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Idiopathic Poikiloderma Atrophicans Vasculare: A Diagnostic Challenge Mimicking Early Mycosis Fungoides - A Case Report

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ABSTRACT

Poikiloderma atrophicans vasculare (PAV) is characterized by epidermal atrophy, telangiectasia, and pigmentary alterations, producing a mottled reticular appearance. We report a young woman with chronic poikilodermatous plaques on the trunk. Clinicopathologic and immunohistochemical findings favored an inflammatory poikilodermatous process; however, because of focal epidermotropism, early poikilodermatous mycosis fungoides could not be excluded with absolute certainty, and clinical surveillance was recommended.

Keywords

Acquired poikiloderma, Congenital poikiloderma, Poikiloderma atrophicans vasculare, mycosis fungoides, poikilodermatous variant.

Introduction

Poikiloderma vasculare atrophicans (PVA) is a clinical term that describes the coexistence of cutaneous atrophy, telangiectasias, and hyperpigmentation, which confer a characteristic mottled-reticular appearance to the skin. Although congenital forms exist, such as Rothmund–Thomson syndrome, most cases encountered in clinical practice are acquired [1,2].

From a histopathological perspective, poikiloderma is characterized by a pattern of predominantly nonspecific changes shared among its various etiologies [3]. Epidermal atrophy with thinning of the Malpighian layer is observed, associated with hydropic degeneration of the basal layer, as well as pigmentary incontinence evidenced by the presence of melanophages in the papillary dermis [4,5].

Concomitantly, dilation of capillaries in the papillary dermis is frequently identified, contributing to the telangiectatic clinical component. In addition, more specific histopathologic findings may be recognized depending on the underlying condition; in this context, epidermotropism of atypical lymphocytes constitutes a distinctive feature of mycosis fungoides, particularly in its poikilodermatous variant [6-8].

Case Report

A 37-year-old woman with a history of atopic dermatitis since 2000 and polycystic ovary syndrome since 2019, as well as prior cholecystectomy (2014), myomectomy, and teratoma resection (2019), reported the onset of a dermatosis involving the trunk in 2020. The eruption presented with similar characteristics to the current lesions and was diagnosed at that time as a fixed drug eruption, for which she was treated with 1% hydrocortisone cream.

In December 2024, she presented to a private dermatology clinic with a disseminated dermatosis involving the trunk (Figure 1A), affecting both the anterior and posterior chest. The lesions were characterized by erythema, atrophy, and hyperpigmentation, forming oval reticulated plaques approximately 8 cm in diameter, slightly infiltrated, consistent with poikilodermatous plaques, and associated with mild pruritus (Figure 1B).

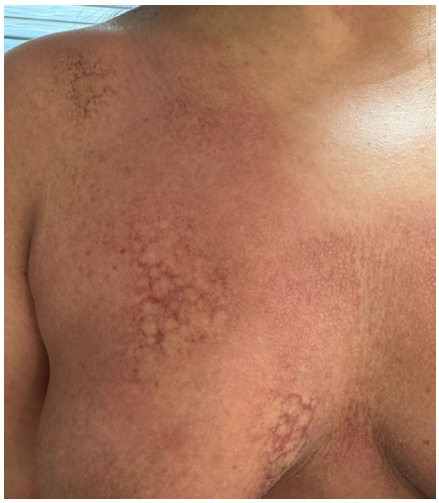

Figure 1A: Topography: Anterior chest, right breast region, showing circular poikilodermatous plaques.

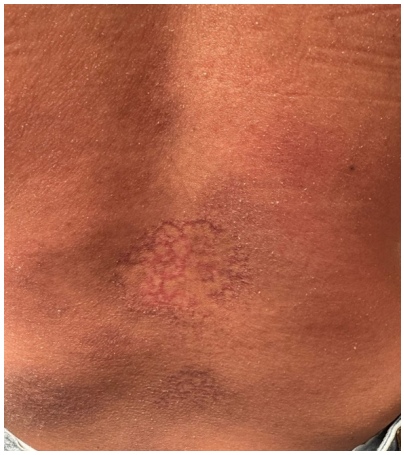

Figure 1B: Morphology: Characteristic reticulated lesions with epidermal atrophy, telangiectasias, and hyperpigmentation in lower back.

