

Poikiloderma: Differential Diagnosis & Treatment

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Poikiloderma

- ⦿ is not a disease,
- ⦿ is a combination of:
 - > hyperpigmentation,
 - > hypopigmentation,
 - > telangiectasias, and
 - > epidermal atrophy

Poikiloderma



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Poikiloderma: (Etiology) Differential Diagnosis

- Congenital

- Acquired

Differential Diagnosis: Congenital Poikiloderma

1. Congenital bullous poikiloderma (Kindler syndrome)
2. Poikiloderma congenitale (Rothmund-Thomson syndrome)
3. Congenital telangiectatic erythema (Bloom syndrome)
4. Dyskeratosis congenita (Zinsser-Engman-Cole syndrome)
5. Cockayne's syndrome
6. Certain mitochondrial disorders

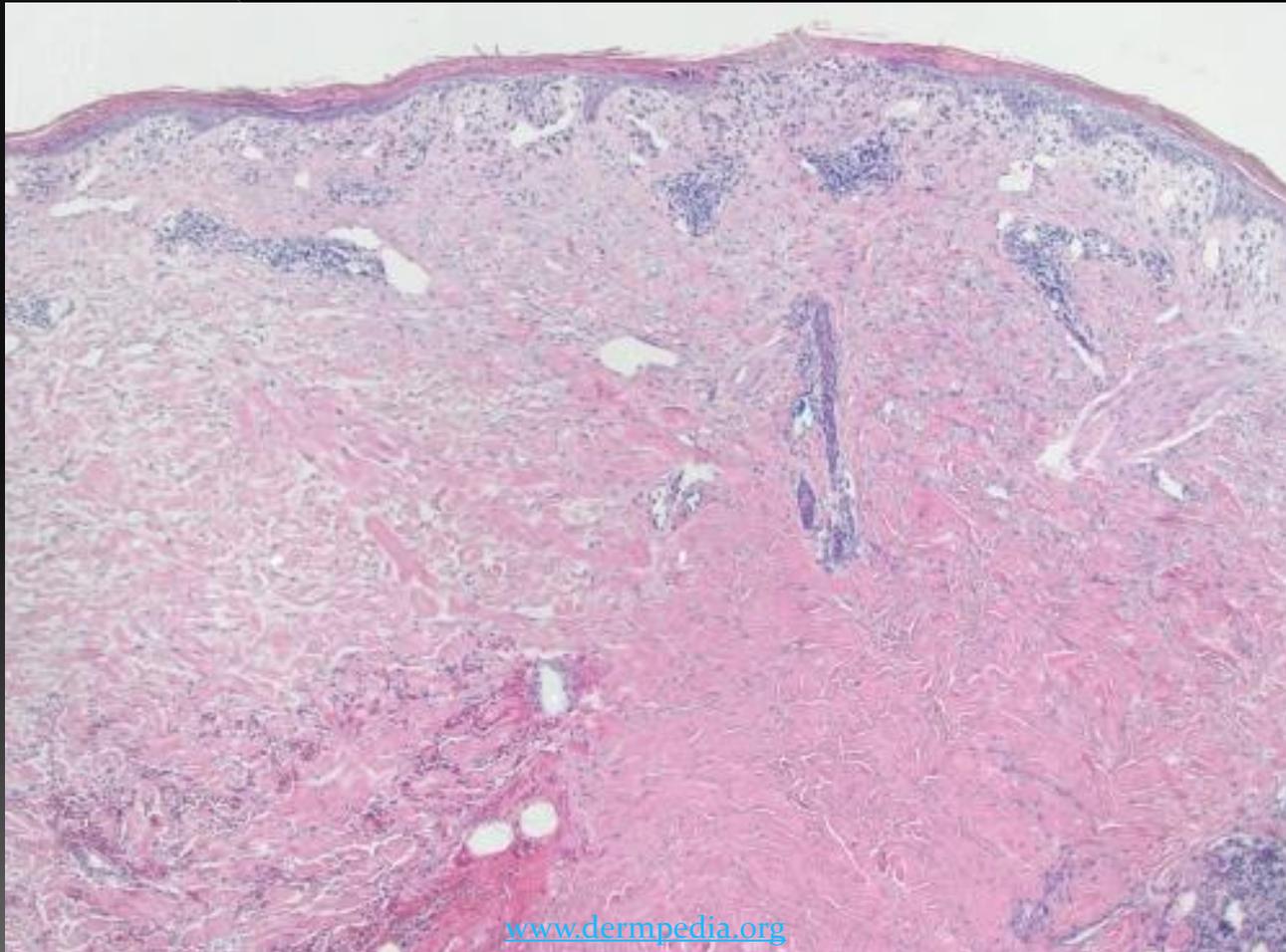
Differential Diagnosis: Acquired Poikiloderma

