Acta Myologica • 2013; XXXII: p. 170-173

MEMORIES OF A MYOLOGIST

Differential diagnosis and treatment of muscle hypertonia as practiced in Zagreb's Centre/Institute for Neuromuscular Diseases

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Because I am a neuromyologist that has dealt for many years with muscle hypertonia, I decided to write my memories in order to motivate younger researchers to try to duplicate the same observations and experiences.

We defined a whole range of conditions and symptoms, partly or in full. That is the first crucial step on the way to suppressing or relieving suffering. In some cases there was nothing we could do. In the other cases, we managed to diminish the uncomfortable symptoms. In still other cases, we cured the diseases, at least for a while. My conclusion is that great and systematic effort is always worth the trial. Maybe someone will follow us?!

Always again, I used to repeat to myself and to the others that, when approaching the patient, always the following rules should be respected: watch, listen and use your own common sense to evaluate what you observe; analyse why the symptoms occur in a concrete case; include the therapy in the logic of symptom development; continue to follow the patient and ask questions of yourself and of your colleagues; consult the literature; find the differences; ask again and again what else could be done … and the solutions will appear unexpectedly.

Key words: Spasticity, cramps, neuromyotony

Introduction

In neurology, we consider the muscle tonus increased if, by passive movement of extremities or parts thereof, a resistance occurs, in spite of the patient being fully relaxed. If we cannot passively change the position of an extremity at all, we are observing contracture. By elimination of the heightened tonus, and especially contracture, the movement is freed. Many decades ago, as a young neuropsychiatry specialist, I started to worry about how to help the patients with increased muscle tonus conditions. A summary of the different clinical conditions and their pathogenesis, diagnosis and treatment are listed in Table 1.

Increased muscle tonus as a consequence of central nervous pathways damage

These were mainly patients with spasticity or rigidity of the Parkinsonian type. When attempts to suppress spasticity by phenol blockades (1) did not lead to the desired effect, I went in 1968 to H.F. Hufschmidt in Frankfurt-Main, Germany, on a DAAD scholarship, to become acquainted with Hufschmidt's method of low-frequency electrostimulation in spasticity. Upon my return, the Neuropsychiatry Department of Rebro Hospital acquired Hufschmidt's machine and we started applying it following his scheme, beginning with spastic patients (2). I tried using it later with other indications. Those were mainly characteristic of the Parkinsonian syndrome, and we achieved very nice effects in a number of cases (3). In the meantime, l-dopa came to Croatia and the positive effects on rigor were now faster and easier achievable, so the electrostimulation for Parkinson's Disease lost its significance. As a side effect of electrotherapy we noticed improvement in retention and incontinence of urine, and even in sexual function. We elaborated Hufschmidt's scheme of muscle stimulation and achieved desired results by an indirect approach (4). The method survived in the Lipik and Varaždin Rehabilitation Centres and in the Urologic Department of the Rebro Hospital in Zagreb.

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Table 1. Features differentiating already used terms.

Names of clinical features of muscle hypertonus	Pathogenesis	Diagnostic examinations	Treatment
Spasticity	Lesion of the corticospinal pathways	Clinical examination, neuroimaging methods	Phenol injection, electrostimulation sec. Hufschmidt, kinetic therapy, baclofen etc
Rigidity	l-dopa deficiency	Clinical examination, neuro-imaging methods	l- dopa, etc
Cramps	Heredity, secondary to various causes		Mg++, etc
Tetanic spasms	Ca++ or Mg++ deficiency	EMG multiplets provoked by ischemia or hyperventilation	Ca++ or Mg++, D3
Myotonia	Slowed muscle relaxation due to myogenic electrical hyperexcitability	Clinical examination, EMG, warm-up test	Sodium channel blockers, carbamazepine
Neuromyotonia	Heredity, peripheral distal nervous lesion	Clinical examination, EMG	Carbamazepine, corticosteroids
Contracture	Various	Clinical examination, local curare test	Neurolysis
Paroxysmal, symmetric, generalised spasms in full consciousness	Spinal lesion	Clinical neurological observation, spinal MR	Corticosteroids

Increased muscle tonus due to peripheral nervous system and muscle damage

With the development of the Centre for Neuromuscular Diseases in Zagreb, patients with increased muscle tonus due to nerve or muscle disease started to arrive much more frequently. As the first entity, we differentiated hereditary distal muscle cramps in one family. Albrecht Struppler considered the finding very interesting and enabled me to take part in the International Congress of Neurology in New York 1969 and to present the results for discussion to the wide scientific audience.

Encouraged by the discussion, I sent my article to the *"Journal of Neurology, Neurosurgery and Psychiatry"*, at that time the most distinguished neurologic journal (5). Later on, we also published a paper on two and three related families with the almost the same syndrome (6, 7). We also made a videotape (8) to better present that transient, painful muscle hypertonus. The activity of involuntary contraction was included in the electromyographic evoked secondary muscle potential to which I paid considerable attention. We published a picture of this phenomenon in the first English article. No medication able to suppress those very uncomfortable symptoms was found.

