

## Poikiloderma: Differential Diagnosis and Treatment

Sehriyar Nazari\*

*Dermatology Unit of Istanbul Safak Hospital, Istanbul, Turkey*

\*Corresponding author: Sehriyar Nazari, Dermatology Unit of Istanbul Safak Hospital, Istanbul, Turkey, Tel: 0212-5306474; E-mail: [sehriyar.nazari@istanbulsafak.com](mailto:sehriyar.nazari@istanbulsafak.com)

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### Letter

As you know, in dermatology textbooks, you may not find the title of poikiloderma in the list of the diseases. It is because poikiloderma is not a disease per se, but a descriptive term featuring a combination of hyperpigmentation, hypopigmentation, telangiectasias, and epidermal atrophy that results in a mottled skin appearance.

Poikiloderma can be caused by both congenital and acquired conditions. Congenital poikiloderma occurs in certain genetically determined syndromes such as Kindler Syndrome (congenital bullous poikiloderma), and Rothmund-Thomson Syndrome; whereas acquired poikiloderma can occur by different causes such as neoplasm, infection, radiation and etcetra.

### Histopathology

Despite the cause of poikiloderma, histopathological findings are generally common and nonspecific in any case of poikiloderma and include thinning of stratum basale and stratum spinosum, hydropic degeneration of the basal cell layer, presence of melanophages in the papillary dermis, and dilatation of the papillary dermal capillaries. However, characteristic pathologic features can be also found additionally in relation to some specific causes such as epidermotropism in case of mycosis fungoides.

I will focus just on the causes & treatment of acquired poikiloderma:

Here, you find a proposed classification for the causes of acquired poikiloderma which was mainly prepared by Nofal and colleagues. As you can see, these causes are summarized in seven major categories, including infection, inflammatory cause, metabolic cause, connective tissue diseases, environmental causes iatrogenic causes, and neoplastic causes.

Nofal A, Salah E. Acquired poikiloderma: Proposed classification and diagnostic approach. *J Am Acad Dermatol* 2013; 69: e129-40.

#### 1. Infection: Acrodermatitis chronica atrophicans of Lyme disease.

Lyme disease is a tick-borne zoonosis caused by strains of a gram-negative spirochete, *B. afzelii*, and sometimes *B. burgdorferi*. ACA is the late stigma of the disease appeared months to years after the tick bite. The extensors of the lower extremities are the most common sites of involvement. It is a biphasic disorder consisting of an early, easily treatable inflammatory stage characterized by bluish red discoloration and skin swelling, and a late, treatment resistant, atrophic stage in which skin has a glistening cigarette-paper appearance. In this stage, the poikilodermatous lesions may occur and slowly extend centrifugally leaving central areas of atrophy. The spirochete may be identified by Warthin-Starry stain and may be cultured from the atrophic skin in some cases.

**2. Inflammatory cause:** Chronic GVHD: GVHD is a multiorgan disease most commonly due to the transfer of hematopoietic stem cells via an allogenic stem cell transplant. Chronic GVHD is a polymorphous condition which occurs within 3 years after the transplant. Poikiloderma is one of the diagnostic cutaneous signs for chronic GVHD and is often present on the face and trunk during sclerodermoid phase of the disease.

**3. Metabolic cause:** Poikiloderma-like cutaneous amyloidosis: Primary cutaneous amyloidosis is the deposition of amyloid in the skin without involvement of internal organs. It is easily diagnosed when presented in its typical manifestation. Poikiloderma-like cutaneous amyloidosis (PCA), a rare variant of primary cutaneous amyloidosis, is characterised by: 1) poikilodermatous skin lesions; 2) lichenoid papules; 3) blisters; 4) cutaneous amyloid deposits in the pigmented and lichenoid lesions; 5) light sensitivity; 6) short stature; and 7) palmoplantar keratosis.

#### 4. Connective tissue diseases:

**-LE:** In SLE, poikiloderma may occur especially as a feature of advanced disease when acute erythematous lesions can proceed to evolve poikilodermatous lesions on sun-exposed areas. In SCLE and DLE, lesions of poikiloderma are rarely reported.

**-Dermatomyositis:** Poikiloderma in dermatomyositis is a late finding and generally occurs 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, and sometimes it is more generalized.

