Poikilodermatous plaque-like hemangioma: Case series of a newly defined entity

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Background: We present a distinctive type of acquired vascular proliferation, for which we propose the name of poikilodermatous plaque-like hemangioma.

Objective: The aim of this study was to summarize the clinical and histopathologic features in a case series of poikilodermatous plaque-like hemangioma.

Methods: Sixteen cases were identified from the routine clinical and referral practices of the authors. Clinical characteristics, including demographic details and clinical morphology, were collated. The salient histopathologic features, including immunohistochemical staining results, were summarized.

Results: The lesions were usually solitary erythematous-to-violaceous poikilodermatous plaques on the lower extremities and pelvic girdle, with an indolent clinical course. Mean age of affected patients was 72 (range 58-80) years, and there was a male predominance. Histology comprised a distinctive band-like proliferation of vascular channels suggestive of postcapillary venules within the superficial dermis with a background of fibrosis, edema, and loss of elastic fibers. Despite the clinical atrophic appearance, acanthosis was a frequent finding.

Limitations: Retrospective study.

Conclusion: Poikilodermatous plaque-like hemangioma is a distinctive and previously undescribed vascular proliferation defined by a constellation of consistent and reproducible clinical and histologic features. (J Am Acad Dermatol 2019;81:1257-70.)

Key words: acquired hemangioma; mycosis fungoides; poikilodermatous plaque-like hemangioma; vascular proliferation.

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Classification and diagnosis of cutaneous vascular proliferations can be challenging; a wide variety of clinicopathologic entities often display overlapping features. Furthermore, the vascular nature of a lesion might not be readily apparent on clinical appearance alone. The dermatologic causes of poikilodermatous clinical morphology are wide ranging, including genodermatoses, inflammatory dermatoses, acrodermatitis chronica atrophicans secondary to *Borrelia* infection, connective tissue diseases, and physical agents, such as ionizing radiation and cutaneous T-cell lymphoma (CTCL), most notably mycosis fungoides (MF). We herein describe the clinicopathologic features of 16 cases of a distinctive type of acquired vascular proliferation, for which we propose the name of poikilodermatous plaque-like hemangioma. This type typically presents as a solitary (or occasionally multiple) erythematous-to-violaceous poikilodermatous plaque on nonsun-damaged skin with a distinctive band-like vascular proliferation in the superficial dermis on histology.

CAPSULE SUMMARY

- The diagnosis of vascular lesions can be challenging to clinicians and pathologists.
- We describe and define a novel entity that presents as an atrophic violaceous plaque with a characteristic band-like vascular proliferation on histology. Poikilodermatous plaque-like hemangioma is a distinctive, acquired vascular lesion with a benign clinical course.

MATERIALS AND METHODS

A total of 16 cases were identified. Most of the cases (n = 13) were retrieved from the consultation files of 1 of the authors (Dr Calonje) or were sent to the St. John's Institute Department of Dermatopathology for second opinion. Additional cases were retrieved from the consultation files of 2 of the other authors (Dr Carr and Dr Taibjee). The clinical picture was similar in all cases. Patients presented with a solitary atrophic erythematous or violaceous plaque or, in rare cases, slightly raised plaque on the extremities or buttocks. We reviewed 4-μL–thick hematoxylin-eosin–stained sections for the presence or absence of the following features: proliferation of thin-walled vascular channels with a plaque-like arrangement in the superficial dermis, inflammation, lymphocyte atypia, background dermal changes (including edema, fibrosis, and elastic fibers), and epidermal changes (including atrophy, ulceration, hyperkeratosis, parakeratosis, acanthosis, spongiosis, exocytosis of lymphocytes, and vacuolar interface tissue reaction). Immunohistochemical studies were available or performed on 4-μm–thick formalin-fixed, paraffin-embedded tissue sections. Monoclonal antibodies used included those specific to CD3, CD4, CD8, CD31, erythroblast transformation-specific–related gene (ERG), smooth muscle actin, glucose transporter 1 (GLUT-1) and D2-40 antibody. Elastic van Gieson stain was performed to assess elastic fiber density. Multiple biopsies taken over the course of several years were available for 2 cases, enabling assessment of the evolution of the histologic features. T-cell gene rearrangement studies were carried out for 2 patients, and 3 patients were tested for antibodies against *Borrelia* subspecies (IgM and IgG).

The hematoxylin-eosin and immunohistochemically stained slides were reviewed independently by 2 authors (Dr Semkova and Dr Calonje). Clinical data and follow-up information including sex, age, site, size, duration, and follow-up course were obtained from the referring pathologist or dermatologist.

RESULTS

Clinical features

Patient age ranged from 58 to 80 (mean 72) years. The lesion duration, available for 13 patients, ranged from 2 months to 6 years (mean 2.5 years). There was a male predominance (male-to-female ratio 14:2). Lesions were typically described as slowly growing, asymptomatic erythematous or violaceous atrophic plaques, ranging in size from 2 to 7 (mean 5.8) cm in diameter. Superficial scale was clinically noted in 4 patients. Almost all lesions were located on the lower extremities (hip, thigh, and lower leg); 1 lesion arose on the shoulder (Fig 1). In all, 14 patients had a single plaque, 1 patient had 2 plaques on the lower extremities, and 1 patient had 2 plaques on the buttock and 1 on the shoulder. Two of these lesions were biopsied and showed similar histologic features. Refer to Table I for a summary of the clinical features.