1. Infection (Lyme disease)
2. Inflammatory cause
3. Metabolic cause
4. Connective tissue diseases
5. Environmental causes
6. Iatrogenic causes
7. Neoplastic causes

Poikiloderma: Histopathology

- ◎ is generally common:
 - > thinning of stratum basale and stratum spinosum,
 - > hydropic degeneration of the basal cell layer,
 - > melanin incontinence, and
 - > dilatation of the papillary dermal capillaries.

Poikiloderma: Histopathology



Acquired Poikiloderma

1. Infection: acrodermatitis chronica atrophicans of Lyme disease
2. Inflammatory cause: chronic graft-versus host disease (GVHD)
3. Metabolic cause: poikiloderma-like cutaneous amyloidosis
4. Connective tissue diseases:
 1. Lupus erythematosus
 2. Dermatomyositis
5. Environmental causes:
 1. Solar radiation
 - I. Poikiloderma of Civatte
 - II. Photo-aging: dermatoheliosis
 2. Heat and infrared radiation: erythema ab igne
 3. Sulfur mustard-induced poikiloderma
6. Iatrogenic causes:
 1. Drugs: corticosteroids and hydroxyurea
 2. Radiotherapy
7. Neoplastic causes:
 1. Poikilodermatous mycosis fungoides
 2. Poikilodermatous (large plaque) parapsoriasis

1. Infection:

Acrodermatitis chronica atrophicans (ACA) of Lyme disease

- ⊙ Lyme: tick borne
- ⊙ Caused by *B. afzelii*, and *B. burgdorferi*, Gram-negative spirochetes
- ⊙ ACA: the late stigma of this infection: the extensors of the lower extremities
- ⊙ Biphasic disorder:
 - > Early inflammatory stage: bluish red discoloration and swelling

Acrodermatitis chronica atrophicans



1. Infection:

Acrodermatitis chronica atrophicans (ACA) of Lyme disease

- ◉ Lyme: tick borne
- ◉ Caused by *B. afzelii*, and *B. burgdorferi*, Gram-negative spirochetes
- ◉ ACA: the late stigma of the Lyme disease: the extensors of the lower extremities
- ◉ Biphasic disorder:
 - > Early inflammatory stage: bluish red discoloration and swelling
 - > Late atrophic stage: glistening cigarette-paper appearance: poikilodermatous lesions occur and slowly extend centrifugally

Acrodermatitis chronica atrophicans



2. Inflammatory:

Chronic graft-versus host disease (GVHD)

- ⦿ GVHD: a multiorgan disease due to the transfer of hematopoietic stem cells via an allogeneic stem cell transplant.
- ⦿ Chronic GVHD: a polymorphous condition occurs within 3 years after the transplant.
- ⦿ Poikiloderma: one of the diagnostic cutaneous signs, often presented on the face and trunk during the sclerodermoid phase of the disease.

Chronic Graft-versus host disease



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3. Metabolic:

Poikiloderma-like cutaneous amyloidosis

- ◎ A rare variant of primary cutaneous amyloidosis; a syndrome characterized by:
 1. Cutaneous amyloid deposits in the lesions
 2. Lichenoid papules
 3. Poikilodermatous skin lesions
 4. Light sensitivity
 5. Short stature
 6. Blisters; and
 7. Palmoplantar keratosis.

4. Connective tissue diseases

1. Lupus erythematosus:

I. SLE

II. SCLE & DLE

2. Dermatomyositis

4. Connective tissue diseases

1. Lupus erythematosus:

I. SLE: poikiloderma occurs especially as a feature of advanced disease: proceeding of acute erythematous lesions to poikilodermatous lesions, sun-exposed areas

II. SCLE & DLE: rarely

Lupus Erythematosus



Carly A. Elston; Dirk M. Elston, MD
<http://reference.medscape.com>

4. Connective tissue diseases

2. Dermatomyositis:

- > Poikiloderma is a late finding, presented on the sun exposed areas of the skin such as the upper aspect of the back (Shawl sign) and V-shaped area of the neck.

