Supplementary Table 1.

Thirteen RCEV cases retrieved from the literature.

Abbreviations: HP: histopathology; F: female; M: male; m: months; w: weeks; ESR: erythrocyte sedimentation rate; CPR: C-Reactive Protein; IgE: Immunoglobulin E; ANA: antinuclear antibodies; ANCA: anti-neutrophil cytoplasmic antibodies; CT: computerized tomography; LDH: lactate dehydrogenase; BID: two times a day.

Nº	Author, year	Sex-Age	Duration	Localization	Description	HP	Blood tests	Treatment	Follow-up
1	Chen 1994(1)	56-F	6m	Generalized	Purpuric papular pruritic lesions, gingivitis, periorbital and perioral edema	Necrotizing vasculitis of small vessels with almost exclusive eosinophil infiltration into vessel walls	ESR 10mm/h; IgE <29ug/L; Eosinophilia $1.3x10^9$/L. No other significant abnormalities (ANCA negative).	Prednisone daily 60mg, then alternate-day prednisone because of recurrence. Hydroxyurea to better control.	2 years without lesions.
2	Chen 1994(1)	18-F	12m	Generalized	Pruritic erythematous and purpuric papules,	Necrotizing vasculitis with marked perivascular eosinophil infiltration	ESR 112mm/h; IgE 218ug/L; Eosinophilia $3.6x10^9$/L. No	Prednisone daily 60mg, but recurred with mucositis. Maintained on	23 years with out lesions.

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					angioedema of hands and periorbital area, gingivitis		other significant abnormalities (ANA negative).	alternate-day methylprednisolone 8 to 16mg to control skin disease	
3	Chen 1994(1)	17-F	12m	Generalized	Pruritic erythematous and purpuric papules, recurrent episodes of periorbital and palmoplantar swelling. Axillary, cervical and inguinal lymph node enlargement	No malignancy or vasculitis was found on lymph nodes; eosinophilic infiltration was observed in the spleen. Skin biopsies with necrotizing vasculitis with striking perivascular eosinophil infiltration.	ESR 61mm/h; IgE 59,280ug/L; Eosinophilia $6.2x10^9$/L. No other significant abnormalities (ANA negative).	Topical glucocorticoids, oral antihistamines.	17 years with out lesions.
4	Launay 2000(14)	81 - F	2w	Lower limbs	Pruritic, infiltrating, necrotic purpuric and papular skin lesions.	Almost exclusively eosinophilic, perivascular infiltration throughout the dermis, with out any leukocytoclastic vasculitis or amyloid deposit.	Eosinophilia 3.9 $x10^9$/L; ESR 70mm/h; CRP 48.8mg/L; fibrinogen 5.1 g/L. No other	1mg/kg prednisone daily for three weeks. Tapered to 10mg for four months. When reduced to <5mg, lesions recurred; then,	One year with out recurrence.

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					Angioedema of right hand.	Necrotizing vasculitis of small artery in deep dermis, with almost exclusive infiltration by eosinophils. IHC positive for eosinophil-derived neurotoxin, eosinophil peroxidase and eosinophil cationic protein in walls of vessels.	significant abnormalities (ANCA negative).	patient was under 5mg prednisolone daily.	
5	Sakuma-Oyama 2003(13)	27 - F	2m	Generalized	Edematous swelling of her face and fingers, palpable purpuric papules on both legs and urticarial plaques on the palms and trunk. Low-grade fever.	Fibrinoid necrosis of dermal vessels with marked infiltration of eosinophils and scattered lymphocytes. Elastica–van Gieson staining revealed that the process was limited to dermal veins. Direct immunofluorescence showed no evidence of immunoglobulin or complement binding.	Eosinophilia 7.2 $x10^9$/L; C1q and C3 complement $27x10^3$ug/L, Antinuclear antibodies 1:320. No other significant abnormalities.	Prednisolone 20mg/day. After tapering to 5mg/day, lesions and laboratory relapsed. Then betamethasone 1.5mg/day with sutaplast tosilate 300mg/day. Finally only sutaplast tosilate 300mg/day.	Two year with out symptoms and stable eosinophils with sutaplast tosilate.

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6	Tsunemi 2005(9)	53-F	36m	Lower limbs	Pruritic erythematous and urticarial plaques with recurrence.	Infiltration of many eosinophils and some lymphocytes around the dermal small vessels and destructive alteration of the vessels. There were few neutrophils and no leukocytoclasia	No significant abnormalities (ANA negative).	Prednisone daily 15mg, with dramatic improvement. Tapered to 7.5mg of prednisolone.	No recurrence of lesions.
7	Tanglertsampan 2007(8)	53 - M	6y	Generalized	Recurrent pruritic papules, nodules, and ulcers of the face, scalp, and hands	Dense mixed inflammatory infiltration of lymphocytes, histiocytes, eosinophils and extravasated red blood cells around necrotic areas. Also collagen fibers coated with eosinophilic granules. Fibrin thrombi and deposition of eosinophilic fibrinoid material in the vascular lumens and walls, respectively. No leukocytoclasia.	No significant abnormalities (ANCA negative).	Prednisolone 60mg/day. Recurrence after discontinuation. Indomethacin 75mg/day, then increased to 150mg/day with omeprazole 20mg/day.	Good response to indomethacin in 1 to 2 weeks.

