Focal myokymia in carpal tunnel syndrome

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A 38-year-old Gravida 4 Para 3 at 28 weeks gestation presented with left hand paresthesias, morning stiffness, and loss of delicate movements of the fingers of 1-month duration. Symptoms began spontaneously about 3 months ago on entering the second trimester of her pregnancy and were not associated with any injury or change in her activities. She had similar symptoms in her left hand 8 years previously during her second pregnancy, which resolved on delivery. There were no further investigations. She was employed as a nurse and was right hand dominant.

She reported discomfort and intermittent twitching in her left thumb involving the thenar eminence. Neurologic examination showed slight weakness of the abductor pollicis brevis (Medical Research Council 4). Both Phalen test and Tinel sign were negative. There was an undulating, rippling, and wave-like involuntary movement consistent with myokymia confined to the left thenar eminence (video 1). The semirhythmic movement was elicited and aggravated by active wrist flexion. Nerve conduction studies showed prolonged latencies of the left median nerve compound muscle action potential and sensory nerve action potential and slowing of sensory conduction velocity consistent with demyelination in carpal tunnel syndrome.

Electromyography (EMG) showed a characteristic audio signal of myokymia known as the “marching soldier in the snow.” These are grouped repetitive discharges of the same motor unit, which occurs in quadruplets and multiplets. The interburst frequency was 2 Hz, whereas the intraburst frequency was about 50–60 Hz (video 2).

On the basis of history, physical examination, and electrophysiologic studies, the diagnosis of carpal tunnel syndrome was made. In view of her pregnancy, she opted for conservative treatment with splinting and a wrist brace that prevents wrist flexion.

The word “myokymia,” derived from the Greek myo–muscle and kymos–wave, describes involuntary, continuous, slow, undulating contractions of muscle fibers with specific clinical and electromyographic characteristics.¹ The pathophysiologic basis of myokymia has not been fully understood and seems to be related to motor axon membrane hyperexcitability and demyelination. An abnormal motor nerve generates spontaneous activity consisting of grouped repetitive discharges of motor unit action potential. Ephaptic excitation of demyelinated axons may also explain generation of myokymia.² Needle EMG shows characteristic myokymic discharges that are defined as spontaneous, semirhythmic bursts of potentials, firing at rates of 2–60 Hz.³

Myokymia has rarely been found in limb muscles and is most commonly associated with radiation-induced nerve injury. Limb myokymia associated with entrapment neuropathy is rare. To date, only 2 cases of focal myokymia associated with chronic carpal tunnel

PRACTICAL IMPLICATIONS

Localized appendicular myokymia is rarely seen distal to a nerve entrapment. Presence of myokymia, especially in specific muscle groups, should prompt clinicians to consider the differential diagnosis of entrapment neuropathies, e.g., abductor pollicis brevis in carpal tunnel syndrome.

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syndrome and another case in distal ulnar neuropathy have been reported.^{4,5} Myokymia has been described in association with diffuse disease of the central and peripheral nervous systems (radiation injury, Guillain Barre syndrome, MS, Bell palsy, rattlesnake venom poisoning, Isaac syndrome, and brain stem tumors) but rarely is seen involving the abductor pollicis brevis muscle in patients with carpal tunnel syndrome.^{6,7} This uncommon observation bears importance to the diagnosis and prognosis of entrapment neuropathies, especially carpal tunnel syndrome. Presence of myokymia, especially in specific muscle groups, should prompt clinicians to consider the differential diagnosis of entrapment neuropathies.

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Appendix 1 Author contributions			
Name	Location	Role	Contribution
Yong Chuan Chee	Hospital Sultanah Bahiyah	Corresponding author	Designed and conceptualized the study, analyzed the data, and drafted the manuscript for intellectual content
Beng Hooi Ong	Hospital Sultanah Bahiyah	Author	Organizing and supervising the course of the project and critical review of the manuscript
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References

1. Gutmann L, Gutmann L. Myokymia and neuromyotonia 2004. *J Neurol* 2004;251:138–142.
2. Rasminsky M. Spontaneous activity and cross-talk in pathological nerve fibers. *Res Publ Assoc Res Nerv Ment Dis* 1987;65:39–49.
3. Gutmann L, Libell D, Gutmann L. When is myokymia neuromyotonia? *Muscle Nerve* 2001;24:151–153.
4. Albers JW, Allen AA II, Bastron JA, Daube JR. Limb myokymia. *Muscle Nerve* 1981;4:494–504.
5. Streib EW. Distal ulnar neuropathy as a cause of finger tremor: a case report. *Neurology* 1990;40:153–154.
6. Gutmann L. AAEM minimonograph #37: facial and limb myokymia. *Muscle Nerve* 1991;14:1043–1049.
7. Zhang YQ. Teaching video NeuroImages: regional myokymia. *Neurology* 2010;74:e103-4.