Acquired poikiloderma: Proposed classification and diagnostic approach

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Poikiloderma is a morphologic and descriptive term referring to a combination of cutaneous atrophy, telangiectasia, and varied macular pigmentary changes that result in a mottled skin appearance. Its etiology includes both congenital and acquired causes. Many studies have reported different causes of acquired poikiloderma; however, no single well-defined classification has been explored to date. Herein, we analyze all the possible causes of acquired poikiloderma and propose an etiological classification that, hopefully, will lead to better characterization for this ill-defined condition. Moreover, this study presents a step-by-step approach to the management of patients with acquired poikiloderma and summarizes the key differentiating features for each individual cause, which may help in easy and precise diagnosis of different causes of acquired poikiloderma. (J Am Acad Dermatol 2013;69:e129-40.)

Key words: mottled pigmentation; parapsoriasis; poikiloderma; poikiloderma of Civatte; poikiloderma vasculare atrophicans; poikilodermatous mycosis fungoides.

Histopathology

Histopathological findings (Fig 3) include common features in any case of poikiloderma such as thinning of the stratum malpighii, hydropic degeneration of the basal cell layer, presence of melanophages in the papillary dermis, and dilatation of the papillary dermal capillaries. Characteristic features in relation to some specific causes such as epidermotropism in case of poikilodermatous mycosis fungoides (MF) have also been reported.

Classification

Infections

Borrelia burgdorferi: Acrodermatitis chronica atrophicans of Lyme disease. Lyme disease is a tick-borne zoonosis caused by strains of the Gram-negative spirochete Borrelia burgdorferi sensu lato. Acrodermatitis chronica atrophicans is the late stigma of Lyme disease with an insidious onset after the initial tick bite. The sun-exposed extensors of the lower extremities are the most common predilection site; however, lesions can also affect the trunk and rarely the face. In a matter of years, poikilodermatous skin may be the end
result of the untreated plaques of acrodermatitis chronicica atrophicans.\(^8\)

In addition to the common histopathological features of poikiloderma, acrodermatitis chronicica atrophicans is characterized by destruction of epidermal appendages, a subepidermal zone of degenerated connective tissue, and a prominent plasma cell dermal infiltrate. Moreover, the spirochete may be identified by Warthin-Starry stain\(^9\) and may be cultured from the atrophic skin in some cases.\(^10\)

### Inflammatory

**Atopic dermatitis.** Poikiloderma is frequently encountered on the neck in severe cases of adult atopic dermatitis. This may be attributed to chronic inflammation and delayed wound healing induced by the prolonged topical corticosteroid application. The histopathological findings include those of poikiloderma and spongiotic dermatitis.\(^11\)

**Chronic graft-versus-host disease.** Chronic graft-versus-host disease may occur within 3 years after allogeneic hematopoietic cell transplantation. Poikiloderma is one of the diagnostic cutaneous signs for chronic graft-versus-host disease and is often present on the face and trunk during sclerodermoid reaction.\(^12\) In addition to the common histologic features of poikiloderma, the sclerodermoid phase is characterized by dermal fibrosis with destruction of adnexal structures.\(^13\)

**Lichen planus–induced poikiloderma.** This is classic lichen planus that is rarely followed, within months or years, by a network of generalized small erythematous papules and poikilodermatous lesions. Histopathologically, the lesions show features suggestive of both lichen planus and poikiloderma.\(^14\)

### Metabolic

**Amyloidosis: Poikiloderma-like cutaneous amyloidosis.** Poikiloderma-like cutaneous amyloidosis combines cutaneous poikilodermatous changes with amyloid deposits.\(^15\) Two different clinical forms were described: ordinary and syndromic.\(^16\) The ordinary poikiloderma-like cutaneous amyloidosis appears around the fifth decade of life and is characterized by the presence of poikilodermatous lesions at sun-exposed or sun-protected sites (Fig 4), lichenoid papules, and blisters, especially on the limbs.\(^17\)

On the other hand, the term “poikiloderma-like cutaneous amyloidosis syndrome” was proposed for patients with poikilodermatous lesions, lichenoid papules, blisters, photosensitivity, palmpoplantar keratoses, short stature, and onset early in life.\(^15,16\)

The histopathological findings in routine sections include the common poikilodermatous changes and eosinophilic amyloid deposits. Moreover, sections stain positive for Congo red stain and fluoresce with apple-green birefringence under polarized light.\(^15,17\)

### Connective tissue diseases

**Lupus erythematosus.** In systemic lupus erythematosus, poikiloderma has been reported mainly as a feature of advanced disease in which acute erythematous lesions can proceed to poikilodermatous lesions on sun-exposed sites.\(^18\) More surprisingly, in 1964 Tuffanelli et al\(^19\) described generalized poikiloderma occurring in an identical twin sister of a patient with systemic lupus erythematosus.

