

Efficacy of Stellate Ganglion Block in Cholinergic Urticaria with Acquired Generalized Hypohidrosis

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Cholinergic urticaria with acquired generalized hypohidrosis, and its pathophysiology is not well known. Autoimmunity to sweat glands or to acetylcholine receptors on sweat glands has been mentioned as one of the possible etiologies. Systemic steroid therapy, antihistamines, anticholinergics, and avoidance of the stimulatory situations are recommended for treatment. We experienced a case of cholinergic urticaria with acquired generalized hypohidrosis in a patient who had no other associated disease, and the symptoms eased after repeated bilateral stellate ganglion block. Stellate ganglion block normalized the elevated sympathetic tone and may relieve symptoms in patients with this condition. (Korean J Pain 2012; 25: 278-280)

Key Words:

cholinergic urticaria, hypohidrosis, stellate ganglion block.

Cholinergic urticaria is a condition characterized by erythematous pruritic wheals and caused by an increase in core temperature, physical and emotional stress, and spicy foods [1]. Cholinergic urticaria with generalized hypohidrosis can be caused by functional abnormality of acetylcholine receptors or by poral occlusion [2]. The authors report a case of cholinergic urticaria with generalized hypohidrosis and systemic pain that were improved through treatment with repetitive stellate ganglion block.

CASE REPORT

A 24-year-old male was admitted to the hospital due to systemic erythema and wheals with an itchy sensation,

which he had had for 9 years. A stinging pain, rated 7 cm on a visual analogue scale (VAS: from 0 to 10 cm), was one of the dermatological symptoms and was predominant when the patient ate hot or spicy foods, engaged in a vigorous workout, took a hot bath, or experienced situational nervousness or stress. The symptoms disappeared within 30-90 minutes of rest or symptomatic relief.

When the patient was exposed to heat, the symptoms worsened, with an excessive hot flush sensation; the symptoms were relieved when he rested in cool conditions. The symptoms were worse during winter than during summer. There were no signs of autonomic dysfunction such as orthostatic hypotension, impotence, and photophobia. There was nothing significant in the family history

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or the physical examination.

The results of the blood laboratory test, liver function test, urinalysis, rheumatoid factor, coagulation test, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and chest X-ray were either within the normal range or negative.

Glycopyrrolate 2 mg/day was administered orally for 1 week for diagnostic purposes, and a symptomatic change was observed. After the reduction in diaphoresis from the administration of glycopyrrolate, the hot flush became severe and the patient still complained of pain with a VAS rating of 7 cm. The medication was ceased under the impression of possible cholineraic urticaria with generalized hypohidrosis, and a right and left stellate ganglion block was carried out by administering mepivacaine 10 cc with a 20-minute interval.

In the 1-week follow-up, the frequency of symptoms was similar, but there was a reduction in the degree of excessive hot flushes, and the systemic pain had a VAS rating of 4 cm. An extra stellate ganglion block was since carried out, and the VAS rating was reduced to 2 cm.

DISCUSSION

Cholinergic urticaria is characterized by systemic erythema and wheals with an itchy sensation and is brought by an increase in body temperature after an exercise or hot bath [3,4]. The diagnosis is generally made from the patient history, and differential diagnosis should be made with respect to exercise-induced urticaria or anaphylactic reaction to foods through a provocation test [5].

The pathology of cholinergic urticaria is not clearly identified but can be divided into 4 large categories according to the cause: poral occlusion, acquired generalized hypohidrosis, sweat allergy, and idiopathic [1]. From these 4 categories, cholinergic urticaria with generalized hypohidrosis is defined by urticaria with reduced sweat secretion even with sufficient stimulation. The etiological factors are occlusion of the sweat glands due to keratinization, autoimmune disease or disappearance of the sweat glands as a congenital defect [2], and a malfunction of the autonomic nervous system [6]. Stimulation of the sympathetic nervous system causes the secretion of acetylcholine, and sweat secretion occurs as feedback of the reaction of acetylcholine receptors in the sweat glands.

Sawada et al. [7] reported through skin tissue biopsies

that patients with cholinergic urticaria with generalized hypohidrosis have reduced cholinergic receptor muscarinic 3 (CHRM 3). They hypothesized that the reduction of CHRM 3 in sweat glands leads to insufficient sweat secretion and an increase in acetylcholine secretion through a feedback mechanism, and the increased serum acetylcholine induces the surrounding master cells to secrete histamine or stimulate sensory nerve terminals, which in turn causes cholinergic urticaria and pain [1,7].

The authors used anticholinergic drugs to differentiate sweat allergy from a hypersensitivity reaction after sweat secretion and cholinergic urticaria with generalized hypohidrosis, and a worsening of the patient's symptoms was observed soon after administration of the medication and the reduction in sweat secretion. Therefore, the reduction of sweat is thought to be the etiological factor in this patient.

In this case, the patient did not have any significant medical history such as specific physical appearances or intellectual abnormalities to suggest the possibility of congenital diseases. Moreover, the patient did not present any abnormalities in the autonomic nervous system except for hypohidrosis. Diagnosis is thus possible, as the sweat glands disappeared due to autoimmune disease or were occluded by keratinization rather than congenital defects.

Antihistamines and anticholinergic drugs are the standard treatments for cholinergic urticaria. However, most patients experience moderate relief from an antihistamine [1,8] and minor relief from anticholinergic drugs [1,9].

Systemic steroids can be administered when the cause of hypohidrosis is destruction or occlusion of sweat glands due to autoimmune disease [2,10]. Keratolysis can be chosen as a treatment for the blockade of the opening of sweat glands due to keratinization. Desensitization through repetitive sweating by bathing and exercising can be treatments for sweat allergy [11].

The stellate ganglion block is effective at stabilizing the hypertonic sympathetic nerve system in the head, neck, and arms [12,13]. It is known to temporarily block the sympathetic nervous system, and it also disconnects the pain circle and recovers the balance of the somatic nervous system and sympathetic nervous system within the area where pain occurs [13].

Cholinergic urticaria with generalized hypohidrosis is a symptom that arises when the sympathetic nerve system is activated due to excessive exercise or bathing and suffi-

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cient sweat secretion does not occur. This increases the serum acetylcholine level and activation of the sympathetic nervous system through a feedback system, and the stellate ganglion block can facilitate improvement of symptoms.

However, persistent nerve therapy and follow-up observations are needed for an accurate determination of efficacy, consideringthat stellate ganglion block also causes hypohidrosis [14] and the low frequency of the stellate ganglion block procedure.

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