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Recurrent cutaneous eosinophilic vasculitis characterized by annular purpuric lesions: A case report

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Abstract

A 71-year-old woman presented with a persistent, intensely pruritic cutaneous eruption localized on the palmoplantar regions, lips and palate. The histological findings allowed to make the diagnosis of recurrent cutaneous eosinophilic vasculitis, a very rare cutaneous vasculitis characterized clinically by multiple erythematous or purpuric erythematous papules or plaques or angioedema with a relapsing course in the absence of systemic involvement and histologically by a necrotizing vasculitis of the dermal small vessels with a dominant eosinophilic infiltration. The patient was treated with oral methylprednisolone and pentoxifylline which led to a rapid resolution of the cutaneous lesions.

Keywords

Recurrent cutaneous eosinophilic vasculitis, annular, purpura

Introduction

Recurrent cutaneous eosinophilic vasculitis (RCEV) is a very rare cutaneous vasculitis characterized clinically by pruritic purpuric papules and plaques and angioedema without systemic involvement and histologically by a necrotizing vasculitis of the dermal small vessels with a dominant eosinophilic infiltration.[1,2] Here, we present a case of RCEV characterized by purpuric lesions with an annular morphology.

Case report

A 71-year-old Caucasian female patient presented with a 1-month history of a persistent, intensely pruritic eruption initially localized on the palmoplantar regions, lips and palate, which had appeared the day immediately after an episode of left retinal vein occlusion. The patient reported neither fever nor constitutional symptoms; however, slight unintentional weight loss (from 65 to 60 kg) was reported in the preceding months. Her past medical history revealed Parkinson's disease, glaucoma and age-related macular degeneration. There was no personal or family history of cutaneous diseases. Current oral medications included carbidopa/levodopa, clonazepam and vitamin D, while topical ophthalmic medications included brimonidine and tafluprost/timolol eye drops. Clinical examination revealed multiple annular purpuric patches and plaques distributed on the palms, soles, lips and hard palate (Figure 1), while systemic examination revealed no abnormalities.

Laboratory investigations revealed eosinophilia (2.0×10^9/L; normal range (NR) < 0.5), mild megaloblastic anemia (hemoglobin 11.8 g/dL, mean corpuscular volume 96 fL; NR 13.8–17.2 and 80–94) due to folate deficiency, mild hyperhomocysteinemia (26 μmol/L; NR 4–14) and mild hyperglycemia (103 mg/dL; NR 70–99). Autoimmune studies showed weak positive antinuclear antibodies (ANA; 1:160), while the other autoimmune markers, including anti-double-stranded DNA (dsDNA) antibodies, extractable nuclear antigen panel and anti-neutrophil cytoplasmic antibodies were negative. Renal and liver function tests, lipid panel, complement levels, erythrocyte sedimentation rate, C-reactive protein, protein electrophoresis, electrolytes and urinalysis were

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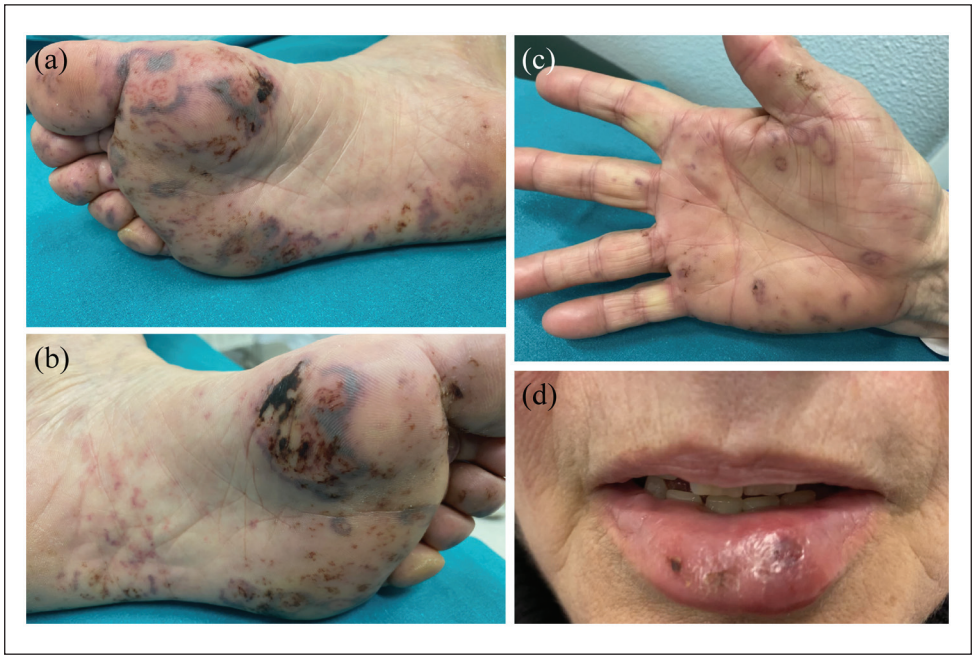