Cases of poikilodermatous subacute cutaneous lupus erythematosus are rarely reported.\(^1\) In one case report from Bulgaria, the patient developed poikilodermatous subacute cutaneous lupus erythematosus after an episode of sunburn, suggesting a role for ultraviolet radiation in the development of this poikiloderma.\(^20\)

In discoid lupus erythematosus, an extremely rare variant consisting of purplish plaques or blotty reticulate telangiectasia is called the “telangiectoid variant” that may develop on many sites, particularly the face and neck.\(^21,22\) This variant is mostly associated with other chronic discoid lupus erythematosus lesions or can replace active inflammation in patients with classic discoid lupus erythematosus. Cases of lupus panniculitis that clear with poikilodermatous features have also been reported.\(^23\)

Regarding neonatal lupus erythematosus, the typical skin lesions usually clear in the first year of life but may leave areas of atrophy, hypopigmentation, and telangiectasia, resulting in a poikilodermatous appearance.\(^24\)

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**CAPSULE SUMMARY**

- Different causes of acquired poikiloderma have been reported in the literature; however, no single well-defined classification has been established to date.
- The study suggests an etiological classification, presents a step-by-step approach, and summarizes the key differentiating features of acquired poikiloderma.
- The proposed classification and diagnostic approach would serve as a practical tool for proper diagnosis, differentiation, and management of acquired poikiloderma.
Crowley and Frieden\textsuperscript{25} reported a patient with neonatal lupus erythematosus and an unusual presentation in the form of generalized poikilodermia, associated with extensive skin erosions, patchy scalp alopecia, absent eyebrows and eyelashes, absent neonatal lupus erythematosus classic skin lesions, and negative Ro and La antibodies associated with a strongly positive antinuclear antibody titer with a speckled pattern in both mother and infant.

**Dermatomyositis.** Poikilodermia in dermatomyositis is often a late finding and it may occur on sun-exposed skin such as the V-shaped area of the neck and/or sun-protected skin such as the upper aspect of the back (shawl sign) (Fig 5) and the upper-lateral aspect of thighs (holster sign).\textsuperscript{26-28} Sometimes the poikilodermia in dermatomyositis is more generalized and this may resemble a poikilodermic cutaneous T-cell lymphoma.\textsuperscript{29} The histopathological alternations found in poikilodermatomyositis (Fig 2) are just those of classic poikilodermatous changes.\textsuperscript{26}

**Systemic sclerosis.** In systemic sclerosis, the most characteristic pigmented change is loss of pigment in a large patch with perifollicular pigment retention. The affected areas become hairless and atrophic.\textsuperscript{30} Mat-like telangiectasia is found mainly on the face but may extend as far as the upper aspect of thighs. Pigmentation occurs also most frequently on the face and to a lesser extent on the legs, thighs, and lower aspect of abdomen.\textsuperscript{31} Therefore, we believe that a mixture of these findings can result in a poikilodermic appearance.

**Environmental Solar radiation.** Poikiloderma of Civatte. Poikiloderma of Civatte is a common condition with a slowly progressive and irreversible course.\textsuperscript{31} According to the predominance of 1 or more of the components of poikilodermia, some classify poikiloderma of Civatte into erythematotelangiec-tatic, pigmented and mixed types.\textsuperscript{32} The classic distribution of the lesions symmetrically involves the side of the face and neck and the upper aspect of the chest (Fig 6). Solar radiation has been proposed as the main culprit besides other factors such as genetics, low estrogen, and phototoxic or photoallergic reactions to chemicals in fragrances or cosmetics.\textsuperscript{32-34} The most prominent and constant histopathological feature, besides the classic poikilodermic changes, is solar elastosis of the papillary dermis.\textsuperscript{35}

