Supplementary Table S1. Full search strategies used for identification of studies

Database	Date searched	Search strategy
PubMed	16 February 2026	("recurrent cutaneous eosinophilic vasculitis"[tiab] OR "recurrent cutaneous necrotizing eosinophilic vasculitis"[tiab] OR "recurrent cutaneous eosinophilic necrotizing vasculitis"[tiab] OR "cutaneous necrotizing eosinophilic vasculitis"[tiab] OR "cutaneous eosinophilic necrotizing vasculitis"[tiab] OR RCNEV[tiab] OR RCEV[tiab]) OR (recurrent[tiab] AND (cutaneous[tiab] OR skin[tiab]) AND eosinophil*[tiab] AND (vasculitis[tiab] OR necrotiz*[tiab] OR necrotis*[tiab] OR leukocytoclastic[tiab]))
Scopus	16 February 2026	TITLE-ABS-KEY("recurrent cutaneous eosinophilic vasculitis" OR "recurrent cutaneous necrotizing eosinophilic vasculitis" OR "recurrent cutaneous eosinophilic necrotizing vasculitis" OR "cutaneous necrotizing eosinophilic vasculitis" OR "cutaneous eosinophilic necrotizing vasculitis" OR RCNEV OR RCEV) OR TITLE-ABS-KEY(recurrent AND (cutaneous OR skin) AND eosinophil* AND (vasculitis OR necrotiz* OR necrotis* OR leukocytoclastic))

Full search strategies for study identification. No date or language restrictions were applied. Reference lists of included articles and relevant reviews were screened. Two non-English full texts could not be retrieved and were documented as “not retrieved” in the PRISMA flow diagram. Line breaks were added for readability only.

Abbreviations: RCEV, recurrent cutaneous eosinophilic vasculitis; RCNEV, recurrent cutaneous necrotizing eosinophilic vasculitis.

Supplementary Table S2. Included studies and study-level characteristics

Study ID (Reference)	First author	Year	Country	Journal	Article type	Terminology used	No. of patients reported (n)	No. of unique cases contributed (n)
S01 (3)	Chen	1994	USA	Arch. Dermatol.	Case series	RCNEV	3	3
S02 (11)	Chen	1995	USA	Semin. Dermatol.	Case series	RCNEV	3	0
S03 (12)	Launay	2000	France	Acta Derm. Venereol.	Case report	RCEV	1	1
S04 (13)	Sakuma-Oyama	2003	Japan	Br. J. Dermatol.	Case report	RCEV	1	1
S05 (14)	Tsunemi	2005	Japan	Acta Derm. Venereol.	Case report	RCEV	1	1
S06 (15)	Tanglertsampan	2007	Thailand	J. Med. Assoc. Thai.	Case report	RCNEV	1	1
S07 (10)	Nakajima	2009	Japan	Clin Exp Dermatol	Case report	Thrombotic eosinophilic vasculitis	1	1
S08 (16)	Kiorpelidou	2011	Greece	Case Rep. Dermatol. Med.	Case report	RCEV	1	1
S09 (17)	Sugiyama	2013	Japan	Ann. Dermatol.	Case report	RCEV	1	1
S10 (2)	Li	2013	China	Diagn. Pathol.	Case report	RCNEV	1	1
S11 (18)	Sawada	2016	Japan	Eur. J. Dermatol.	Case report	RCEV	1	1
S12 (19)	Riyaz	2016	India	Indian J. Dermatol.	Case report	RCEV	1	1
S13 (1)	Quijano-Gomero	2019	Peru	Actas Dermosifiliogr.	Case series	RCEV	4	4
S14 (8)	Lin	2021	Taiwan	Australas. J. Dermatol.	Case report	RCEV	1	1
S15 (20)	Uehara	2022	Japan	J. Dermatol.	Case report	RCEV	1	1
S16 (21)	Gisondi	2023	Italy	SAGE Open Med. Case Rep.	Case report	RCEV	1	1
S17 (22)	Iinuma	2023	Japan	Acta Derm. Venereol.	Quiz (case-based report)	RCEV	1	1
S18 (23)	Li	2023	China	Minerva Gastroenterol. (Torino)	Case report	RCNEV	1	1
S19 (24)	Rodríguez-Troncoso	2025	Spain	Eur. J. Dermatol.	Quiz (case-based report)	RCEV	1	1
S20 (25)	Ohnishi	2025	Japan	J. Allergy Clin. Immunol. Pract.	Case report	RCEV	1	1

Included studies are listed at the report level. Reference numbers correspond to the main reference list in the manuscript. “No. of unique cases contributed (n)” indicates the number of unique patients included in the final patient-level dataset used for Tables I–III. S01 and S02 were retained for report-level transparency; S02 was judged to describe the same cases as S01 and contributes no unique cases (n=0). NR indicates not reported in the source publication.

Abbreviations: RCEV, recurrent cutaneous eosinophilic vasculitis; RCNEV, recurrent cutaneous necrotizing eosinophilic vasculitis; EGPA, eosinophilic granulomatosis with polyangiitis; NR, not reported.