The most frequently suggested clinical diagnosis was mycosis fungoides (CTCL), followed by fixed drug reaction or a granulomatous condition. Morphea, spider bite, and lupus were considered in 1 case each. One patient reported a possible preceding trauma during hiking, but there were no clear precipitating factors in the remaining patients. There were no systemic symptoms or any relevant underlying medical conditions. IgG and IgM antibodies against *Borrelia* subspecies were negative in

Abbreviations used:

ACA:	acrodermatitis chronica atrophicans
AEH:	acquired elastotic hemangioma
CTCL:	cutaneous T-cell lymphoma
ERG:	erythroblast transformation-specific–related gene
GLUT-1:	glucose transporter 1
MF:	mycosis fungoides

the 3 patients tested. Treatment history before biopsy was available for 7 patients: 3 received no treatment, 2 were treated with topical corticosteroids, 1 was treated with a topical combination of corticosteroid with an antifungal, and 1 was on oral prednisolone for concomitant sarcoidosis. The lesions persisted or progressed despite treatment in all cases. Follow-up ranged from 24 to 120 (mean 46.6) months and revealed a typical natural history of initial growth followed by long-term persistence with benign clinical course.

Histopathologic features

A summary of the histopathologic features is presented in Table II. At scanning magnification, all cases showed strikingly similar appearances. The main feature was a diffuse band-like vascular proliferation in the papillary dermis and superficial reticular dermis without a clear Grenz zone. This proliferation comprised numerous thin-walled vascular channels with features of postcapillary venules lined by a single layer of endothelial cells, with only a focal lobular arrangement. The endothelial cells had benign morphology; mitotic activity was absent. A small number of background lymphatic vessels were seen in some cases, which was confirmed by positive staining with D2-40 antibody.

Variable epidermal changes were present in all cases and included mild-to-moderate acanthosis (n = 10), hyperkeratosis (n = 16), focal parakeratosis (n = 3), hypergranulosis (n = 3), spongiosis (n = 5), exocytosis of lymphocytes (n = 6), and an interface vacuolar tissue reaction (n = 2). Significant atrophy was not present, but some cases showed focal mild thinning of the epidermis with flattening of the rete ridges. Additional features noted included dermal fibrosis (n = 12) and dermal edema (n = 12). Most cases showed mild perivascular or interstitial chronic inflammation, consisting of lymphocytes and histiocytes. Lymphocytic atypia was absent. There was no evidence of alignment of lymphocytes at the basal cell layer (or tagging). Red cell extravasation was noted in 4 cases, but there was no evidence of hemosiderin deposition. Solar elastosis was absent. Dermal elastic fibers appeared lost or diminished within the zone of vascular proliferation, in contrast with the adjacent dermis (Figs 2-5).

Two patients had multiple biopsies over a span of 3 years. The initial biopsies showed a more prominent superficial dermal lymphohistiocytic infiltrate and moderate acanthosis. In contrast, subsequent biopsies showed only scattered inflammatory cells and mild acanthosis with focal thinning of the epidermis. The other histologic changes, in particular the vascular proliferation, were noted throughout.

Immunohistochemical studies with antibodies specific to CD31 (n = 6) or ERG (n = 6) highlighted the vascular proliferation. T-cell markers were present in 6 cases; in all these cases, the dermal lymphocytic subpopulation was composed of CD3-positive, CD2-positive, and CD5-positive T-cells with a predominance of CD4-positive cells over CD8-positive cells, with no loss of antigen expression. The infiltrate was negative for CD20 and CD30. The primary vascular proliferation did not stain with the D2-40 antibody (n = 6), but the antibody highlighted scattered background lymphatics. Smooth muscle actin antibody (n = 7) highlighted a rim of pericytes around the majority of vessels. The proliferation was also negative for GLUT-1 (n = 4; refer to Table II for summary of histologic features).

T-cell gene rearrangement studies were performed for 2 cases. One case showed no rearrangement. For the second case, 3 specimens were submitted for analysis. In 2 of the samples, 2 primers were polyclonal and 3 did not amplify because of poor DNA quality, and in the third sample, 3 primers did not amplify, 1 was polyclonal, and 1 was equivocal.

DISCUSSION

We describe a previously unreported vascular proliferation with distinctive and reproducible features. It usually presents as an indolent solitary erythematous or violaceous atrophic plaque during or after the 5th decade of life, predominantly in male patients, most commonly on the lower extremities; this proliferation demonstrated a benign clinical course during the available follow-up period (up to 6 years). Histologically, there is a characteristic superficial dermal band-like vascular proliferation comprising thin-walled vascular channels with features of postcapillary venules, lined by bland endothelial cells, with additional dermal fibrosis or edema. Epidermal changes, such as hyperplasia and spongiosis, might be evident, although atrophy is not a prominent feature histologically despite the poikilodermatous clinical appearance.