**Photoaging “dermatoheliosis.”** Repeated solar injuries can ultimately result in photoaging or what
is called “dermatoheliosis.” This happens more in patients with fair skin, especially outdoor workers. The most common sites affected are the sun-exposed areas, particularly face and scalp in bald men. 36

Cutaneous manifestations of dermatoheliosis (Fig 7) show a combination of atrophy, telangiectases, purpura, spotty depigmentation, hyperpigmentation solar lentigines, and actinic keratoses. 36

Heat and infrared radiation: Erythema ab igne. Erythema ab igne is characterized by localized mottled and reticulated cutaneous hyperpigmentation with variable telangiectasia and skin atrophy resulting from repeated or prolonged exposure to heat and infrared radiation. Therefore poikiloderma is considered the end stage of the cumulative changes caused by repeated and prolonged exposure to infrared radiation. 9

Erythema ab igne can occur at all ages including childhood 37 but it was reported as a common condition on the shins among the elderly using heaters or fireplaces. 38 The distribution of the lesions depends on the direction of the incident radiation, the contour of the skin, and the interposition of clothing. 9

In the early stages, epidermal atrophy, dermal pigmentation, and capillary dilatation are evident. Basophilic degeneration of the connective tissue, focal hyperkeratosis, and epithelial cellular atypia occur later. 39

Chemical intoxication: Sulfur mustard-induced poikiloderma. Sulfur mustard is a chemical weapon that was first used by the German Army in 1917 40 and was widely used in the 1980s during the Iran-Iraq conflict. Based on the lipophilic properties of this gas, it easily penetrates the skin and mucosal surfaces causing several acute and chronic effects on the skin, eye, and respiratory system. 41

In 2011, Emadi et al 42 proposed a diagnosis of sulfur mustard–induced poikiloderma in a 41-year-old man who was injured with sulfur mustard during an Iraq chemical attack in 1988. Their diagnosis was based on the clinical findings of atrophy, pigmentation, and telangiectasia on the upper aspect of the trunk, back of the hands, and genitalia associated with relevant histopathological features of poikiloderma.

Iatrogenic

Drugs. Corticosteroids. Corticosteroids are accused of many undesirable side effects including atrophy, telangiectasia, and cutaneous dyspigmentation. 43 Therefore, it can be expected that a constellation of these features will eventually result in a poikilodermic skin appearance.

