IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 16, Issue 2 Ver. V (February. 2017), PP 14-19
www.iosrjournals.org

Retrospective Analysis of the Clinical Presentation of Progressive Macular Hypomelanosis and Outcome of Its Therapeutic Intervention with Narrow- Band Ultraviolet B Phototherapy

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Abstract: *Progressive Macular Hypomelanosis is often an underdiagnosed hypo pigmented, nonscaly, macular eruption involving the trunk, abdomen, face,extremities and the buttocks with great cosmetic concern to the sufferer. Topical antifungal, corticosteroid has been ineffective in its treatment but phototherapy and topical antimicrobial has proven effective over time.*

Objective: *This study seeks to determine the clinical presentation of Progressive Macular Hypomelanosis in our environment and its response to Narrowband-Ultraviolet B (NB-UVB)*

Methods: *Twenty-three patients who were diagnosed as having Progressive Macular Hypomelanosis and had NB-UVB exposure were retrieved from dermatology unit records. Their pattern of presentation and their treatment outcome analyzed*

Result: *The mean age (SD) of the study population was 28.0 (±13.6) years ranging from 7-55 years. Male to female ratio was 3.6**:** 1. The percentage body surface area covered ranged from 3-60% and areas most affected included the face, trunk, extremities, buttocks and scalp respectively. Significant repigmentation was noticed at 12th sessions of NB-UVB exposure and with almost maximum restoration at 18 sessions of exposure. However, there was a relapse in 4 patients at 4- 12 months of stoppage of NB-UVB.*

Conclusion: *Progressive Macular Hypomelanosis seemed to be more generalized in much younger age group when compare with older adults and NB-UVB exposure was helpful in their treatment.*

Keywords: *Progressive Macular Hypomelanosis, clinical presentation, NB-UVB`*

I. Introduction

Progressive macular hypomelanosis (PMH) was originally described in patients with FitzPatrick's skin type IV-VI from tropical and subtropical countries of the world (1,2). However, it has been found to have a world-wide distribution (2). The precise aetiology and pathogenesis of PMH is yet to be fully understood. Although, Westerhof et al had observed that hair follicles within the lesions fluoresce coral red under wood lamp, but not inter-follicular or non-lesional skin (3). This suggests the presence of a porphyrin producing organism in the follicle, of which they were able to isolate and identify as gram positive nonspore- forming anaerobic rod propronibacterium acnes (3,4). PMH is a common, often misdiagnosed dermatosis with a great cosmetic concern to the sufferer. Presentation is asymptomatic, ill-defined to discoid, non-scaly, symmetrical hypopigmented macules that sometimes coalesce into patches (4,5). The eruptions are often seen on the trunk, face and a satellite extension to the extremities, neck and buttocks. (4,5,6). The clinical course of the lesions varies and some authors have asserted that the lesions tend to stabilize and undergo spontaneous regression over a period of 2-5years (2). While others stated that the normal course is one of a slow progression of the lesion overtime (6). Some researchers have also reported more female preponderance in the ratio 3:1 when compared with male gender, but other studies did not support this observation (3, 5- 9). PMH can easily be distinguished from other hypopigmented disorder such as pityriasis alba, pityriasis versicolor, mycosis fungoides, vitiligo, leprosy and postinflammatory hypopigmentation both clinically and histologically (1,5). For example, it is not associated with initial erythema and scaling of pityriasis alba. In PMH, there is normal sensation, no prior history of inflammation, trauma, itching, pain or infection. Potassium hydroxide mounts of skin scrapings for Malassezia fungi is negative. Histologically, PMH is characterized by diminished pigment in the epidermis and a normal looking dermis (5,6). Presently, there is no agreeable first line treatment for Progressive Macular Hypomelanosis after trials of many treatment options (3).

Phototherapy alone has been attempted for the treatment of PMH with variable outcomes. Psoralen ultraviolet A (PUVA) therapy was noticed to induce repigmentation after six (6) weeks of therapy but hypopigmented spots recurred in the same location within few weeks of stopping PUVA (10). Also, an uncontrolled prospective study of narrowband ultraviolet B (NB-UVB) therapy was performed on 17 patients and repigmentation occurred by a gradual darkening of the lesions after some sessions of exposure. This was in contrast to repigmentation observed in vitiligo which occurred from discrete perifollicular islands (7,11). The

speculative mode of action for the efficacy of NB-UVB phototherapy includes stimulation of melanogenesis and antimicrobial effects (7, 12). This study aims to define the clinical course of progressive macular hypomelanosis and its possible therapeutic outcome with NB-UVB (311nm) in our environment.

