A mysterious white rash

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CASE

A 40-year-old Vietnamese man presented for an asymptomatic rash on his back and chest that had been present for the past decade. Over the years, he was treated by multiple providers, including 7 dermatologists. The rash had not improved despite treatment with topical corticosteroids, calcineurin inhibitors, various bland emollients, ketoconazole cream and shampoo, oral fluconazole, and various over-the-counter antifungal medications. On examination, the patient had nonscaly, hypopigmented macules coalescing into patches on his chest and back (Fig 1). He denied itching, pain, or other symptoms. Potassium hydroxide test result was negative and biopsy revealed no pathologic changes. Wood’s lamp examination revealed perifollicular, punctiform, red-orange fluorescence (Fig 2).

Question 1: What is the most likely diagnosis?

A. Pityriasis alba
B. Pityriasis (tinea) versicolor
C. Postinflammatory hypopigmentation
D. Progressive macular hypomelanosis (PMH)
E. Vitiligo

Answers:

A. Pityriasis alba—Incorrect. Pityriasis alba is a skin condition of hypopigmentation that is associated with atopic dermatitis. It predominantly affects children, and lesions typically involve the face. Wood’s lamp examination does not accentuate the hypopigmentation or exhibit perifollicular fluorescence.

B. Pityriasis (tinea) versicolor—Incorrect. Like PMH, tinea versicolor is characterized by macular areas of hypopigmentation that typically involve the chest and upper portion of the back. However, tinea versicolor would be expected to improve with antifungal medications and does not exhibit red-orange perifollicular fluorescence under Wood’s lamp examination. Instead, black-light illumination of skin affected by tinea versicolor reveals a yellow-orange fluorescence of the yeast. Additionally, potassium hydroxide examination characteristically reveals hyphae and yeast in a “spaghetti and meatballs” pattern.

C. Postinflammatory hypopigmentation—Incorrect. Postinflammatory hypopigmentation describes the loss of pigment in the skin after the resolution of an inflammatory process. The pathogenesis is thought to be due to the loss of functional melanocytes but is not completely understood. Wood’s lamp examination does not accentuate hypopigmentation or reveal perifollicular fluorescence.

D. PMH—Correct. PMH is a skin condition of hypopigmentation that was undescribed in the scientific literature until 1988. It is clinically characterized by macular areas of hypopigmentation, typically on the chest or back, which exhibit perifollicular red-orange fluorescence under Wood’s lamp examination. This characteristic finding with such an examination was first described by Wu et al in 2010. In their study of 21 patients with a previous diagnosis of PMH and with a previous negative potassium hydroxide test result, all 21 had perifollicular fluorescence with the application of black light. This finding has not been shown to be associated with any other hypomelanotic conditions and may therefore be considered pathognomonic for PMH. Currently, there are reports of cases studied with dermatoscopy; however, in hospitals in which there is no such tool or experience is limited, the use of Wood’s lamp is an excellent method of differentiating PMH from other conditions of hypopigmentation, thus avoiding misdiagnoses and unnecessary treatments. This report supports the utility of Wood’s lamp as a diagnostic tool for hypopigmented lesions.

E. Vitiligo—Incorrect. Vitiligo is an acquired disorder characterized by macules and patches of depigmentation that result from a progressive loss of functional melanocytes. Although clinical presentation may be similar to that of PMH, Wood’s lamp examination of areas affected by vitiligo does not reveal red-orange perifollicular fluorescence and demonstrates complete depigmentation rather than the hypopigmentation of PMH.

Question 2: Which of the following is associated with this condition?

A. Malassezia furfur
B. Pernicious anemia
C. Pityrosporum orbiculare
D. Cutibacterium acnes (formerly Propionibacterium acnes)
E. Squamous cell carcinoma

Answers:

A. Malassezia furfur—Incorrect. Malassezia species are associated with pityriasis (tinea) versicolor, not PMH.
B. Pernicious anemia—Incorrect. Noncutaneous autoimmune disorders, such as Grave disease and pernicious anemia, have been shown to occur with higher incidence in patients with vitiligo. No association has been shown to exist between PMH and autoimmune disorders.

C. P orbiculare—Incorrect. P orbiculare has been suggested to be associated with confluent and reticulated papillomatosis because of its prevalence in some lesions in yeast form; however, this is not largely accepted as the pathogenic mechanism of confluent and reticulated papillomatosi because many lesions have not been found to contain the yeast. To our knowledge, it has no role in the development of PMH.

D. C acnes—Correct. In recent years, an association between PMH and C acnes colonization within the pilosebaceous unit has been consistently demonstrated. A certain subtype of the organism, phylogenetic type III, seems to be responsible. The characteristic hypomelanotic lesions are thought to result from bacterial production of a depigmentation factor that interferes with melanin synthesis, distribution, or both. This hypothesis is largely due to histopathologic analysis of lesions, which demonstrate decreased melanin content and melanosome distribution in the setting of a normal quantity and morphology of melanocytes. Additionally, the bacteria produce a type of porphyrin, explaining the diagnostic per-follicular fluorescence on Wood’s lamp examination.

E. Squamous cell carcinoma—Incorrect. The development of squamous cell carcinoma represents a long-term risk in skin affected by erythema ab igne. This association is particularly observed in patients exposed to hydrocarbon-fueled heat sources such as coal. No such association between PMH and cutaneous malignancy has been shown.

Question 3: Which of the following is an effective treatment regimen?

A. Benzoyl peroxide and clindamycin—Correct. Further supporting a bacterial etiology, antimicrobial treatments such as benzoyl peroxide 5% wash and topical clindamycin 1% that are traditionally used in acne management have proven to be successful in clearing PMH lesions. In the case of our patient, the combination of doxycycline 100 mg orally daily, clindamycin lotion, and benzoyl peroxide wash for 4 weeks was successful in achieving complete clearance. No lesions were observed at 3-month follow-up (Fig 3).

B. Ketoconazole shampoo—Incorrect. Ketoconazole shampoo has not been shown to be effective in treating PMH lesions.

C. Psoralen plus ultraviolet A—Incorrect. Psoralen plus ultraviolet A has not been shown to be an effective treatment for PMH.

D. Topical corticosteroids—Incorrect. Topical corticosteroids have not been shown to be effective in clearing PMH lesions.

E. Topical calcineurin inhibitors—Incorrect. Topical calcineurin inhibitors have not been shown to be effective in clearing PMH lesions.

Abbreviation used:
PMH: progressive macular hypomelanosis

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