97.
3. Shibasaki H, Thompson PD. Milestones in myoclonus. Mov Disord. 2011;26(6):1142-8.
4. Borg M. Symptomatic myoclonus. Neurophysiol Clin. 2006; 36(5-6):309-18.