Cutaneous Dermatosis in Persons Living with Albinism in Kaduna — Northwestern Nigeria

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Abstract

Background: Albinism is a congenital autosomal recessive disorder characterized by the complete or partial absence of pigment in the skin, hair, and eyes. Nigeria is ranked amongst the highest in the world with an estimated figure of over two million. Albinism is associated with several skin anomalies such as freckles, thickening, and wrinkling of the skin, yellowish-brown hair, and a high incidence of skin cancers among others. In Nigeria, there are few studies on skin disorders associated with albinism in the north. This study is aimed at identifying skin anomalies among albinos and also to identify factors related to these skin changes.

Materials and Methods: This was a cross-sectional study conducted during the 2022 World Albinism Day at Kaduna. All Fifty-one Albinos who were in attendance were recruited. Records of the patient's age, gender, use of sunscreen, and family history of Albinism were noted. An evaluation of the skin lesions was made clinically. Data was analyzed using SPSS 21.

Results: The mean age of the respondents was 22.4 years ±16.5. The respondents were mostly males. Twenty-nine respondents had more than one skin lesion these include –freckles, solar elastosis, Squamous cell carcinoma of the ear lobe, and a case of congenital ectropion. Thirty-six individuals (71%) had a family history of albinism and ten (20%) used sunscreen consistently and correctly..

Conclusion: Dermatoses are common among individuals living with albinism. Since dermatoses are related to exposure to sunlight, counseling on the use of sunscreens and other measures to reduce sun exposure is essential in people living with albinism

Keywords: Albinism, Sun protection, Kaduna, Squamous cell carcinoma.

Dermatoses chez les Personnes Vivant avec l'albinisme à Kaduna - Nord-Ouest du Nigeria

Résumé

Contexte: L'albinisme est une maladie congénitale autosomique récessive caractérisée par l'absence totale ou partielle de mélanine dans la peau, les cheveux et les yeux. Le Nigeria est classé parmi les premiers au monde avec un chiffre estimé à plus de deux millions d'albinos. L'albinisme est associé à plusieurs anomalies cutanées telles que des taches de rousseur, un épaississement et des rides de la peau, des cheveux bruns jaunâtres et une incidence élevée de cancers de la peau, entre autres. Au Nigeria, il existe peu d'études sur les troubles cutanés associés à l'albinisme dans le nord. Cette étude vise à identifier les anomalies cutanées chez les albinos et ainsi que les facteurs liés à ces modifications.

Matériels et méthodes: Il s'agissait d'une étude transversale menée lors de la Journée mondiale de l'albinisme 2022 à Kaduna. Cinquante et un albinos ont été recrutés. L'âge, le sexe, l'utilisation de crème solaire ainsi que les antécédents familiaux d'albinisme des patients ont été collectés. Un examen clinique des lésions cutanées a été réalisée. Les données ont été analysées à l'aide de SPSS 21.

Résultats: L'âge moyen des répondants était de 22,4 ans ±16,5. Les répondants étaient majoritairement des hommes. Les trois cinquièmes des personnes interrogées présentaient plus d'une lésion cutanée, notamment des taches de rousseur, une élastose solaire, un carcinome épidermoïde du lobe de l'oreille et un cas d'ectropion congénital. Environ les trois cinquièmes avaient des antécédents familiaux d'albinisme et seulement un cinquième utilisait un écran solaire de manière cohérente et correcte.

Conclusion: Les dermatoses sont courantes chez les personnes vivant avec l'albinisme. Étant donné que il s'agit de dermatoses liées à l'exposition solaire, des conseils sur l'utilisation d'écrans solaires et d'autres mesures visant à réduire l'exposition au soleil sont essentiels chez les personnes atteintes d'albinisme.

Mots clés: Modifications cutanées, Albinos, Protection solaire, Kaduna, Carcinome épidermoïde.

Introduction

Oculocutaneous albinism (OCA) encompasses a heterogeneous group of genetic conditions with an autosomal recessive inheritance. It is characterized by hypopigmentation of the skin, hair, and eyes due to a reduced or lack of cutaneous melanin pigment production.¹

It has a global prevalence of 1:20,000² with wide variation from population to population. There are higher figures than the global average ranging from 1 in 15,000 in Eastern Nigeria³ to 1 in 1,000 in the Tonga tribe of Zimbabwe.⁴ It shares an equal occurrence in both genders. There are two major types of OCA; the syndromic and the non-syndromic OCA with the former being more common and of seven different types.