Hydroxyurea (hydroxycarbamide). Hydroxyurea is commonly used for myeloproliferative disorders and rarely for severe recalcitrant psoriasis. 44 Major adverse reactions of hydroxyurea include both
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<td>Achoer dermatitis chronica atrophicans</td>
<td>Months or years after initial tick bite</td>
<td>Extensor surfaces of lower limbs</td>
<td>Arthritis, morphea-like or lichen sclerosis atrophicus—like lesions</td>
<td>Prominent plasma cell dermal infiltrate and destruction of appendages</td>
<td>Positive serology and PCR for <em>Borrelia</em> can confirm diagnosis</td>
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<td>Spirochetes may be detected by Warthin-Starry stain</td>
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<td>Atopic dermatitis</td>
<td>Mostly in adults</td>
<td>Sides of neck</td>
<td>Itching and other atopic manifestations</td>
<td>Spongiotic dermatitis and poikiloderma</td>
<td>Prolonged corticosteroid application may contribute to condition</td>
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<td>Chronic GVHD</td>
<td>Usually within 3 y post-HCT</td>
<td>Face and trunk during sclerodermoid reaction</td>
<td>Lichen planus—like, morphea-like, lichen sclerosis atrophicus—like lesions</td>
<td>Sclerodermoid phase is characterized by dermal fibrosis and destruction of adnexal structures</td>
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<td>Lichen planus—induced poikiloderma</td>
<td>Years after onset of classic lichen planus lesions</td>
<td>May be generalized</td>
<td>Classic lichen planus lesions</td>
<td>Histologic changes of both lichen planus and poikiloderma</td>
<td>Only 1 case is reported</td>
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<td>Poikiloderma-like cutaneous amyloidosis</td>
<td>Around fifth decade of life</td>
<td>Sun-exposed or sun-protected skin</td>
<td>Lichen amyloidosis and cutaneous blisters</td>
<td>Dermal amyloid deposits and positive Congo red Apple-green birefringence with polarized light</td>
<td>Disappointing therapeutic options</td>
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<td>Lupus erythematosus</td>
<td>Patients with SLE and NLE With SCLE after sunburn After clearing lesions of lupus panniculitis</td>
<td>On sun-exposed sites, may be generalized</td>
<td>SLE and NLE: other cutaneous and systemic manifestations SCLE: other classic skin lesions</td>
<td>Patchy lymphocytic infiltrate of LE plus poikilodermatous changes</td>
<td>Poikiloderma indicates advanced SLE Serologic tests can help to confirm diagnosis Anti-Ro and La were in the reported case about NLE and poikiloderma</td>
</tr>
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<td>Dermatomyositis</td>
<td>Usually late finding during disease course</td>
<td>Arm extensors, V-area of neck, upper aspects of back and thighs or generalized</td>
<td>Photosensitivity, myositis Diagnostic signs of dermatomyositis, eg, Gottron sign and papules</td>
<td>Poikilodermatous changes EMG and muscle biopsy specimen showing inflammatory myopathy Elevated CK, AST, ALT</td>
<td></td>
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<tr>
<td>Systemic sclerosis</td>
<td>Progressive</td>
<td>Usually on face</td>
<td>Other systemic manifestation</td>
<td>Dermal fibrosis</td>
<td></td>
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<tr>
<td>Poikiloderma of Civatte</td>
<td>After repeated UV exposure Slowly progressive and irreversible</td>
<td>Sun-exposed side of face and neck, upper aspect of chest with sparing of submental area</td>
<td>Usually asymptomatic May be itching, burning, and flushing</td>
<td>Poikidromatous changes plus solar elastosis</td>
<td>Patch test to diagnose cases induced by certain allergens IPL may help Proper photoprotection can limit occurrence</td>
</tr>
<tr>
<td>Dermatoheliosis</td>
<td>Chronic sun exposure especially in fair skin</td>
<td>Sun-exposed, eg, bald scalp in men</td>
<td>Actinic keratoses are invariably present</td>
<td>Solar elastosis plus poikidromatous changes</td>
<td></td>
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<tr>
<td>EAI</td>
<td>EAI starts 2 wk to several months after heat exposure</td>
<td>Any heat-exposed site can be affected but commonly on shins</td>
<td>Usually asymptomatic Some reported itching and burning sensation</td>
<td>Poikidromatous changes with connective tissue basophilic degeneration</td>
<td>Later, epithelial cell atypia can be found</td>
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<td>Sulfur mustard–induced poikiloderma</td>
<td>Years after exposure to warfare agent with history of blistering after acute exposure</td>
<td>Upper aspect of trunk, back of hands, genitalia</td>
<td>Eyes and respiratory system</td>
<td>Poikidromatous changes</td>
<td>Was widely used in 1980s during Iran-Iraq conflict</td>
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<tr>
<td>Corticosteroids</td>
<td>Depends on potency, site, and presence or absence of occlusion</td>
<td>Sites of prolonged steroid application</td>
<td>Other side effects, eg, acneiform eruptions, hypertrichosis</td>
<td>Poikidromatous changes</td>
<td>Following safe guidelines for corticosteroid use can prevent this poikiloderma Other nondermatologic side effects can be found, eg, bone-marrow suppression Pulsed dye laser can improve telangiectasia</td>
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<td>Hydroxyurea</td>
<td>Usually in patients with myeloproliferative disorders on hydroxyurea therapy</td>
<td>More on sun-exposed sites</td>
<td>Other cutaneous adverse effects, eg, leg ulcers and dermatomyositis-like eruption</td>
<td>Poikidromatous changes</td>
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<tr>
<td>Chronic radiation dermatitis</td>
<td>History of exposure to ionizing radiation source</td>
<td>At radiation-exposed site</td>
<td>Other adverse reactions, eg, alopecia, nail loss, and tissue fibrosis</td>
<td>Poikidromatous changes and dermal fibrosis</td>
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nondermatologic effects such as bone-marrow suppression, and dermatologic effects such as poikiloderma, ichthyosis, Gottron-like eruptions, nail abnormalities, oral ulceration and pigmentation, leg ulcers, actinic keratoses, and squamous cell carcinoma.45-47

