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CUTANEOUS MALIGNANCY IN THE NIGERIAN ALBINO: A STUDY IN A TERTIARY URBAN HOSPITAL IN SOUTHERN NIGERIA

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ABSTRACT

Background: Oculocutaneous albinism is an autosomal recessive disorder of pigmentation involving the skin, eyes, and hair. It is characterized by reduced or complete lack of melanin production. Melanin is protective against the harmful effects of the tropical sun. A lack of it predisposes the melanin deficient skin to cutaneous cancers. **Aim:** It determined the prevalence and pattern of cutaneous cancers in people with albinism living in Benin city. It also determined the relationship if any between cutaneous malignancy in them and the type of occupation, as well as use of sun protection methods. **Methodology:** This was a cross-sectional study conducted via a dermatological outreach albino clinic in the University of Benin Teaching Hospital in Southern Nigeria. A semi-structured questionnaire was used to collect data on sociodemographic characteristics. Also data regarding sun related dermatoses was collected and patients were examined thoroughly for cutaneous malignancy. Dermoscopy was used to boost diagnostic accuracy and skin biopsies were done to confirm diagnosis. **Results:** The prevalence of skin cancers was 9(12.4%) in the albino population and nil in their controls.$p<0.01$. Squamous cell carcinoma was the most common skin cancer observed in 8(11%) of the albinos.**Conclusion:** Cutaneous cancers are common in patients with albinism.The use of sun protective measures may reduce its incidence in them.

KEYWORDS: Albinism; Cutaneous cancers, Melanin

INTRODUCTION

Melanin protects the skin from the deleterious effects of the tropical sun. The absence of this protective pigment in albinos predisposes them to solar lentigenes, photoaging, solar keratosis and subsequent dysplastic changes in the skin which may later progress to cutaneous cancers. African albinos are more predisposed to having skin malignancy when compared to the Caucasian albinos because they live close to the equator where the exposure to ultraviolet radiation of the sun is much. The most common of the skin cancers reported among the albinos has been the squamous cell carcinoma. Oluwafemi and colleagues reported 68%[{1}], Yakubu and Mabogunje documented squamous cell carcinoma of 83% in Northern Nigeria;[2] Luande *et al.* 96% in Tanzania,[3] and Opara and Jiburum had 75% in their study.[4]The incidence of malignant melanoma (MM) is rare in albinos. Although there have been some few reports in the literature, especially in tyrosinase-positive albinism.[3-6] The use of sun-protection methods like avoidance of sun-peak hours, the use of sunscreens and the use of sun protective clothing have been shown to be protective against cutaneous malignancy in them. This study sought to determine the prevalence of cutaneous cancers in albinos in Benin city. Furthermore it sought to determine the relationship between cutaneous malignancy and type of occupation (indoor/outdoor) and finally the relationship between cutaneous malignancy in albinos and the use of sunscreens.

METHODOLOGY

Study Area

The study was carried out in the Dermatology Unit through an albino outreach clinic at the University of Benin Teaching Hospital. The hospital is located in the South-South geopolitical region in Nigeria, Benin City, Edo State.

Study Design

A descriptive cross sectional study was utilized.

Study Population

Clients with albinism, presenting at the Dermatology Outreach Albino Clinics in the University of Teaching Hospital, Benin City.

Selection Criteria

All patients with albinism consenting to the study, attending the albino outreach clinics were included in the study while all patients with albinism who did not consent to study were exempted.

On the other hand, the controls were all non-albino patients consenting to study attending the general

outpatient clinics. Those who did not consent to study were excluded.

Sample Size Determination
Using the fisher's formula,[7]

$$N = \frac{z^2 pq}{d^2}$$

n = minimum sample size
Z = Normal standard deviation 99% confidence level of 2.58
P = prevalence
q =1-p
d = margin of error =0.01

Studies describe a prevalence of 1 in 1000, i.e. 0.1% [3]

$$\text{Therefore } n = \frac{2.58^2 \times 0.001 \times 0.999}{0.01^2} = \frac{0.006649}{0.0001} = 66.49$$

Altrition rate of 10%= 6.65
Hence sample size = 73

Study Population
Study Group - 73
Control Group - 73

A sample size of 73 was utilized for the study population. The controls were age and gender matched persons without albinism.

Ethical Consideration
Ethical clearance was obtained from the hospital for the study and permission was subsequently sought from all clients in whom this study was done, after explaining to them the purpose of the research, drawbacks and benefits of the research. In addition, information received was treated with utmost confidentiality.

