

# A CASE OF ATYPICAL COLD URTICARIA WITH AN UNUSUAL PHENOTYPE

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## Summary

*Acquired cold-induced urticaria is an uncommon form of chronic urticaria, characterized by development of wheals, angioedema or both in response to cold exposure. In this report it is presented a case of atypical cold urticaria with an unusual phenotype: A female patient covering herself with hijab referred to dermatology outpatient clinic with redness and itching only on the exposed regions. In clinical examination there was erythema, strictly delineated by the boundaries of her garment, on her face and hands. She was 34 years old and these manifestations were recurring for three years in winter times with an increasing severity. Total IgE level was 157 IU/mL (normal value < 100 IU/mL). The ice cube test for verification revealed a negative result, which renders the case as atypical according to classification criteria. During one month follow-up with a triple dose antihistamine daily, the attacks were controlled to a great extent. This case, although it was in type-1 severity clinically, the needed antihistamine dose to suppress the manifestations was in the range of type-3 severity. The tendency to increase in clinical manifestations during the course of the disease and the relative higher doses needed to suppress the symptoms should alert the physician for a probable generalized reaction. Our case, besides being an atypical one, is also interesting to show how the cold sensitivity of the skin can be so precise, displaying such sharp borders.*

**Keywords:** cold urticaria, atypical, unusual phenotype.

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## Introduction

Acquired cold-induced urticaria (ACU) is an uncommon form of chronic urticaria [1], with an estimated incidence of 0.05% [2], and higher rates in cold-climate countries [3]. The majority of the patients are young adults with female predominance [4]. It is generally characterized by development of wheals, angioedema or both in response to cold exposure. Patients with ACU may react to various cold triggers, including exposure to cold air, contact with cold fluids or solid surfaces, and the ingestion of cold foods or drinks. Its pathophysiology comprises the cold-induced formation of autoallergens and IgE response to these autoallergens, which provoke

the release of proinflammatory mediators from skin mast cells [3]. The diagnosis should thoroughly be verified via cold stimulation tests (CSTs), the ice cube test being the traditional standardized method. There is a small proportion of patients having the same clinical manifestations which can not be reproduced by standard CSTs. Those cases are classified as atypical ACU and the diagnosis primarily depends on the patient's clinical manifestations or history [1].

## Case presentations

A female Caucasian patient covering herself with hijab referred to dermatology outpatient clinic with redness and itching on the exposed

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(a) (b) (c)  
Figure 1. The forehead (a), left hand (b) and lower two-thirds of the face (c)

regions. She was 34 years old and these manifestations were recurring for three years in winter times. The current winter she was expressing more severe redness and itching compared to previous years. In clinical examination there was erythema, strictly delineated by the boundaries of her garment, on her face and hands (Fig. 1).

Outside temperature was 6 °C on the examination day, and she emphasised that the redness and itching were getting more severe in colder temperatures. She had no history of a co-existing chronic disease, and her family history was also negative. In laboratory examination her differential blood count, C-reactive protein, erythrocyte sedimentation rate, thyroid stimulating hormone, alanine transaminase, aspartate transaminase and creatinine values were normal. Her total IgE level was 157 IU/mL (normal value < 100 IU/mL). The ice cube test for verification revealed a negative result. In therapy, single dose desloratadine as 5 mg q.d. was used for one week, but the dose was increased to 5 mg b.i.d. in the second week and 5 mg t.i.d. in the third week by necessity. During one month follow-up with the triple dose, the attacks were controlled to a great extent, and the patient was recommended to keep the pertinent dose until the spring.

## Discussions

Although different phenotypes of patients with ACU were identified, data about particular phenotypes and prognostic variables of the disease in large series of patients are scarce. The diagnosis is typically made in patients with a clinical history of wheals and/or angioedema after cold exposure [1]. There can be a considerable heterogeneity in clinical presentations,

ranging from mild localized whealing to cold-induced anaphylaxis [5]. The severity of disease was proposed as type-1, localized urticaria or angioedema; type-2, generalized urticaria or angioedema without systemic symptoms; and type-3, severe systemic reactions with hypotensive or respiratory symptoms [1,6]. Our case can be considered as type-1 given the strict delineation of the lesions by the limits of clothing and lack of progression to generalized urticaria. It was reported that lower antihistamine doses were needed to control the disease in milder ACU cases compared to more severe ones [1]. This case, although it was in type-1 severity, the needed antihistamine dose to suppress the manifestations was in the range of type-3 severity according to above mentioned severity scale. The tendency to increase in clinical manifestations during the course of the disease and the relative higher doses needed to suppress the symptoms should alert the physician for a probable generalized reaction. The primary factors influencing systemic reactions appear to be the exposed skin's surface area, outside temperature, and duration of exposure [5]. Considering the probable role of widespread clothing style in controlling the symptoms, the recent increase in severity of manifestations, and relative high dose of antihistamine to suppress the relevant clinical symptoms, the patient was warned against more severe forms of the disease in the future. In different studies the rates of atypical cases were differentiated from 15.5% to 28.4% [1,4,6]. This case as an atypical cold urticaria also displays an unusual phenotype, only erythema without wheal or angioedema, strictly limited to exposed areas. The case is interesting to show how the

cold sensitivity of the skin can be so precise, displaying such sharp borders.

## Conclusions

There can be a considerable heterogeneity in clinical presentations of ACU, ranging from mild localized whealing to cold-induced anaphylaxis. Our case, although it was in type-1 severity clinically, the needed antihistamine dose to sup-

press the manifestations was in the range of type-3 severity. The tendency to increase in clinical manifestations during the course of the disease and the relative higher doses needed to suppress the symptoms should alert the physician for a probable generalized reaction. This case as an atypical cold urticaria also displays an unusual phenotype, only erythema without wheal or angioedema, strictly limited to exposed areas.

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*The text is neither under consideration by any other journal, nor published previously.*

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