RESEARCH PAPER

Urticarial Vasculitis Presenting as Annular Polycyclic Purpuric Plaques Over the Thighs: A Case Report

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ABSTRACT

Background: Urticarial vasculitis (UV) is an uncommon form of cutaneous small vessel vasculitis characterized by persistent urticarial lesions with histopathological evidence of leukocytoclastic vasculitis. Lesions, in contrast to ordinary urticaria, usually remain longer than 24 hours and may go away with purpura or residual pigmentation. Diagnostic difficulties may arise from atypical morphological appearances.

Case Presentation: We report a case of a middle-aged patient presenting with multiple erythematous to violaceous lesions over both thighs of one-week duration. Examination of the skin showed bilaterally symmetrical annular and polycyclic plaques with patches of non-blanchable purpura, center clearing, and serpiginous borders. There was no noticeable pruritus, but the lesions were persistent, slightly sensitive, and accompanied by a burning sensation. Systemic symptoms were absent. Complement levels and autoimmune markers, among other laboratory tests, were within normal ranges. A diagnosis of normocomplementemic urticarial vasculitis was determined based on clinical characteristics.

Discussion: The peculiar appearance of the annular polycyclic purpuric plaques in this case is noteworthy since they might resemble other dermatological disorders including annular erythema and erythema multiforme. The duration of lesions and the existence of purpura were important indicators of vasculitis.

Conclusion: Accurate diagnosis and adequate assessment of urticarial vasculitis depend on the recognition of unusual appearances. In order to prevent misdiagnosis and guarantee prompt treatment, clinicians should take UV into consideration when dealing with chronic annular purpuric lesions.

Keywords: Urticarial vasculitis, annular plaques, polycyclic lesions, purpura, cutaneous vasculitis.

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INTRODUCTION

Urticarial vasculitis (UV) is an uncommon clinicopathological entity characterized by the presence of urticarial lesions with histological evidence of leukocytoclastic vasculitis involving small vessels [1]. The illness, which was first identified by McDuffie et al. in 1973, is a unique subtype of cutaneous vasculitis in which vascular inflammation and complement activation are caused by immune complex deposition [1,2]. In contrast to conventional urticaria, UV lesions usually last longer than 24 hours and are commonly accompanied by burning or pain instead of pruritus, and often goes away with purpura or persistent hyperpigmentation [3,4].

From limited epidermal involvement to systemic illness involving organs such the kidneys, lungs, joints, and gastrointestinal tract, UV exhibits a broad range of clinical manifestations [5]. UV is generally divided into two categories based on complement levels: hypocomplementemic urticarial vasculitis (HUV), which is more commonly linked to systemic symptoms and autoimmune diseases like systemic lupus erythematosus, and normocomplementemic urticarial vasculitis (NUV), which typically has a benign course [6,7]. Lesions in UV may have a wide range of morphologies. Although the most frequent appearance is erythematous wheals that resemble chronic urticaria, there have been reports of atypical forms such as purpuric plaques, annular lesions, and polycyclic

patterns [4,8]. It might be difficult to diagnose such unusual presentations since they can resemble other dermatological disorders including erythema multiforme, cutaneous lupus erythematosus, or other types of small vessel vasculitis [5,8].

It's critical to identify these peculiar morphological variations since delayed or inaccurate diagnosis might result in insufficient testing for systemic involvement. Here, we describe an uncommon case of urticarial vasculitis that presents as annular polycyclic purpuric plaques confined across the thighs, emphasizing the need of clinicopathological association in making a precise diagnosis.

CASE PRESENTATION

A middle-aged patient presented to the dermatology outpatient department with complaints of multiple reddish lesions over both thighs for the past 5–7 days. The lesions began subtly and grew in size and quantity over time. There was no recent drug use, infection, systemic disease, or history of such events. Multiple annular and polycyclic erythematous to violaceous plaques were symmetrically distributed throughout the anterior and medial regions of both thighs, according to a cutaneous examination. The lesions had sections of core clearing with peripheral expansion, creating a remarkable gyrate pattern, and were well-defined with uneven, serpiginous margins. A non-blanchable purpuric component was seen in a number of lesions, which may indicate underlying vascular involvement. The diameter of the plaques varied from 2 to 10 cm, and in certain places, coalescence formed bigger polycyclic patterns.

The lesions had no discernible induration and were just slightly painful when palpated. There was no evidence of necrosis, ulceration, vesiculation, or surface scaling. Purpura was further supported by diascopy, which showed partial non-blanchability. The patient experienced little pruritus and a slight burning sensation over the lesions. Crucially, the lesions did not go away after more than a day, setting them apart from typical urticaria. Systemic signs including fever, arthralgia, stomach discomfort, hematuria, or respiratory problems were absent.

The results of routine laboratory testing, such as liver, kidney, and complete blood counts, were within normal ranges. There was a little increase in inflammatory markers. The complement levels (C3, C4) fell within the typical range. The antinuclear antibody component of the autoimmune workup came out negative. Despite being recommended, a skin biopsy was not carried out because of patient limitations. A clinical diagnosis of urticarial vasculitis (likely normocomplementemic type) was determined based on the persistent nature (>24 hours), purpuric shape, annular polycyclic arrangement, and accompanying burning sensation.

