

Full Length Research Paper

Oculocutaneous albinism and skin cancer in Calabar, Southern Nigeria

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Oculocutaneous albinism (OCA) is a congenital condition causing hypo pigmentation of the hair, skin, and eyes. Albinism and solar radiation are established risk factors for skin cancer, the commonest being squamous cell carcinoma (SCC) of the head and neck in black Africans. We present four OCA patients with histologic diagnosis of skin cancer presented to the University of Calabar Teaching Hospital from January, 2007 and December, 2008. These cases were compared with SCC and total skin malignancy seen during the same period. Forty-five patients presented skin malignancy during the study period. Four OCA patients (2 men and 2 women) with SCC accounted for 21.1% of SCC and 8.9% of skin malignancy. They ranged in age from 21 to 30 years (mean 25 years). All the lesions were on the head and neck. The tumours were excised with a recurrence in a man who also had adjuvant chemotherapy with poor response. During the evaluation period of the 7 and 12th months, the sites appeared healed with no evidence of recurrence in 3 patients. Albinism and exposure to solar radiation were identified as risk factors for skin cancer. Establishment of “albino support group” to finance early implementation of public education strategies on prevention and treatment is recommended.

Key words: Albinism, squamous cell carcinoma (SCC), skin cancer.

INTRODUCTION

Oculocutaneous albinism (OCA) is a congenital condition causing hypo pigmentation of the hair, skin, and eyes (Lund, 2005). OCA 2, the form of albinism found in Africa, is inherited, as an autosomal recessive disorder; OCA tyrosinase, the key enzyme for synthesis of melanin is present and functional. The defect in OCA 2 is in the P gene coding for a membrane protein of the melanosome leading to malfunction of melanin synthesis (Biswas and Lloyd, 1999). Non-melanoma skin cancers, basal cell carcinoma (BCC) and squamous cell carcinoma (SCC) are common among the susceptible group with albinism (Okulicz et al., 2003). SCC of the head and neck are the most common cutaneous tumours in African patients with albinism (Yakubu and Mabogunje, 1993). OCA is a stable condition with no cure and requiring life long management. Dealing effectively with the health and social issues surrounding albinos in low-income countries with limited resources is a challenge (Lund and Taylor, 2008). We

present four OCA patients to highlight SCC as the commonest skin cancer in albinos and the role of solar radiation as a risk factor in our setting.

PATIENTS AND METHODS

Clinical histories and treatment outcomes of four consecutive OCA patients seen at the University of Calabar Teaching Hospital, Calabar, during a 2-year period from January, 2007 and December, 2008 with histologic diagnosis of skin cancer were prospectively studied. The incidence was compared to total skin cancer during the same period. In total, 1978 patients were seen in the surgical out patient during the same period.

RESULTS

Forty-five skin malignancies were presented during the study period. SCC ranked first with 19 patients (42.2%), followed by Kaposi sarcoma (KS) with 15 patients (33.3%). The four albinos all with SCC accounted for 8.9% of skin malignancies and 21.1% of SCC. The ages of the four albinos: two men and 2 women ranged from

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Figure 1. (A) Squamous cell carcinoma, (B) squamous cell carcinoma (same patient).

Table 1. Clinicopathological features and outcome of four albinos with SCC.

Patients No.	Age (years)	Gender	Clinical presentation (site)	Histology	Treatment	Outcome/ Follow-up
1	30	M	Chronic ulcer (Neck) 7 months	SCC moderate differential	Excision skin grafting, adjuvant chemotherapy, referred for radiotherapy	Recurrence 2 months, lost to follow-up
2	23	M	Chronic ulcer (Face) 7 months	SCC well differentiated	Excision	Healed 12 months
3	26	F	Chronic ulcer (Fore head) 15 months	SCC well differentiated	Excision skin grafting	Healed 9 months
4	21	F	Multiple lesions, 2 fleshy masses (Scalp), Chronic ulcer (Neck) 24 months	SCC well differentiated	Excision	Healed 7 months

21 to 30 years (mean 25 years). The ages of the SCC patients with Marjolin's ulcer ranged from 32 to 70 years (mean 48.3 years), while that of the non Marjolin's, non albinos ranged from 30 to 70 years (mean 56.6 years). The mean age of the 45 patients with skin malignancy was 46.6 years and their ages ranged from 5 to 75 years. The lesions in the albinos were located on the head and neck (Figure 1 and Table 1). The commonest site for SCC was the limbs (37%), the head, and neck ranked second (32%), while the third was external genitalia/perineum (26%). All the patients (15 blacks, 4 albinos) were human immunodeficiency virus (HIV) sero negative (Table 2).

