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A Case Report On Aquagenic Urticaria

Ghizlane Jaabouti [a*], Sara Benchidmi [b], Naima El Hafidi [a], Soumia Bencchakroun [a] and Chafiq Mahraoui [a]

[a] Division of Pediatric Immuno Allergology and Infectious Diseases, Ibn Sina University Hospital, Rabat, Morocco.
[b] Children University Hospital, Ibn Sina University Hospital, Rabat, Morocco.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Aquagenic urticaria (AU) is a rare form of physical urticaria that is triggered by exposure to water. Despite its rarity, it can significantly impact patients' quality of life. The underlying pathophysiology of AU is not well-understood, but diagnosis is typically based on clinical history and confirmed by skin tests. Unfortunately, avoidance of the trigger is often impossible, making symptom control the primary approach to treatment.

The lack of awareness and understanding of AU can make diagnosis and management challenging. Therefore, this case report highlights the clinical and etiological features of AU and emphasizes the need for improved management strategies.

The patient in this case is a 15-year-old with a well-controlled asthma background treatment of fluticasone. Since age 7, the patient has experienced urticaria when exposed to both sea and fresh water. A diagnosis of aquagenic urticaria was confirmed through skin tests and patient history. Symptomatic treatment with antihistamines and barrier creams before swimming has been effective, with a limitation on the time spent in the water to manage symptom.

Keywords: Aquagenic urticaria; physical urticaria; fresh water; sea water; antihistamine.

**Corresponding author: E-mail: ghiz.jaabouti@gmail.com;*

1. INTRODUCTION

Chronic urticaria (CU) is a common condition in dermatology, and its chronic form is defined by the presence of urticaria and/or angioedema for more than 6 weeks. There are two categories of CU, spontaneous and inducible, with the latter being characterized by lesions appearing under specific conditions, such as exposure to cold, heat, sun, or water.

Aquagenic urticaria (AU) is a rare form of chronic inducible urticaria, where erythematous and pruritic plaques appear within minutes of contact with water, regardless of temperature or salinity. The exact pathophysiology of AU remains unclear, and while it may be classified as an allergy, it does not involve histamine release as in other forms of urticaria. It is important to note that AU should not be confused with aquagenic pruritus, which presents with no visible skin lesions. Raising awareness of the clinical and etiological characteristics of AU can help improve its diagnosis and management
[1].

2. PRESENTATION OF CASE

The patient is a 15-year-old child who has been suffering from pruritic red or pinkish patches for the past 8 years. The patches are superficial, rounded, well limited and in relief, and are located on the neck, trunk, back and upper limbs. The eruptions appear a few minutes after the start of his shower or after swimming in the sea or in a pool. These patches are sometimes accompanied by a pruritic edema, and the lesions regress spontaneously 30 to 45 minutes after the end of the contact with water. These eruptions were initially intermittent and then became permanent at puberty.

To support the diagnosis, skin tests were performed on the reported patient. The areas chosen for the application of the compresses corresponded to the location of the lesions described by the patient. We applied simultaneously on each cheek a compress soaked in fresh water (on the left cheek) and seawater (on the right cheek) at 37°C for 30 minutes [Fig. 1]. The temperature of 37°C allows us to avoid a reaction that would be rather related to cold urticaria.

After the 30 minutes, we noted the appearance of edema and erythema at the areas of contact with the compresses, which were more marked on the left cheek where we applied a compress soaked in fresh water. The control test consisted of the application of a compress soaked in fresh water at 37°C on the neck for 30 minutes, and the reaction was similar to that present on the cheeks [Fig. 2].

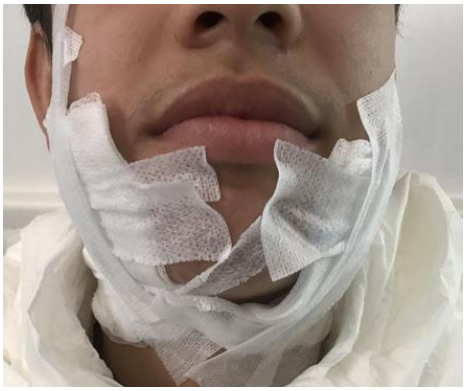

Fig. 1. Skin tests applied simultaneously on each cheek: A compress soaked in fresh water (on the left cheek) and seawater (on the right cheek)

An effort test and an ice cube test were performed and came back negative to eliminate other causes of chronic inducible urticarial [Fig. 3].

We confirmed the diagnosis of aquagenic urticaria based on the patient's clinical history and skin tests. Since it is virtually impossible to avoid the trigger, treatment is based on symptom control. We initially proposed H1 antihistamines during seaside vacations, the application of barrier creams before swimming, and limiting the time spent in the water.

3. DISCUSSION

Chronic urticaria is defined by the daily presence or recurrent flare-ups of superficial urticaria and/or angioedema for a period exceeding 6 weeks [2]. Its prevalence is estimated to be between 0.5 and 5% in the general population [3,4]. It is less frequent (0.1 and 0.3%) in children and adolescents [5].

According to the new 2013 classification, chronic urticaria includes two categories [6-8]: Chronic spontaneous urticaria without the intervention of an external physical stimulus and not reproducible by provocation tests , and inducible urticaria which includes dermographism, vibratory, aquagenic, solar, cholinergic, cold, adrenergic pressure urticaria.

