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ORIGINAL COMMUNICATION

Focal neuromyotonia: do I love you?

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Abstract We present a rare case of focal neuromyotonia in a 73-year-old woman with a follow up of 5 years. The clinical picture showed a fixed contraction of the 3rd and 4th finger of the left hand. Similar to other published cases, our patient suffered from COPD and was treated with beta-2-sympathomimetics. This clinical picture shows a rare but rather salient differential diagnosis of Dupuytren's contracture. EMG of the affected muscles may yield a diagnosis and prevent the patient from a long and ineffective treatment "odyssey".

Keywords Focal neuromyotonia · COPD · Dupuytren · EMG · Neuromyotonic discharges

Case

We present the case of a 73-year-old woman with a gradual onset of painless flexion of her 3rd and 4th fingers on the left hand. In suspicion of a tendovaginitis stenosans, the ringbands of these fingers were operatively opened without any improvement. Examination revealed fixed contractions (Fig. 1). No weakness of the arm and hand muscles, no percussion myotonia, and no loss of sensibility or reflexes were found. Otherwise the neurological examination was normal.

Injections of 50 MU Botulinumtoxin-A into the flexor digitorum superficialis, and 60 MU into the intrinsic hand muscles in a second session, did not result in any improvement. During the EMG-controlled injection, spontaneous muscle activity in the form of neuromyotonic discharges of up to 30 s was found in the following muscles on the left: extensor digitorum communis, biceps brachii, flexor digitorum superficialis, flexor carpi ulnaris and brachioradialis.

The relevant medical history included severe chronic obstructive pulmonary disease (COPD) with centrilobular emphysema, of onset following partial lung resection for squamous cell carcinoma 6 years earlier. In the same year the patient required a pacemaker because of AV block and atrial fibrillation. She also had mild renal impairment and peripheral artery occlusive disease. Her treatment included prednisolone (25 mg per day) and salmeterol (50 µg as needed), aspirin, a statin, and antihypertensives (incl. betablockers).

On follow up after 5 years, the clinical picture (Fig. 1), as well as the neurological examination remained unchanged. Neuromyotonic discharges were found only in the flexor digitorum superficialis on the left side (Fig. 2). There was no pathological spontaneous activity in the extensor digitorum on the left or in the flexor digitorum on the right.

Discussion

We present a rare case of focal neuromyotonia. There are only a few other cases published [1–3]. However, this form of neuromyotonia might well be underdiagnosed. These

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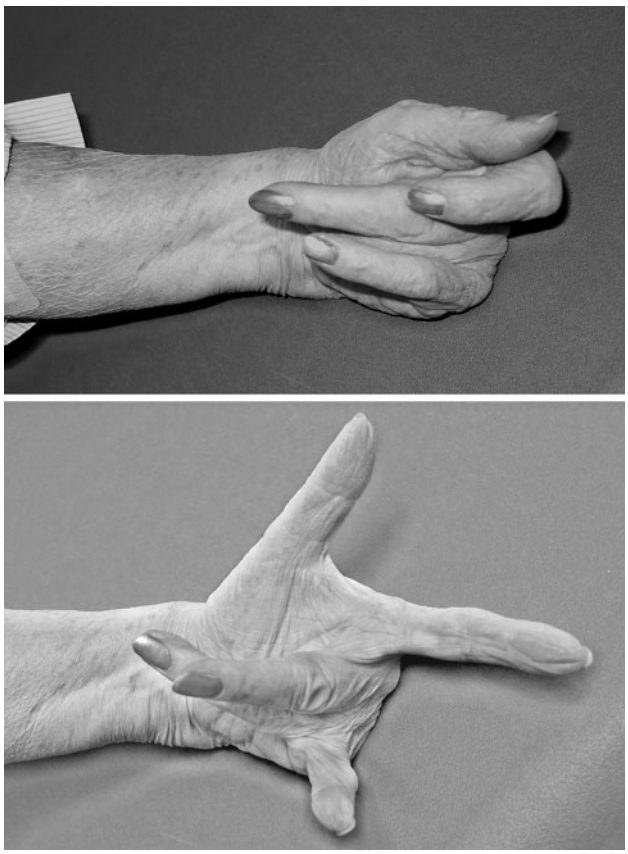