Sampling Technique
An albino outreach clinic was set up in the Dermatology Unit of the University of Benin Teaching Hospital (UBTH), Benin city. A snowball sampling technique was utilized where any client with albinism, presenting at the clinic and who consented to study was encouraged to nominate another albino. The nominated subjects were observed and this continued in the same way until the total number of clients were obtained.[4]

The control subjects were age and sex matched.

3.9 Data Management
An interviewer administered questionnaire containing two aspects was administered to each respondent. This included information on sociodemographics. The socio-demographic section assessed for characteristics like age, sex, marital status, occupation. Information on sunscreen ues was also taken. Respondents were thoroughly examined for cutaneous malignancy Dermatological tools like the dermoscope was used to boost diagnostic accuracy. Clinical diagnosis was confirmed by skin biopsy.

Skin Biopsy Techniques
Skin biopsy involved a process whereby skin tissue was taken for the purpose of histological diagnosis.There were different methods of skin biopsy techniques used in the course of the study and they included:

Punch biopsy
This method involved sterilizing the skin with alcohol swab and then subsequent anaesthesia with lignocaine. A sterile biopsy punch was used to remove a cylindrical core of skin and all layers of skin including subcutaneous fat . Once skin was cored, a scapel blade was used to excise cored skin.[8] This technique was useful for smaller suspicious lesions.

Incisional Biopsy
This was used where a larger piece of skin was required to make a diagnosis. A scapel blade was used after skin had been cleaned and anaesthesized. Stitches were then subsequently applied to secure hemostasis.[9]

Excisional biopsy
This involved the removal of the entire skin lesion with a wide margin. It was done for suspected skin malignancies like squamous cell carcinoma, basal cell carcinoma etc. Stitches were usually required to secure hemostasis.[9]

Data Analysis
All data generated were analysed using statistical package for social sciences (SPSS) version 21.0. Results were presented in tabular form. Discrete variables were presented as frequency and percentages. Continuous variables were presented as mean and standard deviation. Chi-square was used to determine association between categorical variables. Chi square test with yates correction was used for comparisons with small sub-group size of 5 or less. $P < 0.05$ was taken as significant.

RESULTS
The albino group was 73 (41 females and 32 males) with a male to female ratio of 0.8:1. The control group which was sex and age matched was also 73 with (41female and 32males).

Amongst the albino population, 46(63%) were single, 91(2.3%) were married, 1(1.4%) separated and 17(23.3%) were not eligible for marriage, while for the control population, 31(42.5%) were single, 23(31.5%) were married, 1(1.4%) separated and 18 (24.7%) were not eligible for marriage. The married were more frequently observed in the control population than in the albino population. This finding was statistically significant ($p<0.05$).

A monthly income of < N20000 was more frequently observed in the albino population 48(65.8%) than the

control population 29(39.7%). This difference was statistically significant (p<0.001).

The mean age for the albino population was 24.1± 11.3 years with an age range of 3-53yrs while the mean age for the control group was 24.1± 11 years (t= 0.007, p>0.05). {see table 1}

Table 1: Demographic Data.

Age (years)	Albino n=73(%)	Control Group n=73(%)	p-value
≤10	10(13.7)	10(13.7)	0.994
11-20	17(23.3)	17(23.3)	
21-30	27(37.0)	27(37.0)	
31-40	15(20.5)	15(20.5)	
41-50	3(4.1)	3(4.1)	
51-60	1(1.4)	1(1.4)	
Mean±Std. Dev	24.1 ± 11.3 years	24.1 ± 11years	(t=-0.007, p=0.994)
Sex			
Female	41(56.2)	41(56.2)	1.000
Male	32(43.8)	32(43.8)	
Marital Status			
Married	9(12.3)	23(31.5)	0.028*
Separated	1(1.4)	1(1.4)	
Single	46(63.0)	31(42.5)	
Not Eligible	17(23.3)	18(24.7)	
Educational Status			
None	13(17.8)	11(15.1)	0.835
Primary	3(4.1)	3(4.1)	
Secondary	12(16.4)	9(12.3)	
Tertiary	45(61.6)	50(68.5)	

Table 1b: Demographic DATA (Contd.)