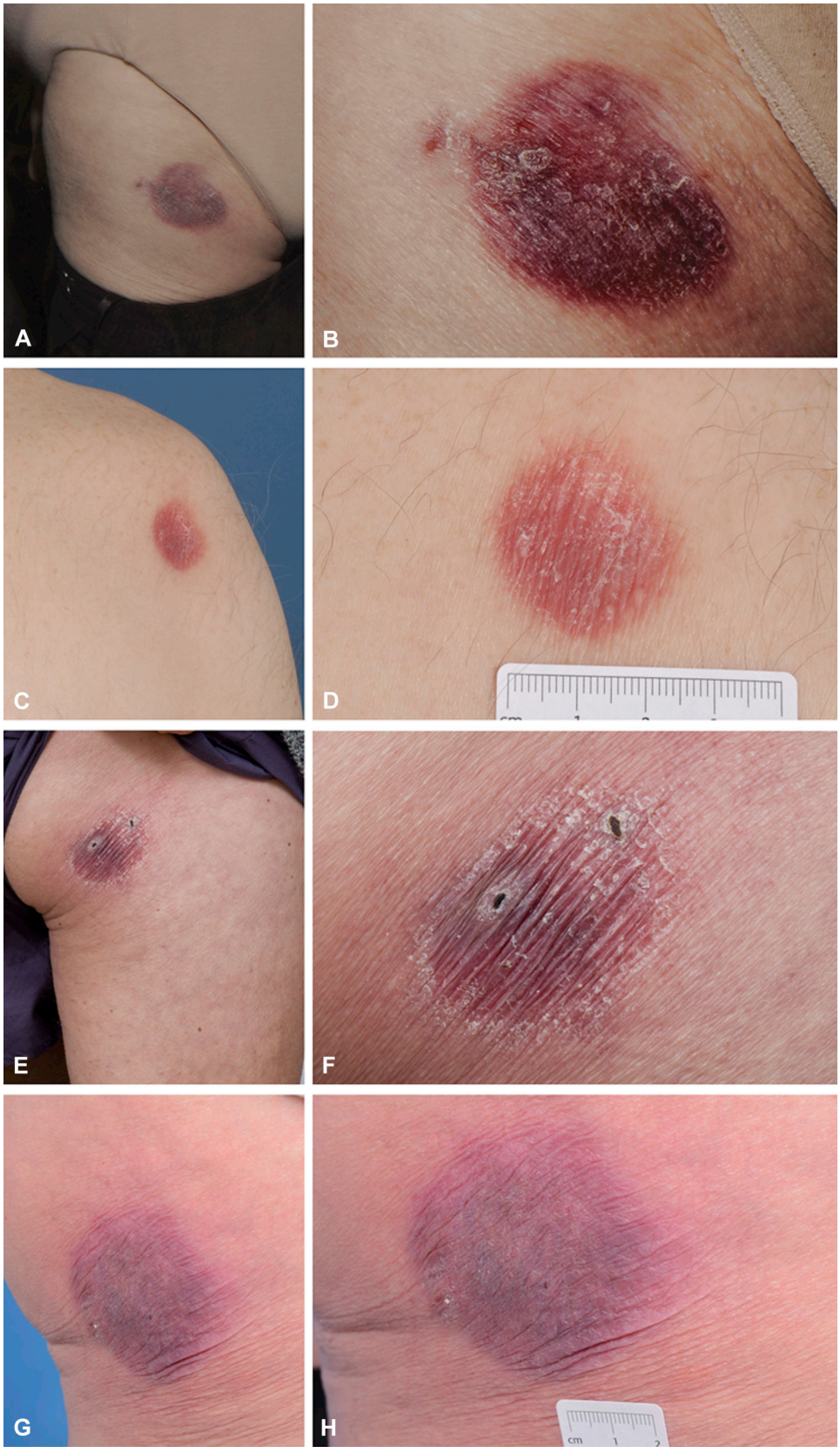

Fig 1. Clinical presentation. **A** and **B**, Case 3. **C** and **D**, Case 4. **E** and **F**, Case 8. **G** and **H**, Case 9.

Table I. Clinical characteristics

Case	Age, y	Sex	No. lesions	Site	Duration	Clinical information	Suggested clinical diagnosis	Ancillary studies
1	75	M	Single	Hip	NA	Fixed atrophic plaque	Mycosis fungoides or fixed drug eruption	Clonality studies
2	58	M	Single	Lower leg	2 years	Purple-red patch with central induration and superficial atrophy; could have been trauma triggered	Foreign body or insect bite reaction, mycosis fungoides, morphea	*Borrelia* antibody negative
3	76	F	Single	Thigh	6 years	Purplish atrophic plaque; history of myelodysplastic syndrome	NA	
4	79	M	Multiple	Shoulder, hip, buttock	5 years	Asymptomatic persistent indurated erythematous plaques, gradually increasing in size	Mycosis fungoides	
5	64	M	Single	Lower leg	3 years	Asymptomatic atrophic purplish plaque, 7 × 4 cm	Mycosis fungoides	*Borrelia* antibody negative
6	60	M	Single	Leg	1.5 years	Purple erythematous patch with evidence of epidermal atrophy but no scale, 2.5 cm; sometimes sore; comorbid pulmonary sarcoidosis	NA	Dermoscopy: hair-pin type vessels
7	75	M	Single	Thigh	1.5 years	Poikilodermatous patch	Mycosis fungoides	
8	79	M	Single	Thigh	1 year	Asymptomatic, mildly scaly, atrophic, purple erythematous patch, 6 × 6 cm	Mycosis fungoides, lupus	
9	69	F	Single	Hip, buttock	1 year	Well circumscribed purple erythematous plaque, tender on palpation, otherwise asymptomatic	Fixed drug eruption	
10	68	M	Single	Hip	2 years	Atrophic plaque, 7 × 6 cm	Granuloma, mycosis fungoides, morphea	
11	75	M	Multiple	Thigh, knee	NA	Several plaques with thickening and atrophy on the lower limbs		
12	75	M	Single	Buttock	6 years	Subtle striate telangiectatic erythema	Mycosis fungoides	Clonality studies
13	78	M	Single	Calf	3 years	Asymptomatic, persistent, erythematous plaque	Insect bite	*Borrelia* antibody negative, negative acid-fast bacilli culture
14	80	M	Single	Hip	8 months	Erythematous patch	Spider bite, granuloma	
15	74	M	Single	Calf	3 months	Well-defined erythematous patch with desquamation	NA	
16	71	M	Single	Calf	NA	Erythematous patch with minimal desquamation	NA	

NA, Not available.