Localized and generalized stiffening, contractures and spontaneous EMG activity

More or less at the same time, tetanies was also a focus of my interest. I brought along French literature on spasmophilia due to the lack of magnesium, a topic that was very up-to-date by the end of the 1960s, especially in Paris at L'Hôpital Salpètiere. I standardised the ischemic and hyperventilation tests for "multiplets" provocation on the EMG screen and of clinically visible distal spasms of tetany. The test results were often positive in different neuromuscular nosological units. Therefore, one of my students in electromyography, who was from Split, was put in charge of elaborating the phenomenon on a larger group for his master's degree work. Unfortunately, he never finished it. My text in the proceedings of the meeting in Ljubljana (9) remained the only one for a long time.

I continued to follow up the symptoms of hypertonus in neuromuscular diseases and, as early as 1972 (10), we published an article on the effect of carbamazepine on stiffening in dystrophia myotonica without suppression of electromyographic serial discharges. In 1976 (11), we described the prolonged effect of intramuscular injections of Lignocaine on dystrophic myotony and stiff-man syndrome, with stiffness in limbs and in swallowing and mastication. (Looking back, we would now call it neuromyotonia instead of stiff-man syndrome). In this neuro-

myotonic patient, we also applied carbamazepine which had a clear positive effect.

In 1982, in the preliminary report in a case of neuromyotonia (12), the authors pointed to the cerebrospinal liquor inflammatory changes, along with reduction of spontaneous electromyographic activity and stiffening by carbamazepine. The stiffness and spontaneous activities were stopped entirely with fluocortolone. The authors also found cerebral atrophy and lively myotatic reflexes after suppression of stiffness by carbamazepine. In experimental studies of the case, the spontaneous EMG activity would disappear only after distal nerve infiltration with Xylocaine. On proximal or intramuscular examination, it did not change significantly. The article was sent to an Austrian periodical but it was rejected with commentary that the immunological analysis was too insufficient to allow the disease to be declared immunologic. We published the case in *Extenso* in 1984 (13).

We continued to discover many patients with similar main symptoms: for example, there was a patient with hypoparathyroidism, generalized stiffness and abundant spontaneous electromyographic activity (14), then there was a baby and a child with neuromyotonia (15, 16) and four adolescent patients - two sporadic and two hereditary (17). All of them improved on carbamazepine. In the two sporadic adolescents, even an extensive remission on corticosteroids was recorded. In one hereditary case, intensive contracture in the arm joint disappeared after a local curare test (18). Pronounced contractures in the wrist area and interphalangeal joints, in four adult patients with dystrophic myotony improved also following a regional curare test. The result pointed to the possible origin in distal nerve irritation (19).

Correctible contractures, with lasting extensive improvement

The observed contractures developed in a short time, limited to the third, fourth and fifth finger and wrist, with some sensibility damage in the ulnar nerve innervation region. In two cases, the compression was differentiated in the elbow region; in the one, it was on the lower arm. Instead of tenotomy or an operation of the muscle, neurolysis was done. The location was indicated by plurisegmental electroneurography. Contracture disappeared very soon after the operation (20).

Persistent contractures

Very precise differential diagnostics of a slowly progressive diffuse contracture of the spine ("rigid spine syndrome") was conducted, associated with thorax deformity, mainly proximal myopathy and delayed sexual maturity, in three unrelated patients (21).

In 1989 (22), I published an article about progredient syndrome in two generations, when the syndrome had not yet been described in the literature. It had been considered as joint disease. However, it was obviously a disease of the muscle. A very slow progredient contracture of the fingers is the dominant symptom. Percussion of muscle causes depression only on the tongue, while on the extremities it causes extreme high skin bulging. On forced, passive extension of muscles, repetitive EMG activity occurred as registered by special bipolar wire electrodes used in my other kinesiologic studies. On the local curare test the contracture disappeared and the percussion response was reduced.

Paroxismal, generalised, very painful spasms in full consciousness

After occurring for five days, at intervals of a few minutes, throughout the day and night, generalised spasms almost led to the death. Individually adjusted and prolonged therapy by methyl-prednisolone led to complete remission with the patient living a normal life for years afterwards. Within the first months of treatment, spinal MR demyelinization symptoms also disappeared (23).

Conclusion

We tried to influence muscle hypertonia, defined clinically as resistance to passive movements of extremities or their parts. The author first refers to hypertonus of central origin which we tried to suppress by subarachnoidal application of phenol, and later on by low frequency electrostimulation according to Hufschmidt's system. Positive effects on Parkinsonian rigidity and akinesia were found as well. The adapted technique was applied with good results even on retention and incontinence of urine.

For the first time, a syndrome of transient painful cramps of peripheral genesis was differentiated as a hereditary disease without the possibility of being improved. At the same time, we developed ischemic and hyperventilation tests for chronic tetany, applying them to different conditions. The resistance in dystrophic myotony was reduced by carbamazepine or Lignocaine with unchanged spontaneous EMG activity. As early as 1982, we differentiated a patient with neuromyotonia, whose symptoms were reduced by carbamazepine; they then completely disappeared on corticosteroids. Patients with neuromyotonia kept appearing.

We differentiated a new neurological symptom of subacute contracture of fingers that disappeared very quickly on ulnar nerve neurolysis. In three unrelated patients, we differentiated slowly progressive contracture of the spine with proximal myopathy, and, until then not described, a syndrome of hereditary progressive contracture of fingers accompanied by extreme muscle percussion symptom and special repetitive EMG activity. In one patient, with spinal MR pathology the frequent, very painful paroxysmal, generalised spasms disappeared fully on corticosteroids.

All these significant results were the consequence of steady application of the basic rules cited above: watch, listen and use your own common sense and experience; ask questions and compare!

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