

Progressive macular hypomelanosis: an epidemiological study of 103 cases in Southeast Brazil

Hipomelanose macular progressiva: estudo epidemiológico com 103 casos da Região Sudeste do Brasil

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Received on: 10 October 2014

Approved on: 2 December 2014

This study was carried out at the Centro de Dermatologia Prof. René Garrido Neves – Fundação Municipal de Saúde de Niterói – Niterói (RJ), Brazil.

Financial support: none

Conflict of interest: none

ABSTRACT

Introduction: Progressive macular hypomelanosis is characterized by the emergence of hypopigmentation, predominantly in the trunk region of young women of mixed racial origin. Although recent studies suggest that *Propionibacterium acnes* may have an important role in the pathogenesis, its etiology remains unclear.

Objective: To evaluate the prevalence of the condition with regards to gender, age, skin phototype, affected areas and disease onset, and the relationship with ordinary acne and oily skin in the city of Niterói, Rio de Janeiro State, in the coastal region of Southeast Brazil.

Methods: Retrospective epidemiological study that uses the analysis of medical records of 103 patients with diagnosis of progressive macular hypomelanosis, from 2001 to 2012.

Results: Eighty-two percent of studied patients were women and 18% men, with a predominance of phototypes III (42.7%) and IV (42.7%). The average age of onset of the disease at diagnosis was 26 years. In 46.0% of patients the disease duration ranged from 1 to 4 years. When two areas were affected (50%), the dorsum was the most frequent (79.0%), followed by the abdomen (20.0%). In 98% of cases, patients had oily skin, and 71.8% had active acne.

Conclusion: Progressive macular hypomelanosis is a chronic condition that primarily affects the skin of the dorsum and abdomen of young women with skin phototypes III and IV, who are also bearers of oily skin and acne. There are still many doubts regarding the true etiology and treatment of progressive macular hypomelanosis. The high percentage of association with skin oiliness and acne vulgaris suggests that these clinical conditions are possibly related.

Keywords: Hypopigmentation; epidemiology; acne vulgaris

RESUMO

Introdução: A hipomelanose macular progressiva se caracteriza pelo surgimento de hipopigmentação predominantemente na região do tronco de mulheres jovens de origem racial mista. Embora estudos recentes indiquem que o *Propionibacterium acnes* possa ter papel relevante na patogênese, sua etiologia permanece incerta.

Objetivo: Avaliar a prevalência da doença quanto a sexo, idade, tipo de pele, áreas afetadas e início da doença, relação com acne vulgar e pele oleosa na cidade de Niterói, RJ, região litorânea no sudeste do Brasil.

MÉTODOS: Estudo epidemiológico, retrospectivo, através da análise de prontuários de 103 pacientes com diagnóstico clínico de hipomelanose macular progressiva entre 2001 e 2012.

Resultados: 82% dos pacientes avaliados eram mulheres, e 18% homens, com predomínio dos fototipos III (42,7%) e IV (42,7%). A idade média de aparecimento da doença no momento do diagnóstico foi 26 anos; em 46% o tempo de doença variou entre um e quatro anos. Quando eram afetadas duas áreas (50%), o dorso foi mais frequentemente afetado (79%), seguido do abdômen (20%). Em 98% dos casos, os pacientes tinham pele oleosa e 71,8% deles apresentavam acne ativa.

Conclusão: A hipomelanose macular progressiva é doença crônica que atinge preferencialmente a pele do dorso e do abdômen de mulheres jovens de fototipos III e IV portadoras de pele oleosa e acne. Existem ainda muitas dúvidas em relação à verdadeira etiologia e ao tratamento da hipomelanose macular progressiva. A alta porcentagem de associação com oleosidade cutânea e acne vulgar indica a possibilidade de essas condições clínicas estarem relacionadas.

Palavras-chave: hipopigmentação; epidemiologia; acne vulgar

INTRODUCTION

Progressive macular hypomelanosis (PMH) is a cutaneous condition characterized by the emergence of hypopigmentation, predominantly in the trunk region of young women of mixed racial origin. It is a common, yet underdiagnosed pigmented disorder. The condition can be found in patients worldwide, however it is more common in those from tropical and subtropical countries.

