A Case of Progressive Macular Hypopigmentation

Mehreen Sheikh, BS,* Jocelyn LaRocque, DO, FAOCD**

*Medical student, 4th year, Lincoln Memorial University DeBusk College of Osteopathic Medicine, Harrogate, TN
**Private-practice dermatologist, Charlotte, NC

Disclosures: None
Correspondence: Mehreen Sheikh, BS; mehreen.sheikh@lmunet.edu

Abstract

Progressive macular hypomelanosis (PMH) is defined as a localized loss of pigmentation seen mostly on the trunk, sometimes with extension into the upper extremities and head/neck region. The pathogenesis of PMH is not known, but it has been speculated that certain strains of P. acnes residing in hair follicles may produce a depigmenting factor, which causes PMH. This case presents a female with PMH lesions located on her back, neck, and lower extremities bilaterally. Herein, we discuss PMH pathogenesis, clinical presentation, differential diagnosis, and patient management.

Introduction

Progressive macular hypomelanosis (PMH) is a localized loss of pigmentation presenting mostly commonly on the trunk, though it may extend to the upper extremities and head/neck region.1,2 Hypopigmented, confluent macules arise without prior onset of injury, inflammation, or known infection.2 They are ill-defined and non-scaly in nature, with a predilection for young females aged 18 to 25 years old.2,3 Affected individuals most commonly have skin type IV, which may be due to the ease of lesion recognition in contrast to the surrounding pigmented skin.4 The pathogenesis of PMH is not known; however, it has been speculated that certain strains of P. acnes residing in hair follicles may produce a depigmenting factor, which causes PMH.1 Treatments have been geared toward P. acnes reduction and repigmentation with the use of antimicrobial, ultraviolet light and anti-inflammatory therapies. Success has been shown in the use of antimicrobials and UV light.1,2 More recent studies have shown the successful use of isotretinoin in PMH lesions, further supporting an underlying P. acnes origin of this disease.3

PMH is not a commonly diagnosed entity. Dermatologists may want to consider the condition when hypopigmented lesions do not respond to typical therapies like topical steroids and anti-fungal treatments. A case of PMH is described here to highlight the importance of recognizing this cutaneous disorder.

Case Study

A 58-year-old female with skin type IV presented with pigment loss of unknown cause, most prominently located on the midline of her lower back, neck, and lower extremities bilaterally (Figures 1, 2). The lesions had been present for months, and previous treatments included clobetasol propionate and betamethasone dipropionate, which did not lead to improvement of the condition. Clinically, she had asymptomatic, ill-defined, hypopigmented, and non-scaly macules that were coalescing into patches. They had rapidly progressed over several weeks. The lesions were not inflammatory. Distribution did not follow Blaschko’s lines. The non-affected skin appeared uniformly tan in comparison to the hypopigmented regions.

The patient’s only past medical history included hypothyroidism and hypercholesterolemia.

A 4 mm punch biopsy was obtained from the right lower back. Pathology showed subtle changes that included mild, scattered superficial perivascular inflammation and dermal pigment deposition (Figures 3, 4). These results clinically and histologically support PMH, where other findings have included a ring of pigment around the dermal papillae, but decreased melanin content.9

Upon follow-up, the patient received benzoyl peroxide wash daily and minocycline HCl 100 mg capsules BID for eight weeks. The patient discontinued minocycline after one week due to GI upset and was switched to doxycycline 100 mg capsules BID for the remaining seven weeks.

Appearance of lesions was improved at the eight-week appointment. The hypopigmented patches were still present, but repigmentation had occurred compared to initial presentation, and the lesions were not spreading.

Discussion

In the 1980s, Guillette et al. first defined PMH as a skin pigment disorder designated for people of mixed ethnicity. It is currently postulated that PMH presents in skin types IV-VI.1 Many terms around the world have been used to describe PMH, such as “Creole dyschromia” and “cutis trunci variata.”

Because of the variation in names of PMH, it is important to elucidate an etiology behind the hypopigmentation. A study conducted by Westerhof et al. showed that biopsies from sites of hypopigmentation contained a mild perifollicular lymphocytic infiltrate as well as positive P. acnes from cultures.2,4 Wood’s lamp illumination over the hypopigmented areas produced a red-colored follicular fluorescence.1 Healthy, pigmented skin, when observed histologically and cultured, did not show signs of infiltrate or bacteria.