Table II. Histologic features

Case	Age, y	Sex	Epidermal changes	Dermal infiltrate	Dermal fibrosis	Dermal edema	Special stains	Lymphocytic atypia	Additional features
1	75	M	Early lesion: hyperkeratosis, acanthosis, parakeratosis (focal), spongiosis (focal), exocytosis of lymphocytes; late lesion: very mild acanthosis	Early lesion: fairly prominent band-like lymphocytes, rare extravasated red blood cells; late lesion: sparse infiltrate, predominantly histiocytes and lymphocytes, rare extravasated red blood cells	Yes	Yes	Lymphocytes positive for CD2, CD3, CD4, CD5; scattered CD8	Minimal	
2	58	M	Hyperkeratosis acanthosis	Minimal	Yes	Yes	NA	No	Obstruction of sweat glands by fibrosis
3	76	F	Mild acanthosis, hyperkeratosis (focal), spongiosis, exocytosis of lymphocytes	Focal, mild-to-focal moderately dense lymphocytes and histiocytes, extravasated red blood cells	No	Yes	Positive for CD31, SMA, D2-40, ERG	No	
4	79	M	Hyperkeratosis, hypergranulosis	Lymphocytes and histiocytes	Yes	Yes	NA		
5	64	M	Hyperkeratosis, mild acanthosis	Scattered lymphocytes, sparse extravasated red blood cells	No	Yes	Positive for SMA		Mild decrease in elastic fibers
6	60	M	Hyperkeratosis, mild acanthosis	Scattered lymphocytes, sparse extravasated red blood cells	Yes		No		
7	75	M	Hyperkeratosis, mild spongiosis, vacuolar interface (focal), exocytosis of lymphocytes	Scattered lymphocytes	Yes	Yes	NA	No	
8	79	M	Hyperkeratosis (mild), parakeratosis (focal), exocytosis of lymphocytes	Mild-to-focally moderate lymphocytes and histiocytes	No	Yes	Positive for CD31, SMA, D2-40, ERG	No	
9	69	F	Hyperkeratosis, acanthosis	Scattered lymphocytes	Yes	Yes	Positive for CD31, SMA, D2-40, ERG; negative for GLUT-1		
10	68	M	Hyperkeratosis, parakeratosis (focal), spongiosis, exocytosis of lymphocytes, focal necrosis	Mild-to-moderately dense mixed inflammatory infiltrate	Yes	Yes	NA		
11	75	M	Hyperkeratosis, hypergranulosis, acanthosis	Scattered lymphocytes and mast cells	Yes	No	Positive for CD31, SMA, D2-40, ERG; negative for GLUT-1	No	

12	75	M	Hyperkeratosis	Mild-to-focally moderate lymphocytes and histiocytes	Yes	Yes	Positive for CD31, SMA, D2-40, ERG; negative for GLUT-1	No	Follicular plugging
13	78	M	Hyperkeratosis, hypergranulosis (focal), focal mild flattening of epidermis with loss of rete ridges	Scattered lymphocytes	Yes	Yes	Positive for CD31, SMA, D2-40, ERG; negative for GLUT-1		Follicular plugging
14	80	M	Hyperkeratosis, acanthosis, vacuolar interface (focal)	Scattered lymphocytes	Yes	No	NA	No	Subepidermal blister formation
15	74	M	Hyperkeratosis, acanthosis	Rare lymphocytes	No	Yes	NA	No	
16	71	M	Hyperkeratosis, acanthosis	No	No	Yes	NA	No	

ERG, Erythroblast transformation-specific–related gene; *GLUT-1*, glucose transporter 1; *NA*, not available; *SMA*, smooth muscle actin.

We propose the name poikilodermatous plaque-like hemangioma to emphasize the striking clinical resemblance and important distinction from poikilodermatous MF. Poikilodermatous MF, historically also referred to as poikiloderma atrophicans vasculare, parapsoriasis variegata, or mycosis fungoides—lichenoid type, is an uncommon presentation of CTCL.[1-3] Clinically, poikilodermatous MF presents with asymptomatic large hypopigmented or hyperpigmented patches or plaques with atrophy and telangiectasia located on the trunk or flexural areas. Histopathologically, poikilodermatous MF demonstrates features in common with other subtypes of MF, including an epidermotropic atypical lymphoid infiltrate, but with additional epidermal atrophy, basal vacuolar degeneration, pigment incontinence, and telangiectatic blood vessels. Both CD8-positive and CD4-positive variants have been reported.[2]

Features that aid in the distinction of poikilodermatous plaque-like hemangioma from poikilodermatous MF include the usual solitary nature of poikilodermatous plaque-like hemangioma and its predilection for the lower extremities. Histologically, the vascular proliferation is distinctive. The usual epidermal reaction is of acanthosis rather than atrophy, despite the clinical appearance, and although a mild lymphocytic infiltrate was noted in some of our cases, there was generally no cytologic atypia and no evidence of clonality for the 2 cases tested.

Other differential diagnoses are summarized in Table III.[1,2,4-12] Fixed drug eruption can resemble poikilodermatous plaque-like hemangioma, but this diagnosis requires a clear history of preceding drug intake and typically resolves on discontinuation of the culprit drug. Histologically, acute fixed drug reaction demonstrates an interface vacuolar degeneration with colloid body formation and marked pigment incontinence. Dyskeratotic keratinocytes and a mild-to-moderately dense lichenoid infiltrate are also seen. In late lesions, only pigment incontinence is seen and vascular proliferation is not a feature.

Acrodermatitis chronica atrophicans (ACA) might present with a similar clinical appearance.[4] However, although a localized form of ACA was considered in the differential diagnosis in 3 of our patients, *Borrelia* (IgM and IgG) antibody testing was negative. Furthermore, there was no clear history of tick bite or other symptoms and signs of Lyme disease. Histologically, ACA is characterized by dilatation and not proliferation of vascular channels and by more prominent, often band-like inflammation that includes lymphocytes, plasma cells, histiocytes, and macrophages.