This entity was first described in 1987 by Borelli² as *cutis trunci variata* and named by Guillet in 1988 as progressive macular hypomelanosis.³ Although this became the preferred terminology, it has been recognized by several authors around the world under a wide variety of denominations, such as: *hypomelanose maculeuse confluent et progressive du metis melanoderme*,⁴ creole dyschromia,⁵ idiopathic macular hypomelanosis of the melanodermic,⁵ and progressive trunk hypomelanosis.⁶

Its etiology remains unknown despite several factors having already been suggested as causes for the lesions: racial origin and, later on, a suggested correlation with tinea versicolor and pityriasis alba.³ The presence of *Propionibacterium acnes* producing a depigmentation factor has also been proposed, as well as disorders in the melanosomes.⁷ Hereditary factors may play an important role, although further investigations are necessary.⁸

PMH is characterized by hypopigmented macules of undefined limits, usually 10–30 mm in diameter, located predominantly in the trunk region of the body. The condition generally affects skin areas rich in sebaceous glands.⁹ There is typically no reference to previous inflammatory occurrences or lesions of an infectious nature.

The diagnosis is mostly clinical and can be confirmed by examination using a Wood's lamp, which shows red fluorescent spots.¹⁰ Observation with confocal microscopy shows that the "pigmented ring" around the dermal papillae in the lesions is intact, however its melanin content is less than the that of the surrounding normal skin.¹⁰ No spores, hyphae or bacteria are observed in the stratum corneum.⁸ Histological investigations show a slight reduction of melanin granules in the basal cell layer with a variable reduction of melanin transfer to keratinocytes. Nonetheless, the histological findings are not sufficient to characterize the definitive diagnosis of this condition.¹¹

It is necessary to exclude the possibility of pityriasis versicolor, pityriasis alba, leprosy and hypochromic fungoid mycosis as differential diagnoses.¹¹

Several treatments are mentioned in the literature: topical and oral antibiotics, benzoyl peroxide, and phototherapy (PUVA and UVB–NB) – all of which are unstable and still produce results that are not fully satisfactory.^{12–15}

The present study is aimed at evaluating the prevalence of the dermatosis in regards to gender, age, skin phototype, affected areas and duration of the disease, and also considering its correlation with acne vulgaris and oily skin conditions.

METHODS

A retrospective study was performed through the analysis of medical records of patients with clinical diagnosis of PMH

treated at private dermatology practices in the city of Niterói (Rio de Janeiro State), in the coastal region in southeastern Brazil, between 2001 and 2012. The study included 103 patients with complete personal and epidemiological data, and whose diagnosis was conclusive for this disease.

RESULTS

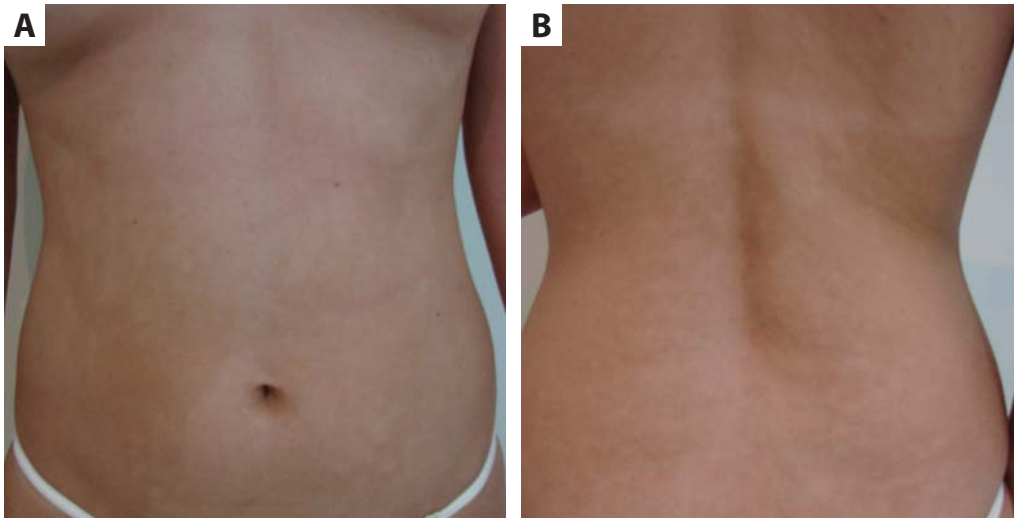
From Table 1 it is possible to note that roughly 82% of the patients were female (Figures 1 and 2), and 18% male (Figures 3 and 4). There was a predominance of phototypes III (42.7%) and IV (42.7%). The patients' mean age at diagnosis was 26 years (min = 13 years, max = 50 years, mode = 22).

According to Table 2, patients had oily skin in 98% of cases, and 71% had active acne, indicating a possible association of the disease with oily skin and the presence of acne.