#### 5. Environmental Causes

In this category, we can find solar radiation, including poikiloderma of Civatte and photoaging, heat and infrared radiation, and sulfur mustard-induced poikiloderma.

**-Solar radiation:** Poikiloderma of Civatte: Poikiloderma of Civatte is a common condition with a slowly progressive and irreversible course. The classic distribution of the lesions symmetrically involves the sides of the face and neck. Solar radiation has been proposed as the main culprit besides other factors such as genetics, low estrogen, and phototoxic reactions to chemicals in fragrances or cosmetics.

**-Photoaging:** Dermatoheliosis: It is the result of repeated solar injuries, and happens more in fair skin patients. The most common sites affected are the sun-exposed areas, particularly the face and scalp in the bald men.

**-Heat and infrared radiation:** Erythema ab igne: Erythema ab igne is characterized by localized mottled and reticulated poikilodermatous area resulting from repeated or prolonged exposure to heat and infrared radiation. The sites and the distribution of the lesions depend on the direction of the radiation; so, sheens are affected commonly as a result of putting stoves nearby.

**-Sulfur mustard-induced poikiloderma:** Sulfur mustard is a chemical weapon that 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 including poikilodermatous lesions.

#### 6. Iatrogenic causes: Drugs and Radiotherapy

**-Corticosteroids:** Corticosteroids are accused of many undesirable side effects including atrophy, telangiectasia, and cutaneous dyspigmentation. Therefore, it can be expected that a constellation of these features will eventually lead to a poikilodermatous appearance. This side effect is especially seen in chronic dermatologic conditions like atopic dermatitis and lichen planus in which topical corticosteroids with different potencies are used repeatedly and /or continuously over the same areas of disease involvement. (-Atopic dermatitis: It is encountered on the neck in the severe cases of adult atopic dermatitis. Lichen planus: It is rarely followed over months to years, by a network of poikilodermatous lesions.)

**-Hydroxyurea (hydroxycarbamide):** Hydroxyurea is commonly used for myeloproliferative disorders and rarely for severe psoriasis. Adverse reactions of hydroxyurea include both nondermatologic and dermatologic effects such as poikiloderma, ichthyosis, nail abnormalities, and skin malignancy.

**-Radiotherapy:** Chronic radiation dermatitis occurs after exposure to ionizing radiation used in tumor radiotherapy. In such conditions, persistent poikilodermatous changes are indicative of significant cutaneous injury.

#### 7. Neoplastic causes:

**-Mycosis fungoides:** Poikilodermatous Mycosis Fungoides (MF) is a variant of MF, which is sometimes identified as poikiloderma vasculare atrophicans by some authors. The lesions are predominantly located on the breast, hips, and gluteal areas, and may be associated with other classic patches and plaques of MF. 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.

**-Poikilodermatous (large plaque) parapsoriasis:** Large plaque parapsoriasis is considered a pre-lymphomatous skin condition that

overlaps early MF. Clinically, it presents with persistent large, 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 predilection sites include the lower aspect of the trunk, upper aspects of thighs, and flexural surfaces.

Approach to the patient with acquired poikiloderma:

Before entering this part, I would like to emphasize that there is no definite and specific therapeutic way to remove poikiloderma completely.

It is proposed a three step approach for the treatment of acquired poikiloderma:

- Prevent the cause: as poikiloderma is not a disease per se, for every given patient presenting with poikiloderma, the primary approach is focused on preventing its cause; for example proper photoprotection and avoidance of perfume use in case of poikiloderma of Civatte, avoidance of prolonged heat or infrared exposure for erythema ab igne, and following the safe guidelines for topical corticosteroid use.

- The second step consists of specific therapeutic modalities:

Laser therapy especially by argon, intense pulsed light (Civatte) and pulsed dye lasers (chronic radiation dermatitis) can relatively improve poikilodermatous lesions.

Hydroquinone-containing preparations may help fade the pigmentation associated with poikiloderma. Exfoliant agents including  $\alpha$ -hydroxy acids and topical retinoids may reverse some of the changes of dermatoheliosis including poikiloderma in some extents.

- Treat the cause: Treatment for poikilodermatous MF and poikilodermatous parapsoriasis includes their standard and classic treatments.

#### Conclusion

Poikiloderma is a divergent condition that has been attributed to different dermatologic conditions. 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 usually unsatisfactory.