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Anais Brasileiros de Dermatologia

version ISSN 0365-0590

An. Bras. Dermatol. vol.84 no.6 Rio de Janeiro Nov./Dec. 2009

doi: 10.1590/S0365-05962009000600016

CASE REPORT

Delayed pressure urticaria with systemic manifestations - Case report*

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ABSTRACT

Delayed Pressure Urticaria is considered a rare disease, with clinical diagnosis different from classical urticaria, with possible systemic manifestations. Therefore, it is frequently underdiagnosed, even by specialists. In this article, the case of a patient with a typical history of pressure-induced lesions is presented. Because the patient had fever and leukocytosis, she was admitted to a hospital for investigation of infection.

Keywords: classification; diagnosis differential; urticaria

INTRODUCTION

Delayed pressure urticaria (DPU) is a relatively rare reaction to physical stimulus and was first described by Urbach, in 1949.² The DPU is clinically characterized by deep, tender and erythematous edema from 30 minutes to 9 hours after static mechanical pressure on the skin,^{1,3,4} and lasting from 12 (twelve) to 72 (seventy two) hours.²

The symptoms last, in average, three years, but the natural course of the disease varies and may persist for many years, with periods of remission and flares.^{2,4-6}

In the isolated form, it accounts for less than 2% of all urticarias. Among patients with chronic idiopathic urticaria, 37% present DPU, whereas over 90% of patients with DPU have associated chronic idiopathic urticaria or angioedema.^{4,6,7} This condition may not be very rare, but little known and underdiagnosed, or even misunderstood with other diseases of systemic nature.⁶

The DPU represents one of the types of urticaria most difficult to control. The primary management is to orient patients to avoid situations that trigger the lesions.⁴ Drug therapy is generally unsatisfactory and includes antihistamines, non-steroidal anti-inflammatory drugs (NSAIDS), topical or systemic corticosteroids, sulphasalazine and dapsone.⁵ The use of H1antihistamines is not very effective but help patients with associated chronic idiopathic urticaria (CIU), giving relief to the most superficial lesions.^{2,8} In some cases the use of the NSAIDS may be useful; however the therapeutic effects have not been established yet and the coexisting CIU may flare up. Good results are achieved in the severe forms of DPU with systemic steroids, but the adverse effects of chronic use of this drug class contraindicate its routine utilization.^{2,3} Topical corticosteroids may be useful when there is edema in selected areas, such as hands or feet.⁹ Sulphasalazine is an alternative for patients with DPU not well controlled with the standard treatment and could be useful to spare corticosteroids.^{1,10} Dapsone has also been used as an alternative drug in severe cases.¹¹

CASE REPORT

A 43-year-old, Caucasian, female, married patient, housewife, presented disseminated painful and pruriginous erythematous and papular lesions, that had appeared four times per week, for approximately two months. She also had fever and arthralgia, mainly in the joints of the hands. She went to emergency services several times and was given antihistamines, antipyretics and analgesic agents with partial relief. In another visit to the emergency department she had a complete blood count due to fever and exacerbation of symptoms, which showed significant leukocytosis (26,700 cells/mm³), with shift to the left (12% of band cells). The patient was admitted to hospital to investigate any systemic infectious disease.

During hospitalization she was submitted to laboratory and imaging tests, which identified no source of infection. Since no cause was found for leukocytosis, one week

after admission the patient was evaluated by one member of our team who made the presumptive diagnosis of DPU and suggested initiating on promethazine 25 mg/day. She progressed with relief of symptoms, and was discharged three days later. She was referred to the Department of Clinical Immunology at the *Hospital Clementino Fraga Filho* - UFRJ (HUCFF-UFRJ) to investigate chronic urticaria (CU).

After evaluation at the Outpatient's Clinic - Chronic Urticaria Program, she underwent the following laboratory tests: complete blood count, erythrocyte sedimentation rate (ESR), ultra sensitive C-reactive protein (CRP), thyroid-stimulating hormone (TSH), free thyroxine (T4), antithyroperoxidase, antithyroglobulin, urine analysis and parasitologic examination of stools. Moreover, the patient was submitted to challenging tests to diagnose physical urticarias and autologous serum skin test.

The tests showed leukocytosis (15,500 cells/mm³) and elevated CRP (15.7 m/L). The Warin test ([Figure 1](#)) was positive, with edema, erythema and pain on the application site four hours after challenge ([Figure 2](#)).