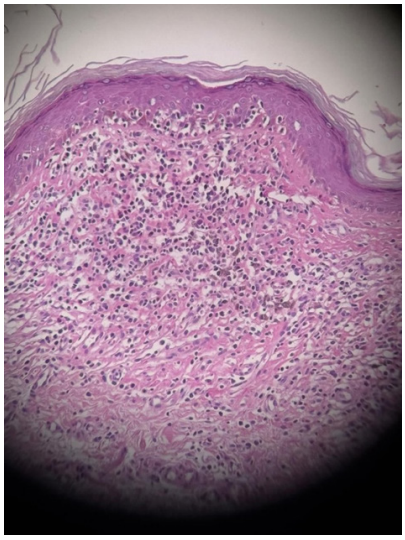

Figure 1C: Photomicrograph (H&E)**:** Lamellar orthokeratosis overlying a thinned epidermis with areas of atrophy and flattening of the rete ridges. Epidermotropism of lymphocytes is observed.

Histopathologic examination revealed lamellar orthokeratosis overlying a thinned epidermis with areas of atrophy and flattening of the rete ridges. Epidermotropism of lymphocytes was observed, with cells displaying enlarged, round to irregular, hyperchromatic nuclei surrounded by a clear halo, arranged in small clusters in focal areas more evident with PAS and Masson's trichrome stains (Figures 1C, 1D).

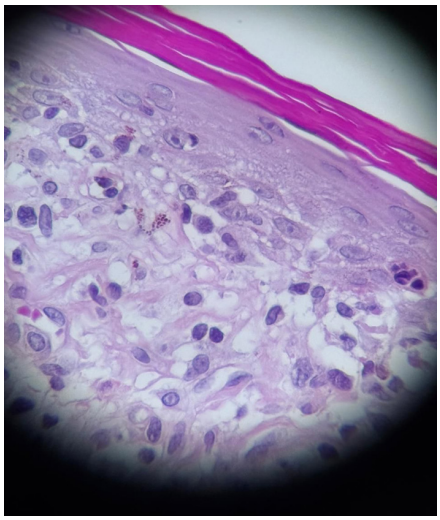

Figure 1D: Photomicrograph (PAS stain and trichrome): Epidermal and dermal lymphocytes displaying enlarged, round to irregular, hyperchromatic nuclei surrounded by a clear halo, arranged in small focal clusters.

In the papillary dermis, a moderate lymphocytic infiltrate with mild cytologic variability/nuclear irregularity. The remaining sections showed dilated capillaries, melanophages, and adnexal structures.

Immunohistochemistry showed a predominantly T-cell infiltrate (CD3+), with CD4- and CD8-positive lymphocytes. CD30 highlighted scattered reactive cells, and CD20 was negative. The overall immunophenotype was not diagnostic of cutaneous lymphoma (Figures 2A-D). CD7 AND CD20 was negative.

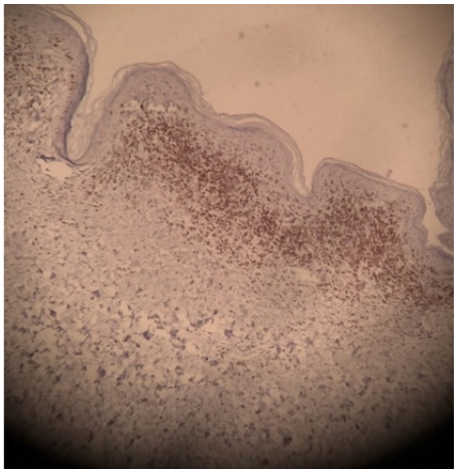

Figure 2A: Immunohistochemistry showing a reactive CD4-positive pattern.

