REVIEW ARTICLE

Recurrent Cutaneous Eosinophilic Vasculitis: A Systematic Review of Clinicopathologic Features and Treatment Outcomes

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Recurrent cutaneous eosinophilic vasculitis is a rare, predominantly cutaneous vasculitis described mainly in isolated reports. We aimed to systematically synthesize published evidence on its clinico-pathological features, treatment and outcomes. We reviewed case reports and case series of recurrent cutaneous eosinophilic vasculitis and related entities identified in PubMed, Scopus and reference lists (last searched 16 February 2026), using structured report- and case-level extraction, duplicate handling, Joanna Briggs Institute critical appraisal and descriptive synthesis. Twenty reports comprising 24 unique cases were included. Most patients presented with relapsing pruritic purpura of the lower extremities. Peripheral eosinophilia was frequent, antineutrophil cytoplasmic antibodies were negative in all reported cases and histopathology consistently showed eosinophil-rich vasculitis, usually with fibrinoid and/or necrotizing change. Systemic corticosteroids often induced improvement, but relapse during tapering or after withdrawal was common. The evidence was limited by small, heterogeneous reports and incomplete follow-up. Overall, the findings support a recognizable cutaneous-predominant, relapse-prone pattern, highlight the importance of assessing extracutaneous involvement in differential diagnosis and support provisional refinement of the previously proposed diagnostic framework.

Key words: eosinophilia; eosinophilic granulomatosis with polyangiitis; glucocorticoids; systematic review; vasculitis.

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SIGNIFICANCE

Recurrent cutaneous eosinophilic vasculitis is a rare skin disease that can resemble more serious illnesses affecting other organs. This review shows that it usually causes repeated itchy purple skin lesions on the lower legs and often flares again when steroid treatment is reduced. These findings may help doctors recognize the disease earlier, check for signs beyond the skin, and monitor patients more safely during treatment. This may improve diagnosis and follow-up in a condition that is easily overlooked.

Recurrent cutaneous eosinophilic vasculitis (RCEV) is a rare, primarily cutaneous disorder characterized by relapsing eruptions and eosinophil-rich vasculitis on histopathology (1). Many reports describe necrotizing or fibrinoid vascular changes, and some use the term recurrent cutaneous necrotizing eosinophilic vasculitis (RCNEV) for cases within this spectrum (2). Since its original description in 1994, the literature has remained limited and consists largely of single case reports and small case series (3). An earlier review summarized 17 patients and outlined the clinical, dermoscopic and histopathological features of the disease (1), but an updated systematic synthesis of published cases has not been available.

Important differential diagnoses include systemic eosinophilic disorders such as eosinophilic granulo-matosis with polyangiitis (EGPA) and hypereosinophilic syndrome (HES), as well as connective tissue disease-associated vasculitis and drug-induced eosinophilic vasculitis, all of which may present with eosinophil-rich cutaneous vasculitic lesions (4–7). Because published reports often provide limited or heterogeneous documentation of systemic evaluation and follow-up, the diagnosis of RCEV often depends on the exclusion of systemic eosinophilic disorders such as EGPA and HES, and distinguishing it from these conditions can be challenging in practice.

Reported clinicopathologic features are hetero-geneous. Common histopathologic descriptions include eosinophil-rich vasculitis with fibrinoid and/or necrotizing change (2). However, the involved vessel size and level within the skin are not uniformly de-scribed, although these features may be clinically important in the differential diagnosis. Reported therapeutic approaches have also been diverse. Systemic corticosteroids are commonly used and often induce clinical improvement; however,

relapse during tapering or after withdrawal, as well as glucocorticoid depen-dence, has been described, whereas evidence for steroid-sparing or adjunctive therapies remains limited (8). As a result, the overall clinicopathologic profile, treatment course and relapse pattern of RCEV remain incompletely characterized.

Therefore, a structured synthesis is needed to better define this condition. Accordingly, we performed a systematic review of published RCEV reports, with structured extraction at both the report and patient levels and descriptive synthesis of clinical characteristics, laboratory and histopathological findings, treatments and outcomes. To our knowledge, this is the first systematic review of published reports of RCEV/RCNEV to include explicit duplicate handling and a structured case-level analysis.

MATERIALS AND METHODS

Study design and registration

We conducted a systematic review of published reports of RCEV and related entities described as RCNEV. The review protocol was registered in the International Prospective Register of Systematic Reviews (PROSPERO; CRD420261308795). No amendments were made to the registered protocol.

Eligibility criteria

We included case reports and case series describing patients diagnosed with RCEV, RCNEV or closely related terms, provided that the reports contained sufficient clinical and histopathological detail to allow case-level data extraction.

We excluded (*i*) review articles without original case-level data; (*ii*) reports primarily described as systemic eosinophilic vasculitis or systemic eosinophilic disease, including EGPA, or reports in which extra-cutaneous organ involvement was a defining diagnostic feature; and (*iii*) drug-induced eosinophilic vasculitis cases in which the vasculitis was primarily attributed to a medication, with a clear temporal relationship and resolution after drug withdrawal, and was described as a distinct secondary entity. Eligibility decisions were based on the diagnostic description and overall clinical context of each report, with the aim of focusing on primarily cutaneous, relapsing eosinophilic vasculitis phenotypes.