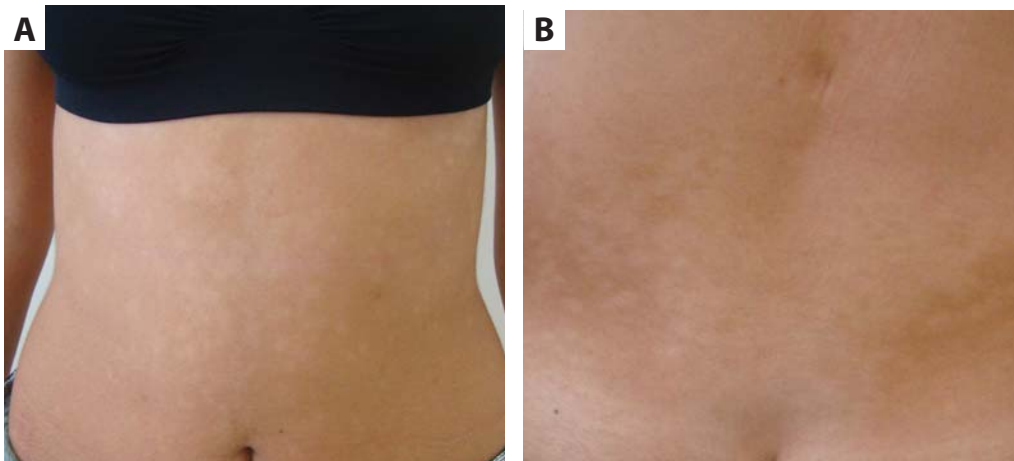
The results in Table 3 indicate that for most evaluated patients, the disease arose between the ages of 15 and 29, with 25% of cases between 15 and 19, 25% between 20 and 24, and 20% between 25 and 29 years. In addition, only 16% had developed the disease less than one year before, and in 46% the time of onset was one to four years before, with 18% with between five to nine years. It is worth noting that some patients had never noticed the lesion or did not exactly know when it had developed, with 4% of patients having more than 15 years of development without remission. One patient reported the development of the picture for about 20 years. Regarding the location, 51 patients had lesions only in one area, and 52 in two or more areas. When the patient had a single lesion, the dorsum was the most often affected area (79%), followed by the abdomen (10 cases, 20%) and chest (one case, 2%). Analyzing the association of affected areas, 47 patients (90%) had concomitant lesions on the dorsum and abdomen, and three patients (6%) had concomitant lesions in the dorsum, abdomen and chest.

TABLE 1: Perfil epidemiológico de pacientes com HMP – fototipo, sexo, idade (n = 103)

	Frequency	Percentage
PHOTOTYPE		
I	-	-
II	7	6,8
III	44	42,7
IV	44	42,7
V	8	7,8
GENDER		
F	84	81,6
M	19	18,4
AGE		
Mean	26,4	
Median	26	
Mode	22	
Standard deviation	7,7	
Minimum	13	
Maximum	50	



FIGURES 1 (A e B): Female patient, 35-years-old, phototype III, with lesions in the abdomen and dorsum after 10 years of development; oily skin, but without active acne



FIGURES 2 (A e B): Female patient, 21-years-old, skin phototype III, with lesions in the abdomen and dorsum after two years of development; oily skin and active acne on the face

DISCUSSION

Despite the fact that PMH is a considerably common disorder, its true prevalence is unknown.¹³ It mainly affects young individuals, predominantly between the ages of 18 and 25 years, with rare reports of cases in patients over the age of 50.¹⁶ The majority of cases occur in women, in proportions ranging from 3:1 to 7:1,^{9,13} though some authors have not found differences in the distribution between the genders.^{2,17} The present study evaluated 103 patients with clinical diagnosis of PMH. Of these, 82% were female, with 70.8% reporting the lesions' onset at between 15 and 29 years of age, thus corroborating the literature findings. It is important to note that in the present study it was possible to observe cases in men, in all skin phototypes and with onset of lesions between 9 and 49 years old.

Regarding the skin phototype, the literature has divergent data: while some authors found an increased incidence in

patients of mixed ethnic origin (Negroids/Caucasoids),^{12, 13, 18} other studies deny this claim,^{1, 2, 12} explaining that the disease is probably more easily seen in patients with pigmented skin. Among the present study's patients, 85.4% had Fitzpatrick phototypes III and IV.

