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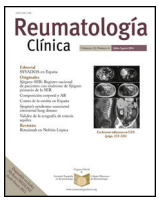

Images in Clinical Rheumatology

Purpuric Component Features that 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.[1] 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[2] and others that it is due to scratching.[3] 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.

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Table 1
Summary of Clinical and Analytical Data.

	Case 1 (Fig. 1a)	Case 2 (Fig. 1b)	Case 3 (Fig. 1c)	Case 4 (Fig. 1d)	Case 5 (Fig. 2a)	Case 6 (Fig. 2b)	Case 7 (Fig. 2c)	Case 8 (Fig. 2d)
Sex/age	M/37 years	F/81 years	F/68 years	F/60 years	M/51 years	M/42 years	M/60 years	M/50 years
Comorbidities	—	Nonallergic bronchial asthma	—	Invasive ductal breast adeno-carcinoma	—	—	—	—
Site	Thighs and abdomen	Chest, abdomen and LL	Abdomen	Chest, arms and thighs	Arms, thighs and abdomen	UL, trunk, LL	UL, trunk, LL	UL, trunk, LL
Time since onset	7 days	3 days	12 days	3 days	12 days	3 days	5 days	7 days
Local symptoms	Pruritus	Pruritus	Pain	Burning sensation	Pruritus	Pruritus	Pruritus	Pruritus
	Dolor	—	—	Pain	—	—	—	—
General symptoms	No	Fever	No	No	No	No	No	No
Histology	Deep and superficial perivascular neutrophilic inflammatory infiltrate. Moderate number of eosinophils. Focal fibrinoid necrosis.	Fibrinoid necrosis, dermal edema, blood extravasation.	Fibrinoid necrosis, dermal edema, blood extravasation.	Superficial perivascular neutrophilic inflammatory infiltrate. Moderate number of eosinophils. Focal fibrinoid necrosis. Nuclear fragments.	Superficial perivascular lymphocytic inflammatory infiltrate with eosinophils, blood extravasation.	Superficial perivascular lymphocytic inflammatory infiltrate, blood extravasation.	Superficial perivascular lymphocytic inflammatory infiltrate with eosinophils, blood extravasation.	Superficial perivascular lymphocytic inflammatory infiltrate, blood extravasation.
	DIF: fibrinogen surrounding the vessels	DIF: negative	DIF: negative	Dermal edema and blood extravasation. DIF: fibrinogen surrounding the vessels	DIF: negative	DIF: negative	DIF: negative	DIF: negative
Analytical study	ANA by IIF/ELISA: negative	ANA by IIF/ELISA: negative	ANA by IIF/ELISA: negative	ANA by IIF: 1:320 homogeneous pattern	ANA by IIF/ELISA: negative	ANA by IIF/ELISA: negative	ANA by IIF/ELISA: negative	ANA by IIF/ELISA: negative
	ANCA: negative	ANCA: negative	ANCA: negative	anti-ENA ab: negative	ANCA: negative	ANCA: negative	ANCA: negative	ANCA: negative
	C3: 50.9 mg/dL (91–190)	C3: normal	C3: normal	ANCA: negative	C3: normal	C3: normal	C3: normal	C3: normal
	C4: 7.1 mg/dL (18–56)	C4: normal	C4: normal	C3: 79.8 mg/dL	C4: normal	C4: normal	C4: normal	C4: normal
	C1q: not done			C4: 15.2 mg/dL C1q: 235 mg/L (100–255)				
Treatment	PRED 0.5 mg/kg/day in tapering regimen over 1 month	PRED 0.5 mg/kg/day in tapering regimen over 1 month	PRED 0.5 mg/kg/day in tapering regimen over 1 month	PRED 0.5 mg/kg/day in tapering regimen over 1 month	H2-antihistamines	H2-antihistamines	H2-antihistamines	H2-antihistamines
Outcome	Frequent relapses over a year of follow-up	Two previous episodes. No relapses over a 1-year period	No relapses over 1 year of follow-up	Lost to follow-up	No relapses over 1 year of follow-up	No relapses over 1 year of follow-up	No relapses over 1 year of follow-up	No relapses over 1 year of follow-up

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;