Information sources and search strategy

We searched PubMed and Scopus on 16 February 2026. No date or language restrictions were applied. The full electronic search strategies are provided in Table SI. In addition, the reference lists of included articles and relevant reviews were manually screened for additional eligible reports.

Study selection and duplicate handling

Records were de-duplicated and screened in 2 stages: title/abstract screening followed by full-text review. Title/abstract screening, full-text eligibility assessment and data extraction were performed independently by 2 reviewers, and disagreements were resolved through discussion and consensus. Reasons for exclusion at the full-text stage were recorded. Full-text articles that could not be retrieved were documented as not retrieved.

Because duplicate publication of the same patients was possible, we assessed potential duplication at both the report and case levels using patient age, sex, clinical morphology and distribution, histo-pathological features, treatment course and publication context. Duplicate or companion publications, defined as multiple reports of the same patients, were retained at the report level for transparency but contributed no additional unique cases at the patient level when overlap was confirmed. The PRISMA 2020 flow diagram was generated using the PRISMA2020 Shiny app (9).

Data extraction and variables

We developed a structured data extraction form and collected both study-level and patient-level variables. Study-level characteristics, including publication year, study design, number of reported patients and contribution to the unique-case dataset, are summarized in Table SII. Variables not explicitly report-ed in the source publication were coded as NR (not reported). For summaries restricted to cases with available data, particularly for laboratory and immunologic variables, only cases in which the relevant item was explicitly reported were included in the denominator.

Patient-level extraction included reported age at presentation, sex, cutaneous morphology, distribution, symptoms and associated systemic features; peripheral eosinophil data (absolute count and/or percentage) and related laboratory markers, including total IgE, antinuclear antibody (ANA), antineutrophil cytoplasmic antibody (ANCA), serum complement, C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR), when available; histopathological features, including eosinophil prominence, fibrinoid and/or necrotizing change, leukocytoclasia and the reported vessel size and depth of involvement when explicitly described; and direct immunofluorescence findings (DIF/IF), when performed. Treatments, including systemic and topical therapies and outcomes, including initial response, relapse/recurrence patterns, relapse thresholds during tapering when explicitly reported and

follow-up duration, were also extracted. Patient-level extracted data are provided in Table SIII.

Quality appraisal and synthesis

Methodological quality was assessed using Joanna Briggs Institute (JBI) critical appraisal tools, with the case report or case series checklist selected according to study design. Appraisal results are summarized at the report level in Table SIV.

Given the rarity of the condition and the heterogeneity of reporting, we performed a descriptive synthesis without meta-analysis. Patient-level findings were summarized using counts and percentages, with reported denominators used when data were missing or not explicitly stated. Reporting bias and certainty of evidence were not formally assessed because of the small number and descriptive nature of the included case reports and case series.

RESULTS

Study selection

A total of 278 records were identified through database searching (PubMed, *n*=73; Scopus, *n*=205). After removal of 69 duplicates, 209 records underwent title/abstract screening, of which 185 were excluded. Full texts were sought for 24 records, of which 2 could not be retrieved (1 Chinese-language article and 1 Korean-language article). Twenty-two full-text articles were assessed for eligibility, and 3 were excluded at the full-text stage (review article, *n*=1; EGPA case, *n*=1; drug-induced case, *n*=1). One additional eligible article was identified through reference list screening (10). In total, 20 reports were included (Table SII), and the selection process is shown in **Fig. 1**.

Included studies and unique cases

Included reports were published between 1994 and 2025 and consisted predominantly of case reports, with 2 small case series (1–3, 8, 10–25). Their study-level characteristics are summarized in Table SII. Terminology varied across reports (RCEV, RCNEV and related terms). At the report level, all included publications were retained for transparency; however, one report (S02) was judged to be a duplicate/companion publication of the S01 cohort and contributed no additional unique cases (*n*=0 in patient-level analyses) (3, 11). After duplicate handling, 24 unique cases were included in the patient-level analytic set used for **Tables I–III**.

Clinical characteristics and cutaneous phenotypes

Clinical features are summarized in Table I. The median age was 58 years (range 17–93), and 17/24 patients (70.8%) were female. Purpura was the most frequent morphology (18/24, 75.0%), with purpuric papules/plaques/patches (14/24, 58.3%) and erythematous papules/plaques (13/24, 54.2%) commonly described. Marked edema/angioedema was reported in half of cases (12/24, 50.0%), whereas necrotic lesions/ulcers (5/24, 20.8%), vesicles/bullae (3/24, 12.5%) and mucosal/oral involvement (3/24, 12.5%) were less frequent. Lesions involved the lower extremities in 18/24 cases (75.0%), with upper extremity involvement in 11/24 (45.8%),

