Images in Clinical Rheumatology

Purpuric Component Features to Differentiate Urticarial Vasculitis and Urticaria Without Vasculitis

Características del componente purpúrico de la urticaria con vasculitis y de la urticaria sin vasculitis

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Case Reports

We studied a series of 8 patients (5 men and 3 women) who presented with urticaria-like wheals and purpura that had developed more than 24 h earlier.

Diagnosis and Disease Course

Four patients were diagnosed with urticarial vasculitis (UV), 2 of them with hypocomplementemia, and another 4 patients with urticaria without vasculitis. None of them had extracutaneous involvement. Their protein profiles and immunoglobulin levels were normal and serological tests for human immunodeficiency virus and hepatitis B and C viruses were negative (other parameters are shown in Table 1).

Discussion

Urticarial vasculitis is characterized in clinical terms by urticaria-like wheals and histologically by leukocytoclastic vasculitis (LV). The rash lasts more than 24 h and leaves residual purpura. In clinical practice, it is not uncommon to see urticaria-like conditions that persist over 24 h that are accompanied by a purpuric component. Biopsy only reveals the presence of a superficial perivascular lymphocytic infiltrate. The cause of purpura in the absence of LV is controversial: some authors maintain that the origin is lymphocytic vasculitis and others that it is due to scratching. Lee et al. combine the 2 types of lesions as characteristics of prolonged urticaria with purpura, granting greater importance to the clinical similarities than to the histological resemblance. In our experience, recent UV has a homogeneous erythematous or purpuric color, and leaves purpura in the entire region of the rash on resolution (Fig. 1). The wheals with the lymphocytic infiltrate have areas of an ecchymotic yellowish color within the acute lesions or extending beyond their bounds (Fig. 2). We consider that, when urticarial wheals persist more than 24 h, the residual purpura surrounding the entire area of the lesions supports the diagnosis of UV, whereas a yellowish or ecchymotic discoloration on the periphery or within the wheal corroborates the diagnosis of what Lee et al. classify as prolonged urticaria with purpura in the absence of LV. This could enable the avoidance of biopsy and other studies in single and self-limiting episodes.

Ethical Disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.
Table 1  
Summary of Clinical and Analytical Data.

<table>
<thead>
<tr>
<th></th>
<th>Case 1 (Fig. 1a)</th>
<th>Case 2 (Fig. 1b)</th>
<th>Case 3 (Fig. 1c)</th>
<th>Case 4 (Fig. 1d)</th>
<th>Case 5 (Fig. 2a)</th>
<th>Case 6 (Fig. 2b)</th>
<th>Case 7 (Fig. 2c)</th>
<th>Case 8 (Fig. 2d)</th>
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</thead>
<tbody>
<tr>
<td>Sex/age Comorbidities</td>
<td>M/37 years</td>
<td>F/81 years</td>
<td>F/68 years</td>
<td>F/60 years</td>
<td>M/51 years</td>
<td>M/42 years</td>
<td>M/60 years</td>
<td>M/50 years</td>
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<tr>
<td>Site</td>
<td>Thighs and abdomen</td>
<td>Chest, abdomen and LL</td>
<td>Abdomen</td>
<td>Chest, arms and thighs</td>
<td>Arms, thighs and abdomen</td>
<td>UL, trunk, LL</td>
<td>UL, trunk, LL</td>
<td>UL, trunk, LL</td>
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<tr>
<td>Time since onset</td>
<td>7 days</td>
<td>3 days</td>
<td>12 days</td>
<td>3 days</td>
<td>12 days</td>
<td>3 days</td>
<td>5 days</td>
<td>7 days</td>
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<tr>
<td>Local symptoms</td>
<td>Pruritus</td>
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<td>Pruritus</td>
<td>Pruritus</td>
<td>Pruritus</td>
<td>Pruritus</td>
<td>Pruritus</td>
<td>Pruritus</td>
</tr>
<tr>
<td>Sex/age</td>
<td>Dolor</td>
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<td>—</td>
<td>—</td>
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<td>—</td>
<td>—</td>
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<tr>
<td>General symptoms</td>
<td>—</td>
<td>Fever</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
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<tr>
<td>Treatment</td>
<td>PRED 0.5 mg/kg/day in tapering regimen over 1 month Frequent relapses over a year of follow-up</td>
<td>PRED 0.5 mg/kg/day in tapering regimen over 1 month</td>
<td>PRED 0.5 mg/kg/day in tapering regimen over 1 month</td>
<td>PRED 0.5 mg/kg/day in tapering regimen over 1 month</td>
<td>H2-antihistamines</td>
<td>H2-antihistamines</td>
<td>H2-antihistamines</td>
<td>H2-antihistamines</td>
</tr>
<tr>
<td>Outcome</td>
<td>Lost to follow-up</td>
<td>No relapses over 1 year of follow-up</td>
<td>No relapses over 1 year of follow-up</td>
<td>No relapses over 1 year of follow-up</td>
<td>No relapses over 1 year of follow-up</td>
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ab, antibody; ANA, antinuclear antibodies; ANCA, antineutrophil cytoplasmic antibodies; ELISA, enzyme-linked immunosorbent assay; ENA, extractable nuclear antigens; DIF, direct immunofluorescence; F, female; IIF, direct immunofluorescence LL, lower limbs; m, male; PRED, prednisone; UA, upper airway; UL, upper limbs;
Fig. 1. Urticarial vasculitis. The 4 patients had undergone a biopsy that confirmed the diagnosis of leukocytoclastic vasculitis. Note the presence of acute urticaria-like wheals and lesions in resolution with purpura (a–d).

Fig. 2. Prolonged urticaria without leukocytoclastic vasculitis. The 4 patients had undergone at least one biopsy that revealed the presence of a superficial perivascular lymphocytic inflammatory infiltrate, with or without eosinophils, in the absence of vasculitis. Note the presence of an ecchymotic component within the acute lesions (a and b) and extending beyond the bounds of the wheals (c and d).

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Conflicts of Interest

The authors declare they have no conflicts of interest.

References