Supplementary Table S3. Patient-level extracted data

S3a. Patient-level extracted data (unique cases): clinical features

Patient ID	Study ID	Age	Sex	Lesion morphology	Distribution	Symptoms	Systemic details	Asthma/allergic rhinitis (history)
S01-P1	S01	56	F	Recurrent generalized pruritic erythematous and purpuric papules; palpable purpura; angioedema; persistent gingivitis	Generalized skin; periorbital/perioral edema; hands	Pruritus; gingivitis; edema/angioedema	No systemic organ involvement described; investigations largely negative/normal.	No
S01-P2	S01	18	F	Recurrent pruritic erythematous/purpuric papules; palpable purpura; urticarial plaques; gingivitis; angioedema	Generalized; hands/periorbital area	Pruritus; gingivitis; angioedema	Hepatic vein occlusion history (surgical shunt); no asthma/allergic rhinitis; no other visceral involvement reported.	No
S01-P3	S01	17	M	Recurrent pruritic purpuric/urticarial lesions; erythematous oedematous plaques/annular lesions; angioedema	Feet/hands initially then generalized; buttocks → whole body	Pruritus; swelling; occasional arthralgia	Lymphadenopathy and mild hepatosplenomegaly reported; no asthma/allergic rhinitis; otherwise no visceral organ involvement noted.	No
S03-P1	S03	81	F	Intensely pruritic infiltrating purpuric papules and necrotic purpuric lesions; lesions at different ages; angioedema	Lower limbs (legs and feet) predominance; right hand angioedema	Intense pruritus; ankle/wrist arthralgia; angioedema	No fever or weight loss; no peripheral neuropathy; no cardiac/pulmonary/ENT/CNS/renal involvement; cultures sterile; imaging normal	No
S04-P1	S04	27	F	Pruritic violaceous swelling of digits; oedematous swelling of face/fingers; palpable purpura; urticarial plaques	Digits/fingers; face (upper eyelids/cheeks); legs; palms; trunk; abdomen (biopsy site)	Pruritus; oedema/angioedema; sore throat; low-grade fever/pyrexia	No palpable lymphadenopathy, hepatosplenomegaly, or neuropathy; pyrexia 38°C with sore throat	NR
S05-P1	S05	53	F	Recurrent intensely pruritic urticarial erythematous plaques; partly fused with annular configuration; resolve with brown pigmentation	Extremities (predominant); right thigh lesion biopsied	Intense pruritus; each lesion lasts ~10 days; frequent recurrence	No systemic symptoms; no asthma or allergic rhinitis; labs normal incl eosinophils, ESR, CRP, IgA/M/G/E, complements, tumor markers; ANA/RF/HBV/HCV negative; systemic exam negative incl malignancy/CTD	No
S06-P1	S06	53	M	Recurrent pruritic papules, nodules, and ulcers	Face, scalp, and hands (later left forearm purpuric papules)	Pruritus; papules/nodules/ulcers	Evaluation negative/normal: CBC, urinalysis, stool exam, ANA, ANCA, RF, ESR, C3/C4, CRP, cryoglobulin, HBV/HCV serology, VDRL, HIV; chest X-ray small calcified spot LUL; bacterial culture Staph epidermidis	NR
S07-P1	S07	93	F	Indurative purpura with gangrene; widespread purpura; microinfarcts	Both calves/thighs; right 2nd toe; left 3rd finger	Severe leg pain; bilateral leg oedema; fever	Persistent fever and bilateral lower-leg swelling; anaemia; PT/APTT prolonged; no thrombocytopenia; no visceral involvement reported	No
S08-P1	S08	82	F	Multiple polycyclic/annular erythematous papules and plaques; urticarial-like lesions with central clearing/scales; heal leaving greyish macules with blood extravasation	Extremities (wrist/hand; leg)	Pruritus; fatigue; anorexia	Abdominal CT revealed chronic periaortitis/retroperitoneal fibrosis with right mild hydronephrosis and ureter dilatation	NR
S09-P1	S09	80	F	Multiple purpuric patches (not palpable) and purpuric papules/plaques	Palms; lower extremities; trunk	Digital pain; low-grade fever; general malaise; weight loss	No systemic organ involvement identified; bone marrow eosinophil-dominant proliferation without atypia/abnormal karyotype	NR
S10-P1	S10	57	M	Needlepoint-to-millet-sized pruritic papules → purpuric plaques with angioedema; some necrotic lesions	Lower legs (anterior tibias) and feet; lower limbs predominant	Intense pruritus; angioedema	No visceral organ involvement on ECG, chest X-ray, abdominal ultrasound, cranial CT; BP normal; no fever/weight loss reported	NR
S11-P1	S11	55	F	Recurrent pruritic mild indurated erythemas (~5 mm); erythematous plaques on palms	Hands (palms)	Pruritus	Budd-Chiari syndrome with ascites and liver dysfunction; angiography showed severe stenosis of middle and left hepatic veins; no asthma/allergic rhinitis; no systemic vasculitis symptoms	NR

