Role of EMG evaluation in muscle hyperactivity syndromes

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Introduction

Muscle hyperactivity can manifest as involuntary twitches of muscle fibers or muscle groups or, more rarely, as a long lasting spasm. Clinically evident muscle hyperactivity must be considered abnormal only when it is consistent. Occasional involuntary twitches such as those experienced in the eyelids or in other muscles are probably only reflecting transient states of general stress or nervousness. Complaints of nonspecific muscle tension or tightness are usually accompanied by almost unapparent signs on clinical inspection or superficial examination. In these instances, however, EMG studies may reveal a fair amount of sometimes unsuspected activity that may present as isolated potentials, bursts, or more complex discharges, and are usually accompanied by peculiar sounds, which call for full attention from the examiner and colleagues around. EMG recordings carry useful information regarding the characteristics of the abnormal muscle hyperactivity, its site of origin, and the underlying pathophysiological mechanisms. When muscle hyperactivity does not have clinical expression, it may become evident only if an EMG examination is carried out because of symptoms or signs that may or may not be related to the same disorder as muscle hyperactivity. In these instances, the physician performing electromyography should be aware of the significance of the finding that, even apparently unrelated to the clinical problem that brings the patient to the examination, may reveal an underlying process of diagnostic relevance.

There is a very wide spectrum of conditions that lead to muscle hyperactivity. One possible classification of these disorders is the one taking into account the site of origin of the hyperactivity. Although in clinical practice the action potential showing abnormal muscle hyperactivity is practically always recorded from muscle fibers, the site in which the impulse has arisen will determine its shape and firing patterns. In this review, we describe the EMG characteristics observed in syndromes featuring muscle hyperactivity and the pathophysiology underlying the abnormal firing of muscle fibers.

Abstract

Muscle hyperactivity can be a clinical feature on its own or, more commonly, an observation on electromyography (EMG) examinations. Whatever manifestation it takes, muscle hyperactivity always means enhanced excitability of muscle, axons or neurons. Clinical findings may be variable, ranging from fasciculations to muscle cramps. Even though clinical examination may lead in most instances to suggest the diagnosis of the underlying disease, EMG studies are necessary to identify the type of abnormal discharges and suggest the site of their suspected origin. Although in clinical studies, the action potential showing abnormal muscle hyperactivity is practically always recorded from muscle fibers, the site in which the impulse has arisen will determine its shape and firing patterns. In this review, we describe the EMG characteristics observed in syndromes featuring muscle hyperactivity and the pathophysiology underlying the abnormal firing of muscle fibers.

Keywords

muscle hyperactivity · EMG · fasciculation · high frequency discharges · myokymia · neuromyotonia · stiff person syndrome
which the abnormal impulses arise will determine the shape and firing patterns of the action potentials. In the following paragraphs, we will consider three possibilities:

- the muscle fiber or motor end-plates
- the terminal branches of the axons, or any point along the axonal membrane
- the motoneuronal axon hillock

**Muscle hyperactivity originating in muscle fibers or motor end-plates**

Table 1 summarizes the syndromes in which muscle hyperactivity can be attributed to muscle membrane hyperexcitability. The simplest form of it is the fibrillation potential, which is due to the depolarization of a denervated muscle fiber, induced by the mechanical stimulus of needle movements or any chemical stimulus, such as small amounts of circulating acetylcholine. Denervated muscle fibers are sensitive to any of these stimuli, as well as to relatively low intensity electrical stimuli. This possibility is worth knowing not only for its physiological content, but also because of some clinical utility: In denervated muscles, hyperexcitable muscle fibers may be responsible for muscle action potentials recorded over the same muscle when trying to stimulate the unexcitable motor nerve. A variant of fibrillation potentials are the positive sharp waves, which arise from muscle fibers damaged by the needle. In this situation, the needle detects only the propagating action potential approaching the site of injury, not being able to pass through to generate the negative spike.

A reverse situation occurs with the end-plate spikes and bursts of insertional activity.

This type of activity is generated in the muscle end plate by irritation of the motor axon terminal twig or the muscle membrane. The action potentials, called end-plate spikes, always begin with a negative rising phase, not preceded by any approaching activity. They are single muscle fiber action potentials that may discharge in a short lasting burst of very high internal frequency, decreasing rapidly in both frequency and amplitude (Fig. 1). These high frequency discharges may be seen in muscles of otherwise healthy subjects, but may also be an early manifestation of diseases presenting with muscle fiber hyperexcitability, such as motor neuron disease, radiculopathies, polyneuropathies or myopathies.

Myotonic discharges are pathognomonic of myotonia (either myotonic dystrophy, myotonia congenita, or paramyotonia). Clinically apparent myotonia may be seen in glycogenosis, centronuclear myopathy, the Schwartz-Jampel syndrome, and other channelopathies. EMG recordings may be of help to distinguish true myotonic discharges from other types of muscle hyperactivity that can resemble myotonia in clinical inspection.

**Table 1** Muscle hyperactivity generated in the muscle fiber

<table>
<thead>
<tr>
<th>Type of activity</th>
<th>EMG correlate</th>
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<tbody>
<tr>
<td>end-plate spikes/insertional activity</td>
<td>Short bursts of single fiber action potentials</td>
</tr>
<tr>
<td>denervation</td>
<td>Fibrillation potentials/Positive sharp waves</td>
</tr>
<tr>
<td>myotonia</td>
<td>Repetitive firing of single fiber action potentials</td>
</tr>
<tr>
<td>complex repetitive discharges*</td>
<td>Repeated bursts of a group of muscle fibers</td>
</tr>
<tr>
<td>contracture</td>
<td>No electrical activity</td>
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</tbody>
</table>

* other terms such as ‘bizarre high frequency discharges’ and ‘pseudomyotonia’ are discouraged

**Fig. 1** Left: High frequency discharges of muscle fiber action potentials due to irritation of the muscle fiber with the needle electrode. The recording was done in a healthy subject. Note the initially negative rising phase, which is consistent with an action potential generated in the vicinity of the needle. This is the case with spike potentials, shown in B, recorded occasionally when the needle is approaching the motor end plate.

10 ms