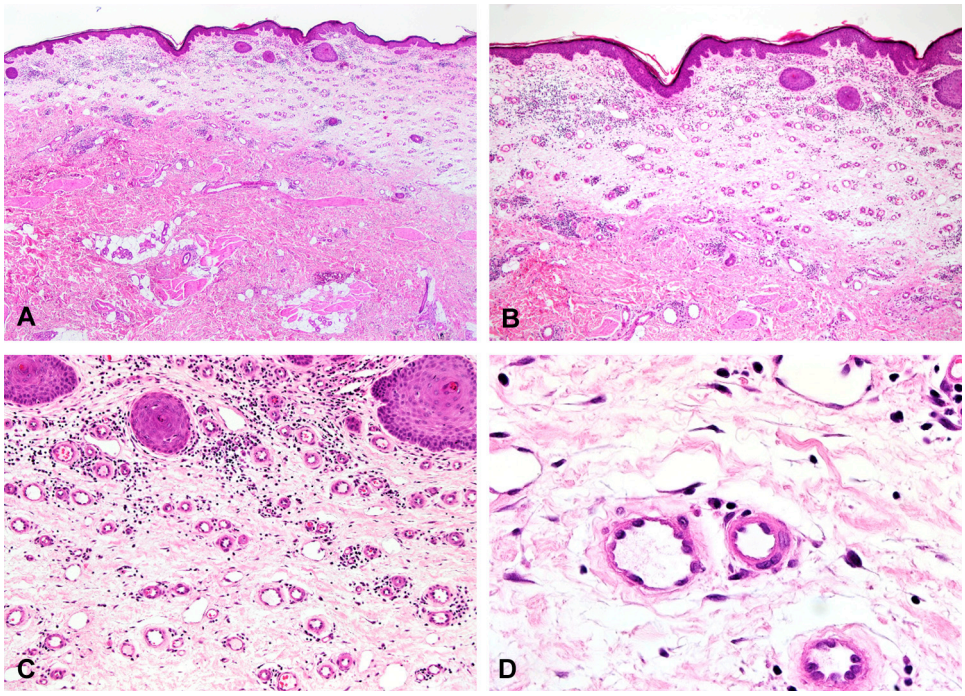

Fig 2. Histologic presentation of case 3. (**A**-**D**, Hematoxylin-eosin stain; original magnification: **A**, ×20; **B**, ×40; **C**, ×100; **D**, ×600.)

Clinically, poikilodermatous plaque-like hemangioma can resemble the pigmented purpuric dermatoses. These diseases represent a group with similar clinical presentation and identical histologic features. On the basis of their clinical manifestation these dermatoses are subclassified as progressive pigmentary dermatosis of Schamberg (bilateral, irregular, nonblanchable, brownish, purpuric macules with pinpoint petechiae), purpura annularis telangiectodes of Majocchi (bilateral, symmetrical, annular purpuric plaques with punctate telangiectasia in the border, predominantly in young female patients), pigmented purpuric lichenoid dermatitis of Gougerot and Blum (purpuric lichenoid papules), eczematid-like purpura of Doucas Kapetanakis (scaly and petechial purpuric macules and papules), and lichen aureus (solitary, well-circumscribed, golden-brown or erythematous patches or plaques often overlying a vein).[13] On histology, all variants show erythrocyte extravasation and hemosiderin deposition in the dermis with variable epidermal changes and dermal infiltrates.[6] In contrast, in poikilodermatous plaque-like hemangioma, there is a proliferation of vascular channels, and extravasated red blood cells with hemosiderin deposition not a feature. Moreover, clinically the plaques of poikilodermatous plaque-like hemangioma are well defined, atrophic, and without pinpoint petechiae.

Acquired elastotic hemangioma (AEH), an additional diagnostic consideration[7] also presents as a solitary, slowly growing, asymptomatic, erythematous-to-violaceous plaque, but in contrast with poikilodermatous plaque-like hemangioma, it is usually small and noted on sun-exposed areas, particularly the upper extremities and neck. Although initially reported to have a female predominance, further studies of AEH showed a roughly equal male-to-female ratio.[8,9] Histology of AEH is characterized by a proliferation of vascular channels in the superficial dermis in a band-like fashion with a Grenz zone. In contrast with poikilodermatous plaque-like hemangioma, the vessels in AEH are often dilated, a prominent solar elastosis is invariably present (lacking in our cases), and background fibrosis or edema is not a feature.[7-9] The histogenesis of the vessels in AEH might be a lymphatic lineage, as suggested by the positive D2-40 staining in 1 study,[8] although this was not reproducible in another study.[9] A further possible distinction between poikilodermatous plaque-like hemangioma and AEH is the loss of elastic fibers in poikilodermatous plaque-like hemangioma, demonstrable by Elastic van Gieson staining.