Radiotherapy. Chronic radiation dermatitis. Chronic radiation dermatitis occurs after exposure to ionizing radiation used in tumor radiotherapy, and during interventional procedures such as fluoroscopic imaging.48 In chronic radiation dermatitis, persistent poikilodermatous changes are indicative of significant cutaneous injury.49,50

Neoplastic

Mycosis fungoides. Poikilodermatous variant. Poikilodermatous MF is predominantly located on the breast (Fig 8), hips, and buttocks. These lesions may be associated with other patches and plaques of classic MF. Rarely, patients may have extensive poikiloderma as a feature of erythrodermic disease.9 The progression of poikilodermatous MF may be similar to that of the patch stage of classic MF, although a greater proportion of cases tend to show spontaneous regression and fewer patients proceed to tumor stage.51

A skin biopsy specimen shows histologic findings similar to those seen in long-standing patch- or plaque-stage MF lesions, in addition to the typical histopathological changes of poikiloderma.52 Immunophenotypic studies typically demonstrate a CD2+, CD3+, CD4+, CD8+, and CD7+ phenotype can be found. It indicates poor prognosis.

Typical changes of MF and poikiloderma

Other patches and plaques of classic MF may be found. It is rare type with no distinct clinical presentation stage. Histopathological features are essential for its diagnosis. Slowly progressive. Lower aspect of trunk and upper aspect of thighs. ± itching.

Poikilodermatous MF

Mainly on breasts and hips. It slowly progresses like patch stage of classic MF but few cases proceed to tumor stage. Histopathological features are essential for its diagnosis. Slowly progressive. Trunk and flexural areas. ± itching.

Poikilodermatous parapsoriasis

Poikiloderma vasculare atrophicans.

Poikiloderma vasculare atrophicans has been considered as poikilodermatous MF but few cases proceed to tumor stage. Histopathological features are essential for its diagnosis. Slowly progressive. Lower aspect of trunk and upper aspect of thighs. ± itching.

CD4+, but rarely CD8+ phenotype can be found. It indicates poor prognosis.

Granulomatous MF

Granulomatous reactions with epidermotrophic changes. Usually shows mild dermatitis. ± itching.

Poikiloderma atrophicans

Poikiloderma atrophicans is a rare type of cutaneous T-cell lymphoma that indicates a poor prognosis. It can only be diagnosed by the histopathological demonstration of granulomatous reaction because of the lack of a distinct clinical presentation.55

In 2008, Morihara et al55 reported a case granulomatous MF presenting clinically with poikiloderma, ichthyosis, and erythematous scaly plaques, and histologically by a granulomatous reaction with giant cells and epidermotropic atypical lymphocytes.

Poikiloderma vasculare atrophicans. Poikiloderma vasculare atrophicans has been considered as poikilodermatous MF56 and, according to Kreuter et al57 in 2005, the term “poikilodermatous variant of cutaneous T-cell lymphoma” has mostly replaced “poikiloderma vasculare atrophicans.” However, poikiloderma vasculare atrophicans has been
known to show a benign course, without progression to the tumor stage of MF.58

Besides poikiloderma, lesions of poikiloderma vasculare atrophicans usually manifest as asymptomatic or mildly pruritic flat-topped, scaly papules coalescing into retiform patterns or zebra-like distributions and associated with telangiectasia. The typical sites of predilection are the trunk and flexural areas.59

The histologic findings include those of poikiloderma and the classic histopathological and immunohistochemical findings of long-standing patch or plaque lesions of the classic MF.55,57

**Fig 3.** Step-by-step approach to the management of the patient with acquired poikiloderma.

**AC4A,** Acrodermatitis chronica atrophicans; **AD,** atopic dermatitis; **B burgdorferi,** Borrelia burgdorferi; **DM,** dermatomyositis; **EAI,** erythema ab igne; **GVHD,** graft-versus-host disease; **HCT,** hematopoietic cell transplantation; **H&E,** hematoxylin-eosin; **IPL,** intense pulsed light; **LD,** Lyme disease; **LSA,** lichen sclerosus atrophicus; **MF,** mycosis fungoides; **NLE,** neonatal lupus erythematosus; **PC,** poikiloderma of Civatte; **PDL,** pulsed dye laser; **PLCA,** poikiloderma-like cutaneous amyloidosis; **PVA,** poikiloderma vasculare atrophicans; **SLE,** systemic lupus erythematosus; **SS,** systemic sclerosis.