	Albino n=73(%)	Control Group n=73(%)	p-value
Monthly Income			
<20,000	48(65.8)	29(39.7)	<0.001*
20,000 to 49,000	10(13.7)	3(4.1)	
50,000 to 99,000	9(12.3)	7(9.6)	
100,000 to 199,000	6(8.2)	8(11.0)	
200,000 to 299,000	0(0.0)	20(27.4)	
300,000 and above	0(0.0)	6(8.2)	

* Statistically significant

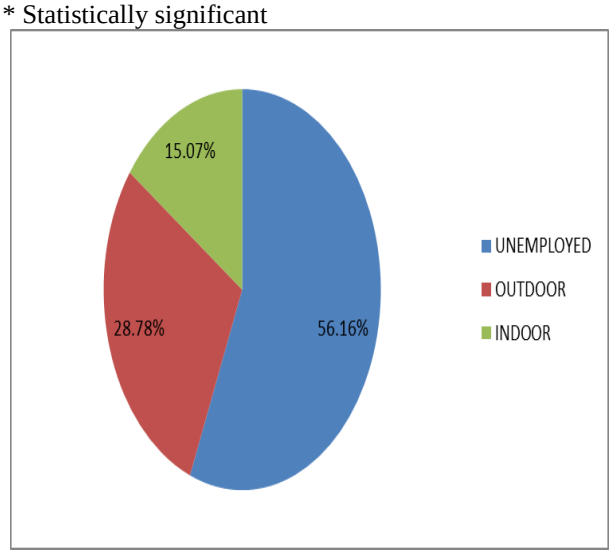

Fig 1: Type of Occupation.

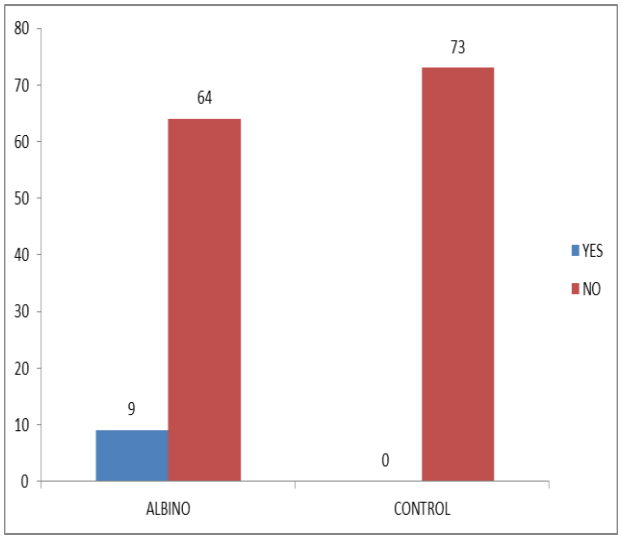

Fig. 2: Prevalence of Skin Cancers in Albinos and Control P=0.002

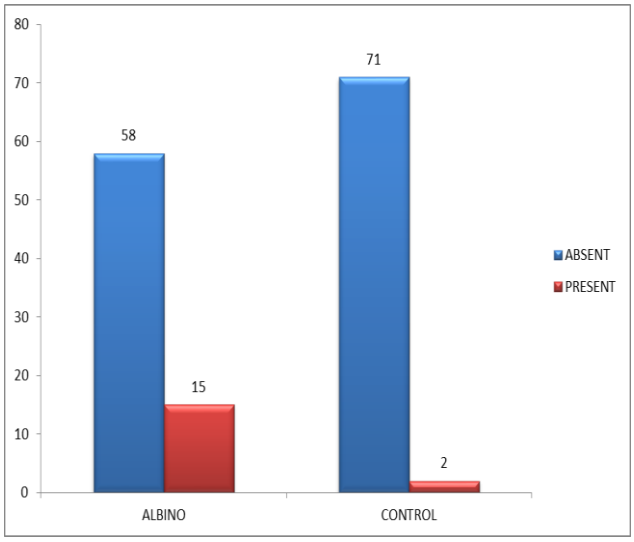

Fig 3: Frequency of Sunscreen Use in Albino and Control Population.

Table 2: Relationship between Skin Cancers, Type of Occupation and Sun-Protection Methods.

Age	Cutaneous Cancers Yes n (%)	No n (%)	p-value*
<30	3(4.1)	45(61.6)	0.055
>/30	6(8.2)	19(26.0)	
Occupation Indoor	1(1.4)	10(13.7)	0.01*
Outdoor	8(11.0)	13(17.8)	
Unemployed	0(0)	41(56.2)	
Sunscreen use Yes	1(1.4)	14(19.2)	
No	8(11.0)	50(68.5)	0.454
Sunprotection clothing			
Yes	3(4.1)	10(13.7)	
No	6(8.2)	54(74.0)	0.194

*statistically significant

A 40yr old school teacher with advanced squamous cell carcinoma Lesion had slowly increased in size over a 2yr period. Patient had initially resorted to herbal medication. Diagnosis was confirmed on histology.