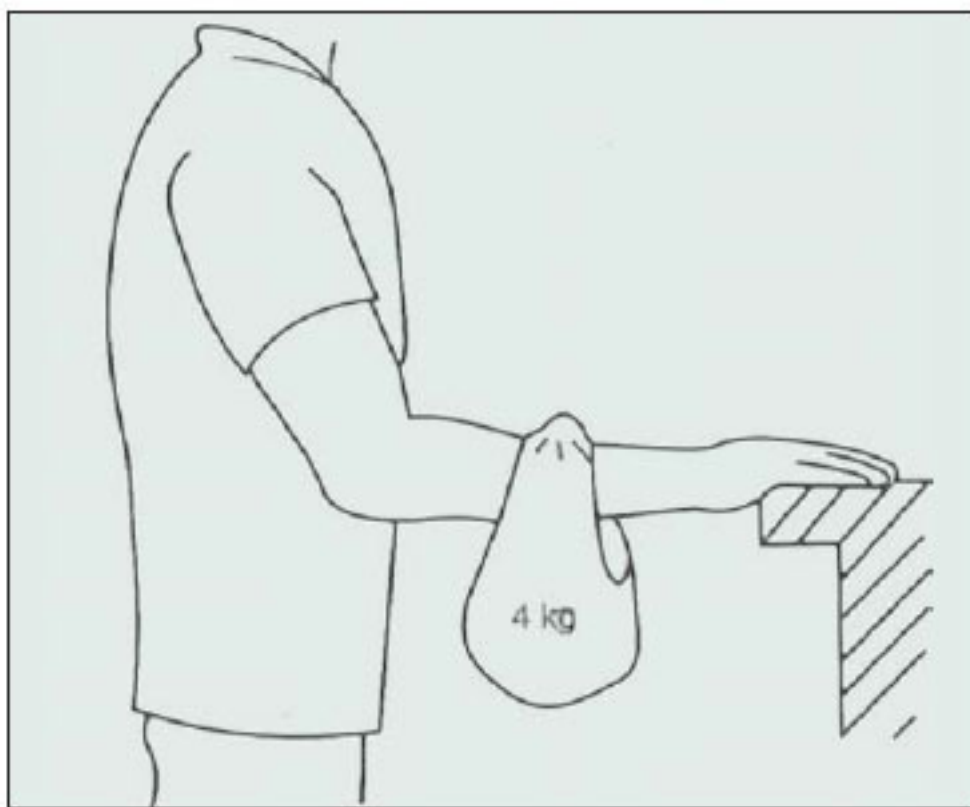


Figure 1: Warin technique



Figure 2: Erythematous and edematous lesion on the extensor region of the forearm, four hours after performing the Warin technique.

The patient was oriented to avoid situations that could trigger the DPU. Hydroxyzine 50 mg/day was prescribed to treat chronic idiopathic urticaria, in addition to amitriptyline 12.5 mg/day and chlordiazepoxide 5 mg/day, and she evolved with improvement of symptoms.

DISCUSSION

The DPU is one type of physical urticaria with distinct characteristics. An important finding is absent or reduced pruritus in the lesions. Local pain and burning sensation are more often observed.^{2,4,12} When the edema involves joints and muscles, the complaint of pain is particularly relevant.¹³ It affects more male individuals and is more often between the third and fifth decades of life.^{3,13}

It is clinically characterized by onset of a deep, tender and erythematous edema, initiating from 30 minutes to 9 hours after static pressure on the skin, and lasting for 12 to 72 hours.^{4,5,13} Pruritus is virtually absent.⁴

The lesions may be localized, affecting any region of the body. The most frequent sites are palmar and plantar regions, lips, shoulders and arms, since they are more submitted to pressure, besides the buttocks after being seated for a prolonged period.

When patients do not associate onset of edematous lesions to localized pressure, it may be erroneously diagnosed as angioedema.⁶

Approximately 50% of patients with DPU present extracutaneous manifestations,¹⁴ such as the case described above. The DPU may be associated to fever, headache, chills, malaise, asthenia, dyspnea and arthralgia, mimicking a viral condition; these symptoms coincide with the peak of edema and are more intense when there are several lesions.^{2,4,6,12,14} Patients may present leukocytosis (20% to 50% of cases) and increased erythrocyte sedimentation rate (in up to 70% of patients).^{2,3,4,12,14}

The pathophysiology of the DPU has not been explained yet, and probably there is more than one factor involved.

Kinins are inflammatory mediators that cause more pain than pruritus. Therefore, they have been considered in the pathogenesis of this type of urticaria, since the lesions are usually associated to local burning or pain, the onset is delayed and there is leukocytosis.²

Kininases inactivate kinins and it has already been demonstrated that a low pH inhibits the activity of these enzymes. Hence, it might be assumed that the drop in local blood flow, generated by local pressure, would reduce the pH of the skin on that site leading to increased activity of kinins.²

It is speculated that interleukin 1 (IL-1) is released on the skin exposed to pressure, and it would be responsible for fever, malaise, myalgia, arthralgia and leukocytosis.¹³

Upon histological examination, the lesions of DPU show eosinophilic infiltrate, edema and deposition of fibrin. According to some studies, these histological findings characterize a reaction of the late cutaneous phase, demonstrating a predominance of neutrophils and eosinophils in a large amount; however, in the initial process, there is massive presence of lymphocytes and eosinophils.¹⁴

Due to this diversity of signs and symptoms, the DPU is frequently underdiagnosed, even by specialists.

The diagnosis is made based on a detailed history, since most patients do not establish a causal relation between the stimulus applied by pressure and the onset of urticaria some hours later. Diagnosis is confirmed by means of a dermatographometer with 100 g/mm², placed perpendicularly on the dorsum of the patient, for 70 seconds, or using the Warin technique (Figure 1), in which the DPU lesion can be reproduced by applying a pressure equivalent to 4 kilograms, for 5 minutes on the skin of the upper external third of the forearm. The reading of the test is performed 4 to 6 hours later, when the edema is usually at most intensity.^{3,6,15}

Since the condition presents peculiar characteristics, the tests for DPU must be conducted in all patients with chronic urticaria, mainly if it associated to systemic signs and symptoms.

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Funding: None.

Conflict of interest: None.

* Setting: Discipline of Immunology, Faculdade de Medicina da Universidade Federal do Rio de Janeiro (UFRJ) and Department of Immunology, Hospital Universitário Clementino Fraga Filho (UFRJ) – Rio de Janeiro (RJ), Brazil.



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