Although some authors incriminate the *Propionibacterium acnes* as the etiologic agent of this condition, the presence of acne vulgaris associated with HMP was an uncommon finding in some studies. It is also relevant to note that with conventional identification methods, there are still practical difficulties in distinguishing the species or subspecies of some specific genres that can play an important role in the pathogenesis of diseases. In this manner, the *Propionibacterium acnes* species causing acne and progressive macular hypomelanosis could be of different subtypes.⁹ In a study by Rodriguez in Spain, of 10 patients with a clinical picture of PMH,



Figure 3: Male patient, 20-years-old, skin phototype III, with lesions in the abdomen and dorsum, after three years of development; oily skin and active acne

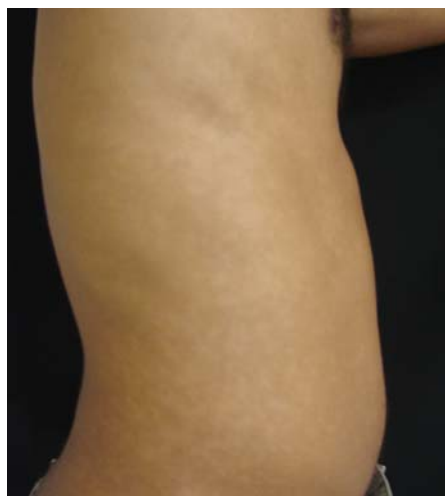


FIGURE 4: Male patient, 35-years-old, skin phototype IV, with lesions in the abdomen and side of the chest, after eight years of development; oily skin without acne

TABLE 2: Epidemiological profile of patients with PMH - oily skin and acne (n = 103)

	Frequency	Percentage
OILINESS		
Yes	101	98,1
No	2	1,9
ACNE		
Yes	73	71
No	29	28
NL	1	1

TABLE 3: Epidemiological profile of patients with PMH - age of lesion onset, disease development, location (n = 103).

	Frequency	Percentage
AGE OF LESION ONSET		
0-11 months	-	-
1-4 years	-	-
5-9 years	1	1
10-14 years	6	5,8
15-19 years	26	25,2
20-24 years	26	25,2
25-29 years	21	20,4
30-34 years	5	4,9
35-39 years	4	3,9
40-44 years	1	1
45-49 years	2	1,9
Did not know	11	10,7
PROGRESSION TIME		
0-11 months	17	16,3
1-4 years	48	46,2
5-9 years	19	18,3
10-14 years	4	3,8
15-19 years	3	2,9
20-24 years	1	1
25-29 years	-	-
30-34 years	-	-
35-39 years	-	-
40-44 years	-	-
45-49 years	-	-
Did not know	11	10,6
LOCATION OF LESIONS		
Single area (n = 51)		
Abdomen	10	19,6
Dorsum	39	76,5
Dorsum (sacrum)	1	2
Chest	1	2
Two or more areas (n = 52)		
Abdomen/dorsum	47	90,4
Dorsum/Thorax Chest	1	1,9
Abdomen/Dorsum/Thorax Chest	3	5,8
Abdomen/Dorsum/Thorax Chest (axillary)	1	1,9

seven reported association with acne, however the author himself questions the possibility of different subtypes of this bacteria.¹⁹ In the present study, 71% of patients had acne vulgaris as an associated complaint. In addition, oily skin was observed in a significant percentage (98%), in line with studies from other authors.⁹

Twenty patients had lesions limited to the dorsum; eight had lesions only in the abdomen; 20 had lesions in both the dorsum and abdomen; two had concomitant lesions in the chest in addition to the dorsum and abdomen; and only one had lesions exclusively in the chest. These findings are consistent with the literature, showing the lesions' predominant distribution to be in the dorsal and abdominal regions.

The progression of the lesions is uncertain. Spontaneous regression usually occurs within 3 to 5 years, though some cases persist for more than ten years.⁴ Menke et al. do not believe in spontaneous remission. On the contrary, they observed stability of the lesions in a 10-year follow up of patients, with no repigmentation being observed in any of them.⁶ Lesuer et al. showed that lesions may persist for in excess of 25 years.²⁰ In the patients evaluated in the present study, the lesions' time progression at diagnosis ranged from two months to 240 months, with 11 of the 103 patients being unable to specify the exact onset time of the disease due to the fact they had not noticed its presence, and because the lesions were only identified at the dermatological examination.

CONCLUSION

PMH is a chronic disease that primarily affects the skin of the dorsum and abdomen of young women with phototypes III and IV, oily skin, and acne.

Although several studies have been previously carried out, there are still many doubts regarding PMH, and its true etiology and effective treatment remain under question. The

present review reinforces the findings described in the literature, with the high number of patients with skin oiliness and acne vulgaris inducing the authors' belief that the condition's etiology is related to these clinical conditions. Further studies are necessary to confirm the actual involvement of *P. acnes* or subtypes of this bacterium in the etiology of PMH and to further elucidate this disease. ●

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