Figure 1. Multiple purpuric patches and plaques with an annular morphology located on soles (a, b), palms (c), and lips (d).

all within normal limits, and coagulation studies—including prothrombin time (PT), activated partial thromboplastin time (aPTT), fibrinogen, protein S and protein C—were also within the NR. Of note, heterozygous methylenetetrahydrofolate reductase (MTHFR) gene mutation was found, while neither coagulation factor mutations (factor V Leiden and factor II) nor antiphospholipid antibodies (anti-cardiolipin antibodies, lupus anticoagulant, and anti-β2 glycoprotein I) were identified.

A 6-mm skin punch biopsy of a right sole lesion was performed, and histology showed, in the epidermis, exocytosis and spongiosis with microvesicle formation. In the superficial and deep dermis, there was a dense infiltrate composed of numerous eosinophils admixed with lymphocytes and histiocytes (Figure 2). The infiltrate exhibited a perivascular and, to a lesser extent, interstitial distribution in the dermis and also involved the subcutaneous tissue where it surrounded the adipose lobules. Dermal and subcutaneous blood vessels exhibited thickened walls infiltrated by eosinophils and endothelial hypertrophy with consequent vascular lumen obliteration, though in the absence of fibrinoid necrosis of the vessel walls (Figure 2). There were extravasated red blood cells in the papillary dermis and focally in the epidermis, and few intravascular neutrophils in the dermis (Figure 2).

The histological findings allowed to make the diagnosis of RCEV. In order to rule out systemic involvement, an 18F-fluorodeoxyglucose positron emission tomography–computed tomography (18F-FDG PET-CT) was performed, which showed increased tracer uptake in the right nasopharynx and lateral cervical lymph nodes bilaterally. These findings were attributed to an upper respiratory tract infection, while the absence of other areas of hypermetabolism excluded systemic involvement of the cutaneous vasculitis. The patient was treated with methylprednisolone at an initial dose of 16 mg/day and pentoxifylline 400 mg/day orally which led to a rapid resolution of the cutaneous lesions. After a complete clinical response was achieved, the steroid dose was tapered to 4 mg/day, which the patient is currently receiving, and no flare-ups have been observed.

Discussion

RCEV is a very rare entity characterized clinically by multiple erythematous or purpuric erythematous papules or plaques or angioedema with a relapsing course and in the absence of systemic involvement.[2] Histology classically reveals a necrotizing vasculitis with predominant eosinophilic infiltration, without granulomas or leukocytoclasia.[2] Although its pathogenesis is still unclear, an abnormal eosinophil-driven response against a still-unknown antigen has been hypothesized to play a causative role.[3] The disease responds promptly to systemic corticosteroids, but long-term steroid treatment is often needed to prevent flare-ups.[2,3]

The clinical presentation of this case was consistent with the diagnosis of RCEV. Of note, annular configurations similar to that of this case have been reported in other cases of RCEV.[1,4] In the present case, peripheral blood eosinophilia also supported the diagnosis as it represents a common finding,[2] while ANA positivity has also been reported in a few cases.[5] Although fibrinoid necrosis of the vessel walls would have been expected at histology,[2] the eosinophilic infiltrate

