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Case Report

Normocomplementemic urticarial vasculitis simulating subacute cutaneous lupus erythematosus

Anant Kumar Singh[1], Naveen Kumar Kansal[1], Ashok Singh[2], Omna Chawla[3]

Departments of [1]Dermatology and Venereology and [2]Pathology, All India Institute of Medical Sciences, Rishikesh, [3]Department of Physiology, Government Doon Medical College, Dehradun, Uttarakhand, India.

ABSTRACT

Urticarial vasculitis is a cutaneous disease characterized by recurring wheals along with histopathological evidence of leukocytoclastic vasculitis, along with potential systemic complications. Urticarial vasculitis clinically presents with erythematous wheals. The measurement of complement levels is useful in classifying urticarial vasculitis. Here, we describe a rare case of normocomplementemic urticarial vasculitis clinically mimicking subacute cutaneous lupus erythematosus in a young female.

Keywords: Complement, Urticarial vasculitis, Leukocytoclastic vasculitis, Lupus erythematosus, Urticaria

INTRODUCTION

Urticarial vasculitis, a form of vasculitis, is distinguished by inflammation of the small cutaneous blood vessels.[1] The condition is more common in females as compared to males. There are only a few population-based epidemiological studies describing the incidence and prevalence. One study conducted in Sweden estimated an annual incidence of 0.7% with a point prevalence of 9.5/million.[2] The condition may be triggered by or associated with hepatitis, rheumatoid arthritis, lupus erythematosus, particular malignancies, thyroid disorders or medications, e.g., angiotensin-converting enzyme inhibitors, penicillin, and non-steroidal anti-inflammatory drugs. However, in more than half patients, the etiology is unknown.[3,4] Here, we report a case of normocomplementemic urticarial vasculitis with a clinical presentation simulating subacute cutaneous lupus erythematosus.

CASE REPORT

A 22-year-old woman, presented with painful, purplish, itchy rashes for the past 1 year. The lesions used to appear bilaterally on the cheeks, followed by involvement of the trunk and limbs lasting for around 7–10 days, which resolved with hyperpigmentation in the following 3–4 days. The current episode was present for the last 3 days. Complaints of photosensitivity, occasional joint pains, and fever were present. However, no history suggestive of recent infections or medications, weight loss, recurrent oral ulcers, burning micturition, abdominal pain, bleeding diathesis, muscle weakness, etc., was present. Past and family history were non-contributory.

On mucocutaneous examination, multiple erythematous figurate urticarial plaques with patchy brownish hyperpigmented macules (suggestive of post-inflammatory pigmentation) were present on the back and arms bilaterally. Erythematous plaques were also noted on both sides of the malar area and nose, sparing the nasolabial folds [Figure 1a and b]. The plaques were non-blanchable, but tenderness was present. A diffuse decrease in the density of the hair was noted on the vertex without any scarring. Oral and genital mucosa and nails were normal. Considering the history and clinical findings, differential diagnoses of subacute cutaneous lupus erythematosus and urticarial vasculitis were considered. Complete blood counts revealed anemia (hemoglobin, 9.2 g% [normal 13–17 g%), along with a microcytic hypochromic blood picture and raised erythrocyte sedimentation rate (75 mm 1st h [normal 0–20 mm 1st h]). Liver and kidney function tests, urine examination, thyroid function tests, blood glucose levels and glycated hemoglobin, chest X-ray (posteroanterior view), human immunodeficiency virus-1 and 2, hepatitis C virus, and hepatitis B surface

***Corresponding author:** Naveen Kumar Kansal, Department of Dermatology and Venereology, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India. kansalnaveen@gmail.com

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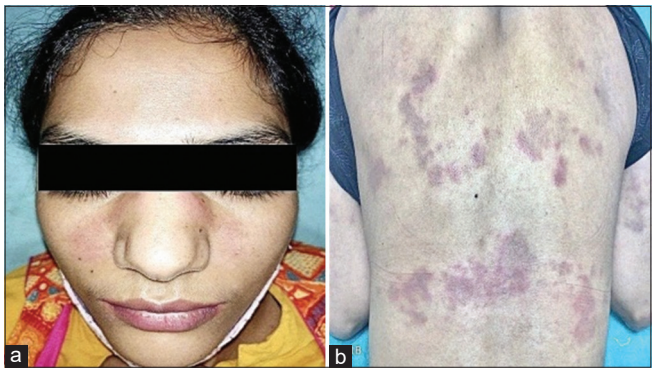