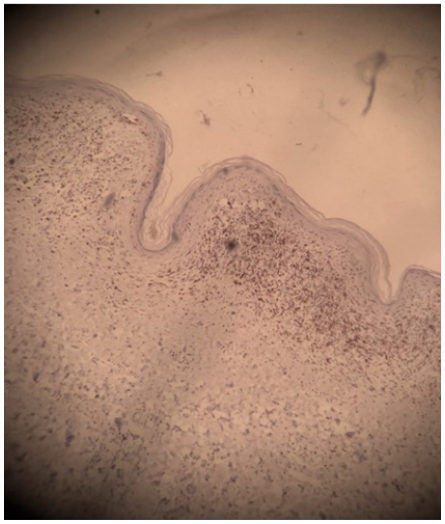

Figure 2B: Immunohistochemistry showing a reactive CD30-positive pattern.

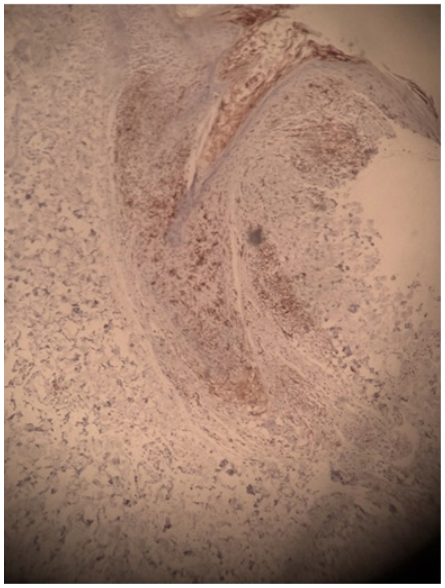

Figure 2C: Immunohistochemistry showing a negative CD7-negative pattern.

Laboratory evaluation, including complete blood count, comprehensive metabolic panel, thyroid function tests, immunologic profile, antinuclear antibodies, anti–double-stranded DNA, anti-Sm, anti–Jo-1, PL-7, PL-12, EJ, OJ, SRP, Mi-2 alpha and beta, MDA5, TIF1-γ, and NXP antibodies, was unremarkable.

Based on clinical, histopathologic, and immunohistochemical correlation, a diagnosis of idiopathic poikiloderma atrophicans vasculare was favored. Given the focal epidermotropism, continued clinical surveillance and repeat biopsy were recommended if the lesions progressed or changed.

Treatment was initiated with phototherapy and high-potency topical corticosteroids, resulting in clinical improvement at 40 days of Clinical follow-up and repeat biopsy were recommended if lesions progressed or changed. However, due to exacerbation of atopic dermatitis, methotrexate 15 mg weekly with folic acid supplementation (50 mcg) was prescribed for 3 months, leading to a reduction in poikiloderma and infiltration, with residual post-inflammatory hyperpigmentation.

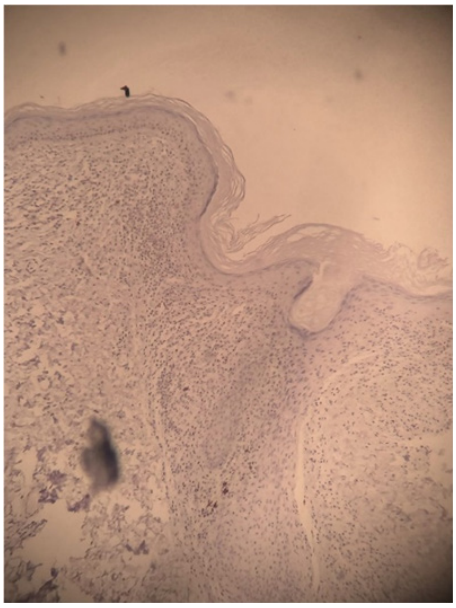

Figure 2D: Immunohistochemistry negative for CD20.

Discussion

Poikiloderma vasculare atrophicans (PVA) represents a complex clinical and histopathologic pattern that may be associated with multiple clinical entities, both benign and malignant. In this case, a young patient with a personal history of atopic dermatitis and hormonal disorders presented with a chronic dermatosis of insidious onset and clinical features consistent with poikiloderma (atrophy, telangiectasias, and hyperpigmentation), which necessitated a thorough evaluation to exclude systemic or neoplastic causes [7-9].