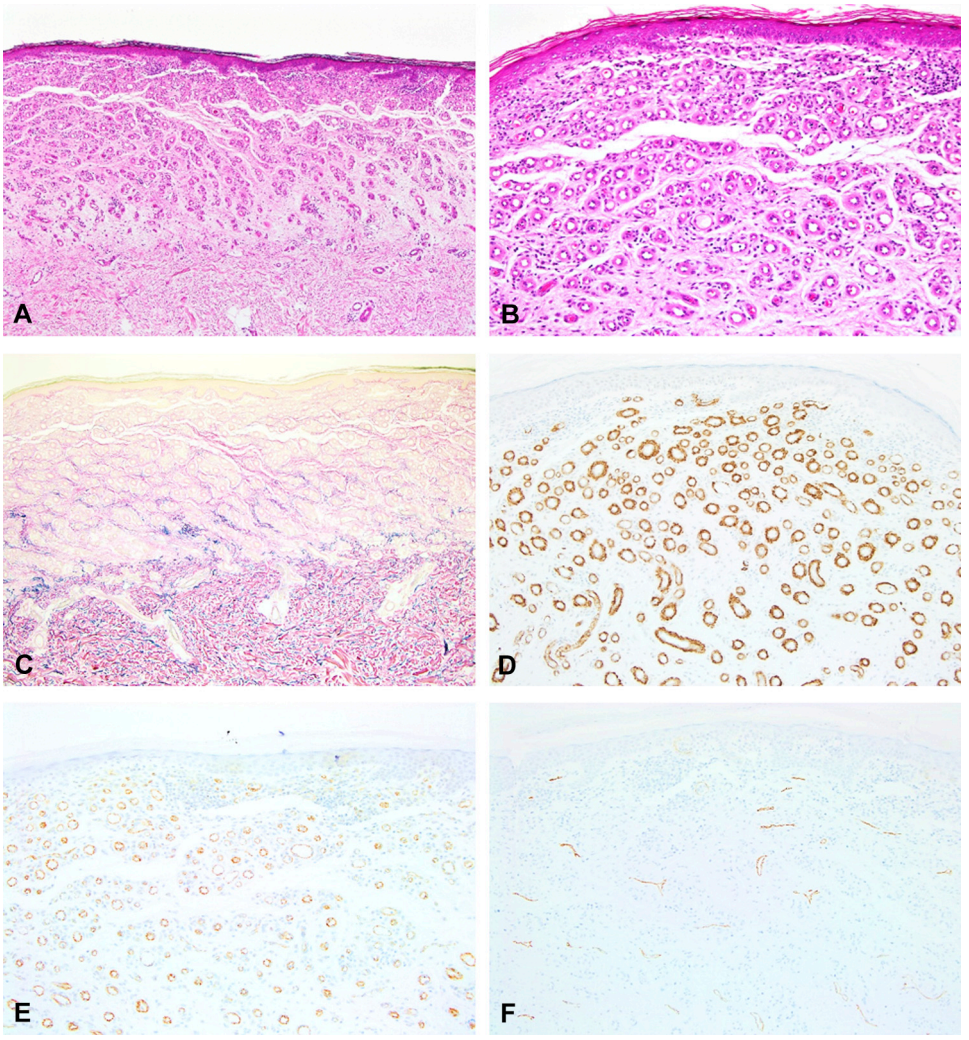

Fig 3. Histologic presentation of case 8. (**A** and **B**, Hematoxylin-eosin stain; **C**, Elastic van Gieson stain; **D**, smooth muscle actin; **E**, CD31; **F**, D2-40; original magnification: **A** and **C**, ×40; **B** and **D**-**F**, ×100.)

Microvenular hemangioma typically presents in younger patients with red or brown macules, plaques, or nodules in a more wide-ranging anatomical distribution. The histology also differs; vessels show a more dissecting pattern, with tracking along adnexae, and the characteristic infiltration of arrector pili muscles.[5]

Hobnail hemangioma (targetoid hemosiderotic hemangioma) presents with a violaceous central papule surrounded by a thin pale ring and an ecchymotic periphery. However, in many cases the classical targetoid appearance is absent and the morphology is of a nonspecific vascular lesion.[11] The age distribution of patients is wide and cases in children, including from birth, have been reported.[12] On histology, a biphasic vascular proliferation can be found in the dermis extending into the subcutaneous tissue with superficial ectatic vascular channels lined by hobnail endothelial cells and with deeper slit-like vessels dissecting through collagen bundles.[11] The vessels in this lesion type are believed to be predominantly of a lymphatic origin. In poikilodermatous plaque-like hemangioma, the lesions are larger and lack the characteristic zonal clinical appearance, and the vascular proliferation is superficial, does not have a hobnail endothelium, and represents mainly microvenules.

Malignant vascular neoplasms can also present as solitary plaques, although the histologic features are readily distinguishable. Kaposi sarcoma (early stage) is characterized by thin vascular lymphatic channels devoid of pericytes, intersecting the collagen bundles, and with accompanying lymphoplasmacytic inflammation.[14] Well-differentiated angiosarcoma

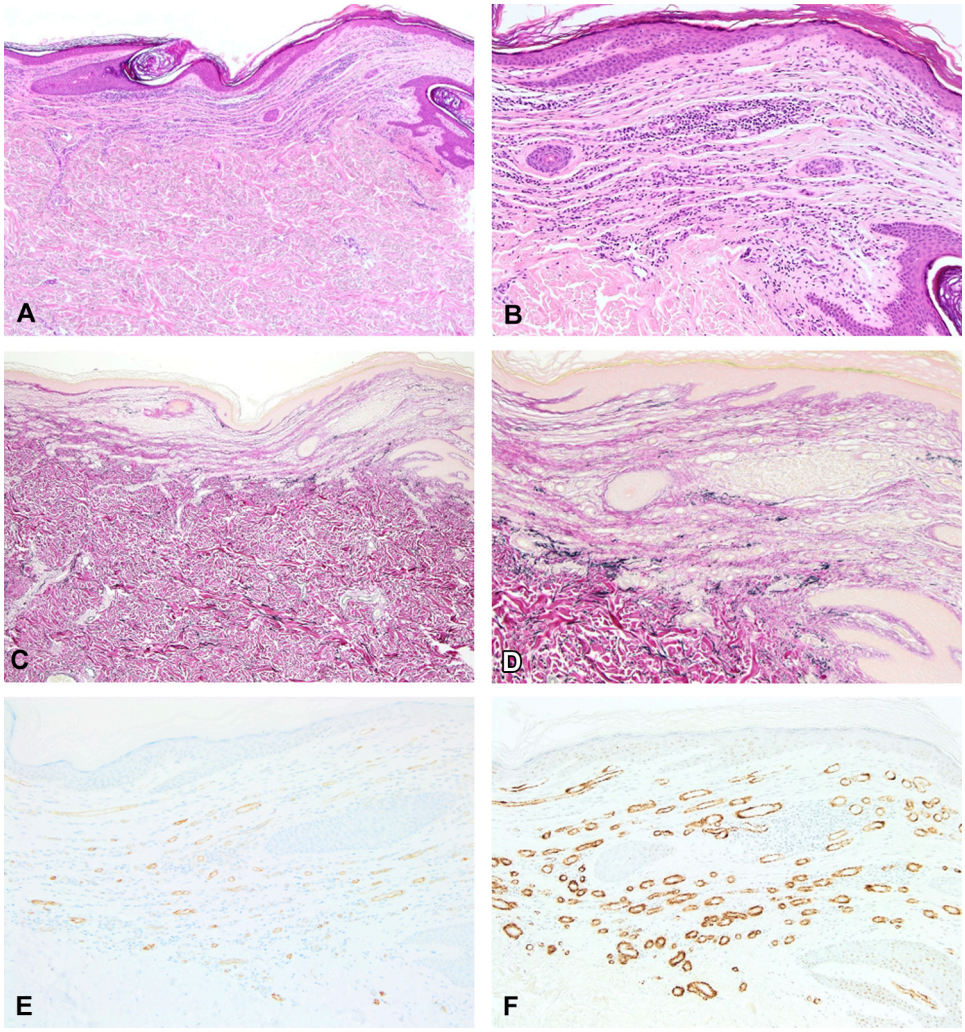

Fig 4. Histologic presentation of case 9. (**A** and **B**, Hematoxylin-eosin stain; **C** and **D**, Elastic van Gieson stain; **E**, CD31; **F**, smooth muscle actin; original magnification: **A** and **C**, ×40; **B** and **D**-**F**, ×100.)

shows dissecting irregular anastomosing vascular channels lined by atypical endothelial cells with mitoses and multilayering.