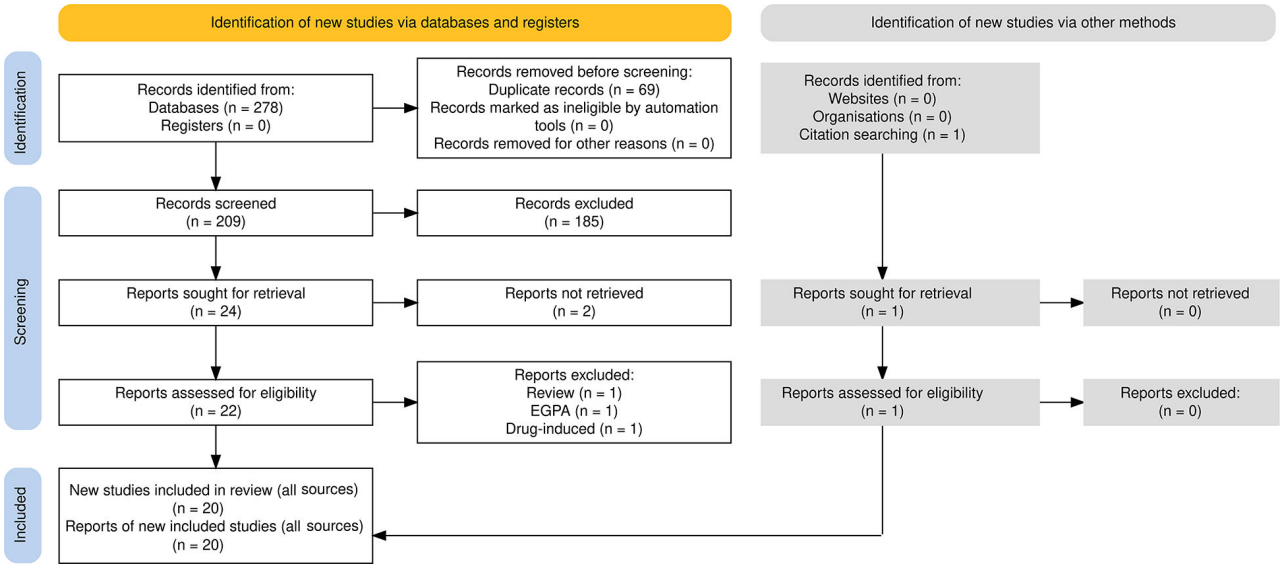

Fig. 1. PRISMA 2020 flow diagram of study identification, screening and inclusion. Flow diagram showing identification, screening, eligibility assessment and final inclusion of studies in this systematic review. Of 278 records identified, 69 duplicates were removed and 209 records were screened. Twenty-two reports were assessed for eligibility, and 20 reports were ultimately included after exclusion of 3 reports and addition of one report identified through citation searching. EGPA, eosinophilic granulomatosis with polyangiitis.

Table I. Clinical characteristics and cutaneous phenotypes

Item	Summary
Patient characteristics	
Age, years (median [range])	58 [17–93]
Female	17/24 (70.8%)
Male	7/24 (29.2%)
Skin lesion morphology	
Purpura (any)	18/24 (75.0%)
Purpuric papules/plaques/patches	14/24 (58.3%)
Erythematous papules/plaques	13/24 (54.2%)
Edematous plaques/angioedema/marked swelling	12/24 (50.0%)
Urticarial plaques/urticarial-like lesions	6/24 (25.0%)
Annular/polycyclic/central clearing	5/24 (20.8%)
Necrotic lesions/ulcers	5/24 (20.8%)
Vesicles/bullae	3/24 (12.5%)
Mucosal/oral involvement (including gingivitis)	3/24 (12.5%)
Distribution of skin lesions	
Lower extremity involvement	18/24 (75.0%)
Upper extremity involvement	11/24 (45.8%)
Trunk/abdomen involvement	8/24 (33.3%)
Head/face/neck involvement (including scalp/periorbital)	7/24 (29.2%)
Symptoms	
Pruritus	19/24 (79.2%)
Pain/tenderness	7/24 (29.2%)
Fever	3/24 (12.5%)
Systemic manifestations/associated findings	
Documented extracutaneous organ involvement/comorbidity relevant to the case	4/24 (16.7%)[a]
Asthma and/or allergic rhinitis history (*n*=8)	0/8 (0.0%)

[a]Includes Budd-Chiari/hepatic vein disease (*n*=2), lymphadenopathy with mild hepatosplenomegaly (*n*=1) and chronic periaortitis/retroperitoneal fibrosis (*n*=1)

Values are *n*/*N* (%), where *N*=24 unique cases, unless otherwise specified. Multiple morphologies, sites, and symptoms may co-occur; categories are not mutually exclusive. Asthma/allergic rhinitis was summarized among cases with explicit reporting (*n*=8). Extracutaneous involvement and comorbidities were determined by manual review of the clinical descriptions.

trunk/abdomen involvement in 8/24 (33.3%) and head/face/neck involvement in 7/24 (29.2%). Pruritus was the most frequent symptom (19/24, 79.2%), followed by pain/tenderness (7/24, 29.2%) and fever (3/24, 12.5%). Extracutaneous organ involvement or clinically relevant comorbidity was described in 4/24 cases (16.7%). A history of asthma and/or allergic rhinitis was not reported in most cases; among cases with explicit reporting (*n*=8), none had asthma/allergic rhinitis (0/8).