Fig. 1 Opening the left hand, one can perceive the sign of the Devil's horns ("mano cornuto"). But, this is also the symbol for "I love you" in sign language

patients are typically seen in an outpatient hand-surgery or rheumatology clinic, and may be misdiagnosed as Dupuytren's contracture or fixed trigger finger. Some are even operated on, albeit unsuccessfully. The neuromyotonic discharges during EMG-assisted botox-injection led to the diagnosis of a focal neuromyotonia. Further diagnostic tests (such as voltage-gated potassium channel antibodies) were declined by the patient. However, the focal phenotype remained unchanged in the 5 years follow up.

In similarity to the other published cases [1, 2], our patient presented suffered from COPD and was treated with beta-2-sympathomimetics. Enhancement of axonal hyperexcitability due to hypoxia (from COPD) and effects on sodium–potassium pumps, leading to hyperpolarisation through activation by sympathomimetics, may be potentially contributory [4, 5]. However, the exact mechanisms by which a few elderly patients with similar disposition develop this rare neuromuscular disease, especially with such a focal phenotype, remain unclear. In our case, we note that the Botox injections had no effect on the contractions. As botulinum toxin A acts through praesynaptic inhibition of acetylcholine release [6], we would hypothesise either a mechanical dysfunction by transformation of muscle into fibrous tissue, or an additional postsynaptic mechanism at the neuromuscular junction or the muscle itself as underlying pathology.

This clinical picture shows a rare but rather salient differential diagnosis of Dupuytren's contracture. In atypical cases, particularly in patients with COPD, a referral to

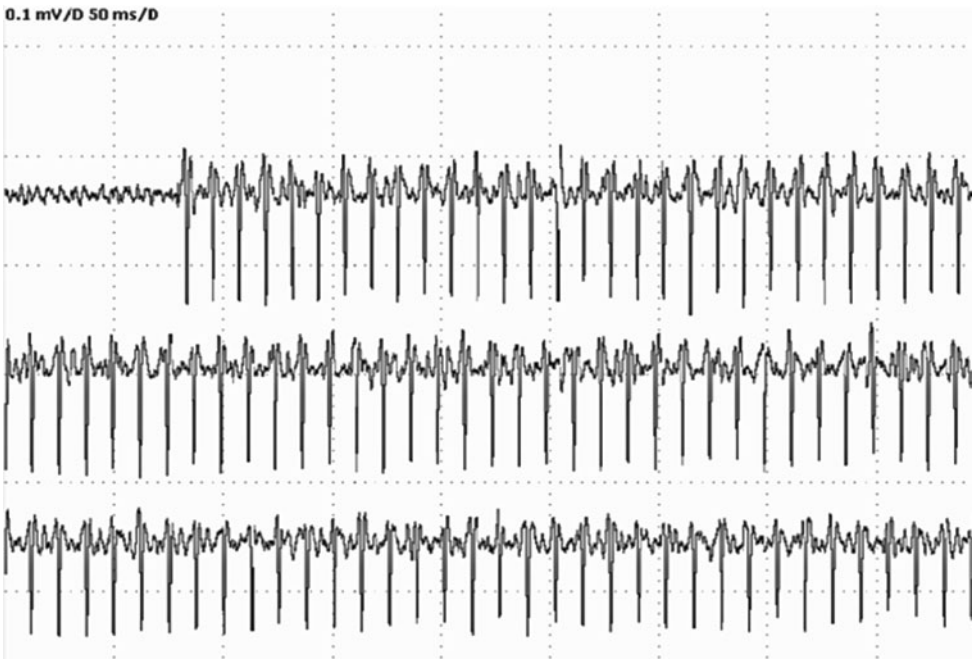

Fig. 2 EMG in the left flexor digitorum superficialis muscle showing neuromyotonic bursts (spontaneous high frequency discharges with waning amplitudes and abrupt ending)

a neurologist might be helpful. EMG of the affected muscles may yield a diagnosis and prevent the patient from a long and ineffective treatment "odyssey".

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