Figure 1: Erythematous urticarial plaques on the face (a) and back with patchy brownish hyperpigmented macule (b).

antigen serology were normal. The 24-h urine protein revealed no proteinuria. The antineutrophil cytoplasmic antibodies were not performed. Antinuclear antibody, anti-double-stranded deoxyribonucleic acid antibody, anticardiolipin antibody, lupus anticoagulant, and anti-Smith antibody were negative. Serum complement measurement revealed normal levels of C3, C4, and C1q. Histopathological examination showed focal exocytosis with spongiosis and mild acanthotic epidermis. Interface dermatitis with periadnexal, perivascular, and perineural moderate-to-dense infiltration of lymphocytes, plasma cells, and neutrophils was present. Foci of small vessel vasculitis with neutrophilic infiltration in the vessel wall were also noted. There was septal inflammation and focal vasculitis in the subcutaneous fat [Figures 2 and 3]. Direct immunofluorescence for immunoglobulin A, immunoglobulin G, C3, and C4 was negative [Figure 4]. Based on the clinical features and pathological findings of vasculitis, we made the diagnosis of normocomplementemic urticarial vasculitis. She was started on oral prednisolone at a dose of 20 mg daily along with oral antihistamines. After getting clearance from ophthalmology, hydroxychloroquine 400 mg/day was added. After 3 weeks of treatment, there was an improvement of around 80% in terms of decreased erythema and there were no new episodes. Later on, she was treated with oral dapsone 50 mg once a day and other medications were gradually stopped. She is on regular follow-up and is in remission [Figure 5].

DISCUSSION

Urticarial vasculitis is a special form of cutaneous vasculitis, in which, in contrast to common urticaria, the lesions last for >24 h. Patients usually experience a burning sensation over these lesions.[5] The lesions may be in the form of red wheals and rarely, may be associated with erythema multiforme, purpura, or occasionally bullous lesions. The lesions usually leave behind residual hyperpigmentation on resolution. The systemic manifestations that may be associated, include angioedema, arthritis or arthralgia,

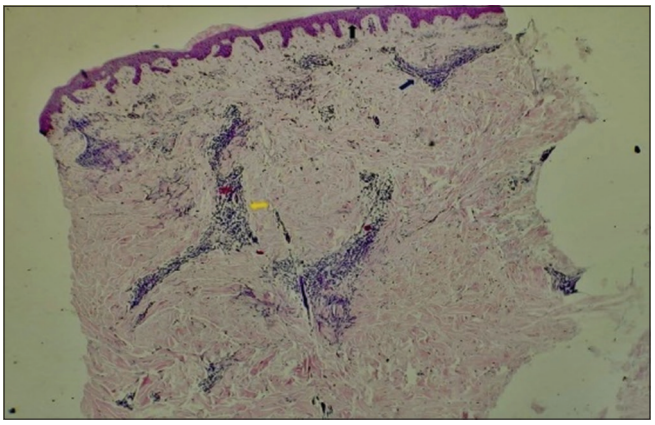

Figure 2: Mild acanthosis (black arrow), interface dermatitis with peri-adnexal (yellow arrow), perivascular (blue arrow) moderate-to-dense infiltration of inflammatory cells (H&E, ×40).