Last, a similar vascular proliferation has recently been described in cases of lichen simplex chronicus and prurigo nodularis on the knees and elbows.[10] However, in contrast with our cases, the clinical impression of these proliferations was of a hyperkeratotic neoplastic or inflammatory process, and additional histologic findings, such as syringosquamous metaplasia, melanocytic hyperplasia, and compressed and increased elastic fibers were also noted. The epidermal hyperplasia was generally more marked. The authors of this recent report speculated that their cases represented a reactive process involving the epidermis, vessels, and sweat ducts, likely resulting from chronic pressure and repeated mechanical stimulation, accounting for the anatomic location. Although the vascular proliferations in our cases had morphologic similarities, our patients had a different and distinctive clinical presentation and a more wide-ranging anatomic distribution. Furthermore, in none of our cases was chronic pressure suggested as a possible etiologic factor because many of the lesions were on nonpressure—bearing sites.

The etiology of poikilodermatous plaque-like hemangioma remains unclear. One patient in the presented series reported a possible insect bite or trauma during hiking before the development of the lesion, another reported a possible spider bite. There were no reported triggers in the remaining patients. The concomitant diseases also varied, with no consistent pattern to suggest a systemic cause. Interestingly, however, the vast majority of patients were living in the North of England. Although this

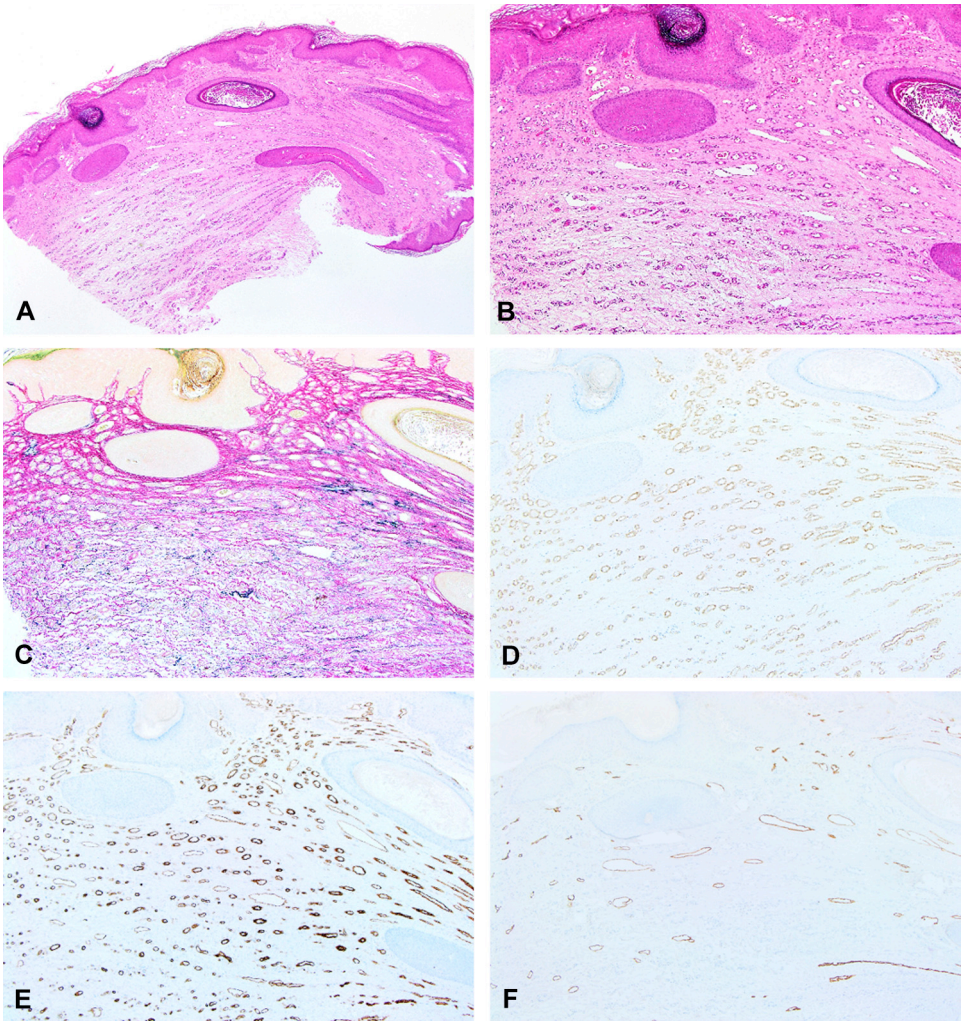

Fig 5. Histologic presentation of case 12. (**A** and **B**, Hematoxylin-eosin stain; **C**, Elastic van Gieson stain; **D**, smooth muscle actin; **E**, CD31; **F**, D2-40; original magnification: **A** and **C** ×20; **B** ×40; **D**-**F** ×100.)

might be considered to represent a referral bias for the biopsies, specimens for consultation are sent to our institution from all regions of the United Kingdom and roughly equal numbers come from the North and South. Hence, it could be speculated that the clustering of cases results from the specifics of the environment or an undetermined infective cause. Most lesions arose on sun-protected areas. The duration of lesions varied 1-6 years with indolent behavior, suggesting a benign clinical course. The reproducible clinical pattern of initial slow growth with persistence of the established lesion and the lack of response to anti-inflammatory treatment support the concept that poikilodermatous plaque-like hemangioma is a true hemangioma and not a reactive lesion.