All the patients had excision. The outcome was satisfactory in three patients (a man and two women) with the sites healed and no evidence of recurrence. However, there was recurrence in a male, noticed 2 months post excision that persisted following adjuvant chemotherapy (cisplatin and methotrexate). He was lost to follow up after referral for radiotherapy. The histological grading of the lesion of the patient with a recurrence was moderate differentiation, while the others with satisfactory outcome were well differentiated (Table 2).

Table 2. Site distribution of SCC (2007 – 2008).

Site	Total (%)	Remark
Head/Neck	6 (32)	2 blacks and 4 albinos
Chest	-	-
Abdomen	-	-
Limbs		
Upper	1	7 (37) All blacks
Lower	6	
External		
Genitalia/vulva	3	5 (26) All blacks
Peritoneum	2	
Anus	1 (5)	Black
Total	19 (100)	15 Blacks and 4 albinos

DISCUSSION

Forty-five patients with skin cancer were observed during the study period (January 2007 and December 2008): 19 of the patients had SCC (42.2%) ranking first, while

Kaposi sarcoma (33.3%) ranked second. The four OCA patients accounted for 8.9% of skin malignancies and 21.1% of SCC. Reports from Datubo-Brown (1991) in Port Harcourt (Southern Nigeria) showed that albinism accounted for 16.7% of skin cancer, Yakubu and Mabogunje (1995) in Zaria (Northern Nigeria) reported 8%, while Kromberg et al. (1989) in South Africa reported 23.4%. These reports highlight albinism as a risk factor for skin cancer. The ratio of African albinos in the general population in our setting has not been reported: however, 1978 patients were seen in the surgical clinics during the period of study.

The mean age for presentation of skin cancer in OCA patients was 25 years (3rd decade), they were presented 2 decades earlier than black patients did with skin malignancy (5th decade). Black patients with Marjolin's ulcer in the SCC subset were presented in the 5th decade, while the non-Marjolin's subset of SCC excluding the albinos was presented a decade later (6th decade). In Tanzania, examining 164 patients with albinism, skin cancer were found in 25% of those with age over 20 years (Lookingbill et al., 1995). Protection from the sun must start at birth and continue throughout life as those who present with skin cancer run a risk of developing another skin cancer later in life (Lund and Taylor, 2008; Asuquo et al., 2008). OCA is a stable condition with no cure requiring life-long management; this is a challenge in a low income setting with limited resources as ours.

The lesions in the four patients were SCC located on the head and neck. The presence of the tumour in these sites highlights the role of solar radiation as a risk factor for skin cancer in albinos. Lund and Taylor (2008) in South Africa, Yakubu and Mabogunje (1993) in Northern Nigeria, Luande et al. (1985) in Tanzania, and Kromberg et al. (1989) in South Africa reported the head and neck as the most commonly affected site. This is in contrast to Caucasians in whom BCC is the most frequent skin cancer of the head and neck with solar radiation as risk factor. Perhaps other non-pigment related factors in African render them prone to SCC (Urbach, 1991; Ochicha et al., 2004). However, the commonest site of SCC in the black patients was the lower limb, followed by the external genitalia/perineum (Table 2) highlighting the role of non-solar risk factors. In white populations, SCC is the second most common cancer after BCC, affecting the head and neck with solar radiation as the risk (Gross and Monroe, 2006).

There was a recurrence in a patient whose histological diagnosis was SCC (moderate differentiation), the outcome in the other 3 patients with satisfactory outcome was SCC (well differentiated). A further study on this is required as this may predict outcome in terms of recurrence and nodal metastasis.

Albinism and environmental exposure to solar radiation were identified as risk factors for skin cancer. OCA patients presented skin malignancy 2 decades earlier than blacks. Establishment of "albino support group" to finance early implementation of public education strategies on prevention and treatment is recommended.

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