Patient ID	Study ID	Age	Sex	Lesion morphology	Distribution	Symptoms	Systemic details	Asthma/allergic rhinitis (history)
S12-P1	S12	45	F	Pruritic purpuric papules and plaques; some annular; angioedema	Forearms, legs, palms, soles; also forehead/neck/abdomen/limbs	Intensely pruritic; lip angioedema	No fever/constitutional symptoms; no asthma/allergic disorders; no precipitating food/drug; extensive evaluation negative (CBC incl AEC normal; LFT/RFT/urinalysis/coagulation normal; ANA/RF/ANCA/cryoglobulin negative; complement and Ig profile normal; CXR/ECG/US abdomen-pelvis/CT thorax/sinus X-ray/NCS normal)	NR
S13-P1	S13	61	F	Pruritic erythematous-purpuric macules/plaques with edema and pain; dermographism/face lesions	Generalized (started lower limbs → trunk/face/arms/thighs)	Pruritus; pain	Stool positive for Giardia lamblia (trigger/infection noted); ANCA negative	NR
S13-P2	S13	66	F	Poorly delimited erythematous-purpuric plaque with local edema; pruritus and pain	Left lower limb	Pruritus; pain; edema	Urine positive for E. coli; ANCA negative	NR
S13-P3	S13	78	M	Well-delimited erythematous-purpuric edematous pruritic plaque(s)	Lower limb (posterior right leg described)	Pruritus; mild edema	No other significant abnormalities; ANCA negative	NR
S13-P4	S13	88	F	Bilateral erythematous-purpuric pruritic painful edematous plaques (poorly delimited)	Lower limbs	Pruritus; pain; edema	No other significant abnormalities; ANCA negative	NR
S14-P1	S14	33	M	Erythematous papuloplaques with central purplish hue and haemorrhagic bullae; progressed to necrosis/ulceration	All four extremities (legs swelling)	Painful eruptions; leg swelling	No extracutaneous symptoms; no fever/oral ulcers/hepatosplenomegaly/genital lesions; extensive work-up negative (ANA/ANCA/complement/RF/cryoglobulins/HCV; urinalysis; stool O&P; echo; CXR; abdominal US)	NR
S15-P1	S15	59	F	Erythematous swelling with multiple subcutaneous nodules (early); later erythema/purpura with bloody blisters and palpable purpura	Lower legs/lower extremities	Edematous swelling; subcutaneous nodules; later palpable purpura/bloody blisters	Bone marrow normal; gallium scintigraphy and enhanced CT (chest–pelvis) showed no organ damage/neoplastic lesions	No
S16-P1	S16	71	F	Annular purpuric patches/plaques; mucosal purpura (lips/palate)	Palms/soles; lips; hard palate	Intensely pruritic eruption	Systemic exam normal; PET-CT findings attributed to URTI; no systemic vasculitis involvement	NR
S17-P1	S17	64	M	Palpable purpura with haemorrhagic vesicles	Lower legs	Pruritic papules/purpura	No clinical signs/symptoms/lab evidence of internal organ involvement	No
S18-P1	S18	62	M	Whole-body recurrent erythematous papules → erythroderma tendency; later violaceous papules (lower extremities spreading to trunk/head)	Whole body; lower extremities → trunk/head	Not stated	No significant lab abnormalities aside from eosinophilia; HIV and viral hepatitis negative; ANA/ANCA/complement/PT/APTT normal	NR
S19-P1	S19	46	F	Erythematous-purpuric papules and plaques; some ulcerated with raised border and perilesional erythema; nodular plaque with fibrinous background	Lower limbs (legs) and abdomen	Pruritus	No fever or systemic symptoms; PCR for Leishmania and bacterial/mycobacterial/fungal cultures negative; ANCA/serologies normal; PAS stain negative; immunohistochemical and clonal rearrangement studies normal (ruled out lymphomatoid papulosis)	NR
S20-P1	S20	48	F	Urticarial-to-purpuric lesions; purpura (palms/soles)	Palms and soles; also abdomen	Tingling → pain; later pruritus	No headache/nasal/respiratory/chest pain/arthralgia/myalgia/neurologic symptoms; urinalysis normal	NR

Legend: NR indicates not reported in the source publication. Multiple fields may apply to a single case; categories are not mutually exclusive. For asthma/allergic rhinitis, “No” was coded only when explicitly stated; otherwise coded as NR.

S3b. Patient-level extracted data (unique cases): laboratory and histopathological findings