Laboratory, immunologic and histopathological findings

Laboratory and histopathological findings are summarized in Table II. Peripheral eosinophil data were available in 18/24 cases, and peripheral eosinophilia (>500 cells/μL and/or >5%) was present in 16/18 cases (88.9%). Among cases with reported absolute eosinophil counts (*n*=18), the median count was 1,700 cells/μL (range 220–10,080). Among cases reporting eosinophil percentages (*n*=9), the median was 31.0% (range 16.1–63.0). Total IgE was reported in 9 cases, and elevation was observed in 4/9 (44.4%), interpreted cautiously because of heterogeneous units and reference ranges. Inflammatory markers were variably reported: CRP was elevated in 8/13 (61.5%) and ESR in 8/13 (61.5%) among cases with reported values. ANA was positive in 4/18 (22.2%), typically weak or low-titer when specified, whereas ANCA was negative in all reported cases (0/22) and hypocomplementemia was not reported (0/16).

Table II. Laboratory, immunologic and histopathological findings

Item	Summary
Peripheral blood eosinophilia	
Peripheral eosinophilia present (>500 cells/μL and/or >5%) (*n*=18)	16/18 (88.9%)
Absolute eosinophil count, median [range] (*n*=18)	1700 [220–10,080] cells/μL
Eosinophil percentage, median [range] (*n*=9)	31.0 [16.1–63.0]
IgE elevated (*n*=9)	4/9 (44.4%)
Inflammatory markers	
CRP elevated (*n*=13)	8/13 (61.5%)
ESR elevated (*n*=13)	8/13 (61.5%)
Autoantibodies and complement	
ANA positive (*n*=18)	4/18 (22.2%)[a]
ANCA positive (*n*=22)	0/22 (0.0%)
Hypocomplementemia (*n*=16)	0/16 (0.0%)
Histopathology	
Eosinophils prominent	24/24 (100.0%)
Necrotizing and/or fibrinoid vasculitis present	22/24 (91.7%)
Leukocytoclasia/leukocytoclastic vasculitis present	2/24 (8.3%)
Affected vessels and depth	
Small-vessel involvement only explicitly described	9/24 (37.5%)
Deep dermis and/or subcutis involvement only explicitly described	3/24 (12.5%)
Both small-vessel and deep dermis/subcutis involvement explicitly described	6/24 (25.0%)
Neither small-vessel involvement nor deep dermis/subcutis involvement explicitly described	6/24 (25.0%)
Tissue immunofluorescence (DIF/IF)	
C3 deposition in vessel walls (*n*=10)	2/10 (20.0%)
IgM deposition in vessel walls (*n*=10)	1/10 (10.0%)

[a]Includes weak/slight positive or low-titer ANA

Values are *n*/*N* (%), where *N*=24 unique cases, unless otherwise specified. For laboratory/immunologic tests, the denominator for each item is the number of cases in which the relevant test was reported (*n* shown). Vessel size/depth categories were assigned based on explicit reporting and are mutually exclusive.

ANA:antinuclear antibody; ANCA:antineutrophil cytoplasmic antibody; CRP:C-reactive protein; DIF:direct immunofluorescence; ESR:erythrocyte sedimentation rate; IF:immunofluorescence.

Table III. Treatments and outcomes

Item	Summary
Treatments (any use during the reported course)	
Systemic corticosteroids	21/24 (87.5%)
Topical corticosteroids	6/24 (25.0%)
Dapsone	3/24 (12.5%)
Oral antihistamines	2/24 (8.3%)
Other systemic immunomodulatory agents (each reported in 1 patient)	4/24 (16.7%) Agents: oral tacrolimus, methotrexate, azathioprine, cyclophosphamide.
Other adjunctive agents (reported in small numbers)	7/24 (29.2%) Agents: colchicine, indomethacin, suplatast tosilate, pentoxifylline, compound glycyrrhizin, warfarin, aspirin, beraprost.
Systemic corticosteroid regimen, response, and follow-up	
Oral prednisone/prednisolone used	13/24 (54.2%)
Prednisone/prednisolone initial dose (mg/day), median [range] (*n*=10)	30 [10–60] mg/day
Prednisone/prednisolone initial dose (mg/kg/day), median [range] (*n*=3)	1.0 [0.3–1.0] mg/kg/day
Prednisone/prednisolone relapse threshold during tapering (mg/day), median [range] (*n*=7)	5 [2–25] mg/day
Response to systemic corticosteroids: rapid/prompt improvement (*n*=21)	14/21 (66.7%)
Response to systemic corticosteroids: relapse/recurrence during taper/withdrawal/discontinuation (*n*=21)	15/21 (71.4%)
Follow-up duration (months), median [range] (*n*=19)	8 [2–276] months
No relapse/recurrence at last follow-up (*n*=19)	7/19 (36.8%)

Values are *n*/*N* (%), where *N*=24 unique cases, unless otherwise specified. Multiagent regimens are counted in multiple rows. Prednisone/prednisolone dose summaries were calculated among oral users (*n*=13) and among cases with explicit dose reporting (n shown). Response outcomes were assessed among systemic corticosteroid-treated cases (*n*=21). Follow-up summaries were calculated among cases reporting follow-up duration (*n*=19).

Histopathologically, eosinophils were described as prominent in all cases (24/24), and necrotizing and/or fibrinoid vasculitic changes were reported in most cases (22/24). Leukocytoclasia or leukocytoclastic vasculitis was explicitly described in 2/24 cases (8.3%). With respect to vessel size and level within the skin, small-vessel involvement only was explicitly described in 9/24 cases (37.5%), deep dermis and/or subcutis involvement only in 3/24 (12.5%), and both in 6/24 (25.0%); in the remaining 6/24 cases (25.0%), neither was explicitly stated. DIF/IF was performed in a minority of cases (*n*=10), with C3 deposition in vessel walls reported in 2/10 (20.0%) and IgM deposition in 1/10 (10.0%).