Treatment does not appear to change the course of the disease and most of our patients did not receive any therapy after biopsy confirmation of the benign nature of the lesion. Most lesions did not progress or regress for many years. However, although a wait-and-see approach appears to be the most reasonable management, adequate treatment recommendations will require analysis of a larger series of patients.

In summary, we describe a new distinctive clinicopathologic entity, and propose the name poikilodermatous plaque-like hemangioma. Awareness and recognition of poikilodermatous plaque-like hemangioma should enable identification of new cases and further characterization. Clinically this condition could be confused with MF

Table III. Clinical and histologic differential diagnosis with discriminative features

Diagnosis	Clinical features	Histologic features
Poikilodermatous plaque-like hemangioma	Erythematous, clinically atrophic patch or plaque; usually solitary lesion; mean age 72 years; male predominance; lower extremities and pelvic girdle area; prolonged duration with minimal progression	Superficial dermal band-like vascular proliferation of postcapillary venules; epidermal changes (hyperplasia, rare focal atrophy, spongiosis, and exocytosis of lymphocytes); expansion of the papillary dermis with loss of elastic fibers, but no elastolysis; no Grenz zone; dermal fibrosis or edema; immunohistochemistry: CD31 and ERG highlight the vascular proliferation, D2-40 stains scattered lymphatics; Perls stain negative for hemosiderin
Clinical differential diagnoses		
Mycosis fungoides[1,2]	Hypopigmented or hyperpigmented patches or plaques with atrophy and telangiectasiae; usually multiple lesions; median age 44 years; trunk and flexural areas	Infiltrate in the papillary dermis comprising lymphocytes with cytologic atypia; epidermotropism, epidermal atrophy, basal vacuolar degeneration; pigment incontinence; telangiectatic blood vessels; immunohistochemistry: variable loss of T-cell markers
Acrodermatitis chronica atrophicans[4]	Erythematous or livedoid atrophic patches; multiple lesions, poorly demarcated, might be widespread; extremities; no age or sex predominance; history of tick bite; other symptoms and signs of Lyme disease	Dilatation of vascular channels (telangiectasia); dense infiltrate of lymphocytes, plasma cells, histiocytes, and macrophages; epidermis commonly hypertrophic with acanthosis or might be atrophic with loss of ridge pattern; immunohistochemistry: CD31 and ERG highlight the telangiectatic vessels, D2-40 stains scattered lymphatics
Fixed drug reaction	Erythematous or livedoid patch or plaque; single recurrent, rarely multiple; no age or sex predominance; preceding drug intake	Vacuolar interface reaction with colloid body formation; dyskeratotic keratinocytes; arked pigment incontinence; mild-to-moderately dense lichenoid infiltrate; proliferation of the vascular channels or changes in the vasculature is not a feature; immunohistochemistry: noncontributory as vascular proliferation is not a feature
Microvenular hemangioma[5]	Erythematous patches, plaque, or nodule; most often solitary but can be multiple; trunk, extremities, or head and neck; young adults and rarely children; sudden onset with short period of rapid growth	Thin-walled blood vessels dissecting through collagen bundles; superficial to mid-dermis; occasional tracking along adnexal structures and invasion of arrector pili; minimal inflammation; immunohistochemistry: CD31 and ERG highlight the vessels, D2-40 negative
Hobnail hemangioma[11,12]	Violaceous central papule surrounded by a thin pale ring and an ecchymotic periphery and nonspecific vascular lesion; wide age distribution; no clear sex predilection	Biphasic vascular proliferation in dermis extending into the subcutis; ectatic vascular channels lined by hobnail endothelial cells in superficial dermis; slit-like or angulated vessels dissecting through collagen bundles; lymphatic origin; immunohistochemistry: D2-40 and CD31 stain most vessels, ERG highlights blood vessels endothelium

Continued

Table III. Cont'd

Diagnosis	Clinical features	Histologic features
Pigmented purpuric dermatoses[6]	Irregular purpuric macules and plaques with pinpoint petechiae; solitary in lichen aureus and multiple in other variants; predominantly lower extremities, rarely on buttocks, trunk, and arms; middle age and older men predominantly; purpura annularis telangiectodes more common in women	Red cell extravasation; hemosiderin deposition with epidermal changes (spongiosis, interface, lymphocyte exocytosis, erythrocyte exocytosis) in over two thirds of cases; special stains: positive Perls stain; immunohistochemistry: noncontributory, vascular proliferation is not a feature
Histologic differential diagnoses		
Acquired elastotic hemangioma[7-9]	Erythematous and violaceous plaque; sun-exposed areas, particularly the upper extremities and neck; solitary, slowly growing, asymptomatic; equal male-to-female ratio	Proliferation of dilated, vascular channels; band-like pattern; separated from the epidermis by a thin Grenz zone; prominent solar elastosis; no background fibrosis or edema; mild, nonspecific; immunohistochemistry: CD31 and ERG highlight vessels, occasionally positive for D2-40
Microvenular hemangioma[5]	See above	See above
Acquired elastotic hemangioma-like changes and eccrine sweat duct squamous metaplasia in lichen simplex chronicus or prurigo nodularis-like lesions of the knee and elbow[10]	Hyperkeratotic erythematous plaque; mean age 74 years; male predominance; elbows and knees	Compact hyperkeratosis, with small foci of parakeratosis, hypergranulosis, and acanthosis, with irregular elongation of thick rete ridges; mild melanocytic hyperplasia; no Grenz zone; correlation between thickness of epidermis and depth of vascular proliferation; syringosquamous metaplasia; milia-like changes, periductal mucinous; compressed increased elastic tissue underlying vascular proliferation; immunohistochemistry: CD31 and ERG highlight vessels, D2-40 positive in 10%-30% of vessels

ERG, Erythroblast transformation-specific–related gene.

or other potentially serious dermatoses, so histologic examination is paramount for the diagnosis. Early recognition of poikilodermatous plaque-like hemangioma will prevent unnecessary investigation and treatment and minimize anxiety for both clinicians and patients.

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