Patient ID	Study ID	Eosinophil count (cells/µL)	Eosinophil (%)	IgE	Inflammatory/immunologic labs	Histology - vasculitis	DIF (Ig/C3)
S01-P1	S01	1400	NR	<29 µg/L	ESR: 10 mm/h; ANA: negative; ANCA: negative; Complement: normal (C3/C4/CH50)	Necrotizing small-vessel eosinophilic vasculitis with fibrinoid necrosis; eosinophils marginating/infiltrating vessel walls; leukocytoclasis minimal/absent	Negative (DIF)
S01-P2	S01	3600	25.5	218 µg/L	ESR: 110 mm/h; ANA: negative; ANCA: negative; Complement: normal (C3/C4/CH50)	Necrotizing small-vessel eosinophilic vasculitis with fibrinoid necrosis; marked perivascular eosinophils; leukocytoclasis minimal/absent	Negative (DIF)
S01-P3	S01	6200	32	59 280 µg/L	ESR: 61 mm/h; ANA: negative; ANCA: negative; Complement: normal (C3/C4/CH50)	Necrotizing small-vessel eosinophilic vasculitis with fibrinoid necrosis; eosinophil-predominant infiltrate; leukocytoclasis minimal/absent	Negative (DIF)
S03-P1	S03	3900	31	81 kU/L	CRP: 48.8 mg/L; ESR: 70 mm/h; ANA: positive (slight); ANCA: negative; Complement: CH50 135%; C3 1380 mg/L; C4 370 mg/L (inflammatory context)	Eosinophil-predominant necrotizing vasculitis of dermal small vessels/artery (deep dermis); early endothelial damage and RBC extravasation in non-necrotic lesion; immunohistochemistry showed deposits of eosinophil granule proteins	NR
S04-P1	S04	6900	48	Within normal limits	ESR: 154 mm/h; ANA: positive (1:320); Complement: NR	Necrotizing eosinophilic vasculitis with fibrinoid necrosis of dermal vessels/veins; no leukocytoclastic vasculitis, granuloma, or giant cells; small arteries intact on EVG stain	Negative (no Ig or complement binding)
S05-P1	S05	NR	NR	NR	CRP: normal; ESR: normal; ANA: negative; Complement: normal	Necrotizing small-vessel vasculitis with predominant eosinophils and some lymphocytes around dermal small vessels; destructive vascular change; eosinophil margination in lumen and infiltration into vessel walls; few neutrophils; no leukocytoclasis	Negative (DIF)
S06-P1	S06	NR	NR	NR	CRP: normal; ESR: normal; ANA: negative; ANCA: negative; Complement: normal	Necrotizing small-vessel vasculitis with eosinophil-rich infiltrate; endothelial necrosis; fibrin thrombi; eosinophilic fibrinoid deposition in vessel walls/lumens; no leukocytoclasis	NR
S07-P1	S07	1072	NR	NR	ANA: negative; ANCA: negative (p-ANCA); Complement: normal	Small-vessel eosinophilic vasculitis with vessel-wall destruction and intravascular thrombosis	NR
S08-P1	S08	NR	NR	NR	CRP: 30.1 mg/L; ESR: 107 mm/h; ANA: negative; ANCA: negative; Complement: normal	Eosinophilic vasculitis of superficial dermal vessels with endothelial swelling, intraluminal fibrin, eosinophils in vessel wall and eosinophilic dust; no leukocytoclastic vasculitis	Negative (DIF)
S09-P1	S09	10080	63	4 mg/dL	ANCA: negative (MPO-ANCA & PR3-ANCA); Complement: normal (not decreased)	Dermal small-vessel vasculitis (necrotizing) with eosinophil-predominant infiltrate	NR
S10-P1	S10	3400	28.7	658.3 IU/mL	CRP: 14.5 mg/L; ESR: 32 mm/h; ANA: negative; ANCA: negative (MPO-ANCA 9 Ru/mL; PR3-ANCA 7 Ru/mL; within normal range); Complement: C3 1260 mg/L (normal 900–1800); C4 430 mg/L (slightly high)	Necrotizing small-vessel vasculitis with eosinophil-predominant infiltrate in upper/deep dermis and subcutis; vessel-wall thickening; fibrinoid degeneration; extravascular RBCs; fibrin thrombi	NR
S11-P1	S11	781	NR	NR	CRP: elevated (mild); ANA: negative; ANCA: negative (MPO-ANCA and PR3-ANCA negative)	Eosinophilic necrotizing vasculitis in dermal superficial vascular plexus (arterioles/venules): eosinophils within/around vessel walls, fibrin deposits, luminal occlusion, RBC extravasation	NR
S12-P1	S12	NR	NR	NR	ANA: negative; ANCA: negative; Complement: normal	Necrotizing small-vessel vasculitis with eosinophil-predominant perivascular infiltrate; fibrinoid necrosis; erythrocyte extravasation	Negative (IgG/IgA/IgM/C3)
S13-P1	S13	700	NR	NR	CRP: 13 mg/L; ANCA: negative	Necrotizing small-vessel vasculitis (fibrinoid necrosis) with abundant eosinophils; no leukocytoclasia	NR
S13-P2	S13	400	NR	NR	CRP: 10 mg/L; ANCA: negative	Necrotizing vasculitis (fibrinoid necrosis) with eosinophils infiltrating vessel walls	NR