Representative unpublished clinical and histopathological images from a previously reported case from our institution (22) are shown in **Fig. 2** to provide visual context for the typical presentation of RCEV.

Treatments and outcomes

Treatment patterns and outcomes are summarized in Table III. Systemic corticosteroids were the most commonly reported therapy (21/24, 87.5%). Other therapies included topical corticosteroids (6/24, 25.0%), dapsone (3/24, 12.5%) and oral antihistamines (2/24, 8.3%). Other systemic immunomodulatory agents were each reported in single patients (oral tacrolimus, methotrexate, azathioprine and cyclophosphamide; 4/24, 16.7%). Additional adjunctive

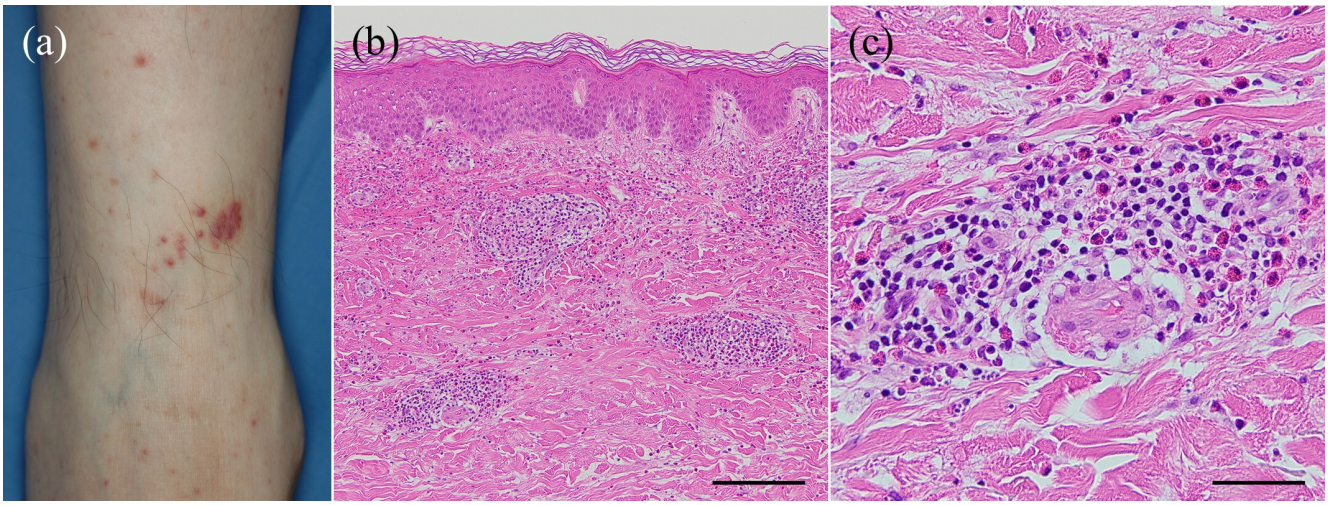

Fig. 2. Representative clinical and histopathological findings in a previously reported institutional case of recurrent cutaneous eosinophilic vasculitis. Representative unpublished clinical and histopathological images from a previously reported case from our institution (22). (a) Clinical photograph showing multiple palpable purpuric lesions on the anterior aspect of the right lower leg. (b) Low-power histopathological image showing superficial to mid-dermal perivascular inflammatory infiltrates (hematoxylin-eosin stain; scale bar = 200 μm). (c) High-power histopathological image showing eosinophil-rich vasculitis with fibrinoid change of dermal vessels (hematoxylin-eosin stain; scale bar = 50 μm).

agents were reported in small numbers (7/24, 29.2%), including colchicine, indomethacin, suplatast tosilate, pentoxifylline, compound glycyrrhizin and antithrombotic/vasodilator therapies.

Among cases treated with oral prednisone/prednisolone (13/24), the median initial dose was 30 mg/day (range 10–60; *n*=10 with explicit dose reporting), and the median relapse threshold during tapering was 5 mg/day (range 2–25; *n*=7). Rapid or prompt improvement after systemic corticosteroid treatment was explicitly described in 14/21 cases (66.7%), whereas relapse/recurrence during tapering or after withdrawal was described in 15/21 (71.4%). Follow-up duration was reported in 19 cases; the median follow-up was 8 months (range 2–276). At last follow-up, 7/19 cases (36.8%) had no reported relapse/recurrence.

DISCUSSION

This systematic review identifies a recognizable clinicopathologic pattern of RCEV despite the rarity of the condition and heterogeneity of the published literature. As the literature consists predominantly of case reports and small case series, the available evidence remains limited. Nevertheless, this review has practical clinical importance because it helps define the typical presentation of RCEV, clarifies laboratory and histopathological patterns relevant to differential diagnosis and highlights the relapse-prone treatment course that clinicians may encounter in practice.

