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SHORT COMMUNICATION

Familial Atypical Cold Urticaria Localized on the Face: A Case Report

Laura Huilaja[1], Riitta Riekki[1], Pekka T. Leinonen[1–3], Aarne Oikarinen[1] and Kaisa Tasanen[1]

[1]Department of Dermatology, Oulu University Hospital and University of Oulu, Aapistie 5A, FIN-90220 Oulu, and [2]Department of Anatomy and Cell Biology, University of Oulu, Oulu, and [3]Department of Dermatology, Kainuu Central Hospital, Kajaani, Finland. E-mail: laura.huilaja@oulu.fi

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Cold urticarias (CUs) are a heterogeneous group of diseases characterized by rapid appearance of itching wheals, and occasionally angioedema after cold stimulation. Diagnosis of CU is based on a suggestive history, excluding other (physical) urticarias and a positive cold stimulation test result. Most often wheal-and-flare type reactions represent acquired CU (ACU), which typically affects young adults (1). ACU is further subdivided into primary and secondary forms (typical ACU); rare forms of atypical ACU in which cold stimulation test (CST) is negative have also been reported (2). The pathomechanisms of sporadic CU remain mostly unknown (1). In addition, very rare hereditary types, i.e. delayed CU, familial cold auto-inflammatory syndrome (2) and, most recently, familial atypical cold urticaria (FACU) (3), are described. With informed consent, we present here a case of FACU in a child.

CASE REPORT

A 6-year-old girl was referred to us with cold-induced symptoms. She had previously been diagnosed with atopic eczema. Family history was positive for autoimmune diseases (multiple sclerosis and rheumatoid arthritis in grandparents). Since the first months of her life she had presented paling and induration on her face when exposed to atmospheric cold and wind year-round. Typically, her facial skin pales for 10–30 min after cold exposure and she experiences urticarial erythema on her face when warming up again (Fig. 1). No concomitant systemic symptoms or generalized erythema has occurred, even after swimming in cool lake water. The duration of her symptoms is 1–2 h. Her father and parental grandmother reported similar facial symptoms lifelong. Her brother, who is 3 years younger, has also had similar symptoms on his face.

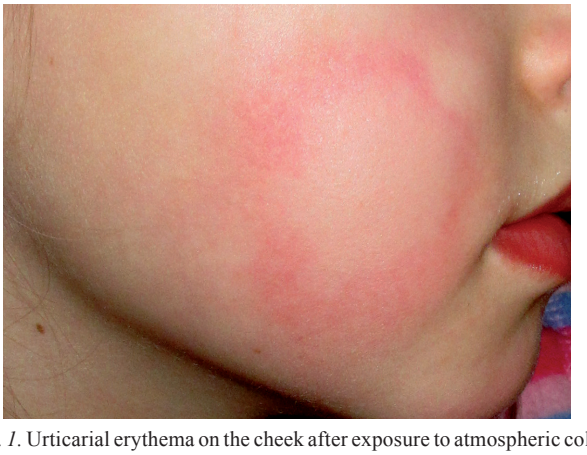

Fig. 1. Urticarial erythema on the cheek after exposure to atmospheric cold.

On clinical examination she had atopic eczema only on her wrists. A CST with an ice cube for 20 min was performed, and it remained negative. Based on family history and her symptoms, a diagnosis of FACU was made. She was advised to protect herself from strong cold wind and massive cold exposure. In addition, desloratadine was prescribed and the use of a symptom diary was recommended to monitor its efficiency. After 2 years of follow-up, she still has symptoms only on her face. The use of desloratadine has been irregular, with an unclear effect on her symptoms.

DISCUSSION

ACU is one of the most common urticarias, and it typically affects young adults (1), but in paediatric patients the mean onset is at around 7 years of age (4). Typically, ACU affects any part of the body exposed to cold, although localized CU in a paediatric patient has been described (5). Our patient has had facial symptoms since infantile age, which suggested familial type of CU. In addition, she has a clear family history of CU. However, her symptoms appeared shortly after cold exposure, distinguishing her disease from delayed familiar CU, in which wheals arise within 9–18 h after exposure (6). She had neither fever nor conjunctivitis nor arthralgias typical to autosomal dominantly inherited familial cold autoinflammatory syndrome (7).

Autosomal dominantly inherited FACU is a recently described entity of CU (3). Lifelong symptoms, such as urticarial erythema and sometimes angioedema after cold exposure, occur early in life. Atmospheric cold is a typical trigger, but handling cold objects or intake of cold food or beverages can also bring on symptoms.

CSTs with ice cubes are negative in patients with FACU (3). In our case, too, it seems that cold alone does not provoke the patient's symptoms, but only when associated with wind. This was also observed by Gandhi and co-workers in evaporative cooling tests. In addition, exposure to humid and cool air was most often reported as a cause of FACU symptoms by Gandhi et al. (3). Patients with FACU presented erythema and urticarial lesions only in areas exposed to cold in outdoor cold challenging tests, and no systemic reactions or spreading of symptoms were seen. Surprisingly, our patient has had symptoms solely on her face, even though other parts of her body have been exposed to atmospheric cold or even to cold water.

In patients with FACU, drying and re-warming the skin is most important when controlling the symptoms. Daily anti-histamines have been reported to decrease

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symptom severity in patients with FACU, similarly to ACU (3). Since our patient has used desloratadine irregularly, we cannot report on its effect.

REFERENCES

1. Siebenhaar F, Weller K, Mlynek A, Magerl M, Altrichter S, Vieira Dos Santos R, et al. Acquired cold urticaria: clinical picture and update on diagnosis and treatment. Clin Exp Dermatol 2007; 32: 241–245.
2. Wanderer AA, Hoffman HM. The spectrum of acquired and familial cold-induced urticaria/urticaria-like syndromes. Immunol Allergy Clin North Am 2004; 24: 259–286.
3. Gandhi C, Healy C, Wanderer AA, Hoffman HM. Familial atypical cold urticaria: description of a new hereditary disease. J Allergy Clin Immunol 2009; 124: 1245–1250.
4. Alangari AA, Twarog FJ, Shih MC, Schneider LC. Clinical features and anaphylaxis in children with cold urticaria. Pediatrics 2004; 113: e313–317.
5. Sciallis GF 2nd, Krych EH. Localized cold urticaria to the face in a pediatric patient: a case report and literature review. Pediatr Dermatol 2010; 27: 266–269.
6. Soter NA, Joshi NP, Twarog FJ, Zeiger RS, Rothman PM, Colten HR. Delayed cold-induced urticaria: a dominantly inherited disorder. J Allergy Clin Immunol 1977; 59: 294–297.
7. Hoffman HM, Wanderer AA, Broide DH. Familial cold autoinflammatory syndrome: phenotype and genotype of an autosomal dominant periodic fever. J Allergy Clin Immunol 2001; 108: 615–620.