

Progressive macular hypomelanosis: a report of three cases

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Abstract Progressive macular hypomelanosis (PMH) is an acquired disorder of skin pigmentation, of uncertain etiology, characterized by asymptomatic hypopigmented macules, predominantly located on the trunk. There are several treatment options available, although topical clindamycin and benzoyl peroxide have been used traditionally. Good results have recently been achieved using narrow-band ultraviolet B (NB-UVB) phototherapy. Herein three cases one male and two females, belonging to ethnic Kashmiri population with progressive macular hypomelanosis are reported in view of the clinical rarity of this condition.

Key words

Progressive macular hypomelanosis, hypopigmented macules, narrow-band ultraviolet B.

Introduction

Progressive macular hypomelanosis (PMH) is an uncommon skin disorder characterized by ill-defined nummular, non-scaly hypopigmented confluent macules on the trunk, often in and around the midline, and rarely extending to the proximal extremities and neck/head region. There is normal sensation without any history of itching, pain, or preceding infection, trauma or inflammation.^{1,2} Progressive macular hypomelanosis has been identified in Black people living in or originating from tropical countries. It is also more often seen in young females. Histologically, PMH is characterized by diminished pigment in the epidermis with a normal-looking dermis. Electron microscopy shows a shift from large melanosomes in normal-looking skin to small aggregated, membrane-bound melanosomes in hypopigmented skin. The exact pathogenesis of

progressive macular hypomelanosis is unknown; however, recent studies suggest hypopigmentation results from decreased melanin formation and altered melanosome distribution in response to *Propionibacterium*. This condition is frequently misdiagnosed and treated inadequately with antifungals or topical steroids resulting in patient frustration. The treatment options that have been described in the literature include phototherapy (PUVA, UVA or narrow band UVB), topical benzoyl peroxide 5%, topical clindamycin 1% and oral doxycycline.

Case Reports

Case 1

A 28-year-old male presented with hypopigmented lesions on his trunk and limbs that had appeared when he was 16 years of age and progressed over these years. There was no history of itching or dysesthesia. There was no history suggestive of eczema, dermatitis, atopy or any fungal infections. Cutaneous examination revealed symmetrically distributed

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Figure 1 Symmetrically distributed hypopigmented macules with well-defined borders and a non-scaly surface.



Figure 2 Well-defined, non-scaly, round or oval, macules more pronounced over trunk and limbs.

hypopigmented macules, about 2-3 mm in size, round or oval in shape with well-defined borders and a non-scaly surface, more pronounced over trunk and proximal aspects of limbs, sparing the head and neck (**Figure 1**).

Case 2

A 30-year-old female presented with multiple small hypopigmented lesions over trunk and limbs from last 5 years. These lesions were



Figure 3 Multiple hypopigmented nummular macules, located symmetrically on the anterior and posterior aspects of trunk.

asymptomatic, not associated with any pain or pruritus and were not preceded by any inflammatory skin lesions. Cutaneous examination revealed multiple small hypopigmented macules 2-4 mm in size, distributed bilaterally over trunk and limbs. These macules were well-defined, non-scaly, round or oval, more pronounced over trunk and limbs, sparing the head and neck (**Figure 2**).

Case 3

A 35-year-old female presented with multiple hypopigmented lesions over the trunk. These lesions appeared at an age of 25 years and since then were increasing in number and size. There had been no precedents of inflammatory skin lesions or any history of pruritus. Cutaneous examination revealed poorly defined, hypopigmented nummular macules, located symmetrically on the anterior and posterior aspects of trunk and proximal aspects of limbs with sparing of head and neck (**Figure 3**).

Discussion

Progressive macular hypomelanosis (PMH) is an idiopathic skin disorder characterized by hypopigmented macules predominantly located on the trunk without scales or any history of skin problems, more common in tropical and subtropical regions. PMH was first published by Guillet *et al.*¹ in 1988 in young women aged 13 to 35 years in the West Indies and in a Caribbean immigrant population in France characterized by spreading hypochromic macules on the trunk. Clinically, PMH is characterized by ill-defined, non-scaly, round-to-oval, asymptomatic, and symmetric hypopigmentation. The lesions may become confluent, forming large hypopigmented macules after an increase in their number. The natural history of PMH is a stable disease or perhaps slow progression over decades, with spontaneous disappearance after mid-life.³ The etiology of this condition is uncertain, although Westerhof *et al.*⁴ recently incriminated *Propionibacterium acnes* as the causative agent. In patients with dark skin types, PMH may be distressing because the contrast between normal skin and hypopigmented macules makes the lesions appear more prominent and the patients feel socially awkward.

Histopathological evaluation reveals that there is no decrease in the number of melanocytes but only a decrease in melanin content which suggests that there is probably a functional defect in pigmentation or a problem in melanin distribution. Electron microscopic examination of lesional skin in these patients has revealed that less melanized aggregated melanosomes instead of single, mature melanosomes are transferred from melanocytes to keratinocytes resulting in a decreased epidermal melanin.⁵

Several differentials need to be ruled out once PMH is considered including pityriasis alba,

post-inflammatory hypopigmentation, idiopathic guttate hypomelanosis and hypopigmented mycosis fungoides.

PMH is known to be resistant to treatment. Topical and systemic antifungal agents, topical steroids, topical antimicrobial agents, and PUVA therapy have been used to treat PMH; however, no consistently effective therapy has been established. Treatment modalities include narrow band UVB,⁶ combination of sunlight and oral tetracycline, combination of UVA1 plus topical clindamycin and benzoyl peroxide or combination of topical benzoyl peroxide and oral doxycycline.⁷

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