**A step – by – step approach to the patient with acquired poikiloderma.**

**Step 1**

**History**
- **Course:** Persistent e.g. in ACA and poikiloderma of Civatte. Gradually regressive e.g. in EAI after cessation of heat exposure. Progressive e.g. in mycosis fungoides.
- **Occupation:** Outdoor workers for dermatoheliosis & poikiloderma of Civatte, bakers for EAI & war soldiers for nitrogen mustard induced poikiloderma.
- **Exposure to tick bites or living in/travelling to endemic areas:** For ACA.
- **Drug:** Ask for hydroxyurea intake & corticosteroid application.

**History of skin disease:** e.g. atopic dermatitis for AD associated poikiloderma & lichen planus for lichen plaques - induced poikiloderma.

**History of medical disease:** e.g. history of malignant tumor and radiotherapy administration, history of myeloproliferative disorders and hydroxyurea intake, history suggestive of SLE, DM or SS.

**Histology:** Itchy poikiloderma can present in cases of e.g. AD associated poikiloderma, lichen planus induced poikiloderma, mycosis fungoides variants, PVA, poikilodermatous parapsoriasis & poikiloderma of Civatte.

**Other associated cutaneous symptoms:** e.g. a roses-like flushing in poikiloderma of Civatte.

**Step 2**

**Examination**
- **Site of poikiloderma lesions:** e.g. lower extremities in ACA, at the sides of the neck in AD associated cases, at the VI area of the neck “double sign” in DM, at sun-exposed lateral face & neck with sparing of submental area in poikiloderma of Civatte, at the shins of legs in EAI, at the breasts & hips in poikiloderma MF and at the lower trunk & upper thighs in poikilodermatous parapsoriasis.

**Distribution:** e.g. bilateral & symmetric in poikiloderma of Civatte, retiform pattern or zebra-like distribution in PVA & poikilodermatous parapsoriasis and bimodal distribution in MF.

**Other cutaneous lesions:** e.g. Morphea-like and/or LSA-like lesions with ACA & chronic GVHD, lichen and/or vascular amyloidosis classic lesions with PLCA, malar rash in SLE, cutaneous papules & sign with DM, Ramsay’s phenomenon in SS, photosensitivity in SLE & DM, acquired ichthyosis with atopia and nail pigmentation in hydroxyurea induced cases, severe skin fibrosis with atopia and nail loss in chronic radiation dermatitis.

**Associated systemic lesions:** e.g. Arthritis with ACA & SLE, acrochordons with lichen sclerosus, diabetes in chronic GVHD, myositis with DM, nephritis with SLE & heart block with NLE, esophageal dysmotility & dysphagia in SS & bone marrow suppression in hydroxyurea induced lesions.

**Possible risks & complications:** e.g. Poikiloderma with SLE indicates advanced disease, dermatoheliosis is invariably associated with pre-cancerous actinic keratoses, PVA can be a true MF & poikilodermatous parapsoriasis can progress finally to frank MF.

**Step 3**

**Investigations**
- **Serological tests:** e.g. for B. burgdorferi, SLE, NLE, DM & SS.
- **H & E sections:** Classic poikiloderma in addition to some characteristic features e.g. prominent plasma cell dermal infiltrate in ACA, amyloid deposits in PLCA, prominent sallow eosinophilic and poikiloderma of Civatte & dermatoheliosis, epithelial atypia in late cases of EAI, atypical lymphocytes in MF variants & PVA and granulomatous reaction in granulomatous MF.
- **Special stainings:** e.g. Congo red for amyloidosis & Warthin Starry for B. burgdorferi.

**Immunohistochernistry:** e.g. a monoclonal antibody, CD4+ T-cell infiltrate in MF variants.

**Step 4**

**Prevention & Treatment**
- **Prevention:** e.g. chemoprophylaxis and vaccination for LD, photoprotection for PC and dermatoheliosis & following the safe guidelines for topical corticosteroid usage.
- **Treatment:** e.g. PDL for poikiloderma of Civatte; IPL for PC, chemotherapy & phototherapy for MF and poikilodermatous parapsoriasis.

