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Variant form of stiff-man syndrome with neck pain: report of a case treated with muscle afferent block

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Introduction

The stiff-man syndrome is characterized by progressive fluctuating muscular rigidity and painful spasm [1]. Rigidity and spasms may dominate in the axial muscles, or in one or more distal limbs. We describe a patient who fulfilled, although not completely, most of the diagnostic criteria for the stiff-man syndrome. Muscle spasm and pain were uncontrollable with diazepam and baclofen. Muscle afferent block (MAB) by injection of local anesthetics and absolute alcohol into the muscle effectively ameliorated the symptoms.

Case report

A 64-year-old man was first aware of stiffness and pain in the neck 5 years ago. He was admitted to the hospital twice and the stiff-man syndrome was diagnosed. He was treated with diazepam and baclofen with no favorable results. The patient was referred to our clinic. Examination revealed marked rigidity of the sternocleidomastoid and trapezius muscles and involuntary muscle contraction of the upper extremities and truncus. The symptoms were increased by rest and emotional stress, decreased by voluntary movement, and disappeared during sleep. The patient had pain on the entire trapezius muscle with no trigger point detected. There were no abnormal findings in muscle power (by sthenometry), deep tendon reflexes, sensation, or mentation. Surface electromyograms showed continuous discharges at rest in the trapezius, deltoid, biceps, and triceps muscles (Fig. 1a).

After obtaining informed consent, we performed MAB on the trapezius muscle, which were the site of severe pain. Bupivacaine (0.125%, 10 ml) was injected into the muscle (MAB with local anesthetics) twice a week. The pain and rigidity of the trapezius muscle were relieved for 2 to 5 h after the block, while voluntary muscle power was preserved. We therefore commenced MAB with alcohol, which has been shown effective for dystonia [2].

The techniques were as follows. The trapezius muscle was identified (the involuntary contracting muscle is identified with echo), and 8 ml of 1% mepivacaine was injected into the muscle belly using a 25-G, 25-mm needle, over 30 s. This was followed by injection of 1 ml of absolute alcohol over 10 s and then injection of 2 ml of 1% mepivacaine over 10 s.

Mepivacaine given before and after alcohol injection is helpful to prevent severe pain from alcohol injection and from possible leakage during withdrawal of the needle. The block was repeated twice a week, mainly on the left side, and a few times on the right side. The symptoms were substantially improved. Induration at the injection site was the only complication.

MAB was performed a total of 40 times, using either bupivacaine alone (30 times) or alcohol in combination with mepivacaine (10 times), resulting in a decrease in pain from 83 mm to 39 mm on a visual analogue scale. Sustained rigidity in the trapezius muscle disappeared, and rigidity was observed only intermittently. Rigidity of the trapezius muscle, which had been observed in the entire muscle, was now confined to a smaller area, and some trigger points became apparent. Although involuntary movement of the muscles continued, pain became controllable with trigger point block once every 2 weeks. Surface electromyograms after the latest MAB showed minimum discharges in the trapezius muscles.