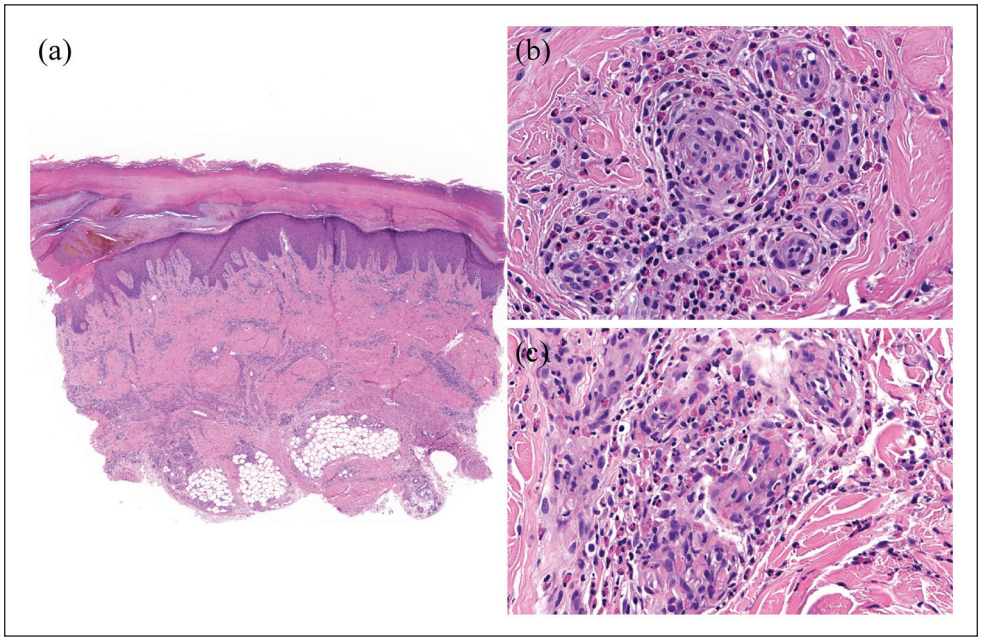

Figure 2. Histology showing a dense infiltrate in the dermis (with a perivascular and interstitial distribution) and surrounding the adipose lobules: (a) dermal and subcutaneous blood vessels show thickened walls infiltrated by eosinophils and endothelial hypertrophy with consequent vascular lumen obliteration (b, c).

invading the walls of dermal and subcutaneous vessels, and red blood cell extravasation were characteristic enough to allow the diagnosis of vasculitis. Furthermore, other differential diagnoses were excluded due to their different clinical or histologic presentation. Hypereosinophilic syndrome, which is associated with eosinophilia $>1500/\mu L$, was excluded due to the lack of visceral involvement.[6] Besides, cutaneous eosinophilic vasculitis is extremely rare in hypereosinophilic syndrome. Eosinophilic granulomatosis with polyangiitis also has peripheral eosinophilia but, unlike our case, it is associated with asthma and histologically by a leukocytoclastic vasculitis with granulomas.[7] Ultimately, hyperhomocysteinemia—which in our patient was explained by the MTHFR gene mutation—failed to explain the cutaneous and histologic presentation as it is associated with thrombosis (such as deep vein thrombosis), livedoid vasculopathy and leg ulcers.[8,9] However, hyperhomocysteinemia may have contributed to the development of retinal vein thrombosis in our patient, in keeping with studies which have shown that hyperhomocysteinemia represents a risk factor for retinal vein occlusion.[10] Of note, given the short time lapse between retinal vein occlusion and the development of the cutaneous manifestations in our patient, a causative role of RCEV in inducing retinal vein occlusion may also be hypothesized. This is in line with two previous cases of RCEV associated with hepatic vein occlusion and Budd–Chiari syndrome, respectively.[5] In conclusion, we described a case of RCEV characterized by purpuric lesions with an annular morphology. Despite its rarity, RCEV should be considered in the differential diagnosis of annular purpuric eruptions.

Declaration of conflicting interests

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Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. Ethical approval is not required for this study in accordance with national guidelines.

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References

1. Chen KR, Pittelkow MR, Su D, et al. Recurrent cutaneous necrotizing eosinophilic vasculitis. A novel eosinophil-mediated syndrome. *Arch Dermatol* 1994; 130(9): 1159–1166.
2. Quijano-Gomero EG, Rodríguez-Zúñiga MJM, Sanz-Montero ME, et al. Clinical, dermoscopic and histologic features of recurrent cutaneous eosinophilic vasculitis cases. *Actas Dermosifiliogr* 2019; 110(7): 590–596.
3. Sakuma-Oyama Y, Nishibu A, Oyama N, et al. A case of recurrent cutaneous eosinophilic vasculitis: successful adjuvant therapy with suplatast tosilate. *Br J Dermatol* 2003; 149(4): 901–903.
4. Tsunemi Y, Saeki H, Ihn H, et al. Recurrent cutaneous eosinophilic vasculitis presenting as annular urticarial plaques. *Acta Derm Venereol* 2005; 85(4): 380–381.

5. Lekić B, Gajić-Veljić M, Bonači-Nikolić B, et al. Penicillin-induced cutaneous necrotizing eosinophilic vasculitis with cryofibrinogenemia. *Acta Dermatovenerol Croat* 2019; 27(1): 28–32.
6. Curtis C and Ogbogu P. Hypereosinophilic syndrome. *Clin Rev Allergy Immunol* 2016; 50(2): 240–251.
7. Trivioli G, Terrier B and Vaglio A. Eosinophilic granulomatosis with polyangiitis: understanding the disease and its management. *Rheumatology* 2020; 59(Suppl. 3): iii84–iii94.
8. Cocuroccia B, Tonanzi T, Menaguale G, et al. Livedoid vasculopathy and skin ulcers in patients with inherited thrombophilia. *Eur J Dermatol* 2002; 12(4): 360–363.
9. Krupa Shankar DS and Akhib S. Leg ulcers due to hyperhomocysteinemia. *Indian J Dermatol Venereol Leprol* 2006; 72: 367–369.
10. Huang X, Yang Y, Duan Y, et al. Homocysteine in retinal artery occlusive disease: a meta-analysis of cohort studies. *Sci Rep* 2017; 7(1): 15708.