Patient ID	Study ID	Eosinophil count (cells/μL)	Eosinophil (%)	IgE	Inflammatory/immunologic labs	Histology - vasculitis	DIF (Ig/C3)
S13-P3	S13	800	NR	161.2 μg/L	CRP: 10 mg/L; ANCA: negative	Necrotizing small-vessel vasculitis (fibrinoid necrosis) with eosinophil-rich infiltrate; minimal/no leukocytoclasia; RBC extravasation	NR
S13-P4	S13	NR	NR	NR	ESR: 32 mm/h; ANCA: negative	Necrotizing small-vessel vasculitis (fibrinoid necrosis) with eosinophil-rich infiltrate; minimal/no leukocytoclasia; RBC extravasation	NR
S14-P1	S14	6760	38	NR	CRP: elevated; ESR: elevated; ANA: negative; ANCA: negative; Complement: normal	Necrotising vasculitis with eosinophil-predominant infiltrate; fibrinoid necrosis; multiple fibrin thrombi; deep dermis/subcutis involved; no leukocytoclasis	Negative (no Ig/complement binding)
S15-P1	S15	840	NR	7925.5 IU/mL	CRP: 0.4 mg/dL; ANA: negative; ANCA: negative (MPO-ANCA)	Eosinophilic vasculitis evolving to RCEV: early subcutaneous eosinophilic perivascular infiltrate without LCV/fibrinoid necrosis; later dermal vessel fibrinoid necrosis with marked eosinophils	NR
S16-P1	S16	2000	NR	NR	CRP: normal; ESR: normal; ANA: positive (1:160); ANCA: negative; Complement: normal	Eosinophil-rich small-vessel vasculitis; perivascular/interstitial eosinophils; endothelial hypertrophy and lumen obliteration; no fibrinoid necrosis	NR
S17-P1	S17	220	NR	NR	ANA: negative; ANCA: negative (PR3/MPO-ANCA); Complement: normal	Leukocytoclastic vasculitis with eosinophil-predominant infiltrate; fibrinoid degeneration; leukocytoclasia; erythrocyte extravasation	IgM and C3 deposition in dermal vessel walls; IgG/IgA negative
S18-P1	S18	4120	21	NR	ANA: negative; ANCA: negative; Complement: normal	Necrotizing vasculitis with fibrinoid degeneration; neutrophil + prominent eosinophil infiltration	NR
S19-P1	S19	NR	NR	NR	ANCA: negative	Epidermal ulceration with intra- and sub-epithelial oedema; perivascular/interstitial lymphohistiocytic infiltrate rich in eosinophils; focal collagen degeneration (“flame figure”); necrotizing vasculitis of small vessels without leukocytoclasis	NR
S20-P1	S20	1330	16.1	NR	CRP: normal; ESR: normal; ANA: negative; ANCA: negative (ELISA/IIF); Complement: normal	Leukocytoclastic vasculitis with nuclear dust around upper dermal vessels, hemorrhage, and prominent eosinophil infiltration	C3 deposition in dermal vessels (IF)

Legend: NR indicates not reported in the source publication. Laboratory values are shown as reported (units may vary). Absolute eosinophil counts are presented as cells/μL (values reported as ×10^9/L were converted; 1×10^9/L = 1000 cells/μL). IgE values are shown with the original units as reported. “Inflammatory/immunologic labs” summarizes CRP, ESR, ANA, ANCA, and complement findings in narrative form when available. Histology and DIF/IF fields summarize key wording from the source publication.

Abbreviations: ANA, antinuclear antibody; ANCA, antineutrophil cytoplasmic antibody; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; DIF, direct immunofluorescence; IF, immunofluorescence; NR, not reported.