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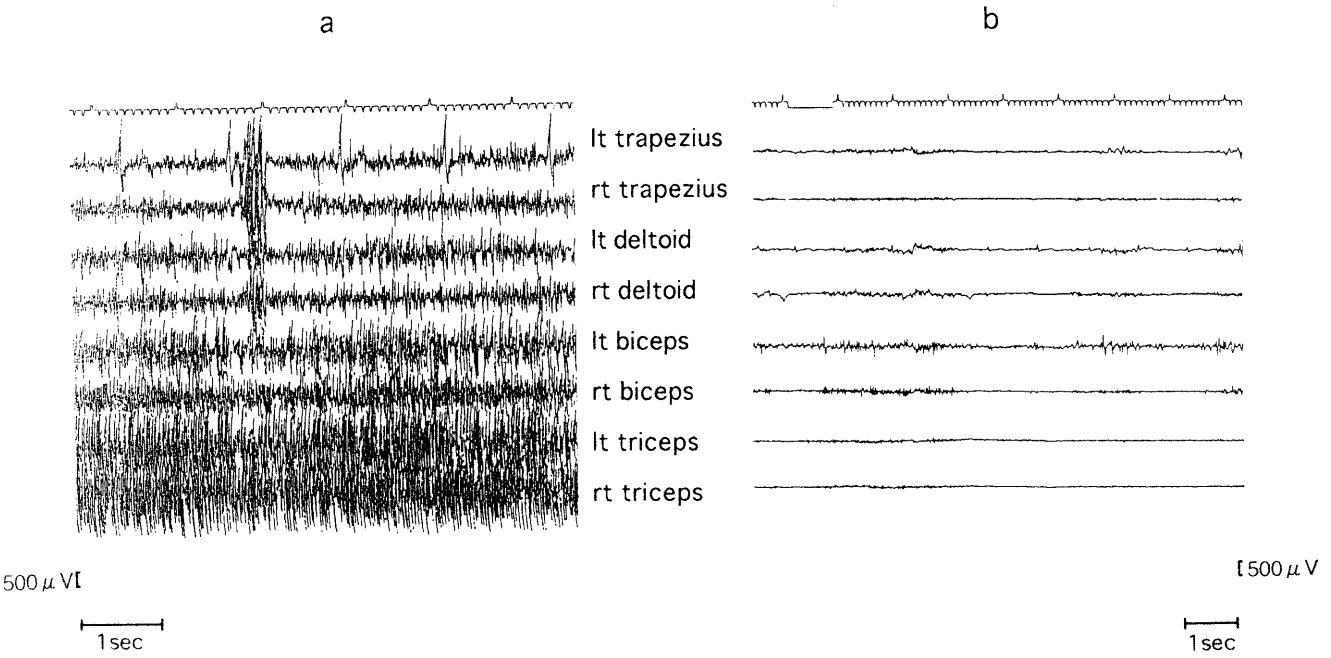

Fig. 1. Surface electromyograms (at rest). **a** Before muscle afferent block, **b** after muscle afferent block with alcohol. Recording chart speed is different between **a** and **b**. Time constant = 0.03 s. Note continuous discharges in trapezius, deltoid, biceps, and triceps muscles. The continuous discharges were markedly decreased after the latest muscle afferent block

Table 1. Values of CPK, LDH, AST, aldolase, and lactate before and after muscle afferent block with alcohol

				After			
Enzyme	Until	Normal range	Before	3 h	2 wk	4 wk	6 wk
CPK	$IU \cdot l^{-1}$	76–251	143	691	242	252	216
LDH	$IU \cdot l^{-1}$	115–217	118	308	298	263	286
AST	$IU \cdot l^{-1}$	12–34	34	47	44	52	26
Aldolase	$IU \cdot l^{-1}$	1.7–5.7	7.6	13.5	10.8	9.7	8.6
Lactate	$mmol \cdot l^{-1}$	0.4–1.3	1.2	1.5	1.9	1.6	1.6

CPK, Creatinine phosphokinase; LDH, lactate dehydrogenase; AST, aspartate aminotransferase.

The discharges on other muscles were also decreased (Fig. 1b).

The results of blood chemistry analysis before and 3 h, 2 weeks, 4 weeks, and 6 weeks after the latest MAB with alcohol (performed 20 months after the preceding MAB with alcohol) are summarized in Table 1. There were moderate increases in creatinine phosphokinase, aldolase, and lactate and slight increases in lactate dehydrogenase and aspartate aminotransferase, with variable degrees of recovery. The patient was negative for the glutamic acid decarboxylase (GAD) antibody.

Discussion

The stiff-man syndrome is characterized by muscle rigidity and painful spasm [1]. This syndrome is thought to be caused by dysfunction of the descending tracts controlling muscle tonus and exteroceptive reflexes [3], or by the existence of autoantibodies to gamma-aminobutyric acid (GABA)-ergic neurons [4,5]. However, the mechanisms of this syndrome are not fully understood.