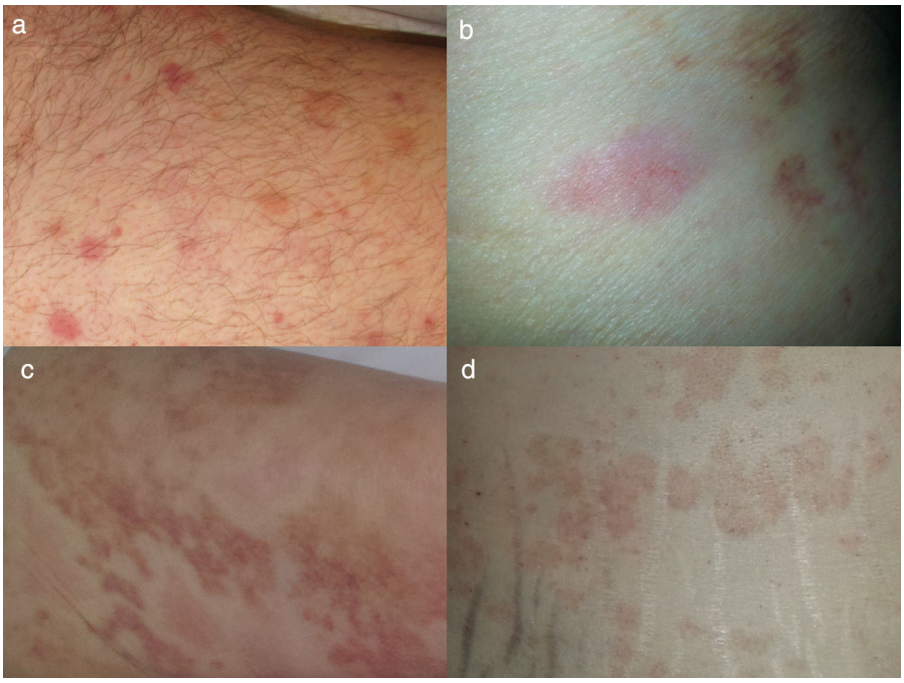

Fig. 1. Urticarial vasculitis. The 4 patients had undergone a biopsy that confirmed the diagnosis of leukocytoclastic vasculitis. Note the presence of acute urticaria-like wheals and lesions in resolution with purpura (a–d).

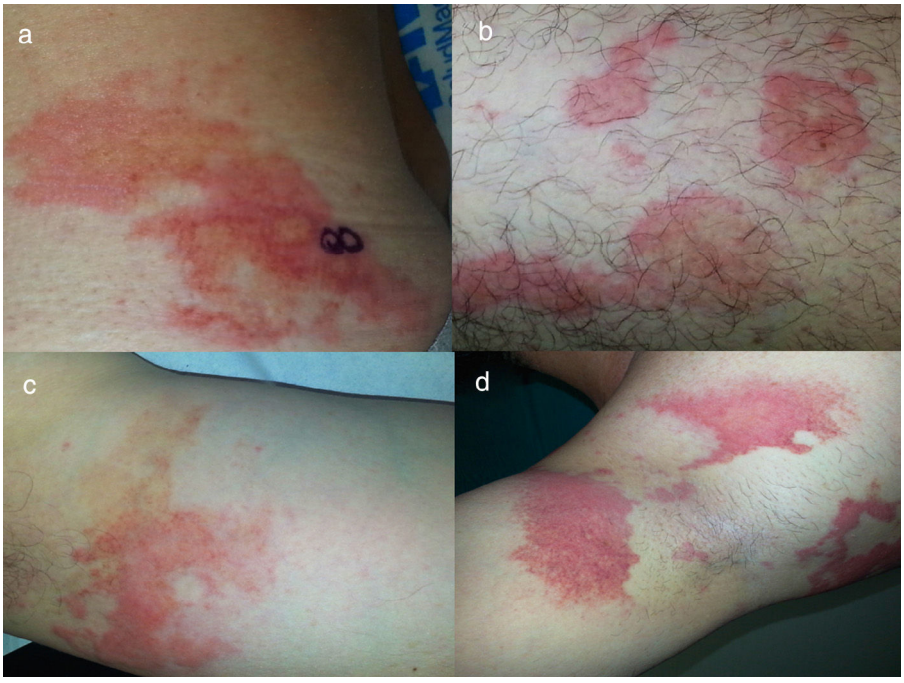

Fig. 2. Prolonged urticaria without leukocytoclastic vasculitis. The 4 patients had undergone at least one biopsy that revealed the presence of a superficial perivascular lymphocytic inflammatory infiltrate, with or without eosinophils, in the absence of vasculitis. Note the presence of an ecchymotic 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.

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