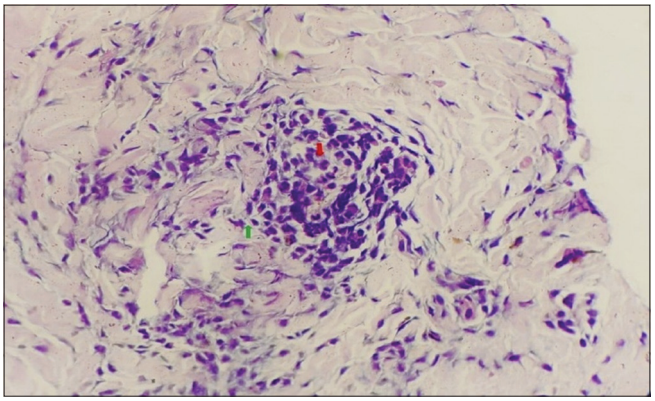

Figure 3: Perivascular (green arrow) and intravascular wall infiltration (red arrow) of lymphocytes, neutrophils and plasma cells in the dermis (H&E, ×100).

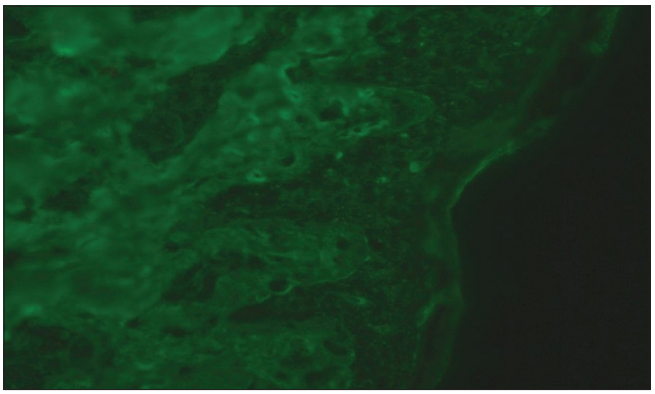

Figure 4: Direct immunofluorescence for immunoglobulin A, immunoglobulin G, C3, and C4 showed no staining.

abdominal pain, fever, pulmonary or renal disease, ophthalmologic involvement (uveitis and/or episcleritis), and Raynaud phenomenon.[6] There are two variants of urticarial vasculitis based on the measurement of the levels

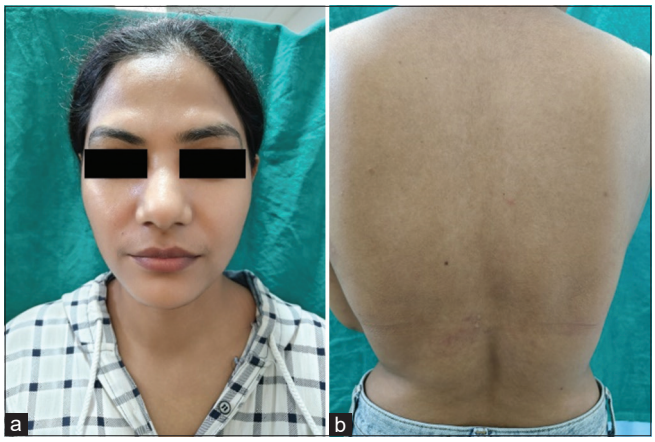

Figure 5: (a and b) Post-treatment images showing the patient in remission.

of complement. Normocomplementemic urticarial vasculitis has normal serum complement levels and is less severe clinically. Hypocomplementemic urticarial vasculitis has low C1q complement levels and elevated anti-C1q antibodies. Hypocomplementemic urticarial vasculitis has a more severe disease course and may be associated with systemic inflammatory diseases.[7,8]

Our patient developed erythematous plaques similar to lesions of subacute cutaneous lupus erythematosus lesions with photosensitivity, occasional joint pain, and fever. Her blood investigations revealed normal levels of complement. The autoimmune profile was negative. Skin biopsy had features of leukocytoclastic vasculitis with negative immunofluorescence findings. Hence, the diagnosis of normocomplementemic urticarial vasculitis was finally made.

The mean duration of urticarial vasculitis has been reported to be about 3–4 years, however, many cases may persist for even decades. Normocomplementemic urticarial vasculitis is associated with a relatively good prognosis. Limited published data indicate that hypocomplementemic urticarial vasculitis also seems to have a good prognosis.[9]

CONCLUSION

Clinically, lesions of urticarial vasculitis may rarely simulate subacute cutaneous lupus erythematosus. As seen in our index case, initial treatment with antimalarials may be indicated. Definitive treatment should be initiated once the patient starts responding.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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