Objectives

The aim of this paper is to determine the clinical presentation of progressive macular hypomelanosis in our environment and its response to narrowband-ultraviolet B (NB-UVB)

II. Materials And Methods

This was a retrospective study involving patients who were seen in our dermatology outpatients clinic from December 2013 to November 2016 in University of Abuja Teaching Hospital. Out of one thousand four hundred and four (1404) new patients seen, twenty three (23) of them were diagnosed as having progressive macular hypomelanosis, clinically and histologically where necessary. A questionnaire was designed to capture the following: bio data, body mass index (BMI), other family members with similar disease, clinical distribution of the lesion, duration of lesion, medication on or before presentation, minimal erythema dose (MED) at the commencement of narrow-band phototherapy (NB-UVB), number of sessions before noticeable repigmentation, percentage of repigmentation, relapse of repigmentation after stoppage of NB-UVB phototherapy, any adjunctive medication while on phototherapy and any noticeable side effect(s).

Patients who had progressive macular hypomelanosis were exposed to narrow-band ultraviolet B (311nm) after explanation and consent duly signed for exposure. The NV-UVB (311nm) used was a Philip TL 01 (1000 series) ultraviolet therapy lamp unit with 8 bulbs by Solarc system incorporated (13). TL/W/01 is a panel with eight (8) fluorescent bulbs protected with a wire guard. Each of those bulbs delivers irradiant energy at the rate of 6.5mw/cm^2

Irradiance = $\frac{\text{Expose Dose (1joule/cm}^2\text{)}}{\text{Exposure time (seconds)}}$

Acceptable distance or positioning at the commencement of treatment was 8inches-12inches (20-30cm) and energy delivered can vary due to time exposure (13, 14). A minimal erythema dose (MED) was obtained for each patient as minimal energy delivered to cause mild redness on the skin of the patient after 24hours. However, because of the difficulty in obtaining MED in black skin an alternative which depends on skin type, previously described was used (14, 15, 16, 17)

Skin type	Dose (mj/cm^2)	Time (seonds)
I	300	45
II	400	60
III	600	90
IV	800	120
V	1000	150
VI	1500	225

Almost all the patients were of the skin type V or VI, thus, their alternative MED were set at 1000 joules/cm^2 and above.Time of exposures or energy delivered was increasingly done at the rate of 20% to 25% of the previous energy exposed or with recourse to any observable side effect. Each patient had a chart where progress of their treatment, total energy delivered (TED) per session per week and side effect were duly documented. Other precautionary measures taken include wearing of opaque goggles during treatment and boxers for male patients during whole-body exposure. Progress of treatment was assessed based on skin colour restoration. Those who had no colour change scored 0-25%, minimal skin colour restoration were scored 26%-49%, moderate skin colour restoration were scored 50%-70% and maximum skin colouration were scored 76%-99% and complete repigmentation score was 100%.The information retrieved was into SPSS 20 and analysed accordingly. Frequency, means, standard deviation and correlations were some of the statistical tools used.

III. Result

A total number of 23 patients with PMB were recruited into the study. Mean age (standard deviation) was 28.0 (13.6) years and ranged -7-55 years. Male to female ratio was 3.6:1. Body surface area covered (in %) ranged 3-60% with a mean of 18.6%. Seven 7 (30.4%) declined treatment while 16 (69.9%) were treated with NB-UVB. Out of the 16, repigmentation was noticed in 13 (81.3%) cases. However, 4 cases of relapse were observed after 4-12 months of follow up. Up to 25%-90% repigmentation was noticed between 9 to $\geq$18 sessions. Mean (SD) MED was 1762.5 (552.4)joules/cm^2, ranged 1000-2400joules/cm^2. Number of sessionswas 6-49times with mean of 14.3 sessions.The duration of disease before presentation was 1.5-12years with a mean duration of 5.2years.