The patient-level clinical profile suggests that the condition most commonly presents as a relapsing, pruritic, purpuric eruption predominantly involving the lower extremities. While purpura and purpuric papules/plaques predominate, the cutaneous spectrum also includes erythematous and edematous lesions, and a minority of cases exhibit vesiculobullous changes, mucosal involvement or ulcerative/necrotic lesions. Although less common, these variants are clinically important because they can obscure the typical presentation of RCEV and broaden the differential diagnosis.

Assessment of extracutaneous organ involvement is particularly important when differentiating RCEV from systemic eosinophilic disorders, especially EGPA. Notably, asthma and/or allergic rhinitis, which are key clinical clues for EGPA, were not reported among cases with explicit reporting, and neurologic manifestations suggestive of EGPA, such as paresthesia or peripheral neuropathy, were also not documented. No included report explicitly described subsequent evolution to EGPA or later ANCA seroconversion, although follow-up was limited and heterogeneous. Given the overlap in cutaneous morphology between RCEV and EGPA, the diagnosis of RCEV remains partly dependent on exclusion of EGPA at presentation. Therefore, baseline systemic assessment and continued clinical reassessment are advisable in practice (26).

Laboratory and immunologic testing was variably reported, with item-specific denominators because not all tests were performed or explicitly reported in all cases. Within these limitations, most cases with available eosinophil data demonstrated peripheral eosinophilia, suggesting that eosinophilia is a common laboratory feature of RCEV. In contrast, immunologic testing suggests a pattern that may help in differential diagnosis: ANCA was negative in all reported cases, and hypocomplementemia was not reported among cases in which complement was documented. ANA positivity was observed in a minority of reported cases and was often weak or low-titer when specified, emphasizing the need for cautious interpretation in clinical context.

Histopathology showed a consistent hallmark across reported cases, with eosinophil-prominent vasculitis and necrotizing and/or fibrinoid change described in most cases, whereas leukocytoclasia was explicitly reported only in a small minority. This contrast may reflect histopathologic differences from classic leukocytoclastic vasculitis patterns. The reported size and level of vascular involvement were variable, ranging from small-vessel involvement only to deep dermis/subcutis involvement only, with some cases showing both patterns. This variability in vascular involvement may contribute to the heterogeneous clinical morphology reported across cases. DIF/IF was performed in a minority of cases. When performed, deposition of C3 and IgM in vessel walls was reported only in subsets. Clinically, these laboratory and pathologic patterns support RCEV and can help guide the differential diagnosis when considering EGPA or immune complex-mediated vasculitides (27).

Although diagnostic criteria for RCEV have been proposed by Quijano-Gomero et al., consistent application across the literature has not yet been achieved (1). The criteria comprise major clinicopathologic items and supportive minor items, and define RCEV as either all 3 major criteria plus at least 1 minor criterion, or 2 major criteria plus at least 3 minor criteria. Major items include pruritic erythematous-purpuric plaques/papules, eosinophil-rich vasculitis with fibrinoid necrosis and response to systemic and/or high-potency topical corticosteroids, whereas minor items include edema/angioedema, eosinophilia, dermoscopic purpuric spots, recurrence after tapering/discontinuation of corticosteroids and exclusion of alternative diagnoses.

Our patient-level synthesis suggests that refinement of the previously proposed criteria may be warranted. In particular, inclusion of treatment response among

the major criteria may introduce incorporation bias in retrospective reports, as cases may be more readily labeled as RCEV when they respond to corticosteroids; accordingly, this feature may be better regarded as a minor supportive item. Conversely, exclusion of alternative diagnoses, particularly EGPA, HES, connective tissue disease-associated vasculitis and drug-induced eosinophilic vasculitis, may be more appropriately considered a major diagnostic requirement rather than a minor supportive item. These findings support a provisional framework in which all major diagnostic requirements should be fulfilled, with treatment response retained as a supportive rather than major feature (**Table IV**). Prospective validation is therefore needed to assess the diagnostic performance and generalizability of this provisional framework. Although retrospective application of both frameworks to all included cases would be of interest, several key items were incompletely reported in the source literature, particularly systematic exclusion of alternative diagnoses, dermoscopic findings and long-term follow-up. Therefore, the relative applicability of the 2 frameworks could not be reliably compared in the present review.

Systemic corticosteroids are the dominant therapy and are often associated with improvement; however, relapse during tapering or after discontinuation is frequent among steroid-treated cases. Among prednisone/prednisolone users with explicit dose reporting, clinical improvement was generally achieved at moderate doses, whereas relapse tended to occur at lower doses during tapering, suggesting corticosteroid dependence in a subset. Clinicians should therefore expect relapse during dose reduction and monitor patients closely, particularly at lower doses.

Table IV. Provisional refinement of previously proposed diagnostic framework for recurrent cutaneous eosinophilic vasculitis

Category	Items
Major diagnostic requirements	1. Pruritic erythematous-purpuric papules/plaques
	2. Eosinophil-rich vasculitis with fibrinoid necrosis and eosinophilic infiltration of the dermis and/or vessel walls
	3. Exclusion of alternative diagnoses, particularly EGPA, HES, connective tissue disease-associated vasculitis, and drug-induced eosinophilic vasculitis
Minor supportive items	1. Edema/angioedema
	2. Eosinophilia
	3. Dermoscopic purpuric spots
	4. Response to systemic and/or high-potency topical corticosteroids
	5. Recurrence during corticosteroid tapering or after withdrawal

Proposed rule: All major diagnostic requirements should be fulfilled. Minor supportive items may increase diagnostic confidence but are not mandatory. This framework is intended as a provisional refinement of the previously proposed criteria based on the present patient-level synthesis and should not be regarded as a validated diagnostic or classification criterion. Prospective validation is required.
EGPA:eosinophilic granulomatosis with polyangiitis; HES:hypereosinophilic syndrome.