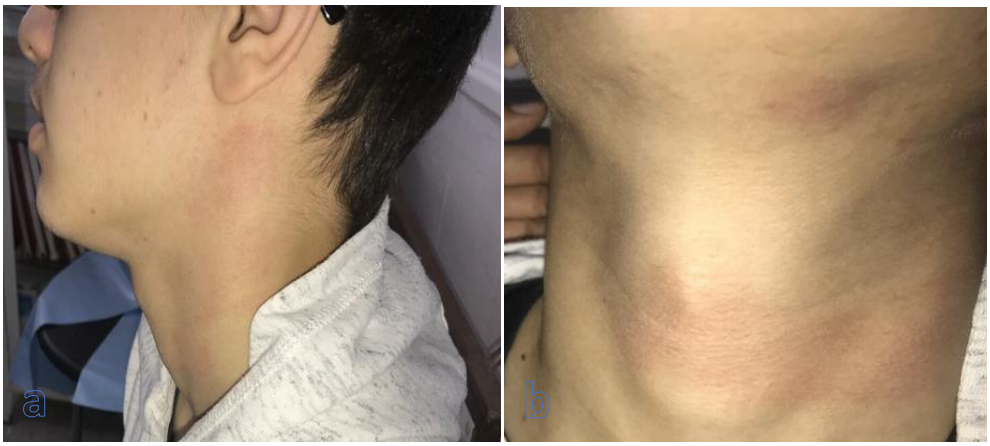

Fig. 2. The appearance of edema and erythema at the areas of contact with the compresses soaked in fresh water at 37◦C

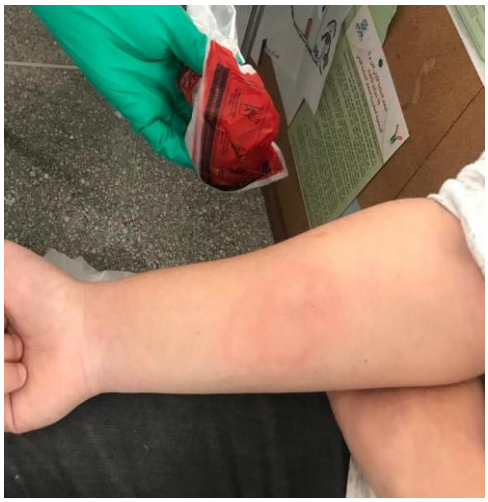

Fig. 3. An ice cube test were performed and came back negative

Aquagenic urticaria is a rare inducible urticaria caused by exposure to water, the salt-dependent form has been described in less than 10 cases [9,10] while temperature and pH have not shown any influence [5]. It was first described by Shelley and Rawnsley [11], and reported three cases in 1964, and since then less than 100 cases have been published in the literature. It is more common in women than men, and it usually first appears during puberty. While most cases of aquagenic urticaria are sporadic, there have been some reported cases of familial aquagenic urticaria.

The exact cause of this condition is not well understood, but there are a few theories that have been proposed. One hypothesis suggests that the interaction of water with sebum (an oily substance produced by the skin) may form a toxic substance that triggers the release of histamine and other chemicals from mast cells in the skin [12]. Another theory proposes that there may be water-soluble epidermal antigens that diffuse into the skin and trigger an immune response [13]. It's also possible that there may be histamine-independent mechanisms involved [14].

In order to diagnose aquagenic urticaria, a water provocation test is typically performed. The test involves applying a 37℃ water compress to the upper body for 30 minutes, while keeping the compress at room temperature to avoid confusion with local cold or heat-induced urticaria. The pretreatment with organic solvents helps to promote the formation of edema and erythema at the areas of contact with the compress, as a result of the enhanced ability of water to penetrate the stratum corneum. Other physical urticarias, such as cholinergic, cold, or heat urticaria, can be excluded by performing stress and ice cube tests.

Antihistamines are the first-line treatment for aquagenic urticaria, but their efficacy varies among patients. In some cases, antihistamines can provide complete control of symptoms, while in other cases, they may not be effective enough to control the symptoms adequately. Therefore, the treatment should be individualized based on the patient's response to therapy. If antihistamines are not effective, other treatments, such as ultraviolet radiation or barrier methods, may be considered [15,16].

Refractory cases of aquagenic urticaria have been treated with a variety of methods, including

ultraviolet (UV) radiation therapy and barrier methods such as the application of emulsion creams to the skin for protection against water. UV radiation therapy involves exposure to UVB or UVA wavelengths, which may help to thicken the epidermis and decrease water penetration, resulting in a reduction of symptoms. However, UV therapy can have potential side effects such as skin damage and increased risk of skin cancer, so it is not recommended as a first-line treatment option. Barrier methods such as the application of emulsion creams can help to reduce water penetration and alleviate symptoms, but may be inconvenient and can cause skin irritation in some cases. It's important to consult with a dermatologist to determine the best treatment options for individual cases of aquagenic urticaria [17].

4. CONCLUSION

The diagnosis of aquagenic urticaria is based on the clinical history and confirmed by a positive water challenge test result. It is important to exclude other types of physical urticaria before making a diagnosis of aquagenic urticaria. Treatment of aquagenic urticaria involves the use of second-generation H1 antihistamines, such as cetirizine or fexofenadine, along with topical emollient cream and phototherapy. In refractory cases, other treatments such as omalizumab, cyclosporine, or mycophenolate mofetil may be considered.

CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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