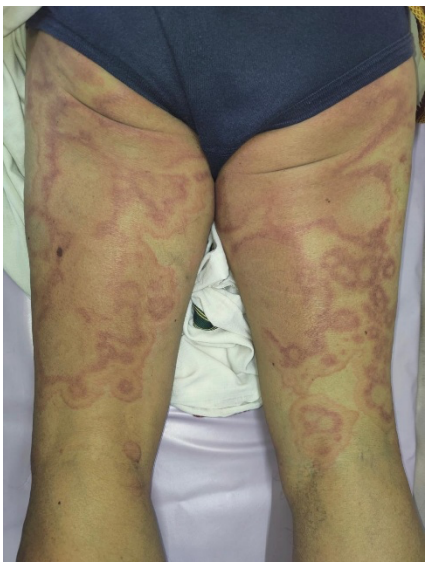

Figure 1: Annular polycyclic purpuric plaques over bilateral thighs

Multiple annular and polycyclic erythematous to violaceous plaques are symmetrically spread throughout the front regions of both thighs in this clinical image. The lesions have a gyrate pattern with irregular serpiginous borders with periods of peripheral expansion and central clearance. A purpuric (non-blanchable) component is present in a number of plaques, which may indicate underlying small vessel vasculitis. The location and form of the atypical annular polycyclic purpuric plaques are consistent with urticarial vasculitis.

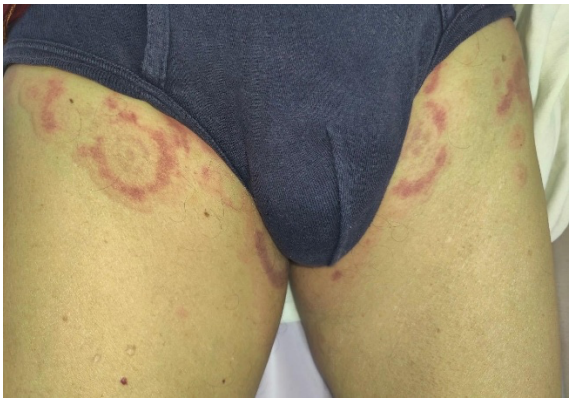

Figure 2: Annular plaques with central clearing over upper thighs

Clinical image displaying annular erythematous to violaceous plaques with distinct boundaries and a central clearing over the upper medial thighs. The

lesions are polycyclic and gyrate in shape, with early purpuric alterations in certain regions that may indicate changing vascular involvement. The diagnosis of urticarial vasculitis with unusual annular presentation is further supported by the distribution and shape.

A skin biopsy was advised; however, it could not be performed due to patient-related constraints. The diagnosis was made based on characteristic clinical features.

Treatment

The patient was started on oral antihistamines (levocetirizine 5 mg once daily) along with a short course of oral corticosteroids (prednisolone 0.5 mg/kg/day). Emollients and other supportive measures were recommended. Although no specific triggering cause was found, the patient was also encouraged to avoid potential triggers.

Outcome and Follow-up

The patient showed significant clinical improvement within 7–10 days, with reduction in erythema and progression of lesions. No new lesions appeared during follow-up. At previously impacted locations, residual post-inflammatory hyperpigmentation was seen. After the corticosteroids were gradually reduced over the course of two weeks, the patient showed no signs of systemic involvement and continued to be asymptomatic at further follow-up appointments.

DISCUSSION

Urticarial vasculitis (UV) is a form of cutaneous small vessel vasculitis characterized by urticarial lesions that histologically demonstrate leukocytoclastic vasculitis [1]. Clinically, it is distinguished from regular urticaria by the lesions' duration for more than 24 hours, the burning or discomfort they cause, and their disappearance with purpura or persistent hyperpigmentation [2, 3]. Atypical annular and polycyclic shape with a significant purpuric component, a rare UV appearance, makes the current example noteworthy. A number of morphological variations, such as annular plaques, retiform purpura, and erythema multiforme-like lesions, have been reported, despite the fact that classical lesions resemble urticaria [4,5]. These differences frequently cause diagnostic confusion with diseases such as erythema multiforme, erythema annulare centrifugum, and other vasculitides [8].

One important characteristic that sets this instance apart is the presence of non-blanchable purpura, which indicates red blood cell extravasation due to vascular inflammation and is not present in basic urticaria [3]. Erythema multiforme was ruled out by the thighs' symmetrical invasion and the lack of mucosal or target lesions. UV is divided into normocomplementemic and hypocomplementemic forms according to complement levels. This patient's normal complement levels and lack of systemic symptoms point to normocomplementemic urticarial vasculitis, which usually has a benign, skin-limited course [6]. However, as hypocomplementemic variations may be linked to autoimmune disorders such systemic lupus erythematosus, assessment for systemic involvement is still crucial [7].

Immune complex deposition in postcapillary venules, complement cascade activation, and neutrophil-mediated vascular injury resulting in enhanced vascular permeability and purpura are all part of the pathophysiology [2,5]. This instance emphasizes how crucial it is to identify unusual UV morphological features, especially annular polycyclic purpuric plaques, in order to guarantee prompt diagnosis and suitable treatment.

CONCLUSION

This case illustrates an unusual presentation of urticarial vasculitis manifesting as annular polycyclic purpuric plaques localized over both thighs, a morphology that can easily mimic other dermatological conditions. Key clinical characteristics that helped distinguish it from simple urticaria and other annular dermatoses were the presence of purpura, the duration of lesions for more than 24 hours, and the accompanying burning sensation. Clinicians must be aware of these unusual patterns since misdiagnosis might postpone proper assessment and treatment. Although this patient's normal complement levels and lack of systemic symptoms point to a cutaneous-limited normocomplementemic variation, close observation is necessary since systemic involvement may occur in some situations. The significance of clinicopathological correlation in suspected instances of urticarial vasculitis is emphasized in this paper. Characteristic clinical signs can direct diagnosis and treatment even in the absence of histological proof. Patient outcomes can be greatly enhanced and problems can be avoided with early detection and adequate treatment. When recurrent annular purpuric lesions appear, dermatologists should be extremely suspicious, particularly if the lesions don't follow the typical urticarial shape.

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