Various causes of acquired poikiloderma have been described in the literature; however, a universally accepted classification has not been established. According to Ahmad Nofal et al., in a study published in 2013, an etiologic classification is proposed based on different categories, including infectious causes (e.g., acrodermatitis chronica atrophicans associated with Borrelia burgdorferi), inflammatory processes (such as lichen planus), metabolic diseases (such as cutaneous amyloidosis), connective tissue disorders (including lupus erythematosus and dermatomyositis), environmental exposures (ultraviolet radiation, infrared heat, or chemical exposures such as mustard gas), iatrogenic factors (medications such as corticosteroids or hydroxyurea, and radiotherapy) and neoplastic diseases, particularly variants of mycosis fungoides (Table 1).

Infectious

This category corresponds to chronic infectious processes that

induce progressive cutaneous damage mediated by persistent inflammation. A classic example is infection with Borrelia burgdorferi, in which Lyme disease triggers a sustained immune response leading to epidermal atrophy, vascular damage, and pigmentary alterations, ultimately resulting in a poikilodermatous pattern [10].

Acrodermatitis chronica atrophicans typically develops months to years after Borrelia infection, most commonly affecting extensor surfaces of the lower extremities; however, involvement of the trunk and, rarely, the face has also been reported. Histopathologically, it is associated with a dermal infiltrate rich in plasma cells, a subepidermal zone of degenerated connective tissue, and progressive atrophy. Diagnosis is supported by serology or polymerase chain reaction (PCR), although the spirochete may also be identified histologically using Warthin–Starry staining [11,12].

Inflammatory

This group is associated with chronic inflammatory dermatoses (such as atopic dermatitis graft-versus-host disease, lichen planus), in which persistent immune activation disrupts epidermal and dermal homeostasis. Sustained inflammation, along with damage to the dermoepidermal junction and defective tissue repair, promotes atrophy, telangiectasias, and pigmentary incontinence.

In the context of atopic dermatitis, poikiloderma may be interpreted as a consequence of chronic and persistent cutaneous inflammation that progressively alters epidermal and dermal architecture [11,12]. This process favors the development of epidermal atrophy, pigmentary incontinence, and superficial vascular dilation, which together may result in a poikilodermatous pattern.

Although our patient had atopic dermatitis, there is no clear association in the literature, other than as a consequence of chronic treatment.

Furthermore, chronic scratching and persistent inflammation contribute to structural skin damage and post-inflammatory hyperpigmentation, reinforcing the characteristic reticular clinical appearance [13,14].

Metabolic

This category is related to deposition disorders or metabolic alterations affecting the skin, such as cutaneous amyloidosis. The accumulation of abnormal substances (e.g., amyloid) within the dermis interferes with normal skin architecture, leading to structural changes that clinically manifest as poikiloderma [14].

Connective Tissue Diseases

This group includes autoimmune conditions such as lupus erythematosus, dermatomyositis, and systemic sclerosis. In these disorders, immune-mediated damage directed against cutaneous components results in vascular alterations, fibrosis, and degeneration of the dermoepidermal junction, favoring the development of a poikilodermatous phenotype [15].

Environmental

Environmental causes result from cumulative damage induced by external physical or chemical agents. Ultraviolet radiation promotes oxidative stress, DNA damage, and solar elastosis; infrared radiation or heat exposure leads to chronic thermal injury with degeneration of connective tissue; and chemical toxins cause direct cytotoxicity and vascular alterations [16,17].

Table 1: Poikiloderma causes and histopathologic findings.