S3c. Patient-level extracted data (unique cases): treatment and outcomes

Patient ID	Study ID	Treatment	Response/relapse	Follow-up (months)
S01-P1	S01	Prednisone 60 mg/day (complete resolution within 72 h); later maintained on alternate-day methylprednisolone 8–16 mg	Rapid response; recurred when glucocorticoids discontinued (steroid-dependent)	24
S01-P2	S01	Topical glucocorticoids + oral antihistamines (slight improvement); systemic glucocorticoids produce prompt response (dose not specified)	Chronic recurrent course; long follow-up	276
S01-P3	S01	Topical glucocorticoids + oral antihistamines (slight improvement); systemic glucocorticoids produce prompt response (dose not specified)	Chronic recurrent course; alopecia totalis; long follow-up	204
S03-P1	S03	Prednisone 1 mg/kg/day; tapered; recurrence when <5 mg/day; maintained 5 mg/day	Dramatic immediate improvement; eosinophils normalized within 12 h; healed necrotic lesions by ~3 weeks; relapse on taper <5 mg/day; controlled on 5 mg/day	12
S04-P1	S04	Prednisolone 20 mg/day → tapered (relapse at 5 mg/day); then betamethasone 1.5 mg/day + suplatast tosilate 300 mg/day; betamethasone tapered/withdrawn; continued suplatast	Rapid improvement on prednisolone; relapse during taper; symptoms gradually subsided with betamethasone+suplatast; no relapse after betamethasone withdrawal; stable eosinophils on suplatast	24
S05-P1	S05	Prednisolone 15 mg/day (0.3 mg/kg) with dramatic immediate response; tapered to 7.5 mg/day then relapse at 8 weeks → switched to betamethasone 0.75 mg	Controlled with maintenance betamethasone 0.75 mg; relapse during steroid taper noted	2
S06-P1	S06	Prednisolone 60 mg/day → good response; relapse after discontinuation for months; indomethacin 75 mg/day → little improvement then 150 mg/day + omeprazole 20 mg/day	Very good response to indomethacin within 1–2 weeks; recurrence after steroid discontinuation noted	NR
S07-P1	S07	Prednisolone 30 mg/day + warfarin 1 mg/day + aspirin 100 mg/day + beraprost 60 µg/day; debridement; relapse on taper → prednisolone 30 mg/day + cyclophosphamide 100 mg/day (stopped due to haematuria)	Fever/purpura resolved within 1 week; relapse when prednisolone reduced to 25 mg/day; improved with re-escalation; extensive ulcers after gangrene removal; died of heart failure	5
S08-P1	S08	Oral methylprednisolone 0.5 mg/kg/day + colchicine 1 mg/day; stopped after 1 month by patient → relapse 1 month later; restarted and tapered slowly over 3 months	Gradual resolution; relapse after stopping; then lesion-free at 6 months and remained so at 12 months	12
S09-P1	S09	Prednisolone 30 mg/day → tapered; relapse at 15 mg/day → prednisolone 30 mg/day; tacrolimus 2 mg/day added at prednisolone 20 mg/day; tapered prednisolone to 2.5 mg/day	Rapid response to prednisolone; relapse on taper; remission maintained with tacrolimus; renal insufficiency at tacrolimus 3 mg/day improved after reducing to 2 mg/day	NR
S10-P1	S10	Prednisone 1 mg/kg/day + compound glycyrrhizin 150 mg/day; mupirocin ointment to necrotic lesions; taper prednisone slowly	Marked improvement within days; lesions healed by 1 month with superficial scars; no new lesions at 4 months; recurrence with attempts to discontinue → continued prednisone 10 mg/day	4
S11-P1	S11	Balloon angioplasty for hepatic veins + warfarin potassium; no systemic corticosteroid	Ascites improved and skin symptoms disappeared within 1 week; eosinophils fluctuated up to 1929/µL without correlation with rash	NR
S12-P1	S12	Prednisolone 30 mg/day → resolved in 7–10 days; recurrence when tapered below 5 mg/day; maintained on 5 mg/day	Controlled on prednisolone 5 mg/day; recurrence on withdrawal/too-low dose	4
S13-P1	S13	Oral deflazacort 30 mg + topical clobetasol 0.05%; intermittent steroids for flares	Good response; relapse after steroid taper/withdrawal; controlled with intermittent therapy	6
S13-P2	S13	Deflazacort 30 mg/day + topical clobetasol 0.05% (2 wks) → recurrence after stopping; then dapsone 100 mg/day + deflazacort (tapered)	Controlled on dapsone; no recurrence reported	8
S13-P3	S13	Deflazacort 30 mg (2 wks) + topical clobetasol 0.05% twice daily (2 wks); then maintained on deflazacort 6 mg chronically	Remitted; maintained without recurrence reported	6
S13-P4	S13	Dapsone 100 mg/day	Controlled; no recurrence reported	6
S14-P1	S14	Dexamethasone 5 mg twice daily → rapid improvement; later relapse when prednisolone tapered to 5 mg/day at 4 weeks → increased to 10 mg/day; maintained on 10 mg/day	Improved rapidly; relapse on taper; controlled on prednisolone 10 mg/day	NR
S15-P1	S15	Prednisolone 30 mg/day → improved; relapse after taper/discontinuation; repeated relapses on taper (2–6 mg) managed by increasing dose; recurrence at 12 years →	Improves with prednisolone but relapses during taper; controlled on 5 mg	156

Patient ID	Study ID	Treatment	Response/relapse	Follow-up (months)
		prednisolone 30 mg again; tapered to 5 mg		
S16-P1	S16	Methylprednisolone 16 mg/day + pentoxifylline 400 mg/day; tapered to 4 mg/day	Complete clinical response; no flare-ups reported on 4 mg/day	NR
S17-P1	S17	High-potency topical corticosteroids	Recurrent but improved with topical corticosteroids	120
S18-P1	S18	Azathioprine 75 mg/day + methylprednisolone 40 mg/day → improved in 2 weeks; discharged on methylprednisolone 28 mg/day + azathioprine 75 mg/day; tapered to 16 mg/day by 6 months	Stable control during taper	6
S19-P1	S19	Prednisone 20 mg/day for 2 weeks → recurrence after discontinuation; prednisone 30 mg/day for 2 weeks with gradual taper → frequent relapses; dapsone 50 mg/day started as maintenance	Lesions resolved at 2-month follow-up on dapsone; no relapses to date	2
S20-P1	S20	Prednisolone 10 mg/day → improved; tapered off in 2 months; methotrexate 8 mg/week added for prevention	Remission maintained on methotrexate	36

Legend: NR indicates not reported in the source publication. Treatment and response/relapse summaries are transcribed from narrative descriptions; cases may have sequential/concomitant therapies. Follow-up is shown in months when reported.

Abbreviations: NR, not reported.