Figure 1

Figure 1. Right flank, showing active, hypopigmented, confluent, non-scaly macules.

Figure 2

Figure 2. Lower back, showing hypopigmented, confluent macules that do not follow Blaschko’s lines.

Figure 3

Figures 3 and 4. Pathology shows subtle changes including mild, scattered superficial perivascular inflammation and dermal pigment deposition.
PMH is often unrecognized in the clinical setting and mistaken for other skin disorders that present similarly. Once a diagnosis of PMH is considered, several other differentials must be ruled out, including, but not limited to, pityriasis alba, vitiligo, tinea versicolor, lichen sclerosis, and mycosis fungoides. Pityriasis alba also presents with patches of hypopigmentation. However, it is most commonly seen in children and adolescents, whereas PMH is mostly seen in females aged approximately 20 to 30 years old.6 Furthermore, pityriasis alba lesions present with fine scale, which is not present in PMH.7 Vitiligo may also be part of the differential; however, this is a process attacking melanocytes that results in loss of pigmentation, rather than a decrease in the amount of pigmentation.8 Histologically, this patient had melanocytes present that were confirmed by immunohistochemical staining, further arguing against a diagnosis of vitiligo. Tinea versicolor can also cause pale patches distributed on the trunk; however, these lesions are scaly, whereas the lesions of PMH are not. Tinea versicolor is also fungal in origin, and the patient presenting with PMH will fail traditional topical anti-fungal treatments for suspected tinea versicolor. Other considerations for hypomelanosis include mycosis fungoides and lichen sclerosis, but pathology did not support either of these.

PMH proves to be a diagnosis of exclusion. Recent studies have argued in favor of a pathogenesis of Propionibacterium acnes as the underlying cause of PMH, and the variation in subtypes that can cause the hypopigmentation.9 P. acnes is found in pilosebaceous glands, which are in high concentration on the trunk. However, acne is not a nidus for PMH nor does it worsen in those who have PMH.10 Different strains of P. acnes could explain why those who have facial acne do not experience the hypopigmentation found from the strains of PMH. One study conducted by Relyveld et al. employed amplified fragment length polymorphism for the 16S rRNA gene of P. acnes to elucidate the differences between strains in acne and PMH. Of the 14 patients studied, eight P. acnes strains were substantially different from those with acne.10

With the possible pathogenesis of P. acnes, treatment has been geared toward resolving the underlying acne strains. A study conducted by Relyveld et al. used a sample size of 45 patients to test the effectiveness of repigmentation between the treatments of 5% benzoyl peroxide hydrogel/1% clindamycin lotion in combination with UVA irradiation compared to 0.05% fluticasone propionate cream in combination with UVA irradiation. The results showed that antimicrobial treatment for repigmentation was superior (photometric measurements P=.007, patient assessment P<.0001, and dermatologist assessment P=.0001).1 This sheds further light on the source behind PMH.

Another study employed narrowband UVB (NB-UVB) treatment for patients with PMH. More than 90% repigmentation was documented for nine of the 16 patients who received biweekly NB-UVB.2 Though the sample size was small, this may suggest another means of therapy for those with PMH.

More recently, a case study showed improvement of PMH pigmentation with oral isotretinoin treatment. The patient had been treated for PMH and rosacea with NB-UVB without success of repigmentation. Once 10 mg daily oral isotretinoin was added to the regimen for treatment of rosacea, the patient experienced resolution of PMH lesions within one month. These lesions did not return once isotretinoin was discontinued. This could be due to the reduction of sebum found in the pilosebaceous unit alongside P. acnes,11 further supporting P. acnes as the pathogenesis of PMH.

One PMH study has shown positive effects with the use of isotretinoin. This patient was prescribed benzoyl peroxide as well as 100 mg doxycycline to treat her hypopigmented lesions. At eight-week follow-up, repigmentation was evident on the trunk and back, again lending credence to the P. acnes theory.

### Conclusion

PMH is a diagnosis of exclusion, and it is often misdiagnosed. Patients are often prescribed corticosteroids or antifungals. The pathogenesis behind PMH is not completely understood, but the identification of P. acnes strains within the pilosebaceous units of affected skin indicates a possible source. Further studies are needed to understand the relationship between P. acnes and the pigmentation changes of PMH; however, dermatologists should consider this diagnosis in a patient presenting with dyschromia.

### References