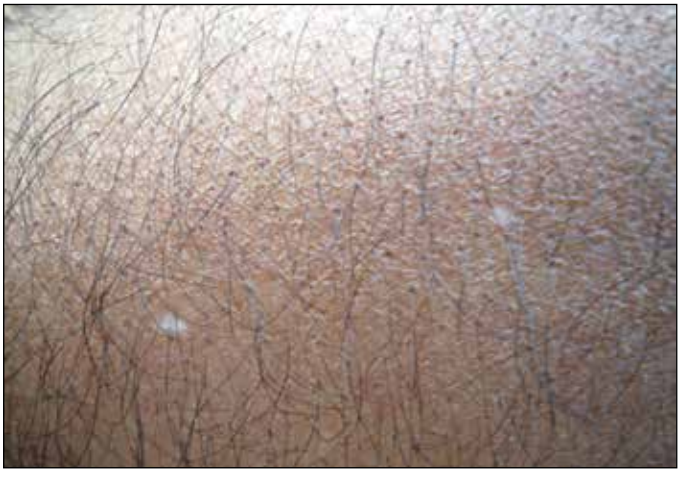
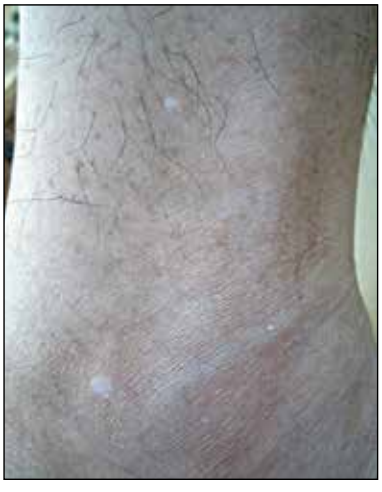
What Is Your Diagnosis?





A 58-year-old man presented with disseminated, hypopigmented, asymptomatic lesions on the right arm (top) and left leg (bottom) that had been present for approximately 6 years. The patient reported that the lesions had become more visible and greater in number within the last year. Multiple circular hypopigmented macules of various sizes ranging from 1 to 3 mm in diameter were identified. No scaling was seen. Physical examination was otherwise unremarkable.

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The Diagnosis: Idiopathic Guttate Hypomelanosis

A biopsy of the largest lesion from the left leg superior to the lateral malleolus was performed. Histopathologic examination revealed solar elastosis, diminished number of focal melanocytes and pigment within keratinocytes compared to uninvolved skin, and presence of hyperkeratosis with flattening of rete ridges. The clinical presentation along with histopathologic analysis confirmed a diagnosis of idiopathic guttate hypomelanosis (IGH). The lesions were treated with short-exposure cryotherapy, which resulted in partial repigmentation after several treatments.

Idiopathic guttate hypomelanosis is a common but underreported condition in elderly patients that usually presents with small, discrete, asymptomatic, hypopigmented macules. The frequency of IGH increases with age.[1] Frequency of the condition is much lower in patients aged 21 to 30 years and does not exceed 7%. Lesions of IGH have a predilection for sun-exposed areas such as the arms and legs but rarely can be seen on the face and trunk. Facial lesions of IGH are more frequently reported in women.[1] The size of lesions can be up to 1.5 cm in diameter. The condition generally is self-limited, but some patients may express aesthetic concerns. Rare cases of IGH in children have been associated with prolonged sun exposure.[2]

The etiology of IGH is unknown but an association with sun exposure has been noted. Patients with IGH frequently show other signs of photoaging, such as numerous seborrheic keratoses, solar lentigines, xeroses, freckles, and actinic keratoses.[1] Short-term exposure to UVB radiation and psoralen plus UVA therapy has been shown to cause IGH in patients with chronic diseases such as mycosis fungoides.[3-5] One small study that examined renal transplant recipients determined an association between HLA-DQ3 antigens and IGH, whereas HLA-DR8 antigens were not identified in any patients with IGH, indicating it may have some advantage in preventing the development of IGH.[6] Shin et al[1] reported that IGH was prevalent among patients who regularly traumatized their skin by scrubbing.

Clinically, IGH should be differentiated from other conditions characterized by hypopigmentation, such as pityriasis alba, pityriasis versicolor, postinflammatory hypopigmentation, progressive macular hypomelanosis, and vitiligo. Aside from clinical examination, histopathologic studies are helpful in making a definitive diagnosis. The differential diagnosis of IGH is presented in the Table.

Histopathology of IGH lesions usually reveals slight atrophy of the epidermis with flattening of rete ridges and concomitant hyperkeratosis. A thickened stratum granulosum also has been noted in lesions of IGH.[2] The diminished number of melanocytes and melanin pigment granules along with hyperkeratosis both appear to contribute to the hypopigmentation noted in IGH.[7] Ultrastructural studies of lesions of IGH can confirm melanocytic degeneration and a decreased number of melanosomes in melanocytes and keratinocytes.[2,8]

There is no uniformly effective treatment of IGH. Topical application of tacrolimus and tretinoin have shown efficacy in repigmenting IGH lesions.[8,9]

Differential Diagnosis of Idiopathic Guttate Hypomelanosis

Condition	Clinical Presentation	Histopathologic Presentation
Idiopathic guttate hypomelanosis	Well-circumscribed areas of hypopigmentation commonly presenting on the extremities	Solar elastosis, diminished number of focal melanocytes and melanin, along with presence of hyperkeratosis with flattening of rete ridges; atrophy of the epidermis may be present
Pityriasis alba	Hypopigmented patches with fine scaling; association with atopic dermatitis (eczema) is frequent	Spongiosis within the epidermis with accompanying hyperkeratosis and local areas of parakeratosis in addition to decreased and irregularly distributed melanin within the stratum basale; no notable change in melanocyte count; a perivascular lymphocytic infiltrate may be visible with exocytosis of lymphocytes

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Condition	Clinical Presentation	Histopathologic Presentation
Pityriasis versicolor	Hypopigmented or hyperpigmented macules with a predilection for the upper trunk and shoulders; fine scales can be seen	Slight parakeratosis and hyperkeratosis, inflammatory infiltrate; melanin pigment is decreased in hypopigmented variants and increased in hyperpigmented macules; melanocyte count is not changed; "spaghetti and meatballs" hyphae and spores can be seen in the stratum corneum
Postinflammatory hypopigmentation	Lesions appear after inflammatory insult; a mixture of hypopigmented and hyperpigmented macules may be seen	Scant inflammatory infiltrate with melanophages; decreased or absent melanin granules with normal melanocyte count
Progressive macular hypomelanosis	Well-circumscribed areas of hypopigmentation on the trunk, typically without scale; often responds to treatment with benzoyl peroxide, suggesting a pathogenic role for *Propionibacterium acnes*	Hypopigmentation, hyperkeratosis, and flattening of rete ridges; diminished melanin pigmentation with normal melanocyte count
Vitiligo	Well-circumscribed, depigmented macules and patches	Absence of melanocytes in the stratum basale with concomitant loss of melanin in keratinocytes

Short-exposure cryotherapy with a duration of 3 to 5 seconds, localized chemical peels, and/or local dermabrasion can be helpful.[10-12] CO_2 lasers also have demonstrated promising results.[13]

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