OCA1 and OCA2 are variants mostly seen with the latter accounting for one-third of cases worldwide and more common in Africa. ⁵ This study aimed to document the skin findings in people with albinism living in Kaduna, Northwestern Nigeria.

Methods

This was a cross-sectional study that was conducted during the 2019 World Albinism Day in Kaduna –Northwestern Nigeria. There are 23 local government areas in Kaduna state, with Kaduna North being one of the largest. During World albinism day, most individuals living with albinism usually attend the outreach at Kaduna North where lectures are delivered by different professionals a mongst which are dermatologists, ophthalmologists, and meteorologists.

Ethical approval and informed consent were obtained from the ethical committee of the hospital and Ministry of Humanitarian Services and respondents respectively before the commencement of the study.

Fifty-one persons with albinism who were in attendance and consented were recruited for the study. All those who did not give their consent were excluded from the study. An interviewer-administered questionnaire. Records of the patients' age, gender, family history of Albinism, and use of sunscreen were noted. An evaluation of the skin lesions was made clinically after examination with a

dermoscope. Those with suspicious lesions were referred to our center for further evaluation.

Data was analyzed using a statistical package for Social Sciences version 21. Data cleaning was carried out and errors were corrected. Quantitative variables were summarized using means and standard deviation. The association between age and skin lesions was tested using a chi-square test. The level of statistical significance was $P \le 0.05$.

Ethical approval and informed consent were obtained from the ethical committee of the Hospital, the Ministry of Humanitarian Services, and the respondents respectively before the commencement of the study. (NHREC/30/11/21A)

Results

A total of 51 respondents with Albinism were recruited for the study. The majority of the respondents were males 30 (58.82%). The youngest respondent was 1.5 years and the oldest was 60 years with a mean age of 22.4 ± 16.56 . (Table 1). About three-fifths (71%) of the respondents had a family history of ocular or oculo-cutaneous albinism. Three-fifths (59%) of the respondents had skin lesions while 22 (41%) had no clinically evident skin lesion except albinism.

The most frequent skin changes seen were freckles, which were observed in 28 (54.9%) of the respondents, other abnormalities seen in association with freckles are solar elastosis which was observed in 10 (19.6%) of the respondents, and actinic keratosis 5 (9.8%). There was one case each of squamous cell carcinoma (following histology), Folliculitis keloidalis nuchae, and congenital ectropion (Table 1). Only 10 (19.6%) of the respondents used sunscreen creams regularly. There was a statistically significant relationship between increasing age and skin lesions (P<0.000). (Table 2)

Discussion

Albinism is an autosomal recessive disorder due to partial or total absence of melanin pigment. It is characterized by the deficiency of the enzyme tyrosinase which is involved in the conversion of tyrosine to dopa and then to melanin. ^{6,7,8,9} This melanin produced protects the skin from harmful



ultraviolet rays which can lead to skin cancers. In this study, more than half of the respondents have one form of skin disorder and only one-fifth of the respondents used of sunscreen correctly and consistently.

Males constituted three-fifths of the population with a male-to-female ratio of 1.4.:1 This is comparable to the study in south-eastern Nigeria by Okoro E where he got a male: female ratio of 1.3:1³ but one Lagos study had a female preponderance of (M/F) 1:1.25.¹⁰

The mean age of the respondents was 22.4 years ±16.5, the youngest being 1.5 years and the oldest being 60 years. This is similar to the mean ages seen in a Lagos and Southern Nigeria study. ^{10,11} In another study by Okoro in Enugu carried out 50 years ago, he found that very few persons living with Albinism were above 30 years. ³This age disparity may be due to modernization and increased awareness over the years.

More than 71% of the respondents had a family history of albinism. This is slightly higher than what was found in Enugu, where 63.3% had at least one family member with albinism.³ The higher figure observed in our study may be due to the small sample size of 51 persons compared to 1000 in the Enugu study. Albinism being an autosomal recessive trait has a 1 in 4 chance of being inherited.