Body surface area mostly affected was the face. While, 20 patients had hypopigmentation both on the face and other parts of the body, three had hypopigmentation on the face alone. Other parts of the body affected alongside the face include, the trunk, upper and lower limb, scalp, and buttocks. The part of the face mostly affected was the malar area and the nose (Table 1).

Table 1: Demographic characteristics and body surface area covered

Variables	N=23 Frequency	100% Percent (%)
Age group (years)		
□15years	6	26.1
>15years	17	73.9
Sex		
Male	18	78.3
Female	5	21.7
Education Status		
Primary	6	26.1
Secondary	5	21.7
Tertiary	12	52.2
Treatment		
No treatment	7	30.4
Treatment	16	69.6
Body surface area covered		
Face, Trunk and Upper Limb	11	47.8
Face and Trunk	4	17.4
Face	3	13.0
Face, Trunk and Scalp	2	8.7
Face and Thigh	1	4.3
Face, Trunk, Upper limb and buttocks	1	4.3
Buttocks and extremities	1	4.3

N =Total number of patients with PMH
Face (Malar =4, nose=3, forehead =1, chin=1)

There were 18 (78.3%)males and 5 (21.7%) females. Six 6 (26.1%) were aged ≤15years and 17 (73.9%) were older than 15years. Those who had primary education were 6 (26.1%), 5 (21.7%) and 12 (52.2%) had secondary and tertiary education respectively. The number of patients treated were 16 (69.6%) while 7 (30.4%) declined treatment. Eleven 11 (47.8%) of the patients had hypopigmentation on the face, trunk and upper limb, 4 (17.4%) of them had hypopigmentation on the face and trunk only while 3 (13.0%) of the patients had hypomelanosis on the face only(Table 1).

Table 2: Relationship between Body surface area involvement of the lesions and age

Age (years)	Mean BSA ± SD	t	P
□15years	35.3 ± 23.5	3.208	0.004**
>15 years	13.8 ±9.4		
Parameter	**Mean ± SD**	**Rho**	**P**
Age (years)	28.0 ± 13.6	1	0.031**
BSA (%)	19.4 ± 16.8	-0.451	

****Statistically significant (p<0.05)**

Mean BSA (%) involvement of the lesions in patients with ages ≤15years was 35.3%. This was significantly higher than was observed in patients older than15years (mean BSA of13.8) at p=0.004. There was a negative moderate correlation between age and percentage BSA. This indicates that as the ages increased, the body surface area involvedwith PMH is reduced and this relationship was statistically significant (p=0.031).

Table 3: Relationship between number of sessions and other variables

	Rho	P	Decision
Number of session	1	-	-
Age	-0.017	0.939	Not significant
Body mass index	-0.034	0.877	Not significant
Duration of disease	-0.282	0.192	Not significant
Minimal Erytherma Dose	0.650	0.001	Highly significant
Total TED	0.866	<0.0001	Highly significant
Body surface area covered (%)	-0.001	0.996	Not significant
No of weeks on treatment	0.819	<0.0001	Highly significant
Repigmentation	0.617	0.001	Highly significant

TED- Total Energy delivered

There was strong a correlation between number of session and minimal erythermal dose, total energy delivered, number of weeks on treatment and repigmentation (Table 3). These relationships were highly significant($p<0.05$).The summary of Patients' treatment outcome is documented on table 4. Repigmentation was noticed between 9^{th} sessions and 18^{th} sessions of narrowband UVB phototherapy.

Table 4: Outcome of Treatment Response of progressive macular hypomelanosis and NB-UVB phototherapy