Supplementary Table S4. JBI critical appraisal of included reports (report-level)

S4a. JBI critical appraisal of case reports

Study ID	First author	Year	n (patient-level)	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8	Overall appraisal
S03	Launay	2000	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S04	Sakuma-Oyama	2003	1	Y	Y	Y	Y	Y	Y	Y	Y	Include
S05	Tsunemi	2005	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S06	Tanglertsampan	2007	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S07	Nakajima	2009	1	Y	Y	Y	Y	Y	Y	Y	Y	Include
S08	Kiorpelidou	2011	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S09	Sugiyama	2013	1	Y	Y	Y	Y	Y	Y	Y	Y	Include
S10	Li	2013	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S11	Sawada	2016	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S12	Riyaz	2016	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S14	Lin	2021	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S15	Uehara	2022	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S16	Gisondi	2023	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S17	Iinuma	2023	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S18	Li	2023	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S19	Rodríguez-Troncoso	2025	1	Y	Y	Y	Y	Y	Y	U	Y	Include
S20	Ohnishi	2025	1	Y	Y	Y	Y	Y	Y	U	Y	Include

S4b. JBI critical appraisal of case series

Study ID	First author	Year	n (patient-level)	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8	Q9	Q10	Overall appraisal
S01	Chen	1994	3	U	Y	Y	U	U	Y	Y	Y	U	U	Include
S02	Chen	1995	0	U	U	U	U	U	U	U	U	U	U	Retained; no unique cases
S13	Quijano-Gomero	2019	4	U	Y	Y	U	U	Y	Y	Y	Y	Y	Include

Report-level JBI critical appraisal (S4a: case reports; S4b: case series). “n (patient-level)” indicates the number of patients in the final patient-level dataset used for Tables 1–3. S01 and S02 were appraised separately; S02 is a duplicate/companion report of S01 and contributes no unique cases (n=0).
Notes: Y=yes; N=no; U=unclear; NA=not applicable.

Abbreviations: JBI, Joanna Briggs Institute.

PRISMA 2020 Main Checklist

Topic	No.	Item	Location where item is reported
TITLE			
Title	1	Identify the report as a systematic review.	Title page
ABSTRACT			
Abstract	2	See the PRISMA 2020 for Abstracts checklist	Abstract
INTRODUCTION			
Rationale	3	Describe the rationale for the review in the context of existing knowledge.	Introduction, paragraphs 1–3
Objectives	4	Provide an explicit statement of the objective(s) or question(s) the review addresses.	Introduction, final paragraph
METHODS			
Eligibility criteria	5	Specify the inclusion and exclusion criteria for the review and how studies were grouped for the syntheses.	Materials and Methods, Eligibility criteria
Information sources	6	Specify all databases, registers, websites, organisations, reference lists and other sources searched or consulted to identify studies. Specify the date when each source was last searched or consulted.	Materials and Methods, Information sources and search strategy
Search strategy	7	Present the full search strategies for all databases, registers and websites, including any filters and limits used.	Supplementary Table S1
Selection process	8	Specify the methods used to decide whether a study met the inclusion criteria of the review, including how many reviewers screened each record and each report retrieved, whether they worked independently, and if applicable, details of automation tools used in the process.	Materials and Methods, Study selection and duplicate handling
Data collection process	9	Specify the methods used to collect data from reports, including how many reviewers collected data from each report, whether they worked independently, any processes for obtaining or confirming data from study investigators, and if applicable, details of automation tools used in the process.	Materials and Methods, Study selection and duplicate handling; Data extraction and variables
Data items	10a	List and define all outcomes for which data were sought. Specify whether all results that were compatible with each outcome domain in each study were sought (e.g. for all measures, time points, analyses), and if not, the methods used to decide which results to collect.	Materials and Methods, Data extraction and variables; Results; Tables I–III
	10b	List and define all other variables for which data were sought (e.g. participant and intervention characteristics, funding sources). Describe any assumptions made about any missing or unclear information.	Materials and Methods, Data extraction and variables
Study risk of bias assessment	11	Specify the methods used to assess risk of bias in the included studies, including details of the tool(s) used, how many reviewers assessed each study and whether they worked independently, and if applicable, details of automation tools used in the process.	Materials and Methods, Quality appraisal and synthesis; Supplementary Table S4

Topic	No.	Item	Location where item is reported
Effect measures	12	Specify for each outcome the effect measure(s) (e.g. risk ratio, mean difference) used in the synthesis or presentation of results.	Not applicable; descriptive synthesis only
Synthesis methods	13a	Describe the processes used to decide which studies were eligible for each synthesis (e.g. tabulating the study intervention characteristics and comparing against the planned groups for each synthesis (item 5)).	Materials and Methods, Eligibility criteria; Study selection and duplicate handling
	13b	Describe any methods required to prepare the data for presentation or synthesis, such as handling of missing summary statistics, or data conversions.	Materials and Methods, Data extraction and variables; Quality appraisal and synthesis
	13c	Describe any methods used to tabulate or visually display results of individual studies and syntheses.	Materials and Methods, Quality appraisal and synthesis; Figure I; Figure II; Tables I–IV; Supplementary Tables S2–S4
	13d	Describe any methods used to synthesize results and provide a rationale for the choice(s). If meta-analysis was performed, describe the model(s), method(s) to identify the presence and extent of statistical heterogeneity, and software package(s) used.	Materials and Methods, Quality appraisal and synthesis
	13e	Describe any methods used to explore possible causes of heterogeneity among study results (e.g. subgroup analysis, meta-regression).	Not applicable; no formal heterogeneity analyses performed
	13f	Describe any sensitivity analyses conducted to assess robustness of the synthesized results.	Not applicable
Reporting bias assessment	14	Describe any methods used to assess risk of bias due to missing results in a synthesis (arising from reporting biases).	Materials and Methods, Quality appraisal and synthesis
Certainty assessment	15	Describe any methods used to assess certainty (or confidence) in the body of evidence for an outcome.	Materials and Methods, Quality appraisal and synthesis
RESULTS			
Study selection	16a	Describe the results of the search and selection process, from the number of records identified in the search to the number of studies included in the review, ideally using a flow diagram.	Results, Study selection; Figure I
	16b	Cite studies that might appear to meet the inclusion criteria, but which were excluded, and explain why they were excluded.	Results, Study selection; Figure I
Study characteristics	17	Cite each included study and present its characteristics.	Results, Included studies and unique cases; Supplementary Table S2
Risk of bias in studies	18	Present assessments of risk of bias for each included study.	Supplementary Table S4