Our patient had severe pain in the spastic muscles, mainly in the whole trapezius muscle, with no obvious trigger point. Muscle spasms were increased by rest or emotional stress, decreased by voluntary movement, and disappeared during sleep. These findings preclude reflex myospasm, myofascial pain syndrome, and fibromyalgia syndrome. Some of our patient's symptoms differ from those classically reported for stiff-man syndrome, i.e., muscle rigidity was confined to the neck, upper extremities, and trunk, and the rigidity decreased

Table 2. Clinical features and findings in subdivisions of stiff patient syndrome [7]

1. Stiffness and painful spasm of limb, usually of the calf and foot, with difficulty walking
2. Spasms may be spontaneous, reflex, or provoked by voluntary action
3. Fixed abnormal posture of distal limb
4. Relapsing and remitting course
5. Upper limb involvement
6. Sphincter involvement
7. EMG during action and reflex-induced spasm is abnormally segmented, consisting of the grouped discharges of many motor units
8. Poor response to baclofen and diazepam
9. No insulin-dependent diabetes mellitus (IDDM)
10. Anti-glutamic acid decarboxylase (GAD) negative

with voluntary movement. Several subdivisions of stiff-patient syndrome have been reported, including stiff-limb syndrome, in which muscle stiffness and spasms are confined to the distal limbs [6–8]. There have been no established criteria for the diagnosis of stiff-man syndrome, but the clinical characteristics shown in Table 2 were reported by Baker et al. [7]. Our patient exhibited six of those characteristics fully (4, 5, 7, 8, 9, and 10) and partially exhibited three of them (1, 2, 3). Thus, our patient may have a variant form of this disorder.

Our patient was negative for anti-GAD antibody. However, most series to date report that only around 60% of patients are positive for this antibody [7]. Furthermore, in patients with stiff-limb syndrome, the prevalence of this antibody is around 15% [7].

Diazepam and baclofen are the drugs of choice [9,10]. Plasmapheresis or administration of adrenocortical hormones has been advocated in view of the existence of autoantibodies against GABA-ergic neurons [11,12]. Our patient was treated with diazepam and baclofen, with no satisfactory results. Plasmapheresis or immunosuppressive therapy was not performed, because the effectiveness of these therapies is not certain in the nonclassical type of the syndrome [7]. We decided to perform MAB on this patient, because his symptoms were localized and severe pain was confined to the trapezius muscle and was uncontrollable by medication.

Botulinum toxin has recently been used and has proved effective as a local treatment for abnormal muscle contraction. However, antibodies to the toxin are easily produced when large doses of toxin are injected into large muscles, and the treatment becomes difficult after antibodies to the toxin have been produced. In our patient, MAB was effective without apparent loss of muscle power. Walsh [13] first reported the injection of large amounts (35–50 ml) of 1% procaine into the muscles of the upper extremities in a patient with Parkinson's disease. Kaji et al. [14], who used 0.5% lidocaine instead of procaine, termed this block MAB and used it to treat writer's cramp. MAB reduces muscle rigidity with relatively little effect on muscle power or the electrically elicited M wave. MAB is supposed to be a partial blocker of the γ-loop, which tonically controls the sensitivity of the muscle, although the precise mechanism of action is unknown [2]. Since the smaller γ-fibers supplying the muscle spindles may be more selectively blocked by low concentrations of local anesthetics, the therapeutic mechanism of the so-called MAB may be the result of blockade of γ-motor nerves. The effectiveness of MAB in our patient suggests that hyperactivity of γ-motor neurons may be involved in the pathology of the stiff-man syndrome and its variant forms.

In our patient, MAB with alcohol caused formation of induration at the puncture site, which made interruption of the therapy necessary. However, we were able to solve this problem by substituting local anesthetics for alcohol for about 1 month, with no worsening of symptoms. A moderate increase in myogenic enzymes and lactate was observed in our patient. This may suggest that the muscles are somewhat affected by alcohol, but the changes are mild, with a tendency toward recovery. The risk of neuropathic pain fron MAB with alcohol cannot be completely excluded, even if absolute alcohol is diluted with local anesthetics, but it was not seen in our patient. Obtaining informed consent is necessary to perform MAB with alcohol.

In summary, MAB with local anesthetics and/or alcohol was effective for pain and muscle rigidity in a patient with a possible variant form of stiff-man syndrome. Further study is desirable to determine the optimal mixture of alcohol and local anesthetics and the usefulness of dibucaine or phenol, as well as the possibility of applying this block to other diseases in which abnormal muscle contraction and pain are found, such as spasmodic torticollis and focal muscle cramp.

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