The prevalence of skin cancer was 9(12.4%) in the albino population and nil in their controls.*p<0.01*. There were two types of skin cancers observed in this study. They included 8 (11%) squamous cell carcinoma and 1(1.4%) basal cell carcinoma. Seven of the squamous cell carcinoma occurred in the head region while one occurred on the chest. The basal cell carcinoma occurred in the head.

There were 41(56.2%) unemployed albinos, 13(28.8%) engaged in outdoor occupation like farming while 10(15.1%) were involved in indoor occupation Fifty-eight (79.5%) of the albinos did not use sunscreens while 15(20.5%) used sunscreens. Only one (1.4%) of the persons who used sunscreens developed skin cancer while 8(11%) did not use sunscreens. This finding was not statistically significant (0.454). The controls had 97.3% (71) who did not use sunscreens while only 2.7% (2) used sunscreens. Sixty (82.2%) persons in the albino population did not use sun-protective clothing while 13(17.8%) did. Six out of the nine who had skin cancers did not use sunprotective clothing.*p=0.194*

DISCUSSION

Melanin protects from the harmful effects of the ultraviolet radiation of the sun. Nigeria is radiation is more in the countries closer to the equator like Nigeria and predisposes Nigerian albinos to higher incidence of cutaneous cancers. The two main types of albinism are OA and oculocutaneous albinism. All the patients in this study are oculocutaneous type as expected. Skin cancers occurred in 9(12.4%) of the albino respondents but did not occur in the controls. Skin cancers were more common in those were engaged in outdoor occupation 8(11%) compared to those who were engaged in indoor occupation 1(1.4%). This was a statistically significant finding. It was in keeping with studies by Joseph et al on

skin cancers in albinos in north western Tanzania where, it was found that exposure to light appeared to be the single most important risk factor for the development of skin cancers in albinos.[10] Squamous cell carcinoma was the most common cancer seen in this study 8(11%). Seven occurred on the head while one occurred on the chest. This is similar to the trend seen by Yakubu et al,[11] and Krombergetal,[12] where, squamous cell carcinoma was found to be the most common skin cancer seen in albinos. The life time risk of squamous cell carcinoma in an albino is said to be 1000 fold compared to the general population and the most favoured site is the head and neck.[6,7] They presented with florid type of lesions due to neglect, poverty, and ignorance. In fact, in some communities in Sub-Sahara Africa, they are considered to demonic or reservoir of evil spirit, and so they abandoned by the parents to be care for by the aged grandparents in the remote places.[4] Our findings of 12.4% was similar to published reports by Asuquo and Ebughe who reported 18.4% in Calabar, Southern Nigeria[13] and 16.7% in Port Harcourt, Nigeria.[14] Olufemi reported a higher prevalence of 50% which they attributed to their small sample size of just 22 albinos.[1] Yakubu and Mabogunje reported a smaller prevalence of 2.5% which was attributed to increase awareness to treatment among the albinos and their caregivers.[3] Cutaneous malignancy was more common after the third decade in our study. This was similar to published reports by Oluwafemi in Irrua.[1] There was a rare incidence of cutaneous cancers in the first two decades of life in albinos, which was also reported by Yakubu and Mabogunje.[3] This is because it takes some time for a malignant transformation of the solar (actinic) keratosis in these patients.[1] The cutaneous cancers are the major risk associated with albinism and are thought to be major cause of death in African albinos.[4] Cutaneous cancers in albinism could be preventable to a reasonable extent, so there should be early education of the albinos, about the care for their skin. The skin can be protected from ultraviolet radiation by avoiding sun-peak hours, avoid wearing sleeveless shirts or blouse, wearing of wide brim hats, application of sunscreen creams with high sun protective factor and always using umbrella whenever there is need to enter under the sun. Provision of subsidized or free health care for albinos in our country will almost reduce the burden of cutaneous cancer significantly by encouraging early presentation of skin lesions and cheaper treatment. The establishment of the Albino foundation or groups in the locality, networking with the national body will go a long way to improve the general health care and quality of life of the albinos.

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