Topic	No.	Item	Location where item is reported
Results of individual studies	19	For all outcomes, present, for each study: (a) summary statistics for each group (where appropriate) and (b) an effect estimate and its precision (e.g. confidence/credible interval), ideally using structured tables or plots.	Supplementary Tables S3a–c; Tables I–III
Results of syntheses	20a	For each synthesis, briefly summarise the characteristics and risk of bias among contributing studies.	Results, Included studies and unique cases; Supplementary Tables S2 and S4
	20b	Present results of all statistical syntheses conducted. If meta-analysis was done, present for each the summary estimate and its precision (e.g. confidence/credible interval) and measures of statistical heterogeneity. If comparing groups, describe the direction of the effect.	Not applicable; no meta-analysis performed
	20c	Present results of all investigations of possible causes of heterogeneity among study results.	Not applicable
	20d	Present results of all sensitivity analyses conducted to assess the robustness of the synthesized results.	Not applicable
Reporting biases	21	Present assessments of risk of bias due to missing results (arising from reporting biases) for each synthesis assessed.	Materials and Methods, Quality appraisal and synthesis
Certainty of evidence	22	Present assessments of certainty (or confidence) in the body of evidence for each outcome assessed.	Materials and Methods, Quality appraisal and synthesis
DISCUSSION			
Discussion	23a	Provide a general interpretation of the results in the context of other evidence.	Discussion, paragraphs 1–8
	23b	Discuss any limitations of the evidence included in the review.	Discussion, strengths and limitations paragraph
	23c	Discuss any limitations of the review processes used.	Discussion, strengths and limitations paragraph
	23d	Discuss implications of the results for practice, policy, and future research.	Discussion, final paragraph
OTHER INFORMATION			
Registration and protocol	24a	Provide registration information for the review, including register name and registration number, or state that the review was not registered.	Materials and Methods, Study design and registration; Ethics declarations & trial registry information
	24b	Indicate where the review protocol can be accessed, or state that a protocol was not prepared.	Materials and Methods, Study design and registration; PROSPERO registration record
	24c	Describe and explain any amendments to information provided at registration or in the protocol.	Materials and Methods, Study design and registration
Support	25	Describe sources of financial or non-financial support for the review, and the role of the funders or sponsors in the review.	Acknowledgements

Topic	No.	Item	Location where item is reported
Competing interests	26	Declare any competing interests of review authors.	Disclosure statements
Availability of data, code and other materials	27	Report which of the following are publicly available and where they can be found: template data collection forms; data extracted from included studies; data used for all analyses; analytic code; any other materials used in the review.	Data availability statement; Supplementary Tables S1–S4

PRIMSA Abstract Checklist

Topic	No.	Item	Reported?
TITLE			
Title	1	Identify the report as a systematic review.	Yes
BACKGROUND			
Objectives	2	Provide an explicit statement of the main objective(s) or question(s) the review addresses.	Yes
METHODS			
Eligibility criteria	3	Specify the inclusion and exclusion criteria for the review.	Yes
Information sources	4	Specify the information sources (e.g. databases, registers) used to identify studies and the date when each was last searched.	Yes
Risk of bias	5	Specify the methods used to assess risk of bias in the included studies.	Yes
Synthesis of results	6	Specify the methods used to present and synthesize results.	Yes
RESULTS			
Included studies	7	Give the total number of included studies and participants and summarise relevant characteristics of studies.	Yes
Synthesis of results	8	Present results for main outcomes, preferably indicating the number of included studies and participants for each. If meta-analysis was done, report the summary estimate and confidence/credible interval. If comparing groups, indicate the direction of the effect (i.e. which group is favoured).	Yes
DISCUSSION			
Limitations of evidence	9	Provide a brief summary of the limitations of the evidence included in the review (e.g. study risk of bias, inconsistency and imprecision).	Yes
Interpretation	10	Provide a general interpretation of the results and important implications.	Yes
OTHER			
Funding	11	Specify the primary source of funding for the review.	Yes
Registration	12	Provide the register name and registration number.	Yes

From: Page MJ, McKenzie JE, Bossuyt PM, Boutron I, Hoffmann TC, Mulrow CD, et al. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. MetaArXiv. 2020, September 14. DOI: 10.31222/osf.io/v7gm2. For more information, visit: www.prisma-statement.org