Total No. of sessions	**TED In J/cm²**	**% Repigmentation**	**Relapse**	**Age (years)**
18	53200	60-70%Repigmentation noticed on the face and trunk at 12 sessions. At the 18 sessions, up to 70-90% repigmentation noticed on the face and trunk and 40-60% noticed on the buttocks	No relapse after 10months of follow up	≤15years
18	55800	Repigmentation noticed on the face at the 9 sessions and at the 18 sessions, up to 70-80% repigmentation was noticed on the face, trunk and extremiites	10% relapse noticed after 6 months of follow up	≤15years
12	24700	Up to 40-50% repigmentation noticed on the face at the 9 sessions	No relapse after 6 months of follow up	≤15years
9	27200	No observable repigmentation noticed	Loss to follow up	≤15years
15	49600	80% repigmentation noticed on the 12 sessions and 90% repigmentation noticed at the buttocks and upper limb	No relapse after 7 months of follow up	> 15years
6	10100	No observable repigmentation noticed	Loss to follow up	≤15years
9	24480	40-60% repigmentation noticed on the face at the 9 sessions	No relapse after 10months of follow up	> 15years
15	47200	40-60% repigmentation noticed on the face and 20-40% repigmentation noticed on the trunk at the 12 sessions	Noticed relapse after 4 months of follow up	> 15years
15	47200	60% and 80% repigmentation noticed on the face at the 12 sessions and 15 session respectively	Noticed relapse after 8 months of follow up	> 15years
9	24000	25% repigmentation noticed on the face at the 9 sessions	No relapse after 8 months of follow up	> 15years

12	35200	40% repigmentation noticed on the 12 sessions	No relapse after 7 months of follow up	> 15years
16	52600	40% and 50% repigmentation noticed on the face and posterior trunk at the 12 sessions and 15 session respectively	Nil relapse of repigmented skin	> 15years
49	67800	40% and 50% repigmentation noticed on the face at the 15 sessions and 18 session respectively	Relapsed repigmentation on the face	> 15years
6	12800	No observable repigmentation noticed	Loss to follow up	> 15years
9	23400	Up to 25% repigmentation noticed on the face at 9 sessions	No relapse after 6 months of follow up	> 15years
10	13400	Up to 35% repigmentation noticed on the face and trunk at 9 sessions	No relapse after 7 months of follow up	≤15years

Age- Patients'age as at the start of the study; TED-Total Energy Delivered

Figure I, II, III, IV respectively shows different clinical presentations of PMH

IV. Discussion

Progressive macular hypomelanosis is a rarely diagnosed disorder which poses challenges in relevant differential diagnosis of hypopigmentations.(1) Guillet etal had described the disorder as a primary, acquired, non-scaly macular hypopigmentation affecting mainly women from 18 years to 25 years of age (1). Young women of West Indies extract and Caribbean immigrant population in France were seen having a widespread hypochromic macules on the trunk and less frequently on the abdomen (1, 2).

In this study, there was predominance of male patients (78.3%) when compared with females. This is in agreement with Martinique et al who revealed that the disease is more widespread in men than women with mean age of 25 years (18). Other studies have shown that progressive macular hypomelanosis can occur on any skin type but more frequently on Fitpatrick skin type IV- VI (2,3,5) and this was consistent with our findings. More than 90% of our patients had presentation of lesion on the face (mostly on the malar area, the nose, the forehead and the chin respectively). Other researchers had reported more of the lesions occurring on the trunk and the abdomen with rare involvement of the face and extremities (1, 2, 3, 6). We found more percentage body surface area involvement in patients below 20 years when compared with older adults 30 years and above. About 12 (52.2%) of the patients who were 30 years and above have had the lesions present for more than 5 years without any sign of clinical improvement. This was in contrast with earlier observation by previous researchers (4, 12, 19).

There have been variable outcomes of repigmentations in patients that have PMH who were exposed to narrowband ultraviolet B (NB-UVB). Duarte et al and Kim with his co researchers had reported more than 50% clinical improvement at the 16th session of NB-UVB (7,20). In the same study 39% had their lesion completely cured, while 26% had clinical improvement of their conditions up to 80-90% (20). Our findings showed upto 50% of our patients on NB-UVB had about 60% of their skin colour restored by12 sessions of exposure while

those who had NB-UVB exposure for onand above 18 sessions achieved more than 75% maximum skin colour restoration. Although, 4 patients had a relapse of their repigmentation at 4-12 months after stoppage of NB-UVB exposure, response to NB-UVB were noticed to improve speedily from face, trunk, upper limbs, buttocks and lower limbs in that order. Notably, lesions on the scalp were noticed to have not responded to NB-UVB phototherapy.

V. Conclusion

In conclusion, this study has helped to highlight the fact that PMH is an emerging cosmetic embarrassing disorder in our environment. It is more generalized when it appears in much younger age group as compared with older adult. NV-UVB is helpful when a reasonable number of sessions of exposure are done and maintenance dose needful to avoid relapse of repigmentation.

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