Adjunctive steroid-sparing therapy may be considered in relapse-prone cases to minimize cumulative glucocorticoid exposure, but the supporting evidence remains limited. Reported agents were used only sporadically and usually in single patients, making meaningful comparisons across therapies difficult. At present, the diversity of attempted therapies appears to reflect clinical need in relapse-prone disease rather than evidence-based selection. Prospective data are needed to clarify when adjunctive therapy should be introduced and which agents offer the best balance of efficacy and safety.

Strengths and limitations

This review has several strengths. It used a structured search strategy with transparent documentation, extraction at both report and patient levels with explicit duplicate handling and report-level JBI appraisal. It also provides a provisional refinement of the previously proposed diagnostic framework based on structured patient-level synthesis. At the same time, the findings should be interpreted with important limitations in mind. The evidence base consisted almost entirely of case reports and very small case series; reporting was heterogeneous; and missing data were common for clinical history, laboratory variables, immunologic testing, histopathological details, treatment details and follow-up. Publication bias is also likely. A further limitation is that, because RCEV is diagnosed in part by excluding EGPA and HES, some reported cases may have represented early or incompletely evolved EGPA or HES in the setting of limited systemic evaluation or follow-up. Given these limitations, the reported clinicopathologic patterns, diagnostic implications and treatment outcomes should be interpreted with caution.

Conclusion

Overall, 2 findings emerge from the patient-level synthesis: a characteristic clinicopathologic pattern and a relapse-prone course during corticosteroid tapering. Taken together, these observations support evaluation for extracutaneous involvement and close monitoring during corticosteroid dose reduction. Future RCEV research should focus on prospective, standardized collection of clinically relevant clinicopathologic features, laboratory data and longitudinal treatment outcomes to enable validation of diagnostic criteria and improvement of clinical management. Collaborative case series or registry-based studies in RCEV may be particularly valuable.

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Ethics committee: Ethics approval and informed consent were not required for this study because it was a systematic review of previously published reports. The review protocol was registered in PROSPERO (CRD420261308795). Written informed consent for publication of the clinical images in Fig. 2 was obtained.

The authors have no conflicts of interest to declare.