Dermatomyositis



Shawl sign



Jeffrey P Callen, MD. Poikiloderma
www.medisuite.ir

5. Environmental causes

1. Solar radiation:
 - I. Poikiloderma of Civatte
 - II. Photo-aging: Dermatoheliosis
2. Heat and infrared radiation: Erythema ab igne
3. Sulfur mustard-induced poikiloderma

5. Environmental causes

1. Solar radiation:

I. Poikiloderma of Civatte

- Common, slowly progressive and irreversible, the sides of the face and the neck.
- Solar radiation, genetics, and phototoxic reaction to chemicals in fragrances or cosmetics.

Poikiloderma of Civatte



5. Environmental causes

1. Solar radiation:

II. Photo-aging: Dermatoheliosis

- Repeated solar injuries, fair skin patients, sun-exposed areas like the face and the bald scalp.

Dermatoheliosis



<http://quizlet.com/12937912/benign-skin-conditions-flash-cards/>

5. Environmental causes

2. Heat and infrared radiation: Erythema ab igne (toasted skin syndrome)
 - > Localized reticulated poikilodermatous areas
 - > Repeated or prolonged exposure to the sources of heat and infrared radiation: heaters, stoves, and laptops!
 - > The sites and the distribution of the lesions depend on the direction of the radiation: shins because of putting the stoves nearby.

Erythema ab igne



5. Environmental causes

3. Sulfur mustard-induced poikiloderma

- > Sulfur mustard: a chemical weapon used in the 1980s.
- > Lipophilic gas: easily penetrates the skin and mucosal surfaces causing several acute and chronic effects on the respiratory system, the eyes and the skin including poikilodermatous lesions.

6. Iatrogenic causes

1. Drugs

I. Topical corticosteroids

II. Hydroxyurea

2. Radiotherapy: Chronic radiation dermatitis

6. Iatrogenic causes

1. Drugs

I. Topical corticosteroids:

- are accused of many undesirable side effects: skin atrophy, telangiectasia, and cutaneous dyspigmentation: poikilodermatous appearance
- repeated and/or continuous usage in chronic dermatologic conditions like atopic dermatitis (on the neck) and lichen planus

II. Hydroxyurea: (hydroxycarbamide):

- is used for myeloproliferative disorders
- nondermatologic and dermatologic adverse reactions: ichthyosis, nail abnormalities, poikiloderma, and skin malignancies

2. Radiotherapy: Chronic radiation dermatitis:

- > After frequent exposures to ionizing radiation used in tumor radiotherapy
- > Poikiloderma: indicative of significant cutaneous injury.

Chronic radiation dermatitis



7. Neoplastic causes

1. Poikilodermatous mycosis fungoides (MF)
2. Poikilodermatous(large) plaque parapsoriasis

7. Neoplastic causes

1. Poikilodermatous MF:

- > Poikiloderma vasculare atrophicans?
- > Breasts, hips, and gluteal areas
- > ± other classic patches and plaques of MF
- > Progression is similar to that of the patch stage of classic MF ± spontaneous regression

Mycosis fungoides



7. Neoplastic causes

2. Poikilodermatous(large) plaque parapsoriasis
 - > A prelymphomatous skin condition, early MF?
 - > Persistent large, scaly atrophic patches and thin plaques.
 - > Appears finely wrinkled as a result of epidermal atrophy, then telangiectasia and mottled pigmentation can be observed: poikilodermatous.
 - > The lower aspect of the trunk, the upper aspects of the thighs, and flexural surfaces

Large plaque parapsoriasis



Acquired Poikiloderma: Treatment

- I. Prevent the cause:
 - > Photoprotection and avoidance of perfumes in case of poikiloderma of Civatte
 - > Avoidance of heat and infrared exposure for erythema ab igne
 - > To be careful with topical corticosteroid use
- II. Specific treatments:
 - I. Laser: Argon, IPL (Civatte), and pulsed dye laser (chronic radiation dermatitis)
 - II. Hydroquinon-containing preparations
 - III. Exfoliants (α -hydroxy acids and topical retinoids): dermatoheliosis
- III. Treat the cause: poikilodermatous MF and large plaque parapsoriasis

Conclusion

- ◎ Poikiloderma is a divergent condition that has been attributed to different dermatologic disorders.
- ◎ Although the histopathological features are nonspecific, a precise evaluation can lead to the diagnosis of serious diseases like MF.
- ◎ Treatment of poikiloderma is that of the cause and laser therapy may be beneficial in some cases; however the results are not completely satisfactory.