Category	Causes	Histopathologic Findings
Infectious	*Borrelia burgdorferi* (Acrodermatitis chronica atrophicans of Lyme disease)	Destruction of cutaneous adnexal structures, degenerated subepidermal connective tissue, and dense dermal infiltrate rich in plasma cells. *Borrelia* spirochetes may be identified using Warthin–Starry stain.
Inflammatory	Atopic dermatitis	Pigmentary incontinence (melanophages in the papillary dermis), and capillary dilation. Spongiotic dermatitis.
	Chronic graft-versus-host disease	Dermal fibrosis and destruction of adnexal structures.
	Lichen planus–induced poikiloderma	Poikiloderma, hyperkeratosis, wedge-shaped hypergranulosis, and dense band-like lymphocytic infiltrate in the papillary dermis.
Metabolic	Poikilodermatous cutaneous amyloidosis	Poikiloderma with eosinophilic amyloid deposits. Congo red positive with apple-green birefringence under polarized light
Connective Tissue Diseases	Lupus Erythematosus	Irregular lymphocytic infiltrate with poikilodermatous changes.
	Dermatomyositis	Poikilodermatous changes
	Systemic sclerosis	Dermal fibrosis
Environmental	Ultraviolet radiation (Civatte poikiloderma, photoaging)	Solar elastosis with poikilodermatous changes.
	Dermatoheliosis	
	Infrared radiation (erythema ab igne)	Poikiloderma with basophilic degeneration of connective tissue.
	Chemical exposure (mustard gas)	Poikilodermatous changes.
Iatrogenic	Drugs: topical/systemic corticosteroids, hydroxyurea	Poikilodermatous changes.
	Radiotherapy (chronic radiation dermatitis)	Poikilodermatous change and dermal fibrosis
Neoplastic	Poikilodermatous mycosis fungoides Poikilodermatous parapsoriasis	Poikilodermatous changes. Atypical lymphoid infiltrate in the papillary dermis with epidermotropism without significant spongiosis. CD4+ T lymphocytes with cerebriform nuclei, Pautrier microabscesses, and dermal fibrosis.

The common denominator among these factors is chronic damage affecting the dermal matrix and microvasculature.

Iatrogenic

Iatrogenic causes are secondary to medical interventions, particularly pharmacologic treatments such as corticosteroids or hydroxyurea, which induce cutaneous atrophy, vascular alterations, and melanocyte disruption. Another common cause is radiotherapy, which produces direct DNA damage, dermal fibrosis, and telangiectasias due to vascular injury—mechanisms that converge into structural changes consistent with poikiloderma.

Neoplastic

This category is primarily associated with cutaneous lymphomas, particularly mycosis fungoides. In this context, infiltration by neoplastic T lymphocytes with epidermotropism disrupts normal skin architecture, leading to characteristic atrophic, vascular, and pigmentary changes [18] (Figure 3).

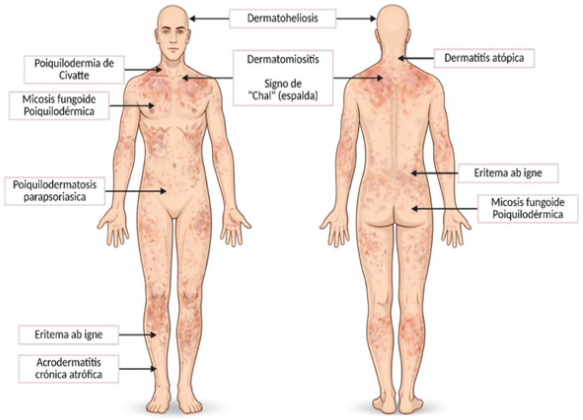

Figure 3: Schematic representation of the distribution of acquired poikiloderma.

Conclusion

Idiopathic PAV is a diagnosis of exclusion that requires careful clinicopathologic correlation because its clinical and histologic features may overlap with early poikilodermatous mycosis fungoides. In this patient, the overall findings favored a reactive/inflammatory poikilodermatous process, and the immunohistochemical profile did not provide sufficient evidence for cutaneous lymphoma. Nevertheless, focal epidermotropism warrants continued clinical surveillance and repeat biopsy if lesions evolve. The favorable response to anti-inflammatory treatment further supports a non-neoplastic process, although treatment response alone cannot definitively exclude early MF.

Acquired PAV is a cutaneous manifestation with diverse etiologies that may mimic serious conditions such as cutaneous lymphomas. A careful clinicopathologic approach did not provide sufficient evidence to diagnose cutaneous lymphoma at this time. Treatment with high-potency topical corticosteroids and phototherapy proved effective, with significant resolution of inflammatory lesions.

This wording is safer scientifically and consistent with the revised interpretation: inflammatory/idiopathic PAV is favored, but early MF is not claimed to be absolutely excluded.

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