**Fig 3.** Step-by-step approach to the management of the patient with acquired poikiloderma. ACA, Acrodermatitis chronica atrophicans; AD, atopic dermatitis; B burgdorferi, Borrelia burgdorferi; DM, dermatomyositis; EAI, erythema ab igne; GVHD, graft-versus-host disease; HCT, hematopoietic cell transplantation; H&E, hematoxylin-eosin; IPL, intense pulsed light; LD, Lyme disease; LSA, lichen sclerosus atrophicus; MF, mycosis fungoides; NLE, neonatal lupus erythematosus; PC, poikiloderma of Civatte; PDL, pulsed dye laser; PLCA, poikiloderma-like cutaneous amyloidosis; PVA, poikiloderma vasculare atrophicans; SLE, systemic lupus erythematosus; SS, systemic sclerosis.
parapsoriasis. Large-plaque parapsoriasis (or poikilodermatous parapsoriasis) is considered a prelymphomatous skin condition because of its biologic behavior and the presence of clinical, histologic, and/or genotypical alterations that overlap early MF. In a large series of patients with poikilodermatous parapsoriasis, 11% developed definite MF but whether these cases were MF from the start or not remains unclear.

Clinically, patients present with persistent large, flat, red or brown, scaly atrophic patches and thin plaques. Lesions appear finely wrinkled as a result of epidermal atrophy, then telangiectasia and mottled pigmentation can be observed, hence the synonym “poikilodermatous parapsoriasis.” Common sites include the lower aspect of the trunk, upper aspect of thighs, and major flexural surfaces. Lesions at the breast and buttock should suggest MF.

Most skin biopsy specimens only show a mild dermatitis and a superficial bandlike dermal lymphocytic infiltrate beneath atrophic epidermis. Single, rarely atypical, lymphocytes without Pautrier microabscesses can be observed in the epidermis in the absence of spongiosis.

**TREATMENT**

Treatment of acquired poikiloderma includes the use of a specific therapy, cosmetic management, or both. Moreover, some causes can be prevented such as...
as the use of chemoprophylaxis and vaccination in Lyme disease—endemic regions, proper photo-protection and patch testing for poikiloderma of Civatte, avoidance of prolonged heat or infrared contact for erythema ab igne, and following the safe guidelines for topical corticosteroid use.

Topical and intralesional corticosteroid, hydrocolloid dressings, topical dimethylsulfoxide, calcineurin inhibitors, acitretin, cyclophosphamide, dermabrasion, phototherapy, and pulsed-dye laser have been all used for poikiloderma-like cutaneous amyloidosis, but are usually disappointing.

Laser therapy using argon or pulsed dye laser can improve poikilodermatomyositis, whereas the results in poikiloderma of Civatte are unsatisfactory. Also, hydroquinones, chemical peels, electrosurgery, and cryotherapy all failed in treating poikiloderma of Civatte. The new intense pulsed light sources are more promising with clearance of more than 75% of telangiectasias and hyperpigmentation in different studies.

Topically applied retinoids may reverse some of the changes of dermatoheliosis. In chronic radiation dermatitis, hyperbaric oxygen can reduce pain and erythema but it has no effect on telangiectasia for which pulsed dye laser may be beneficial.

The standard treatment for early poikiloderma-tous MF includes topical steroids, topical chemotherapy, psoralen plus ultraviolet A, narrowband ultraviolet B, interferon alfa-2a, and oral retinoids. Systemic chemotherapies, denileukin diftitox, anti-CD52 antibody, and interleukin-12 are reserved for advanced stages.

Therapeutic options for poikilodermatous parapsoriasis include high-potency topical steroids, phototherapy, and topical nitrogen mustard.

CONCLUSION

Acquired poikiloderma is a divergent condition that has been attributed to different etiological factors. We hope that our proposed classification and diagnostic approach, including the key differentiating points, may help in easy and precise diagnosis of poikiloderma and its underlying causes. Although the histopathological features are nonspecific, a precise evaluation can lead to the diagnosis of serious diseases as is the case with poikilodermatous MF. Treatment of poikiloderma is that of the cause and laser therapy may be beneficial in some cases; however, the results are usually unsatisfactory.

REFERENCES

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