Only 20% of the respondents used sunscreen correctly and consistently and these individuals were noticed to have less photodamage. Similar, values were obtained by Madubiko in southern Nigeria where only 20.5% used sunscreens and had few lentigines. This was statistically significant.¹¹

More than half (58%) of the respondents had one form of skin lesion or another. This is lower than that seen in the Lagos (95.8%) and Benin study (78%). ^{10,11} The low value seen in this study may not be unrelated to the majority of the respondents in our study being young less than 40 years. Most of these changes have been noticed to increase with age and sun exposure. This is in contrast to what was observed in a study by Samuel et al in Tanzania where none of the respondents had skin lesions. The respondents in the Tanzanian study used sunscreen regularly due to an efficient surveillance program for albinos by the

government. The use of sunscreens has been found to reduce the appearance of photo-aging spots. ¹² The most common skin lesions observed were those related to sun damage- freckles, solar elastosis, and actinic keratosis. Solar lentigines (freckles) were found in about half (54%) of the respondents in this study. This is lower than the 63% seen in the Benin study- southern Nigeria. This disparity may be due to the smaller sample size used in our study in the Northwest.

Other skin changes recorded in this study were solar elastosis (20%), actinic keratosis (10%), and squamous cell carcinoma (1%). These figures are lower than those found in the Benin study with values of 45%, 29%, and 12.3% respectively. The wide differences seen in these studies may be due to the selection processes involved in the north where all the respondents present during the outreach were used for the study, the younger age group formed the bulk of the study accounting for 55% of the total population and also the dress code in the north, which involved covering most of the body due to religious reasons. The cutaneous malignancy (squamous cell carcinoma) noted in this study was on the ear lobe, previous studies have recorded similar findings. 13,14,15

There was a case of congenital ectropion seen in a 57-year-old woman. There was no associated chemosis or corneal scarring. Ectropion is said to be due to the eversion of the eyelids. No report of this finding in persons living with Albinism has been recorded previously. Although very rare, but more common in Africans. It may be associated with Down's syndrome, ichthyosis, and blepharophimosis syndrome. Our patient did not have any of these as such it could be said to be a case of an isolated finding. IT-20

Only one person had folliculitis (acne) keloidalis nuchae (FKN). These are itchy round small bumps at the back of the neck which later form scars and then keloidal masses. In a study by Ogunbiyi et al in southwestern Nigeria, a prevalence of 1.5% was found in men aged 35-44 years. These were individuals with normal skin pigmentation A similar case was reported in Mali-Bamako, where a 27-year-old albino presented with folliculitis

keloidalis nuchae.²³

There was a statistically significant relationship between increasing age and the presence of skin lesions, (p<0.000) with the younger age group (0-20) having no skin changes. The main limitation is the small sample size used in this study.

In conclusion, the importance of skin monitoring and protection cannot be over-emphasized in these groups of individuals since most of the skin findings seen are related to sun exposure. Similarly, patients with albinism are at a high risk of developing skin cancers as such primary preventive measures like enlightenment, free drugs, and accessible screening facilities are highly recommended.

Declaration of patient consent

Appropriate patients' consent was obtained in writing and consent was also given for their images and medical to be used in the journal The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil

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Character	Frequency (%)	
Gender	requency (70)	
Male	30 (58.82)	
Female	21 (41.18)	
Age (years)		
0-20	28 (55)	
21-40	12 (23)	
41-60	11 (22)	
Family history of Albinism		
Yes	36 (71)	
No	15 (29)	
Use of sunscreen		
Yes	10 (20)	
No	41 (80)	
Lesions		
Freckles	11 (21.6)	
Freckles/solar elastosis	10 (19.6)	
Freckles/Actinic keratosis	05 (9.8)	
Freckles/Ectropion	01 (1.96)	
Freckles/Squamous cell Ca	01 (1.96)	
Acne keloidalis nuchae	01 (1.96)	
No skin lesion	22 (43.1)	

Variable	Grade		
Age (years)	Present	Absent	P
0-20	5	23	0.000
21-40	12	0	
41-60	11	0	



Figure 1: Solar lentigines (Freckles) on the face



Figure 2: Solar Elastosis- characterized by furrowing and pebbly appearance on the neck of this Albino





Figure 3: Ectropion in a respondent with Albinism



Figure 4: Folliculitis keloidalis nuchae (Infected) on the back of the neck