REFERENCES

1. Quijano-Gomero EG, Rodríguez-Zúñiga MJM, Sanz-Montero ME, Durand-Buse D. Clinical, dermoscopic and histologic features of recurrent cutaneous eosinophilic vasculitis cases. Actas Dermosifiliogr 2019; 110: 590–596. https://doi.org/10.1016/j.ad.2017.12.021
2. Li W, Cao W, Song H, Ciu Y, Lu X, Zhang F. Recurrent cutaneous necrotizing eosinophilic vasculitis: a case report and review of the literature. Diagn Pathol 2013; 8: 185. https://doi.org/10.1186/1746-1596-8-185
3. Chen KR, Pittelkow MR, Su D, Gleich J, Newman W, Leiferman KM. Recurrent cutaneous necrotizing eosinophilic vasculitis. A novel eosinophil-mediated syndrome. Arch Dermatol 1994; 130: 1159–1166. https://doi.org/10.1001/archderm.1994.01690090083012
4. Ishibashi M, Kawahara Y, Chen KR. Spectrum of cutaneous vasculitis in eosinophilic granulomatosis with polyangiitis (Churg-Strauss): a case series. Am J Dermatopathol 2015; 37: 214–221. https://doi.org/10.1097/DAD.0000000000000192
5. Lefèvre G, Leurs A, Gibier JB, Copin MC, Staumont-Sallé D, Dezoteux F, et al. "Idiopathic eosinophilic vasculitis": another side of hypereosinophilic syndrome? A comprehensive analysis of 117 cases in asthma-free patients. J Allergy Clin Immunol Pract 2020; 8: 1329–1340. https://doi.org/10.1016/j.jaip.2019.12.011
6. Chen KR, Su WP, Pittelkow MR, Conn DL, George T, Leiferman KM. Eosinophilic vasculitis in connective tissue disease. J Am Acad Dermatol 1996; 35: 173–182. https://doi.org/10.1016/s0190-9622(96)90318-7
7. Lekić B, Gajić-Veljić M, Bonači-Nikolić B, Nikolić M. Penicillin-induced cutaneous necrotizing eosinophilic vasculitis with cryofibrinogenemia. Acta Dermatovenerol Croat 2019; 27: 28–32.
8. Lin TL, Yang CS, Chen YJ. Recurrent cutaneous necrotising eosinophilic vasculitis. Australas J Dermatol 2021; 62: e102–e106. https://doi.org/10.1111/ajd.13451
9. Haddaway NR, Page MJ, Pritchard CC, McGuinness LA. *PRISMA2020*: an R package and Shiny app for producing PRISMA 2020-compliant flow diagrams, with interactivity for optimised digital transparency and Open Synthesis. Campbell Syst Rev 2022; 18: e1230. https://doi.org/10.1002/cl2.1230
10. Nakajima H, Nakamura T. Thrombotic eosinophilic vasculitis. Clin Exp Dermatol 2009; 34: e69–71. https://doi.org/10.1111/j.1365-2230.2008.03178.x
11. Chen KR, Su WP, Pittelkow MR, Leiferman KM. Eosinophilic vasculitis syndrome: recurrent cutaneous eosinophilic necrotizing vasculitis. Semin Dermatol 1995; 14: 106–110. https://doi.org/10.1016/s1085-5629(05)80005-7
12. Launay D, Delaporte E, Gillot JM, Janin A, Hachulla E. An unusual cause of vascular purpura: recurrent cutaneous eosinophilic necrotizing vasculitis. Acta Derm Venereol 2000; 80: 394–395.
13. Sakuma-Oyama Y, Nishibu A, Oyama N, Saito M, Nakamura K, Kaneko F. A case of recurrent cutaneous eosinophilic vasculitis: successful adjuvant therapy with suplatast tosilate. Br J Dermatol 2003; 149: 901–903. https://doi.org/10.1046/j.1365-2133.2003.05568.x
14. Tsunemi Y, Saeki H, Ihn H, Tamaki K. Recurrent cutaneous eosinophilic vasculitis presenting as annular urticarial plaques. Acta Derm Venereol 2005; 85: 380–381. https://doi.org/10.1080/00015550510030113
15. Tanglertsampan C, Tantikun N, Noppakun N, Pinyopornpanit V. Indomethacin for recurrent cutaneous necrotizing eosinophilic vasculitis. J Med Assoc Thai 2007; 90: 1180–1182.
16. Kiorpelidou D, Gaitanis G, Zioga A, Tsili AC, Bassukas ID. Chronic periaortitis (retroperitoneal fibrosis) concurrent with recurrent cutaneous eosinophilic vasculitis. Case Rep Dermatol Med 2011; 2011: 548634. https://doi.org/10.1155/2011/548634
17. Sugiyama M, Nozaki Y, Ikoma S, Kinoshita K, Funauchi M. Successful treatment with tacrolimus in a case of the glucocorticoid-dependent recurrent cutaneous eosinophilic vasculitis. Ann Dermatol 2013; 25: 252–254. https://doi.org/10.5021/ad.2013.25.2.252
18. Sawada C, Taniai M, Kawashima M, Ishiguro N. Recurrent cutaneous eosinophilic vasculitis. Eur J Dermatol 2016; 26: 108–109. https://doi.org/10.1684/ejd.2015.2680
19. Riyaz N, Sasidharanpillai S, Hazeena C, Aravindan KP, Bindu CS, Silpa KN. Recurrent cutaneous eosinophilic vasculitis: a rare entity. Indian J Dermatol 2016; 61: 235. https://doi.org/10.4103/0019-5154.177794
20. Uehara A, Endo Y, Kosaka K, Oka A, Ishikawa O, Motegi SI. Case of recurrent cutaneous eosinophilic vasculitis with subcutaneous nodules in the early stage of the disease. J Dermatol 2022; 49: e217–e218. https://doi.org/10.1111/1346-8138.16342
21. Gisondi P, Geat D, Colato C, Girolomoni G. Recurrent cutaneous eosinophilic vasculitis characterized by annular purpuric lesions: a case report. SAGE Open Med Case Rep 2023; 11: 2050313X231163636. https://doi.org/10.1177/2050313X231163636
22. Iinuma S, Kobayashi T, Ishida-Yamamoto A. Pruritic palpable purpura on the lower legs: a quiz. Acta Derm Venereol 2023; 103: adv18366. https://doi.org/10.2340/actadv.v103.18366
23. Li J, Lin J, Ma L. Azathioprine combined with corticosteroids for recurrent cutaneous necrotizing eosinophilic vasculitis secondary to eosinophilic dermatitis. Minerva Gastroenterol 2023; 69: 596–597. https://doi.org/10.23736/S2724-5985.23.03424-1
24. Rodriguez-Troncoso M, Ruiz-Villaverde R, Pegalajar-García MD. sQuiz your knowledge! recurrent ulcerated papuloplaques on the legs: a diagnosis not to be missed. Eur J Dermatol 2025; 35: 162–164. https://doi.org/10.1684/ejd.2025.4897
25. Ohnishi Y, Zenke Y, Arai S, Suyama Y. Recurrent cutaneous eosinophilic vasculitis. J Allergy Clin Immunol Pract 2025; 13: 3420–3421. https://doi.org/10.1016/j.jaip.2025.09.033
26. Emmi G, Bettiol A, Gelain E, Bajema IM, Berti A, Burns S, et al. Evidence-based guideline for the diagnosis and management of eosinophilic granulomatosis with polyangiitis. Nat Rev Rheumatol 2023; 19: 378–393. https://doi.org/10.1038/s41584-023-00958-w
27. Frumholtz L, Laurent-Roussel S, Lipsker D, Terrier B. Cutaneous vasculitis: review on diagnosis and clinicopathologic correlations. Clin Rev Allergy Immunol 2021; 61: 181–193. https://doi.org/10.1007/s12016-020-08788-4