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8	Kiorpelidou 2011(15)	82 - F	2m	Extremities	Multiple urticarial, polycyclic, annular, erythematous papules, and plaques, with central clearing and scales.	Moderately dense perivascular and interstitial infiltrate in the upper dermis of plentiful eosinophils. Endothelial swelling, intraluminal fibrin, eosinophils within vessel walls and eosinophilic dust around them. Direct immunofluorescence studies were negative.	ESR 107mm/h; CRP 30.1mg/L; Chronic periaortitis on abdomen CT-scan. No other significant abnormalities (ANCA negative).	0.5mg/kg/day methylprednisolone oral with 1mg/day colchicine. After one month, patient was discontinued of the treatment and lesions relapsed. The oral corticosteroids were restarted, and tapered over three months.	Twelve months with out recurrence of lesions.
9	Palazzolo 2012(5)	24 - F	2w	Trunk	Itchy erythematous-brownish, exudative papules within a large plaque, with desquamation, in the left sub mammary region.	Necrotizing vasculitis with eosinophils infiltrating the small vessels. No leukocytoclasia.	Eosinophilia 0.6 x10^9/L. No other significant abnormalities (ANCA negative).	High-potency topical corticosteroids.	1 month with out recurrence of lesions.
10	Sugiyama 2013(12)	80 - F	1m	Generalized	Multiple purpuric patches (not	Dermal vasculitis accompanied by	Eosinophilia 10.1 x10^9/L;	30mg prednisolone daily, tapered over	No adverse effects with

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					palpable) on her palms, the lower extremities and trunk. Also fever.	inflammatory cell infiltration, consisting largely of eosinophils and lymphocytes.	LDH 314 U/L; Antinuclear antibodies 1:80. No other significant abnormalities (ANCA negative).	several months. When reduced to 15mg daily, symptoms recurred. Oral tacrolimus 2 mg once daily was combined with 20mg prednisolone, and then tapered to 2.5mg daily.	tacrolimus 2mg/day. No recurrence of lesions.
11	Li 2013(4)	57 - M	1m	Lower limbs	Started as itchy needlepoint- to millet-sized papules; then purpuric plaques with angioedema, some of them necrotic.	Perivascular infiltration of numerous eosinophils and few neutrophils, some into vessel walls, in the upper and deep dermis, and in the sub- cutaneous tissue. Thickening of the vessel walls, numerous extravascular erythrocytes, fibrin thrombi in the lumens, and fibrinoid degeneration.	Eosinophilia $3.4x10^9$/L; ESR 32 mm/h; CRP 14.5mg/L; IgE 658.3ug/L. No other significant abnormalities (ANCA negative).	1mg/kg/day prednisone combined with glycyrrhizin 150mg/day. Tapered over one month to 10mg daily. Any attempt to decrease was followed with relapse of symptoms.	Four months with out recurrence of lesions.

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12	Fernandez 2014(7)	36 - F	6m	Generalized	Pruritic erythematous and purpuric ulcerative plaque, with edema and pain in right dorsal aspect of foot. Also similar plaques in extremities and trunk.	Dense perivascular infiltration of eosinophils, some lymphocytes with fibrinoid necrosis in wall vessels, and edema of adjacent tissue.	Eosinophilia $4.3x10^9$/L; Antinuclear antibodies 1/120; No other significant abnormalities.	Prednisone 50mg/day. Recurrence after tapering doses of 15mg/day, therefore, patient was maintained with 20mg/day.	Three months with out recurrence.
13	Sawada 2016(2)	55 - F	12m	Upper limb	Bilateral recurrent pruritic erythemas with induration in palms	Infiltration of eosinophils within and around the walls mainly of arterioles or venulae in in the dermal superficial vascular plexus. Fibrin deposits in the vessel walls with occlusions of lumen of the vessels.	Eosinophilia 1.9 $x10^9$/L. Mild elevation of CRP, fibrinogen and D-dimer. No other significant abnormalities (ANCA negative).	Patient concurred with Budd-Chiari syndrome, and lesions resolved after angioplasty. No systemic steroids were used.	Lesions disappeared after a week of treatment.

Comentado [DBML(1]:

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